Children change. Despite the truism of this statement, the dynamics of developmental change are frequently absent from studies of child disorders. Why is this? We believe that the reason lies in the strong influence of adult neuropsychology, in which researchers and clinicians focus on brains that have developed normally and become consolidated by adulthood prior to the brain insult. Since the adult brain is highly specialized, it is unsurprising that models of adult brain function focus on special purpose, independently functioning modules, whose components could be damaged or left intact by a specific brain trauma: the metaphor of boxes in the brain to be crossed through when damaged. While the adult framework can be informative about the end-state of development, it is inappropriate for understanding developmental disorders or even typical development because it ignores the dynamics of developmental change (Karmiloff-Smith, 1997, 1998). Indeed, the start-state of development is very different from the adult end-state.

The normal infant cortex is initially highly interconnected (Huttenlocher & Dabholkar, 1997; Neville, 2006), and it is only with time and with the processing of different kinds of inputs that the child brain becomes increasingly specialized and localized for function (Johnson, 2001). In other words, the brain does not start out with independently functioning modules: Modules are emergent from a gradual and complex process of modularization (Karmiloff-Smith, 1992). This means that a tiny impairment early on in, say, the developing visual system might have cascading effects on the subsequent acquisition of, say, number or vocabulary. Such impairments may or may not be compensated for, depending on the severity and the specialization of the impairment in question. It also means that one cannot take a single snapshot of, say, middle childhood, describe the phenotype of a developmental disorder, and from that suggest an intervention program. This would not only be clinically imprecise for a given child, but likely to be inappropriate for the syndrome in general. In our view, to assist clinical diagnosis and subsequent intervention, it is crucial to ascertain how the current phenotype...
originated at the beginning of a developmental trajectory, as well as knowing where it will lead in the future of that developmental trajectory.

This chapter will therefore concentrate on the importance of tracing and tracking full developmental trajectories, as well as focusing on associations between domains and between syndromes, rather than the current focus on dissociations. For illustrative purposes, we will concentrate mainly on autism spectrum disorder (ASD), Down syndrome (DS), fragile-X syndrome (FXS), and Williams syndrome (WS).

Prenatal Learning

Fetal development starts very early, at the onset of zygote formation, with the first neurons of the human forebrain present at a very early stage (Bystron, Rakic, Molnár, & Blakemore, 2006). Moreover, for the cognitive neuroscientist, learning also starts very early. From about the seventh month of pregnancy onward, the healthy fetus is actively processing various forms of auditory input (Hepper, 1995; Moore, 2002). Fetuses who hear a specific piece of music in the womb will discriminate that particular music from other pieces at birth. Newborns also recognize their mother's voice at birth, despite the fact that in the womb it was filtered through the amniotic fluid and sounds very different ex utero. Yet, during intrauterine life the fetus forms some abstract representation of mother’s voice and is able to distinguish her voice from other female voices at birth (Kisilevsky et al., 2003).

Furthermore, fetuses also learn the beginnings of the speech patterns of their mother tongue while in the womb. Research using acoustic spectroscopy has shown that, at 27 weeks, a fetus’s cry already contains some features of his or her mother’s speech, such as rhythms and voice characteristics. Also, DeCasper and colleagues showed that fetuses at 33–37 weeks’ gestation demonstrated memory of children’s rhyme, while still in the womb, in response to mothers repeatedly reading a certain rhyme to their unborn baby (DeCasper, Lecaunet, Busnel, Granier-Deferre, & Maugeais, 1994).

For the moment, we lack any knowledge about the learning capacities of the atypically developing fetus. However, for a truly full understanding of the developmental trajectory of a child with a disorder, this is where we should in the future be grounding our field of enquiry. For the time being, we must begin with postnatal development.

Neuroconstructivism and Postnatal Learning

From the moment the child is born, he or she is bombarded with interesting stimuli—faces, voices, objects, and so forth—and, as a result of the repeated processing of these different stimuli, the infant brain becomes slowly but increasingly specialized (Johnson, 2001). Elsewhere, we have argued that a middle way is needed between staunch nativism, on the one hand, in which the infant brain is thought to be prespecified for each of its
modular abilities, and behaviorism, on the other, in which a single, general purpose learning mechanism is invoked. Neuroconstructivism, an intermediate way between nativism and behaviorism, holds that a small number of domain-relevant learning algorithms jump-start the infant brain (Elman et al., 1996). Initially, all algorithms attempt to process all inputs, but with time the one that is most domain-relevant (say, to rapid sequential processing) wins out in the competition between algorithms and becomes domain-specific over developmental time (Karmiloff-Smith, 1998). We speculate that this is the case for the typically developing infant. However, we do not know whether the atypically developing infant brain displays the same level of interconnectivity early on, and whether subsequent pruning leads to specialization and localization of function in children with developmental disorders. But, theoretically, we can already ask what the implications of early interconnectivity would be for the atypically developing brain.

Within the theoretical assumptions of neuroconstructivism, the interconnectivity of early cortical development means that a tiny deficit could permeate all parts of the cortex. But, given the interaction between different algorithms and different structures in the environmental input, some parts of the brain would be more seriously affected by the deficit than others. This could give rise, over developmental time, to a seemingly isolated domain-specific impairment and the apparent preservation of other domains (i.e., scores “in the normal range”). In other words, what seems in the end-state to be a domain-specific deficit may have originated in the start-state as a more domain-general deficit (Annaz & Karmiloff-Smith, 2005; Karmiloff-Smith, 1997, 1998; Karmiloff-Smith et al., 2004). We therefore strongly advocate the importance of investigating not only domains of weaknesses, but also domains in which individuals show proficiency; that is, reach scores comparable to controls. Indeed, if changes to domain-relevant properties are initially widespread, and some properties are less relevant to a given domain, then that domain might exhibit lesser, more subtle impairments (Karmiloff-Smith, 1998; Karmiloff-Smith, Scerif, & Ansari, 2003). Ideally, then, an explanation of developmental deficits consists in identifying how these initial domain relevancies have been altered in the disorder, and then how the subsequent process of emergent modularization has been perturbed.

“Spared” versus “Impaired” Processing?

In the literature on developmental disorders, one frequently encounters terms such as “spared,” “intact,” and “preserved” when describing atypical development (e.g., Hoffman, Landau, & Pagani, 2003; Rouse, Donnelly, Hadwin, & Brown, 2004; Tager-Flusberg, Plesa-Skwerer, Faja, & Joseph, 2003). The notion of a selective deficit implies the impairment of a single process or domain together with the preservation (i.e., normal development and functioning across time) of others. When a brain has developed normally, resulting over time in specialized, localized functions, it is possible that after consolidation subsequent brain injury may produce selective damage(s) while other
components continue to operate normally. Hence, it might be appropriate to consider them as spared, intact, or preserved.

However, in the case of a developmental disorder of genetic origin, the use of such terms is questionable. They imply that the purported intact function has *developed normally* from infancy, through childhood to adulthood, with no interactions with other developing parts of the brain. Yet, as we mentioned above, the infant brain starts out highly interconnected (Neville, 2006), so it is unlikely that one part of the brain can develop normally in total isolation, without being affected (even subtly) by other parts of the atypically developing brain. The use by clinicians of the intact–impaired dichotomy in characterizing developmental aspects of functioning has problematic implications for intervention (A = intact, no intervention required; B = impaired, intervention required). Such dichotomies, then, could actually hinder rather than enhance the study of the dynamics of atypical development. By contrast, if one considers development as a dynamic process of interactions and competition, it could be, for instance, that training in rapid sequential movements in the assumed “preserved” motor system could impact another nonmotor domain which is impaired, rather than direct training in that nonmotor domain.

**Concrete Examples from Developmental Cognitive Neuroscience**

Studies that have taken the neuroconstructivist developmental approach to behavioral phenotypes have shown, for instance, that areas of purported relative strength at one stage of development (middle childhood or adolescence) were not necessarily stronger at earlier stages of development (Paterson, Brown, Gsödl, Johnson, & Karmiloff-Smith, 1999). For example, Paterson and colleagues (1999) showed that infant cognitive profiles in Williams syndrome and Down syndrome cannot be predicted from the adult end-state pattern of their cognitive functioning. One of the most compelling examples is vocabulary learning in toddlers with Williams syndrome, which is very poor and as delayed as vocabulary acquisition in toddlers with Down syndrome. By contrast, when individuals with Williams syndrome reach adolescence or adulthood, their language vastly outstrips that of their counterparts with Down syndrome. The same differences between the infant start-state and the adult end-state exist for number (Paterson et al., 1999; Paterson, Girelli, Butterworth, & Karmiloff-Smith, 2006). Infants and toddlers with Williams syndrome are sensitive to differences in small numbers, whereas those with Down syndrome perform even more poorly than younger mental age-matched infant controls. By contrast, in adulthood, scores for Down syndrome in the number domain outstrip those for Williams syndrome (Paterson et al., 2006). This highlights the importance of examining an entire developmental trajectory rather than a snapshot of development in childhood or adulthood.

Another example comes from studies of children with unusual genetic mutations. We have for several years been examining the cognitive phenotypes of children with
deletions within the Williams syndrome critical region but which are smaller than the typical Williams syndrome deletion on chromosome 7 (Karmiloff-Smith et al., 2003; Tassabehji et al., 1999). Our aim is to delineate the functions of various genes in expressing the full Williams syndrome phenotype. Here again, developmental trajectories have played a crucial role. In the case of one partial deletion child (HR), we found on initial testing that she did not differ from normal controls on the Bayley Scales of Infant Development. We could have concluded that the genes deleted in her case played no role in the Williams syndrome phenotype. However, as we began to trace her trajectory over developmental time, we found that, although she had a milder phenotype, she none the less progressively approximated the Williams syndrome phenotype and drew away from the typical trajectory. This was true at both the level of facial dysmorphology (Hammond et al., 2005) and that of her cognitive phenotype (Karmiloff-Smith, 2004). Figure 2.1 provides an illustration of this changing pattern.

Another example from developmental cognitive neuroscience is provided by Scerif and colleagues (2004) who investigated visual search in toddlers with fragile-X syndrome and those with Williams syndrome. These researchers demonstrated how important it is to go beyond mere scores to examine patterns of errors. While both groups of atypically developing toddlers reached a similar overall level compared to mental age-matched controls, their pattern of errors was very different. Toddlers with Williams syndrome made the highest number of erroneous touches on distractors. They were more affected than the other groups by the combination of larger display size and target–distractor similarity (conjointly increasing the perceptual load of the search task). By contrast, the toddlers with fragile-X syndrome made more errors of perseverance to targets already visited. In other words, where performance scores did not distinguish between the two syndromes, their respective patterns of error did.

![Figure 2.1](image.png)

**Figure 2.1** Changes in scores on the Bayley Scale of Infant Development over time for a child (HR) with Williams syndrome with deletion on chromosome 7, compared to typically developing (TD) children and children with Williams syndrome (WS).
A third example comes from face processing in Williams syndrome. There is no doubt that face processing is a relative strength in this syndrome: On some standardized tests, individuals with Williams syndrome achieve scores in the normal range (Bellugi, Marks, Bihrlle, & Sabo, 1988; Udwin & Yule, 1991). However, it would be erroneous to maintain that face processing develops normally in this clinical group. Several behavioral and electrophysiological studies point to atypical development of face processing in Williams syndrome compared to controls (Grice et al., 2003; Karmiloff-Smith et al., 2004). The general consensus (but see Tager-Flusberg et al., 2003, who continue to maintain that Williams syndrome face processing is no different from healthy controls) is that the behavioral proficiency in Williams syndrome face processing (and in autism spectrum disorder face processing) is underpinned by different cognitive processes (Annaz, 2006; Deruelle, Mancini, Livet, Cassé-Perrot, & de Schonen 1999; Karmiloff-Smith, 1997, 1998; Karmiloff-Smith et al., 2004; Rossen, Bihrlle, Klima, Bellugi, & Jones, 1996). This was further corroborated by our event-related potential (ERP) study comparing the brain processes of healthy controls versus adolescents and adults with Williams syndrome when processing faces and cars (Grice et al., 2003), as well as in another study of cerebral integration (Grice et al., 2001). The face-processing findings highlighted the fact that although healthy controls processed both human and monkey faces in a relatively similar way, their brains treated cars very differently. By contrast, the brains of participants with Williams syndrome displayed no differences between faces and cars.

Moreover, unlike the healthy controls, who showed a right hemisphere dominance for upright faces, the clinical group failed to display any difference in hemispheric activation (Karmiloff-Smith et al., 2004). This highlights two facts about the deviant trajectory of Williams syndrome face processing, despite their behavioral proficiency. First, there is a lack of specialization: Individuals with Williams syndrome show similar electrophysiological responses for both faces and cars; that is, they have not progressively restricted the brain circuits responsible for face processing uniquely to face stimuli, but process all manner of visual stimuli in a similar way. Second, there is a lack of localization: Healthy controls show stronger processing for faces in the right hemisphere, whereas the clinical population displayed equivalent bilateral activation. The lack of specialization and localization in Williams syndrome face processing indicates that, despite enormous daily experience with faces, a face-processing module fails to emerge over developmental time in this clinical population. In other words, their proficiency on some standardized tasks is achieved through different cognitive and brain processes than in normal development. It follows that clinicians need to be cautious when they encounter scores in the normal range, given that these may camouflage different cognitive and brain processes from healthy controls.

The sensitivity of standardized tests is, as we saw above, open to discussion, raising the risk that scores in the normal range may be achieved by atypical brain and cognitive processes. It also raises questions about what is being matched when researchers do group or individual mental age matching. Annaz (2006) illustrated these problems by testing children with Williams syndrome and those with high functioning autism (HFA) on the Benton Facial Recognition Test (Benton, Hamsher, Varney, & Spreen, 1983). Figure 2.2 illustrates how both groups score within the normal range on this
face-processing task. However, to examine whether both groups were processing faces like their typically developing (TD) counterparts, Annaz carried out an in-depth examination of their face-recognition skills. She found that both clinical groups performed significantly more poorly on face-specific tasks in which configural processing was manipulated, suggesting that the Benton task can be solved by featural processing and does not require the configural processing used by normal controls.

We have illustrated in the case of face processing just how crucial it is to differentiate “normal” scores at the behavioral level from underlying cognitive and brain processes. It is also obvious that the choice of a matched control group has theoretical implications. If one matches on IQ, it implies that intelligence affects the domain in question, say, language. If one matches, say, Williams syndrome with Down syndrome, this is not theory neutral because a match on their overall IQ camouflages the fact that in one case the score is brought down by the spatial component and in the other case by the verbal component. These differences will clearly affect all subsequent measures. So, what is the best way to gain a deeper understanding of developmental disorders? We believe that it is by building task-specific, full developmental trajectories.

## The Need for Developmental Trajectories

Of course, the most informative way of gaining an insight into how developmental changes occur over time in clinical groups (or, in fact, typically developing children also) is to conduct longitudinal studies. However, these studies are highly time-consuming and may put parents, children, and teachers under unnecessary pressure. Drop-out rates are indeed high in longitudinal studies. An alternative to the longitudinal method is to
build developmental trajectories by means of a cross-sectional design. This approach has been successfully used in recent studies (Annaz, 2006; Karmiloff-Smith et al., 2004; Thomas et al., 2001). The developmental trajectories approach seeks to build a task-specific typical developmental trajectory by first measuring performance across a wide range of ages in the normal population. Then, given an individual with a disorder, one can next establish whether his or her performance fits anywhere on the typical trajectory. Unlike the use of matched controls, this comparison is theory neutral.

Secondly, one can assess whether the individual fits on the trajectory at the position predicted by his or her chronological or mental age. Additionally, one can use a variety of other predictors (for example, language age, nonverbal reasoning age, and so on) to assess whether the individual fits on the normal trajectory according to any aspect of their cognitive profile. Indeed, one will often discover that predictors differ between healthy controls and the clinical group. So, for instance, whereas language predicts scores in numerical cognition in Williams syndrome, it is spatial scores that predict numerical outcome in the typical group (Ansari et al., 2003; Ansari & Karmiloff-Smith, 2002).

Tracing developing trajectories is not only possible for the normal population. Given a group of individuals with a certain disorder, with a wide age range, it is also possible to construct atypical task-specific developmental trajectories for a particular disorder and contrast this with the typically developing group. So, rather than comparing scores at a single point in development, the trajectories approach offers a more direct way of addressing the question “Does the target behavior develop normally or atypically in the disorder?” Such an approach also makes it possible to reconsider the notions of delay versus deviance.

An illustration comes from a cross-syndrome study (WS, DS, and ASD) by Annaz (2006). She and her colleagues investigated the development of featural and configural face recognition using the Jane Faces Task that had been extensively tested on the normal population (Mondloch, Le Grand, & Maurer, 2002). In the low functioning ASD group, atypical U-shaped performance was observed on the inverted featural face trials: Younger children with autism displayed better performance on inverted trials, followed by a decrease in accuracy at around 9 years of age (Figure 2.3). Yet for the whole developmental period, accuracy on upright featural trials continued to increase. However, performance on the configural trials did not significantly increase with chronological age or when assessed against other predictors (such as language). Had the research used the traditional mental age-matched individual or group comparisons, these effects would actually have been masked.

The use of the trajectories approach makes it possible to go beyond describing behavior as delayed. In other words, it becomes possible to provide more in-depth descriptors in terms of a delayed onset, which implies a normal rate (not statistically different from a normal trajectory); a delayed rate of developmental trajectory, which implies a normal onset but a slower rate of increase in performance; a delayed rate and onset; or, finally, a zero rate (a gradient not significantly different across time; Thomas et al., in press). It is also possible to examine intra- and intergroup variability using the trajectories approach.
The use of developmental trajectories is not only necessary at the behavioral level but it also needs to be complemented at the neuroanatomical level (Shaw et al., 2006). Shaw and colleagues used a longitudinal design to examine the relationship between cortical development and cognitive variation. They found a marked developmental shift from a predominantly negative correlation between intelligence and cortical thickness in early childhood to a positive correlation in late childhood and beyond. This study indicates that the neuroanatomical expression of intelligence in children is dynamic. Many other studies also indicate that IQ levels are not static but change with brain development and are impacted by environmental factors. Indeed, environmental factors may play a more important role than research often grants them (Mareschal et al., 2007). A child must always be considered within the environment that he or she lives because, as soon as parents are told that their child has a developmental disorder, their behavior changes subtly. They may unwittingly impede their child from freely exploring the environment and/or they may help the child avoid making mistakes, whereas the natural process of learning actually involves erroneous overgeneralizations and so forth.

**Conclusion**

No approach to developmental disorders is without its inherent problems, and neuroconstructivism is no exception. For example, Thomas (2005) highlights two unanswered problems associated with the theoretical assumptions of neuroconstructivism.
First, a clearer picture is needed of the initial domain relevancies that predate a particular domain and of the nature of the process that eventually delivers domain-specific functional structures. The second difficulty is related to methodological issues of building developmental trajectories from infancy through to adulthood. It cannot be assumed that the same task is treated in the same way across developmental time (i.e., using the same brain and cognitive mechanisms at very different ages). Clearly, these questions need to be at the heart of new research within the developmental trajectories approach.

In our view, the notions of interactivity, competition, compensation, redundancy, specialization, localization, and modularization will be key in characterizing in more depth how atypical development proceeds at the cognitive level, notions that have significant implications for the formation and functioning of mechanisms over developmental time (Karmiloff-Smith, 1992, 1998; Karmiloff-Smith & Thomas, 2003; Scerif & Karmiloff-Smith, 2005; Thomas, 2003, 2005), and as yet these mechanisms have in the main been neglected in developmental cognitive neuroscience.

We reiterate the importance of examining more closely “scores in the normal range.” It is also important to recall that phenotypic outcomes at the cognitive level could stem from much lower level deficits. Indeed, a very small difference in developmental timing, gene expression, neuronal formation, migration, and density, and many other genetic and biochemical factors, can impact on development over time and result in much greater deficits in the phenotypic outcome. This is why we contend that the task-specific developmental trajectories approach, starting wherever possible at the very outset of infant development, constitutes a first but important step toward gaining a deeper understanding of both the dissociations and associations across different syndromes.

References


Brain Plasticity: Evidence from Children with Perinatal Brain Injury

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Until very recently, our understanding of brain organization in humans has relied primarily on the classical neuropsychological approach of associating a specific lesion site with a behavioral deficit. Using this approach, studies of adults with strokes have informed our understanding of the neural underpinnings of various cognitive systems in the adult steady state, that is, after the brain has already acquired critical cognitive systems and the individual is an “expert user.” Functional imaging studies have broadly confirmed these findings, while adding to and refining our understanding of adult brain organization. However, how the brain reaches this mature state is a topic of significant interest and rich debate.

The development of the young brain is a dynamic process (Stiles, Paul, & Hesselink, 2006) and children’s brains continue to develop well into adolescence (e.g., Blakemore & Choudhury, 2006). A corollary of this protracted development is that children with early localized brain injury suffer less severe effects than adults with comparable lesions (e.g., Basser, 1962; Lenneberg, 1967), suggesting that this dynamic developmental process continues even in the presence of an early injury. For example, in language studies, children’s impressive development after early stroke led Lenneberg (1967) to propose that both hemispheres of the child’s brain were equipotential for language. Such plasticity, first systematically studied by Kennard in the 1930s (Kennard, 1940), has led to the widely held view that the brain is remarkably malleable or plastic early in life, and that, over development, there is a decrease in this flexibility. However, development is neither linear nor unidimensional, and different cognitive systems develop on distinctive timetables. For example, whereas children are generally producing short sentences by their second birthday, they do not draw recognizable objects, such as houses, until about the age of 4.

In this chapter on brain plasticity, the principal questions we pose concern the degree to which particular brain regions may be privileged for certain functions, and the degree to which there is neuroplasticity for differing cognitive systems as they develop. The context for this discussion will be prospective studies of children’s development
Brain Plasticity

following a perinatal (PL) injury (i.e., before higher cognitive systems have developed). This will permit us to witness how the developing brain adapts; that is, how the brain generates alternative organizations for behavioral functions. Following such children will illuminate both the degree to which initial biases exist, as well as the extent to which the developing brain is flexible in adapting and acquiring behavioral functions in the face of early injury and an overall, decreased capacity. Investigating development both within and across cognitive systems will permit us to identify gradients of plasticity for various systems as well as their component processes.

The chapter begins with an introduction to the population and then an overview of general intellectual functioning in children with perinatal stroke. We then turn to profiles of development within cognitive systems that in the adult model reflect distinct neural profiles: language, spatial cognition, and emotion processing. We close by exploring two contexts where these systems intersect: labeling emotions and the development of literacy in the PL group. As a framework to evaluate cognitive development in the PL group, we propose four questions that have guided our research over the past 20 years:

1. Do children show specific deficits early in development?
2. Do such profiles of deficit map onto those of adults with comparable lesions?
3. Do these deficits persist or is there development over time?
4. If they persist, do these deficits remain stable or change with development?

Children with Perinatal Brain Injury: Lesion Characterization

Perinatal strokes occur in 1 in 4,000 term infants (Nelson & Lynch, 2004). Stroke in the full-term infant can occur during the third trimester, during labor and delivery, or during the neonatal period (birth to the end of the first month of life)—in other words, the pre-perinatal period. Term infants generally have large strokes involving the middle cerebral artery, damaging both cortical and subcortical regions (Bax, Tydeman, & Flormark, 2006; Kirton & deVeber, 2006; Wu, Croen, Shah, Newman, & Najjar, 2006). Thus, this population provides an opportunity to examine issues of specialization and plasticity in children with large lesions that compromise much of one cerebral hemisphere.

Most pre-perinatal strokes involve the left hemisphere (LH), a finding attributable to anatomic or hemodynamic differences between the left and right common carotid artery (Volpe, 2000). The majority of the strokes have an embolic etiology and the placenta (Pathan & Kittner, 2003) is probably the most common source of these emboli. About three-quarters of term infants with stroke present with seizures in the neonatal period. However, some children with perinatal stroke do not present until several months of age when decreased hand use is noted and imaging (CT or MRI) reveals an old stroke which is presumed to have occurred in the pre-perinatal period. Because proximal arm and leg muscles have bilateral innervation and because infants do not
really start to use their hands until a few months of age, the hemiparesis is not immediately apparent. Newborns whose injury occurs during labor and delivery have acute signs of brain insult like seizures, while those whose stroke occurred even a few days before birth have “recovered” and are usually asymptomatic as newborns.

Neonatal seizures are routinely evaluated with an EEG and a CT and/or an MRI scan. Imaging of children presenting with a hemiparesis during the first year documents the extent of the lesion and confirms the unilaterality of the pathology. The medical workup at all ages includes an assessment of the heart, studies of blood clotting, ruling out infection, and studies evaluating for metabolic/genetic problems which can cause stroke (Kirton & deVeber, 2006). This workup assures exclusion of a child with bilateral involvement. Between 30% and 50% of children who have had a neonatal stroke appear neurologically normal (Lee, Croen, & Lindan, 2005; Nelson & Lynch, 2004). Hemiparetic cerebral palsy (which rarely prevents walking) occurs in 40–60% of cases, epilepsy in about 40%, visual problems in 25%, and behavior problems in 20%. About 80% have a problem in at least one domain. Risk factors for poor outcome include: abnormal examination at discharge from the nursery, neonatal seizures, abnormal initial EEG, blood clotting abnormalities, and late presentation of hemiparesis (Bax et al., 2006; Mercuri et al., 1999; Sreenan, Bhargava, & Robertson, 2000; Wu et al., 2004). Imaging data about site, side, and timing of stroke (pre- versus perinatal) provides additional information predictive of outcome (Kirton & deVeber, 2006; Kwong & Wong, 2004; Nass, Peterson, & Koch, 1989; Staudt et al., 2004).

Epilepsy is associated with poorer cognitive outcomes in children with early unilateral brain injury (Carlsson, Hagberg, & Olsson, 2003; Isaacs et al., 1996; Muter, Taylor, & Vargha-Khadem, 1997; Vargha-Khadem, Isaacs, Van der Werf, Robb, & Wilson, 1992). Consistent with these findings, an abnormal neonatal EEG predicts poorer outcomes at 15 months (Mercuri et al., 1999). When neonatal seizures occur concurrently with the stroke, they are generally few in number and easy to control. Moreover, the majority of children who have seizures as newborns never have them again; about 25–40% have recurrent seizures later in life that are easily controlled with medication. A small proportion of children with early unilateral brain injury have persistent seizures, and a small proportion of these children develop intractable seizures. Rarely, surgery is required to control the seizures.

**Overall Intellectual Performance: Performance IQ and Verbal IQ**

While it is well known that focal brain lesions incurred early in life have very different functional consequences than apparently similar lesions incurred later in life, our knowledge of early functional plasticity comes mainly from studies that examine the functioning of PL children at only one developmental time-point. Such studies provide only a snapshot of how an early lesion affects behavioral outcomes. With respect to the IQ of PL children, the majority of studies are based on assessments that were obtained at only a single time-point. These studies leave us with a somewhat mixed
pattern of results: Some studies report that the average IQ of children in this population is more than one standard deviation below that of control children (e.g., Levine, Huttenlocher, Banich, & Duda, 1987; Perlstein & Hood, 1955; St. James-Roberts, 1981; Woods, 1980), whereas other studies report that there is no significant IQ deficit (e.g., Aram & Ekelman, 1986; Bates, Vicari, & Trauner, 1999; Nass et al., 1989). These apparent discrepancies may reflect differences in the age of the children at the time of assessment as the studies that report little or no impact on IQ tend to assess children at earlier time-points than those that report IQ deficits.

Animal studies indicate that the functional consequences of early lesions change over the course of development (Goldman, 1971, 1974; Goldman-Rakic, Isseroff, Schwartz, & Bugbee, 1983; Kolb & Wishaw, 1985). For example, a series of studies of monkeys with early bilateral lesions of the dorsolateral frontal lobe revealed no impairment on a delayed response task at 12–18 months but a marked impairment at 2 years of age, in both cases in comparison to age-matched control monkeys (Goldman, 1971, 1974). Goldman-Rakic et al. (1983) suggest that this pattern reflects maturational changes in the frontal lobe. That is, deficits are only apparent in the lesioned monkeys when the dorsolateral frontal cortex, the region that is damaged in the lesioned monkeys, becomes sufficiently mature in the intact monkeys to contribute to the target behavior, their delayed response behavior. This level of frontal lobe maturity is apparently attained by 2 years of age, but not yet by 12–18 months of age. Thus, a comparison of performance between lesioned and intact monkeys reveals a deficit at the later but not at the earlier time-point.

A recent study examining the IQ level of PL children uncovered a similar developmental pattern (Levine, Brasky, & Nikolas, 2005). In this study, a group of children with pre- or perinatal unilateral brain injury was administered a standardized IQ test both before and after the age of 7. The results showed that PL children had significantly lower verbal and performance IQ levels at the later time-point than at the earlier time-point ($M_{\text{pre-7 verbal IQ}} = 95.13; M_{\text{post-7 verbal IQ}} = 85.73; M_{\text{pre-7 performance IQ}} = 94.67; M_{\text{post-7 performance IQ}} = 87.87$). Importantly, the lower IQ levels at the later assessment time-point do not reflect a loss of knowledge, but rather, a slower rate of development than occurred in the normative sample (Banich, Levine, Kim, & Huttenlocher, 1990). Individual subtest scaled scores were compared across time to examine whether the decline in IQ level was specific to particular subtests. Although all of the subtests that were administered at both time-points showed a decline, the decline was significant for Arithmetic, Vocabulary, Picture Completion, and Block Design, but not for Similarities or Information. Overall, this analysis revealed a rather broad decline that was not specific to verbal or spatial subtests. Levine and colleagues also examined whether the degree of falloff on subtests tapping visuospatial skills was greater for those tests with a motor requirement (Levine, Kraus, Alexander, Suriyakham, & Huttenlocher, 2005). We viewed this as a possibility since most of our PL children had some degree of hemiparesis. This question was addressed by comparing the degree of falloff on two performance subtests, Block Design, which requires some dexterity in manipulating pieces, and Picture Completion, which has minimal or no motor demands. This comparison
revealed that the degree of decline did not significantly differ on these two subtests, suggesting that the falloff on performance subtests is not attributable solely to motor difficulties.

As in the monkeys with early frontal lesions whose deficits were only apparent at the later assessment time-point, it is possible that the increasing IQ gap between PL and typically developing children reflects maturational changes in the developing brains of non-brain-injured children. That is, particular brain regions may only contribute to task performance in typically developing children once they have reached a sufficient level of maturity. If such regions are damaged and fail to develop normally in the PL children, the IQ gap between typically developing and brain-injured children will become increasingly apparent over time. The falloff in IQ level in brain-injured children can also be viewed as reflecting limitations that early lesions place on the brain’s overall processing capacities. Assuming that these processing capacities are increasingly taxed as task demands augment, the gap between the IQ level of typical and brain-injured children would be expected to widen over time. Thus, the degree of functional plasticity observed after an early lesion may vary with the age of the child at the time of assessment. It may also vary depending on the specific task and domain of functioning being examined. Since IQ tests were designed to assess overall levels of intellectual functioning rather than functioning in particular cognitive domains, it is critical to examine performance using more specific tasks in order to address this question; thus, we begin with an overview of language acquisition in the PL group.

Language Development in Children with Perinatal Stroke

In mapping brain–language relations, 160 years of research in a broad range of languages, including sign languages, has confirmed the original findings of Paul Broca: For the vast majority of adults, the formal and core aspects of language (phonology, morphology, and syntax) are mediated by the left hemisphere of the brain. With respect to the development of such relations, early observations of both Basser (1962) and Lenneberg (1967) noted that children who suffered brain injury did not display the same persistent and marked deficits as did adults with injuries to homologous regions. In the past 40 years, a number of studies have investigated language development in both typically developing children and in children with different neurodevelopmental disorders to better understand the process of language development and its neural substrates. A growing literature attests to Lenneberg’s observation: Children with brain injury fare much better than adults on tasks of language performance (Bates et al., 1999, 2001; Eisele & Aram, 1995; Feldman, 2005; Reilly, Losh, Bellugi, & Wulfeck et al., 2004; Vargha-Khadem, Isaacs, & Muter, 1994; Vicari et al., 2000, Weckerly, Wulfeck, & Reilly, 2004; Wulfeck, Bates, Krupa-Kwiatkowski, & Saltzman, 2004). For those studies that included children who have suffered lesions after their first birthday, that is, once the process of language acquisition is underway, the findings are mixed: Some studies show no significant differences in language performance according to lesion
site (e.g., Vargha-Khadem, Gorman, & Watters, 1985), whereas others have noted subtle persistent deficits for children with left hemisphere injury or hemidecortication (Dennis & Kohn, 1975; Dennis & Whitaker, 1976; Eisele & Aram, 1995; Riva & Cazzaniga, 1986; Vargha-Khadem et al., 1985). Here we address these issues by focusing on children who incurred their lesions in the perinatal period, before the onset of higher cognitive functions.

Traditionally, neuropsychologists have used standardized language tests (such as the Clinical Evaluation of Language Function [CELF]; Semel, Wiig, & Secord, 1995) to assess language proficiency. Such tests are designed to evaluate specific aspects of both receptive and productive language: vocabulary, morphology, and specific syntactic structures. Others subtests, rather than testing individual linguistic structures, ask children to use language to solve problems. For example, in the subtest Formulating Sentences (CELF), the child is shown a picture and given two words. His or her job is to make a coherent sentence about the picture using those two words. Such subtests are similar to those that constitute the verbal IQ score where language is a tool to solve problems, rather than testing mastery of specific grammatical structures. Two studies of PL children that used the CELF (Ballantyne, Spilkin, & Trauner, in press; MacWhinney, Feldman, Saccio, & Valdes-Perez, 2000) found that the performance of the PL group was significantly worse than age-matched controls. MacWhinney et al. (2000) also reported that the children with left hemisphere injury had particular difficulty on the subtests of formulating sentences and oral directions, similar to studies looking at overall intellectual performance (Carlsson et al., 2003; Isaacs et al., 1996; Muter et al., 1997; Vargha-Khadem et al., 1992). Ballantyne et al. (in press) reported that children with seizures performed significantly more poorly than the rest of the PL group on the standardized language measures. Seizure status has also been noted by other researchers as a factor in mediating language performance (e.g., Dall’Oglio, Bates, Vologa, Di Capua, & Pezzini, 1994). To complement these studies using standardized tests, below we present studies targeting the acquisition of core aspects of language and their development. The vast majority of studies have been conducted with English-speaking children, but studies of children acquiring other languages are included where available.

Early communication and language in children with perinatal brain injury

Early development of language in the PL group has been the focus of several research groups: All have noted initial delay in the emergence of language with subsequent progress following the typical developmental trajectory. Importantly, delay in the PL group is characteristic of children with lesions to either the left or the right hemisphere and thus does not map onto the language profile of adults who incur strokes later in life.

Several investigators have asked whether language development proceeds in the same way in brain-injured and in typically developing children by focusing on the relation of early gesture use and language development. As in typically developing children, early gesture milestones predate and predict subsequent vocabulary and syntactic milestones in the PL group (e.g., Bates & Dick, 2002; Sauer, Gripshover, Harden, Meanwell, & Levine,
2006). For example, the number of gestures produced at 14 months of age predicts subsequent word production and comprehension in both typically developing and PL children (Sauer et al., 2006). Similarly, gesture–speech combinations (e.g., “Mommy” + point to shoe) predict the subsequent emergence of parallel argument structures in speech (Levine, Özcülskan, & Goldin-Meadow, 2007). Such findings are particularly noteworthy in that the integral relationship between language and gesture is maintained even though children with PL are frequently hemiparetic. Another early study (Marchman, Miller, & Bates, 1991) followed a small group of PL children, and found delay in the onset of babbling, gesture, and first words in the PL group regardless of lesion site.

With the advent of the MacArthur Communicative Inventory, a parental report form (MCDI; Fenson et al., 1993), tracking children’s lexical development became more convenient and efficient. Bates and colleagues (1997) used the MCDI to chronicle early language development in a group of 40 PL children aged between 8 and 30 months. They found overall delay in the PL group as a whole, both for comprehension and for the production of first words. However, within this general context of delay, site-specific deficits emerged: In the group of PL children between 10 and 17 months, children with early right hemisphere injury (RPL) demonstrated greater delays in word comprehension than the rest of the PL group; between the ages of 19 and 31 months, children with left temporal damage (LPL) showed the greatest delays for productive vocabulary; a transient bilateral frontal effect appeared from 19 to 31 months. To directly evaluate children’s language production, Bates and colleagues also collected and analyzed language data from spontaneous parent–child interactions from 30 children with PL aged 19–44 months of age. Focusing on mean length of utterance (MLU), they found that children with left posterior damage on average used shorter utterances than controls or children with injury to other brain regions. This production profile is consistent with that of the parental reports, but is at odds with the typical adult profile of deficits in comprehension after left temporal injury.

Several longitudinal studies have also focused on early lexical development: Feldman and colleagues (Feldman, 1994; Feldman, Holland, Kemp, & Janosky, 1992) found both initial delay and wide variability in performance. In this smaller group of PL children, some performed in the normal range whereas others were delayed. However, after an initial delay, in most of the children progress was steady. Looking at morphological and syntactic development, they analyzed free-speech language samples using the IPSYN (Index of Productive Syntax; Scarborough, 1990). Again, they found wide variability in performance with half of those with LH injury and half of those with RH injury falling well below the norm. A longitudinal study of PL children by Thal and colleagues (Thal, Reilly, Seibert, Jeffries, & Fenson, 2004) also analyzed language samples using the IPSYN, and they too found wide variability in performance with delay in PL children with either RPL or LPL. These data are consistent with, and complemented by, a large longitudinal study of 40 PL children conducted by Levine and colleagues (Brasky, Nikolos, Meanwell, Levine, & Goldin-Meadow, 2005; Levine, Kraus et al., 2005) where the investigators found overall delay in the onset of language, but that lesion size rather
than site modulated the developmental trajectory for both productive vocabulary and syntactic development.

These findings are broadly consistent with those of other groups, including those studying languages other than English. Using the Italian MCDI, Vicari et al. (2000) also noted early delay in language onset with some site-specific profiles: Children with LH injury were more delayed in lexical and grammatical production than those with RH injury. Chilosí’s studies (Chilosí, Cipriani, Bertuccelli, Pfanner, & Cioni, 2001; Chilosí et al., 2005) of Italian PL children report similar findings. Further, as in typically developing children (Hart & Risley, 1995), Rowe and Levine (2007) found that the language a child hears (i.e., caregiver input) mediates language development of children with PL. This effect is sufficiently significant that children with lesions who have high language input from parents (more than 1 SD above the mean) have more rapid vocabulary development than control children with low language input from parents (more than 1 SD below the mean) at least until 3 years of age. In sum, as in typically developing children, input modulates language development; however, early brain injury to either the right or the left hemisphere will delay the onset of language. These findings suggest that the process of acquiring a language is different from maintaining an already functional system, as in adults, and, further, acquiring language draws on both the right and left cerebral hemispheres.

Language performance during the school years

By the age of 4–5 years, typically developing children have access to the majority of the morphosyntactic structures of their language (Slobin, 1985, 1992, 1995). None the less, conversations with a 5-year-old are markedly different from those with a child of 10. How do they differ? Whereas we assume a larger and more diverse vocabulary with older children, there are structural differences as well (Nippold, 1998). Language development from kindergarten onward entails refining particular components; for example, complex auxiliary verb morphology (for English) and developing the knowledge and facility to use structures effectively in various discourse contexts, such as telling a story, giving directions, or persuading one’s parents. Rather than focusing on morphology intraclausally (e.g., noun–verb agreement), children during the school years acquire more and diverse logical connectives (e.g., “although”), and they are learning how to exploit these elements to establish both cross-clausal connections (“If I had known you were coming, I would have baked a cake”) and hierarchical relationships (as in integrating the local episode of a narrative with its overarching theme (e.g., “While he was searching for his lost frog, he climbed up the tree to look into the hole and an owl swooped out and frightened him”; Bamberg, 1987; Berman & Slobin, 1994; Reilly et al., 2004). Researchers have proposed that this entails a re-organization in which old forms now assume new discourse functions (Berman & Slobin, 1994; Slobin, 1973) or a re-representation of particular structures (Karmiloff-Smith, 1992). To illustrate
discourse development in the PL group, below we present data from two discourse genres: a biographical interview and a storybook narrative.

An interview is a locally organized interaction: The experimenter poses a question and the child responds using the vocabulary and grammatical structures of his or her own choosing. Such quasi-naturalistic measures permit us to capture not only the child's everyday language, but also how he or she uses the language; for example, the appropriateness or relevance of a response. In one of the few studies directly comparing language performance of PL children (aged 5–8) to that of adults with comparable lesions, Bates et al. (2001) report data from a biographical interview. Unsurprisingly, the adults displayed the predicted adult profile: Those with LHD displayed contrastive profiles of impairment (including classic differences between fluent and nonfluent aphasia); those adults with RHD (and three nonaphasic adults with LHD) showed fluent but disinhibited, and sometimes empty speech. In contrast, the spontaneous language of the children with PL did not differ from the typically developing (TD) group. Both groups made very few morphological errors, and they used complex sentences with comparable frequency and diversity. As such, the earlier site-specific differences noted above were resolved. With respect to lexical production and diversity, morphology and syntax, that is, core aspects of language, the language of the PL group was comparable to their age-matched typically developing peers in this conversational context.

In this same data set of biographical interviews, we also examined anaphora, or the use of pronouns, as adults with RHD often have difficulty with the coherence of their discourse. Since a pronoun can function as a link to a preceding noun phrase, pronouns represent one linguistic means to connect utterances (Halliday & Hasan, 1976). Interestingly, in examining the children's use of pronouns (Dardier, Reilly, Bates, Delaye, & Laurent-Vannier, 2005), we found that both PL and TD children had many pronouns for which the referent was unclear. However, in the younger group (ages 5–6), those with RPL had a significantly greater frequency of ambiguous pronouns than either those with LPL or the TD group (see Figure 5.1).

![Figure 5.1 Pronominal use in biographical interviews: proportion of non-ambiguous pronouns used in interview. In the younger group, children with RPL use significantly fewer pronouns for which the referent is clear than either children with LPL or the control group. However, by the age of 7–8 years, there are no longer differences between groups.](image-url)
By the age of 7–8 years, this side-specific difference had resolved, even though both PL and TD children still have many unspecified referents for their pronouns. Considered together, these findings suggest that toddlers and preschoolers with PL are making good progress by school age in acquiring the structures of language, and, unlike the profile for adults with later acquired lesions (e.g., Rasmussen & Milner, 1977; Strauss, Satz, & Wada, 1990), a bilateral brain network is implicated in acquiring language functions. In contrast, as the PL children reach school age, RH injury appears to be associated with difficulties in discourse cohesion; this is true for both children with early lesions and adults with later acquired lesions. However, for the PL children, this deficit appears to resolve with development.

Continuing to investigate language production in school-aged children with PL, Reilly and colleagues (Reilly, Bates, & Marchman, 1998; Reilly et al., 2004) studied spoken narratives by asking children to tell the story from Mayer’s wordless picture book, *Frog, where are you?* (Mayer, 1969). Narratives constitute a more challenging discourse genre because they are monologic, as opposed to the interview (above) which is a dialogue. In telling a picture story, the child must choose what information to convey and then use appropriate linguistic structures to organize and convey the significant events in the story. The child must infer the motivations of the characters as well as the theme of the story; and these must be integrated linguistically into the recounting of the narrative. Reporting data from 52 children with PL, Reilly and colleagues found that the younger PL group (4–6 years) told shorter stories, and made significantly more morphological errors than age-matched controls. They also used complex syntax less frequently and used fewer distinct complex syntactic constructions than controls. Overall, there were no significant side-specific differences. As children reach the age of 8–9 years, performance by the PL group was in the low normal range for all linguistic measures (morphology and syntax) as well as those of narrative structure: the number of episodes included in the stories and explicit mention and integration of the theme of the story.

It is important to note that the types of morphological errors of the PL children were similar to those made by younger controls; that is, we did not find any extraordinary errors in the stories of the PL group. These findings suggest that the language acquisition process in the PL group follows a similar developmental course to that of TD children despite being mediated by different underlying brain structures. The findings for productive morphosyntax in this quasi-naturalistic narrative task are mirrored in experimental data reported by Weckerly et al. (2004). They tested English verb morphology using a Tag Question task (“He loves chocolate, doesn’t he?”) and reported performance in the low normal range for the PL group with no left-right differences. To summarize so far, with respect to the acquisition of morphology and syntax in naturalistic and experimental contexts, the PL group as a whole, regardless of lesion side, is performing in the low normal range by mid-school age. Further, to illustrate the degree to which children with PL have mastered English, and how children with LPL do not mirror adults with acquired LH injury, given below are some expressive language samples from the *Frog* narrative and biographical interview from a child with a large
perinatal, left hemisphere, middle cerebral infarct that involves frontal, temporal, parietal, and occipital regions.

*Frog story (4 years, 11 months)*
The boy is looking at the f'og.
The dog is looking at the frog too.
The ... the boy was s'leeping and the f'og comed out. Then he woked up and he didn't see the f'og.

*Biographical interview (7 years)*
Oh we really had fun ...
[there's] a dragon ride that's fun and it goes um fast but it doesn't go as fast as a roller coaster.

