The Cognitive and Behavioral Manifestations of Blindness in Children

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Introduction

Ground-breaking progress in the field of neuro-developmental disorders has allowed us far more insight into blindness and visual impairment (VI). The field of cognitive neuroscience has now established itself solidly in the literature, combining the knowledge from cognitive psychology, clinical studies related to brain damage and neuroscience to open the way to significant advances in understanding. In recent years the optimism engendered from the adult studies has played a large part in providing the impetus for developmental studies and in particular developmental neuroscience (Johnson, 2005; Tager-Flusberg, 1999). It is from this developmental neuroscience perspective that we can begin to understand the cognitive and behavioral manifestations associated with blindness and visual impairment; albeit with the proviso that children with VI present particularly heterogeneous developmental patterns when compared to typically developing children (Fraiberg, 1971).

In considering the effects of blindness and visual impairment this chapter will focus on social understanding, language, cognition and motor-development. However it begins with brief introductions to epidemiology and the effects of blindness on the functional and structural organization of the brain, which it is hoped will provide a useful context in which to consider the development of children who cannot see.
Today the number of blind people in the world stands at some 45 million even though up
to 75% of blindness could be avoided either by treatment or by prevention. The number
of people with avoidable blindness will have doubled from 1990 to 2020 unless there is
rapid and effective intervention, and the total number of the blind is projected to be as
many as 76 million by 2020. To prevent this scenario, the World Health Organization
(WHO) and the IAPB International Agency for the Prevention of Blindness (IAPB) have
jointly initiated Vision 2020, a project that aims to eliminate the main causes of
avoidable blindness by the year 2020 with the ultimate long-term goal of a world in
which all avoidable blindness is eliminated and in which everyone with unavoidable
vision loss reaches their full potential.

The top priority of Vision 2020 is the prevention of childhood blindness. At present there
are 1.4 million children under the age of 15 who are blind. Around 500,000 children
become blind each year, 75% of them in developing countries. Shockingly, up to 60% of
these die within a year of losing their sight. The survivors will not only have a lifetime of
blindness to contend with but will also be adversely affected in terms of their emotional,
social and psychomotor development. Blindness in children is complex, requiring multi-
disciplinary collaboration from community, educational and medical services. Sight
restoration and blindness prevention programs are among the most cost-effective
interventions in health care and some 40% of the causes of childhood blindness are
preventable or treatable.
The epidemiology of pediatric blindness clearly reflects socio-economic development. The prevalence ranges from 3/10,000 in affluent societies to 15/10,000 in the least affluent. The main cause of childhood blindness in the developing world is corneal opacification resulting from measles, Vitamin A deficiency and the use of traditional eye medicines whereas the main causes in the USA are cortical visual impairment, retinopathy of prematurity and optic nerve hypoplasia. At the other end of the spectrum in older age commonly reported causes of eye problems are cataracts, glaucoma, general ill-health and diabetes.

Congenital impairment certainly has different outcomes from late blindness and it is important for clinicians to distinguish cerebral from peripheral disorders (Dale & Sonksen, 2002). The cerebral congenital disorders of the visual system are more common and are associated with additional disabilities, including learning difficulties and cerebral palsy. Congenital disorders of the peripheral visual system (CDPVS) can be further subdivided into two groups. The first group is referred to as ‘potentially complicated’ CDPVS, which involves children in whom the peripheral eye disorder is a part of diagnosed paediatric disorder, including underlying damage to the central nervous system. Examples of ‘potentially complicated’ CDPVS are cataracts in Down Syndrome and retinal dystrophy in peroxysomal disorders (i.e., a group of congenital diseases characterized by the absence of normal peroxisomes in the cells of the body, such as Joubert Syndrome). The second group is referred to as ‘potentially uncomplicated’ CDPVS and involves children in whom there is no known involvement of the central nervous system in the visual disorder diagnosis. In the ‘potentially complicated’ CDPVS the incidence of additional disabilities is higher than in the ‘potentially uncomplicated’ where only 17% of global learning difficulties has been reported (Sonksen & Dale, 2002), which is relatively low for the general VI population. Because of lower expected confounding variables of learning difficulties and motor impairments, the ‘potentially uncomplicated’ CDPVS group is a target population that is particularly useful for
psychological investigations of vision-loss. Example diagnoses falling under the ‘potentially uncomplicated’ CDPVS classification are: Glaucoma, Mycopthalmia, Aniridia, Coloboma, Persistent hyperplastic primary vitreous, Familial exudative, vitreoretinopathy (Norrie’s Syndrome), Cataracts, Leber’s Congenital Amaurosis, Cone Dystrophy, Optic Nerve Aplasia and Optic Nerve Hypoplasia.

