Mental health in children with severe hearing impairment

Nicoletta Gentili & Andrew Holwell

SUMMARY
Deafness does not in itself cause emotional/behavioural or cognitive problems or psychiatric disorders. However, children with hearing impairment are at greater risk of developing emotional/behavioural problems and neurodevelopmental disorders. The incidence of both seems to be higher in deaf children from hearing families. Most prelingual deafness is caused by recessive genes; hence, most deaf individuals come from hearing families, the majority of whom do not use sign language. Numerous studies, in both hearing and deaf populations, show how the lack of access to language has an impact on the emotional development of children. This article focuses on the mechanisms by which early language deprivation mediates emotional/behavioural difficulties and consequent emotional dysregulation, and may produce behaviours and symptoms that can be misdiagnosed as neurodevelopmental disorders in deaf children and adolescents (from infancy to 18 years of age).

DECLARATION OF INTEREST
None.

Hearing impairment from birth or early infancy may have lifelong consequences for an individual’s social adjustment and mental well-being in terms of communication, education, identity and employment (Hauser 2006; Woll 2008). A number of comorbid conditions that often accompany this sensory impairment might contribute to the development of mental disorders encountered in deaf children (Roberts 1999). Specific environmental factors, including familial, social, cultural and educational influences, can be both the direct and the indirect basis for the development of emotional/behavioural problems and psychiatric disorders through a diversion of the child’s developmental pathway. Lack of effective communication with carers, typically in families with hearing parents and deaf children, and consequent lack of or delayed language development is one of the most important environmental aetiological factors, albeit not the only one. Language deprivation has an impact on the quality of the shared emotional environment with the carer and potentially on the attachment relationship. It may also affect the individual’s cognitive ability and executive functions (Hauser 2006) and their ability to understand and share the emotional states of others and develop friendships (Dunn 2004). In this article, using the body of knowledge and research carried out on brain development, language and intersubjectivity, we focus on two aspects of mental health problems in individuals between infancy and 18 years of age with severe hearing impairment (whom we refer to here as deaf children, for conciseness): the intricate mechanisms that could be the foundation of the development of emotional/behavioural problems and how early language deprivation can cause symptoms that mimic attention-deficit hyperactivity disorder (ADHD) and autism-spectrum disorders.

Epidemiology of psychiatric disorders and communication
The total number of deaf children aged 0–15 years in the UK is around 20 000 (Woll 2008), and around 95% of them will have been born to hearing parents (Mitchell 2004). In sensorineural deafness (Box 1), at least 50% of cases have a genetic aetiology (Steel 1999); non-hereditary causes of sensorineural hearing loss include: congenital infections such as cytomegalovirus and rubella, perinatal anoxia and postnatal infections such as bacterial meningitis. Complications as a result of premature birth can also result in deafness (Wills 2005). These non-hereditary forms of deafness are often associated

<table>
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<th>BOX 1 The two main types of hearing loss</th>
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**Conductive**
- Involves the middle/outer ear
- Rarely causes severe to profound hearing loss
- The main cause is otitis media with effusion

**Sensorineural**
- Involves the inner ear, most frequently the cochlea, with damage to Corti’s organ or to the auditory nervous system
- Has a genetic cause in at least 50% of cases
- In most cases the gene is recessive
with other neurological sequelae that may affect behavioural, emotional and cognitive functioning (Hauser 2006).

Before the mid-1990s, the prevalence of psychiatric disorders in deaf children and adolescents was unclear. This was largely because the assessment instruments used had not been validated in the deaf population and the children themselves were rarely interviewed (Hindley 1997). A pilot study of new psychiatric screening questionnaires designed for this age group suggested that about 50% would have a mental health problem at some time before adulthood (Hindley 1994). Many subsequent studies (e.g. Hindley 1997; Van Eldik 2004) show a higher incidence and prevalence of mental health problems in deaf children compared with their hearing counterparts. Specifically, emotional and behavioural problems seem to be twice as common among deaf children (Vostanis 1997). Neurodevelopmental disorders appear to have a different prevalence: the incidence of ADHD in children with inherited deafness is the same as in their hearing counterparts but higher than in children with non-inherited deafness (Hindley 1998). This might be because medical conditions such as cytomegalovirus and rubella can cause both deafness and ADHD (Hindley 2005). Studies in the hearing population estimate the prevalence of autism-spectrum disorders to be between 58 and 67 per 10 000 (Tidmarsh 2003).