Returning to the pragmatic use of language, in addition to the pronominal use mentioned above, another index of discourse cohesion is how one uses complex sentences, as these make the relations between propositions explicit. For example, it is grammatical to say: "The boy was sleeping. The frog escaped." However, by adding a subordinate conjunction—for example, "While the little boy was sleeping, the frog escaped!" —the simultaneous relation between the events is clear. As they get older, typically developing children use complex syntax more frequently and with more diversity. In the cross-sectional data, the results show a trend wherein the TD and LPL group use more complex syntax in the older groups, but those with RPL appear to plateau and use complex syntax less frequently than those with LPL or the TD group. Complementing their cross-sectional narrative data, Reilly and colleagues (Reilly, Stiles, Wulfeck, & Nass, 2005) reported corroborating longitudinal narrative data from children at three data points (see Figure 5.2): Those with RPL use significantly less complex syntax than those with

![Figure 5.2](https://example.com/figure5.2.png)

**Figure 5.2** Use of complex syntax in narratives: a longitudinal study. In the youngest group, the children with both LPL and RPL are significantly below the TD group in their use of complex sentences. However, the children with LH injury make impressive developmental progress, and at the two older data points, only the differences between the RPL and TD group are significant.
LPL or controls. Why might this be the case? One function of complex sentences in a narrative is to tie or link different episodes in the story or to integrate the ongoing local event with the theme of the narrative, searching for the lost frog; for example: “When the boy climbed the rock to call for his frog, he grabbed the branches, but they were a deer.”

Thus, we propose that, similar to the increased use of ambiguous pronouns of the younger children with RPL in the interview data, this decrease in the use of complex syntax in narratives reflects an impairment in using language for discourse cohesion. Problems in discourse cohesion are also reported in adults with RHD (Gardner, Brownell, Wapner, & Michelow, 1983; Hough, 1990; Joanette, Goulet, & Hannequin, 1990; Kaplan, Brownell, Jacobs, & Gardner, 1990). However, just as the early pronominal impairment appears to resolve with age for the children, we expect a similar profile of development in the use of complex syntax as well. In responding to questions in an interview or telling a story from pictures, it is the child who chooses the words and structures he or she uses.

Online measures represent an alternative means of assessing language comprehension and grammaticality knowledge. Since online measures not only assess accuracy but also reaction time, they may be more sensitive measures of morphosyntactic abilities. Such tasks provide a measure of processing time and thus provide another dimension to our understanding of children's linguistic abilities. MacWhinney et al. (2000) administered an online battery of detection, recognition and word repetition, and picture-naming tasks and found that, overall, the PL group showed slight impairments and slower reaction times, although these differences resolved with age. Dick and colleagues (Dick, Wulfeck, Krupa-Kwiatkowski, & Bates, 2004) focused on complex syntax and assessed sentence comprehension in PL children; they focused their investigation on active and passive sentences as well as subject and object cleft sentences. For the easier sentences, active and subject clefts, the performance of the PL group was comparable to controls, but they were less accurate on the passive sentences, and their accuracy was significantly lower for the object clefts; reaction times mirrored accuracy. Wulfeck et al. (2004) investigated what school-aged children know about their language—that is, grammatical sensitivity—by asking children to make grammaticality judgments on sentences, half of which included determiner or auxiliary verb violations. The PL group showed lower grammatical sensitivity than controls, but they scored well above chance. Further, both the controls and the PL group improved with age.

In sum, these findings suggest subtle impairments in the processing of complex language for the children with PL; however, the differences largely resolve with age. As in the language production data, the children appear to catch up in individual aspects of language learning, and performance overall resembled that of younger, healthy controls. The similarity of the error patterns in both language production and the online interpretation tasks to those of younger, typically developing children suggests two notable points: (a) the process of language acquisition itself is fairly rigid and constrained, and (b) children with PL are approaching the problem of acquiring a language in a similar manner to typically developing children despite the recruitment of different brain areas.
To summarize: First, in the acquisition of grammatical structures, children with early focal brain injury do surprisingly well; their functional everyday use of language is in the normal range by the age of 8–9 years. The trajectory of language development in these children does not map onto that for adults with comparable damage; rather, we see initial delay in children with either right or left hemisphere injury, suggesting that acquiring language as opposed to maintaining the system, requires a bilateral cerebral network. After the early delay, language development in the PL group follows the same trajectory as seen in TD children: Early site-specific profiles largely resolve, and by mid-school age, the PL group performs in the low normal range. It is important to note that acquisition is not just "catching up;" rather, at each new level of linguistic challenge, we see initial delay and then subsequent development in mastering grammatical structures.

Second, in contrast to acquiring language, if we focus on how the PL group uses language, a different picture is emerging. Evidence is accruing that the right hemisphere mediates discourse integration in children, as is typical of adults. Thus, pragmatically—that is, in recruiting and employing language structures for discourse purposes—the PL children with RH injury subtly mirror the profile of adults with RH injury. Moreover, as in the acquisition of linguistic structures, an iterative profile is apparent with development: Early deficits resolve with age to appear again in new guises in more challenging discourse contexts.

Visuospatial Cognition in Children with Perinatal Brain Injury

Visual pattern processing

The ability to understand and interpret a visually presented pattern requires assessment of both the parts and the overall configuration, and analysis of how the parts are related to form a whole. In adults, the neural system for processing visual pattern information is organized bilaterally in the ventral occipitotemporal lobes (OT). The two cerebral hemispheres work cooperatively, but contribute differently to visual pattern processing. Within this bilateral network, the left OT region is dominant for processing parts or features, and right OT is dominant in processing pattern configuration. Studies with adults have shown that localized injury to this region results in specific disorders of visuospatial functioning (e.g., Arena & Gainotti, 1978; Delis, Kiefner, & Fridlund, 1988; Delis, Robertson, & Efron, 1986; Lamb & Robertson, 1988; McFie & Zangwill, 1960; Piercy, Hecaen, & De Ajuriaguerra, 1960; Ratcliff, 1982; Robertson & Delis, 1986; Robertson, Lamb, & Knight, 1988; Swindell, Holland, Fromm, & Greenhouse, 1988; Wasserstein, Zappulla, Rosen, & Gerstman, 1987). Injury to left OT brain regions results in disorders involving difficulty in defining the parts of a spatial array. Patients with left injury tend to oversimplify spatial patterns and to rely upon overall configural cues while ignoring specific elements. By contrast, patients with right OT lesions have difficulty with the configurational aspects of spatial analysis.
They fail to maintain a coherent organization among the elements, focusing on the parts of the pattern without attending to the overall form.

Children in the PL population present with a similar pattern of deficit as those observed among adult stroke patients (Stiles et al., 2006; Stiles, Reilly, Moses, & Paul, 2005; Stiles, Trauner, Engel, & Nass, 1997; Stiles-Davis, Janowsky, Engel, & Nass, 1988; Vicari, Stiles, Stern, & Resca, 1998). Specifically, children with right-sided injury have impairments of configural processing that are evident on a wide variety of tasks, while children with left-sided injury have difficulty with pattern detail. However, their deficits are milder than those observed among adults, and children appear to be able to compensate for their deficits to a degree that adults do not. The fact that children with PL manifest specific deficits of visuospatial processing suggests that the basic organization of the OT system is established early. But the subtle nature of their deficits and their capacity for compensation suggest that the neural system is also capable of adaptive organization. Studies examining visual pattern processing among children ranging in age from the preschool period through adolescence provide supporting evidence for subtle, persistent, but lesion site-specific deficits (Stiles & Nass, 1991; Stiles, Stern, Trauner, & Nass, 1996; Stiles-Davis, 1988; Stiles-Davis et al., 1988; Stiles-Davis, Sugarman, & Nass, 1985).

Studies of visual pattern processing have used “hierarchical” stimuli to examine part–whole processing in both typical and atypical populations (Figure 5.3, model). Hierarchical stimuli are large “global level” forms composed of appropriately arranged “local level” elements (e.g. a global level “H” made up of smaller local level “S”s). When asked to copy such patterns from memory, adults with right OT injury have difficulty with the global level form, while local level processing is impaired by left OT. Children with right or left hemisphere injury show similar patterns of deficit. Among children with right-sided injury, accuracy scores for the local level are comparable to those of age- and IQ-matched control children, but their global level accuracy is significantly lower (Stiles, 2008; Stiles et al., 2006). The children with left-sided injury showed the reverse pattern. These data suggest a dissociation in global–local processing deficits associated with early lateralized brain injury that is similar to that observed with adult onset injury. Further, data from a smaller cohort of children studied longitudinally confirmed the persistence of subtle processing deficits lasting into early adolescence.

Other studies of visuospatial processing confirm the early patterns of impairment, but also suggest that children may compensate for their deficits by developing alternative processing strategies. For example, at the age of 5 years, children with right-sided injury have difficulty with the simple task of drawing a house (Stiles et al., 1997). With development, they master the task but notably the houses they produce are very similar from year to year, suggesting that they have developed a graphic formula. Moreover, when the task is made more challenging, the limitations of that strategy emerge. When typical school-aged children, or children with LH injury, are asked to draw “an impossible house, a house that could not be,” the most common response is to alter the spatial configuration of the parts of the house. However, children with right-sided injury
rarely respond in that manner. Rather, they draw a house using their graphic formula, and provide verbal descriptions about why this house is impossible ("the kitchen is so hot you can't go in"); "it is on the moon"), or they draw a dot and say it is a tiny house, or they hand back a blank sheet of paper and say it is invisible. Altering the configuration of the house to make it "impossible" taxes a spatial weakness for which the graphic formula provided compensation. Their solutions to the task of making a house "impossible," while all reasonable and appropriate, diverge from that of typically developing children who most commonly employ a spatial solution.

Evidence for the use of alternative strategies is observed among older children using a more difficult construction task, the Rey–Osterrieth Complex Figure (ROCF; Plate 4, model). The ROCF was originally developed to examine spatial planning in adult patients with neurological disorders. The ROCF is a complex 2-D geometric pattern which is organized around a central rectangle that is symmetrically divided by a set of three bisecting lines. Additional pattern details are positioned both within and surrounding the core rectangle.

In the standard administration, patients are first asked to copy the ROCF with the model present; later, they are asked to reproduce the form from memory. The most
efficient strategy for copying the ROCF, the one adopted by most typical adults, is to begin with the core rectangle and bisectors, and then add pattern details. Studies of typically developing children (Akshoomoff & Stiles, 1995) have documented that they do not regularly use this advanced copying strategy until about 10–12 years of age. Younger children parse the figure into smaller units, but the size of the unit increases with age. Six-year-olds typically use a very piecemeal strategy, drawing each small subdivision separately, while older children use progressively larger subunits (quadrants, halves), until finally, by about 10–12 years, their organization strategy centers around the core rectangle.

Children with PL, regardless of side of brain injury, have particular difficulty with the ROCF (Akshoomoff, Feroe, Doyle, & Stiles, 2002). On the copying version of the task, the performance of all the children in the PL group was worse than normal age-matched controls. Deficits were particularly evident among the 6 to 7-year-olds, the youngest age tested. Their drawings were more poorly organized and included less detail than those of typical controls. Interestingly, there were few notable differences between the drawings of the children with left-sided injury and those with right-sided injury. Data were collected from this sample of children longitudinally between the ages of 6 and 12 years. Across the developmental period, the performance of all children improved considerably, such that by the age of 10 these children produced accurate copies of the ROCF. However, the strategies children used did not change significantly over the developmental period. At 10–12 years of age, the children with early injury persisted in using the piecemeal, immature strategy to generate their copy of the ROCF (see Plate 4, for example). The fact that the left and right lesion groups did not differ in their performance was interesting and unexpected. The ROCF is a challenging task, even for adults. It is likely that the underlying task demands tax both segmentation and integration processes. Thus, a deficit in either process would disrupt performance on the task.

Interestingly, while the copying task did not differentiate the lesion groups, the memory task did. Examination of the copy and memory data obtained when the children were 11–14 years old revealed a striking difference in performance on the two tasks for the children with left- but not right-sided lesions. The memory and copy reproductions for the children with right-sided injury were remarkably similar. For both tasks, children produced accurate copies of the ROCF using a piecemeal processing strategy. The memory data from the children with left-sided injury differed dramatically from their copy data. While the children produced accurate reproductions on the copying task by using a piecemeal strategy, in their memory reproductions the children organized their drawings around the core rectangle and provided relatively few additional details. The particular processing demands of the memory task thus revealed a characteristic left hemisphere visuospatial processing deficit. Specifically, the children encoded the core, global form but very few other details. By contrast, the copying task captured the children's strategies for compensating for that deficit. With a model available, the children adopted the most immature drawing strategy, but with that approach were able to produce relatively accurate copies of the target form.
Coding of spatial location in children with early focal lesions

Much less work has been done on the processing of dorsal stream spatial information in PL children. A number of studies that have been carried out on normal adults and children indicate that there are hemispheric differences in the coding of locational information. These lateralization studies indicate that the left hemisphere is advantaged at coding categorical spatial relations (e.g., above/below), whereas the right hemisphere is advantaged in coding coordinate spatial relations that retain precise metric information about the location of objects and the distance between objects (e.g., Hellige & Michimata, 1989; Koenig, Reiss, & Kosslyn, 1990). Consistent with these findings, Laeng (1994) reports that adult patients with right hemisphere damage are more impaired on coordinate spatial tasks and those with left hemisphere damage are more impaired on metric spatial tasks.

In a recent study, Lourenco and Levine (2007) asked whether young children with pre- or perinatal unilateral lesions show this same pattern. On their task, 3-, 4-, and 5-year-old PL children were shown the locations of different toys on a long, narrow rug, 5 ft (1.5 m) by 8 in (20 cm). Each toy was placed on the rug, and then removed by the experimenter. The child (who stood in different positions relative to the rug) was then asked to place that toy in the location that it had been previously placed. On a similar location reproduction task, typically developing 16 to 24-month-olds showed evidence of hierarchical coding of spatial location information (Huttenlocher, Newcombe, & Sandberg, 1994). That is, not only did they represent information about the exact location of the target object (e.g., fine-grained distance information), they also incorporated information about the larger spatial region (e.g., categorical information) in which the target was located. While the responses of typically developing children are generally highly accurate on this location reproduction task, the responses are also consistently biased toward the center of the space. The bias would only occur if children treated the long, narrow space as a single category with a prototypic location at the center. Although such hierarchical coding leads to bias in responses, when there is uncertainty about the fine-grained distance information, using information about a prototypic location within a category has the effect of increasing overall accuracy (for review, see Huttenlocher & Lourenco, 2007).

Studies of 3 to 5-year-old typically developing children show that they are highly accurate on this task. They show category effects, but unlike younger children, they divide the space into two categories (the left half and the right half), each with a prototype at the center. Thus, they show bias toward the center of each of these categories for object locations within each half (see Huttenlocher & Lourenco, 2007). Similar to typically developing children, 3 to 5-year-olds with early left hemisphere injury showed very accurate performance. However, rather than reflecting two prototypic centers (one for the left half of the space and one for the right half), their category effects were similar to those shown by typically developing 16 to 24-month-olds (i.e., bias toward the center of a single category). Three- and 4-year-old children with RH injury showed much less accurate performance than those with LH injury, but also showed marked
category effects of the kind shown by typically developing 16 to 24-month-olds. By 5 years of age, the RH children, like the LH children, were highly accurate and they, too, continued to show the kind of category effects shown by typically developing toddlers. Interestingly, even at 5 years of age, neither the LH nor the RH children showed the more complex category effects that were apparent in the typical 3 to 5-year-old children. We are currently exploring whether this more complex kind of hierarchical coding emerges later in development in the brain-injured children. Thus, similar to the pattern processing deficits described above, children with early brain injury show site-specific deficits in the coding of spatial location. There is also evidence of plasticity as children with RH injury are initially delayed in coding the locations of objects in space, but show improvement across age. It remains an open question whether there is plasticity in the nature of the hierarchical coding used.

Lourenco and Levine (2007) also gave these same PL children a mapping task. On this task, the children were given a strip of paper indicating the location of a toy and were asked to place the toy on the larger rug in the corresponding location. Interestingly, the results obtained on this mapping task were markedly different from those obtained on the spatial location task. In particular, 3- and 4-year-old children with left and right hemisphere injury showed evidence of contralateral neglect. That is, they placed the objects on the side of the rug that was ipsilateral to their lesions, regardless of the location indicated on the map. By 5 years of age, this neglect had resolved in children with RH and LH lesions. Again, resolution of an initial deficit provides evidence of plasticity. However, the asymmetrical neglect evident on the mapping task differs from the preponderance of neglect following right hemisphere lesions in adult patients (see also Trauner, 2003). Further, the neglect observed on the mapping task was not observed on the object location task, suggesting a dissociation that depends on how the locations were initially coded.

In summary, these longitudinal data on spatial location coding and visual pattern processing indicate that, with development, children with left- and right-sided injury show both early deficits and considerable behavioral improvement, eventually achieving ceiling level performance on many spatial tasks, at least when accuracy is used as the measure of performance. However, the time course over which this improvement occurs is often protracted. Further, when process is used as the metric of performance, it becomes evident that the PL children frequently adopt compensatory strategies to facilitate performance, and when those strategies are taxed, persistent deficits in performance are again observed (Akshoomoff & Stiles, 2003).

**Emotion Processing**

Studies of patients with unilateral lesions acquired during adulthood, as well as those from healthy adults, overwhelmingly find that the right hemisphere, especially frontoparietal regions, are critical for emotion recognition (e.g., Adolphs, Tranel, & Damasio, 2003; Blonder, Bowers, & Heilman, 1991; Borod, 1992, 1993; Borod et al.,
Face processing

Faces are critical in both conveying and interpreting emotional signals. As such, how one processes faces per se can influence the processing of emotional information on the face. Typically, face processing is mediated by occipitotemporal regions of the brain, including the fusiform gyrus: As a spatial pattern, such processing relies more on holistic and configural processing than featural processing, at least by school age (Carey & Diamond, 1977; de Haan, 2001). As such, many studies report an RH bias for face processing; in fact, infants have shown an RH bias for face processing as early as 4 months of age (de Schonen & Mathivet, 1990). A study of face processing in a small sample of children with PL (Mancini, de Schonen, Deruelle, & Massoulie, 1994) suggests long-term face-processing deficits. In a larger sample of PL children and adolescents, Paul et al. (2007) conducted a reaction-time study of face matching to examine whether the ability to identify faces might be differentially affected in children with PL. Children were presented with a series of three black-and-white face photographs, then two memory set items, followed by a probe. Participants were asked to indicate whether the probe face matched one of the faces in the memory set. Data from 42 participants between 9 and 23 years of age matched for age and IQ were studied (RH = 11; LH = 15; controls = 16). The groups differed significantly on both accuracy and reaction time. Children with RH injury were significantly less accurate and slower than controls, while children with LH injury were marginally less accurate than controls, but did not differ in speed. The two lesion groups did not differ from one another in accuracy or reaction time. These data suggest a face-processing deficit in both groups, which is somewhat more severe in the RH group.

Expression of emotion in infants and toddlers with perinatal brain injury

As typically developing children approach their first birthday, they produce their first words and use facial expressions both to convey and to interpret emotions. To characterize emotional expression in infants and toddlers with PL, we have conducted naturalistic studies of emotional facial expression and vocalizations using videotaped
recordings of mother–infant dyads in free play (Reilly, Stiles, Larsen, & Trauner, 1995). Focusing on facial expressions, 24 infants (6–22 months; 6 with RPL; 6 with LPL; and 12 age- and gender-matched controls) were videotaped. The Facial Action Coding System (FACS; Ekman & Friesen, 1978) was used to code infant and maternal facial expressions. As an index of expressivity, infants’ and toddlers’ positive responses to their mothers’ bids for interaction were coded. All children smiled in response to their mothers; however, those with left posterior injury and controls smiled easily and often, but those with right posterior injury smiled infrequently (Figure 5.4). These data were further confirmed by longitudinal case studies; together they suggest that the right hemisphere mediates emotional expression from as early as 6 months of age and that valence (positive/negative) is a significant factor in the early organization of emotions. Such findings are consistent with infant perceptual data that show a right hemisphere bias for face perception from 4–9 months of age (de Schonen & Mathivet, 1990).

Infants and toddlers use not only the face, but also the vocal channel to express emotions. Reilly and colleagues (Reilly, Anderson, & Martinez, 2007) have used free-play mother–infant interactions as a context to collect infant vocalizations from 23 infants with PL (13 LPL, 10 RPL) aged 9–16 months. Listening to 2,200 vocalizations extracted from videotapes, independent judges evaluated the children’s vocal productions as positive, negative, or ambiguous in quality. As in the facial-expression data, control infants and infants with LPL produced more positive than negative vocalizations. The RPL group produced significantly more negative vocalizations than either of the other groups. The children with RPL who also had basal ganglia involvement were most responsible for this profile. These findings are complemented by a study of temperament in the PL group that found increased negative affect in infants and toddlers with RH injury (Nass & Koch, 1987). In sum, the expressive data are consistent across

Figure 5.4 Expressing positive emotions: smiles in response to maternal bids for interaction. Typically developing children and children with LPL smile significantly more often than children with RPL. Note also the range of expressivity in the TD group compared to the children with brain injury. Both the LPL and the RPL groups demonstrate more homogeneous profiles than their typically developing controls.
productive channels for emotion: face and voice. They suggest that the neural substrates for emotion are established at least by the middle of the first year of life, and, importantly, that profile of deficit is not modality specific, but rather general to the domain of emotion.

The previous sections of this chapter were devoted to the developmental profiles in the PL group of specific cognitive domains: language, space, and emotion. In the following sections, we examine two areas in which two or more of these cognitive systems are recruited: labeling emotions and developing literacy.

**Language, Spatial Cognition, and Emotion: Labeling Emotions**

A traditional measure of evaluating emotion processing involves supplying emotional labels for static photographs of people's faces conveying different emotional expressions. As such, this task draws on three cognitive systems, requiring (a) spatial analysis, (b) recognition of the emotion, and (c) recruiting the appropriate linguistic form. Free labeling data from 24 PL and 24 TD children aged 4–8 demonstrate how characteristic behavioral profiles in each of these systems play a role (Reilly, Charten, Stiles, del Guercio, & Nass, 2005b). In this task, the stimuli included photographs of the same woman demonstrating the following emotional expressions: happy, sad, angry, afraid, surprised, and neutral. For the TD children, there is a slight improvement in overall performance with age, and, to our surprise, the children with RPL generally perform like controls. Interestingly, it is the younger children (aged 4–5) with LPL who fare the worst (see Figure 5.5) in labeling emotional expressions. Even though those children with RPL appear to have deficits in producing emotional facial expressions in infancy, it is the young children with LPL who are impaired in labeling emotional expressions.

![Figure 5.5](image)

**Figure 5.5** Labeling facial expressions: all emotions together. In the younger age group (4:00–5:11 years), the children with RPL perform comparably to controls, but those with LPL perform significantly worse than either children with RPL or TD. However, at the older data point (age 6:00–7:11 years) performance is comparable in all groups.
Several possible explanations come to mind: The performance of the LPL group may reflect a subtle deficit in lexical production as noted above, or perhaps it may be a reflection of their problems with isolating and identifying features within a spatial array, as discussed above (Stiles et al., 2006). Importantly, the early deficit evident at 4–5 years is no longer apparent in the older group (ages 6–7), again demonstrating the plasticity of development following early lesions.

**Literacy: The Intersection of Language and Space**

Literacy is the major academic challenge for school-aged children. A traditional definition of literacy includes the ability to read and write. Thus, the acquisition of literacy involves learning to map spoken language onto its visual counterpart. For beginning readers, the task involves learning letter forms, learning letter–sound correspondences, and using these skills, along with language comprehension skills, to decode and understand written texts. Overall, reading involves the coordination of orthographic, phonologic, semantic, syntactic, metalinguistic, and comprehension skills (e.g., Snow, Burns, & Griffin, 1998). Thus, reading achievement is related to both the acquisition of code-related skills (letter–sound correspondences) and also to oral language comprehension skills (e.g., Gough & Tunner, 1986; Joshi, Williams, & Wood, 1998; NICHD Early Child Care Research Network, 2005; Storch & Whitehurst, 2002). Code-related skills in the preschool years, including phonologic awareness and letter identification, are significant predictors of later reading achievement (e.g., Whitehurst & Lonigan, 1998) as are early language skills, including lexical, syntactic, and narrative skills (e.g., Dickinson & Tabors, 2001; NICHD, 2005; Storch & Whitehurst, 2002).

Whereas a large body of literature exists on reading and its development in typically developing youngsters, much less is known about the development of reading in children with early unilateral brain injury. The vast majority of studies have focused on the development of language skills, mainly in the toddler and preschool years, whereas literacy acquisition occurs during primary school. The little information that exists about the development of literacy skills in this population presents a mixed picture, perhaps due to the variety of different kinds of reading assessments that have been used. Using the Woodcock–Johnson Psycho-Education Battery, Aram and Ekelman (1988) found that children with right hemisphere lesions performed significantly lower than control children on the Reading and Math Clusters as well as the Written Language Cluster (dictation and proofing), whereas children with left hemisphere lesions performed significantly lower than control children only on the Written Language Cluster. Woods and Carey (1979) administered a reading test (Sentence Completion test from Boston Diagnostic Aphasia Examination) and a spelling test to children with pre- or perinatal left hemisphere injury. Relative to controls, the children with early left hemisphere injury were significantly impaired on the spelling test, but not on the reading test. Of note, the reading comprehension tests used by Aram and Ekelman and Woods and Carey involved reading a sentence and selecting the best completion from multiple
alternatives. This type of "cloze" procedure has been shown to load more heavily on word recognition/decoding skills than on language comprehension skills, whereas the reverse loading pattern is found for measures involving passage reading followed by comprehension questions (e.g., Bowey, 1986; Cutting & Scarborough, 2006; Francis, Fletcher, Catts, & Tomblin, 2005). Thus, it is possible that children with PL have greater difficulty with the kind of reading tasks that load more heavily on oral language comprehension skills, but this issue is in need of investigation.

Booth et al. (2000) found that children with large pre- or perinatal left hemisphere lesions scored lower on reading decoding tasks (e.g., Woodcock–Johnson Word Identification and Word Attack) than those with smaller lesions, even though the receptive vocabularies of the children with large lesions were in the normal range. Based on this finding, Booth et al. suggest that early developmental plasticity may be of more benefit to earlier developing language skills than to reading. Consistent with this possibility, Levine and Fisher (2007) have found that school-aged children with early brain injury (n = 17; 13 with left hemisphere injury, 4 with right hemisphere injury) performed significantly below age norms on reading decoding and comprehension tests even when they performed within the normal range on verbal IQ tests at 6–9 years of age.

Whereas reading is the recognition of language in the visual modality, writing requires its creation and production. In typically developing children, preschoolers already distinguish writing from drawing and numerals (Tolchinsky, 2003), but it is not until the age of 5–6 that they acquire the alphabetic principle (Tolchinsky, 2003; Treiman & Bourassa, 2000). This is the concept that letter strings (words) have particular meaning, and specifically that graphemes (letters) represent the phonemes of a word. Typically, children map the sounds of English onto their graphic forms (letters) in a series of stages (Sprenger-Charolles & Bechennec, 2004) wherein, first, entire syllables are represented by one letter (e.g., car may be written as k), then by multiple consonants (car is now kr); next, vowels appear, (kar); and, finally, English spelling conventions emerge: car. Unlike Spanish or Turkish, where sound–letter correspondences are regular with one-to-one mapping, the English spelling system is a holdover from the pronunciation of fifteenth-century English. Our pronunciation has changed dramatically, but our spelling has not. The result is an opaque relationship between the spoken and written forms (consider knife, lamb, phone, ghost, wreath vs read, where vs wear, and box vs socks or weight vs wait).

Earlier studies investigating writing in PL groups have used standardized achievement tests to assess writing. As noted above, on standardized tests, both the Aram and Ekelman (1988) and Woods and Carey (1979) studies found that children with left hemisphere injury had more difficulty with dictation and spelling than those with right hemisphere injury or controls. A more recent study by Frith and Vargha-Khadem (2000) looked at both French- and English-speaking children with unilateral brain injury (both early and during childhood). They found no significant differences in spelling performance according to lesion site, or the age at which the lesion was acquired; however, they did identify a sex-by-side interaction. Boys with left hemisphere
injury performed more poorly than those with right hemisphere injury (who performed like controls), whereas girls showed the reverse profile but did not differ significantly from controls. Such findings suggest that sexual dimorphism may play a role in literacy skills from early in life.

Taking a broader view of written discourse, we have begun to trace writing development in a group of PL children aged 8–16 (Reilly, Stiles, Fenson, & Nass, 2005) by asking them first “to tell about a time when they had a problem either at home or at school, how it started, what happened, and how it ended.” After the child has told a story, the child is asked to write the same story. Constructing a narrative spontaneously is a more challenging task than telling a story from pictures (as in the frog story above). As in the frog story data, in this older group of children, there were no significant right–left differences; moreover, in the spoken personal narratives there were no significant differences between the stories of the PL and TD groups with respect to story length (in propositions), frequency of complex sentences, diversity of complex syntax, nor in the structure of the narratives, although the children with PL required more prompts to provide a resolution to their spoken narrative.

The most striking differences in the profiles of PL and TD children become apparent when spoken and written narrative texts are compared. Whereas the spoken stories do not differ significantly across groups with respect to linguistic measures, in the written form, the stories of both groups are shorter than the spoken form, but those of the PL group are significantly shorter than those of the TD group. With respect to recruiting complex sentences, we found that written stories from the PL group have proportions of complex syntax that are in the low normal range. Interestingly, when we calculated the types of complex syntactic structures recruited by both groups, we found no differences across groups in either spoken or written forms.

An analysis of narrative structure (that is, the degree to which children include basic components of narrative: setting, problem, complication, and resolution) reveals no site-specific differences. Moreover, neither group, PL or TD, tells or writes narratives that include all components. However, in the spoken modality, the stories from the PL group do not differ significantly from those of the TD children. In contrast, in the written modality, the TD group performs slightly better than in the oral modality, while the PL group fares worse in the written than the oral, reflecting the challenges that writing presents to these children. Thus, whereas writing as a process permits planning and revision, without the online pressures of spoken language, it appears that only the typically developing children were able to take advantage of the possibilities offered by the written modality.

Gradients of Plasticity: Language, Space, and Emotion

Looking across the three behavioral domains discussed above—language, spatial cognition, and emotion—the profiles of the PL group are distinctive, and responses to our original questions will differ for each of the domains.
For language acquisition (that is, acquiring the morphology and syntax of language), children with either right or left hemisphere injury are initially delayed, and a bilateral network is implicated for the acquisition of language. Their profile is unlike adults with late acquired strokes. Site-specific profiles evident early in development resolve, and by mid-school age, language performance for the PL group overall is in the low normal range. Importantly, children with early left hemisphere injury do not mirror the pattern for adults with acquired left hemisphere injury. However, if we consider how language is used in discourse, children with RPL are more likely to resemble the pattern seen in patients with right hemisphere injury acquired in adulthood, albeit more subtly. At different developmental stages, there is evidence of impairment with subsequent development in the integration of discourse. Although the basic nature of the integrative deficit appears to be persistent, how it manifests is dependent on the child’s level of development.

In the domain of spatial cognition, although we see clear evidence of development, the children’s profiles much more closely mirror that of adults with similar injury. This profile is evident for spatial analysis of patterns and for the coding of spatial location. The profile is one of deficit and subsequent development to achieve specific spatial milestones; however, the characteristic deficits are again revealed with increasingly challenging tasks.

Similar to the visuospatial profile, for the domain of face processing and emotion, the PL children again follow the adult path for both simple face matching and for processing emotions. Children with right hemisphere injury are significantly more impaired on simple face-matching tasks than controls, although it is notable that children with LH injury also have subtle face-processing deficits. Early RH injury also impacts at least the early development of affective processing as is evident in a decrease in the production of positive and an increase in the production of negative facial expressions and vocalizations in this group of children.

Finally, considering tasks that tap multiple cognitive systems (e.g., labeling emotions), if we probe at the particular developmental point at which the necessary skill is still developing, deficits within the individual domains become apparent. Decrements in later developing tasks drawing on multiple cognitive systems, such as reading and writing, may reflect declining plasticity and/or the result of decreased functional neural tissue to handle increased processing demands. By looking at development across several cognitive systems, we see strong evidence for development in all three cognitive domains. None the less, the three functional systems reflect differing degrees of developmental flexibility with language acquisition showing delay regardless of lesion site and then developing fairly rapidly; in contrast, spatial cognition and emotion mirror the adult model, albeit more subtly. Such contrastive behavioral profiles suggest differing degrees of brain plasticity: The findings for language acquisition, but not language use, provide clear evidence that, although perhaps not optimal, multiple brain regions can assume core language functions. This does not appear to be the case for either visuospatial cognition or for emotion processing.

Several possible explanations come to mind for these differential profiles. The first is an evolutionary perspective: Emotion and space are older systems, whereas language
represents an evolutionarily newer invention. As our late colleague, Elizabeth Bates often noted, “language is a new system invented from old parts.” It may also be the case that systems that are dependent on a particular sensory modality (e.g., face perception) are by definition less flexible than those, such as language, that are amodal (language exists in spoken, signed, or written forms). Another finding that emerged from a cross-domain perspective is the possibility of a more general characterization of the children with RH injury. If we consider their recurrent problems with discourse cohesion and their visuospatial impairments, both might be characterized as an integrative deficit that persists across cognitive domains, reflecting the global processing of the right hemisphere.

In sum, children with PL provide a unique context for investigating neuroplasticity. Tracing development across cognitive systems in the PL group has demonstrated that neuroplasticity is not a unidimensional construct. Rather, a gradient of plasticity exists, both across, as well as within, cognitive systems. For example, for language acquisition, the developing brain shows impressive flexibility; however, even within language, if we focus on its use in discourse, we see differential degrees of adaptive flexibility. As such, chronicling behavior in the PL group provides us with important clues as to which specific brain areas might be privileged for specific cognitive functions, as well as those for which the developing brain is more flexible.

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References


The Neuropsychology of Language Development

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At its most fundamental, language can be defined as a system composed of “words and the methods of combining them for the expression of thought” (New Oxford American Dictionary, 2005). In general, typically developing children will rapidly and comprehensively master at least one of the over 6,000 languages that exist around the globe. The complexity of these language systems, and the speed and apparent facility with which children master them, have been the topic of philosophical and scientific speculation for millennia. In AD 397, in reflecting upon his own acquisition of language, St. Augustine wrote “as I heard words repeatedly used in their proper places in various sentences, I gradually learnt to understand what objects they signified; and after I had trained my mouth to form these signs, I used them to express my own desires” (quoted in Wittgenstein, 1953). St. Augustine’s intuitions notwithstanding, more recent thinking and research on children’s language acquisition suggest that the problem facing a child is much more intricate than simply remembering the association between a sound and an object and learning to reproduce the word’s sound. The rich and multilayered nature of this problem—and the many and varied paths to its solution (Bates, Bretherton, & Snyder, 1988)—make the process of language acquisition a unique window into multiple low and high level developmental processes. Indeed, studies of language development have provided unparalleled views into broad neural and behavioral change in response to input and consolidation, to injury, and the process of emergent organization and reorganization that unfolds over developmental time.

In this chapter, we will chart the multiple waves of change in language comprehension and production, beginning at birth with studies of speech perception, moving through babbling, phoneme and word discrimination into the dawn of word comprehension and production, and the subsequent emergence of syntactic and pragmatic abilities. We will also look at language’s “fellow travelers,” skills such as social cognition, gestural communication, and environmental sound recognition that appear to presage or accompany linguistic milestones. We will also consider the neural bases underlying early (mostly electrophysiological studies) and later language development
(predominantly functional magnetic resonance imaging). In particular, recent neuro-imaging literature increasingly demonstrates the importance of experience and learning on the development of the neural correlates of language development, as well as the absence of any straightforward and task-independent, language-specific neural substrates. The section on typical development will focus, in part, on the impressive degree of individual differences in language learning—something that is of prime importance when evaluating language development in atypical populations. We will also highlight the importance of the structure and statistics of the input to multiple levels of language learning.

In the second half of the chapter, we turn to language acquisition in the face of disease, injury, or congenital disorder. First, we review studies of “exogenous” challenges to children’s language learning. These challenges include early exposure to neurotoxins or pathogens, as well as enforced sensory or social deprivation. The subsequent section presents a brief overview of the known effects of childhood traumatic brain injury (TBI) and surgical hemispherectomy on language learning. In the third section, we consider the trajectory of language development in four congenital developmental disorders (autism, Down syndrome, fragile-X syndrome, and Williams syndrome), with a special focus on the particular linguistic strengths and weakness observed in each population at different epochs of development. Finally, we survey the field of “specific” language impairment and its relationship to other developmental disorders. In all four sections, we explore the impact of genetic, neurological, and environmental influences on the developing system, showing how children’s language acquisition can sometimes be dramatically delayed or deviant in the seeming absence of severe neurological, genetic, or environmental abnormality, but also how language acquisition is in some cases amazingly resilient to such insults. We close with a short overview of the literature on best clinical practice for the treatment of language disorders, and the potential directions for future research.

Language Acquisition in Typical Development

Precursors to language

The onset of language development is not signaled by the child's first word. Rather, even before birth infants are adapting to their language environment, mastering the necessary prelinguistic building blocks that support later language learning. As we will see in this section, over the first year of life children make huge strides in constructing the social, perceptual, and attentional tools that language needs to get off the ground.

1 Please see also Chapter 5 of this volume for language and visuospatial development following perinatal focal lesions.
Even before and shortly after birth it is possible to see the effects of experience-dependent speech discrimination. For instance, the heartbeats of at-term fetuses tend to increase in response to hearing their mother’s voice (Kisilevsky et al., 2003). Infants as young as 4 days old can use rhythm to discriminate between familiar and unfamiliar languages (Nazzi, Bertoncini, & Mehler, 1998); similar skills have also been noted in tamarin monkeys (Ramus, Hauser, Miller, Morris, & Mehler, 2000). Newborn infants also demonstrate evidence of phonetic categorical perception for the vowels and consonants from all human natural languages (Eimas, Siqueland, Jusczyk, & Vigorito, 1971; see Kuhl, 2004, for a review). Infants, like adults, classify sounds into different categories. That is to say, as some physical characteristic of a phonetic contrast varies along a continuum (such as voice-onset time), we do not hear gradual variation in the sounds but instead a sharp change from one sound to another. Again, this ability is not specific to humans: Monkeys (Kuhl & Padden, 1983) and chinchillas (Kuhl & Miller, 1975) demonstrate similar phonetic categorical perception as infants. One suggestion is that human phonetic categories have evolved around more general characteristics of mammalian sensory systems (Dick, Saygin, Moineau, Aydelott, & Bates, 2004; Kuhl, 1986; Smith & Lewicki, 2006; but see also Fitch & Hauser, 2004, and Tomasello, 1999, for evidence of more human-specific abilities that may play a part in language development).

Starting around the half-year mark, infants’ ability to distinguish between all phonemes begins to disappear, so that by around the first year infants form a strong bias toward native language-specific phonetic perception, beginning with vowels and subsequently extending to consonants (for a review, see Werker & Desjardins, 1995). For instance, at 6 months an infant exposed to a Japanese-speaking environment can distinguish between the English /r/ and /l/ sounds, but by 12 months the same child discerns only a single phoneme, unlike an infant reared in an English-speaking environment. Furthermore, children’s abilities to make such phonetic classifications in their native language at 7 months positively predicts language outcomes, such as word production, mean length of utterance (MLU), and sentence complexity between 14 and 20 months, whereas ability on non-native phonetic contrasts is inversely related to later language measures (Kuhl, Conboy, Padden, Nelson, & Pruitt, 2005).

What underlies these developmental changes in infants’ auditory discrimination? There is growing evidence that in the second year of life, infants’ phonetic discriminations in their native language increasingly rely on the stochastic distributional information available in natural speech. Although the actual examples of any given phoneme that an infant hears vary considerably along many acoustic dimensions, they tend to conform to general statistical distributions that the infant can use to identify the most informative boundaries for distinguishing phonemes (Jusczyk, Luce, & Charles-Luce, 1994; Kuhl, Williams, Lacerda, Stevens, & Lindblom, 1992; Maye, Werker, & Gerken, 2002; Saffran & Thiessen, 2003).

While infants do lose the ability to make discriminations for phonetic contrasts in all the world’s languages at around the first birthday, this is not a straightforward example of a “critical” or “sensitive” period in brain maturation. An elegant combined behavioral and fMRI study by Pallier and colleagues demonstrated that Korean-speaking
children who were adopted into French-speaking families between the ages of 3 and 8—importantly with no further exposure to Korean—did not differ from children born into French-speaking families when both were tested on phonetic contrasts in Korean and French as adults (Pallier et al., 2003).

In addition to learning to segment the speech stream into meaningful language, children are also faced with the task of producing meaningful speech themselves. The early precursors of productive language start with infants’ preverbal vocalizations. From around 3 months, infants begin producing vowel sounds, and appear to be able to imitate adult-modeled vowel sounds (Kuhl & Meltzoff, 1996). From 6 to 8 months infants start babbling—making consonant–vowel combinations, for example, ba, ata. This early babbling is not obviously communicative, often occurring when the infant is on his or her own. As with phonetic perception, over the first year the sounds the infant produces move from being “universal” (with respect to all of the world’s languages) to increasingly resembling the sounds of the language(s) spoken around them. Infants produce native language-specific vowel and consonant sounds before they produce their first words, thus internalizing the acoustical or phonetic patterns of the language they are exposed to (Boysson-Bardies, Halle, Sagart, & Durand, 1989; Boysson-Bardies & Vihman, 1991).

Although social development is not a direct precursor of word or syntax comprehension or production, it is entwined with language across early development. Indeed, the early beneficial effects of social context on language learning are evident in infants’ vocalizations and phonetic discriminations. For instance, Bloom, Russell, and Wassenberg (1987) showed that turn-taking can alter very young infants’ vocalizations, and Goldstein, King, and West (2003) showed that parental feedback increases infant vocalizations. In addition, Kuhl and colleagues (Kuhl, Tsao, & Liu, 2003) found that North American infants learned non-native Mandarin phonemic contrasts in the presence of a Mandarin speaker but not from a video recording of the same information.

At around 3–6 months, social cognition in infants is perhaps most evident in gaze following, first directed at nearby targets (D’Entremont, Hains, & Muir, 1997) and expanding to further targets by around the first birthday (Corkum & Moore, 1995). (As with other skills, this feature is not uniquely human, and is present in at least several other nonhuman primate species; Tomasello, Call, & Hare, 1998). Starting around 9 months, we also see a change from dyadic interactions (i.e., the infant interacting with another object or another person) to triadic interactions (i.e., the infant and a caregiver jointly attending to each other and an object; Treharthen & Hubley, 1978).

From the end of the first year and beyond, children become increasingly adept at understanding and directing other people’s attention, using this information to make the task of language learning more tractable (Bates, 2004). In a longitudinal study of infants from 9 to 15 months, Carpenter, Nagell, and Tomasello (1998) showed a linked progression of gesture and joint attention, from infants initially sharing attention, to subsequently following an adult’s attention, to directing another’s attention (for a detailed review, see Tomasello, Carpenter, Call, Behne, & Moll, 2005). Slightly older
infants use an adult’s communicative intent to rapidly attach meaning to novel words (for a review, see Tomasello, 2001). Given the prominence of joint attention and its relation to language development, it is not surprising that the quantity and type of joint attention between infant and caregiver predict children’s early communicative abilities, with particular gains when the caregiver focuses on the object of the infant’s attention (Carpenter et al., 1998; Tomasello & Todd, 1983).

First words

As the infant’s native language discrimination improves, the child faces the daunting task of using these phonemes to segment speech into words, and attach some meaning to them. While adult listeners tend to perceive the speech stream as a series of discrete words presented one after another (at least in their native language), human speech actually affords no such luxury, in that there is generally no one-to-one mapping between pauses or silences and word boundaries. Despite this fact, by 7½ months, infants can detect words that they have been familiarized with from a stream of natural speech (Jusczyk & Aslin, 1995), and furthermore demonstrate longer-term memory for these words (Jusczyk & Hohne, 1997). The speech stream contains a number of different clues as to the location of word boundaries, such as syllabic stress patterns (Jusczyk, Houston, & Newsome, 1999) and phonetic transitional probabilities (i.e., the likelihood that one phonetic segment follows another; Saffran, Aslin, & Newport, 1996; see also Saffran, Johnson, Aslin, & Newport, 1999, and Hauser, Newport, & Aslin, 2001, for evidence of similar statistical pattern learning for non-speech stimuli and in nonhuman primates). Infants use a combination of these different cues to segment speech into words, with the relative weighting of cues varying over development (Johnson & Jusczyk, 2001; Thiessen & Saffran, 2003).

The purpose of this segmentation is, of course, to identify the chunks of speech (words) to which meaning can be attached and/or extracted. Infants become increasingly rapid and skillful at forming these word-to-meaning associations. Indeed, by around 2 years (and possibly earlier), infants demonstrate fast mapping (Carey, 1978) whereby word-to-meaning mappings are learnt after a single exposure. As with previous examples, this skill is neither specific to language (Markson & Bloom, 1997) nor specific to humans (Kaminski, Call, & Fischer, 2004).