Neurological abnormalities

**Imaging**

Vision is a powerful sensory modality which integrates and co-ordinates the information provided by other senses, allowing the features of the external world to be consolidated as a unified experience (Rock, 1985). Congenital blindness has offered philosophers and scientists alike the opportunity to speculate on how humans respond without vision. The interactive nature of development suggests that it is simply not that easy to ask how someone with sight differs from someone who became blind either at birth, in early, or later childhood. Nevertheless we can ask: how do areas of the brain that we know are normally associated with vision adapt if vision itself is lost? Recent research based on technological advances in neuroimagery have opened the way to a new era of understanding and now, through lesion, structural and functional imaging, we find that neural pathways are surprisingly ‘plastic’ in response to being deprived of sight. Neural circuitry is highly adaptable and if experience changes, for example, with vision-loss then the brain responds to other experiences from other sensory channels. For those congenitally deprived of visual input there is evidence of adaptive compensatory cortical reorganization. Functional magnetic resonance imaging (fMRI) and electro-
encephalography (EEG) techniques have shown that, for those with early blindness, cross-modal sensory re-organization occurs such that tactual sensory input and also tactual imagery activate cortical areas traditionally associated with visual processing. For example occipital cerebral blood flow and metabolism in both primary and secondary visual cortices during a rest period and during auditory and tactile discrimination tasks is increased in participants with congenital blindness when compared to sighted controls (Sadato, Pascual-Leone, Grafman, et al. 1996). Additionally, tactile imagery tasks (containing both sensory and cognitive components) recruit both visual cortex and parietal association cortex (Uhl, Kretschmer, Lindinger, et al. 1994).

Furthermore, studies, using a variety of auditory tasks, have reported a higher activation level in the occipital brain areas of people who are blind (e.g. Röder, Rösler & Neville, 2000). The research also shows that in some auditory tasks people who are blind outperform those who are sighted. For example they have better auditory localization (attending to sounds in peripheral auditory space) and auditory discrimination (detecting a rare target tone among frequent standard tones) abilities than sighted controls.

The research in connection with Braille reading has a long history and the results have not always been consistent. Braille reading by individuals with visual impairment activates the inferior parietal lobule, superior occipital gyri, primary visual cortex, fusiform gyri, ventral premotor area, superior parietal lobule, cerebellum and primary sensorimotor area bilaterally, as well as the right dorsal premotor cortex, right middle occipital gyrus and right prefrontal area. During tactile but non-Braille discrimination
tasks, in such participants, the ventral occipital regions, including the primary visual
cortex and fusiform gyri bilaterally were activated while the secondary somatosensory
area was deactivated. The reverse pattern was found in sighted subjects where the
secondary somatosensory area was activated while the ventral occipital regions were
suppressed. Thus, tactile processing pathways usually linked in the secondary
somatosensory area appear to be rerouted in blind subjects to the ventral occipital cortical
regions originally reserved for visual shape discrimination (Sadato, Pascual-Leone,
Grafman et al. 1996). The suggestion has been made that the task difficulty in Braille, as
well as its learning component, may explain its differential effects found in comparison
with simple tactual discrimination which shows reduced activation in the primary cortex.

In terms of age of blindness, there has been some uncertainty as to whether there is a
critical period of brain development after which neuro-plasticity is markedly constrained.
Some evidence seems to show that restriction of developmental progress in the early
years (up to the age of 16) limits the extent of possible changes later on, but this evidence
is largely dependent on studies of Braille readers and does not entirely accord with more
recent findings. Although recently some interesting evidence related to spatial
localization has thrown up a striking effect, related to touch, where early sight exerts a
lifelong influence on external and anatomical reference systems (see the section on
Spatial representation). There are other studies however, looking at different patterns of
neural transmission, that have shown that altering signals to different visual pathways and
the visual cortex can effect ‘temporary changes’ even after just a short period of vision
loss. Only quite recently has it become clear that changes in brain structure ( e.g.
Noppeney et al, 2005) as well as brain function ( e.g. Merbet et al, 2005; Maculuso &
Driver, 2005; Roder et al, 1999, 2002; Sadato et al, 1996, 2002) are mediated by changes in vision much later in life than was at first considered possible and furthermore, that these can occur on a temporary as well as a permanent basis.

With respect to blindness then compensatory recruitment of neural circuitry occurs and affects the way spared sensory modalities are processed and integrated. Progressive recruitment of parietal and then occipital cortex for auditory attention for example provides evidence for cross-modal sensory reorganization in the blind, although there have been some recent suggestions that the activated areas in the occipital cortex are functioning in a ‘task specific’ rather than a ‘sense specific’ way (Roder, Teder-Salejarvi, and Steer et al, 1999; Sadato et al, 2002). Thus spatial processing or a supramodal function may be involved in the regular functioning of the traditional ‘visual cortex’ (Macaluso, Frith & Driver, 2002). Furthermore, the changes that are seen to occur do not only happen as a result of disease or accident but though the use of investigative studies using transcranial magnetic stimulation (TMS). This is a noninvasive method by which neuroscientists can alter brain activity, producing ‘temporary’ lesions and in this case, mimic vision-loss. This methodology will herald a new phase in our understanding of the effects of blindness.

Neuropsychological profile:

Cognitive profile

Intelligence
Kolk (1977) reviewing many studies of intelligence concluded that ‘in general, average IQ scores do not differ significantly’ for blind children as compared to sighted children. Gerhradt (1982) reported that in terms of early categorical classification of form and function, as would be expected in free play, visually impaired infants of 14, 16 and 18 months followed the expected developmental path as would be predicted for sighted infants. Amongst the early studies though Tillman (1967) and Zweibelson and Barg (1967) refer to a more concrete concept bias at the cost of an understanding of abstract terms in the early childhood of blind children.

There have however been some suggestions that differential visual impairment diagnoses are linked with specific cognitive strengths and weaknesses. While no definitive evidence has been published in this respect, an ongoing investigation suggests that the exception may exist in a superior intelligence exhibited by those with a diagnosis of Retinoblastoma (Tobin, personal communication forthcoming).

**Language and verbal cognition**

Language has generally been seen as playing a powerful role in the development of children born with severely impaired vision (Landau & Gleitman, 1985; Warren & Hatton, 2003). Pérez- Pereira (1994) and colleagues have maintained over the years that language provides a privileged tool for children with VI, who rely on it and benefit from it to a greater extent than children who are sighted.
Verbal reasoning and intelligence helps children to develop strategies to cope with the loss of a sensory channel. So linguistic competence is an important factor not only in terms of knowledge acquisition where it clearly plays an important role but also that it helps mediate social outcomes in children with severely impaired vision. For children who are visually impaired language-based measures are commonly used to assess their general intellectual level, making it difficult to isolate the contribution of language irrespective of a child’s general cognitive ability. With regards to the “regular language” skills of children who are visually impaired from birth, research generally shows that these are developed with relative ease. A number of studies have demonstrated some specific delays and irregularities in early vocabulary acquisition and production, syntactic knowledge and acquisition of semantic concepts in children with VI (Andersen, Dunlea, & Kekelis, 1984; Dunlea, 1989), but generally speaking the development and use of “regular language” is largely in line with that of sighted children (e.g. Landau & Gleitman, 1985). An interesting example concerns the use of color terms. Studies with school-aged children have found that blind children do understand that vision endows color information and that this information is associated with objects and scenes. They have learnt then that bananas are generally ‘yellow’ and that the sky is ‘blue’ and show the same expectations or predictions of the use of such color terms in verbal prose along with understanding the subtleties which the color terms are associated.

While “regular language” skills such as articulation of speech, use of grammar, vocabulary level and conceptual understanding of the vocabulary in question, may enable a person to converse fluently they are not sufficient for achieving a successful socio-
communicative interaction with another person. For this, one must also master pragmatic language skills, i.e. use language appropriately in a given context. Vision is implicated in language development in general, as visually-driven joint attention experiences in early childhood are seen as providing a framework within which language learning occurs (Tomasello & Farrar, 1986). For this reason visual input may be of particular importance in the development of pragmatic language skills which are a cardinal feature of social communication.

The picture is somewhat unclear regarding language use for social and pragmatic purpose in children with VI. Research studies looking primarily at preschool children with VI, have raised concerns that children with congenital VI tend to use stereotyped language, show impoverished use of gestures for communicative purposes and use questions, sometimes inappropriately and to a greater extent than typically developing sighted children. It has been suggested that pragmatic language of children with VI has features that are similar to those of children with pragmatic language impairment (PLI) (Mills, 1993). Although it has been argued that such features of pragmatic language use of children with VI may have an important function in promoting their cognition and social interaction by providing an adaptive strategy to gather information, analyze speech, reduce memory load and avoid isolation. We ourselves have found that there are some irregularities in language presentation of a group of 15 children that we studied with congenital VI age range 6-12 yrs (Tadic, Pring & Dale, 2008a). Our findings were based on a structured language assessment (The Clinical Evaluation of Language Fundamentals – 3 : CELF- 3; Semel, Wiig, & Secord, 2000). We also used parental ratings of language
and communicative behaviors using the Children’s Communication Checklist (Bishop, 2003). The checklist targets both structural and pragmatic language behaviors observable in an everyday context, but also social interaction skills evident from everyday language use. The children in this study were matched with typically developing children with the same age, gender and verbal IQ scores. The findings suggested that there is a discrepancy in presentation of language ability in children with VI; that is, average to good and potentially superior regular structural language skills, but weaker use of language for conversational and social purpose. The pragmatic language difficulties in the VI group were observed in a substantial proportion of the children, these together with the checklist scores on social interaction and restricted and repetitive actions combined to suggest that many were of clinical concern and consistent with autistic spectrum disorder (discussed below).

**Memory**

Some early studies on memory performance found that children and adults who were blind did not forget their experiences in quite the same way as their sighted counterparts. They retained the details of the sensory or narrative experience. (e.g. Pring, 1995). By contrast the process of learning in sighted people reveal a tendency to forget the exact material, the learning episode itself, but instead remember the gist or the overall meaning of the material.

Several studies have reported significant advantages for short-term memory (STM) in children born with severely impaired vision, compared to sighted peers (e.g. Hull &
Mason, 1995; Smits & Mommers, 1976). Indeed advantages in STM have been noted in a wide variety of domains from pitch memory, sentence recall, auditory recall and memory for Braille and tactile illustrations (see Pring, 2008). Thus, the reliance and attention to auditory/verbal material and associations may be linked with maintaining information for longer in an ‘active store’, such as, for example, a phonological short term memory before dispensing with the information as it might be for sighted individuals. This may relate to the suggestion that individuals without sight have a higher incidence of absolute pitch ability than is normally found in the population.