As any proud (yet weary) parent will attest, young children are not just consumers of language, but also use it increasingly productively to communicate their needs, desires, and interests with others. Whereas word comprehension typically starts around 9–10 months, word production typically follows several weeks later (Fenson et al., 1994). In general, the size of a toddler’s early receptive vocabulary maintains a healthy numerical superiority over his or her productive vocabulary, although there is considerable individual variability in the extent of this relationship (Fenson et al., 1994). Infants’ early productive vocabulary is mostly composed of nominal labels for objects or people, although they also produce non-nominal words (for instance, the relational label “up”). However, straightforward classifications of infants’ language in terms of adult linguistic
categories, such as verbs or nouns, are probably inaccurate. Since infant speech is driven primarily by the desire to communicate, Tomasello (2006) argues that many of the early one-word utterances are actually “holophrases”—expressing a holistic communicative function with a single label. For instance, an utterance such as “up” might serve as the infant’s shorthand for an adult phrase such as “pick it up.” Thus, the infant may be copying a part of the adult phrase as a way to express the communicative intent of the phrase as a whole.

Individual variability, developmental trajectories, and the vocabulary “burst”

As noted at the start of this chapter, in order to understand the mechanisms underlying both typical and atypical language development, it is vital to have an understanding of the trajectory of that development, both in the “average” child and in individual children. The MacArthur–Bates Communicative Development Inventory (CDI) provides an excellent and carefully normed method of tracing an individual child’s linguistic developmental trajectory from the tentative start of meaningful communication around the first birthday (for the “average” child) through the advent of complex sentence production and comprehension. An instrument based on parental report (and validated through laboratory observation; Fenson et al., 2000), the CDI is extremely useful for comparing typical and atypical populations, and for assessing individual variation within a given sample.

One interesting finding of the initial MacArthur–Bates CDI norming studies (Fenson et al., 1994, 2000) is that there is little overall difference between girls and boys in terms of the trajectory of language development. Girls are on average one month ahead of boys, but this difference accounts for only 2% of the variation within and across age groups. Thus, these gender differences are relatively insignificant compared to the much greater variation between individuals. It is worth emphasizing how much individual variation there is between “typically” developing (TD) children—indeed, the idea of “typical” language development is something of a useful fiction. As an illustration of this, we reproduce here the cross-sectional growth curves from the Fenson et al. (1994) monograph showing the receptive and productive vocabulary size of children in the 10th, 25th, 50th, 75th, and 90th percentiles (Figures 8.1 and 8.2); we also show longitudinal growth curves for three different typically developing children for comparison (Figure 8.3).

The use of statistical averages to simplify complex and highly variable time series can sometimes mask more interesting phenomena. A case in point is the sudden acceleration at 16–20 months in a child’s vocabulary, the so-called vocabulary burst that follows a period of very gradual increases in vocabulary size following the first few words. This vocabulary acceleration involves not just an increase in the total number of words a child produces, but also changes in the content of the words, with a shift to a greater proportion of adjectives and verbs (Bates et al., 1988; Feason et al., 1994; Hampson & Nelson, 1993; Nelson, 1981; Tomasello, 2006). This sudden change in vocabulary size has often been considered an indicator of the onset of a new cognitive ability, such as

developing a "naming insight" (Dore, 1974). However, this "average" picture masks the wide gamut of individual developmental trajectories observed with the CDI for so-called "normal" children (i.e., children without obvious language problems).

There is wide individual variation in productive vocabulary size at the point when the "average" child launches his or her vocabulary burst. At this time in chronological development (~16 months), children in the highest 10th percentile produce about 180 words, whereas those in the lowest 10th percentile produce fewer than 10 words. It is very important to note that, despite this early variation, most of these children—including those in the 10th percentile who are slow getting language off the ground—will go on to have similar language outcomes as adults as their initially more able peers. There is also massive variation in the shape of the growth curves for different individuals' productive vocabulary with age. Some children show a recognizable burst, whereas others' vocabulary appears to grow at a much steadier pace, with still others advancing in a series of small successive bursts. A strong possibility is that the relationship between vocabulary size and age is inherently nonlinear, rather than the more frequently assumed form of one linear relationship giving way to another linear relationship with the onset of each new skill. Indeed, the variability across individual vocabulary growth curves can be captured most parsimoniously using nonlinear models (Bates & Carnevale, 1993; see also Elman et al., 1996, for a more general discussion of nonlinearity in development). Irrespective of the cause, recognizing the tremendous individual
variability is paramount when assessing atypical populations and in understanding in what ways these children differ from typically developing infants.

Early language and its relationship to nonlinguistic abilities

Meaningful language production and comprehension develop in tandem with a raft of nonlinguistic cognitive and motor abilities, in particular gestural communication and environmental sound recognition. The close developmental relationship between gesture and language (see Bates & Dick, 2002, for a review) appears to begin from around 6 months with a correlation between the onset of babbling and the onset of rhythmic hand banging. Toward the end of the first year, first word comprehension tends to co-occur with the start of deictic gestures (e.g., pointing and showing gestures) and gestural routines (e.g., waving goodbye; Bates, Benigni, Bretherton, Camaioni, & Volterra, 1979). In a similar vein, the later onset of productive vocabulary also co-occurs with—or is slightly preceded by—early recognitory gestures, e.g., putting a phone to the ear, or a brush to hair (Volterra, Bates, Benigni, Bretherton, & Camaioni, 1979).

Infants’ word comprehension also shares a similar developmental trajectory to their understanding of familiar environmental sounds (i.e., meaningful, nonlinguistic sounds such as a cow mooing or a car starting). Cummings, Saygin, Bates, and Dick (2007) found that 15 to 25-month-old infants’ accuracy in comprehending environmental sounds and spoken phrases was roughly equivalent (with a slight advantage for environmental sound recognition early in development). These results suggest that in fact speech does not appear to start out as being “privileged” as an acoustical transmitter of referential information.

Over development, the relationship between language, environmental sound comprehension, and gesture production changes as the infant gains more linguistic experience. Recognitory gesture is eclipsed by the exponential increase in a child’s productive vocabulary as language “wins custody” over gesture as the prime means of expressive communication (Bates & Dick, 2002). Similarly for environmental sound comprehension, infants with larger productive vocabularies show a significant accuracy advantage for comprehending spoken words over environmental sounds; this advantage for words over environmental sounds is not revealed when infants are grouped by chronological age. None the less, throughout the lifespan there remains a close relationship between language and gesture (Saygin, Dick, & Bates, 2005) and language and environmental sound (Borovsky et al., 2007; Cummings et al., 2006; Dick et al., in press; Saygin, Dick, Wilson, Dronkers, & Bates, 2003). In sum, language is not an isolated ability, “fenced off” from the rest of cognition. Instead, language appears to emerge from the interactions of many domain-general cognitive processes, including memory, attention, object recognition and categorization, social and emotional abilities, as well as the nonlinguistic motor and acoustical abilities just mentioned (Bates, Thal, Finlay, & Clancy, 2003). Language may be best described as “a new machine that Nature has constructed out of old parts” (Bates, 2004, p. 250).
Neural correlates of word learning

How is the child’s brain reorganizing itself during this period of profound language development? A series of electrophysiological studies by Debra Mills and colleagues suggests that “cerebral specialization for language emerges as a function of learning, and to some extent depends on the rate of learning” (Mills & Sheehan, 2007). Mills and colleagues have shown that the relative lateralization of electrophysiological (EEG) components (P100, N200–400) in the first years of life is intimately related to language learning and expertise. In particular, the lateral distribution of the N200–400 component for known versus unknown words is related to the overall size of the infant’s vocabulary in a particular language. Conboy and Mills (2006) showed that in 20-month-old bilingual toddlers classed as having high or low total vocabulary sizes, the N200–400 difference between known versus unknown words was lateralized only in children with higher vocabularies, and only in their dominant language. Conversely, this known versus unknown word N200–400 difference was bilaterally distributed in the nondominant language, and in the lower-total-vocabulary toddlers. (A similar finding was reported in a rapid-word-learning experiment by Mills, Plunkett, Prat, & Schafer, 2005.) Thus, changes in the large-scale topography of neural responses to words were driven by infants’ expertise with words in general, as well as by their knowledge of specific word exemplars.

More generally, in their review of the infant and child EEG language literature, Mills and Sheehan point out that the relative lateralization of EEG components changes dynamically over the lifespan. As an example, they cite the case of the P1 component evoked in response to auditory stimuli, which shows an early left lateralization from 3 months to 3 years, a symmetrical distribution from 6 to 12 years, and a right-lateralized distribution from 13 years into adulthood. The existence of such complex developmental trajectories demonstrates that any putative “early lateralization” for speech or language stimuli—such as reported in neonates and 3-month-olds (Dehaene-Lambertz, Dehaene, & Hertz-Pannier, 2002; Peña et al., 2003)—must be understood in the context of changes over the lifespan. (For an alternative view of earlier neural commitment to language processing, as well as a very useful summary of recent results, see Friederici, 2005).

The relationship between vocabulary and grammar

The sudden acceleration in vocabulary growth is accompanied or followed by the first two-word combinations at 18–20 months. Early word combinations mark the start of a second “burst” in the child’s abilities, this time in the realm of productive grammatical complexity. As with previous language milestones, this rapid increase in syntactic

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2 Note that in terms of comprehension, infants younger than 12 months can discriminate patterns analogous to simple grammars (Gomez & Gerken, 1999, Marcus, Vijayan, Bandi Rao, & Vishnun, 1999).
sophistication does not occur in a vacuum. Rather, toddlers' burgeoning syntactic abilities during the middle of the second year are closely yoked to their productive vocabularies—that is, syntax is not independent of the lexicon (Bates & Goodman, 1997).

Bates et al. (1988) found significant positive correlations between productive vocabulary size at 20-28 months and mean length of utterance (MLU) in the same period, with the strongest correlation between vocabulary at 20 months and MLU at 28 months when the "average" child's complex grammatical language is changing most rapidly. It is important to note that this tight synchronous and diachronous relationship between lexical size and grammatical complexity is not driven by a latent variable, like "maturational." In this vein, Bates and Goodman (1997) used data from the large CDI norming study (Penson et al., 1994) to demonstrate that total vocabulary size correlates with grammatical complexity equally as strongly as grammatical complexity correlates with itself—and that this relationship held true when chronological age was partialled out. In fact, calculated over the entire CDI sample, hierarchical stepwise regressions revealed that age uniquely accounted for only 0.8% of the variance for grammatical complexity, while vocabulary size accounted for 32.3% of unique grammatical variance. These results suggest a law-like relationship whereby total vocabulary size, irrespective of age, predicts grammatical complexity. Furthermore, there is very little variability between individuals around this relationship—including in all but one clinical population where this question has been investigated (see Figure 8.4 and below).

Remarkably, the law-like relationship between grammar and the lexicon also appears to hold over languages, despite the fact that languages differ tremendously in terms of the morphosyntactic cues that provide "clues" to meaning. For example, in English the most reliable grammatical cue to agency ("who is doing what to whom") is word order, whereas for some other languages (for example, Italian), these sentential roles are often imparted through inflectional morphology. Prima facie, such languages show somewhat different grammatical developmental trajectories: For instance, in a highly inflected, regular, and transparent language like Turkish, the use of morphological particles is observed much earlier in development than in English (Slobin, 1985). This does not entail, however, that the relation between the lexicon and syntax need be fundamentally different. Indeed, the CDI has been used cross-linguistically to compare English and a language with rich inflectional morphology for tense, aspect, number, and gender—namely Italian. Despite the obvious differences in the languages, the same law-like predictive link between vocabulary size and grammatical complexity exists for Italian and English (Caselli, Casadio, & Bates, 1999), demonstrating the generality of

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3 MLU is a frequently used measure of grammatical complexity calculated by taking the average number of morphemes (the smallest units of meaning) per phrase. MLU is a somewhat problematic measure for comparing children's grammatical knowledge across ages and across languages (Bates et al., 1988). The CDI includes measures of grammatical complexity which have been normed across ages and languages, against benchmark laboratory studies, and so provide a more robust mechanism for investigating the grammar explosion seen in the third year.

This finding (see Figure 8.4). The implication of this work is that grammar is not a completely separate process from word learning, nor does grammar simply require some word knowledge to get started. Instead, grammar and the lexicon are interwoven throughout early development.

The nature of children’s early grammar

One long-standing position in developmental psycholinguistics is that young children and adults fundamentally share the same syntactic “competence” (see Tomasello, 2000a, for a detailed critical review). This “continuity assumption” is one offshoot of the theory that all human languages are built upon a single innate universal grammar, with languages essentially differing only in the words they employ (Pinker, 1984, 1987, 1989). An alternative developmental hypothesis postulates that children’s early syntax

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4 Please note, however, that there are many “flavors” of this basic proposal; for example, Clahsen and Fels (2006) and Crain and Pietroski (2001) to name but two.
is item based. That is, young children initially produce grammatical language not through the utilization of general and abstract linguistic structures (e.g., subject, verb, noun), but rather through reproduction and very conservative and gradual tweaking of individual and specific linguistic "constructions" that they have learnt from others’ speech (Goldberg, 1995, 2006; Tomasello, 1992, 2000a, 2000b).

There is increasing evidence that at least some of young children’s grammar is item based. A number of observational studies of children’s early language production (Pizutto & Caselli, 1992, 1994; Tomasello, 1992) have revealed that children’s early production of verbs does not reveal a systematic pattern of usage. Instead, young children produce many verbs in only a single form with no transfer of structure from verb to verb. For instance, for the verb cut, a child will only produce phrases of the form cut—(e.g., cut apple or cut bread). This phenomenon, termed the “verb island” hypothesis (Tomasello, 1992), has been reported cross-linguistically (e.g., Pizutto & Caselli, 1992, 1994; Rubino & Pine, 1998).

Experimental studies have also investigated the item-based nature of early syntax by considering how well children produce novel verb constructions. In a series of studies, Tomasello and colleagues (see Tomasello, 2000a, 2000b for reviews) investigated what linguistic forms a 2- to 3-year-old child produces when he or she is given a novel verb to use in a variety of linguistic situations. For instance, if a novel verb like “tan” is only modeled for the child in the intransitive form, will a child produce the transitive form of the verb given an appropriate context? These studies repeatedly demonstrate that before the age of about 3 years children will generally base their verb productions on the input that they have heard. In other words, children will not transfer the transitive structure to a novel verb that they encountered in the intransitive form, even when explicitly asked to do so. Akhtar (1999) demonstrated an even more extreme example of how the structure of the input determines children’s linguistic productions. Exposing younger children (2- and 3-year-olds) to novel verbs in different word orders (i.e., subject, verb, object [SVO], SOV, and VSO) led to framing the novel verb in ways that reflected the exposed verb order—even for noncanonical English word orders like SOV and VSO. In contrast, older children (at 4 years) generalized from their knowledge of SVO word order and used the novel verb only in a “canonical” English way, thus suggesting that they were abstracting the verb away from its syntactic frame.

In general, these observational and experimental studies call into question the existence of abstract adult grammatical categories in children’s early syntactic development. These novel verb production studies also indicate that early language is highly sensitive to statistical patterns in the ambient language; for example, the frequency of a word order, such as SVO, is a key determinant of its production by a child. Gradually, as the typical child develops, by 3–4 years of age he or she will be increasingly able to generalize to novel verbs using existing templates such as the transitive SVO structure. However, it is important to note that these word-order preferences are not immutable: Even college-age adults are exquisitely sensitive to changes in the relative frequency of word orders (Dick, Butler, St. John, Gernsbacher, & Ferreira, 2007).
Language development in older children

Children by the age of 3–4 years are increasingly proficient language users with large productive vocabularies, and are able to fluently use and comprehend complex grammatical constructions (Bates & Goodman, 1997). This milestone, however, does not mark the end of the development of language. Instead, children’s language abilities keep gradually improving into adolescence and beyond (Nippold, 1998). One obvious area of improvement is vocabulary growth which continues throughout childhood, increasing by approximately 3,000 words per year (for a review, see Graves, 1986). Similarly, auditory and speech perception continues to improve into adolescence (Cepioniene, Rinne, & Naatanen, 2002). Most surprisingly, perhaps, syntactic abilities also continue to develop into later life.

As with infants, school-aged children remain sensitive to the frequency of syntactic constructions they hear. For instance, Huttenlocher and colleagues (Huttenlocher, Vasilyeva, Cymerman, & Levine, 2002) demonstrated that the proportion of complex syntactic constructions used by a child’s primary school teacher predicts how well the child produces and understands difficult syntactic structures, over and above chronological age. Even approaching adolescence, children’s syntactic comprehension can be demonstrated to vary from that of adults. Leech and colleagues (Leech, Aydelott, Symons, Carnevaie, & Dick, 2007) explored children’s (ages 5–17) and adults’ (ages 18–51) comprehension of morphosyntactically diverse sentences under varying degrees of attentional demands, auditory masking, and semantic interference. The results indicated that perceptual masking of the speech signal has an early and lasting impact on comprehension, particularly for more complex sentence structures, and that young children’s syntactic comprehension is particularly vulnerable to disturbance. This study not only demonstrated that syntax follows an elongated developmental trajectory, but that other more general attentional and perceptual skills continue to play an important role in syntactic processing across the lifespan (see also Hayiou-Thomas, Bishop, & Plunkett, 2004).

One of the advantages of working with older children is the possibility of investigating developmental changes in the neural underpinnings of language processing using fMRI. At least from the ages that are currently accessible with fMRI (about 5 years and up), the relatively small fMRI literature on language development does not show any simple maturational trends: For example, brain regions do not go “on-” and “offline” with increasing age. Rather, there are regionally specific increases and decreases in functional activation over development that tend to be nonlinear (Brown et al., 2005; Fair, Brown, Petersen, & Schlaggar, 2006; Schlaggar et al., 2002) and task-dependent (Saccuman et al., 2007a). For instance, in a comprehensive study of the development of overt verbal fluency from ages 7 to 32, Brown et al. (2005) showed “performance-independent” changes in 40 functionally defined brain regions, where 30 of 40 regions showed age-related decreases ("growing down"), and 10 showed age-related increases ("growing up"). The majority of the regions showing activation “growing down” were in bilateral occipital and temporal cortex, whereas the age-related increases were found
in left frontal and parietal regions. Importantly, both the slope and shape of these increases and decreases in activation differed over regions.

Age-related increases in left frontal activation in word production tasks have also been observed by Gaillard et al. (2003) for word production and Saccuman et al. (2007b) for both word production and sentence comprehension (see also Koelsch, Fritz, Schulze, Alsop, & Schlaug, 2005, for similar frontal trends in the development of music perception). Saccuman et al. (2007a) also found “growing-down” regions in the development of auditory sentence comprehension in several left superior temporal and inferior frontal regions. The one relatively consistent finding over most developmental fMRI language studies is that of relatively early overall left lateralization of activation for production (Gaillard et al., 2003; Holland et al., 2001; Saccuman et al., 2007a) and comprehension (Saccuman et al., 2007a), although again some studies fail to find this effect, at least for auditory speech comprehension (Ulualp, Biswal, Yetkin, & Kidder, 1998). In sum, even the relatively small developmental fMRI literature confirms and extends the notion that the development of language-processing neural networks is an extended, variable, and nonlinear affair that continues well into the later school and adolescent years.

Language Acquisition in Developmental Disorders

The milestones of the “average” child’s language development, as detailed in the previous section, constitute a necessary starting-point for considering how language learning and eventual language outcome is changed by atypical development. As we will show below, language learning can be disrupted by a wide variety of physical, social, and neural challenges to the developing child. However, each challenge tends to affect the trajectory of language acquisition at different points, and for different lengths of time. Historically, atypical development is often studied with a view to (a) the extent to which language learning in children with developmental disorders resembles the language of normally developing children; and (b) the changing patterns of language impairments between disorders. Such differences are usually described in terms of delay or deviation from the normal pathway, and permit us to establish relative strengths and weakness across subdomains of language, such as phonology, syntax, semantics, and pragmatics.

As noted above, the potential causes of delays or difficulties in language development are many, and diverse. For example, Down syndrome, Williams syndrome, and fragile-X syndrome are all defined in terms of genetic anomalies that are fairly well characterized at the molecular level. None the less, we must be particularly cautious in establishing mappings between genotype and phenotype; as we shall show, not only is there substantial phenotypic variability, but the behavioral phenotype changes over developmental time. In contrast to these genetic disorders, syndromes such as autism and specific language impairment are behaviorally defined, and are most likely the result of combinations of genetic and/or environmental factors (possibly
including viral infection and disruptions during prenatal development). All of these developmental disorders are characterized by frequent co-occurring problems with working memory, balance, coordination, as well as perceptual differences and learning difficulties. In addition, social and behavioral problems may also be manifest, although it is unclear whether these are exacerbated by or contribute to ongoing language problems.

In the following sections we review the impact of four different challenges to the development of language. First, we sketch the effects of a number of “extrinsic” factors on language development, such as chronic disease or long-term sensory or social deprivation. Second, we briefly review the consequences of overt brain damage, disease, or malformation for language development (but see Chapter 5 of this volume for a thorough review of development following early focal brain injury). Third, we more thoroughly explore the trajectory of language development in a quartet of congenital developmental disorders—autism, Down syndrome, Williams syndrome, and fragile-X syndrome—that serve to demonstrate some of the “boundary conditions” of language learning. Finally, we discuss developmental language disorder (also known as “specific language impairment”; Leonard, Eyer, Bedore, & Grela, 1997), a behaviorally defined disorder where a child’s language abilities are significantly lower than would be expected given his or her overall cognitive profile. We close the chapter by pointing the reader toward the latest guides for best practice in diagnosis and treatment of developmental language disorders.

Effects of extrinsic factors

As we have emphasized above, the child’s environment plays a crucial role in language development. This is true not only for the kinds of “constructive” environmental influences we have discussed (such as structured language input, joint attention, and so forth), but also for “destructive” environmental effects, such as disease, toxins, and social or sensory deprivation. We review some of the complex consequences of these environmental factors in the following section.

Teratogenic and pathological influences on language development

Neurotoxins such as pesticides may have effects on language development, although there is a paucity of data on this topic. Eskenazi et al. (2006) showed significant negative, dose-related relationships between DDT blood serum levels and Bayley MDI scales (a global measure of early mental development that includes language abilities). Lead exposure has been tied to poorer phonological, lexical, and sentence processing even in randomly sampled asymptomatic adolescent boys, but only when tested on the more complex variants of each task (Campbell, Needleman, Riess, & Tobin, 2000). Indeed, growth curve analyses (Coscia, Ris, Succop, & Dietrich, 2003) suggest that children with higher lead levels show generally lower vocabulary abilities at 15 years of age, and also a slowed rate of vocabulary growth. Interestingly, this trend is not echoed in the development of perceptual organization abilities.
Language Development

Fetal alcohol spectrum disorders (FASD) appear to have variable effects on language, which are modulated by concomitant factors that are difficult to parcel out in small samples (Cone-Wesson, 2005). None the less, as noted in a recent review by Riley and McGee (2005), children with prenatal exposure to alcohol are likely to show poorer profiles of language comprehension and production than their age-matched peers. This is perhaps unsurprising given these children's performance across a wide range of cognitive, emotional, and perceptual measures.

In contrast to what was originally feared, prenatal cocaine exposure does not appear to have severe deleterious effects on language development (Frank, Augustyn, Knight, Pell, & Zuckerman, 2001; Frank et al., 2005). Some studies have reported transient delays in expressive and/or receptive language development, but these effects are often difficult to tease apart from other comorbidities, which appear to have a significant and more lasting effect. For instance, Beeghly et al. (2006) found that, unlike prenatal cocaine exposure, exposure to violence or victimization predicted lower language scores, whereas preschool enrichment predicted greater language skill (see also below).

Chronic and/or severe infectious disease during early childhood may also cause delays in language acquisition, although there is relatively little research specifically on different aspects of language production and comprehension. None the less, several studies suggest effects of disease on language acquisition. For instance, Carter and colleagues (Carter, Murira, Ross, Mung’ala-Odera, & Newton, 2003) showed that Kenyan children who had undergone a severe bout of malaria early in life tended to have poorer comprehension and production scores years after their illness relative to matched control children. A further study by the same group (Carter et al., 2005) showed that cerebral malaria-induced epilepsy is particularly detrimental to language development. Abubakar and colleagues (2005) found that the productive vocabulary of Kenyan toddlers exposed to HIV in utero was almost half that of control children, when both were tested on an adaptation of the MacArthur–Bates CDI.

Social and sensory deprivation
As suggested by the results of Beeghly et al. (2006), chronic emotional neglect, physical mistreatment, or extreme stress during childhood may lead to significant, if possibly transient, delays in language development. In psycholinguistics, conclusions regarding the effect of social deprivation on language and cognition are often driven by a handful of case studies of extreme deprivation and/or abuse in early childhood (reviewed in Skuse, 1993). However, as summarized by Hough (2005), there is evidence from several fairly large group studies that severe neglect—as opposed to abuse alone—can cause serious delays in language comprehension and production (see also Allen & Oliver, 1982; Culp, Lawrence, Letts, Kelly, & Rice, 1991; Fox, Long, & Anglois, 1988).

The long-term effects on language development of early (orphanage) institutionalization are still in dispute, although a comprehensive survey by Frank, Klass, Earls, and Eisenberg (1996) suggests that deficits “in verbal skills and associated academic delays in reading seem to persist after early orphanage care into school age and adolescence,
with the most severe deficits found in children institutionalized at the youngest ages for the longest amounts of time” (p. 572). A yet unpublished doctoral thesis on Eastern European adoptees from deprived orphanages (Hough, 2005) is perhaps the most thorough investigation of this question. Using a wide range of language measures, Hough showed that 29 of 44 Eastern European orphans adopted into North American families would be classed as overall language-impaired, although comprehension tended to be more resilient than other language skills.

It is important to note that the effect of institutionalization on language is likely to be contingent upon the quality and type of interactions between children and staff. In a multi-institution study, Tizard, Cooperman, Joseph, and Tizard (1972) showed that orphaned children in high-quality nurseries exhibited no overall impairment in their language abilities (in contrast to previous studies). However, they did show that the type of language and social interaction between staff and children had a significant effect upon the children’s language comprehension skills, where the more interactive and informative the conversational style of the staff, the better the children’s language comprehension tended to be. Similar findings were reported in a large study of very low birthweight (VLBW) children (Landry, Smith, & Swank, 2002). This eight-year longitudinal study showed that not only were VLBW children on average about 5–8 months delayed in their language development (particularly those with medical complications at birth), but that both full-term and VLBW children’s language benefited from mothers who “built on their interests and topics of conversations” as opposed to simply directing those conversations (Landry et al., 2002, p. 199). These results presage those of Huttenlocher et al. (2002, discussed above), showing that the syntactic complexity of teachers’ speech predicts that of their students. Landry et al. (2002) also found a very striking effect on language of socioeconomic status, above and beyond what would be predicted by the children’s nonverbal abilities, thus suggesting that the conditions associated with extreme poverty may have a particularly pernicious effect on language development (see also Hart & Risley, 1995, for further discussion of this topic).

Language delays associated with early institutional privation or neglect may also be reversible, at least to some degree. For instance, Duyme, Dumaret, and Tomkiewicz (1999) showed that a group of low-IQ, neglected French children adopted later in childhood showed significant increases in verbal and performance IQ when retested as adolescents. Furthermore, the more “enriched” the adoptive home environment (as measured in terms of socioeconomic status), the greater the longitudinal increase in IQ. However, unlike matched early adoptees, who showed equivalent verbal and performance IQ outcomes, late adoptees’ verbal IQ tended to lag behind performance IQ. Extrapolating from these results, the authors suggest that a “supportive environment will be especially valuable for VIQ at the time that language develops most rapidly, between age 1.5 and 4 years” (Duyme et al., 1999, p. 8792).

Auditory deprivation, in the form of early, severe, and untreated sensorineural hearing loss, has significant repercussions for the trajectory of language learning, although the longer-term effects of hearing loss require clarification. In a landmark study, Yoshinaga-Itano, Sedey, Coulter, and Mehl (1998) demonstrated that infants
with severe or total hearing loss who were identified before 6 months of age showed significantly better language production and comprehension than those identified after 6 months of age. Indeed, the magnitude of the language advantage for the early intervention children (at least to 36 months) has prompted the implementation of quasi-universal hearing screening at birth in the United States.

Follow-up studies on these children and other samples should allow for a better understanding of long-term effects. Indeed, several studies of children with mild to moderate hearing loss (Briscoe, Bishop, & Norbury, 2001; Halliday & Bishop, 2006; Norbury, Bishop, & Briscoe, 2001) suggest that less severe hearing loss, while having an effect on some perceptual and metalinguistic skills, may not have a particularly significant effect on later language skills or literacy, at least for a majority of children with such hearing losses. In addition, the first studies of language outcomes in children with cochlear implants suggest that even very coarse auditory input may allow for good language development. In this regard, Geers (2004) found that a substantial minority of early implanted children (with implantation before 24 months) achieved language scores within the normal range. Spencer, Barker, and Tomblin (2003) found that language comprehension, reading comprehension, and writing accuracy scores for a sample of children with cochlear implants fell within one standard deviation of the mean for matched normal-hearing peers, although these children’s expressive scores were significantly lower than would be expected in a normal-hearing sample. (Similar deficits were found in grammatical comprehension for children with cochlear implants; Nikolopoulos, Dyar, Archbold, & O’Donoghue, 2004.)

It is important to differentiate between the potential effects of more severe and permanent sensorineural hearing loss from those associated with transient periods of hearing difficulty, such as those caused by persistent otitis media with effusion (also known as glue ear). The standard clinical teaching is that children who have prolonged or numerous bouts of otitis media are at significantly elevated risk for language impairments or delay. Despite this, there is strong evidence that there are minimal to no lasting effects on affected children’s language or intellectual development, both in prospective studies—addressing causality—and when studies control for covariates such as socioeconomic status and schooling (Paradise et al., 2001, 2005). This does not necessarily mean that there are no possibly transient linguistic consequences of otitis media; for example, Rvachew, Slawinski, Williams, and Green (1999) report that children with persistent otitis media babble less than controls. However, the final language outcome for these children is very close or identical to that of unaffected children.

Effects of hemispherectomy and traumatic brain injury

A second variety of “extrinsic” factor that can radically change the course of language and cognitive development is acquired neurological damage as a result of traumatic brain injury or invasive surgery. In preface, we should note that there is a high degree

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5 For instance, see www.nhsdirect.nhs.uk/articles/article.aspx?articleId=178&sectionId=10822.
of variability in the etiology, comorbidities, and clinical sequelae of hemispherectomy and traumatic closed-head brain injury. Thus, it is very difficult to draw firm conclusions about brain and language development from these cases. None the less, hemispherectomy, in particular, is often cited in theoretical debates on language development as it relates to neural development and plasticity. As such, it is important to present what is currently known about language outcomes following closed-head brain damage or surgical removal of part of the brain.

Hemispherectomy
In the past decade, there has been considerable attention surrounding celebrated cases of extraordinary late language acquisition following removal of an entire cerebral hemisphere, such as the case of “Alex” who acquired fluent complex language following surgery at the age of 8 (Vargha-Khadem & Mishkin, 1997). Conversely, there are also many case studies of poor outcome following the procedure. Hemispherectomy is normally carried out in order to control untreatable epilepsy, meaning that hemispherectomy patients have usually received years of anti-seizure drug treatment, which may have dramatically altered the course of their development (Vargha-Khadem & Polkey, 1992). More importantly, the underlying etiology of the epilepsy is highly variable, including early onset epilepsies resulting from large-scale dysmorphologies in the laminar structure of the cortex, such as cortical dysplasias and hemimegalencephaly, congenital vascular disorders, such as Sturge–Weber syndrome, and disorders with much later onsets, for example, Rasmussen encephalitis. These etiological factors, combined with the small number and highly variable nature of the documented cases, confound a straightforward summary of the effects of hemispherectomy on language development (although, for recent medical reviews of cognitive and linguistic outcomes, see Jonas et al., 2004; Kossof, Buck, & Freeman, 2002; Pulsifer et al., 2004).

Language outcome following hemispherectomy is dependent on the extent to which the surviving hemisphere is unaffected, and thus varies massively with underlying etiology. Consequently, etiology appears to be the strongest predictor of postoperative language outcome: Children with cortical dysplasias and hemimegalencephaly perform far worse than those with Rasmussen encephalitis, while children with Sturge–Weber syndrome achieve the best linguistic outcome (with a sizable proportion of children in mainstream education; Kossof et al., 2002).

Etiology interacts substantially with the side of resection. Left hemispherectomy is only predictive of worse language outcome (as would be predicted by the adult language model) in the Rasmussen encephalitis children, who only present with seizures later in development, around 4 years, and so have followed a normal developmental trajectory till this point (Curtiss, de Bode, & Mathern, 2001; Pulsifer et al., 2004). Similarly, it is very hard to pull apart the differential effects of etiology, age of onset, and age of surgery upon language outcomes, since etiology strongly predicts symptom onset. Thus, given the current data, it is unwise to draw general conclusions about the positive or negative effects of early onset/surgery for language development.
Traumatic brain injury
Childhood closed-head traumatic brain injury (TBI)—as caused by traffic accidents, falls, sports injuries, or blows to the head—typically results in both multifocal injury and diffuse damage in the form of axonal shearing and long-term changes in neurotransmitter function and balance (Ewing-Cobbs & Barnes, 2002). In general, most children with TBI do not show frank aphasic symptoms. However, there do tend to be lasting effects on lexical and sentential production and comprehension, particularly when the injury is suffered earlier in life. Much recent research has focused on the more overt disruption of discourse level processes in TBI children, such as difficulties in drawing inferences, understanding metaphors, and establishing links between different elements of a story. As summarized by Dennis and colleagues (Dennis, Purvis, Barnes, Wilkinson, & Winner, 2001), children with TBI have particular problems with “nonliteral” language use, including figurative expressions, humor, implicature, and idioms. Problems in “higher-level” language use, such as meta-cognition and inference, are particularly notable in children with severe injuries, particularly afflicting the frontal lobes (Dennis, Barnes, Wilkinson, & Humphreys, 1998). Our understanding of the neural sequelae of TBI and its effect on neural and cognitive development at various points during childhood should increase markedly in the coming years with the advent of richer and more precise neuroimaging methods (reviewed in Hurley, McGowan, Arfanakis, & Taber, 2004).

Congenital Language Disorders
In contrast to the case of acquired frank brain injury, the changes to brain structure and function that result from congenital developmental disorders tend to begin very early in development, and be more pervasive and yet more enigmatic. Language development in the face of neurodevelopmental disorder has been a topic of particular interest for both clinicians and psycholinguists, and the relative richness of the data on linguistic and nonlinguistic processing in congenital language disorders reflects this. Of particular import is the question: To what extent are cognitive abilities a limiting factor in the process of language development? Children with learning difficulties typically show a later onset of language learning, and the overall level of language ability that they achieve may be limited. Differential profiles of language ability across disorders may be attributed to (a) the social elements of communicative ability (e.g., joint attention); (b) the computational mechanisms that underpin language (e.g., audiomotor attention); and (c) the use of language in interacting with—and manipulating—other people and the environment (e.g., social pragmatics). These strengths and weaknesses can often be most easily identified in the early stages of development, although their impact upon language development may only be evident later “downstream” in development.
Early language learning

As in our description of typically developing children's language abilities, we will take a roughly chronological approach to developmental language disorders, highlighting the diverging developmental paths in Williams syndrome (WS), Down syndrome (DS), fragile-X syndrome (FXS), and autistic spectrum disorders (ASD), first in infancy and toddlerhood, and then into the school years and adolescence. An "early/late" comparison is particularly informative when exploring these disorders, in that the disorder-specific profile of relative strengths and weaknesses is not constant over development.

Problems with social cognition

In the section on typically developing children, we saw that early language is heavily dependent on social cognition to get off the ground, in particular the ability to share attention. Indeed, even in the first months of life, these important social precursors to language already distinguish between infants with developmental disorders and typically developing infants (TD). For example, TD infants show a preference for their mother's speech (Morse, 1972), whereas infants with ASD show no such preference, and seem to have very little interest in people and social interaction (Klin, 1991; Kuhl, Coffey-Corina, Padden, & Dawson, 2005). These infants, like those with FXS, show poor levels of eye contact and gaze following, and may even attempt to avoid the gaze of a caregiver (Cohen et al., 1988). In contrast, infants with WS are typified by their strong desire for social interaction and keen interest in faces. However, this preference also leads to poor gaze-following behavior through the tendency of infants with WS to prefer to look at the face of the caregiver rather than follow the direction of the gaze (Bellugi, Bihlre, Neville, Jernigan, & Doherty, 1992).

Infants with DS also enjoy interacting socially, though there is a delay in mutual eye contact, which may have an impact upon subsequent dyadic interactions between infant and caregiver (Berger & Cunningham, 1981; Jansow et al., 1988). Moreover, once these initial problems resolve, infants with DS seem to prefer to focus on the eyes, as opposed to visually exploring the facial features of the caregiver as TD infants do (Berger & Cunningham, 1981). Deviations in early interactive behavior can also be seen in more complicated triadic interactions between infant, caregiver, and object. For example, infants with WS struggle to switch their attention from the caregiver to an object. This problem with triadic interactions is thought to hinder acquisition of the conceptual knowledge of objects, contributing to a subsequent delay in vocabulary development (Laing et al., 2002; Mervis et al., 2003).

Precursors of expressive language in developmental disorders

Just as for social cognition, children with developmental disorders and TD infants diverge in terms of their prelinguistic productive abilities (e.g., babbling, gesturing). Although all the developmental disorders considered here are delayed in some way
relative to TD infants, the exact patterns of these delays, and the mechanisms underlying them, may vary from population to population. For instance, for infants with DS, there is usually a delay in the onset of babbling by an average of two months. This delay in babbling and in subsequent phonological development may be due to the articulatory difficulties that frequently occur in conjunction with the disorder. They may also be due in part to the frequent problems DS children have with hearing loss due to chronic and severe otitis media (but see Shott, Joseph, & Heithaus, 2001, for work suggesting that early and aggressive medical treatment may stem almost all hearing loss due to infection).

In contrast to their profile with babbling, young children with DS produce far more communicative gestures than those seen in TD, a behavior that may serve as a compensatory mechanism for delayed and error-prone phonological production (Singer Harris, Bellugi, Bates, Jones, & Rossen, 1997). In contrast, young children with ASD and WS gesture very little. In stark contrast to typical development, use of referential pointing in WS does not appear until after the naming explosion (Laing et al., 2002; Mervis & Bertrand, 1997).

Despite the differences between DS and WS, children with WS show a similar delay in the onset of babbling (Masataka, 2001). Furthermore, children with WS show early problems with speech segmentation (Nazzi, Paterson, & Karmiloff-Smith, 2003). For instance, toddlers with WS detect words with strong–weak stress patterns, but fail to detect weak–strong words, despite the fact that the average typically developing child accomplishes this feat at 10.5 months (Nazzi et al., 2003). These findings are somewhat surprising given the relative strengths of the language and phonological abilities of older children with WS, and highlight the fact that the pattern of deficits and relative strengths in the adult does not mean that these same deficits and strengths are present earlier on in development.

Word comprehension and production and early grammatical development

Just as prelinguistic expressive skills in developmental disorders are delayed, so is the onset of first word comprehension and production (Bates & Goodman, 1997; Singer Harris et al., 1997). Both WS and DS children are severely delayed in word production, reaching a vocabulary size of about 50 words about two years later than a TD child (who reaches this point at around 16 months). After reaching this critical vocabulary size, the language profiles of DS and WS children diverge. On the MacArthur-Bates CDI, WS follows the established law-like relationship between vocabulary size and grammatical complexity demonstrated by typically developing children, albeit one that is considerably delayed. In contrast, children with DS show an atypical relationship, with their grammatical complexity poor relative to their vocabulary (Rice, Warren, & Betz, 2005). Therefore, for WS—unlike DS—there is no obvious dissociation between

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6 Phonological difficulties are also shown by children with FXS (Dykins, Hodapp, & Leckman, 1994).

lexical and grammatical ability at this point in development (see Figure 8.5). Indeed, children with WS are no better at a range of linguistic tasks than their mental age would predict (Vicari, Caselli, Gagliardi, Tonucci, & Volterra, 2002).

Furthermore, the types of construction produced by children with a developmental disorder also vary between populations. In DS, there is a varying rate of change in MLU, with a protracted two-word stage. Some DS children do not extend beyond this stage until 4 years of age, or as late as 5 or 6 years (Fowler, 1988). Moving beyond the two-word stage, the use of grammatical knowledge develops rapidly until an MLU of approximately 3.5 is reached. Subsequent grammatical development can be limited, particularly for those children with a low IQ (below 50) who may never develop complex language (Miller, 1988; Rice et al., 2005). Similar variability in the rate of change of MLU has also been seen for children with ASD. However, in contrast to DS, children with ASD may develop more complex language skills (Bellugi et al., 1992; Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000).

By combining MLU with a more detailed measure of syntactic development—such as the index of productive syntax (Scarborough, 1990)—it is possible to compare syntactic abilities across populations in more detail. Importantly, one can determine
whether children with disorders develop their knowledge of grammatical constructs in the same sequence as that observed in TD children. For children with DS and FXS, syntactic development is slow and does not generally result in these children consistently producing more complex forms. For at least some children with ASD, explanations for restricted growth in grammar point to the limited range of grammatical constructs used by these children, and their tendency to ask fewer questions. Overall, in spite of the differences in the rate of grammatical development, it has been suggested that children with disorders develop this knowledge in approximately the same sequence as TD children (Tager-Flusberg & Sullivan, 1998; see also Tager-Flusberg, Lord, & Paul, 2005).

Perhaps not surprisingly, there are profound differences in the ways in which children with these developmental disorders use language to communicate and interact within a social context. For example, both children with DS and WS generally enjoy the opportunity to interact within a social environment, and their use of language is motivated through social exchanges in conversation. By contrast, language in many children with ASD, although quite fluent and social, appears to lack a sense of reciprocity in conversational norms. Such a language profile is often linked to a significant discrepancy between verbal and nonverbal IQ (Tager-Flusberg & Joseph, 2003; this article is also a valuable overview of language abilities observed in several phenotypes of ASD). Children with FXS also show difficulty in communicating through conversation, exhibiting dysfluent, rapid speech that is described as cluttered (Dykins et al., 1994). Overall, communicative styles differ considerably between disorders.

Later language learning

We have seen how early language learning in atypically developing populations varies from that of TD children; however, the same patterns of strengths and weaknesses are not constant throughout the child’s life. Instead, the relative strengths and weaknesses of early and late language abilities can be quite different as the multiple interacting factors underlying language acquisition unfold. For children with developmental disorders, the overall language ability attained in later childhood and into adolescence is predicted by a number of interacting factors. These include:

1. Mental age (traditionally assessed by standardized testing).
2. Severity of language impairment during the early stages of learning.
4. Frequency of communicative acts.
5. Ability to understand the thoughts and intentions of others.7

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7 It is important to note that there is not necessarily a unidirectional relationship between language development and theory-of-mind ability in either typically developing or autistic children. See Hale and Tager-Flusberg (2003) for a study showing that training on sentential complements improves TD children’s performance on a false belief task.
With these factors in mind, it is worth revisiting the linguistic outcomes of the distinct developmental disorders in later life.

**Williams syndrome**
The main feature of language development in WS is delay (Brock, in press; Thomas et al., 2001, in press). In spite of this, relative to mental age-matched controls, these children may develop good semantic knowledge, an extensive vocabulary, and develop complex syntax, though their vocabulary skills generally exceed that of their syntactic ability in terms of mental age (Karmiloff-Smith et al., 1997). In this regard, Reilly, Losh, Bellugi, and Wulfeck (2004) showed that when narrating a picture-book story, children with WS show a very similar profile of slowed morphosyntactic development to children with specific language impairment (LI), with particular problems in inflectional morphology (but cf. Rice et al., 2005, for differing results). Interestingly, while Reilly et al. (2004) found that, by ages 10–12, children with WS (unlike LI) produced the same proportion of complex syntactic structures as typically developing controls, the children with WS failed to establish ties between episodic and thematic narrative events, whereas children with LI were very similar to typically developing children in this regard. Finally, as might be expected given their hypersocial nature, children with WS far outstripped even typically developing children in their use of socially engaging cues when telling a story.

Thomas and Karmiloff-Smith (2003) characterized two types of hypothesis regarding the developmental profile of WS: (a) a series of imbalance hypotheses, which accounts for the profile shown in WS as being due to some form of imbalance between phonological and semantic processing, and (b) a conservative hypothesis, which proposes that language development in WS is delayed but not fundamentally altered. Thus, the language profile of children with WS might be accounted for by the nonlinguistic characteristics of the disorder—such as a strong desire for social interaction and poor visuospatial skills (Brock, in press).\(^8\)

**Down syndrome**
In general, children with DS show the same sequence of developmental “events” as do TD children, but are generally very delayed. For example, children with DS usually produce the same pattern of phonological errors produced by younger TD children (Stoel-Gammon, 1980). However, when we compare children with WS and those with DS across early and later childhood, we see a divergence of DS and WS developmental trajectories in different language domains. For instance, in early language development, both children with WS and those with DS have a similar level of vocabulary, but by adolescence the vocabulary of children with WS exceeds that of those with DS (Paterson, Brown, Gsödl, Johnson, & Karmiloff-Smith, 1999). Thus, the performance of children with DS appears to asymptote at a lower level.

\(^8\) As with all atypically developing populations, it is important to keep in mind that there may be variability in terms of the relative strengths and weaknesses found in WS linguistic and cognitive skills (Porter & Coltheart, 2005).
Comparisons between children with DS and TD children suggest that the difficulty in DS may be associated with the encoding of verbal information, with children with DS showing better visuospatial than verbal encoding, a pattern opposite to that shown by TD children (Marcell & Weeks, 1988; Varnhagan, Das, & Varnhagan, 1987). Pragmatic abilities indicate that children with DS do have an awareness of the thoughts and intentions of others, though the strength of these pragmatic abilities is believed to be closely tied to mental age and IQ (Fowler, 1998).