**Spatial representation and concepts**

Vision is extremely helpful in understanding space, spatial relationships, internal spatial representations and spatial imagery - but studies have shown that it is generally not essential. For example while some children with VI opt to code their world sequentially, for example in terms of a 'route' (i.e. the path used for walking) others seem able to code for Euclidian space. Although not common, pointing is utilized by young blind children and when asked to point to an upstairs bedroom one child with congenital blindness could point appropriately to the room above and behind the child's location (Lewis et al, 2000a, in Lewis, 2002) while others were reported as more commonly pointing to the route they would *walk* to that room (Bigelow, 1996). The physical coding of movements in space; ie kinesthetic representation, seems to develop as efficiently in blind as in sighted children (Millar and Ittyerah, 1991).

The role of anatomically and externally anchored reference systems in blind and sighted people is currently being investigated by Roder, Focker, Hötting & Spence (2008). Spatial localization of tactile stimuli appears to be influenced by their ‘familiar’ location, with respect to the visual field, thus, when the hands are placed in an unfamiliar posture (crossed over the midline) mistakes are made in localising tactile stimuli presented to the hands (due to the mismatch between tactile and visual familiar co-location). Roder,
Rossler & Spence (2004) have uncovered evidence that early visual experience may play a key role in establishing this influence of the visual spatial frame of reference on tactile space. They found that whereas sighted subjects showed poorer accuracy at locating tactile stimuli with crossed hands, the congenitally blind subjects demonstrated no such impairment. Furthermore, the late blind subjects (one of whom had been blind for 40 years) showed a similar crossed-hands impairment to the sighted subjects, suggesting that early visual experience is necessary in the typical development of tactual space perception.

The imagery abilities of children with VI can be underrated. There is a developmental delay perhaps in being able to mentally rotate objects (Landau, 1991) but certainly by adulthood this ability can be achieved in an elaborate and creative way comparable to sighted controls (Eardley & Pring, 2007). Blind children can understand tactile drawings (Pring & Rusted, 1980) and draw with raised-line drawing materials. John Kennedy’s research has indicated the depth, sophistication and metaphorical artistry that can be found if adults and children with VI are given the tools to express themselves not only with sculpture but also in two-dimensional raised-line drawings (e.g. Kennedy, 2007).

Research has shown that in many situations involving mental spatial imagery such as when dealing with pictures or maps a featural analysis is emphasized at the expense of the overall global impression (e.g. Ungar et al, 1995, 1996). Raised outline maps were given to children to learn and the sighted children tended to use the spatial relationships between different landmarks and their relationship to the edges of the map to reproduce routes. Whilst the children who were blind did not perform as well, they had focused their attention on tracing the routes and naming the landmarks. The featural strategy is often less efficient, but research has shown that when required children and adults who are blind are able to use such methods. Indeed, recent research by Vecchi and colleagues
(eg. 2006) has shown that individuals with blindness can integrate very complex spatial mental images presented sequentially into a single integrated mental representation. This work too dovetails with the findings of Röder and colleagues mentioned above in relation to brain organisation changes in the visually impaired.

**Motor development**

Vision is implicated in balance, posture, gross and fine motor functions and although there is large individual variation significant motor delays have been reported. Hatton et al. (1997) looked at motor delays in 113 children aged between 12 and 73 months with a range of visual impairments but no additional disabilities. On the motor scale of the Battelle Developmental Inventory it was clear that the children with severe and profound visual impairment were very delayed in development, at 30 months of age their score was equivalent to 18 months, for example. However, sight made a significant impact since the children with some form perception, at the same chronological age scored at the level of 22 months and this trend continued with the availability of more sight.

Generally, the suggestion is that achievements that require self-initiated mobility are most significantly delayed such as elevating on arms in prone position, raising to a sitting position, pulling to a stand and walking alone. Vision seems to afford the impetus to cue a change in behavior and especially reaching out and grasping. Sound-initiated interest and the role of sound-making play objects in establishing the attention to and interest in objects to be grasped for the blind child are important. However research continues to find that there is some delay in both gross and fine motor development. In a study of 40 children with severe visual impairment, Levitzion-Korach et al. (2000) found that in all 10
aspects of motor development studied the children were slower than the sighted controls and the measurements predicted by the Bayley Developmental Scale (1993). For instance, they found that the children were delayed in standing alone with support (14.4 months compared to sighted children on average at 8.1 months). Not surprisingly climbing stairs with help (28.8 and 16.1 months) and standing on one foot (52.4 and 22.7 months) were amongst the greatest discrepancies reported.

Methodologically it is a challenge to gain insight into the meaning of motor-movements or the absence of movements in the young blind infant and child. For example, Lewis (2002) in her book on disability points out that the baby may turn her head, not to locate the sound, but in order to equalize the time at which the sound reaches both her ears. Another example would be the ‘freezing’ movement which is also a very common behavior in VI and initiated in response to a noise or some interesting stimulus, reflecting an increased attentional focus on sound-based information by a child with VI (even though there may be no head movement).

Stereotypies

One of the most noticeable behavioral abnormalities found among children with SVI and PVI is that of “stereotypies” – these are perseverative or ritualized movements or postures (Brambring & Tröster, 1992; Hobson, Lee, & Brown, 1999; Wills, 1968). According to parental reports, the most prevalent stereotypies are eye poking, body rocking, hand and finger movements and manipulation of objects. Such repetitive and stereotypic behaviors are also a striking feature of autism and in that context have been linked with compromised mental flexibility (e.g., Lopez, Lincoln, Ozonoff, & Lai, 2005). Amongst children with visual impairment raised levels of such repetitive
behaviors and stereotypic mannerisms in the school years correlate with certain attentional aspects of pre-school behavior such as the ability of the child to shift attention when so directed by an adult (Tadic, Pring & Dale, 2008b). They are to be distinguished from similar behaviors seen in mental retardation (Burack et al, 2000, p 265).

**Academic achievements**

Children who have VIIs often perform remarkably well in secondary schooling level after initial delays in primary school. One cause of such a delay is the difficulty inherent in mastering Braille. Reading Braille by touch with its 2X3 matrix of raised dots is hard because of the demands on tactual acuity. Each Braille cell (character in Braile) represents a letter and reading speed is increased by the ‘contraction; of key Braille words, for example, there are individual characters to represent the word ‘and’ and the group of letters ‘ing’. Braille teaching and perception is primarily ‘sound-based’ and beginning Braille readers often make 'mirror image' reversal errors and other similar confusions of global shape similar to those seen in developmental dyslexia. Children with VI and dyslexia have rarely been studied (though see Arter, 1998). In reading Braille the salient information is taken in while scanning the line (in contrast to vision and print), so it is important to watch the deployment of the hands and the precise timing of the fingers over small details as well as larger amounts of text as Susanna Millar has done (Millar, 1997). Children with VI can vary as to whether they use predominantly the left or right hand; they often use both hands together - the right hand first, followed along the Braille line by their left hand which has a place-marking and confirmatory role. In connection with lexicality, tactile letter identification and the verbally motivated role of reading as a rule of thumb it is probably correct to say that there is a right hand – left hemisphere advantage in adults (for example, Sadato et al, 1995), though, as Millar (ibid) points out,
it is foolhardy to attend to hand preferences in Braille reading when left hand advantage has been found for letter naming, where the spatial and pattern recognition aspects of the task are emphasized (Hermelin & O.Connor 1971; Rudel et al, 1977) and no hand advantage has been found for a variety of other reading related tasks (Millar, 1984).

Behavioral profile

Social emotional functioning

Social understanding
In recent years, there has been a particular emphasis on the deleterious effects of visual impairment on the social communication and social understanding of children with severe or profound visual impairments. The increasing prominence of this potential vulnerability is caused partially because it affects many other aspects of cognition and impact on behavior. Certainly a number of researchers and clinicians have noted striking behavioral resemblances between children with congenital VI and children with autism spectrum disorders (ASD) for whom the use of visually-based information has been called into question (see Pring, 2005 for a general overview). Some preschool children who are blind can display a range of ‘autistic-like’ clinical features, including poor sociability and communicative competence, repetitive and restricted patterns of play, unusual sensory preoccupations, unusual mannerisms, stereotypes of behavior patterning and echolalia.
The mechanisms underlying specific social difficulties and the autistic-like presentation shown by some children with VI, as well as the mechanism by which many children with VI are able to overcome such developmental challenges, still remain poorly understood. Early social functioning and later social understanding in children with typical development and children with autistic spectrum disorder is outlined briefly below in order that the behavior of children with VI can be put into context.

Early social functioning in typical development

Infants benefit from varied and stimulating social lives from the earliest stages of their development. Early social experiences are dyadic in nature, with an infant taking part largely in face-to-face interactions only with one social partner at a time. Typically developing sighted infants demonstrate responsive conscious appreciation of the adult’s communicative intentions and signaling by engaging in mutual eye-gaze, vocalization and rhythmic turn-taking patterns of behaviors (e.g., such as in social games like ‘peek-a-boo’). From around six months of age the new patterns of communication emerge, as the child moves from the purely dyadic interactions with one social partner into the world of objects. The main characteristic of these novel experiences is the infant’s awareness that their experiences of objects, people and events can be shared with others.

The coordinated sharing of attention (known as joint attention) between the child, an adult and objects in space has been the subject of much research. Its behavioral manifestation encompasses a complex set of actions, such as eye-gaze directing and
following, point following, showing and pointing, the purpose of which is to negotiate and share the mutual focus of interest with a social partner. Research evidence (Tomasello & Farrar, 1986) suggests that such behaviors emerge typically between six and twelve months and consolidate by eighteen months of age. These shared experiences between infants and their caregivers are largely driven by visual modality, hence they are often referred to as ‘joint visual attention’. Later on these young children begin to show more complex aspects of social understanding. By between 18 and 24 months of age a child may engage in pretend play. Pretend play involves the child understanding that one object can stand for another, that pretend properties can be attributed to real objects and that pretend interaction can be carried out with a non-real object. Certain ways of thinking then, that follow on from joint attention and precede theory of mind, (discussed below) underlie the child’s ability to reason about hypothetical situations (e.g., pretending that a banana is a telephone).