**Autistic spectrum disorders and fragile-X syndrome**

In contrast to DS, children with ASD exhibit **severe pragmatic difficulties** in comparison to mental age-matched controls (Tager-Flusberg, 1981). Though children with ASD will use language to manipulate their environment, their use of language to engage in conversation or express thoughts and emotional states is limited. The pragmatic problems exhibited in ASD are thought to reflect a more general social-cognitive deficit related to theory of mind (Baron-Cohen, Tager-Flusberg, & Cohen, 1993), and it has been suggested that autistic children may actually use their language knowledge to bootstrap their understanding of mental states (Frith & Happé, 1994).

In terms of pragmatic ability, children with ASD struggle to understand figurative speech, such as “it’s raining cats and dogs,” taking literal meanings from such sentences (Singer Harris et al., 1997; Tager-Flusberg et al., 2005). Other characteristics of autistic speech include a flat pattern of intonation, and **echolalia**, where children repeat the verbal utterances of others (Prizant, 1983; Roberts, 1989). These may occur when the child is required to respond to a question or in the form of preconstructed speech during conversation. Children with FXS also exhibit echolalia, which is accompanied by further unusual pragmatic characteristics, such as perseveration, increased use of jargon, and poor conversational ability.

In sum, there are clear differences between disorders in terms of the level of language ability that may be achieved (and in terms of the mechanisms underlying these language problems) and the proficiency with which this language is used within a social context. From the example of WS, it is clear that complex language may be achieved in spite of cognitive limitations; however, this ability may be acquired through atypical means. Overall, language ability in these children may be restricted not only by general cognitive limitations, but also by characteristics of the disorder, which may or may not be language specific, but impede the ability of the child to develop effective communication skills. In the following section, we look at the trajectory of language development in a developmental disorder where language difficulties seem to occur in the absence of frank cognitive impairments, namely specific language impairment.

**Specific Language Impairment**

Specific language impairment (LI) is defined behaviorally as delay or abnormality in expressive and/or receptive language skills in the absence of overt neurological impairment,
mental retardation, hearing loss, ASD, or severe social or emotional problems. Studies attempting to define the nature and specificity of the language deficit affecting these children have revealed problems in many areas, including phonological skills, lexical-semantic development, morphosyntax, and pragmatics (Johnston & Kamhi, 1984; Lahey & Edwards, 1999; Leonard, 1998; Leonard, Schwartz, Allen, Swanson, & Loeb, 1989; Schwartz & Leonard, 1985). Children with LI tend to produce their first words significantly later than normally developing children, and are slower and less accurate in retrieving lexical items (Lahey & Edwards, 1999; McGregor, Newman, Reilly, & Capone, 2002; Trauner, Wulfeck, Tallal, & Hesselink, 1995).

Grammatical morphology is an area of particular weakness. Children with LI have difficulty marking verb inflections, using the auxiliary system, and detecting grammatical violations (Bishop, 1994; Leonard et al., 1997; Marchman, Wulfeck, & Weismer. 1999; Marton, Abramoff, & Rosenzweig, 2005; Wulfeck, Bates, Krupa-Kwiatkowski, & Saltzman, 2004). The ability to use language effectively and flexibly in social contexts, and to use and understand figurative language may also be impaired (Marton et al., 2005; Norbury, 2004; Vance & Wells, 1994). These language difficulties hinder the development of reading and writing skills (Bettinger, Faragher, Simkin, Knox, & Conti-Ramsden, 2001; Stothard, Snowling, Bishop, Chipchase, & Kaplan, 1998), and the calculation skills of these children may also be affected (Cowan, Donlan, Newton, & Lloyd, 2005).

LI is also associated with accompanying nonlinguistic deficits, which have brought into question the label of specific in the strong sense (see Leonard, 1998, for an extensive review). These deficits include working memory problems, impairments in motor skill and speed (Schul, Stiles, Wulfeck, & Townsend, 2004), particularly those involving sequencing, timing, and balance (Hill, 2001), as well as poor auditory frequency discrimination, and sequential auditory processing (Hill, Hogben, & Bishop, 2005; McArthur & Bishop, 2005). Phonological memory problems, as assessed by nonword and sentence repetition, seem to be a particular point of difficulty for LI children (Briscoe, et al., 2001; Conti-Ramsden, Botting, & Faragher, 2001; Dooloughan & Campbell, 1998; Gathercole & Baddeley, 1990). However, the causal role of non-linguistic skills, especially auditory ones, in the emergence of language impairments is hotly debated, and has generated much interesting research (Benasich & Tallal, 2002; Bishop, Adams, Nation, & Rosen, 2005; Bishop, Adams, & Norbury, 2004; Bishop & McArthur, 2004, 2005; Choudhury, Leppanen, Leevers, & Benasich, in press; Halliday & Bishop, 2005, 2006; McArthur & Bishop, 2004a, 2004b; Mengler, Hogben, Michie, & Bishop, 2005; van der Lely, Rosen, & Adlard, 2004; Viding et al., 2004).

The profile of relative strengths and weaknesses in both linguistic and nonlinguistic skills in children with LI is highly variable, characterizing LI as a heterogeneous disorder. The prognosis for young children with LI is that they will develop functionally complex language, but with pervasive underlying difficulties in grammar and nonword repetition (Newbury, Bishop, & Monaco, 2005).

Although LI is a behaviorally defined disorder, there does appear to be a genetic component associated with language difficulties, indicated by twin studies and genomic
screening (reviewed in Newbury et al., 2005; SLI Consortium, 2002, 2004). To date, loci on chromosomes 3 and 19, as well as a gene on chromosome 16, have all been implicated. Specifically, anomalies on chromosomes 3 and 16 are suspected to influence systems related to phonological short-term memory and articulation\(^9\) (SLI Consortium, 2002, 2004), although the mechanisms by which this occurs remain to be clarified. These data indicate that multiple genetic factors may contribute to language difficulties.

In addition, a new, large-scale (556 twin pairs) study of the genetic and environmental influences on language skills showed that the same genetic and environmental factors underlie individual differences on a wide variety of language skills, such as receptive and expressive syntax, vocabulary and lexical semantics, and verbal memory (Hayiou-Thomas et al., 2006). These results suggest that the development of these disparate language abilities may rely upon the same underlying cognitive and perceptual mechanisms. However, in another large twin study, Bishop, Adams, and Norbury (2006) showed that the nonlinguistic task most associated with language impairments—nonword repetition—did not show much phenotypic or etiological overlap with verb-tense impairments, the other “gold standard” hallmark of LI (Rice et al., 2005).

A tantalizing link between quite severe language impairments and genetics was discovered in the KE family (Lai, Fisher, Hurst, Vargha-Khadem, & Monaco, 2001). Here, over half of the family members were affected by an expressive and receptive language and articulation disorder, one that is linked to mutations and deletions of the FOXP2 gene. Subsequent work in magnetic resonance imaging identified structural brain abnormalities in the preSMA/cingulate cortex and Broca’s area, as well as both structural and functional abnormalities in the caudate nucleus, associated with FOXP2 gene mutations (Vargha-Khadem et al., 1998). However, it is extremely important to point out that subsequent studies have shown no involvement of FOXP2 in large-scale samples of language-impaired children, ruling this gene out as a unitary causal factor in language impairments more broadly.

Until recently, only a handful of studies (Gauger, Lombardino, & Leonard, 1997; Jernigan, Hesselink, Sowell, & Tallal, 1991; Trauner, Wulfeck, Tallal, & Hesselink, 2000) had attempted to delineate the neurological, neuroanatomical, or neurophysiological status of LI children revealing significant abnormalities. For example, anomalous asymmetries in prefrontal regions have been identified (Jernigan et al., 1991). Plante and colleagues (Plante, Boliek, Binkiewicz, & Erly, 1996; Plante, Swisher, Vance, & Rapcsak, 1991) found similar anomalies. Trauner and colleagues conducted extensive neurological examinations and observed a higher than normal occurrence of neurological “soft signs” in children with LI compared to control children, and a number of structural (MRI) abnormalities were identified (Trauner et al., 2000).

Two studies using quantitative analyses of structural MRI data have shed more light on the anatomical factors that might underlie language disorders. Herbert et al. (2004)

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\(^9\) It is worth pointing out that LI is not necessarily comorbid with articulatory problems (see Shriberg, Tomblin, & McSweeney, 1999).
reported white-matter abnormalities in a group of 5- to 11-year-old children with a developmental language disorder. In particular, overall white-matter volume was found to be enlarged in impaired children compared to controls. The enlargement was observed in the radiate white-matter compartments, especially in later myelinating frontal and prefrontal regions, suggesting a process modulated by time rather than a disruption linked to specific circuits or processes. Abnormalities were also reported in gray matter. De Fosse et al. (2004) found patterns of reverse rightward asymmetry of cortical volume in the inferior frontal gyrus (pars triangularis and pars opercularis) in boys with LI. The asymmetry was correlated with verbal IQ scores, and not with non-verbal IQ. In sum, while these early studies have advanced our understanding of some of the global differences between TD children and children with LI, a great deal of work remains to be done in elucidating the neural bases of developmental language impairments.

**Treatment of Language Disorders**

Research on developmental language disorders is extremely useful for understanding how the brain develops to process language, but is primarily carried out with the intent to inform and develop new and better treatments. The literature on the treatment of language disorders is a vast field, and we will not attempt to survey it here. Rather, we point the reader to recent, government commissioned reviews on "best practice" principles and findings in the United States and Great Britain. In the US, Nelson, Nygren, Walker, and Panoscha (2006) conducted a systematic evidence review for the US Preventive Services Task Force on the efficacy of screening for speech and language delay in preschool children. This is an analysis of results from 745 articles published between 1966 and 2004 that reported results from either screening or intervention programs. A slightly earlier review by Law, Garrett, and Nye (2004), prepared for the UK-based Cochrane Collaboration, specifically analyzes the outcomes of speech and language interventions for children with language delays or disorders (see also the commentary on this meta-analysis by Johnston, 2005). Both surveys generally conclude that interventions and screening can be effective and valuable, but that much work remains to be done. The commentary by Johnston serves as a salutary reminder that it is nontrivial even to define what constitutes useful evidence for evaluating intervention and therapy programs.

**Conclusion**

Research into how the human child acquires language has come a long way over the past few decades, although this review demonstrates how much is still to be understood. We still know too little about the exact profiles and etiologies of developmental disorders, and the study of the neural bases of language development is only now
beginning in earnest. Language development is inherently a process of change. Simple explanations in terms of static dissociations between developmental disorders are inevitably somewhat inadequate. Exploring the multiple and varied trajectories of language in both typically and atypically developing populations adds not only to our understanding of developmental disorders, but also provides us with insights into the development of more general cognitive processes.

Studies of language development have been particularly useful in helping us to understand the emergence of modularity and expertise, and the scale and flexibility of cognitive processes during learning. In exploring the mechanisms of language development, we cannot underestimate the multiple levels of interaction between the individual and his or her environment. Importantly, the environment should not be viewed as a static influence, but a force that itself may change in response to the child. In short, exploring the parameters affecting the trajectory of development will prove informative in terms of understanding not only developmental disorders but also the process of normal development itself.

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References


The Neuropsychology of Visuospatial and Visuomotor Development

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“Visual spatial development” has many definitions and levels of analysis with different perspectives from neuropsychology, developmental cognitive neuroscience, pediatric neurology and ophthalmology, education, and rehabilitation. It covers many areas, including brain processing related to spatial representations, an understanding of relative motion, of objects in space, the processes underpinning acts of mental rotation and location memory, reaching, grasping, and spatial attention. The neurobiological approach taken in this chapter will attempt to link findings from normal adults and patients with neurological damage, animal neurophysiology, and psychological studies on spatial development in infants and children, so as to identify relationships between changing behavior and brain function and to devise models of these processes.

In some areas, attempts have been made to separate “perception” from “cognition” in spatial development. For example, some developmental psychologists have defined cognitive acts as those in which infants demonstrate “intentionality,” while “perception” is more automatic and less goal oriented. Some argue for limiting “visual perception” to lower centers in the eye and occipital lobes, while “cognition” is mediated by “higher centers”—parietal, temporal, and frontal cortices. Such divisions are somewhat artificial in that many visual spatial actions involve complex circuitry running from subcortical centers, such as the superior colliculus and basal ganglia, to occipitoparietal areas for spatial representations, including cortical and subcortical motor areas, specialist temporal lobe areas for object recognition, top-down attentional control from frontal areas, and feedback loops between all of these networks. In fact, it has been estimated that over half of the brain is involved in even simple spatial tasks such as pouring water from a jug into a glass. However, in many developmental studies there is an attempt to delineate and separate processes within a network by designing stimulus conditions with dimensions known to elicit responses in specific populations of tuned neurons within particular brain areas—these we might call “designer stimuli.” Other studies have adapted tests for children and infants from adult visual spatial tasks, where failures have been related to specific locations of brain damage in the patient.
Of course, in human development, immaturity of processing in any one or more of these networks may limit behavioral performance on any specific visuospatial task. If analogies are found between infant and adult patient behavior, this does not necessarily mean that the cascading processes of human development follow the same course as the adult with specific brain damage. It can only be used as a first approximate step.

This chapter is divided into eight sections which consider:

1. Current neurobiological models of normal infant development of spatial vision.
2. Development of spatial selective attention for action in infancy.
3. Development of dorsal and ventral streams.
5. Development of action modules for locomotion and navigation.
7. Summary of abnormal spatial development.
8. Summary of the developmental model of visual spatial development.

Examples of abnormality of development and their neurobiological underpinnings are briefly reviewed in each section, and are summarized in section 7.

1. Current Neurobiological Models of Normal Infant Development of Spatial Vision

For further details on much of this section see Atkinson (2000).

1.1. Two visual systems: subcortical and cortical

Early models of visual spatial development started from the idea of two visual systems, a phylogenetically older, retinotectal system and a newer, geniculostriate system. The tectal system defines where an object is located to trigger orienting, while cortical mechanisms define what is actually in the selected location (Schneider, 1969; Sprague & Meikle, 1965). Bronson’s (1974) model for human visual development was based on this dual system. Since newborns orient by head and eye movements to conspicuous stimuli, but show little evidence of pattern discrimination, he proposed that vision is subcortically controlled for deciding where a stimulus was located, with the cortex maturing postnatally for deciding what was in the fixated location.

Extensive studies with typically developing infants have allowed more detailed models to be devised for these two systems. These studies have included behavioral measures (e.g., preferential looking) and electrophysiological measures of brain waves (EEGs, visual-evoked potentials or event-related potentials [VEP/ERP]). In Atkinson and Braddick’s initial model (Atkinson, 1984; Braddick, Atkinson, & Wattam-Bell, 1989) distinct functional modules or channels for the different visual attributes for
color, pattern, and motion were proposed, which had their underpinnings in populations of cortical neurons with distinct profiles of response to different visual stimuli.

The visual cortex undergoes very rapid development in neuronal selectivity in the first six months after birth, the beginning of a period during which the number of synapses in visual cortex increases dramatically (Huttenlocher, de Courten, Garey, & van der Loos, 1982), providing the rich connectivity on which the selective cortical processing proposed in the model depends. Atkinson and Braddock’s VEP and behavioral studies with normal infants suggested that the various attributes of selectivity do not emerge together in this wiring-up of the cortex; rather, there is a clear developmental ordering. Mechanisms or channels for orientation for shape discrimination become functional closely after birth, followed by direction selectivity for visual motion, and then selectivity to binocular disparities for stereoscopic vision.

1.2. Magnocellular and parvocellular systems

Sensitivity to motion direction (directionality) and stereo depth information (binocularity) is associated with input to the cortex from the magnocellular pathway, and processing within the cortex by the dorsal stream (Livingstone & Hubel, 1988). The initial development of this pathway may be slower than that of the parvocellular ventral pathway which specializes in processing form (orientation or slant) and color (Atkinson, 1992). Motion and disparity processing both require the comparison of information between separated locations in the visual field and hence the establishment of relatively long, orderly, horizontal connections in cortex. The fact that, after the initial onset, there is a development of infants’ directional sensitivity to progressively greater displacements (Wattam-Bell, 1992, 1996), and of binocular sensitivity to progressively greater disparities (Wattam-Bell, 1995), suggests that the range of these connections, increasing with age, may be a limiting factor in the development of these aspects of cortical selectivity.

1.3. Dorsal and ventral streams

Pioneering electrophysiology starting in the 1970s and 1980s mapped distinct brain areas beyond primary visual cortex containing neurons responding to particular visual attributes, including an area selective for motion information (V5/MT) and a color-specific area (V4; for reviews, see Felleman & Van Essen, 1991; Zeki, 1993). Ungerleider and Mishkin (1982) proposed two broad, functionally distinct, processing streams, the “dorsal” and “ventral” streams. The dorsal stream, including area V5/MT, transmits information to parietal lobe networks for localizing objects within a spatial array (where) and is intimately linked to eye-movement mechanisms of selective attention. The ventral stream processes information for the temporal lobe (including V4), concerned with the what aspects of objects, such as form, color, and face recognition. Supporting evidence came from other studies on primates (e.g., Boussaoud, Ungerleider, & Desimone, 1990; Merigan & Maunsell, 1993; Van Essen & Maunsell, 1983) and from
clinical observations of patients with specific deficits of spatial processing (e.g., Damasio & Benton, 1979), movement perception (Zihl, von Cramon, & Mai, 1983), or object recognition (Milner & Goodale, 1995).

Figure 9.1 shows the current model of Atkinson and Braddick, based on both developmental studies and neurophysiology from other species. In this figure, there are some additional divisions between onset of functioning in “local” and “global” processing (discussed below).

Milner and Goodale (1995; Goodale & Milner, 2003) suggested that the ventral and dorsal cortical streams have different functions in the visual control of behavior: The ventral stream is concerned with perceptual processing (including, for example, object recognition) and the dorsal with visual control of action. This is a functional description of the two streams rather than one based on the types of selectivity of cells.

1.4. Action modules in the dorsal stream

Substantial information now exists about many distinct action modules in primates. Figure 9.2 shows a schematic model of some of these dorsal circuits, drawing on the
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Figure 9.2 Schematic model of different action modules within the dorsal stream. *Abbreviations.* AIP, anterior intraparietal area; BA, Brodmann areas; FEF, frontal eye field; FST, fundal superior temporal; IT, inferior temporal areas; LGN, lateral geniculate nucleus; LIP, lateral intraparietal area; MDP, medial dorsal parietal area; MIR, medial intraparietal area; MST, medial superior temporal; PO, parietal occipital area; STS, superior temporal sulcus; TEO, temporal occipital area; V1, primary visual cortex; V2–V6, extrastriate visual cortical areas; VIP, ventral intraparietal areas.

extensive reviews of Milner and Goodale (1995) and Jeannerod (1997). For example, Jeannerod (1997) has argued that the dorsal stream, transmitting visual information to primary motor cortex (M1), has separate divisions for reaching and grasping.

Both dorsal and ventral streams project to prefrontal cortex, and in the mature brain the two systems are heavily interconnected. Visual spatial development can be divided into processes more dependent on dorsal stream functions, such as visual control of action, and those dependent on ventral, such as recognition. However, for many visuo-spatial tasks, processing across two or more modules must be combined. For example, in reaching for an object, the object must be identified as the correct object for the goal of the action using ventral stream networks before the appropriate action is planned using dorsal stream processing. In looking at development of the first stages of dorsal and ventral stream processing, stimuli have been designed to artificially separate one stream from the other, but even here there can be some debate about their separation.

The eye-movement control systems are the first to develop in young infants. These action systems control saccadic tracking and smooth pursuit of objects of interest and switches of attention involving head and eye movements to scrutinize different objects
of interest sequentially. These two systems are schematized in Figure 9.2. However, before discussing development in each of these early action streams, mechanisms for controlling attention must be considered, as the action- and attention-processing systems are heavily interlinked in development.

1.5. Links between attention and spatial action

Mechanisms of attention play an essential role in perceptually "filtering out" irrelevant stimuli and enhancing those of interest. Traditionally, visual attention has been viewed as a unitary, supramodal mechanism subserved by anatomical systems separate from those involved in sensory and perceptual processing (e.g., LaBerge & Brown, 1989; Posner, 1980; Posner & Petersen, 1990). More recently, two attentional brain systems have been postulated, a posterior system that subserves spatial attention and an anterior system involved in various complex cognitive tasks related to executive function (Posner & Dehaene, 1994).

Effects of "covert attention" are seen in subjects' advantages in responding to a stimulus when they know where it will appear, even when they do not move their eyes to fixate the relevant place (Posner & Cohen, 1980). Attention has also been described as a mechanism that enables "selection for action" (e.g., Allport, 1989), the action being either an eye movement (a saccade to fixate the object) or a bodily movement such as a reach toward the object. Such motor acts have been taken as indicators of overt attention shifts. Rizzolatti and others (e.g., Berthoz, 1996; Rizzolatti, 1983; Rizzolatti & Camarda, 1987) have proposed a "premotor" theory of attention, according to which covert attention (without fixating the object of interest) exploits the same selection-for-action mechanism. In this theory, selective attention to a spatial location would involve a number of action modules. In Figure 9.2 areas related to selective attention overlapping with areas in the dorsal stream are highlighted in square boxes. This suggests considerable overlap between the attention and action systems. Whether or not the premotor theory can provide a complete account of adult attention, it is valuable in a developmental context where evidence for attention control comes from overt orienting acts. In the next section, development of these linked attention and action systems in early infancy is discussed.

2. Development of Spatial Selective Attention for Action in Infancy

The most basic visual spatial action system to develop in young infants involves the control of eye movements to track moving objects and people and to stabilize objects on the retina.

2.1. Newborn eye movements

With a large visual field of conspicuous vertical stripes, moving horizontally at a constant speed of around 10 deg/s, optokinetic nystagmus (OKN)—cycling eye movements, each cycle showing a brief period of smooth pursuit following in the direction of the motion,
followed by a rapid saccadic return in the opposite direction—can be observed in newborns. These stabilizing eye movements act to keep the head and eyes fixating on objects of interest in the real world. OKN is the first directional action system to operate in newborns, but does not seem to be one of which there is conscious awareness.

2.2. Smooth pursuit eye movements

Differences have also been found developmentally between newborns and 3-month-olds in the extent and gain of smooth pursuit, which implies that at least in human development, there are different stabilizing mechanisms for smooth pursuit, enabling targets to be tracked, and saccadic movements, enabling orienting. Detailed studies of the development of these stabilizing mechanisms in infants have been made by Claes von Hofsten and his colleagues (see, for example, von Hofsten & Rosander, 1996, 1997). They recorded smooth pursuit eye movements even in newborns, if targets of sufficient size and contrast were used and their velocity was kept relatively low. However, saccadic tracking is much more commonly observed than pursuit in the newborn. They also looked at the development of the initial coupling of eye and head movements, as the infant develops these stabilizing mechanisms. Fairly accurate coupling and a mature vestibular ocular response is achieved in the first few months of life, although many of the tracking eye movements observed in everyday situations in this period are saccadic, rather than continuous smooth pursuit. This changeover from passive saccadic tracking to smooth pursuit eye movements has been taken to imply anticipation of the end-point of the object in space and a planning mechanism used to control eye movements. Further evidence for this comes from the infant’s ability to anticipate where an object will be in space when moving behind an occluder. There are extensive studies in this area concerning the factors, such as path trajectory, velocity, and target appearance, that will change this anticipatory behavior (for a review, see von Hofsten, 2005).

2.3. Eye/head movement systems for switching attention

For switching attention, there is general agreement that the newborn has a “where?” system, which is largely under subcortical control, and is used for orienting the head and eyes to abrupt and significant changes in the world. In the visual domain these are usually changes in luminance or movement. This system has been studied using the fixation shift paradigm (e.g., Atkinson & Braddick, 1985; Atkinson, Hood, Wattam-Bell, & Braddick, 1992). Newborn infants make a shift of the head and eyes from a central target to a target in the peripheral fields appearing at the moment when the central target disappears. The orienting system used when only one target is visible at one time (noncompetition) is likely to operate supramodally, across domains and sensory modalities, as a nonspecific alerting system. The superior colliculus is strongly implicated within this system, although there may be a number of subcortical circuits involved in different components of the responses (see Figure 9.2 for likely areas involved).
2.4. Disengagement and switching attention when targets compete

The crude subcortical system, described above, will orient to a single salient target. However, it works much less effectively in the fixation shift paradigm when a peripheral stimulus appears but the central target remains visible. Responses to a peripheral stimulus when the central target continues to engage fixation require modulation and disengagement of this orienting system by cortical processes. Both right and left parietofrontal areas linked to subcortical eye-movement systems have been implicated as necessary for development of these attention-switching systems in infancy (Atkinson & Braddick, 1985; Atkinson & Hood, 1994; Atkinson et al., 1992). The cortical system appears to become functional around 3–4 months of age in normally developing infants. This modulation by the cortex can be mapped out by varying the interval between offset of one target and onset of another (Hood & Atkinson, 1993). In some infants who have suffered perinatal brain damage involving both cortical and subcortical areas (and in particular the basal ganglia) even the primitive orienting system may not be functional (Atkinson & Hood, 1994; Mercuri, Atkinson, Braddick, Anker, Cowan, et al., 1997; Mercuri et al., 1996; Mercuri, Atkinson, Braddick, Anker, Nokes, et al., 1997).

2.5. Abnormalities of switching attention

The cortical contribution is vulnerable to brain damage, particularly in the parietal lobes. Two infants who underwent hemispherectomy (removal of one complete hemisphere), one at 4 months of age and the other at 8 months, to relieve intractable epilepsy, postoperatively failed to show disengagement and shifts of eyes and head from a centrally viewed target to one appearing in the periphery when the central target remained visible while the peripheral target appeared (i.e., fixation shift under competition). Failure to shift under competition was on the side of space opposite the removed hemisphere, but good shifts of gaze were made on the side controlled by the remaining functional hemisphere, and to either side when only one target was visible; that is, fixation shifts without competing targets (Braddick et al., 1992). In other studies, infants with either focal lesions or diffuse hypoxic-ischemic damage showed this "sticky fixation," an inability to easily switch visual attention from one target to another when two targets are present at once (Atkinson & Hood, 1994; Hood & Atkinson, 1990; Mercuri et al., 1996). This behavior resembles the problems often seen in adult stroke patients as part of a "visual neglect" syndrome.

In this section, the first spatial selective attention systems have been discussed. However, attention is not a unitary function. Besides neural systems controlling selective attention and switches of attention, there are those for sustaining attention and those for inhibiting actions and learning new ones (the latter sometimes being called "attentional control"). Many of these attentional systems mature in later childhood. Development of attention in spatial tasks in childhood will be considered later in the chapter, together with abnormalities in these component systems.
3. Development of Dorsal and Ventral Streams

3.1. Development of cortical motion systems: local and global processing

The first stage of development of the dorsal stream underpinning spatial development is the development of sensitivity to motion. Behavioral and VEP/ERP studies of infants and young children have been designed to distinguish "local" from "global" processing in the dorsal and ventral streams. Local motion processing can be defined as the sensitivity to direction in a small region of the image, such as a short segment of contour, while global motion processing allows the representation of motion over extended regions that may correspond to surfaces and objects. In adults and in other species, global processing has been identified with the integrative properties of neurons in visual area V5, while local processing is identified with neurons in V1 (Braddick & Qian, 2000).

Several aspects of infant performance indicate that global processes operate at an early stage of development. In a dot pattern containing a proportion of randomly moving dots, processing the motion of individual dots cannot yield the overall direction of motion; this requires integration of motion signals over many dots, a process that can be assessed in terms of the motion coherence threshold, the proportion of coherently moving dots required for detecting the global direction. Such thresholds can be measured using preferential looking where it can be shown that by about 3–4 months of age, a strip of coherently moving dots is preferred over an area of random motion; this closely follows the emergence of local direction discrimination at around 2 months (Wattam-Bell, 1994). These results suggest that very soon after local motion signals are first available in the developing brain, the processes that integrate them into global representations are operating quite efficiently. It may be that connectivity between V1 and extrastriate areas including V5, on which this integration is based, exists early at least in a crude form, awaiting the organization of local directional selectivity in V1—perhaps because the latter requires some minimum level of temporal precision before it can function. Deficits in global motion processing, which have been called "dorsal stream vulnerability," may originate in problems processing temporal information at these very early stages of motion processing.

3.2. Comparison of global form and global motion processing

The development of global motion processing—a function of extrastriate dorsal stream processing—can be compared with global processing of form in the ventral stream, where analogous thresholds can be measured. Here subjects must detect the organization of short line segments into concentric circles, with "noise" introduced by randomizing the orientation of a proportion of the line segments. Neurons responding to concentric organization of this kind have been reported in area V4 in macaques (Gallant, Braun, & Van Essen, 1993), an extrastriate area at a similar level in the ventral stream to V5 in the dorsal stream. In infants, form coherence discrimination is apparent
from 4 to 5 months of age from preferential looking and VEP/ERP studies (Braddick, Curran, Atkinson, Wattam-Bell, & Gunn, 2002), with children’s coherence thresholds reaching adult levels at around 7–8 years (Gunn et al., 2002). When dynamically rotating and static versions of the same circularly organized stimulus are compared, later maturity is found in children for the moving dynamic stimulus (global motion coherence stimulus) than the static stimulus (global form coherence stimulus). This is a reversal of earlier development in infancy where static global form detection appears to be earlier than global motion detection (Atkinson & Braddick, 2005).

Specific areas associated with form and motion coherence tasks have been identified for comparable stimuli in functional magnetic resonance imaging (fMRI) studies of normal adults (Braddick et al., 2001; Braddick, O’Brien, Wattam-Bell, Atkinson, & Turner, 2000). This work has shown that anatomically distinct circuits are activated in global processing of form and motion, although each circuit involves parts of both the parietal and temporal lobes, and cannot therefore be said to be strictly “dorsal” and “ventral” in the human brain. However, the activated areas do include dorsal stream areas V5 and V3A for motion, and anatomically ventral areas for form. It has also been found that brain activity measured on fMRI increases linearly with the degree of coherence in an area analogous to V5 (Rees, Friston, & Koch, 2000) and that areas in the lingual/fusiform gyrus, which may include V4, similarly show a linear response for form coherence (Braddick, O’Brien, Rees, et al., 2002).

In summary, although local orientation sensitivity emerges earlier in development than directional selectivity, global organization based on form, pattern, or orientation is found to be less effective in determining infant behavior than global organization based on motion. This may reflect the importance of global motion for segmentation and depth organization of the visual world for early spatial tasks. Such segmentation arises both from the independent movement of objects and from parallax due to self-motion; the latter is effective for infants (e.g., Kellman & Spelke, 1983), even though their self-motion is largely passive rather than actively controlled in the first six months. Later in childhood, sensitivity to form coherence attains adult levels rather earlier than sensitivity to motion coherence, and is less sensitive to developmental impairments; this “dorsal stream vulnerability” is discussed in the following section.

3.3. Abnormalities of dorsal and ventral stream development

The broad division between the functions of dorsal and ventral cortical streams is reflected in abnormal development. We have studied development of these functions in a number of groups of young children with atypical developmental profiles (children with Williams syndrome, autism, fragile-X syndrome, perinatal brain damage resulting in focal lesions and hemiplegia). Across all these groups, a general finding has been that in tasks designed to compare the two streams, the development of the dorsal action stream is more likely to be affected than the ventral. This has led us to a general hypothesis of “dorsal stream vulnerability” (Atkinson et al., 2001; Atkinson et al., 1999; Spencer et al., 2000). Children with Williams syndrome (or infantile hypercalcemia) typically
show a very uneven profile of neuropsychological development, with relatively strong expressive language abilities combined with unusual semantics, good face recognition, but severely impaired spatial cognition (see, e.g., Bellugi, Bihrl, Trauner, Jernigan, & Doherty, 1990; Bellugi, Lichtenberger, Mills, Galaburda, & Korenberg, 1999; Bellugi, Sabo, & Vaid, 1988; Bellugi, Wang, & Jernigan, 1994; Bertrand, Mervis, & Eisenberg, 1997; Karmiloff-Smith, 1998; Klein & Mervis, 1999; Pezzini, Vicari, Volterra, Milani, & Ossella, 1999). They reach motor milestones later than typically developing children, are often delayed in learning to walk and in the development of fine motor skills, and on a standardized test of motor function (Movement ABC; Henderson & Sugden, 1992) they show an average delay of at least 2 years (Atkinson, Braddick, Anker, et al., 1996). Problems that persist into later life include uncertainty when negotiating stairs or uneven surfaces (Atkinson, Braddick, Anker, et al., 1996), awkward gait and joint contractures in some children (Kaplan, Kirsschner, Watters, & Costa, 1989), and difficulty with the use of everyday tools.

This neuropsychological profile is consistent with the possibility that ventral stream processes (e.g., face recognition) are relatively unimpaired, but dorsal stream function for visual control of action is developmentally abnormal. We have explored this possibility in several ways. First, we have compared tests of motion and form coherence which require global integration by extrastriate visual areas. Many children with Williams syndrome (WS) have considerable difficulty with the motion task relative to the form task (Atkinson, Braddick, Anker, Curran, & Andrew, 2003; Atkinson et al., 1997). In WS, these deficits are found across the age range from 3 years to adulthood (Atkinson, Braddick, Rose, et al., 2006). The same pattern is found in some younger, typically developing children (4–5 years) and so, although the results are in line with the “dorsal vulnerability” hypothesis, they suggest a more general delay with ability never approaching adult levels, but asymptoting at the 4–5 year level.

3.4. Use of motion information for more complex discriminations in infancy

In the previous section, the psychophysical sensitivity of infants to simple stimuli varying in form, pattern, and motion has been measured. These abilities are useful to young infants for understanding the spatial world around them. For example, recognizing the pattern of motion of a human being rather than a rocking chair is fundamental to separating objects in space, and for separating one’s own motion from the intrinsic motion of objects and people. Detecting three-dimensional (3-D) information from the distribution of optic flow information is also a necessary starting-point for segmenting objects from their background.

A diverse range of studies have tested infants for complex discriminations for motion information in spatial tasks. Discrimination has been demonstrated between rigid and nonrigid transformations of a 3-D object (Walker, Owsley, Megaw-Nyce, Gibson, & Bahrick, 1986). Others have shown quite subtle discrimination of 3-D structure from motion (Arterberry & Yonas, 1988, 2000; e.g., the presence of an interior corner on a cube, represented by random dot kinematograms), and kinetic depth information
leading to recognition of a 3-D shape subsequently provided through disparity cues. Infants' sensitivity to the temporal direction of apparently causal event sequences can be demonstrated (e.g., Leslie, 1984), and a preference has been reported for dynamic event sequences with simple shapes that adults categorize as social interactions (Rochat, Morgan, & Carpenter, 1997).

Infants have also shown sensitivity to the patterns of point-light motion that characterize biological motion (e.g., Bertenthal, Proffitt, Spetner, & Thomas, 1985; Booth, Pinto, & Bertenthal, 2002). Most of these discriminations have been in tests of 3- to 6-month-old infants, and none have been shown so early that they require us to revise the view that the general ability for directional discrimination emerges after 7 weeks of age. On the other hand, they demonstrate that, only a few weeks after infants acquire any ability to discriminate motion directions, they can exploit this ability in a wide variety of complex perceptual functions. Together with the findings on global motion, these results support the idea that the emergence of motion processing in development is constrained by the development of relatively low-level directional mechanisms, rather than by immaturity of processes that elaborate and integrate motion information.

3.5. Dorsal and ventral pathways beyond extrastriate areas

The possibility of different courses of development for dorsal and ventral streams has already been mentioned, but there are also important developmental differences between different modules within a stream. The major milestones of exploratory head and eye movements, directed reaching and grasping, and locomotion each involves integrated function of a different spatial action module processing dorsal stream information. All these action programs must involve some spatial analysis of the visual layout, but the different systems need representations at different scales and with different frames of reference. For reaching and grasping, the infant only needs representation of space near to the body and an egocentric frame of reference to match object locations to hand actions. For locomotion, the child needs to represent the environment on a scale beyond arm's length, and with a reference frame that remains stable in space as the body moves. Next in this chapter, we will briefly describe studies on the development of reaching, grasping, and locomotion which are all underpinned by dorsal stream circuitry.


4.1. Reaching and grasping in infancy

Typically developing infants usually start to reach and grasp successfully for objects in near distance at around 4–6 months of age. Two kinds of visual information must be processed within the visual action system controlling reaching and grasping. First, the
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location of the object, laterally and in distance, must be identified. Second, visual analysis is required to determine whether the object is a suitable target for reaching and grasping.

For the first, the coincidence in timing between the development of binocularity around 4 months (Braddick, 1996) and the emergence of visually guided reaching suggests that binocular disparity information, associated with convergence, is a key input to the visuomotor module for reaching. This is confirmed by the finding that binocular information is critical in determining the kinematics of infants’ reaches (Braddick, Atkinson, & Hood, 1996).

From 6 to 9 months, reaching appears as a quite compulsive behavior for small objects presented within range. This behavior raises the question of the visual information by which an infant determines that an object is graspable and hence a suitable target for reaching.

4.2. Preferential looking and preferential reaching

The development of distinct visuomotor modules, and their ultimate integration, is illustrated by experiments which combine preferential looking with preferential reaching (Newman, Atkinson, & Braddick, 2001). In preferential looking, infants make an orienting response of head and eyes toward the most salient object or region in the visual field. Presentation of paired stimuli allows the relative salience of these stimuli to be assessed. Depending on the development of various cortical modules, salience is a function of luminance, color, motion, or depth contrast, and of spatial structure defined by such contrast. It is also modulated by novelty (as in habituation tests) and by the special significance of certain stimuli such as faces. Salience, so defined, is the visual computation characteristic of the set of cortical modules which contribute to the orienting system (providing output through the superior colliculus). In the case where an infant is presented with two 3-D objects, similar in shape and surface but different in size, the infant tends to orient to the larger object (King, Atkinson, Braddick, Nokes, & Braddick, 1996; Newman et al., 2001).

However, reaching is only an appropriate response for objects that are small enough to be grasped. Thus the visual modules that provide information controlling this response need to compute, as well as the spatial direction for the reach, the size of the potential target. This computation is not necessarily possible for the infant at the age when the motor schema of reaching becomes available, but when it is possible, it will cause reaching to be preferentially directed to the smaller object of a pair, when the larger is beyond the span on the infant's hand.

Thus the two visuomotor dorsal stream systems for orienting and reaching may be driven by different visual information from the same pair of objects. The studies of King et al. (1996) and Newman et al. (2001) showed that these systems interact differently at different ages; a possible organization of the two systems at each stage is schematized in Figure 9.3. When infants first start to reach (up to 8 months), they do not show a significant reaching preference between large and small objects. However, their
reaching is predominantly directed to the object they initially fixate. We infer that processing of the specific visual attributes signifying graspability is not yet linked into a visuomotor module for reaching, and that at this stage there is a substantial coupling between the system controlling reaching and that determining orienting.
Between 8 and 12 months a strong preference emerges for reaching for the smaller object, which is within the span of the infant's hand. Thus an effective visual analysis of graspability has developed and serves as an input to the reaching control system. Infants at this age show a noticeable decoupling of reaching and initial orienting: They are more likely than younger or older infants to first fixate one object and then reach for another. This decoupling can be emphasized by manipulating visual salience: Marking a schematic face on one object increases preference for looking at that object, without altering its graspability and hence without a corresponding increase in the tendency for it to elicit reaching in competition (Newman et al., 2001).

After 12 months, reaching becomes less selective toward the smaller object, perhaps because it has become a less compulsive behavior, and also because the infant's grasp can encompass larger objects. At the same time, reaching and initial looking become more congruent again. It appears that the orienting and reaching systems can be integrated into a single piece of goal-directed behavior.

There is an extensive literature on factors affecting reaching in infants (for reviews, see Berthier & Keen, 2006; von Hofsten, 1991). A number of studies have found changes in development during the first two years of life in the kinematics, dependent on the size of the target (e.g., Fagard, 2000; Fagard & Jacquet, 1996). For example, in a recent study cubed objects were varied across a wide range of sizes (Braddock & Atkinson, 2003). For infants between 4 and 6 months, smaller objects elicited more grasping actions than nongrasp contacts. Between 6 and 10 months, the proportion of bimanual reaches for the larger objects increased, with different kinematics of the speed profiles in grasp compared to nongrasp reaches. Some of these differences in kinematics may reflect an understanding of the "graspability" of the object. This suggests that areas such as the anterior intraparietal area may be operating to identify salient object properties for guiding actions in the first year of life.

4.3. Bimanual coordination

Alongside reaching and grasping with one hand comes bimanual coordination for the many tasks requiring two hands to reach a goal. Early bimanual coordination patterns are typically similar for the two hands in that both hands reach or grasp synchronously and involve proximal control. With improvements in posture and visuomotor skill at around 6 months of age, the infant starts to explore objects placed in his or her hand and transfers the object from one hand to the other. These actions form the basis of later developing complementary bimanual patterns of coordination. In these early object explorations, only one hand is active at a time, but toward the end of the first year the two hands are capable of carrying out two different actions simultaneously; for example, lifting and holding open the lid of a box in order to retrieve a toy. The first bimanual attempts are often poorly timed and clumsy, but by 18 months of age the infants perform these tasks smoothly and efficiently with good spatial and temporal organization (e.g., Bruner, 1970; Diamond, 1991; Fagard, 1994). Bojczyk and Corbetta (2004) demonstrated that with repeated exposure (training) infants were able to
develop well-coordinated bimanual actions by 8–9 months and thus outperform age-matched infants with no previous experience of the task.

4.4. Visual information in motor planning: abnormalities in Williams syndrome

For everyday motor planning, many visual factors need to be taken into account. For example, for an object to be grasped, its distance, size, and orientation must be judged accurately. One experimental paradigm that gives insight into more complex visual control of action is the “postbox task” used by Goodale and colleagues (Goodale, Milner, Jakobsen, & Carey, 1991) to study a ventral stream impaired patient. Goodale and colleagues found a striking dissociation. This patient could accurately post a card through an oriented slot (dependent on dorsal-stream control of action) but failed on perceptual matching of the slot orientation (dependent on ventral-stream processing for perceptual judgments). On a task of this kind, children with Williams syndrome showed much greater inaccuracy in posting the card than in matching the card’s orientation to that of the slot, compared with normally developing children (Atkinson et al., 1997). This supports the account of a dorsal-stream deficit, although again the degree of deficit was quite varied between individuals.

When picking up square shapes, children with WS were also impaired on adjusting grip aperture to target size, and made slower reaches, with more movement segments, suggesting that they were less able to program accurately the reach as a whole (Newman, 2001). There thus appears to be a continuing immaturity in WS children in the dorsal stream units controlling reaching and grasping. However, they also show poor performance on a matching test (judgment for the size of the squares without picking them up), indicating that problems in WS appear in both “ventral” and “dorsal” aspects of the task (Newman, 2001).

In the postbox task discussed above (Atkinson et al., 1997), children with WS often found their hands in awkward postures as they rotated the card, suggesting inefficiency in end-state planning, the ability to take into account the intended use or end-state of the object. End-state planning is likely to involve the integration of dorsal-stream information with prefrontal areas involved in inhibiting inappropriate actions and coordinating the elements of action sequences. An end-state planning deficit in WS was explicitly tested by Newman (2001) using a handle rotation task adapted from adult studies by Rosenbaum and colleagues (Rosenbaum, Vaughan, Barnes, & Jorgensen, 1992). Results from this study indicate that children with WS either do not attempt end-state planning or are unable to make the spatial transformations required to predict the end-state correctly.

Overall, these studies found subtle and variable deficits in the use of dorsal-stream information to control manual action, although these were not always dissociated from ventral-stream performance. The deficits were most striking beyond the early stages of visual processing for actions, in the use of visuospatial information for end-state planning. There may well be a “cascade” effect, with early abnormalities in more basic parts
of the dorsal pathway affecting later development of complex feedback loops involved in visuomotor planning, which show deficits even if the lower level effects are overcome. The individual variability highlights the degree to which adaptive strategies may lead to differently configured systems even if there is a common initial developmental deficit.

5. Development of Action Modules for Locomotion and Navigation

The integration of different dorsal-stream modules is required when locomotion becomes part of the infant’s behavioral repertoire. For instance, a desired object is processed in far space as a target for locomotion, and brought into near space as a target for reaching. The child must become able to switch attention from near to far, and to engage the visual processes required at these two scales. Other aspects of visual behavior also require such shifts; for example, joint attention invoked by an adult’s pointing gesture, which is also achieved around the beginning of the second year (Butterworth & Grover, 1990). It remains a challenge for further research to characterize and understand these processes of integration and switching.

A key component of successful locomotion is balance. The use of visual information to maintain balance starts as young as 15 months (Lee & Aronson, 1974) and develops throughout childhood (Shumway-Cook & Woollacott, 1985). Real-world locomotion also depends on avoiding obstacles or accommodating movement to them (Patla, 1991). The visual decisions involved are: Can I step over this or should I walk round it? And, if I step over this, what height should I raise my feet to? These considerations are important when planning both single steps and longer routes.