As the child develops and has more varied experiences of the world and people, s/he develops a critical milestone of social understanding – Theory of Mind. ‘Theory of mind’, ‘mind reading’ and ‘understanding of others’ minds’ have been used synonymously in psychology to refer to the child’s ability to understand and attribute a range of mental states to self and others in order to explain and predict their actions and behaviors (Leslie, 1987). In other words, to make sense of the sophisticated social environment that surrounds them, children must be able to understand that other people have intentions, desires, thoughts, beliefs and feelings which are different from their own and that such states of mind will influence people to act and behave accordingly. Our
actions then can best be understood by a child if s/he can guess what is in our mind but can be baffling if s/he is unable to do this. Understanding that people’s actions can be caused by their intentions is typically acquired by the age of five. Between the ages six and eight the child’s awareness becomes more sophisticated not just in terms of appreciating that people have beliefs about the world (which may be different from the child’s own beliefs), but also a growing sensibility/realization that they have beliefs about the content of others’ minds (i.e., about others’ beliefs), and similarly, that these too may be different or false. Over the later school years more complex and sophisticated use of theory on mind abilities are developed including, for example, the use of irony (Happé, 1994).

Development in social understanding in VI

Research has shown that children with VI can develop free from any cognitive, social or behavioral difficulties, and where difficulties do exist, these may be overcome, being viewed simply in terms of a delay. Nevertheless, it has been reported that some children with blindness continue to experience problems, in particular in the areas of social interaction and communicative competence; emotional expressiveness and emotional recognition; symbolic and functional (i.e., pretend) play; behavioral mannerisms, rituals and stereotypies; repetitive and unusual patterns of language use (i.e., echolalia and pronoun reversal) and autistic-like developmental regression (Cass, Sonksen & McConachie, 1994).
In typically developing sighted children joint attention ability is believed to develop spontaneously, evolving out of a natural context of routine child-caregiver interactions; the caregiver’s sensitivity and responsiveness to the child are the key ingredients to the child’s developing interpersonal engagement. Vision is likely to facilitate the caregiver’s involvement, the manifestation of which is likely to be different for children whose attention cannot be directed through eye-contact and visual gestures. However, Preisler (1991) while watching very young children with VI, noted that the children’s interactions at first seemed to be developing well, but from around their first birthday they had a notable difficulty with establishing the ability to engage in joint attention. Although they could share themselves with their mother, aided by the mother’s affect attunement, the children were unable to co-ordinate their attention at the same time towards an object in the external world. The triangulation then between the two actors and the object was not apparent. Interestingly, Preisler also noted that the infants with VI were attentive to the sounds in the environment and reacted to those sounds by establishing frozen bodily and facial postures. However, while these subtle signs, in addition to distinctive body pointing towards the sound, may provide the means of ‘attention directing’ from the visually impaired child’s perspective, such behaviors may be too subtle and ambiguous for the parents to interpret or notice.

Rogers and Puchalski (1984) commented that where the child is visually impaired, both partners in the child-mother interaction are disadvantaged. While the child is deprived of visual information and the lack of effective communication by the mother, who cannot interpret the child’s signals, the mother is deprived of positive and responsive cues from
her child that would let her know that she is doing the right thing. The study by Rogers and Puchalski highlighted the poverty of responsive social exchanges and initiations in mother-child dyads in cases of children who are visually impaired in contrast to the interactions of sighted children and their mothers. Presumably, this ‘vicious circle’ of impoverished parent-child responsiveness is likely to be both a cause and a result of impoverished joint attention capacity in children with VI.

However, in a study of two infants with congenital profound VI, Urwin (1978) showed that the nature of caregiver-child responsiveness is largely adaptive; once the mother has discovered particular cues that elicit the response of their child with VI, they were able to use these cues repeatedly: “[They] used phased touching routines to alert the babies’ attention; they would trace their fingers around the babies’ mouths, blow on their faces, and encourage them to explore their own body parts. [They] would mock-imitate the babies’ fusses, coughs, splutters and sneezes to ‘dramatize’ the babies’ actions” (Urwin, 1978, p. 88). However, despite the effective socio-interactive routines that facilitated the dyadic relationships between the children with VI and their mothers, both infants studied by Urwin showed difficulties and delays in their triadic interactions that require children to incorporate objects into their interactions with adults and establish reversible exchanges of actions on objects. Neither child exhibited spontaneous ‘showing’ behaviors to initiate joint interaction with the mother; if any reverse actions of ‘giving and taking’ emerged, they were largely a result of specific training provided by the mother.
It must not be forgotten that the effects of vision are extremely powerful and as Bigelow (2003) argues, some behaviors will serve a different function in children with VI compared to sighted children. This serves to exemplify the challenges of research in this field. Children with severely impaired or absent functional vision depend developmentally on tactile information and memory, as well as auditory input such as sound changes, air currents, echolocation (Millar, 1988) and verbal guidance by others. Such experiences must at least to an extent allow them to learn to co-ordinate the spatial placement of objects and establish a shared focus on such objects with others. However, despite the evidence of some joint attentional engagement in children with VI, it generally appears that the nature of such engagement is qualitatively different from what is known about joint attention capacity of sighted children, and this is particularly evident at the level of joint attention.