We know that children avoid objects based on the visual information they have about their size. For example, the classic “visual cliff” experiment showed that young infants avoided crawling on an area they perceived to be dangerously deep (Gibson & Walk, 1960). Some new work considers how visual information is used to accommodate movements appropriately. This work shows that step descent is divided into “transport” and “placement” phases. The transition between phases is marked by the lead leg swinging in to contact the step. The vertical distance traveled by the knee during the transport phase increases in proportion to the step’s riser height. This pattern is lost when vision is removed. Strikingly, this ability to scale movement to riser height is present in children as young as 3 years old. However, these children depend more than adults on online visual control to accomplish this (Cowie, 2007). These studies illustrate that children use vision to control accommodative processes as well as avoidance decisions.

Successful locomotion involves not only using visual information but also combining it with information about one’s own locomotor abilities and current body state. Toddlers take locomotor decisions based on their own walking skill and experience (Kingsnorth & Schmuckler, 2000) and body dimensions (Adolph & Avolio, 2000). In a
new “stepping stones” task children must choose to follow one of two paths of “stepping stones” across a pretend “river.” Children as young as 3 years are influenced in their planning by their own body states—in this case the need to make anticipatory changes in foot position before the start of the task (Cowie, Smith, Braddock, & Atkinson, 2006a; Cowie, Smith, Braddock, Atkinson, & Nardini, 2006).

As with other visuomotor tasks, the visual control of locomotion is likely to be mediated by the dorsal stream. There is little work on this, but the patient D.F., with damage to the ventral stream but a spared dorsal stream, can accurately scale her stepping up movements (Patla & Goodale, 1996), though not her verbal estimates of step height; while young children with Williams syndrome, who show other dorsal-stream impairments, cannot scale stepping down movements in the task reported above (Atkinson, Braddock, Nardini, Cowie, et al., 2006; Cowie, Smith, Braddock, & Atkinson, 2006b).

6. Development of Spatial Localization in Location Memory Tasks

Further important aspects of spatial development depend on the systems underpinning accurate spatial localization in memory tasks. From studies of adults using psychophysics, brain imaging, and transcranial magnetic stimulation (TMS), and from animal lesion studies, dorsal stream parietal networks are the primary processing areas for basic spatial localization, while the hippocampus, parahippocampal gyrus, and entorhinal cortex are involved in more complex spatial memory tasks (see review by Stiles, Paul, Ark, & Akshoomoff, in press).

6.1. Location memory in infants

In the classic Piagetian “A not B” task, an object is repeatedly hidden at location A before a test trial on which it is hidden in novel location B. Infants aged around 9 months continue to search incorrectly in the “familiar location” (A); this is the “A not B” error. AB tasks have been used to test object permanence, that is, the infant’s knowledge that objects exist independently over space and time. However, they also reveal the spatial framework that infants use to define the location of an object.

Children’s ability to overcome the AB error depends on many factors. Self-locomotion reduces the likelihood of the error (Bertenthal & Campos, 1990; Horobin & Acredolo, 1986; Kermoian & Campos, 1988), and looking-time measures indicate that children are aware of the difference between the A and B locations before they are able to avoid the AB error in their reaching responses (e.g., Baillargeon & DeVos, 1991; Hofstadter & Reznick, 1996).

In nonhuman primates, bilateral lesions of dorsolateral prefrontal cortex disrupt AB performance, but bilateral lesions to parietal cortex (Diamond, 1991; Diamond, Werker, & Lalonde, 1994) or hippocampus (Diamond, Zola-Morgan, & Squire, 1989) have little effect. These findings suggest that children’s solution of the task depends on
frontal maturation. Further evidence from changes in frontal metabolic activity in the first year (Chugani, Phelps, & Mazziotta, 1987; Jacobs et al., 1995) supports this view, as do data from EEG and near infrared spectroscopy measures (Baird et al., 2002; Fox & Bell, 1990). Bell and Fox (1992) found increased anterior-posterior EEG coherence associated with improved performance on the AB task, suggesting that long-range axonal connections may contribute to the change in performance. As discussed earlier, these pathways may be critical for the control of reaching. Thus maturation of the anterior-posterior system may account for the differences in performance on reaching and looking tasks.

Infants make a related error when looking for an object after changing position. Infants aged 6–12 months old often search egocentrically after they have moved; that is, they seem not to appreciate that a target that was previously on their left is now on their right (e.g., Acredolo, 1978). As with the AB error, infant egocentrism is moderated by many factors. It can be overcome, particularly when a visual cue that directly indicates the target is available (e.g., Rieser, 1979). Crucial for mature navigation, however, is the later developing ability to use indirect landmarks, which do not directly indicate a target.

6.2. Development of location memory in childhood

Improvements in location memory in mid-childhood follow prefrontal, posterior parietal, and hippocampal maturation. Children’s use of different spatial frames of reference to remember a location develops gradually. Egocentric representations, using the body as a reference, provide a good basis for immediate action toward objects. More robust representations are provided by encoding where objects are relative to stable landmarks (using an allocentric reference frame). This would enable objects to be found even when the viewer changes position. Another way to deal with a change of position is to track where an object is while the observer is moving (“updating” the egocentric representation as the observer moves); adults use these different frames of reference and updating processes in complementary ways.

Representations using external landmarks are reliably used for action by the second year (Huttenlocher, Newcombe, & Sandberg, 1994). At 16–36 months, children retrieve objects hidden in a sandbox after walking around to the other side (Newcombe, Huttenlocher, Bullock Drummey, & Wiley, 1998), showing coding relative to landmarks and/or spatial updating with self-motion. A more difficult problem is processing a change of viewpoint caused by the rotation of an object, rather than one’s own displacement. In this case, the self-motion information that could be used to maintain an accurate egocentric representation while walking is not available. Judging what would be where if the viewer’s viewpoint changed presents a “perspective problem” (Huttenlocher & Presson, 1973), potentially a test of children’s ability to use a purely viewpoint-independent or allocentric frame of reference. Piaget and Huttenlocher’s perspective-taking studies were not usually solved until around 10 years, although similar tasks have been solved earlier with a modified procedure (Newcombe & Huttenlocher, 1992).
Nardini and colleagues (Nardini, Burgess, Breckenridge, & Atkinson, 2006) tracked both egocentric coding, suitable for simple spatial recall, and allocentric codings, capable of solving the “perspective problem,” in 3–6-year-olds within the same task. Children saw a toy hidden under one of 12 cups on a board with landmarks attached to two of its edges, and had to find it after a manipulation. The task systematically varied whether the same view of the board was seen at hiding and test (i.e., whether the toy kept its place relative to the body) and whether the array was rotated (i.e., whether the toy kept its place relative to the room). In 3–6-year-olds, as in adults (Wang & Simons, 1999), there were parallel, additive effects of both body and room frames of reference. The performance improvement when a familiar view allowed egocentric coding was already seen at 3 years. From 5 years, children showed successful recall after changes of viewpoint caused by rotation, which could only be solved by attending to the array and its local landmarks (a “perspective problem”). This ability may depend on codings relative to landmarks, supported by the hippocampus (O’Keefe & Burgess, 1996), and may also include mental rotation, which has elsewhere been demonstrated from 5 years (Kosslyn, Margolis, Barrett, Goldknopf, & Daly, 1990; Marmor, 1975), and shows activation including parietal areas (e.g., Booth et al., 2000) in children and adults.

A separate question concerns how children combine different kinds of visual information to maintain their sense of orientation. The human environment includes discrete landmarks, such as trees or buildings, which could be individuated by color or shape, as well as elements of layout, such as the shape of a room or a field, whose geometric aspects could be coded. Hermer and Spelke (1994, 1996) found that 18–24-month-olds, disoriented in sparsely featured enclosures, re-established their orientation using geometry (enclosure shape), but not the colors of the walls. They argued that early reorientation depends on a specialized geometric module “encapsulated” with respect to color, and that the eventual solution of the task depends on language (Hermer-Vazquez, Spelke, & Katsnelson, 1999).

Solution of the task by nonlinguistic animals (reviewed by Cheng & Newcombe, 2005), and children in larger enclosures (Learmonth, Nadel, & Newcombe, 2002), argues against this account. The ability of 18–24-month-olds to reorient using wall colors in small square enclosures (Nardini, Atkinson, & Burgess, 2008) also demonstrates that reorientation is not encapsulated with respect to color. Nevertheless, the early dominance of room geometry over color for spatial orientation, when both are available, remains an interesting phenomenon that has not been fully explained. Geometric judgments might be favored in small, but not large rooms, as relative wall lengths are easier to judge when standing at a corner (Sovrano & Vallortigara, 2006). In addition, children’s poor use of color may not be specific to disorientation, but could represent a more general phenomenon. Oriented 18–24-month-olds searching in boxes on a table top were poor at using box color alone as a cue, and were especially likely to disregard colors when they had to be combined in memory with actions (Nardini et al., in press). At 30–36 months, children no longer showed this disregard for color, but did show it for monochromatic textures. “Disregard of color” may therefore be part of a
more general developmental phenomenon linked to the uneven development of links between the dorsal and ventral visual streams and prefrontal mechanisms for working memory.

6.3. Impairments of spatial representation in memory in atypical development

Early focal cortical injury in the right hemisphere is associated with deficits in organizing spatial elements coherently, while left hemisphere injury is associated with poor encoding of detail in complex forms (Stiles, Stern, Trauner, & Nass, 1996; Stiles-Davis, Sugarman, & Nass, 1985). Although these patients often show remarkable recovery compared with those obtaining similar injuries in adulthood, fine-grained analysis indicates persistent deficits (Stiles, 2000).

Children born very premature, who show a range of cognitive deficits by school age (Bhutta, Cleves, Casey, Craddock, & Anand, 2002; Marlow, Wolke, Bracewell, Samara, & EpiCure Study Group, 2005), have especially marked deficits in the visuospatial and visuomotor domains (Atkinson, Braddick, Nardini, Anker, et al., 2006). On a spatial memory task assessing recall using different frames of reference, 6-year-olds born at 25–30 weeks' gestation had an average delay of more than one year across conditions (Nardini, Atkinson, et al., 2006). On average, this group's deficit was as large for external frames of reference (landmarks) as for egocentric recall. However, correlations with other cognitive and motor tests indicate subgroups with differential patterns of impairment. Impairments to spatial updating for changes of viewpoint produced by walking may be predicted by poor detection of coherent motion (related to visual processing of optic flow), while performance on the "perspective problem" (changes of viewpoint produced by movement of the array) is predicted by "frontal" tests of inhibition and response selection, suggesting that a "frontal" inhibition or selection process is involved in this task.

In addition to their visuomotor problems (Atkinson et al., 1997), children and adults with Williams syndrome (WS) show deficits on the purely perceptual task of egocentric localization for locations on a screen (Paul, Stiles, Passarotti, Bavar, & Bellugi, 2002; Vicari, Bellucci, & Carlesimo, 2006). Impairments to representations of location may therefore underlie their visuomotor and constructional difficulties. To compare the development of egocentric and landmark-based recall in Williams syndrome, we tested children and adults with WS on the "frames of reference" task. Overall, individuals with WS showed parallel, additive use of body- and room-based reference frames; however, these were combined anomalously in development, and performance in adulthood was not better than at 4 years in typical development (Nardini, Atkinson, Braddick, & Burgess, 2008). Crucially, adults with WS showed only marginal ability to use local landmarks to solve the "perspective problem," solved by typical children at 5 years. Visuomotor and constructional deficits in WS may thus be caused in part by the unusual integration of different frames of reference in development, and the poor ability to select local frames of reference even in adulthood.
7. Summary of Abnormal Spatial Development

Abnormalities of development have been seen in every aspect of spatial vision. Examples have already been given in each section of this chapter; these are summarized here together with a brief description of a number of relatively new tests for diagnosing spatial deficits in infants and very young children.

7.1. Deficits in the development of spatial attention linked to action

Earlier in the chapter, results from studies of infants who underwent hemispherectomy showed abnormal attentional control of actions, such as foveating or reaching for a target in peripheral vision on the side of space opposite the hemisphere lacking a normal cortex. This was only seen in conditions where two targets were competing for attention (Braddick et al., 1992). This failure to disengage and to make eye and head movements to a salient peripheral target when a central target is still visible is a common finding in many children with perinatal brain damage involving parietal and frontal areas. When damage extends to subcortical networks, then even without competition, shifts of gaze (and presumably attention) can be absent and/or slow.

7.2. Deficits in spatial attention in childhood

In school-aged children there are many studies of deficits related to attentional loss. In normally developing children there is considerable improvement in attentional capacities throughout childhood and adolescence. Three different components of attention have been identified from adult studies and patient populations, each with rather different neural underpinnings. The first component is linked to selective visual attention in visual search tasks. The second component is "sustained attention," which can be measured in vigilance tasks, and the third component involves inhibition of a prepotent response to switch and make a new association. Studies indicate that developmental trajectories differ for different attention components. Earlier maturity in selective attention (before 6–7 years), for example, contrasts with the continued development of sustained attention into adolescence, and rapid development of executive function between 7 and 11 years (Kelly, 2000; McKay, Halperin, Schwartz, & Sharma, 1994; Rueda et al., 2004). One test battery used to study attentional development is the Test of Everyday Attention for Children (TEACH; Manly et al., 2001), which examines components of attention between the ages of 6 and 16 years. Data from this battery support the notion that distinct components of this function exist in childhood, and suggest differential impairment of these components in clinical samples including ADHD (Heaton et al., 2001; Manly et al., 2001) and traumatic brain injury (Anderson, Fenwick, Manly, & Robertson, 1998).

A number of tasks have been developed to examine executive function in preschool children (e.g., Backen-Jones, Rothbart, & Posner, 2003; Gerstadt, Hong, & Diamond,
1994; Hood, 1995; Hughes & Russell, 1993; Kirkham, Cruess, & Diamond, 2003; Zelazo, Frye, & Rapus 1996); these show significant changes in the ability to inhibit prepotent responses and shift attention flexibly between the ages of 3 and 4 years. Many of these inhibitory tasks involving inhibition of a prepotent response are thought to have their underpinnings in frontal lobe circuitry. In one such test, “counterpointing,” the child first points as rapidly as possible to a target which appears to either the left or right of a fixation spot. The rule is then changed and the child is asked to point as rapidly as possible to the opposite side to where the target appears. On this test, inhibitory control is achieved on average by 4 years of age in typically developing children, but can be considerably delayed in clinical populations (Atkinson et al., 2003). Measures have also emerged in recent years that allow for more formal assessment of selective and sustained attention in preschoolers, demonstrating improvement in these abilities between the ages of 3 and 6 years (Corkum, Byrne, & Ellsworth, 1995; Mahone, Pillion, Hoffman, Hiemenz, & Denckla, 2005; Prather, Sarmento, & Alexander, 1995).

Despite the clear changes in attention through the preschool age range, however, comprehensive batteries of attention measures are not generally available for children between 2 and 6 years. A new battery of attention measures is being developed for typically developing 3–6-year-olds and children in this mental age range, such as chronologically older children with Down syndrome or Williams syndrome (Breckenridge, 2007).

7.3. Deficits in development of the dorsal stream relative to ventral stream

The general hypothesis of “dorsal stream vulnerability” (Atkinson et al., 1999; Braddick, Atkinson, & Wattam-Bell, 2003; Spencer et al., 2000) was discussed in section 3.3. This refers to the general finding that across a number of clinical populations with very diverse etiologies (e.g., Williams syndrome, fragile-X syndrome, congenital cataract patients, autism, children with hemiplegia), when tasks are carefully designed to compare and isolate responses related to the dorsal and ventral streams, the development of the dorsal stream is more likely to be affected than the ventral (for a review, see Braddick et al., 2003).

7.4. Planning and execution of actions in spatial tasks

Taken together with a comparison of dorsal- and ventral-stream function in the postbox task (see section 4.4) and in other motor-planning tasks, Atkinson and Braddick have proposed that Williams syndrome is an example of broader dorsal stream vulnerability. Support for this relative deficit in dorsal-stream networks in WS comes from a structural MRI study with 2-year-olds (Mercuri, Atkinson, Braddick, Rutherford, et al., 1997) and from structural and functional MRI studies with WS adults (Meyer-Lindenberg et al., 2004; Reiss et al., 2004). Frontal executive functions have been found to be an additional area of deficit for WS, even relative to verbal IQ. The extent of this deficit depends on the nature of the task and is much greater when it requires inhibition
of a prepotent spatially directed response, notably in counterpointing (see above) and also in a spatial detour task (Biro & Russell, 2001). Inhibition of a verbal response is considerably less affected. Thus the transmission of spatial information to frontal systems within the dorsal stream seems to be specifically disrupted in WS.

7.5. New tests of spatial development

Shape-matching and block-construction copying tasks have been normalized and standardized as part of a battery (Atkinson Battery of Child Development for Examining Functional Vision [ABCDEFV]) to measure functional vision in children of mental ages between birth and 5 years (Atkinson, Anker, Rae, Hughes, & Braddick, 2002). All tests have been standardized with typically developing children, failure on a particular subtest usually corresponding to a score below the 15th percentile for the appropriate age. The battery is divided into tests of “core vision,” requiring minimal saccadic tracking eye movement, including measures of acuity, refraction and field perimetry, and additional visuomotor, visuocognitive, and spatial tests (requiring minimum motoric skills of reaching and pointing or grasping with one hand). Some subtests measure spatial vision related to more ventral or more dorsal stream processing, whereas some involve integration across both processing streams.

The ABCDEFV has been used to test spatial development in a number of clinical populations. In a large-scale population infant vision screening program (n = 5,000) for detection of strabismus and refractive errors in 9-month-old infants and prevention of later onset strabismus and amblyopia through spectacle correction in infancy (Atkinson, Braddick, Bobier, et al., 1996), children who had significant hyperopia in the first year of life were significantly worse on many of the visuospatial ABCDEFV tests throughout the preschool years. However, none of the items on the Griffiths scales of pediatric development showed a difference, arguing against a general developmental delay in this clinical group. Overall, these results indicate mild deficits in the hyperopic group, concentrated in areas of visual perception and visuomotor control related to both dorsal and ventral stream development (Atkinson, Anker, Nardini, et al., 2002).

A second battery that has been developed is the Movement ABC (Henderson and Sugden, 1992), a standardized assessment of everyday visuomotor and spatial competence for 4-16-year-olds. The Movement ABC includes tasks to assess visuomotor development within three categories: manual dexterity, balance, and ball skills. At 3 and 5 years, children who had been significantly hyperopic as infants scored lower on these tests (Atkinson et al., 2005). The pattern did not show a subgroup of poorly performing children, but suggested a mild but widespread deficit in the formerly hyperopic group. There was no significant difference between children who had worn spectacles to correct refractive errors in infancy and those who had not, which suggests that there may not be a direct causal connection between poorer sensory vision in infancy and poorer preschool visuomotor and visuocognitive abilities, but rather that abnormal refraction and strabismus in infancy is a soft sign of poorer brain development, affecting not only specific eye–brain networks for sensory vision, but those of
selective attention and spatial cognition involving temporal, parietal, and frontal lobe areas with links to subcortical networks.

In longitudinal studies of very premature infants (under 32 weeks' gestation), a comparison has been made between their visual spatial development, brain imaging at birth and term, and their general neurological and cognitive development over a period from birth to 5 years of age. Diffuse excessive high signal intensity (DEHSI) is a common feature of white matter abnormality in very premature infants when scanned around term. The presence and degree of white matter damage (DEHSI or periventricular leukomalacia) correlated with the measures of early attention on the fixation shift test, deficits in onset of cortical functioning measured with orientation-reversal VEPs, and deficits on spatial tasks in the ABCDEFV and preschool attentional tests of frontal lobe function (Atkinson & Braddick, 2007).

In a second cohort, children who were born very prematurely were intensively studied between 6 and 7 years of age. Across the group, deficits in visuomotor, visuospatial, and attention tasks are found, with relative sparing of verbal performance and language. From this analysis, an overall model of brain development for premature infants is that, as in other developmental disorders, there is vulnerability in the more dorsally controlled areas with attentional deficits arising from poor frontal lobe connections, whereas development of language and communication and ventral-stream function is relatively good (Atkinson & Braddick, 2007). For some children in the premature group these deficits are relatively mild, but many are likely to have subtle learning difficulties (e.g., mild ADHD) which persist and have cascading effects on later academic abilities.

8. Summary of the Developmental Model of Visual Spatial Development

Figure 9.1 illustrates Atkinson and Braddick's (2003) account of the developmental sequence and the broad neural processes corresponding to it. From an initial subcortical stage, there is development of functioning in specific cortical channels, followed by development of integrative processes across channels within a single stream so that the infant can build up internal representations of objects and individuals. This aspect of the developing processes takes place largely in the ventral stream, with dynamic online information contributed from the dorsal stream to control orienting by eye and head movements. Of course, for objects to be represented, information about color, shape, and texture must be integrated with motion information at a relatively early stage so that objects can be segregated from each other and from their background. These processes provide object representations which must be integrated with dorsal-stream spatial information to allow, later, emergence of the visual action systems associated with reaching/grasping and with locomotion. These action systems are a combination of visual, attentional, and motor systems. It is over-simple, however, to show this as a linear sequence; there are likely to be important feedback loops, by which the consequences of a new development can affect the way in which earlier established processes
work. Furthermore, a description of the sequence is only the start. We still cannot explain timing differences in functional onset and plasticity in one system as opposed to another: Why do some processes in certain networks start to function earlier than others? How far are we seeing the unfolding of a maturational sequence, and how far does the developmental trajectory depend on exposure to the environment, including the kinds of exposure made possible by earlier networks becoming functional? There are still many unanswered questions.

A model of deficits in spatial development has been suggested: “dorsal stream vulnerability.” This is based on the general finding that tasks related to more dorsal stream areas show higher levels of deficit that those related to ventral across many different clinical conditions including children with developmental disorders related to genetic abnormalities such as Williams syndrome and fragile-X. We still need to find the basic cause of this difference in plasticity between dorsal stream and ventral stream modules. It may have its origin in very low-level timing mechanisms in subcortical or early cortical areas; it may depend on a misbalance between the number of functional magnocellular and parvocellular cells and their integration; or it may depend on integration of information from processing in many different occipital, parietal, and frontal areas before the planning and decision for action is taken. Of course, we do not know whether a deficit which seems similar behaviorally arises because of identical faults in its neural processing. It is to be hoped, with improved imaging methods and specific behavioral measures, that we may be able to answer these questions in the future and provide appropriate interventions, tuned to the individual, to alleviate spatial difficulties in children.

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References


The Neuropsychology of Attention Development

Maxine Sinclair and Eric Taylor

We are constantly bombarded with more complex sensory information than our cognitive systems can fully process at one time. Although the sensory organs perform parallel processing, the capacity is severely limited and sensory information competes for representation in multiple brain systems. The ability to direct attention toward sensory events in the extrapersonal space is of crucial importance for adaptive behavior. Evidence based on imaging studies and studies of brain-lesioned organisms suggests that the human brain has the ability to select a limited subset of the available information for detailed processing, to filter out distracting, irrelevant information, and to switch this focus to some other part of the environment when necessary.

Whilst there is universal consensus that the brain has inherent limitations with regard to the amount of information it can simultaneously process, theorists have found agreement on an operational definition of attention, and the scope of the phenomena to which it applies, for the most part elusive. Attention has variously been regarded as an active mechanism that selects or binds (e.g., Treisman, 1988), a resource that is shared between competing processes (Norman & Bobrow, 1975), a factor that amplifies signals and improves sensitivity (Lu & Dosher, 1998), or a conglomerate of diverse mental functions (Driver, Vuilleumier, Eimer, & Rees, 2001). What is universally accepted is that attention is not a one-dimensional construct (i.e., a single resource process or mechanism), but complex, multidimensional processes involving several overlapping components (Heuer, 1996) that form the basis of, and are affected by, all cognitive activity.

In this review, we will consider current neurological and neuropsychological models of attention. We will consider in some detail the formal constructs of attention, providing definitions of each as well as evidence from functional neuroimaging studies that have enabled researchers to examine the neural basis of attention in the intact human brain with relatively high spatial resolution. Space does not allow us to catalogue the particular attention disturbances associated with every medical and neuropsychological condition associated with impaired attention. We will instead outline the core
components of the development of attention, focusing on particular implications for attention deficit hyperactivity disorder.

This review will consider attentional mechanisms and development from an almost exclusively visual attention perspective. This correlates with the extant literature relating to attention which has focused almost exclusively on visual attentional processes, although theorists speculate that findings should and do generalize to other sensory modalities (Richards, 2003).

The Neuroanatomy of Attention

Contemporary cognitive neuroscience paradigms propose that attention involves a defined neural network that performs highly specific computations (Fuentes, Vivas, & Humphreys, 1999). In the most elaborated of these anatomical models proposed by Posner and Petersen (1990) and Mesulam (1981, 1990), attention is specified in terms of neural networks corresponding to brain regions activated during imaging tasks that require attention, and regions which, when damaged, produce attention deficits.

Mesulam’s model has been influential in so far as it accentuates the anatomical specificity within the neural network, whilst Posner’s model has dominated research conceptualization in this area because it emphasizes the cognitive functions performed by the subcomponents of the network (Webster & Ungerleider, 1998). Importantly, both models identify similar neural substrates for attention and are based on the traditional view of attention as a factor that amplifies signals, thereby enhancing the processing of the target stimuli.

Mesulam’s distributed network for spatial attention

Mesulam (1999) suggests that at the psychological level attention refers to the preferential allocation of the limited processing resources and response channels to events that have become behaviorally relevant; and at the neurological level it is the reversible modulation in the selectivity, intensity, and duration of neuronal responses to such events. His model was informed by convergent findings from neuropsychological and brain imaging studies. Mesulam (1981) proposed that visuospatial attention was coordinated by a large-scale distributed network that includes three monosynthetically interconnected cortical areas. The epicenters of this neural network were the posterior parietal cortex around the intraparietal sulcus, the frontal eye fields in the premotor cortex, and the anterior cingulated cortex. These local networks have extensive interconnections with each other and the reticular activating system that together form the larger neural network for attention.

The parietal cortex

Mesulam identified the sulcus separating the superior and inferior parietal lobes as the primary anatomical region for attention within the parietal cortex. Its influence on
other areas within the attentional circuitry is explained in terms of reciprocal interconnections with the premotor cortex, the frontal eye fields, the superior colliculus and the paralimbic areas allowing for the potential for both top-down and bottom-up control of attention. Converging evidence from neuroimaging studies (Corbetta, Miezin, Shulman, & Petersen, 1993; Gittleman et al., 1999), behavioral analysis in neurological patients with focal cortical lesions (Posner & Dehaene, 1994), and neurophysiological recordings in nonhuman primates (Colby & Goldberg, 1999), implicate inferior and superior parietal regions neighboring the sulcus in the voluntary allocation of visuospatial attention. Experiments in the macaque have suggested that the parietal component of the attentional network is specialized for mapping the spatial location of salient sensory events and also for compiling motor strategies that would target such events for attentional behaviors (Andersen, 1995; Gottlieb, Kusunoki, & Goldberg, 1998; Robinson & Kertzman, 1995; Snyder, Batista, & Andersen, 1998).

The lateral prefrontal cortex/frontal eye fields
The frontal eye fields and surrounding premotor areas, on the other hand, appear to play a more prominent role in the selection, sequencing, and execution of attentional behaviors related to the foveation, scanning, exploring, reaching, and grasping of salient events (i.e., overt motor exploratory aspects of spatial attention; Goldberg & Bushnell, 1981). Tasks requiring shifts of attention without eye movements, as well as tasks requiring focused attention, activate the frontal cortex in the region of the frontal eye fields (Gittleman et al., 1999; Hopfinger, Buonocore, & Mangun, 2000). It is speculated that this region of the brain has a relative specialization for providing a mental map for the distribution of overt and covert exploratory behaviors. The cortical and subcortical inputs to the frontal eye fields and adjacent motor regions direct the eyes, head, and limbs, permitting exploration of the environment.

Cingulate gyrus
The cingulate gyrus is a major component of the “paralimbic belt.” It provides a zone of cytoarchitectonic transition between core limbic areas and front parietal neocortex (Mesulam, 2000). The location of the cingulate gyrus within the limbic system contributes to the coordination of spatial attention by modulating the distribution of expectancy and emotional/motivational valence (Mesulam, 1981, 1990).

The relevance of the cingulate gyrus to spatial attention has been inferred from reports showing that damage to this part of the brain gave rise to contralesional neglect in monkeys and humans (Heilman, Watson, Valenstein, & Damasio, 1983; Watson, Heilman, Cauthen, & King, 1973). Anterior cingulotomy causes persistent impairments of focused and sustained attention. Cognitive research has demonstrated that the interference condition of the Stroop test elicits robust activation in the anterior cingulate region (Pardo, Pardo, Janer, & Raichle, 1990). Anterior, but not posterior, cingulate activation has been reported when subjects had to divide attention (Posner & Petersen, 1990; Taylor, Huang, Tandon, & Koeppe, 1998). The posterior cingulate gyrus, in contrast, monitors postsaccadic shifts in the direction of overt visual targets and promotes
the speed of spatial target detection, especially when attentional shifts are influenced by such reflexive and volitional cue-induced anticipatory biases (Mesulam, Nobre, Kim, & Parrish, 2001).

Evidence from imaging, physiological, and behavioral investigations in nonhuman primates and humans suggests that each of these regions makes a differential contribution to spatial attention (Chedru, Lebranc, & Lhermitte, 1973; Mesulam, 1981, 1985, 1990; Posner, Walker, Friedrich, & Rafal, 1984). Accordingly, Mesulam’s model posits the idea that effective apportionment of attention across the extrapersonal space requires the integrity of all three of these cortical areas, as well as their connections with each other and with specific subcortical regions in the thalamus and striatum. Damage to any one of these three cortical areas causes contralesional neglect (Heilman & van den Abell, 1980; Mesulam, 1981); that is, a loss of salience in the mental representation and conscious perception of contralateral space and a reluctance to direct orienting and exploratory behavior to the region. Lesions in only one component of this network are associated with partial unilateral neglect, whilst those that encompass more than one component result in more profound deficits (Mesulam, 1981). The evidence also indicates that lesions located within the right hemisphere produce more impairment than lesions within the left hemisphere. This led Mesulam to propose that the right hemisphere was more effective in the execution of attentional tasks; it appeared to coordinate attentional deployment across the entire extrapersonal space (spans both hemispheres). In contrast, the left hemisphere primarily contains the neural apparatus influencing contralateral orientation in the right hemisphere (Mesulam, 1981).

Mesulam believed that this model was sufficient to explain all aspects of spatial attention regardless of modality of input or output. However, the exact functions of this network remain partially unspecified, though visual attention studies have suggested that one possible role of such a control network is to generate biases that influence activity in lower-level visual areas (Corbetta, Kincade, Ollinger, McAvoy, & Shulman, 2000).

Posner’s model

The model of attention proposed by Posner and his colleagues incorporates the same brain regions as Mesulam’s, but these are organized into different functional networks.

Posner and Petersen (1990) discuss three hypotheses about attention. First, the attention system of the brain is separate from the data-processing system. Second, attention is carried out by a network of anatomical areas. Third, the areas involved in attention carry out different functions that can be specified in cognitive terms. The whole network serves to enhance information that occurs at a selected location. Posner and Petersen (1990) propose that the sources of attention form a specific system of anatomical areas, which can be further broken down into three interrelated networks (Table 11.1).

Alerting network

The alerting or sustained attention system is responsible for providing an adequate level of arousal. The network is comprised of the mesencephalic reticular formation
Plate 1 Neurulation in human embryos. A, a five-somite embryo at Carnegie stage 10; B, a ten-somite embryo at stage 10; C, another ten-somite embryo, in which the neural tube has closed at the future cervical level. Another initiation site of neural tube closure can be seen at the mesencephalic-rhomencephalic junction (lower arrow), whereas the medial walls of the mesencephalon appear to make contact with each other (upper arrow). Note. Reproduced with permission from “Neural Tube Closure in Human Initiates at Multiple Sites: Evidence from Human Embryos and Implications for the Pathogenesis of Neural Tube Defects,” by T. Nakatsu, C. Uwabe, and K. Shiota, 2000, Anatomica Embryologica (Berlin), 201, 455–466.
Plate 2  Semilobar holoprosencephaly (HPE): sagittal (A), coronal (B), and axial (C) T2-weighted images of a neonate with a dorsal sac (A); in B, the thalami (T) are separated below a single ventricle. Note. Images kindly provided by Ton van der Vliet, Nijmegen.
Plate 3  MRIs of some malformations of the cerebral cortex. A, tuberous sclerosis with subependymal tubers (arrows) in a 4-month-old girl; B, hemimegalencephaly in a 5-year-old girl with neurofibromatosis with a unilaterally enlarged ventricle; C, X-linked bilateral periventricular nodular heterotopia in an 8-year-old girl presenting with seizures (arrows point at various nodules, lining the lateral ventricles);
Plate 3 (continued) D, type 1 lissencephaly (isolated form in a male neonate); E, subcortical band heterotopia in a 4-month-old girl (double cortex between arrows); and F, schizencephaly in a 5-year-old girl: a complete cleft from the pial surface to the lateral ventricle of the left ventricle. Note. MRIs kindly provided by Berit Verbist, Leiden (A, C, F); Willy Renier, Nijmegen (B, D); and Henk Thijsen, Nijmegen (E).
Plate 4 The Rey–Osterrieth figure: model and longitudinal data from a child with RPL on both the copy and memory conditions.

Plate 5 Frontal brain regions mediating emotion regulation processes. Our particular focus is on the V-PFC, which is the hub of ventral regulatory networks, and the ACC, which is the hub of dorsal control networks. The ACC is often co-activated with dorsolateral PFC and other frontal regions in effortful self-strategies.
Plate 6  Antisocial children who showed the highest levels of behavioral flexibility in interactions with parents showed greater frontal scalp activations, associated with self-regulation, in a go/no-go task following a negative mood induction. Here we used patterns of scalp activation to model the cortical generators of the N2 for the seven most flexible and seven least flexible children in our sample. The model shows a source in the ACC region that is active only in the more flexible children.

Plate 7  Brain regions involved in processing socially relevant stimuli. Medial and inferior frontal and superior temporal cortices, along with the amygdala, form a network of brain regions that implement computations relevant to social processes. Note. The image in the left panel was drawn using BrainTutor (www.brainvoyager.com/BrainTutor.html); the images in the right panel were adapted from “Autism: A Window onto the Development of the Social and the Analytic Brain,” by S. Baron-Cohen and M. Belmonte, 2005, Annual Review of Neuroscience, 28, 109–126.
Plate 8 Brain areas involved in typical reading development measured with functional MRI. The images show the early reliance on left posterior superior temporal cortex in beginning readers and the expansive involvement of left parietal, temporal, and frontal cortices in adult readers (top). Correlations between brain activity during reading and reading ability (measured on standardized tests) demonstrate increased temporal and frontal involvement as reading develops (bottom). Right hemisphere activation declines as reading is acquired. Note. Reproduced from “Development of Neural Mechanisms for Reading,” by P. E. Turkeltaub, L. Gareau, D. L. Flowers, T. A. Zeffiro, & G. F. Eden, 2003, Nature Neuroscience, 6(7), 767–73, courtesy of Guinevere Eden, Center for the Study of Learning, Georgetown University, Washington DC, USA.
Plate 9 Capacity-test screen shots from the *Dyscalculia Screener* (Butterworth, 2003) showing keyboard response arrangement, simple reaction-time test where dot appears at random intervals and learner must press a key as quickly as possible, dot counting ("spots") where learner has to judge true or false, and number comparison where learner has to select left or right keys. Note. From *Dyscalculia Screener*, by B. Butterworth, 2003, London: NFER-Nelson.
Table 11.1 Posner and Petersen's Three Attentional Brain Networks and Suggested Functional Operations.

<table>
<thead>
<tr>
<th>Neural system</th>
<th>Functional cognitive mechanism</th>
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<tr>
<td>Vigilance system alerting</td>
<td>Alerting</td>
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<td>Sustained attention</td>
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<td>Posterior attention system</td>
<td>Reflexive orienting</td>
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<td>Early or perceptual selection</td>
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<td>Anterior attention system</td>
<td>Strategic orienting</td>
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<td>Late or cognitive selection</td>
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<td>Conflict detection</td>
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and the amygdala, which are responsible for maintaining a state of alertness. The effect of different senses is weighted with respect to activating properties, and it is supposed that the system acts as a filter allowing the brain to concentrate only on those stimuli that are pertinent to the organism's activity at that particular time (select attention). The sensory stimuli, through multisynaptic chains of neurons, terminate in a group of nuclei located in the thalamus, which then relays the sensory impulses to the cerebral cortex. A descending pathway in the reticular system allows the cortical structures to "subordinate lower structures to the control of programs (axons) arising in the cortex and requiring modification and modulation of the state of waking for their performance" (Luria, 1974, p. 46).

Any disruption in the ascending or descending reticular activating system, or damage to the processes and structures that activate this functional unit, will result in an insufficient state of waking or cortical tone and subsequently will result in an organism that cannot sufficiently interact with its environment. Visual and acoustic stimuli activate the reticular activating system but bilateral damage or diffuse disruption to the reticular formation results in coma (Aston-Jones, Rajkowski, & Cohen, 1999). The patient is still capable of receiving sensory stimuli but there is no conscious processing of this information.

**Posterior attention system**
The neuroanatomy of the posterior or sensory orienting (facilitation of processing at specific locations in space), selective (filtering of information once a location is attended) attention network has been suggested by lesion studies (Posner, 1988; Sapir, Soroker, Berger, & Henik, 1999), evidence from neurophysiological research in animals, and from functional neuroimaging and electrophysiological techniques in man (Corbetta et al., 2000; Posner, Petersen, Fox, & Raichle, 1988). The results consistently implicate the superior parietal lobes, the midbrain, more specifically the lateral pulvinar nucleus, and the lateral posterior nucleus of the thalamus.
Sensory impulses travel from the reticular activating system through a distributed network that includes modalities within the posterior attentional network. The posterior attention system receives dense noradrenergic innervation from the locus coeruleus (Bush et al., 1999). Noradrenalin functions to inhibit the spontaneous discharge of neurons, resulting in the enhancement of the signal-to-noise ratio of target cells, thus priming the posterior system to orient to and engage novel stimuli (Faraone & Biederman, 1998).

**Anterior attention system**

The anterior system is endowed with the responsibility for executive control of attention. This conceptualization is supported by neuroimaging studies that reveal increased activity in prefrontal and cingulate areas when there is an executive or decision-making component to attentional control, such as when attention must be divided across a number of different stimulus attributes (e.g., Corbetta, Miezin, Dobmeyer, Shulman, & Petersen, 1991).

The anterior network incorporates the frontal lobes (dorsolateral and ventromedial prefrontal cortex), anterior cingulated, and basal ganglia, and is activated when processing and/or responding requires cognitive control; for example, during tasks when conflict is present (requires suppression of a competing response choice or suppression of attention to a salient stimulus) and when the production of a nonhabitual (novel) response is necessary (Rothbart & Posner, 2001), that is, executive attention requires the mobilization of inhibitory attentional processes which are elements of “cognitive control” (Casey, 2001; Casey, Giedd, & Thomas, 2000).

Research suggests that the anterior attentional network exerts control by excitatory and inhibitory processes that are cooperatively engaged to enable the perceptual segregation of relevant and irrelevant information (Desimone, Wessinger, Thomas, & Schneider, 1990). Stuss and colleagues (Stuss, Shallice, Alexander, & Picton, 1995) postulate that this control is reflected in several types of attentional processes, including sustained attention, cognitive flexibility, inhibition, and divided attention. The purpose of this inhibitory pathway is to keep the system clamped down by inhibition to prevent stimulus-bound eye movements.

**The priority network**

Attention has been viewed as a mechanism for biasing the competitive interactions among mutually inhibitory sensory representations in cortex so that an attended object “wins” the competition and thereby comes to enter awareness. Attended objects thus produce more robust cortical activity than unattended objects. The attentional system is thought to have at least two subsystems that give rise to the attentional biasing signal that initiates the sensory enhancement of the selected stimulus. One is a bottom-up system involved in automatic and rapid shifts of “exogenous” (involuntary and stimulus driven, “attentional capture”) attention to abrupt onsets that are sufficiently salient to divert attention from the current focus (Irwin, Colcombe, Kramer, & Hahn, 2000;
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Jonides & Yantis, 1988; Remington, Johnson, & Yantis, 1992). The other is a top-down subsystem involved in volitional shifts of “endogenous” attention (goal directed and voluntary). The bottom-up subsystem is thought to involve circuitry in the superior colliculus (SC), and the top-down subsystem is thought to involve circuitry in the frontal lobe (Mesulam, 1981; Posner & Petersen, 1990). Both goal-directed and stimulus-driven mechanisms have ecological significance such that, under certain circumstances, attention in one or the other fashion has adaptive value. When processing demands are higher for the anterior task, the anterior attention network will modulate the action of the posterior attention network (Posner, Inhoff, Friedrich, & Cohen, 1987); the opposite occurs when detection of target location (i.e., the posterior task) has priority (Fuentes, 2004).

Normal Neuropsychological Development of Attention

The human attention system is immature at birth both anatomically and functionally, and goes through substantial changes, especially during the first few months of postnatal life. Development occurs in a hierarchical order. The critical period seems to vary in onset and duration between different brain regions. Subcortical regions mature earlier than cortical areas, and different functions emerge at different times and develop at different rates with an extended time period for executive attention underpinned by frontal lobes. The different developmental trajectories of the two attention systems potentially serve as the basis for marked individual difference in the rate and profile of attention development.

The differential time course of development of different cortical regions has been studied using positron emission tomography (PET; Chugani, Phelps, & Mazziotta, 1987). In infants under 5 weeks of age, glucose uptake is highest in sensorimotor cortex, thalamus, brain stem, and the cerebellar vermis, whereas by 3 months of age, there are considerable rises in activity in the parietal, temporal, and occipital cortices, basal ganglia, and cerebellar cortex. Maturational rises are not found in the frontal and dorsolateral occipital cortex until approximately 6–8 months of age, and the distribution of resting activity within and across brain regions does not approach adult levels until the end of the first year. As different subcortical and cortical modules are in differential states of maturation early in life, it seems likely that different properties of the attentional system will have primacy during these developmental transitions. Assuming that the same neural areas in adults are responsible for attentional control in infants, this maturational progression predicts attentional primacy that proceeds from the alerting network to the posterior network before ascendency to the anterior network between 6–12 months.

Ruff and Rothbart (1996) hypothesized that the posterior and anterior attention systems could be developmentally conceptualized as (a) an early system in which attention is highly influenced by novelty of objects and events, and (b) a later one in which self-generated and goal-oriented schemes and tasks are a major incentive for sustained,
focused attention. An important aspect of the transition should occur around 12 months as children begin to habituate more readily to novel objects and events, making novelty a less potent determinant of attention. Thus, the duration of attention based on the first system should decline from 12 months onward. However, the second, higher-level system of attention develops slowly with increasing cognitive sophistication and improved self-regulatory skills. As a result, attention may not be strongly governed by the second system until 6 or 7 years of age.

Colombo's model: development of alerting/arousal

This neural hierarchy is reflected in the conceptual framework for the development of visual attention in infancy advanced by Colombo (2001). This model, informed by recent advances in the research on visual attention in infancy, considers that the development of the alert state is the most fundamental attentional process in the neonate. The progression in the neonate from a sleep–wake cycle characterized by six distinct states, only two of which are in the alert phase, to being able to attain more extended periods of alertness (Berg & Berg, 1979) is seen as a prerequisite for other mental activity (Luria, 1974) including attention. Dahl (1996) describes sleep and arousal as the polar extremes of a single continuum where sleep is "a categorical diminution of awareness and responsiveness to the environment." In contrast, alerting is defined as achieving and maintaining a state of high sensitivity to incoming stimuli and is conceived as the initial component in the attention process. Colombo's (2001) ideas of the primacy of the alerting mechanism in the neonatal period concurs with the developmental imaging findings above that indicate greatest cortical activity within Posner and Petersen's (1990) alerting attentional system (the brain stem and thalamus) in infants under 5 weeks of age. This arousal/sustained alertness system permits attention and maintenance of an alert and vigilant state during periods of wakefulness in the neonate.

The reticular activating system's general arousal mechanism has an ascending influence on cortical areas that produces enhanced processing and arousal and a descending influence on heart rate through a parasympathetic outflow that decreases heart rate in infants and children, or decreases heart rate variability in adults. Research using neonates as subjects have used extended heart-rate slowing as an index of a state of general arousal in the brain. Investigators have found that stimuli that evoke heart-rate decreases in awake newborns have produced the reverse effects, heart-rate accelerations, during sleep (Clifton & Nelson, 1976; Pomerleau & Malcuit, 1981; Pomerleau-Malcuit & Clifton, 1973). Only in the awake state can newborns exhibit sustained decreases in heart rate (Graham, Anthony, & Zeigler, 1983). Further, stimulus complexity and intensity appear to interact in determining the direction of heart-rate responses in newborns. Simple auditory stimuli presented at 75–80 dB elicit heart-rate decelerations, while complex auditory stimuli, which require greater neural analysis, of equivalent intensity elicit heart-rate accelerations (Clarkson & Berg, 1983; Fox, 1979). The implication is that the neural substrate associated with complex analysis of the stimulus is insufficiently developed during this neonatal period. Also, heart-rate responses
have been found to change with age. Infants are more likely to display heart-rate decelerations following stimulus exposure with increasing age across the first year of postnatal development which has been interpreted as reflecting the developing maturity of the attention system.