In terms of play in young children with VI there have been mixed reports. Fraiberg (1971) was perhaps the first one of many who mention the lack of “pretend” or symbolic play as opposed to functional play among blind children. Symbolic play involves the substitution of one object for another, for example when a cardboard box becomes a ‘car’ or a wooden spoon takes on the features of a ‘baby’. According to the results of a parental survey by Tröster and Brambring (1994) blind children and sighted children who engaged in ‘undifferentiated manipulation’ of objects were aged 16 and 8 months respectively, those relating to objects were 26 and 13 months respectively, those manipulating objects appropriately were 40 and 24 months respectively and those playing
symbolically were 55 and 35 months respectively. While Hughes et al. (1998) demonstrated in a study of young pre-schoolers that children with profound visual impairments (n=6) spent significant amounts of time in indiscriminate mouthing and manipulating of the toys (up to 75% of their time), while children with only severe sight loss (n=7) but some form vision far less so (44% of their time). It is clear from the above that some children who were blind demonstrated extremely delayed play behaviors though some contrasting results from Pérez-Pereira and Castro’s (1992) report that twin 3 year-old girls, one of whom was blind, frequently engaged in imaginative play, and Chen’s (1996) observation of symbolic play between blind children aged 20-30 months and their parents. Lewis et al (2000b) studied 18 children with VI and found some impairment in functional and symbolic play, but when they removed from the sample the 4 children who met the diagnostic criteria for autism then a different picture emerged, one where symbolic play was at a comparable level to children with sight. Bishop, Hobson & Lee (2005) also removed children who met a diagnosis of autism from their study of play in a group of congenitally blind children. They found that while some, socially able children were able to use symbolic play and were very similar to sighted children, the less socially able group showed significant poverty of symbolic play when compared to a matched mental age and IQ sighted group. Children with VI are restricted by their vision-loss and are likely to be vulnerable to developmental delay as a result but we have still a long way to go to see why some children with the same degree of blindness seem to be influenced by, what we could call, protective factors.

Emotion
Children who have profound visual impairments do have an understanding of cause-effect relationships that evoke basic emotions (i.e., happiness, sadness, fear and anger). More specifically, they are as able as sighted children to identify such emotions as they occur typically in specific situations, from their own perspective (e.g., *How do you feel when you receive a new gift?*) (Roch-Levecq, 2006) and from the perspective of others (e.g., *Susan is given a new bicycle for her birthday? What will Susan feel?*) (Dyck, Farrugia, Shochet, & Holmes-Brown, 2004). Moreover, Dyck et al. (2004) reported that, when asked explicitly to explain the meaning of emotions (e.g., *What does the word ‘angry’ mean?*), the semantic knowledge of children with VI even exceeded the knowledge of the sighted controls. However, both studies found that in the task which required children to represent mental states more implicitly the children with VI were not as proficient. Whilst being able to explain the meaning of basic emotions, the children with VI studied by Dyck et al. (2004) were less able than their sighted peers at recognizing vocal intonations specific to different categories of emotion. A similar difficulty among children with congenital VI with recognizing vocally expressed emotions has been reported by others; this was in comparison to recognizing environmental sounds in school-aged children (Minter, Hobson, & Pring, 1991).

Research suggest that the facial expressions of children and adults with VI are less florid than sighted counterparts. When asked to voluntary mimic emotional expressions on their faces Galati and colleagues (Galati, Miceli, & Sini, 2001) found that the same groups of muscles were activated to imply expression, but in a less marked way than sighted children. Their research showed that both spontaneous and voluntary expressions were
more ambiguous to sighted observers, probably because they lack experienced-based support and feedback. Gallese (2003) has suggested that perceived actions in others are internally simulated or replayed automatically via motor, cognitive and emotional representations. A number of brain systems may be involved in such a process but a candidate neural network is the mirror neuron system. It is interesting to speculate on the importance of the mirror neuron system and internal simulations in development and indeed to consider what the impact of an impairment to such a system might mean, for example in autism (Oberman & Ramachandran, 2007), or indeed, in the case of blindness.

Autism and Autism spectrum disorder

Theoretical and empirical advances in the understanding of the development of social milestones such as joint attention and theory of mind in the past twenty years have gained additional momentum largely as a consequence of the clinical emergence of the concept of autism spectrum disorders (ASD). Autism is a pervasive neurodevelopmental disorder and although biologically based, with a clear genetic component, the disorder is defined and diagnosed on the basis of a triad of behavioral difficulties, namely in social interaction, verbal and non-verbal communication, and repetitive behaviors and restricted interests.

The deficits in theory of mind understanding in autism have been related to disruptions in joint attention in early childhood and the lack of behaviors such as gaze and point following, showing and pointing (Charman, 2003). Absence of these behaviors in children with autism forms one of the criteria for diagnosing the disorder (DSM-IV-TR,
2000). Descriptively, such children generally seem socially aloof and distant – they do not experience the social world, they lack the behaviors of eye gaze detection or point following, they do not watch faces to help them understand meaning and intention. Thus, in many ways they appear as if they are unable to see the social world that surrounds them and within which different mental states and feelings arise. They have difficulty in adopting pretend play on their own or with playmates. They exhibit then a constellation of behaviors (to a greater or lesser extent) which place them apart from typically developing children.