Development of the posterior attention system and orienting

In infancy, attention is typically parsed into two broad components: stimulus orienting and sustained attention (Cohen, 1973; Graham & Clifton, 1966; Ruff & Rothbart, 1996). Stimulus orienting occurs when individuals detect change or become aware of stimuli in their environment (also known as the orienting response, Sokolov, 1963, or “attention-getting,” Cohen, 1973; Sokolov & Cacioppo, 1997). During this process, information on the importance, or the novelty, of the stimulus is encoded and a decision is made to maintain attention or disengage from the stimulus (Neisser, 1967; Sokolov, 1963).

The biophysiological data suggest that subcortical activity, including those areas that subserve the orienting or posterior attention circuitry, is high during early infancy. According to Posner’s model, the act of shifting attention is achieved by a sequence of partially overlapping although separate operations. First, the target stimulus produces a general alerting state that interrupts ongoing activity; this is not spatially selective but works to potentiate other targets. The coordinates of the target stimulus are generated by the orienting system prior to the shift of attention, and orienting is achieved first by disengaging from the current task, next moving the “spotlight,” and then engaging attention to the new location. Consequently, the allocation of visual attention or orienting is closely linked to the generation of saccadic eye movements (Corbetta et al., 1998; Kustov & Robinson, 1996) and the development of its neural substrates.

Correspondingly, the control of the eye has been used as a model system for studying the development of visual attention beyond the most basic arousal functioning. Over the first few weeks of life, most saccades generated by infants appear to be automatic, and are triggered in response to exogenous factors. The sudden appearance of the stimulus, which heightens its saliency, leads to the prepotency of the reflexive saccade. The short latency reflex saccade pathway is primarily subcortical and mature at birth (Schiller, 1985) and derives from a fast, reflexive pathway that initiates saccades to novel stimuli even when such saccades are inappropriate. These saccades are mediated by the parietal lobes which underlie the disengage function to facilitate engagement to the new coordinate or object.

Developmental changes consistent with neural maturational predictions in fixation and disengagement in infants have been examined in a number of studies. Visual behavior in 1–2-month-olds is characterized by a phenomenon called “obligatory attention” (Stechler & Latz, 1966) or “sticky fixation” (Hood, 1995); that is, during this period neonates have difficulty disengaging fixation from a central stimulus to orient to a new stimulus. Some have attributed this to the onset of competition between cortical and subcortical visuomotor pathways (Johnson, 1990). Other authors have compared
obligatory attention with Balint’s syndrome in adults (Hood, 1995). Patients with Balint’s syndrome have acquired bilateral parietal cortex damage and experience similar difficulties in “disengaging” from one stimulus to another, indicating that immaturity of the parietal cortex in the infant has similar behavioral consequences. By 2–3 months of age, these markers of subcortical control are replaced by behavioral advances, such as acquiring the ability to disengage easily from one stimulus to orient to another, consistent with the maturation of regions of the parietal cortex and associated structures. Johnson and colleagues (Johnson, Dziurawiec, Ellis, & Morton, 1991) found that 4-month-olds showed a much greater propensity than 2- or 3-month-olds to disengage fixation from a central target in order to orient to a peripheral target. Similarly, the ability to make anticipatory saccades during a regular visual stimulus sequence has been demonstrated in 3-month-olds (Haith, Hazan, & Goldsmith, 1988) but not 2-month-olds (Canfield & Haith, 1991). Researchers have concluded that this pattern reflects the developmental improvements in disengagement resulting from brain maturation.

Johnson’s model (1990, 1995), a synthesis of research on infant visual attention, oculomotor control, and neuroanatomical maturation, posits that developments in the layers of the primary visual cortex act as a limiting factor for visual attention controlled by neural systems. Influenced by Conel’s (1939–1967) studies of cortical maturation in human infants, Johnson (1990) noted that cells in the early maturing layers 5 and 6 of primary visual cortex project only to subcortical structures, including the superior colliculus and the basal ganglia. Conel’s research indicated that although cells in layers 2 and 3 (the major origin of associative and collosal fibers in the prefrontal cortex) eventually project to the prefrontal cortex, they would be insufficiently mature to contribute to information processing until infants are about 4–6 months of age. This led Johnson (1990; Johnson, Posner, & Rothbart, 1994) to conclude that frontally mediated oculomotor control, and therefore endogenous attention (subject directed), would not appear before this period, and attention control before the 4–6-month period must be under subcortical control. Subcortical control would simply allow visual stimuli to be detected, localized, and bought to the fovea but is controlled by exogenous (directed to salient characteristics of the environment) events and strongly governed by novelty.

Between 2 and 4 months of age, selectivity is influenced by the previous experience of the infant, and orienting is most associated with novelty. Around 9 months of age, there is evidence of a reduction in the orienting response to novel visual stimuli. It has been suggested that this decrease in orienting response may be important for the development of directed attention as it may help reduce distraction by irrelevant stimuli (Ruff & Rothbart, 1996).

Development of the anterior attention system: sustained attention, cognitive flexibility, and inhibitory control

Maturational changes in the neurophysiological organization of visual control between 3 and 6 months of age allow for voluntary shifting of attention (late selection) versus obligatory attention (Rothbart, Posner, & Boylan, 1990). Evidence from the work of
Rothbart and colleagues with infants indicates that 4-month-olds are capable of disengaging attention much more easily than younger infants, suggesting a developmental shift in attentional capacities at this age (Johnson, Posner, & Rothbart, 1991). This stage of development brings attention under volitional control and coincides with the maturation of the frontal lobes. The anterior system is responsible for saccades that are voluntary or planned. Correspondingly, developments at 4–6 months old, such as gaining the ability to inhibit reflexive saccades (Johnson, 1995) in order to sustain attention to a target stimulus and to make saccades in anticipation of a visual target being shown in a particular location (Csibra, Tucker, & Johnson, 2001; Gilmore & Johnson, 1995), have been associated with developments in the frontal cortex. Research evidence supports the proposition that these endogenous (internally directed) functions are mediated by frontal areas. Directed or voluntary attentional saccades are associated with the cingulate cortex in the medial frontal area (Posner, 1995; Posner & Petersen, 1990), and the maintenance of attention and inhibition of shifting have been linked with the frontal eye fields.

Whilst most theorists agree that there is a shift from predominantly subcortical to cortical control over saccades in early infancy, data collected in the 1990s have challenged the traditional views on the timing of the maturational status of the prefrontal cortex. Research utilizing intrastimulus shifting, associated with active and purposeful comparison of paired stimuli (therefore under endogenous control), reports that this ability is four times more likely to be observed in 4-month-old than 3-month-old infants (Colombo, Mitchell, Coldren, & Freeseman, 1991; Frick, Colombo, & Allen, 2000), whereas shifts in attention (cognitive flexibility) was more reliable in 7-month-olds than 4-month-olds across a one-week test/retest period suggestive of a critical period for endogenous control between 4 and 7 months.

Other researchers using different methodologies have reported endogenous attention abilities in much younger infants. Behavioral studies using the visual expectation paradigm (Canfield & Haith, 1991; Haith, Wentworth, & Canfield, 1993) and event-related potential data (Snyder, Batista, & Anderson, 2000; Wentworth & Haith, 1992) have reported evidence of predictive saccades (i.e., planned and voluntary interocular movements) in 3 to 3½-month-old infants. Also, sustained attention, which is dependent on the infant’s ability to inhibit attentional shift (suppression of saccadic eye movement/antisaccade), has been demonstrated in infants as young as 8 weeks (Richards, 1989a). There is, however, considerable evidence that there are significant changes in the amount, depth, and frequency of sustained attention from this age compared to 3- and even 6-month-olds (Richards, 1989b; Richards & Casey, 1992). Richards found that maintained heart-rate decelerations, characteristic of sustained attention, occurred for longer durations in older infants than in younger infants. Furthermore, changes in sustained attention continue through to the latter half of the first year (Lansink, Mintz, & Richards, 2000; Ruff, Capozzoli, Saltarelli, & Dubiner, 1992), and well into the infant’s second and third years (Ruff & Lawson, 1990). Taken together, these results suggest that the contributions of prefrontal cortex to endogenous attention control may emerge early in a rudimentary form but become increasingly differentiated and more efficient as the frontal lobes mature.
The period from 18 months through mid-adolescence is accompanied by an extended development of executive attention, but seems to undergo a particularly rapid development between 2 and 7 years of age. It has been traditionally believed that cognitive development during late childhood and adolescence is subserved primarily by the relatively late incorporation of the prefrontal cortex either by its intrinsic late structural maturation (Bourgeois, 1993; Sowell, Thompson, Holmes, Jernigan, & Toga, 1999) or by the maturation of other neocortical regions (Chugani, 1998; Rakic, 1995) that influence their functional integration with prefrontal cortex (Thatcher, Walker, & Giudice, 1987). Functional magnetic resonance imaging and positron emission tomography data demonstrate prolonged development in both the visual cortex and the frontal lobes which involve structural changes and maturation even in adulthood (Chugani et al., 1987; Sowell et al., 1999).

One aspect of executive attention is the ability to allocate resources in a way that is consistent with self-established goals and plans. The anterior circuitry enhances attention to the particular aspects of the environment that are consistent with these, and inhibits attention to other stimuli. Voluntary response suppression/inhibitory control and the ability to filter out distracters are both crucial for choosing a course of action based on a cognitive plan/goal-directed behavior over alternative task-irrelevant behaviors that hamper adaptive functioning (Bjorklund & Harnishfeger, 1995; Dempster, 1992). Voluntary response suppression has been found to develop throughout childhood (Fischer, Biscaldi, & Gezek, 1997; Luciana & Nelson, 1998; Munoz, Broughton, Goldring, & Armstrong, 1998) and adolescence (Ridderinkhof, Blanch, & Logan, 1999; Ridderinkhof, van der Molen, Band, & Bashore, 1997). The antisaccade task (Hallett, 1978) has been used successfully to characterize development of the ability to voluntarily suppress prepotent responses. The brain systems subserving performance on the antisaccade task have been well delineated in monkeys (Burman & Bruce, 1997) and adult humans (Doricchi et al., 1997; O’Driscoll et al., 1995). These areas include the frontal eye fields, supplementary eye fields, dorsolateral prefrontal cortex, posterior parietal cortex, anterior cingulate cortex, basal ganglia, thalamus, and superior colliculus.

Researchers (Casey, Castellanos, & Giedd, 1997; Casey, Trainor, et al., 1997; Luna et al., 2001) report both significant overlap and qualitative differences in the pattern and extent of brain activation across age groups, paralleling the enhanced voluntary control of behavior throughout adolescence. However, the presence of robust activation in several cortical regions in children indicates that these regions already participate in voluntary response suppression in childhood. Using other measures of response inhibition (go/no-go, stop tasks), researchers have also reported age-related increases in activation in frontal lobes, with children showing significantly greater intensity of activation than adults. Durston et al. (2002) showed that normally developing children aged 7 to 12 years activated regions in the frontal cortex more than adults and that activation was correlated with age. Schachar and Logan (1990) examined differences in both children and adults on another inhibition task (stop task). Findings indicated that normal inhibition is well developed early on, with children evidencing a similar rate of errors as adults
by the second grade. However, younger children had more variability in their responding rates and slower response times overall than did either older children or adults.

Theorists have concluded that this pattern is due to children being less efficient at generating an inhibitory signal because of the immaturity of their frontal lobes. Researchers suggest that although the capacity to suppress a response is present early (all children could perform a correct antisaccade on at least one trial), as has been seen in infancy (Johnson, 1995), efficiency (the ability to perform correct antisaccades consistently) continues to improve into adolescence. Similarly, studies using dual task paradigms indicate that top-down control over attention is still maturing during childhood and adolescence, even though the process of allocating attention is adult-like quite early in childhood (Atkinson, Hood, Wattam-Bell, & Braddock, 1992; Karatekin, Granholm, & Steinhauser, 2004). These data are consistent with the proposition that relatively automatic attentional processes develop earlier and complete their development at a much younger age than anterior or cognitively demanding attentional processes, commensurate with hypotheses that neural development proceeds along a posterior-to-anterior gradient.

**Attention Psychopathology Associated with Attention Deficit Hyperactivity Disorder**

Attention deficit hyperactivity disorder is a neurodevelopmental condition characterized by persistent age-inappropriate symptoms of attention, impulsivity, and hyperactivity. Behavior that suggests difficulties with attention, difficulty sustaining attention in tasks, being easily distracted by extraneous stimuli, and often having difficulty organizing tasks and activities (DSM-IV criteria) are central components of attention deficit hyperactivity disorder (ADHD); however, the neurological basis of these behaviors has not been conclusively demonstrated. Consensus exists regarding the behavioral phenotype of the disorder, the basis of diagnosis. The cognitive basis of this, however, has resisted attempts to analyze it into a single fundamental problem. Rather, there are moderate associations with several types of cognitive change.

**Neuroanatomical correlates of ADHD**

The neuroanatomical correlates of attention deficit hyperactivity disorder, the prefrontal corticostriatal pallidal pathways, approximately parallel Posner's anterior/executive attention network. Specifically, volumetric reductions in total cerebral volume (Castellanos, et al., 2001; Castellanos, Giedd, & Marsh, 1996; Castellanos, Lee, & Sharp, 2002; Filipek, Semrud-Clikeman, Steingrad, Kennedy, & Biederman, 1997), cerebellum (Castellanos et al., 2001, 2002), prefrontal cortex (Castellanos et al., 1996; Filipek et al., 1997; Hynd, Semrud-Clikeman, Lorys, Novey, & Eliopoulos, 1990), striatal structures—caudate and pallidum (Aylward et al., 1996, Castellanos et al., 1996, 2002), and the splenium of the corpus callosum (Hynd, Lorys et al., 1991; Hynd, Semrud-Clikeman,
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Lorys, Novey, & Eliopoulos, 1991) have consistently implicated the right prefrontal corticostriatal-pallidal in the pathophysiology of ADHD (Barkley, 1998; Giedd, Blumenthal, Molloy, & Castellanos, 2001). Atypical structural volumes are assumed to relate to atypical function; however, the role of the deficits in brain morphology associated with ADHD remains unclear. Studies of cognitive function in ADHD have focused on different aspects of attentional processing in affected children.

Alerting in ADHD

There exists a plethora of methodologies for assessing alerting in children with ADHD, most of them relating to reaction time or physiological response in the first seconds after encountering a stimulus. Two different kinds of performance problem are consistent with an alerting deficit: slow and variable responses to fast tasks, and hasty, inaccurate responses to slow, careful tasks. Both kinds of response pattern have been described in children with ADHD (see Oosterlan, Logan, & Sergeant, 1998, for a meta-analysis; Sergeant & Scholten, 1985, for review; Leth-Steensen, Elbaz, & Douglas, 2000; Swanson et al., 2000, for randomized controlled trial). However, Huang-Pollock and Nigg's (2003) review of orienting studies did not find a dependable slow and variable response pattern even though most of the individual studies reported this effect. Further, EEG slow wave findings and early evoked response potential (ERP) are consistent with central nervous system (CNS) hypoarousal in a percentage of children with ADHD-C as well as ADHD-I (for a review of EEG, see Barry, Clarke, & Johnstone, 2003a; for ERP, Barry, Clarke, & Johnstone, 2003b).

Sustained attention in ADHD

The ability to maintain a state of alertness and wakefulness during “prolonged mental activity” (Weinberg & Harper, 1993) or sustained attention has been conceived as a central impairment in ADHD. Performance decrement (slower and more variable responses, more errors) over time is universal (Parasuraman, 1998); therefore, ADHD-related deficits in sustained attention should result in an excess decline over time versus controls, yielding a group-by-time interaction. However, the research does not support this notion. Whilst it is true that children with ADHD appear to lose interest in tasks quickly, they do not show a decline in accuracy (or in speed) over time on laboratory tasks (Sergeant, Oosterlaan, & van der Meere, 1999; Sergeant & Sholten, 1985; Sergeant & van der Meere, 1990). Typically, deficits in performance are evident from the very beginning of a task or response (Sergeant & van der Meere, 1990). This pattern suggests a problem in alerting or arousal, not sustained attention per se.

Posterior attention system in ADHD

In terms of Posner's attention network model, abnormalities of the neuroanatomical substrate of ADHD should implicate executive but not primary orienting and select attention which are under the control of the posterior attention system.
Attention Development

Orienting
Huang-Pollock and Nigg (2003) conducted a meta-analysis of the extant literature to determine the degree to which attention deficit hyperactivity disorder is associated with impaired attention. They reviewed 14 studies of visuospatial orienting in children with ADHD, as measured using Posner’s (1980) Covert Orienting of Visuospatial Attention Task (COVAT). In the qualitative analysis reported by Huang-Pollock and colleagues (Huang-Pollock, Nigg, & Carr, 2005), group differences were found between children with ADHD and controls in each of the four endogenously cued studies (Carter, Krener, Chaderjian, Nortcott, & Wolfe, 1995; McDonald, Bennett, Chambers, & Castiello, 1999; Pearson, Yaffee, Loveland, & Norton, 1995; Tomporowski, Tinsley, & Hager, 1994) analyzed with some suggestion of right hemisphere anterior deficit but no replicated specific deficits were found. Huang-Pollock and her colleagues (2005) report that, despite intriguing single-study findings of weaknesses in the posterior disengage function in ADHD-C, meta-analytic effect sizes across studies have been small to nonexistent.

Select attention
Similarly, research does not support difficulties with perceptual select attention in ADHD (Berman et al., 1999; Sergeant & van der Meere, 1988). Nigg and colleagues (Nigg, Hinshaw, & Huang-Pollock, 2006) and Douglas (1999) have raised concerns regarding the validity of the conclusions that can be drawn from these investigations. The most significant reservation relates to the lack of systematic control of perceptual load; the omission is particularly critical given the contemporary theories of load and selection (Lavie, 1995). However, when perceptual load has been controlled more systematically using contemporary paradigms, no evidence of perceptual selection problems was evident in ADHD (with respect to combined type; Huang-Pollock et al., 2005).

There is a dearth of evidence concerning the ADHD predominantly inattentive subtype (ADHD-I) and attention mechanisms, despite the putative suggestion that under-focused selection in the posterior system might be implicated in the manifestation of this subtype. However, the limited data set on exogenous visual-spatial orienting in ADHD-I has not revealed impaired performance (Huang-Pollock & Nigg, 2003; Huang-Pollock et al., 2005), which would appear to negate dysfunctions within the posterior attention system. However, studies are so few that it might be premature at this stage to accept this conclusion.

Anterior attention system in ADHD
Neuropsychological research provides evidence of modest anterior attention dysfunction, although it is unclear whether these deficits are as robust or as large as might be needed for necessary and sufficient core deficit in ADHD.

Interference control (“late selection”)
An inhibition deficit, including poor interference control (producing late selection), has been implicated as one of the core deficits in ADHD (Barkley, 1997a, 1997b).
Interference control is usually assessed using the Stroop Color–Word Interference Test and numerical Stroop test. An influential rationale for the increased Stroop effect in ADHD is a deficit in attentional modulation of the distributed parallel pathways processing, for example, word and color stimuli (Cohen, Dunbar, & McClelland, 1990; Peterson et al., 1999).

Qualitative (Nigg, 2001) and quantitative (Homack & Riccio, 2004; Van Mourik et al., 2005) reviews have concluded that Stroop interference is at most minimally impaired in ADHD-C in childhood. In the literature, studies employing the Stroop test have been considered as providing evidence for poor interference control in ADHD. However, most studies that report poor Stroop task performance have reported performance on the interference condition without controlling for performance on the color-naming condition. A recent meta-analysis shows that when noninterference aspects of the task are taken into account, children with ADHD do not demonstrate poor interference control on this task (Van Mourik et al., 2005). Van Mourik et al. (2005) concluded that the Stroop color test, in standard form, does not provide strong evidence for a deficit in interference control in ADHD. However, the Stroop color test may not be a valid measure of interference control in ADHD and alternative methodologies may be needed to test this aspect of the inhibitory deficit model in ADHD.

Set shifting
Classical measures of set shifting include trail making, Wisconsin Card Sorting Test (WCST), and Creature Counting (Test of Everyday Attention for Children). Computerized models include the switch task (Maudsley Attention and Response Suppression battery) and the Cambridge Neuropsychological Test Automated Battery (CANTAB). Empirical studies indicate evidence of rapid, controlled shifting deficits (Cepeda, Cepeda, & Kramer, 2000; Hollingsworth, McAuliffe, & Knowlton, 2001; Oosterlaan & Sergeant, 1998; Perchet, Revol, Fournieret, Mauguire, & Garcia-Larrea, 2001; Rubia & Smith, 2001; Schachar, Tannock, Marriott, & Logan, 1995), but studies to date are too few and varying in method to allow firm conclusions about the magnitude of the effect (Nigg et al., 2006).

These results suggest that the early maturing posterior attention network is intact in children with ADHD, whereas the later maturing executive or anterior attentional processes are implicated in its manifestation. However, its contribution is at best modest, representing perhaps secondary rather than primary etiological dysfunction. Meanwhile, arousal and related operations of the moderating noradrenergic vigilance network may well offer a promising candidate for a core within-child causal mechanism.

ADHD Case Study

Background

X is an 8-year-old, right-handed boy diagnosed with functional difficulties with attention, impulse control, and hyperactivity at home and at school. X’s overactivity was first
noticed when he was 2½ years old and had begun nursery school. At that time, there were no concerns about impulsivity or inattention. When he transferred to his reception class it was noted that he lacked motivation for academic activities. His inattentiveness and impulsive behaviors became more apparent in year 1, leading to his receiving an Individual Education Plan as these externalizing behaviors were interfering with his learning. Early infant milestones were unremarkable and there was no consanguinity and no other history of developmental disorders or learning disability within the family.

Hypothesis

Informed by the dominant neuropsychological models of ADHD, it was hypothesized that X would have:

1. Central impairment with behavioral inhibition.
2. Secondary impairment in working memory, planning, and attention.
3. Specific impairments in achievement tasks secondary to executive difficulties.

Behavioral observations

X’s behavior during the assessment was certainly consistent with the primary behavioral characteristics of ADHD. Problems with attention were evident in his difficulty in sustaining attention to activities, careless errors, reluctance to engage in tasks that required sustained mental effort, and internal distractibility. He was restless and fidgety, especially in situations of low stimulation, and there was evidence of poor impulse control, especially anticipating failure before he had tried to solve a problem.

Measures

Together with tests of general cognitive ability (Wechsler Intelligence Scale for Children, fourth edition, UK [WISC IV(k)]) and academic attainment (Wechsler Individual Achievement Test II), the Test of Everyday Attention for Children (TEACH) and the Maudsley Attention and Response Suppression (MARS) battery were used to provide a profile of attentional capacity. The MARS battery is a computer-based research neuropsychological battery that provides measures of inhibitory and attentional control that are collectively considered to be components of executive functioning. An additional task looks at time perception.

Results

X’s current general cognitive ability, as estimated by the WISC IV(k) Full Scale IQ, fell within the average range. He obtained FSIQ of 92 (confidence interval = 87–98). The indices were internally consistent, meaning that the Full Scale IQ provided a meaningful composite of his general intellectual ability.
Attention
X completed all nine subtests from the Test of Everyday Attention for Children (TEACH), which provides separate measures of selective (focused) attention, sustained attention, inhibition, and attentional control/switching. His scores ranged from the exceptionally low range to the high average range (from percentiles <0.2 to 79.8–87.7).

Selective attention: The Sky Search and Map Mission subtests measure selective attention. X’s scores for the number of targets correctly identified and time-per-target were average for his age (percentile band 43.4–56.6). The time-per-target scaled score provides a measure of strategy efficiency indicating that X’s use of a systematic left-right and top-down search strategy helped him find an above-average number of targets in a time that was appropriate for his age. When graphomotor speed is taken out of the equation to provide a purer measure of selective attention, X’s attention score on Sky Search placed him in the high average range (percentile band 79.8–87.8). His performance on the analogous test of visual select attention Map Mission was average to high average (69.2–79.8) and commensurate with his performance on Sky Search.

Sustained attention: The Score, Score DT, Sky Search DT, and Code Transmission subtests measure sustained attention, which is the capacity to self-maintain an actively attentive stance to a task, goal, or one’s own behavior despite there being little inherent stimulation for such continued processing. The Sky Search DT subtest is a dual task that involves the capacity to sustain attention on two tasks simultaneously and monitor one’s allocation of attention to each task. Performance on this test is compared with performance on the earlier, simpler Sky Search subtest to determine the extent of any decrement in performance brought about by increasing the complexity of the task. X’s dual task decrement score fell within the exceptionally low range (percentile band <0.2), indicating significantly greater decrement on this task when compared with others of his age. X found only 12 of the 20 visual targets and was successful in only one of the three auditory targets attempted, indicating difficulties on both components of the task.

X’s score in the low ability range (percentile band 6.7–12.2) on the Score subtests indicates relative difficulties in actively maintaining attention to auditory stimuli/sound when this involves waiting for long periods before anything happens. He was similarly impaired on the Code Transmission subtest which, whilst deliberately monotonous, provides a consistently greater level of auditory stimulation than the Score subtest but is of considerably longer duration. His performance on the Score DT subtest, which incorporates a much more interesting auditory task to compete for the child’s attention, was commensurate with his other sustained attention scores (percentile band 12.2–20.2). Task analysis indicated that X had a tendency to attend to the more interesting task. These results suggest that X’s ability to maintain his attention to task will be greatly influenced by its ability to capture his attention.

Behavioral inhibition: The Walk/Don’t Walk subtest provides a measure of sustained attention to one’s own actions and intentions. To succeed on this task, one must actively resist an automatic, routine form of responding to facilitate a competing though less frequent response. X scored within the impaired range on this task (percentile band
<0.2), suggesting significant difficulty in inhibiting automatic responding compared with other young people of the same age. X found this activity particularly difficult and admitted to not putting any mental effort into the last five trials even though he agreed that it was a good task.

Attentional control/cognitive flexibility: The Creature Counting and Opposite Worlds subtests provide measures of X’s ability to control his attention. X performed within the high average range for accuracy and was average on the timing element on the Creature Counting subtest, which was consistent with his performance within the average range on the Same World/Opposite World subtest.

Executive function
The Switch task of the MARS quantifies cognitive flexibility. When successive responses are governed by the same rule, X’s response rate was indistinguishable from controls. However, his rate of responding was more variable than controls. His omission percentage indicates that this was not due to poor attention. His much larger switch effect, despite his percentage switch success, indicates significantly greater need for more processing time than controls to produce similar success.

The Stop task consists of two elements. Reaction time to go can be considered to be analogous to a behavioral activation system. X’s responses are indistinguishable from controls, but his rate of responding, comparable to his performance on the Switch task, was more variable than controls. His much larger stop signal reaction time suggests difficulty with behavioral inhibition in that he needs longer than controls to successfully inhibit a preprogrammed/prepotent response.

Time perception
The timing task indicates that X has more problems with time perception than age-matched controls.

Conclusion
X is a right-handed, 8-year-old boy whose performance on assessment suggested specific impairment in some executive functions in the context of average intellectual functioning. X is developing along an age-appropriate trajectory with regards to his select attention and cognitive flexibility, but the cognitive control required for the latter was more variable than non-ADHD controls. He has relative difficulties with sustained attention but was most impaired on the tests of behavioral control and time perception.

Conclusion
Although we have focused on ADHD, it is worth noting that problems with inattention are implicated in a number of psychiatric and clinical conditions. Mirsky and Duncan
(2001) listed some 17 conditions associated with disordered attention, including schizophrenia, autism, fetal alcohol syndrome, and closed-head injury. To this list could be added other medical conditions and psychopathology: Tourette syndrome, anxiety disorders, depression, and obsessive compulsive disorder. Attention is a multidimensional process and not all such functions are equally affected across these various conditions. Thus, behavioral problems that appear as "inattention" may or may not be related to dysfunction in attentional mechanisms defined more formally. A range of causal mechanisms within the child could, in fact, be related to inattentive and otherwise dysregulated behavior (Nigg et al., 2006).

Furthermore, the cognitive changes in ADHD are by no means exhausted by the attentional changes considered here. Case-control studies have found alterations in accuracy of time perception (Smith, Taylor, Rogers, Newman, & Rubia, 2002), visuospatial memory (Rhodes, Coghill, & Mathews, 2005), delay discounting of reward (Sagvolden, Johansen, Aase, & Russell, 2005), delay aversion (Sonuga-Barke, 2005), and several tests of planning and foresight (Nigg et al., 2006). Each test considered singly shows a rather modest effect size (typically around 0.5 SD) for the discrimination. Combinations of tests, however, show much better discriminations, with specificity and sensitivity rising toward 80% (Solanto, Arnsten, & Castellanos, 2001). Individual children may show a large impairment on one test. Indeed, in clinical practice it is common to find that individual children show big discrepancies on tests such as speed of information processing or short-term memory. The implication is that cognitive testing does not (yet) make the diagnosis of ADHD, but may be very useful in delineating the strengths and weaknesses of an individual with a view to understanding the condition and advising educators.

The recent advances in the understanding of how attention develops have been striking and are likely to continue. The revolution of functional imaging is allowing the localization and study of progressively more subtle aspects. The mechanisms involved in the motivational and emotional control of attention will be particularly relevant for understanding psychopathology.

Several obstacles, however, still need to be overcome. At the level of basic understanding, the relatively weak temporal resolution of fMRI does not allow incisive analysis of how information is passed around different brain regions in the analysis of incoming information and the organization of response. Technical advances, such as those of electromagnetography, should contribute. The charting of normal developmental function has so far been done only for a limited number of functions and a limited range of ages. Further exploration is needed, especially of the functions most likely to be involved in pathology. Clinical application is severely limited by the lack of standardized tests that have been satisfactorily normed (TEACH and CANTAB are significant exceptions), but the field has advanced to the point where it should be possible to do this and to identify children on the basis of attentional change for behavioral investigation, rather than vice versa as at present in psychopathology. Standardization should advance to the point of understanding changes in people with intellectual disability, so that a concept of specific attentional disability could come to have operational meaning. The value of
such analyses for remediation remains unknown: The training of attentional abilities is still a topic for the future. We can look forward to exciting and unpredictable changes.

References


Each New Year finds many of us making mistakes when writing the date on letters or checks. This kind of “capture error,” first described in William James’s (1890) famous example of going upstairs to change and discovering himself in bed, supports cognitive psychologists’ classical distinction between actions that require conscious effortful control and those that are executed automatically (e.g., Atkinson & Shiffrin, 1968; Schneider & Shiffrin, 1977). This distinction is not simply a contrast between simple and complex actions, as well-learned complex actions can be automatic (e.g., driving a car); nor between externally and internally driven actions, as the latter can be based on automatic processes (e.g., memory recall). Instead, the distinction between controlled and automatic actions hinges upon three key features: (a) the execution of novel versus familiar action sequences; (b) making a choice between alternative responses versus executing a single action sequence; and (c) the execution of acts that do versus those that do not require access to consciousness.

The term “executive function” (EF) therefore refers to a complex cognitive construct encompassing the whole set of processes underlying these controlled, goal-directed responses to novel or difficult situations. More specifically, EF is held to be necessary in situations that involve: (a) planning and decision-making; (b) error correction or troubleshooting; (c) initiation of novel sequences of actions; (d) danger or technical difficulty; or (e) the need to overcome a strong habitual response (Norman & Shallice, 1980, 1986; Shallice & Burgess, 1991). This view of EF as responsible for programming, monitoring, and regulating behavior has been expressed computationally in Shallice and Burgess’s (1991) model of the supervisory attentional system (SAS) illustrated in Figure 12.1.

Alongside its functional definition described above, the term EF is often defined in structural terms: Clinical researchers typically view EF as essentially synonymous with the functioning of the prefrontal cortex (PFC), the regions of which are shown in Figure 12.2. From this clinical viewpoint, EF is most conspicuous by its absence
in individuals with prefrontal lesions who, together with behavioral and personality changes, demonstrate impairments in functions such as planning or abstract thinking. The cognitive importance of the PFC first became apparent through studies of World War I veterans, which demonstrated that soldiers with frontal lobe injuries were unimpaired on routine tasks, but had difficulty mastering new tasks or grasping the whole of a complicated task (Goldstein, 1936, 1944). This led to the view that EF was important for abstract or high level thought, abilities only manifested in adulthood. Support for this view of EF as emerging late in development came from neuroanatomical work suggesting that the PFC only becomes functionally mature around adolescence (Golden, 1981). In addition, primate studies and early research on head injuries suggested that the consequences of juvenile lesions to the PFC did not become apparent until adulthood (the so-called “Kennard effect”). Together, the above factors led to a chronic neglect of EF in childhood for much of the past century.
Figure 12.2 Surface and medial views of the brain, showing key regions of the prefrontal cortex.

Measurement: Problems and Solutions

In contrast with the consensus described above regarding the theoretical definition of EF, an operational definition of EF is much harder to establish. Despite its long history, EF research therefore continues to be riddled with practical challenges. The first obstacle to obtaining accurate, valid, and reliable measurements of EF is that the contrast between automatic and controlled actions is not absolute. Rather, these two types of actions are at the opposite ends of a continuum. Thus, the processes underlying an individual’s performance on a novel task will shift gradually over time from controlled
to automatic. In addition, small changes in task demands may lead to a collapse of automatic performance and a return to controlled performance. As a result, it is often difficult to distinguish “executive” from “nonexecutive” tasks. Indeed, re-administration of any particular test will never tap EF to the same extent as on the first presentation, leading to poor test/re-test reliability. In children, however, the shift from controlled to automatic processes is likely to be more extended, leading to greater stability in both underlying processes and overall performance. In other words, compared with adults, children may perceive (and process) a new task as novel for longer, so that both the validity of identifying a task as an EF test and the reliability of individual differences in task performance are potentially better in studies of children than in studies of adults.

A second obstacle stems from the complexity of standard EF tasks. This complexity means that measures of task performance often represent the pooled outcome of several distinct underlying processes. So, for example, an EF task may be presented as a “planning” task, but also involve other EF processes. Such complex tasks are also susceptible to problems of task impurity, in that their execution may trigger additional nonexecutive processes unrelated to the task at hand. Indeed, since everyday terms such as “planning” are poorly specified, there is no guarantee that the main psychological process actually underlying a “planning task” has any relationship to our folk notion of planning. Furthermore, for EF tasks that involve the simultaneous coordination of a variety of different processes (Kimberg & Farah, 1993), any attempt to isolate an index of a single functional process by applying classical experimental techniques to control task demands is unlikely to be successful.

However, in order to be developmentally appropriate for use with children, standard EF tasks need considerable simplification, and so problems of task impurity are likely to be reduced. Also, given children’s relatively limited processing capacities, it is not necessary to tax several processes simultaneously in order to tap controlled processing. As a result, classical experimental techniques may be more successful. In addition, manipulating task parameters may be especially fruitful in studies of children, since their relatively limited processing capacity makes children more sensitive to effects of increased demands for particular functions. By enabling the components within a specific task to be manipulated directly, this approach provides an important step forward from studies that rely upon within-child correlations in performance across tasks. However, the corollary is that establishing standardized methodological procedures is especially important in studies of children.

A final obstacle for researchers is that of low process–behavior correspondence. Many psychological processes manifest themselves only in one narrow type of situation and not at all in others (e.g., the face recognition system is activated when a subject is shown photos of famous faces, but not a list of words). By contrast, executive processes manifest themselves in a range of different situations. This leads to a low correspondence between process and behavior: A specific process impairment can result in a variety of behaviors, and conversely a specific behavior can be caused by a variety of process impairments. However, compared with adults, children are more transparent in their
behavior (e.g., they are more likely to “think aloud” and to make overt displays of response suppression), and so once again this problem may be less of an issue for research with children. In particular, EF studies of children may benefit particularly from combining quantitative experimental measures of task performance (e.g., percentage correct, time taken) with qualitative observational ratings of task behavior (e.g., frequency of rule violations, displays of frustration/distractibility). For example, one early method of studying rehearsal in children was to record overt signs such as lip movements and whispering. This approach indicated that children begin to use strategies of rehearsal around the age of 7 (Flavell, Beach, & Chinsky, 1966), confirming the results of more formal and inferential analyses in subsequent studies.

Thus, many of the difficulties in measuring EF in adult populations may, at least in part, be overcome when working with children, so that developmental studies of EF may provide special insights into the organization of EF. Nevertheless, researchers interested in charting the development of EF are confronted by a number of problems specific to childhood, particularly as a result of language limitations. For example, by taxing a child’s verbal comprehension (e.g., via complex instructions) a task places demands upon peripheral functions that may well influence overall performance. In addition, fluent literacy emerges relatively late in development and so many adult EF tasks are inappropriate for children, simply because they depend upon over-learned (i.e., automatic) written language skills. Examples include: (a) the Stroop test, in which reading the word “blue” (say) written in a differently colored ink is assumed to be an automatic prepotent response; (b) the trail-making test, in which an alphabet sequence must be repeatedly interrupted to interleave a number sequence (and a tendency to complete either sequence is assumed); (c) random-sequence generation tasks, in which frequent letter/digit/word sequences (e.g., ABC) are again assumed to be automatic prepotent responses; (d) the FAS verbal fluency test, in which individuals are required to name as many words beginning with F, A, or S, and so a rudimentary spelling ability is assumed; and (e) the Hayling sentence completion task, in which an individual must complete a sentence in an unexpected way (so that, again, strong associations between certain word combinations are again assumed to be prepotent).

To minimize this load upon verbal comprehension, EF tests for young children must therefore be kept as simple as possible. A seminal figure who developed many classic EF tests suitable for use with children is the Soviet psychologist, Alexander Luria (1966). These tasks include the go/no-go test (in which the child must execute a response to stimulus A, but withhold this response when presented with stimulus B), and a variety of nonverbal Stroop tasks for children, including the picture-based day/night task (in which the child must say “day” for a picture of the moon, and “night” for a picture of the sun; Diamond & Taylor, 1996; Luria, Pribram, & Homskaya, 1964; Passler, Isaac, & Hynd, 1985), the tapping game (in which the child must tap once in reply to two taps, and twice in response to a single tap), or the hand game depicted in Figure 12.3.

In the same tradition, Frith (1972; Hughes, 1998a) devised a pattern-making task that required children to avoid simple repeats (e.g., dropping black [B] and white [W] marbles into a tube, but avoiding sequences such as BBB/WWW/BWBW). Child-friendly
fluency tasks have also been devised: These involve generating names of objects in a particular category (e.g., animals/transport/clothes; White, Nortz, Mandernach, Huntington, & Steiner, 2001) rather than naming words beginning with a particular letter. Other EF tasks, simple enough even for infants, have also been developed. These tasks (described more fully later) include (a) Diamond’s version of Piaget’s A-not-B test; (b) the object reversal test in which an established reward contingency is reversed; and (c) object retrieval tasks that tap the ability to perform a means–end action, such as making a detour around a barrier to retrieve a desired object (Diamond, 1990; Diamond & Goldman-Rakic, 1989).

These simple tasks have also led to dramatic improvements in our understanding of the development of EF. For instance, it is now known that EF: (a) begins to emerge in the first few years of life; (b) shows stage-like, age-related changes; (c) becomes fully mature in late adolescence and declines with normal ageing; (d) subdivides in children and adults in similar ways (in each case the three most widely reported factors are inhibitory control, attentional flexibility, and working memory/planning); and (e) has important consequences for other cognitive functions. Below we discuss the fractionation and development of EF in more detail.
Neurological Substrate and Fractionation of Executive Function

It is widely accepted that EF is dependent on the functioning of the PFC, conventionally defined as the portion of the frontal cortex that receives projections from the mediiodorsal nucleus of the thalamus. Based on the internal divisions of this nucleus, the prefrontal cortex may be further subdivided into three main regions: the orbitofrontal PFC, the dorsolateral PFC, and the frontal eye fields (the frontal eye fields are involved in the higher control of eye movements and will not be discussed further here). The PFC is also closely connected with neighboring brain regions such as the striatum, anterior cingulate cortex, and the limbic system, with these connections arranged to form a number of parallel circuits. In addition to these parallel anatomical systems, there are parallel neurochemical systems connecting to the PFC, chiefly ascending dopaminergic and serotonergic pathways. There is some overlap between anatomical and neurochemical systems: Dorsolateral prefrontal cortex, for example, is particularly richly innervated by ascending dopaminergic neurons. A full account of the neuroanatomical and neurochemical evidence in support of a fractionated model of prefrontal function is beyond the scope of this chapter, but useful reviews can be found in Casey, Galvan, and Hare (2005) and Robbins (2005).

Working along these anatomical lines, clinical researchers into EF have repeatedly attempted to differentiate between the functions of the orbitofrontal PFC and the dorsolateral PFC. In brief, dorsolateral PFC is particularly connected with regions associated with motor control (e.g., the basal ganglia) and performance monitoring (e.g., the anterior cingulate cortex). Ventromedial PFC, by contrast, is reciprocally connected with regions involved in emotional processing (e.g., the amygdala) and memory (e.g., the hippocampus). In its simplest form, this amounts to a distinction between "hot" versus "cool" EF (i.e., emotional regulation and behavioral control versus affectively neutral higher order cognitive processes).

Exactly how the PFC supports EF is a matter of greater controversy. This is a rapidly expanding field of research, increasingly founded on novel, noninvasive techniques such as event-related potential (ERP) studies, electroencephalography (EEG), and functional imaging studies involving either magnetic resonance imaging (MRI) or positron emission tomography (PET). Many suggestions have been put forward, including Norman and Shallice's attentional control model, Duncan's adaptive coding model, Damasio's somatic marker hypothesis, and Goldman-Rakic's working memory model. At present, no consensus has been reached (for a comprehensive review of competing contemporary models of PFC function, see Wood & Grafman, 2003).

Within cognitive psychology, a multicomponent view of EF is attractive because it helps to avoid the philosophical problem of the homunculus or "ghost in the machine" (Ryle, 1949). Many psychologists have observed that the functions ascribed to the central executive such as "planning," "choosing," "deciding," and other "higher cognitive processes" are poorly defined and all too often imply some sort of conscious, overarching system or entity making these plans, choices, and decisions. This has led investigators
to decompose EF into smaller fragments—a collection of automatic tools, perhaps, rather than an engineer (Shallice & Burgess, 1991).

Empirically, the distinction between different components of EF is also important to developmental cognitive psychologists, for a number of reasons. First, interpreting the results from any EF task depends upon a precise specification of task demands, which in turn depends on a systematic outline of the components of EF. Second, in order to develop age-appropriate versions of adult EF tasks, it is desirable to simplify tasks along the lines of their natural components rather than in an ad hoc fashion. Third, given the variety of developmental disorders in which executive impairments are observed, a finer-grain level of analysis is required to draw useful distinctions between clinical groups and to advance hypotheses about the underlying mechanisms in each case. Here it is likely to be qualitative differences in profiles of EF performance, rather than differences in overall EF performance, that prove most informative. Nevertheless, studying the fractionation of EF in adult populations can be difficult, for a variety of reasons. In particular, fractionation may be either overestimated or underestimated, depending on which processes and behaviors are under investigation (Burgess, 1997).

Paradoxically, because of their relatively limited executive capacities, young children may be ideal candidates for evaluating theoretical predictions concerning the relationship between specific EFs. Compared with adults, tapping EF in children is less dependent upon demands for a sequence of acts, while experimental manipulations have a more pronounced effect; thus, interactive effects can be investigated directly using within-task designs. However, one should avoid the assumption that a specific EF task poses the same type of cognitive demands for children of different ages.

Normal Neuropsychological Development of Executive Function

What do we know about the development of EF in childhood? First, the onset of EF development is much earlier than was traditionally thought (e.g., by Golden, 1981; by Luria, 1973). This does, of course, depend on utilizing the appropriate tools to study EF at an early age. In particular, an impressive set of behavioral and comparative studies by Adele Diamond and colleagues has shown that the functioning of the dorsolateral PFC underpins the performance of infants on Piaget's A-not-B task (Piaget, 1976). In this task, a toy is conspicuously hidden in one place (A); the infant is then allowed to search for it. This is repeated for a few trials and then the object is visibly moved to location B. At this stage in the task, 8-month-old infants persist in searching at location A, but 12-month-olds search correctly at B on the first trial and show few if any perseverative errors on subsequent trials, even if a delay between concealment and search is introduced, making the task more difficult (Diamond, 1988). Support for the involvement of the dorsolateral PFC in this task comes from the finding (from a group of infants tested longitudinally between 7 and 12 months of age) that task performance is correlated with EEG activity and coherence in this brain region (Bell & Fox, 1992).
A second finding to emerge from several independent studies is that EF in children subdivides in a similar manner to adults. Using principal components analyses, Welsh and colleagues (Welsh, Pennington, & Groisser, 1991) showed that the performance of both 8- to 12-year-olds and adults on a battery of EF tasks displayed the same three-factor solution. Similarly, Levin and colleagues (1991) found that the performance of 7- to 15-year-olds on a battery of EF tasks showed the same three-factor solution for all ages. Although the task loadings were not identical in the two studies, in both cases the first factor appeared to be a measure of cognitive flexibility, whilst the second factor tapped into problems of inhibitory control and the third factor captured planning (and working memory). A large-scale study of 537 school-aged children confirmed this three-factor structure (Pennington, 1997), which has also been extended to 3- and 4-year-olds in a study that employed simplified task variants that were adapted for preschoolers (Hughes, 1998a).