Theory of Mind

Hobson (e.g. 1993) has been the most influential in recognizing and trying to explain the importance of vision for early development of relationships and social understanding. His experimental studies and that of others have indicated the serious difficulties confronting children with VI in developing Theory of Mind understanding (e.g. McAlpine and Moore, 1995). Peterson, Peterson, & Webb (2000) for example assessed two groups of children with differing levels of VI and across differing ages (averaging six, eight and twelve years). The findings of the study showed that, while the majority of the six year olds failed all four false belief tasks, the false belief performance improved with age, although significant difficulties could be seen in some eight year olds and to a lesser extent the twelve year olds. Certainly, many children with congenital VI lag behind and then catch up with their sighted counterparts- some need to take as longs as 6 or 8 years, and a subset of children with VI have longer-term difficulties. Such findings were most recently supported by Roch-Levecq (2006) who also demonstrated that primary school
aged children with congenital and profound vision loss who have normal intelligence have a significantly poorer false belief understanding than developmentally matched sighted controls.

While the majority of the studies on theory of mind in children with VI assessed the early belief understanding, Pring, Dewart and Brockbank (1998) used the Strange Stories paradigm (designed by Happé, 1994) to assess their more advanced theory of mind understanding. The task consisted of presenting children with a number of stories about everyday situations where the story protagonists say things that they do not literally mean (i.e., tapping advanced mental state elements, such as sarcasm, misunderstanding, persuasion, pretence and deceit). Pring et al. found that the children with congenital VI were poorer than age-matched sighted controls in predicting whether the protagonist’s statements were true and giving contextually-appropriate mental state justifications for these statements. This suggested that the previously observed socio-cognitive difficulties, based on the children’s false belief performance, persist into later childhood in children with congenital VI (i.e., age 9-12). The authors also reported a significant relationship between the children’s general intellectual levels and the frequency of their appropriate mental state justifications, suggesting that children with VI who are intellectually more able may also be more able to compensate for difficulties in social cognition than children with lower intellectual levels.

The issue of the link between autism and blindness is a thorny one. However, there is increasing evidence that congenitally blind children are ‘at risk’ of presenting with autism.
or an autistic-like condition (ALC). It may be more parsimonious to refer to the condition as ALC because as yet we cannot say whether such children show the neurotypical profile consistent with autism. While co-morbidity is likely to be as prevalent as it is in the sighted population it is likely that vision-loss itself interferes very significantly with those aspects of development that are impaired in autism. Thus we see the triad of impairments in varying degrees in children with congenital profound or severe blindness. There is no doubt that IQ may be a protective factor, along with a pre-disposition to show strengths in social ability – the research work of Hobson and his group working within a strong theoretical framework are likely to continue to make a significant contribution to out understanding (e.g. Bishop, Hobson & Lee, 2005). At the same time the studies that provide overviews of groups of children with VI, broken down by diagnosis, severity of vision-loss as well as behavioral measures, also helps to elucidate the nature of the relationship (e.g. Mukaddes et al, 2007).

In terms of diagnosis it is useful to know that the autistic-like clinical features in children with congenital VI were initially observed in small groups of children with specific diagnoses such as congenital Rubella, Leber’s Amaurosis and retinopathy of prematurity. However, the prevalence found across different etiologies implies that such psychopathology in children with congenital VI is not confined to any specific ophthalmologic disease. Instead it is the severity of VI and brain damage, with its associated intellectual impairment that are seen as the most important mediating factors along with the recent report implicating cerebral palsy (see Mukaddes et a, 2007).
Summary

There are multifactorial reasons for children without sight to present with similarities and differential aspects of cognition and behavior when compared to the typical child. At one end of the continuum we can see significant advances in brain research demanding new ways of thinking about neural plasticity and brain functioning and at the other end we see how children’s social interaction is modulated by their experiences. There are some areas of research that are not mentioned here because they lead to many imponderable questions such as the impact of diagnosis on the infant –caregiver dyad. Additionally, there has been a conscious effort to focus primarily on developmental issues rather than sum up the all the literature concerned with the loss of vision itself. Methodological considerations are also critically important but exacting standards are hard to maintain in an area of research with such a rare population. The amount of sight is hard to assess and can change in the samples of populations that are often reported, and other factors and problems linked to the site of brain impairment has been discussed above, yet these are all important considerations. Finally, behaviors exhibited by children who are VI can be hard to understand for sighted parents and clinicians alike. The visual channel is so important in integrating the senses – those with sight accept it unthinkingly. So that some behaviors may be hard to understand on the basis of current knowledge, for example why finger movements and manipulation might be delayed in blind babies, whose fists are often balled in the early months. Other behaviors, especially in relation to the development of social understanding, may be more successfully understood by learning from the findings with typically developing children. Although language and other non-
visual channels of information can compensate for much, it seems that sight-based knowledge, founded on the massively varied array of visual stimuli (including people) in the natural and man-made world, has a surprisingly critical influence on growing up. The degree of blindness suffered by an infant has a significant impact on their behaviors, and yet there are also reports to the contrary, of behavior comparable to sighted counterparts. No doubt this is where the interactive nature of development, and the multiple factors that mediate changes, have an effect. The vulnerabilities have been outlined in this chapter but there are precious few reports of the protective factors which lead to the most positive outcomes – it is to this that intervention and research studies need to turn their attention.

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