Together, these findings suggest remarkable continuity in the structure of EF from preschool to adulthood. However, as noted by Zelazo and Muller (2002), the story is more complicated than this commonality suggests: Correlations among tasks can result not only from similarities in the mechanisms underlying task performance, but also from shared method variance and shared sensitivity to individual differences at specific ages. Nevertheless, given the above consensus in findings, we shall in the remainder of this section present the key developmental changes for each of the three main factors identified above. For reasons of space, we shall not attempt an exhaustive review of EF tasks that have been used with children (the interested reader is referred to Zelazo & Muller, 2002, for a discussion of some of the tasks that are not reviewed here).

Cognitive flexibility

The most widely used measure of “cognitive flexibility” is Grant and Berg’s (1948) Wisconsin Card Sorting Test (WCST), which involves a deck of picture cards (varying in color, shape, and number), and four target cards. Children are asked to place each card on one of the targets; through feedback on each trial they can, by a process of trial and error, deduce the sorting “rule” as to which dimension is salient. After six consecutively correct responses, the rule is changed (e.g., sort by shape to sort by color). Key measures from the WCST are the numbers of (a) rules deduced and (b) perseverative errors. Chelune and Baer (1986) reported a steady improvement in WCST performance from 6 years, achieving adult levels of performance by around 10 years of age (see also Levin et al., 1991; Welsh, Pennington, & Groisser, 1991). Unfortunately, like other traditional EF tasks, the WCST falls far short of providing a pure measure of cognitive flexibility. In particular, because the same cards are used for each rule (and the child is not told that the rule has changed), perseverative errors on the WCST may reflect either a genuine failure to shift mental set to the newly relevant dimension or simply a failure to inhibit a previously reinforced response to a specific exemplar (e.g., triangles). Thus, the WCST provides a sensitive but nonspecific index of EF impairment.
To rectify this problem of nonspecificity, a computerized variant of the WCST has been developed as part of the Cambridge Automated Neuropsychological Task Battery (CANTAB; Robbins et al., 1997). Two features of the ID/ED test (intradimensional/extradimensional shift test) from the CANTAB enable a finer grain of analysis: (a) a "total transfer" design (i.e., new exemplars are presented when the child achieves criterion on the key stages, alerting the child to the change in rule), and (b) graded multiple stages. Stage 1 tests rule learning; stage 2 tests rule reversal; stage 3 introduces distracter elements that in stage 4 become superimposed upon the salient shape; in stage 5, the rule is again reversed; in stage 6, the shapes and distractors are changed but the rule remains the same (this ID shift tests transfer of learning); in stage 7, the rule is again reversed; in stage 8, new exemplars are presented and the previously irrelevant distractors now become salient (this ED shift is crucial as it provides a relatively pure test of problems in set-shifting); in stage 9, the new rule is reversed.

Hughes, Russell, and Robbins (1994) showed that children with autism typically fail the ED stage of the ID/ED task, and this finding has recently been replicated in a large autistic sample (Ozonoff et al., 2004). Following this intriguing finding, which indicates a marked autistic impairment in higher order set-shifting, Luciana and Nelson (1998) found that most typically developing children did not succeed on the ED shift stage of the task before the age of 6 years. This highlights the importance of adopting developmentally appropriate EF tasks to investigate rudimentary EF skills in young clinical groups.

For example, total transfer set-shifting tasks that are failed by typically developing preschoolers (Hughes, 1998a) are too taxing to be sensitive to impairments in set-shifting among autistic preschoolers (Shearer, 2002); however, simpler rule-switching tasks, such as the Dimensional Change Card Sorting (DCCS) task do appear sensitive to autistic impairments in attentional flexibility (Colvert, Custance, & Swettenham, 2002; Zelazo, Jacques, Burack, & Frye, 2002). It should be noted that the authors of the DCCS view this task as a test of cognitive complexity rather than cognitive flexibility. Specifically, Zelazo and Frye (1997) argue that in order to switch between two rules, a child needs to be able to represent embedded if–if–then structures. According to this "cognitive complexity and control" (CCC) account, 3-year-olds fail the DCCS task because they are unable to distance themselves sufficiently from a particular way of conceptualizing a card so that they can select the right conceptualization when the time comes.

However, findings from a study by Perner and Lang (2002) challenge this view. Specifically, by systematically varying the component demands, Perner and Lang (2002) were able to show that 3-year-olds only displayed a performance deficit on a variant of the DCCS task that included both visual capture and an extradimensional shift. While they discuss this finding in terms of the questions it raises for both CCC and inhibitory control accounts, in the current context this interaction effect highlights the importance of a developmental perspective. That is, the 3-year-olds (but not the 4-year-olds) in Perner and Lang’s study were clearly affected by both the novelty and pragmatic complexity of specific rule-switching tasks. This finding highlights the need to avoid the assumption that the same task poses the same type of cognitive demands for children of different ages.
Inhibitory control

Findings from both clinical and developmental research indicate that the construct of inhibitory control can itself be subdivided. In particular, there is a clear difference in demands between tasks that simply require the child to withhold a prepotent response (e.g., the stop task, or the go/no-go task) and tasks that require the child to withhold a prepotent response in order to execute a rule-guided action. With regard to the first type of task, recent findings suggest that the go/no-go task (widely used with school-aged children) also gives reliable and valid results with preschoolers (Mahone, Pillion, & Hiemenz, 2001). Specifically, 3-year-olds completed a computerized go/no-go task with few omission or commission errors, and from age 3 to age 6 there was a significant reduction in errors, response latency and variability (which correlated significantly with parental ratings of attentional problems).

The second, more complex, type of inhibitory control is tapped by nonverbal Stroop tasks (e.g., Luria’s day/night, hand game, knock/tap) and by the detour-reaching task (Hughes & Russell, 1993). For example, Hughes and Russell (1993) reported that 3-year-olds and older children with autism (but not typically developing 4-year-olds) failed to retrieve a marble inside a box when this required not only the inhibition of a direct reach but also an indirect and arbitrary means–end action (flicking a switch before reaching into the box). Likewise, the majority of 3-year-olds (but not 4-year-olds) also fail the day/night task (Gerstadt, Hong, & Diamond, 1994), the hand game (Hughes, 1996, 1998a), and the knock/tap task (Perner & Lang, 2002). Significant improvements in both simple and complex inhibitory control are therefore evident across the preschool years. Developmental improvements in inhibitory control also continue throughout childhood, as demonstrated by findings from studies with school-aged children using the stop and go/no-go tasks and the Opposite Worlds Stroop task (e.g., Manly et al., 2001). Interestingly, findings from an fMRI study (Casey et al., 1997) suggest that children and adults show similar patterns of brain activation during the go/no-go task, and demonstrate a negative association between test indices for inhibitory control (e.g., errors of commission) and ventromedial PFC activity.

Planning

The most widely used paradigm for testing planning skills is the Tower of Hanoi task (Piaget, 1976), and its simplified variant, the Tower of London (Shallice, 1982). The differences between these two tasks are subtle, but important. In the Tower of Hanoi, a set of doughnut-like discs, graded in size to form a pyramid-like structure, must be moved from one of three equally sized pegs to another, following the following constraints: move only one disc at a time; move only the top disc on a peg; place no disc on top of a bigger disc. A “perfect” solution requires \(2^n-1\) moves, where \(n\) is the number of discs. So a very simple 2-disc problem would take 3 moves to solve, whereas a 6-disc problem would require a minimum of 63 moves. Performance is measured by the number of attempts the child (or adult!) requires to achieve a perfect
solution: The Tower of Hanoi task therefore taps not only planning ability but also rule following and procedural learning.

The Tower of London provides a purer test of planning ability: Discs are replaced by three differently colored balls (removing much of the need for rule constraints); the pegs vary in length (reducing the size of the “problem space”); the full tower-to-tower transfer is replaced by a graded set of novel 2- to 5-move subproblems (minimizing practice effects). Key performance measures at each level include the number of perfect solutions and the mean number of extra moves. The computerized version of the Tower of London in the CANTAB also includes a yoked set of “follow” tasks; by subtracting individual response times for each of these from those on the corresponding full task, both thinking and execution times can be estimated. These modifications make the Tower of London a much more satisfactory test of planning than the Tower of Hanoi.1

For reasons of space, we shall therefore restrict our report to findings from the Tower of London. Note that in a study using positron emission tomography (PET), the planning demands of the CANTAB Tower of London task were associated with activation of the dorsolateral PFC (Baker et al., 1996).

Using a manual version of the Tower of London, Hughes (1998b) found that 4- and 5-year-olds showed a steep drop in performance across problem levels: 62% produced perfect solutions to all three 2-move subproblems, as compared with 22% at the 3-move level, and just 16% at the 4-move level. Interestingly, overall scores correlated significantly with the children’s performances on the detour-reaching task a year earlier. Together, these results indicate that the Tower of London is developmentally appropriate for preschool-aged children, and also highlight the magnitude and validity of early individual differences in planning ability. Finally, Luciana and Nelson (1998) demonstrated that developmental improvements in planning continue at a steady rate through the school years.

Abnormal Development

Impairments in EF are now thought to play a key role in a variety of developmental disorders, though the evidence is strongest for attention deficit hyperactivity disorder (ADHD) and autism (Pennington & Ozonoff, 1996). For reasons of space, and because the evidence for a specific EF impairment appears stronger for ADHD, we shall in this section only consider EF in relation to ADHD. This syndrome is relatively common, affecting 2–7% of school-aged children (mostly boys), but is difficult to diagnose before the age of 6, perhaps because its three cardinal symptoms (distractibility, hyperactivity, and impulsivity) tap what is often “normal” behavior in very young children. Children

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1 Nevertheless, planning ability clearly depends on several component executive skills (Goel & Grafman, 1995). According to Shallice (1982), success on the Tower of London involves focused and sustained attention, goal recognition/selection, generation of plans, and appropriate response to feedback.
with ADHD often show other comorbid problems such as oppositional defiant disorder, conduct disorder, reading disability, and (less frequently) depression or anxiety.

There is good evidence for PFC involvement in ADHD, in terms of both structure (e.g., delayed myelination of the PFC) and function (e.g., dopamine depletion). Increasingly, children with ADHD are medicated with amphetamine-based stimulants (most commonly, Ritalin) that affect the dopaminergic system and lead to improvements in both behavior and cognitive performance (e.g., Kempton et al., 1999). Group differences between children with ADHD and ability-matched controls show large effect sizes (d = 1) and are most evident on tasks that require inhibitory control (Barkley, 1997; Sergeant, 2000). Studies have also suggested that children with ADHD lack strategic flexibility, display poor planning and working memory, and are poor at monitoring their own behavior (Cepeda, Cepeda, & Kramer, 2000; Clark & Rutter, 1981), but none the less it is generally impairments in inhibitory control that are emphasized.

Impairment of attention in ADHD is clearly a diagnostic feature, although historically the nature of this impairment has been poorly specified. Manly et al. (2001) reported findings from 24 boys with ADHD (not yet receiving medication) tested on an age-normed battery of manual tests of attention and EF (the Test of Everyday Attention in Children). Compared with several comparison groups, the ADHD boys showed global deficits, but group differences were especially robust on the tests of sustained attention and suppression of prepotent responses. In contrast, a study using the CANTAB with a group of “hard to manage” 7-year-olds (Brophy, Taylor, & Hughes, 2002) showed no deficits on any of the formal task measures, despite significantly elevated rates of observed rule violations and perseverative errors whilst performing the tasks.² However, positive findings have been reported for another computerized battery, the Maudsley Attention and Response Suppression (MARS) battery which includes three response-inhibition tasks: go/no-go, stop, and rule reversal. Children with ADHD (aged 7–15 years, and off medication for at least 48 hours) performed significantly worse than IQ-matched psychiatric and typical controls on all three tasks (Rubia et al., 2001). Moreover, fMRI of a subset of participants during the response-inhibition tasks revealed reduced right prefrontal activation for the ADHD group.

The above findings support the conceptualization of ADHD as an executive disorder, in which inhibitory control is especially affected. However, an alternative view has also been posited, namely that the behaviors that typify ADHD may reflect deviance in motivational style rather than executive dysfunction (e.g., Haelelein & Caul, 1987; Johansen, Aase, Meyer, & Sagvolden, 2002; Zentall & Zentall, 1983). Specifically, children with ADHD appear motivated to avoid or escape delay, perhaps because of an altered perception of time (Sonuga-Barke, Houlberg, & Hall, 1994). If so, children

² This may be because the CANTAB software is programmed to ignore irrelevant responses in order to obtain “pure” measures of specific functions. Whilst clearly a laudable aim in adult studies, this design feature does appear to limit the use of CANTAB with younger children.
with ADHD may appear inattentive, overactive, or impulsive (and perform poorly on executive tasks) without any primary underlying executive impairments.

The true picture may be that ADHD is a heterogeneous disorder, and includes both executive and motivational deficits. In support of this “dual path” model (Sonuga-Barke, 2002), several experiments (Sonuga-Barke, Houbreg, & Hall, 1994; Sonuga-Barke, Taylor, Semb, & Smith, 1992; Sonuga-Barke, Williams, Hall, & Saxton, 1996) demonstrate a double dissociation between preference for delayed rewards and inhibitory control; in addition, a recent multicenter study of children with ADHD (Solanto et al., 2001) revealed dual but unrelated impairments on a standard stop signal task (indicating poor inhibitory control) and a choice delay task (implicating motivational factors). Investigations of EF deficits may therefore be more informative at the level of symptoms than at that of diagnostic category.

Behavioral Genetics of Executive Function

The range of possible genetic influences on prefrontal function and EF is vast, and findings in this area are largely preliminary. In summary, there is good evidence that both general and specific genetic factors are involved in the determination of prefrontal function (Winterer & Goldman, 2003). Many of the general factors appear to play a wider role in the regulation of central nervous system function and are also implicated in the heritability of attributes such as general intelligence or processing speed. These general genetic factors are insufficient, however, to explain the heritability of more specific executive functions such as working memory.

Efforts to investigate the genetics of higher cognitive functions in more detail are constrained by the lack of an operational definition for EF, and studies to date have therefore concentrated, understandably, on either relatively narrow aspects of EF, such as the executive attention network (Fan, Wu, Fossella, & Posner, 2001), or the co-occurrence of EF impairment in heritable and operationally defined clinical disorders such as ADHD. For example, in individuals with ADHD, population-based twin studies show a highly significant overlap between ADHD symptoms/diagnosis and IQ, a co-association that appears to be almost entirely carried by shared genetic influences (Kuntsi et al., 2004). Similarly, structural equation modeling studies suggest a substantial overlap between the genetic liabilities to the disorder of ADHD, accompanying EF deficits, and comorbid conduct disorder or oppositional defiant disorder, although there is also support for additional genetic influences underlying EF variability independent of those shared with ADHD (Coolidge, Thede, & Young, 2000).

Perhaps the most specific genetic factor yet identified concerns polymorphisms in the gene for the enzyme catechol-O-methyltransferase (COMT), which is involved in the degradation of dopamine, and performance on tasks of working memory (Malhotra et al., 2002). In a significant proportion of individuals, the COMT gene contains a variation in its coding sequence at position 472, leading to a single amino acid substitution of methionine for valine. This substitution leads to a dramatic increase in the
temperature lability of the enzyme, such that individuals with the \textit{met} allele have only one-quarter of the enzyme activity of individuals with the \textit{val} allele (Lachman et al., 1996). Remarkably, enzyme activity is in turn correlated with performance on a standard test of EF, the Wisconsin Card Sorting Test, individuals homozygous for the \textit{val} allele showing the best performance, heterozygous \textit{met/val} individuals showing intermediate performance, and individuals homozygous for the \textit{met} allele showing the worst performance (Egan et al., 2001). Although studies screening further candidate genes for their influence on prefrontal function have so far yielded few clear results, this may in large part be due to a lack of power in early studies, a problem that is likely to be overcome by the rapid pace of methodological advances in the field of behavioral genetics.

\section*{Clinical Implications and Current Research}

The rapid progress in our understanding of the basic development of EF has exciting consequences, and there are now several hot topics for research. First, impairments in the control of action contribute to the behavioral problems that set children on a trajectory towards deviance, delinquency, and antisocial conduct. For instance, in a study of "hard to manage" 3- and 4-year-olds, individual differences in EF were significantly associated with antisocial behavior (Hughes, White, Sharpen, & Dunn, 2000). Studies that deepen our understanding of normative age-related improvements in EF may therefore help to identify children with poor regulatory control who could benefit from intervention programs, and so has clear societal importance. Thus, interest in early EF is closely tied to the growth of the new discipline of \textit{developmental neuropsychology}. In particular, impairments in EF are thought to play a key role in several childhood disorders, including attention deficit hyperactivity disorder (ADHD) and autism, though the latter is more controversial.

Second, studies with children offer the promise of differentiating the components of EF. In particular, the technique of manipulating task parameters may be especially fruitful in studies of children, since their relatively limited processing capacity makes them more sensitive to effects of increased demands for particular functions. This provides a direct solution to the low \textit{discriminant validity} shown by traditional EF tasks. Because such tasks are typically complex and multicomponent, different clinical groups may perform equally poorly for different reasons. For example, ADHD and autism have quite different clinical presentations, and yet both groups show substantial EF deficits (scoring \(\approx 1\) \textit{SD} below control groups). At first glance, one might therefore expect EF impairments to be rather nonspecific. However, studies that adopt an information-processing approach (involving simplified tasks that allow comparisons based on specific rather than global performance measures) have revealed both quantitative and qualitative distinctions between EF impairments in these two disorders.

Third, there is converging evidence for a functional link between EF and "theory of mind" (ToM) defined as the ability to attribute mental states to oneself and others.
This evidence includes: (a) pronounced impairments in both EF and ToM among children with autism; (b) the developmental synchrony of improvements in both EF and ToM among typically developing preschoolers; and (c) robust correlations between individual differences in EF and ToM, even with effects of age and IQ controlled. Since the topic of ToM continues to attract intense research interest, the nature and significance of its association with EF is a matter of considerable debate. Empirically, there is longitudinal evidence for a predictive association between individual differences in EF at age 4 and in ToM one year later (even controlling for initial ToM), but no association between early ToM and later EF (Hughes, 1998b). This asymmetry suggests a direction of influence (EF → ToM), but findings from intervention studies are needed to establish a causal path.

In addition, it may be that associations between EF and ToM are specific rather than global. For example, Carlson and Moses (2001) have reported particularly strong associations between inhibitory control and ToM. Further support for this view comes from the findings of several imaging studies, demonstrating that ToM tasks activate the ventromedial PFC (the subregion previously identified as important in inhibitory control). Since individuals with autism are known to show profound impairments in ToM, this finding suggests that research into EF impairments in autism should focus on EF tasks that are associated with the ventromedial PFC (previous research in this field has generally employed traditional EF tasks that are typically associated with the dorsolateral PFC). Thus, one positive consequence of the debate surrounding the relation between ToM and EF is the integration of brain-based research with studies of both normative and atypical development.

**Methodological Challenges and Future Directions**

Despite the various advances outlined above, it should be emphasized that EF research remains besieged by methodological problems. In particular, much more work is needed to achieve a fine-grained analysis of the distinct components of EF. This fractionated approach is also important as a solution to the *homunculus problem*, raised by terms such as “effortful control,” since it allows EF to be compared with a set of automatic tools rather than an engineer. In addition, it is very difficult to design “pure” tests of EF, or even tasks that show good test/re-test reliability (an inherent problem with EF research is that any task is only truly novel once). Progress in our conceptual understanding of EF depends critically upon innovative and rigorous solutions to these methodological challenges.

The challenges for future research in the above areas are vast and varied. However, one that deserves special mention is the need to investigate whether contrasts in EF help explain differences in the form and severity of behavioral symptoms. Research on this topic may enable us to elucidate specific links between EF and behavior. Such links may be best described in terms of distinct behaviors, such as reactive versus proactive forms of aggression (for disruptive behavior disorders) or catastrophic responses to
change versus ritualistic routines (for autism). Alternatively, links between EF and behavior may be clearest for specific contexts (e.g., peer interactions that are not scaffolded by familiar routines). Addressing the relation between variability in EF and in behavior also provides a promising alternative to simply comparing diagnostic groups, in that disorders with overlapping symptoms (e.g., ADHD and conduct disorder) may show relative rather than absolute differences in EF. For many reasons, then, research that combines innovative task manipulations with valid and reliable observational methods is vital.

References


In this chapter, we take a key concept in neuroscience, namely empathy, and consider it in terms of neurocognitive developmental mechanisms and in terms of individual differences. The first part of the chapter deals with the development of empathy; the second part presents some empirical evidence on a quantitative trait measure of empathy. Trait empathy is intrinsically connected with our perception of, and response to, others’ emotions. We therefore briefly review the literature on emotion processing from neuroimaging studies that suggest that discrete basic emotions are subserved by different neural regions and networks (Panksepp, 1998). Finally, we describe a recent study that investigates whether and how an individual’s level of empathy affects the way in which their brain processes discrete emotions.

What is Empathizing?

Empathizing is the drive to identify another person’s emotions and thoughts, and to respond to these with an appropriate emotion (Davis, 1994). We use the term “drive” but recognize that it also overlaps with the concept of a skill or ability. We also focus on the definition of empathy given by Davis (1994), whilst recognizing that other authors may have a slightly different definition. Empathizing does not just entail the cold calculation of what someone else thinks and feels (or what is sometimes called “mind-reading”). Psychopaths can do that much. Empathizing is also about having an appropriate emotional reaction inside you, an emotion triggered by the other person’s emotion. Empathizing is done in order to understand another person, to predict their behavior, and to connect or resonate with them emotionally. Imagine you could recognize that “Jane is in pain” but this left you cold, or detached, or happy, or preoccupied. This would not be empathizing. Now imagine you do not only see Jane’s pain, but you also automatically feel concern, wincing yourself, and feeling a desire to run across and help alleviate her pain. That is empathizing. And empathizing extends to recognizing
and responding to any emotion or state of mind, not just the more obvious ones, like pain. Empathy is a skill (or a set of skills). As with any other skill, such as athleticism or mathematical or musical ability, we all vary in it. In the same way that we can consider why someone is talented or average or even disabled in these other areas, so we can think about individual differences in empathy.

Empathy is a defining feature of human relationships. Empathy stops you doing things that would hurt another person’s feelings. Empathy also stops you inflicting pain on a person or animal. Empathy allows you to tune into someone else’s world, setting aside your own world—your perception, knowledge, assumptions, or feelings. It allows you to see another side of an argument easily. Empathy drives you to care for, or offer comfort to, another person, even if they are unrelated to you and you stand to gain nothing in return. Empathy also makes real communication possible. Talking “at” a person is not real communication. It is a monologue. Real conversation is sensitive to this listener at this time. Empathy also provides a framework for the development of a moral code. Moral codes are built out of fellow-feeling and compassion.

**Fractionating Empathy**

Philosophical (Stein, 1989) and evolutionary (Brothers, 1990; Levenson, 1996; Preston & de Waal, 2002) accounts have suggested that empathizing is not a unitary construct. Possible constituent “fractions” of empathy include (a) “emotional contagion/affective empathy,” (b) “cognitive empathy,” and (c) sympathy. Cognitive empathy is involved in explicit understanding of another’s feelings and switching to take their perspective. Piaget referred to empathy as “decentering” or responding non-egocentrically (Piaget & Inhelder, 1956). More recent developmental psychologists refer to this aspect of empathy in terms of using a “theory of mind” or “mindreading” (Astington, Harris, & Olson, 1988; Whiten, 1991). Essentially, the cognitive component of empathizing entails setting aside your own current perspective, attributing a mental state (sometimes called an “attitude”) to the other person, and then inferring the likely content of their mental state, given their experience. The cognitive element also allows you to predict the other person’s mental state or behavior.

The second aspect to empathy is the “affective” component (Hobson, 1993). A similar component in other accounts has been called “emotional contagion,” defined as the tendency to automatically mimic and synchronize facial expressions, vocalizations, postures, and movements with those of another person, and, consequently, to converge emotionally (Hatfield, Cacioppo, & Rapson, 1992). This may be the most primitive component of empathy. For example, if, when witnessing someone else in a state of fear, you “catch” a similar state of fear, this acts as a quick and easy route to alerting you to environmental dangers without having to face the dangers yourself. A third component involves a “concern mechanism” (Nichols, 2001) often associated with a prosocial/altruistic component, also termed “sympathy.” This is distinct from emotional contagion as it does not necessarily involve matched states between the observer and the
person experiencing the emotion, and is possibly specific to a certain class of emotions (sadness and pain, but not disgust or happiness) in the other person. It represents a case where the observer feels both an emotional response to someone else's distress and a desire to alleviate their suffering.

**Development of Empathy**

In 1994, Baron-Cohen proposed a model to specify the neurocognitive mechanisms that comprise the “mindreading system” (Baron-Cohen, 1994, 1995). “Mindreading” is defined as the ability to interpret one’s own or another agent’s actions as driven by mental states. The model was proposed in order to explain (a) the ontogenesis of a theory of mind, and (b) the neurocognitive dissociations that are seen in children with or without autism. The model is shown in Figure 14.1 and contains four components: ID, or the intentionality detector; EDD, or the eye direction detector; SAM, or the shared attention mechanism; and, finally, ToMM, or the theory of mind mechanism.

ID and EDD build “dyadic” representations of simple mental states. ID automatically represents or interprets an agent’s self-propelled movement as a desire or goal-directed movement, a sign of its agency, or an entity with volition (Premack, 1990). For example, ID interprets an animate-like moving shape as “it wants x” or “it has goal y.” EDD automatically interprets or represents eye-like stimuli as “looking at me” or “looking at something else.” That is, EDD picks out that an entity with eyes can perceive. Both ID and EDD are developmentally prior to the other two mechanisms, and are active early in infancy, if not from birth.

SAM is developmentally more advanced. SAM automatically represents or interprets whether the self and another agent are (or are not) perceiving the same event. SAM does this by building “triadic” representations. For example, where ID can build the

dyadic representation “Mother wants the cup” and where EDD can build the dyadic representation “Mother sees the cup,” SAM can build the triadic representation “Mother sees that I see the cup.” As is apparent, triadic representations involve embedding or recursion. (A dyadic representation [“I see the cup”] is embedded within another dyadic representation [“Mother sees the cup”] to produce this triadic representation.) SAM takes its input from ID and EDD, and triadic representations are made out of dyadic representations. SAM typically functions from 9 to 14 months of age, and allows “joint attention” behaviors such as protodeclarative pointing and gaze monitoring (Scaife & Bruner, 1975).

ToMM is the jewel in the crown of the 1994 model of the mindreading system. It allows epistemic mental states to be represented (e.g., “Mother thinks this cup contains water” or “Mother pretends this cup contains water”), and it integrates the full set of mental state concepts (including emotions) into a theory. ToMM develops between 2 and 4 years of age, and allows pretend play (Leslie, 1987), understanding of false belief (Wimmer & Perner, 1983), and understanding of the relationships between mental states (Wellman, 1990). An example of the latter is the seeing-leads-to-knowing principle (Pratt & Bryant, 1990) where the typical 3-year-old can infer that if someone has seen an event, then they will know about it.

The model shows the ontogenesis of a theory of mind in the first four years of life, and justifies the existence of four components on the basis of developmental competence and neuropsychological dissociation. In terms of developmental competence, joint attention does not appear possible until 9–14 months of age, and joint attention appears to be a necessary but not sufficient condition for understanding epistemic mental states (Baron-Cohen, 1991; Baron-Cohen & Swettenham, 1996). There appears to be a developmental lag between acquiring SAM and ToMM, suggesting that these two mechanisms are dissociable. In terms of neuropsychological dissociation, congenitally blind children can ultimately develop joint (auditory or tactile) attention (i.e., SAM), using the amodal ID rather than the visual EDD route. They can therefore go on to develop ToMM. Children with autism appear able to represent the dyadic mental states of seeing and wanting, but show delays in shared attention (Baron-Cohen, 1989b) and in understanding false belief (Baron-Cohen, 1989a; Baron-Cohen, Leslie, & Frith, 1985)—that is, in acquiring SAM and ultimately ToMM. It is this specific developmental delay that suggests that SAM and ToMM are dissociable from EDD.

The 1994 model of the mindreading system was revised in 2005 because of certain omissions and too narrow a focus. The key omission is that information about affective states, available to the infant perceptual system, has no dedicated neurocognitive mechanism. In Figure 14.2, the revised model (Baron-Cohen, 2005) is shown and now includes a new, fifth component: TED, or the emotion detector. But the concept of mindreading (or theory of mind) makes no reference to the affective state in the observer triggered by recognition of another’s mental state. This is a particular problem for any account of the distinction between autism and psychopathy. For this reason, the model is no longer of “mindreading” but is of “empathizing,” and the revised model also includes a new sixth component, TESS, or the empathizing system. (TESS is spelt as it is to playfully populate
the mindreading model with apparently anthropomorphic components.) Where the 1994 mindreading system was a model of a passive observer (because all the components had simple decoding functions), the 2005 empathizing system is a model of an observer impelled toward action (because an emotion is triggered in the observer which typically motivates the observer to respond to the other person).

Like the other infancy perceptual input mechanisms of ID and EDD, the new component of TED can build dyadic representations of a special kind, namely, it can represent affective states. An example would be “Mother—is unhappy,” or even “Mother—is angry—with me.” Formally, we can describe this as agent—affective state—proposition. We know that infants can represent affective states from as early as 3 months of age (Walker, 1982). As with ID, TED is anodal, in that affective information can be picked up from facial expression or vocal intonation, “motherese” being a particularly rich source of the latter (Field, 1979). Another’s affective state is presumably also detectable from their touch (e.g., tense versus relaxed), which implies that congenitally blind infants should find affective information accessible through both auditory and tactile modalities. TED allows the detection of the basic emotions (Ekman & Friesen, 1969). The development of TED is probably aided by the simple imitation that is typical of infants (e.g., imitating caregiver’s expressions) which in itself would facilitate emotional contagion (Meltzoff & Decety, 2003).

When SAM becomes available, at 9–14 months of age, it can receive inputs from any of the three infancy mechanisms, ID, EDD, or TED. Here, we focus on how a dyadic representation of an affective state can be converted into a triadic representation by SAM. An example would be that the dyadic representation “Mother is unhappy” can be converted into a triadic representation “I am unhappy that Mother is unhappy” or “Mother is unhappy that I am unhappy,” and so on. Again, as with perceptual or volitional states, SAM’s triadic representations of affective states have this special embedded or recursive property.
TESS in the 2005 model is the real jewel in the crown. This is not to minimize the importance of ToMM, which has been celebrated for the past 20 years in research in developmental psychology (Leslie, 1987; Whiten, 1991; Wimmer, Hogrefe, & Perner, 1988). ToMM is of major importance in allowing the child to represent the full range of mental states, including epistemic ones (such as false belief), and is important in allowing the child to pull mentalistic knowledge into a useful theory with which to predict behavior (Baron-Cohen, 1995; Wellman, 1990). But TESS allows more than behavioral explanation and prediction (itself a powerful achievement). TESS allows an empathic reaction to another’s emotional state. This is, however, not to say that these two modules do not interact. Knowledge of the mental states of others made possible by ToMM could certainly influence the way in which an emotion is processed and/or expressed by TESS. TESS also allows for sympathy. It is this element of TESS that gives it the adaptive benefit of ensuring that organisms feel a drive to help each other.

To see the difference between TESS and ToMM, consider this example: I see you are in pain. Here, ToMM is needed to interpret your facial expressions and writhing body movements in terms of your underlying mental state (pain). But now consider this further example: I am devastated—that you are in pain. Here, TESS is needed since an appropriate affective state has been triggered in the observer by the emotional state identified in the other person. And where ToMM employs M-representations (“M” stands for “mental”; Leslie, 1994) of the form agent—attitude—proposition (e.g., Mother—believes—Johnny took the cookie), TESS employs a new class of representations, which we can call E-representations (“E” stands for “empathy”) of the form self—affective state—(self—affective state—proposition); for example, “I feel sorry that—I feel sad about—the news in the letter” (Baron-Cohen, 2003). The critical feature of this E-representation is that the self’s affective state is appropriate to, and triggered by, the other person’s affective state. Thus, TESS can represent “I am horrified—that you are in pain,” or “I am concerned—that you are in pain,” or “I want to alleviate—that you are in pain,” but it cannot represent “I am happy—that you are in pain.” At least, it cannot do so if TESS is functioning normally. One could imagine an abnormality in TESS leading to such inappropriate emotional states being triggered, or one could imagine them arising from other systems (such as a competition system or a sibling rivalry system), but these would not be evidence of TESS per se.

Before leaving this revision of the model, it is worth discussing why the need for this has arisen. First, emotional states are an important class of mental states to detect in others, and yet the earlier model focused only on volitional, perceptual, informational, and epistemic states. Second, when it comes to pathology, it would appear that in autism TED may function, although this may be delayed (Baron-Cohen, Spitz, & Cross, 1993; Baron-Cohen, Wheelwright, & Joliffe, 1997; Hobson, 1986), at least in terms of detecting basic emotions. Even high functioning people with autism or Asperger syndrome have difficulties both in ToMM (when measured with mental-age appropriate tests; Baron-Cohen, Joliffe, Mortimore, & Robertson, 1997; Baron-Cohen, Wheelwright, Hill, Raste, & Plumb, 2001; Happé, 1994) and TESS (Attwood, 1997; Baron-Cohen,
O’Riordan, Jones, Stone, & Plaisted, 1999; Baron-Cohen, Richler, Bisarya, Gurunathan, & Wheelwright, 2003; Baron-Cohen, & Wheelwright, 2004; Baron-Cohen, Wheelwright, Stone, & Rutherford, 1999). This suggests that TED and TESS may be fractionated.

In contrast, the psychiatric condition of psychopathy may entail an intact TED and ToMM, alongside an impaired TESS. The psychopath (or sociopath) can represent that you are in pain, or that you believe that he is the gasman, thereby gaining access to your house or your credit card. The psychopath can go on to hurt you or to cheat you without having the appropriate affective reaction to your affective state. In other words, he or she does not care about your affective state (Blair, Jones, Clark, & Smith, 1997; Mealey, 1995). Lack of guilt or shame or compassion in the presence of another’s distress are diagnostic of psychopathy (Cleckley, 1977; Hare et al., 1990). Separating TESS and ToMM thus allows a functional distinction to be drawn between the neurocognitive causes of autism and psychopathy.

Developmentally, one can also distinguish TED from TESS. We know that at 3 months of age, infants can discriminate facial and vocal expressions of emotion (Trevathan, 1989; Walker, 1982), but that it is not until about 14 months that they can respond with appropriate affect (e.g., a facial expression of concern) to another’s apparent pain (Yirmiya, Kasari, Sigman, & Mundy, 1990) or show “social referencing.” Clearly, this account is skeletal in not specifying how many emotions TED is capable of recognizing. Our recent survey of emotions identifies that there are 412 discrete emotion concepts that the adult English-language user recognizes (Golan, & Baron-Cohen, 2006). How many of these are recognized in the first year of life is not clear. It is also not clear exactly how empathizing changes during the second year of life. We have assumed that the same mechanism that enables social referencing at 14 months old also allows sympathy and the growth of empathy across development. This is the most parsimonious model, though it may be that future research will justify further mechanisms that affect the development of empathy.

**Sex Differences in Empathizing**

Some of the best evidence for individual differences in empathizing comes from the study of sex differences, where many studies converge on the conclusion that there is a female superiority in empathizing. Sex differences are best viewed as summated individual differences on multiple dimensions that include genetic and epigenetic factors. Some of the observed behavioral differences are reviewed here:

1. **Sharing and turn-taking.** On average, girls show more concern for fairness, whilst boys share less. In one study, boys showed fifty times more competition, whilst girls showed twenty times more turn-taking (Charlesworth & Dzur, 1987).
2. **Rough and tumble play** or “rough housing” (wrestling, mock fighting, and so on). Boys show more of this than girls do. Although there is a playful component, it can hurt or be intrusive, so it needs lower empathizing to carry it out (Maccoby, 1999).
3. **Responding empathically to the distress of other people.** Girls from 1 year old show greater concern through more sad looks, sympathetic vocalizations, and comforting. More women than men also report frequently sharing the emotional distress of their friends. Women also show more comforting, even of strangers, than men do (Hoffman, 1977).

4. **Using a “theory of mind.”** By 3 years old, girls are already ahead of boys in their ability to infer what people might be thinking or intending (Happé, 1995). This sex difference appears in some but not all studies (Charman, Ruffman, & Clements, 2002).

5. **Sensitivity to facial expressions.** Women are better at decoding nonverbal communication, picking up subtle nuances from tone of voice or facial expression, or judging a person’s character (Hall, 1978).

6. **Questionnaires measuring empathy.** Many of these find that women score higher than men (Davis, 1994).

7. **Values in relationships.** More women value the development of altruistic, reciprocal relationships, which by definition require empathizing. In contrast, more men value power, politics, and competition (Ahlgren & Johnson, 1979). Girls are more likely to endorse cooperative items on a questionnaire and to rate the establishment of intimacy as more important than the establishment of dominance. Boys are more likely than girls to endorse competitive items and to rate social status as more important than intimacy (Knight, Fabes, & Higgins, 1989).

8. **Disorders of empathy** (such as psychopathic personality disorder or conduct disorder) are far more common among males (Blair, 1995; Dodge, 1980).

9. **Aggression,** even in normal quantities, can only occur with reduced empathizing. Here again, there is a clear sex difference. Males tend to show far more “direct” aggression (pushing, hitting, punching, etc.), whilst females tend to show more “indirect” (or “relational,” covert) aggression (gossip, exclusion, bitchy remarks, etc.). Direct aggression may require an even lower level of empathy than indirect aggression. Indirect aggression needs better mindreading skills than does direct aggression because its impact is strategic (Crick & Grotpeter, 1995).

10. **Murder** is the ultimate example of a lack of empathy. Daly and Wilson (1988) analyzed homicide records dating back over 700 years from a range of different societies. They found that “male on male” homicide was 30–40 times more frequent than “female on female” homicide.

11. **Establishing a “dominance hierarchy.”** Males are quicker to establish these. This, in part, may reflect their lower empathizing skills because often a hierarchy is established by one person pushing others around to become the leader (Strayer, 1980).

12. **Language style.** Girls’ speech is more cooperative, reciprocal, and collaborative. In concrete terms, this is also reflected in the ability of girls to keep a conversational exchange with a partner going for longer. When girls disagree, they are more likely to express their different opinion sensitively, in the form of a question, rather than an assertion. Boys’ talk is more “single-voiced discourse” (the speaker presents their own perspective alone). The female speech style is more “double voiced discourse” (girls spend more time negotiating with the other person, trying to take the other person’s wishes into account; Smith, 1985).
13. *Talk about emotions.* Women's conversation involves much more talk about feelings, whilst men's conversation with each other tends to be more object or activity focused (Tannen, 1991).

14. *Parenting style.* Fathers are less likely than mothers to hold their infant in a face-to-face position. Mothers are more likely to follow through the child's choice of topic in play, whilst fathers are more likely to impose their own topic. And mothers fine-tune their speech more often to match what the child can understand (Power, 1985).

15. *Face preference and eye contact.* From birth, females look longer at faces, and particularly at people's eyes, and males are more likely to look at inanimate objects (Connelan, Baron-Cohen, Wheelwright, Ba'tki, & Ahluwalia, 2001).

16. Finally, females have also been shown to have better *language ability* than males. It seems likely that good empathizing would promote language development (Baron-Cohen, Baldwin, & Crowson, 1997) and vice versa, so these may not be independent.

Leaving aside sex differences as one source of evidence for individual differences, one can see that empathy is normally distributed within the population. Figure 14.3 shows the data from the Empathy Quotient (EQ), a validated, 60-item, self-report questionnaire (Baron-Cohen & Wheelwright, 2004). It has been factor analyzed in two independent studies (Lawrence, Shaw, Baker, Baron-Cohen, & David, 2004; Muncer & Ling, 2006) to suggest the existence of three distinct components, which roughly correspond

![Figure 14.3](image)

*Figure 14.3* The normal distribution of empathy in the population (dotted line). Also shown is the distribution of empathy scores from people with Asperger syndrome (AS) or high functioning autism (HFA). Note. From "The Empathy Quotient (EQ): An Investigation of Adults with Asperger Syndrome or High Functioning Autism, and Normal Sex Differences," by S. Baron-Cohen and S. Wheelwright, 2004, *Journal of Autism and Developmental Disorders*, 34, 163–175.
to the three-component model of empathy. Scores on the EQ show a continuous
distribution in several populations, with scores from people with autism spectrum
conditions (ASC) clustering toward the lower end (see Figure 14.3). The EQ is associ-
ated with significant sex differences (Goldenfeld, Baron-Cohen, Wheelwright, Ashwin, &
Chakrabarti, 2007).

The search for the neural correlates of empathy has had two traditions of research,
one focusing on theory of mind studies (largely involving intention attribution or
emotion attribution) and another focusing on action understanding. The latter has
gained considerable importance in recent years since the discovery of mirror neurons
(Rizzolatti, Fadiga, Gallese, & Fogassi, 1996).

On finding increasing evidence of sex differences in the EQ in the "normal" popula-
tion, we sought to investigate the neural correlates of this trait measure of empathizing
across the population. Since trait empathy, by definition, influences how we perceive
and respond to emotions, we attempted to marry the two fields of emotion perception
and empathizing. The following section briefly introduces the current state of the lit-
erature on the neural bases of basic emotions as well as the different processes that
contribute to the development of empathy. We then discuss a recent neuroimaging
study from our laboratory that explicitly addresses this question.

**Neuroimaging Studies of Empathizing and Emotion**

Neuroimaging studies, conducted mostly on adults, have implicated the following dif-
ferent brain areas for performing tasks that tap components of the model of empathy
proposed above, presented in order of their development (see Plate 7).

1. Studies of emotional contagion have demonstrated involuntary facial mimicry
   (Dimberg, Thunberg, & Elmehed, 2000), as well as activity in regions of the brain where
   the existence of "mirror" neurons has been suggested; for example, the inferior frontal
   gyrus, the inferior parietal lobule, and the superior temporal sulcus (Carr, Iacoboni,
   Dubeau, Mazziotta, & Lenzi, 2003; Jackson, Meltzoff, & Decety, 2005; Wicker et al., 2003).
   Dapretto and colleagues (2006) have recently demonstrated that children with ASC show
   a lower response in inferior frontal gyrus both during observation and explicit imitation
   of facial expressions of emotion, when compared to typically developing controls.

2. ID has been tested in a PET study in a task involving attribution of intentions to
cartoon characters versus predicting physical causality using the same set of characters
(Brunet, Sarfati, Hardy-Bayle, & Decety, 2000). Significantly activated regions included
the right medial prefrontal (BA 9) and inferior frontal (BA 47) cortices, superior tem-
poral gyrus (BA 42), and bilateral anterior cingulate cortex. In an elegant set of exper-
iments that required participants to attribute intentions to animations of simple
geometric shapes (Castelli, Happé, Frith, & Frith, 2000), it was found that the "inten-
tionality" score attributed by the participants to individual animations was positively
related to the activity in the superior temporal sulcus (STS), the temporoparietal
junction, and the medial prefrontal cortex. In a subsequent study (Castelli, Frith, Happé, & Frith, 2002), a group difference in activity in the same set of structures was demonstrated between people with ASC and typical controls.

3. EDD has been studied in several neuroimaging studies on gaze direction perception (Calder et al., 2002; Grosbras, Laird, & Paus, 2005; Pelphrey, Singerman, Allison, & McCarthy, 2003), and have implicated the posterior STS bilaterally. This evidence, taken together with similar findings from primate literature (Perrett & Emery, 1994), suggests that this area is a strong candidate for the anatomical equivalent of the EDD.

4. A recent imaging study (Williams, Waiter, Perrett, & Whiton, 2005) investigated the neural correlates of SAM and reported bilateral activation in anterior cingulate (BA 32, 24) and medial prefrontal cortex (BA 9, 10) and the body of caudate nucleus in a joint attention task, when compared to a control task involving non-joint attention (Frith & Frith, 2003).

5. Traditional “theory of mind” (cognitive empathy) tasks have consistently shown activity in the medial prefrontal cortex, the superior temporal gyrus, and the temporoparietal junctions (Frith & Frith, 2003; Saxe, Carey, & Kanwisher, 2004). This could be equated to the brain basis of ToM.

6. Sympathy has been relatively less investigated, with one study implicating the left inferior frontal gyrus, among a network of other structures (Decety & Chaminade, 2003). Work on “moral” emotions has suggested the involvement of a network comprising the medial frontal gyrus, the medial orbitofrontal cortex, and the STS (Moll et al., 2002).

**Neuroimaging of Discrete Emotions**

An increasing body of evidence from lesion, neuroimaging, and electrophysiological studies suggests that these affect programs might have discrete neural bases (Calder, Lawrence, & Young, 2001). Fear is possibly the most well-investigated emotion. Passive viewing of fear expressions as well as experiencing fear (as induced through recalling a fear memory or seeing fear stimuli) reliably activates the amygdala, orbitofrontal cortex, and the anterior cingulate cortex (Blair, Morris, Frith, Perrett, & Dolan, 1999; Hariri, Mattay, Tessitore, Fera, & Weinberger, 2003). There is considerable evidence from nonhuman primates (Kalin, Shelton, & Davidson, 2001) and rats (LeDoux, 2000) to suggest a crucial role for these regions in processing fear. Passive viewing of disgust faces as well as experiencing disgust oneself is known to evoke a response in the anterior insula and globus pallidus as reported in several studies (Calder et al., 2001; Phillips et al., 1997; Wicker et al., 2003). An increasing consensus on the role of the ventral striatum in processing reward from different sensory domains (receiving food rewards: O’Doherty, Deichmann, Critchley, & Dolan, 2002; viewing funny cartoons: Mobbs, Greicius, Abdel-Azim, Menon, & Reiss, 2003; remembering happy events: Damasio et al. 2000) concurs well with studies that report activation of this region in response to viewing happy faces (Lawrence et al., 2004; Phillips, Baron-Cohen, & Rutter, 1998; Phillips et al., 1998).
Perception of angry expressions has been shown to evoke a response in the premotor cortex and the striatum (Grosbras & Paus, 2006) as well as the lateral orbitofrontal cortex (Blair & Cipolotti, 2000; Blair et al., 1999). The results of studies on the processing of sad expressions are comparatively less consistent. Perception of a sad face and induction of a sad mood are both known to be associated with an increased response in the subgenual cingulate cortex (Liotti et al., 2000; Mayberg et al., 1999), the hypothalamus in humans (Malhi et al., 2004) and in rats (Shumake, Edwards, & Gonzalez-Lima, 2001), as well as in the middle temporal gyrus (Eugene et al., 2003). There have been very few studies on the passive viewing of surprise. One study by Schroeder and colleagues (2004) reported bilateral activation in the parahippocampal region, which is known for its role in novelty detection from animal literature.

While the discrete emotions model holds well for these relatively “simple” emotions, the dimensional models (Rolls, 2002) become increasingly relevant as we consider the more socially complex emotions—for example, pride, shame, and guilt—since it would not be economical to have discrete neural substrates for the whole gamut of emotions. These two models, however, need not be in conflict, since the more complex emotions can be conceptualized as being formed from a combination of the basic ones (i.e., with each of the “basic” emotions representing a dimension in emotion space).

Two major meta-analytic studies of neuroimaging literature on emotions highlight the role of discrete regions in primarily visual processing of different basic emotions (Murphy, Nimmo-Smith, & Lawrence, 2003; Phan, Wager, Taylor, & Liberzon, 2002). Some studies using other sensory stimuli (olfactory: Anderson et al., 2003; gustatory: Small et al., 2003; auditory: Lewis, Critchley, Rothstein, & Dolan, 2005) have shown the possibly dissociable role of the amygdala and the orbitofrontal cortex in processing emotions along the two dimensions of valence and arousal.

The relative absence of neuroimaging studies of “complex” emotions could be possibly due to the increased cultural variability of the elicitors, as well as the display rules that these expressions entail. Among the few exceptions, guilt and embarrassment have been investigated by Takahashi and colleagues (2004), who reported activation in the ventromedial prefrontal cortex, the left superior temporal sulcus, and higher visual cortices when participants read sentences designed to evoke guilt or embarrassment. This, taken together with the areas underlying the ToMM system, suggests an increased role of “theory of mind” to make sense of these emotions.

**Empathizing with Discrete Emotions**

Returning to the concept of individual differences in empathizing, this poses an interesting question for the brain basis of perception of discrete emotions. Do we use a centralized “empathy circuit” to make sense of all emotions? If so, can one detect differences in how discrete emotions are processed among individuals at different points on the EQ continuum?
A direct approach to investigating individual differences in empathizing has been to test for sex differences in the perception of emotions. Using facial electromyography, one study (Helland, 2005) observed that females tend to show increased facial mimicry to facial expressions of happiness and anger when compared to males. In a meta-review of neuroimaging results on sex differences in emotion perception, Wager and colleagues (Wager, Phan, Liberzon, & Taylor, 2003) report that females show increased bilaterality in emotion-relevant activation compared to males, though this is not always found (Lee et al., 2002; Schienle, Shafer, Stark, Walter, & Vaitl, 2005). One of the reasons for this might be the fact that sex differences are summated individual differences. Instead of such a broad category-based approach (as in sex-difference studies), an approach based on individual differences in self-report personality scores (Canli, Sivers, Whitfield, Gotlib, & Gabrieli, 2002) or genetic differences (e.g., Hariri et al., 2002) may be more finely tuned.

To test this model of individual variability, we asked whether an individual's score on the EQ predicted his or her response to four basic emotions (happy, sad, angry, disgust). If empathizing was modulated by a unitary circuit, then individual differences in empathizing would correlate with activity in the same structures for all basic emotions. Twenty-five adult volunteers (13 female, 12 male), selected across the EQ space, were scanned in a 3T fMRI scanner on a passive viewing task using dynamic facial expressions as stimuli. It was found that activity in different brain regions correlated with EQ scores for different basic emotions (Chakrabarti, Baron-Cohen, & Bullmore, 2005).

Using a whole-brain analysis with permutation-based techniques (XBAMM, www-bmu.psychiatry.cam.ac.uk/software/docs/xbamm/), we found that different regional responses were correlated with the EQ for different emotions. Specifically, for the perception of happy faces, a parahippocampal-ventral-striatal cluster response was positively correlated with the EQ. The role of this region in reward processing is well known (O'Doherty, 2004). This suggests that the more empathic a person is, the higher is his or her reward response to a happy face. Interestingly, the response from the same region correlated negatively with the EQ during the perception of sad faces. This fits perfectly with the earlier results; that is, the more empathic a person is, the lower is his or her reward response to a sad face.

For happy and sad faces, therefore, empathizing seems to involve mirroring. The higher a person's EQ, the stronger the reward response to happy faces and vice versa for sad faces. This is in concordance with suggestions from earlier studies on pain (Singer et al., 2004) and disgust perception (Wicker et al., 2003), where observation and experience have been shown to be mediated by the same set of structures. One of the issues with the previous studies is a possible confound between "personal distress" and empathizing. The novel element in our study is that we explicitly tested for the personality trait of empathizing in relation to the perception of specific emotions.

However, empathizing does not appear to be purely an index of mirroring. For the perception of angry faces, EQ correlated positively to clusters centered on the precuneus/inferior parietal lobule, the superior temporal gyrus, and the dorsolateral prefrontal cortex. The posterior cingulate region is known to be involved in self/other
distinction (Vogt, 2005), and the superior temporal gyrus is known for its role in ToM tasks (Saxe et al., 2004). This suggests that higher EQ corresponds to higher activation in areas related to the distinction of self versus other, as well as those that are recruited to determine another person's intentions. The dorsolateral prefrontal cortex is known for its role in decision-making and context evaluation (Rahm et al., 2006). Higher EQ would therefore predict better evaluation of the threat from an angry expression. Since expressions of anger are usually more socially urgent for attention than those of either sadness or happiness, it is essential that highly empathic persons do not merely "mirror" the expression. A high empathizer's perception of an angry face would therefore need to be accompanied by an accurate determination of the intentions of the person as well as an evaluation of the posed threat.

In response to disgust faces, a cluster containing the dorsal anterior cingulate cortex and medial prefrontal cortices is negatively correlated with EQ, suggesting that the areas involved in the attribution of mental states (primarily required for deciphering the "complex" emotions) are selectively recruited less by people of high EQ. This is what might be expected, since disgust as an emotion is less interpersonal than anger or sadness, so resources for decoding complex emotional signals need not be utilized. Another cluster that includes the right insula and inferior frontal gyrus (IFG) is negatively correlated with EQ. Given the well-established role of this region in processing disgust, this was a surprising result. We expected that an increased ability to empathize would result in an increased disgust response to facial expressions of disgust. The negative correlation suggests that people with high EQ had a lower insula-IFG response to disgust expressions. A re-examination of the behavioral literature on disgust sensitivity reveals a similar result since Haidt, McCauley, and Rozin (1994) suggested that increased socialization leads to lower disgust sensitivity. Individuals with high EQ may socialize more than those with low EQ.

The results suggest that empathizing with different basic emotions involves distinct brain regions. While some of the emotions involve more "mirroring" (the same areas show activation during recognition and experience; e.g., the striatal response to happy faces correlating positively with EQ), others require an increased distinction between one's own emotional state and another's (e.g., the superior temporal gyrus and precuneus/inferior parietal lobule response to angry faces correlating with EQ). While this explanation fits the discrete emotions model, it did not explicitly test whether there was any region that was common to all four correlation maps. To explore this, we performed a conjunction analysis for all four (emotion-neutral) versus EQ correlation plots. Using a hypothesis-driven region of interest analysis, we found a significant overlap in the left IFG-premotor cortex. This region was positively correlated with EQ for all four (emotion-neutral) contrasts.

The IFG-premotor cortex is a fundamental part of the "mirror systems" discussed earlier (Keysers & Perrett, 2004; Rizzolatti & Craighero, 2004). Several studies have shown involvement of "mirror systems" during perception of facial expressions (Buccino et al., 2001; Carr et al., 2003; Dapretto et al., 2006) and actions (Johnson-Frey et al., 2003; Molnar-Szakacs, Iacoboni, Koski, & Mazziotta, 2005) in humans. This fits
well with predictions from heuristic models that integrate perception and action (Hurley, 2005). The lower IFG-premotor response to all expressions as a function of trait empathy corroborates similar findings (Dapretto et al., 2006; Nishitani, Avikainen, & Hari, 2004). However, some studies (Carr et al., 2003; Dapretto et al., 2006) have used paradigms involving perception and explicit imitation of facial expressions and did not report any analysis for possible differences between emotions. Our analysis takes these possible differences into account and the IFG-premotor cluster emerges as a candidate region that correlates with empathy, independent of which emotion is being perceived.

This result provides a putative biomarker for empathy, a trait distributed continuously across the general population, with people with autism spectrum conditions (ASC) clustering toward the low end (Baron-Cohen & Wheelwright, 2004).

**Common and Discrete Neural Substrates of Empathy**

Comparing the results from the conjunction analysis (showing a common neural substrate of EQ across different emotions) with those from the whole-brain analysis (showing varying spatial patterns of correlation of EQ with different emotions) shows that there are both common regions that underlie empathy across different emotions and regions specific to certain emotions.

We interpret this using a model of face processing (Haxby, Hoffman, & Gobbini, 2000) applied to a discrete-emotions framework (see Figure 14.4a). At its simplest, the model proposes a core visual system for face perception. This constitutes the inferior occipital gyrus (IOG, for low-level facial feature analysis), the lateral fusiform gyrus (FG, for higher-order invariant aspects of faces such as identity), and the superior temporal sulcus (STS, for variable aspects of faces, such as lip movement and speech comprehension). This then interacts with an extended system, which involves different structures for different emotions (Haxby, Hoffman, & Gobbini, 2002). Focusing specifically on the perception of dynamic facial expressions of emotion, we propose that an intermediate module for action perception is involved, in line with similar suggestions from others (Gallese, 2003; Keysers & Perrett, 2004; Preston & de Waal, 2002; Rizzolatti & Craighero, 2004; see Figure 14.4b).

Focusing on the left of the dotted line in Figure 14.4b shows the processes that are equally influenced by trait empathy across all emotions. This includes the regions involved in face perception and the fundamental “mirror systems” used for action perception. This is revealed by the conjunction analysis, which shows a cluster that includes the IFG-premotor cortex. The common element in different facial expressions of emotion is the fact that they involve movement of eyes and mouth, which are possibly coded for by the generic “mirror systems” used for action perception. However, on investigating the interaction of each emotion with empathy, we move over to the right-hand side of the dotted line, which gives us emotion-specific correlation maps, in accordance with the discrete emotions model. We interpret these in light
of their evolutionary function. It is worth noting, though, that we do not propose a strict temporal sequence of activation from left to right of this model; nor do we represent subcortical pathways from the visual areas to the emotion-related structures. As in the original model, several of these regions are reciprocally connected and the temporal progression of activation could be mediated through reafferent projections (Iacoboni et al., 2001). These can be investigated through methods that allow better temporal resolution (e.g., magnetoencephalography) and forward-model-based connectivity analysis (e.g., dynamic causal modeling: Friston, Harrison, & Penny, 2003).

This study reveals how empathy at the neural level is subtle and complex: Neural networks activated by perception of discrete emotions depend on the observer’s EQ.
At the molecular level, empathy is likely to be determined by other individual differences, such as fetal testosterone (Castelli et al., 2000; Knickmeyer, Baron-Cohen, Raggatt, & Taylor, 2005), genetic variation (Chakrabarti, Kent, Suckling, Bullmore, & Baron-Cohen, 2006; Skuse et al., 1997), as well as early care or neglect (Fonagy, Steele, Steele, & Holder, 1997). We conclude that more basic research into the neuroscience of empathy will enrich our understanding of this most fundamental human quality.

Acknowledgments

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References


Literacy encompasses both reading and spelling. It is essentially the ability to comprehend and communicate using language expressed in visual form. The visual form may comprise an alphabet, a code in which individual speech sounds are represented by individual symbols, as in English or Russian. The visual form may comprise characters, which may represent whole words or individual syllables in the spoken form, as in Chinese or Japanese. In either case, the visual symbols represent speech written down. To become literate, the child must learn the code used by their culture for representing spoken language visually.

Accordingly, the neuropsychological development of reading is highly dependent on competent visual and auditory processing. A child who is blind needs a specially adapted script in order to read, a script that can be decoded using another sensory system (such as Braille). A child who is deaf may struggle to attain a reading age or spelling skills in line with chronological age because of oral language problems. For children without gross sensory impairments of this nature, individual differences in reading and spelling development are mainly governed by phonological skills, and not by visual skills. Although originally conceived as a visual disorder, believed to involve letters reversing themselves on the page (Hinshelwood, 1896), developmental dyslexia across languages and scripts is usually a phonological disorder. The child with developmental dyslexia has a serious and specific difficulty with the neural representation of the sounds that make up words.

In this chapter, I will first discuss the development of phonological representations in typically developing children. I will demonstrate how phonological skills determine and are then shaped by literacy acquisition across languages. The focus will be on data gathered by cognitive developmental psychologists. To date, there is very little genetic and imaging data concerning the development of reading in typically developing children. Most genetic and imaging data have come from studies of adults with developmental dyslexia. I will thus first discuss developmental dyslexia within the cognitive framework developed for typical reading development, and then discuss associated
imaging and genetic studies. As will become clear, the view that individual differences in literacy development are governed by individual differences in a child's awareness of phonology is accepted across languages and cultures.

Typical Neuropsychological Development of Reading across Languages

The phonological structure of language

The child's awareness of the phonological structure of their language, measured before schooling, is the strongest predictor that we have of how well a particular child will learn to read and to spell. The construct of "phonological awareness" is usually defined as the ability to detect and manipulate component sounds in words. Component sounds can be defined at a number of different linguistic levels or "grain sizes." For example, a single word can comprise a number of syllables (caterpillar has four syllables, wigwam has two syllables). One word can rhyme with another. A word like fountain rhymes with mountain because the words share their phonology after the first sound (this sound is called the onset). A word like street rhymes with eat even though street has an onset made up of three sounds before the part of the word that rhymes with eat. This onset comprises three phonemes. The rhyming part of the syllable is called the rime by linguists. There are two shared rimes in fountain and mountain, the soundsount andain. There is one shared rime in street and eat, the sound eet. Phonemes are the smallest units of sound in words that change meaning. For example, street differs in meaning from treat because it has one extra phoneme (the phoneme /s/). Phonemes usually correspond to alphabetic letters: The alphabet is a code that works primarily at the phoneme level of phonology. However, the primary phonological processing unit across most of the world's languages is the syllable.

In most languages of the world, syllables follow what linguists call a simple structure. Syllables consist of a consonant (C) and a vowel (V). The CV syllable structure characterizes spoken languages as diverse as Finnish, Italian, and Chinese. Clearly, not all languages with a CV syllable structure use the alphabet as their visual code, although many do. Cross-language research suggests that it is easier to become phonologically aware in languages like Spanish and Italian. This is because dividing a syllable into its component sounds is relatively easy. Most syllables contain two sounds, an onset comprising a single phoneme, and a rime comprising a single phoneme. In fact, for languages with a CV syllable structure, there is no distinction between phonemes, onsets, and rimes. This makes it easier to become phonologically aware.

For English monosyllables, only 5% follow the CV pattern (De Cara & Goswami, 2002). Examples are words like sea and go. The majority of monosyllables in English (43%) follow a CVC structure (dog, soap, hill). The next most frequent structure, CVCC, accounts for an additional 21% of monosyllables (last, felt, jump). A further 15% of monosyllables follow a CCVC structure (trip, clap, broom). Dividing up complex syllables like
these into their constituent sounds is more challenging for young children. Even for CVC syllables, there are two phonemes in the rime. Consonant clusters pose particular difficulties (jump, trip, street, spring); some clusters may contain three phonemes. These aspects of English phonology make it more challenging for English-speaking children across English-speaking countries to develop phonological awareness.

The development of phonological awareness across languages

Most theorists assume that phonological awareness develops from the phonological representations that underpin spoken language. During the first four or five years of their linguistic development, children are acquiring spoken language, not written language. Their focus is communication, and not phonological awareness, and they know about phonology at an implicit level, by being competent users of their language. Whereas the average 1-year-old might have a productive vocabulary of around 100 words, by the age of 6 it is estimated that the average child's vocabulary contains around 14,000 words (Dollaghan, 1994). All of these words need to be represented as phonologically distinct from each other. Clearly, most children achieve this, as typically developing children seldom produce the wrong word during communication, apart from slips of the tongue (which adults produce as well). A pre-reading child in fact recognizes phonological similarities that, as literate adults, we no longer notice. For example, a pre-reading child will judge correctly that chair and train share more phonological similarity at onset than train and tip (Read, 1986). As literate adults, we have lost this insight. This is because orthography tells us that train and tip begin with the same phoneme, /t/.

There seems to be a universal sequence in the development of phonological awareness, even though the phonology of languages differs. Despite the differences in syllable structure and in the absolute number of phonemes found in the phonological inventories of different languages (e.g., 44 phonemes in English compared to 21 phonemes in Finnish), children in all languages so far studied appear to follow a similar developmental pathway in terms of phonological awareness. Children first become aware of relatively large sounds in words, such as syllables. They then become aware of the onset/rime division of the syllable (str-eet, j-ump, tr-ip). Awareness of phonemes develops later in children learning to speak languages with a complex phonology, such as English and German. Pre-readers in English and German are aware of onsets and rimes, but they cannot recognize or manipulate phonemes (Goswami, Ziegler, & Richardson, 2005). However, pre-readers in languages like Italian and Spanish do not have a problem. For them, onsets and rimes are also phonemes because of the simple phonology of the CV syllable. The hierarchical organization of syllable structure is shown in Table 15.1.

The development of phonological awareness appears to respect this hierarchical organization. Empirical data for a developmental progression comes from studies that measure children’s phonological skills at different points in development in different languages. Some of these studies have also demonstrated longitudinal
Table 15.1 Hierarchical Phonological Structure for the Early Acquired Word “Girl” in English and Italian.

<table>
<thead>
<tr>
<th>Linguistic level</th>
<th>English</th>
<th>Italian</th>
</tr>
</thead>
<tbody>
<tr>
<td>Word</td>
<td>Girl</td>
<td>Ragazza</td>
</tr>
<tr>
<td>Syllable</td>
<td>Girl</td>
<td>Ra ga za</td>
</tr>
<tr>
<td>Onset/rime</td>
<td>G irl</td>
<td>Ra ga za</td>
</tr>
<tr>
<td>Phoneme*</td>
<td>G  ir  l</td>
<td>Ra ga za</td>
</tr>
</tbody>
</table>

*The phoneme represented as “ir” is phonetically a single vowel.

connections between phonological awareness measured at time 1, and reading measured at time 2. These longitudinal connections have been found across languages. A few studies have demonstrated that the connection between phonological awareness and literacy is a causal one. These studies involve training children's phonological skills, and demonstrating an impact on literacy. As space precludes a thorough discussion of the relevant empirical data, a more comprehensive review can be found in Ziegler and Goswami (2005).

The sequence of phonological development

A large variety of tasks has been invented to measure the development of phonological awareness in young children. For example, children may be asked to monitor and correct speech errors (e.g., sie to pie), to select the “odd word out” in terms of sound (e.g., which word does not rhyme: cat, bat, sit), to make a judgment about sound similarity (e.g., do these two words share a syllable? compete, repeat), to count sounds in words by tapping with a stick (e.g., tap out the component sounds in soap = 3 taps), and to blend sounds into words (e.g., d-ish or d-i-sh to make dish; see, for example, Bradley & Bryant, 1983; Chaney, 1992; Liberman, Shankweiler, Fischer, & Carter, 1974; Metsala, 1999; Treiman & Zukowski, 1991). These different tasks also make differing cognitive demands on young children. For example, “same—different” judgments (compete—repeat = same) are often considered to be easier than oddity tasks (see Treiman & Zukowski, 1991). The best way to investigate the sequence of phonological development is to equate the cognitive demands of the chosen task across linguistic level. For example, an oddity task or a same—different judgment task can be used to compare both onset/rime and phoneme levels of awareness (see Goswami et al., 2005; Treiman & Zukowski, 1991).

Surprisingly, it is rare to find research studies that have used the same cognitive task to study the emergence of phonological awareness at the different linguistic levels of syllable, onset/rime, and phoneme. The most comprehensive studies in English are those conducted by Anthony and his colleagues (Anthony & Lonigan, 2004; Anthony et al., 2002; Anthony, Lonigan, Driscoll, Phillips & Burgess, 2003). For example, Anthony et al. (2003) used blending and deletion tasks at the word, syllable, onset/rime, and phoneme
level. They studied a large group of more than 1,000 children, and included a much wider age range than many studies (2–6 years). Anthony et al. (2003) found that the development of children’s phonological awareness followed the hierarchical model shown in Table 15.1. English-speaking children generally mastered word-level skills before they mastered syllable-level skills; they mastered syllable-level skills before onset/rime-level skills; and they mastered onset/rime-level skills before phoneme skills.

This developmental progression from syllable awareness via onset/rime awareness to phoneme awareness has been mirrored by many other studies conducted in English using a broad variety of tasks (see Goswami & Bryant, 1990, for a survey). Counting and oddity tasks have been particularly useful for comparisons across languages. Focusing usually on syllable versus phoneme awareness (because of the typical CV syllable structure), such studies demonstrate that syllable awareness emerges prior to phoneme awareness in children learning all languages so far studied. For example, Cossu and colleagues studied the development of syllable versus phoneme awareness in Italian preschoolers and school-aged children using a counting task (comparing groups of 4-year-olds, 5-year-olds, and 7–8-year-olds). Syllable awareness was shown by 67% of the 4-year-olds, 80% of the 5-year-olds, and 100% of the school-aged sample (Cossu, Shankweiler, Liberman, Katz, & Tola, 1988). Phoneme awareness was shown by 13% of the 4-year-olds, 27% of the 5-year-olds, and 97% of the school-aged sample. A similar study was carried out by Liberman and her colleagues (1974) with American children. Children aged from 4 to 6 years were asked to tap once for words that had either one syllable or phoneme (dog, i), twice for words that had two syllables or phonemes (dinner, my), and three times for words that had three syllables or phonemes (president, book). Syllable awareness was shown by 46% of the 4-year-olds, 48% of the 5-year-olds, and 90% of the 6-year-olds. The 4- and 5-year-olds were pre-readers, and the 6-year-olds had been learning to read for about a year. Phonemic awareness was shown by 0% of the 4-year-olds, 17% of the 5-year-olds, and 70% of the 6-year-olds. Liberman et al. (1974) concluded that whereas syllabic awareness was present in pre-readers, phonemic awareness was dependent on learning to read. This finding for English has now been replicated many times (see Ziegler & Goswami, 2005).

Longitudinal connections between phonological awareness and reading

The existence of a longitudinal connection between individual differences in children’s phonological awareness measured prior to schooling and their later progress in reading and spelling was first demonstrated in a seminal study in Danish carried out by Lundberg, Olofsson, and Wall (1980). Similar results were reported for English by Bradley and Bryant (1983), who demonstrated the importance of onset/rime awareness measured in pre-readers for subsequent reading development using the oddity task. Longitudinal studies in other languages are also finding significant relationships between phonology and reading (e.g., German: Schneider, Roth, & Ennemoser, 2000; Norwegian: Høien, Lundberg, Stanovich, & Bjaalid, 1995; Chinese: Ho & Bryant, 1997). I will only describe the study in English by Bradley and Bryant here.
In their study, oddity tasks at the onset and rime level were administered to 400 preschoolers when they were aged 4 and 5 years. The same children’s progress in reading and spelling was then measured 2–3 years later. Examples of the oddity tasks are *sun*, *sock*, *rag* (onset) or *pin*, *win*, *sit* (rime). Bradley and Bryant reported that onset/rime awareness was a significant predictor of the children’s progress in reading and spelling when measured at 8 and 9 years of age. This longitudinal correlation remained significant even when other factors such as IQ and socioeconomic status were controlled in multiple regression equations. It was also specific to reading, as no significant longitudinal correlations were found for development in mathematics. Subsequently, MacLean, Bryant, and Bradley (1987) reported a significant connection for English between rhyming skills at the age of 3 measured via nursery rhyme knowledge and single word reading at 4 years and 6 months. When the same sample was followed up two years later, Bryant, MacLean, Bradley, and Crossland (1990) found a significant relationship between nursery rhyme knowledge at the age of 3 and success in reading and spelling at the ages of 5 and 6. This relationship was significant even after factors such as social background and IQ were controlled in multiple regression equations.

While these controls for possible intervening variables are very important, however, they do not in themselves guarantee that the longitudinal connection between phonology at time 1 and reading at time 2 is a causal one. In order to demonstrate a causal connection, a training study is required. If early phonological awareness has a direct effect on how well a child learns to read and spell, then training children to discover and attend to the phonological structure of language should have a measurable impact on their reading progress. In fact, Bradley and Bryant (1983) included a training component in their longitudinal study. They selected for training the 60 children in their cohort of 400 who had performed most poorly in the oddity task at 4 and 5 years of age. The children were given two years of intervention, which largely comprised grouping words on the basis of sounds by using a picture sorting task. The children were taught to group words by onset, rime, vowel, and coda (syllable-final) phonemes (for example, placing pictures of a *hat*, a *rat*, a *mat*, and a *bat* together for grouping by rime). A control group learned to sort the same pictures into semantic categories (e.g., farmyard animals). In addition, half of the experimental group also learned to match plastic letters to the shared phonological units in the words in the pictures; for example, making the spelling unit *at* for words like *hat*, *rat*, and *mat*.

Following the intervention, the children in the experimental group which had had plastic letters training were 8 months further on in reading than the children in the semantic control group, and 12 months further on in spelling, even after adjusting post-test scores for age and IQ. A second control group was “unseen,” comprising children who had spent the intervening period receiving normal classroom teaching without an additional intervention. Compared to these children, the experimental group was a remarkable 24 months further on in spelling, and 12 months further on in reading. Positive effects of intervention have also been reported by large-scale training studies with Danish children (Lundberg, Frost, & Petersen, 1988) and German children (Schneider, Kuespert, Roth,
Vise, & Marx, 1997). In both the English and German studies, particularly strong effects of phonological training were reported for progress in spelling.

In summary, cross-sectional, longitudinal, and training studies conducted across languages have shown that the links between phonological awareness and literacy appear to be language-universal. In all languages so far studied, phonological awareness progresses from an awareness of large units of sound, such as syllables and rimes, to an awareness of small units of sound—phonemes. In all languages so far studied, including character-based scripts, individual differences in phonological awareness are predictive of individual differences in literacy. Finally, in all languages so far studied, providing children with training in phonological awareness which is coupled with training in how letters represent sounds has a measurable positive impact on progress in literacy. Let us now consider how differences in the phonological structure of different languages might affect the ways in which early phonological awareness can support the acquisition of literacy.

The acquisition of reading and spelling skills across languages

As noted at the beginning of this chapter, the primary phonological unit in language is the syllable. Furthermore, many of the world's languages have a simple syllabic structure, comprising largely CV syllables. For children learning to speak these languages, the onset/rime level of phonological structure and the phoneme level represented by the alphabet are the same. Other languages, like English and German, have a complex phonological structure. These languages allow syllables to contain clusters of consonant phonemes. These clusters can be either before the vowel (spray, street), after the vowel (jump, sand), or both (stamp, clasp). For languages like these, the onset/rime level of phonological structure and the phoneme level are not the same. In languages like these, children typically arrive in school with good onset/rime awareness, but lacking phoneme awareness (Goswami & Bryant, 1990; Goswami et al., 2005). Phoneme awareness must be learned, and it is usually learned via letters.

Learning letters helps to develop phoneme awareness in all languages. This is easily shown by cross-language studies demonstrating phoneme awareness in pre-readers versus readers of different ages (see Ziegler & Goswami, 2005). A few studies have also demonstrated that illiterate adults, who have never learned to read, lack phoneme awareness (e.g., Morais, Cary, Alegría, & Bertelson, 1979). However, for most European languages, acquisition of phoneme awareness is very rapid once children begin learning to read. For English-speaking children, it is not. The reasons appear to be twofold. It is relatively easy to learn about phonemes when your language has a CV syllable structure, and a transparent orthography. It is relatively difficult to learn about phonemes when your language has a complex syllable structure, and an ambiguous orthography. Unfortunately for English-speaking children, cross-language analyses show that English is particularly ambiguous with respect to both spelling-to-sound and sound-to-spelling relations (Ziegler, Stone, & Jacobs, 1997). In English, a single letter or letter cluster can have multiple pronunciations (e.g., the letter a in car, cat, cake, call; the
cluster *ough* in *cough, bough, through*. A phoneme can also have multiple spellings (consider the vowel sound in *hurt, dirt*, and *Bert*; or the sound /f/ in *photo* versus *off* versus *cough*). Some languages, like Italian, are remarkably consistent in both directions. Other languages, like German, have a complex phonology but an orthography that is transparent for reading, although not for spelling.

Cross-language studies of literacy acquisition show that these factors have systematic effects on the acquisition of reading. Children who are learning to read a language with a simple CV syllable structure and a transparent orthography, like Finnish, learn to read simple words efficiently within weeks of arriving in school (Seymour, Aro, & Erskine, 2003). Children learning to read English take much longer to become competent at decoding, despite their earlier start. In England, we begin to teach reading at the age of 4 years. In some European countries, such as the Scandinavian countries, reading instruction can commence as late as 7 years. Nevertheless, children in such countries become highly efficient at decoding quite rapidly following the onset of teaching. This was shown, for example, by the largest and most consistent cross-language study of early reading acquisition to date, the study reported by Seymour et al. (2003). They reported the outcome of a 14-language study carried out by scientists participating in the European Concerted Action on Learning Disorders as a Barrier to Human Development.¹ These scientists developed a matched set of simple real words and nonwords across languages that were given to children to read during their first year of schooling in the different EU member states. Participating schools all used a “phonics-based” instructional approach to reading. A summary of the results is reported in Table 15.2.

As is clear from Table 15.2, decoding accuracy approached ceiling levels in many European languages during the first year of schooling (Greece, Finland, Italy, Spain). All of these languages have a transparent spelling system. It was not close to ceiling in four languages: French, Portuguese, Danish, and English. All of these languages have less transparent spelling systems. English, in particular, has a very inconsistent orthography, as described above. The English-speaking children (a Scottish sample—Scotland traditionally has strong phonics teaching for early reading) performed particularly poorly, reaching 34% accuracy for real word reading and 29% accuracy for simple nonsense words (like *eb* and *fip*). It is interesting to compare the performance of the English children with that of the German children, as English and German have the same linguistic root. In fact, many of the words in English and German are the same words (*wine/Wein*, *mouse/Maus*, *garden/garten*). Nevertheless, German has a consistent orthography. The German children attained 98% accuracy for real word reading, and 94% accuracy for nonsense word reading. These differences in reading acquisition by

¹ National representatives of this action were: H. Wöhrer, T. Reinelt (Austria), J. Alegria, J. Morsis, J. Leybaert (Belgium), C. Elbro, E. Arnbak (Denmark), H. Lyttingen, P. Niemi (Finland), J.-E. Gombert, M.-T. Le Normand, L. Sprenger-Charolles, S. Valdois (France), A. Warnke, W. Schneider (Germany), C. Porpodas (Greece), V. Gsepe (Hungary), H. Ragnarsson (Iceland), C. Cornoldi, P. Giovanardi Rossi, C. Vio, P. Tressoldi, A. Parmeggiani (Italy), C. Firma (Malta), R. Licht, A. M. b. De Groot (Netherlands), F. E. Tonnessen (Norway), L. Castro, L. Cary (Portugal), S. Defor, F. Martos, J. Sainz, X. Angerri (Spain), S. Stromqvist, A. Olofsson (Sweden), P. Seymour, P. Bryant, U. Goswami (United Kingdom).
Table 15.2 Data (% correct) from the COST A8 Study of Grapheme–Phoneme Recoding Skills for Monosyllables in 14 European Languages.

<table>
<thead>
<tr>
<th>Language</th>
<th>Familiar, real words</th>
<th>Nonwords</th>
</tr>
</thead>
<tbody>
<tr>
<td>Greek</td>
<td>98</td>
<td>92</td>
</tr>
<tr>
<td>Finnish</td>
<td>98</td>
<td>95</td>
</tr>
<tr>
<td>German</td>
<td>98</td>
<td>94</td>
</tr>
<tr>
<td>Austrian German</td>
<td>97</td>
<td>92</td>
</tr>
<tr>
<td>Italian</td>
<td>95</td>
<td>89</td>
</tr>
<tr>
<td>Spanish</td>
<td>95</td>
<td>89</td>
</tr>
<tr>
<td>Swedish</td>
<td>95</td>
<td>88</td>
</tr>
<tr>
<td>Dutch</td>
<td>95</td>
<td>82</td>
</tr>
<tr>
<td>Icelandic</td>
<td>94</td>
<td>86</td>
</tr>
<tr>
<td>Norwegian</td>
<td>92</td>
<td>91</td>
</tr>
<tr>
<td>French</td>
<td>79</td>
<td>85</td>
</tr>
<tr>
<td>Portuguese</td>
<td>73</td>
<td>77</td>
</tr>
<tr>
<td>Danish</td>
<td>71</td>
<td>54</td>
</tr>
<tr>
<td>Scottish English</td>
<td>34</td>
<td>29</td>
</tr>
</tbody>
</table>


English versus German children have also been demonstrated in smaller studies with careful cognitive matching of the participants (Frith, Wimmer, & Landerl, 1998; Wimmer & Goswami, 1994).

Clearly, the normal neuropsychological development of reading differs depending on the language that you are learning to read. The same cognitive factor underpins successful reading acquisition in all languages so far studied, namely phonological awareness. However, phonological skills interact with orthographic transparency. Children who are learning to read languages with a simple phonological structure and a transparent orthography do particularly well (Finnish, Spanish). Children who are learning to read languages with a complex phonological structure but a transparent orthography also do well (German, Welsh—not in Table 15.2, but see Ellis & Hooper, 2001). Children who are learning to read a language like English, which has a complex phonological structure and a particularly inconsistent orthography, take longer to acquire efficient decoding skills. But for typically developing children, this longer learning process is easily explained. Literacy is a more difficult “learning problem” in a language like English. By the age of around 10 years, and sometimes earlier, differences in decoding efficiency between English children and those learning to read in other languages have disappeared (e.g., Goswami, Gombert, & de Barrera, 1998). In fact, the better English readers have done rather well in recent international comparisons. We turn now to consider children who have specific difficulties with reading, children with developmental dyslexia. For these children, having to learn to read a language like English has rather different consequences.
Atypical Neuropsychological Development of Reading across Languages

The cognitive framework developed above to explain the typical neuropsychological development of reading across languages makes some clear predictions regarding atypical development. One key prediction is that a specific difficulty in representing the phonological structure of words should manifest itself differently in reading depending on the language that you are learning to read. For example, if you are learning to read a language like Italian, which has a simple CV syllable structure and a transparent orthography, having a specific difficulty with phonology might not have a very large impact on your reading. Once you learn about letters, the 1:1 relationship between these letters and sounds might help you to improve your deficient phonological representations. The highly predictable relationship between spelling and sound also enables reading accuracy, as children with developmental dyslexia are of average intelligence. They can learn letter–sound relationships if the relationships are 1:1, even if they do so rather slowly. If you are learning to read a language like German, which has a complex syllable structure but a transparent orthography, having a specific difficulty with phonology might also have a relatively minor impact on your eventual ability to read. Again, the 1:1 relationship between letters and sounds should help to improve your phonological abilities and enable accurate reading. However, if you are learning to read a language like English, which has a complex syllable structure and a highly inconsistent orthography, a specific difficulty with phonology might have rather profound consequences. Learning letters will not be particularly helpful for phonological awareness, and recoding letters to sound seems likely to remain problematic given the highly variable relationships that will be encountered. The research base in developmental dyslexia supports exactly this picture of atypical development.

Phonological awareness in developmental dyslexia

Cognitive studies across languages demonstrate that children with developmental dyslexia in all languages so far studied have difficulties with phonological awareness tasks. They find it difficult to count syllables, to recognize rhyme, to decide whether words share phonemes, and to substitute one phoneme for another (e.g., Korean: Kim & Davis, 2004; German: Wimmer, 1996; Greek: Porpodas, Pantelis, & Hantziou, 1990; Hebrew: Share & Levin, 1999; see Ziegler & Goswami, 2005, for a comprehensive review). Numerous studies in English, in particular, have shown that children with developmental dyslexia remain poor at tasks such as deciding whether words rhyme (Bradley & Bryant, 1978), making accurate judgments in counting, or same–different judgment tasks at the different linguistic levels of syllable, onset/rime, and phoneme (Swan & Goswami, 1997), making oddity judgments about phonemes (Bowey, Cain, & Ryan, 1992), and Spoonerism tasks (Landerl, Wimmer, & Frith, 1997). These difficulties persist into the teenage years (e.g., Bruck, 1992).
However, for dyslexic children who are learning to read transparent orthographies, reading has an impact on phonological awareness. Learning consistent letter–sound relationships appears to help children to specify phonological similarities and differences between words. For example, German dyslexic children show age-appropriate phonological skills in some phonological awareness tasks (those that can be solved using letters) by the age of 10 years. A Spoonerism task is an example of a phonological awareness task that can be solved using orthography. In Spoonerism tasks, the child has to swap onsets in words (like Reverend Spooner, who told students “You have hissed all my mystery lectures”). For example, the child may have to say “Dob Dylan” instead of “Bob Dylan.” German dyslexic children eventually became able to do such tasks as well as control children (Wimmer, 1993). However, when German dyslexic children are compared to matched English dyslexic children, the Germans perform much better in Spoonerism tasks (Landerl et al., 1997).

Acquisition of literacy skills in developmental dyslexia

Despite measurable phonological difficulties in children with developmental dyslexia across languages, for most of the world's languages, differences in the accuracy of decoding print compared to age-matched peers is only found in the very earliest stages of reading. Studies of young Greek and German children who later turned out to have specific reading difficulties showed that word and nonsense word reading was significantly poorer than that of age-matched controls in the first year of reading instruction, but this difference soon disappeared (Porpodas, 1999; Wimmer, 1993). Although difficulties with phonology remain for children learning to read transparent orthographies, they do not impede reading accuracy. Rather, they impede reading speed and spelling accuracy. Developmental dyslexia in most languages other than English is usually diagnosed on the basis of extremely slow and effortful reading, and strikingly poor spelling.

For English children, however, developmental dyslexia is characterized by both inaccurate and effortful reading and by inaccurate spelling. Even dyslexic adults in English remain poor at decoding words accurately (Bruck, 1992). Hence the same cognitive deficit can manifest differently in different languages. In fact, English children with developmental dyslexia perform significantly more poorly in tasks such as nonsense word reading compared to German children with developmental dyslexia, even when they are trying to read the same items (e.g., nonsense words like grall, see Landerl et al., 1997). This demonstrates the pervasive effects of learning to read an inconsistent orthography on reading development. The consequences of having a phonological deficit are more profound in developmental terms for literacy in English.

Behavioral genetics

The phonological deficit that characterizes dyslexia across languages appears to be heritable. The heritability of dyslexia has been demonstrated by a number of family and
twin studies, particularly in English (e.g., Gayan & Olson, 2001; Gayan et al., 1999). Linkage studies have been used to try to determine where in the human genome the critical genes for dyslexia are located. The most promising findings so far concern the short arm of chromosome 6, and sites on chromosome 15 (e.g., Grigorenko et al., 1997). These studies depend on definitions of the dyslexic phenotype that are based on deficits in phonological awareness tasks and single or nonsense word reading. The dyslexic “phenotype” is defined in terms of phonology. Of course, there cannot be a “gene” for dyslexia in the sense that there is a gene for eye color, as reading is a culturally determined activity. Individuals at genetic risk for dyslexia who develop in a favorable early environment as far as reading is concerned (for example, children whose carers actively develop their phonological awareness via language games, nursery rhymes, and so on; and who read books to them and model and encourage literacy activities by reading and writing extensively themselves) may be able to compensate to some extent for their genetic predisposition to dyslexia. Other individuals with a lower degree of risk but relatively adverse early environments may be more handicapped. Nevertheless, no candidate gene so far identified can be described as a “gene for reading.” Levels of association reported so far in behavioral and molecular genetics are not strong enough to translate into reliable predictors of risk for a single individual (Fisher & Francks, 2006).

**Neuroimaging of Typical and Atypical Readers**

Reading and phonological processing in adults depends on a left-lateralized network of frontal, temporoparietal, and occipitotemporal regions. Studies of aphasia long ago revealed the importance of Broca’s area (inferior frontal gyrus) and Wernicke’s area (posterior superior temporal gyrus) for the motor production and receptive aspects of speech, respectively, and these areas appear to be recruited along with visual and frontal areas by literacy. In normally reading adults, remarkable overlap is found in the neural networks underpinning reading, whether the adult has learned to read a consistent orthography like Italian, an inconsistent orthography like English, or a character-based orthography like Chinese (Paulesu et al., 2001; Siok, Perfetti, Jin, & Tan, 2004). This has led researchers such as Paulesu to propose that a common neurological network underpins dyslexia across languages. Studies of adults with developmental dyslexia suggest that phonological mechanisms are localized in the temporoparietal junction (see Eden & Zeffiro, 1998, for review). When performing tasks like rhyme judgment, rhyme detection, and word and nonsense word reading, dyslexic adults show reduced activation in temporal and parietal regions, particularly within the left hemisphere.

Work by Turkeltaub and colleagues has explored the neural underpinning of phonological processing in children (Turkeltaub, Gareau, Flowers, Zeffiro, & Eden, 2003; see Plate 8). Phonological awareness appears to depend on a network of areas in the left posterior superior temporal cortex. Activity in this region is modulated by the level of children’s phonological skills. The rapid output of phonological information appears to depend on a different, bilateral network, including right posterior superior temporal
gyrus, right middle temporal gyrus, and left ventral inferior frontal gyrus. The left posterior temporal sulcus is also the primary area recruited by young children at the beginning of reading development. As reading skills develop, an area termed the “visual word form area” becomes more engaged (Cohen & Dehaene, 2004). This area is also active for nonsense word reading, hence it probably stores orthography–phonology connections at different grain sizes (Goswami & Ziegler, 2006). Children with developmental dyslexia show reduced activation in the normal left hemisphere sites, and atypical engagement of right temporoparietal cortex. If targeted remediation is provided, usually via intensive tuition in phonological skills and in letter–sound conversion, activity in the left temporal and parietal areas appears to normalize (e.g., Simos et al., 2002). These studies have all been carried out with dyslexic children learning to read in English. Studies with children learning to read in other languages have yet to be done.

Implications of Current Research

Research suggests that while brains are similar across languages, orthographies are not. In all languages so far studied, one aspect of language acquisition is the development of phonological awareness. Individual differences in phonological awareness determine individual differences in the development of reading and spelling. Literacy, in turn, results in the further development of phonological awareness. Indeed, the demonstration of the importance of phonological awareness for literacy has been hailed as the success story of developmental psychology (see Adams, 1990; Lundberg, 1991; Stanovich, 1992). Specific characteristics of this developmental relationship vary with language, but the variation is systematic, depending on phonological complexity and orthographic consistency. One important factor is the “grain size” or unit of analysis (e.g., the syllable is a large grain size, the phoneme is small grain size). There are predictable developmental differences in the ease with which phonological awareness at different grain sizes emerges across orthographies, in the grain size of lexical representations across orthographies, and in developmental reading strategies across orthographies (see Ziegler & Goswami, 2005). Future research in additional languages using this “psycholinguistic grain size” framework will be very useful in pinpointing universal causal mechanisms for literacy.

The importance of phonology for reading and spelling means that clinical assessments of reading and reading difficulties rely on phonological tests. One of the most widely used test batteries is the Phonological Assessment Battery (PhAB), which provides standardized age norms for different measures of phonological awareness such as rhyme fluency and Spoonerisms (Fredrickson, Frith, & Reason, 1997). Other tests which purport to measure a wider range of skills, such as the Dyslexia Early Screening Test (Nicolson & Fawcett, 1996) also rely heavily on phonological subtests. The development of single word reading, spelling, and reading comprehension skills can also be measured by a range of standardized tests, such as the British Ability Scales (Elliott, 1996) and the Neale Analysis of Reading Ability (Neale, 1989). In terms of intervention,
the most successful interventions remain those that work directly on the component skills of phonology and letter–sound relationships. Although claims are sometimes made for interventions based on remediating motor skills (e.g., the Dyslexia Dyspraxia Attention Deficit Treatment [DDAT]; Reynolds, Nicolson, & Hambly, 2003) or disturbances of the visual system (e.g., Irlen lenses), research on their effectiveness remains equivocal (e.g., Snowling & Hulme, 2003). Most children who present in the clinic with developmental dyslexia are best served by systematic teaching of phonological skills, letter–sound relationships, and the larger orthographic patterns that enable consistency in English spelling–sound relations.

References