An association between a number of causes of deafness (such as CHARGE syndrome, rubella, cytomegalovirus and autism) has been observed (Jure 1991). Studies have found that the incidence of deafness is increased among children with autism-spectrum disorders (Rosenhall 1999). Conversely, the albeit limited research indicates that the incidence of autism-spectrum disorders among deaf students is also significantly higher, with one study identifying an estimated incidence of 1 in 76 (Gallaudet Research Institute 2007) and another of 4% (Jure 1991).

Deaf children are at greater risk of being physically and sexually abused than the hearing population (Sullivan 1991) and this in itself could trigger the development of emotional/behavioural problems or other psychiatric disorders (MacMillan 2001). Studies involving hearing children have shown how the emotional experience of physical abuse has an impact on their attentional abilities (Pollak 2003).

It has been estimated that 90% of deaf children who experience mental health problems come from hearing families (Moores 1987). Often these families have difficulties accepting deafness and in turn learning to use an adequate level of communication with their child (du Feu 2003).

It should be noted that when many of these studies were carried out, the average age at which deafness was diagnosed among children in the UK was around 20 months, and families were therefore unaware of their child’s needs for a crucial period of his or her development. Consequently, the incidence of early language deprivation was potentially higher. Medical advances and improvements in technology implemented in the mid-2000s have reduced the average age of diagnosis in the UK to under 3 months (Woll 2008).

The value of early intervention

A study in Finland found that the incidence of mental health problems among deaf children of parents who learnt to communicate adequately with them from infancy was much the same as among hearing children: 18.75 v. 15.8% (Sinkkonen 1994). Interestingly, all the hearing mothers of the children in this study and 94% of the hearing fathers had sign language skills and this was the preferred mode of communication in all the families sampled. This suggests that deaf children who grow up in psychologically healthy families in which they are not abused and with parents who have learnt to sign fluently will have the same incidence of mental health problems as their hearing peers. Other studies have shown that families who are helped to learn sign language can establish a more effective way of communicating with their children, thus improving their social and emotional development (Vaccari 1997).

There is some evidence that if early diagnosis is accompanied by intervention and support before the baby reaches 6 months of age, parental adjustment is better and the child’s language development is improved (Yoshinaga-Itano 2000).

These findings suggest that deafness is not in itself the direct cause of developmental, emotional/behavioural and cognitive problems or psychiatric disorders and that it is extremely important for parents to learn how to communicate effectively with their deaf child from a very early age.

Language and emotion

As discussed above, it is clear that deaf children from hearing families where there is not an adequate level of communication have a higher incidence of mental health problems than their hearing counterparts. This often seems to be related to a lack of language development caused by either a delayed diagnosis of deafness and/or families failing or being unable to learn how to communicate effectively with their deaf child. This language deficit will have an impact on the essential development, understanding and regulation of
emotions. This developmental process is generally thought to occur through language acquisition, the close link between language and executive functions, and the intersubjective relationship between the parents and the child, within which attachment and the pattern of emotions develop (Box 2).

Language in humans is uniquely complex. All species are able to communicate, but humans have the ability to discuss objects even in their absence, even when they are temporally distant. Human communication implies time and space. It is through the use of language that we can pass our experience on to others, enabling the process of learning to be more efficient and less dangerous. Language also enables the identification and labelling of emotions, which allows individuals to develop the ability to decode their own emotional states and achieve better emotional regulation. Language enables the disambiguation of unclear situations and allows a greater abstraction of thinking through the articulation of past experience and future goals. Studies on the interaction between language and emotions highlight how the labelling of emotions increases activity in the inferior frontal gyrus and decreases the amygdala response (Box 3) (Lieberman 2007). Research illustrates that children from families in which feelings are discussed with them are especially successful in performing on tests of social understanding (Meins 1999). In essence, language is the fundamental instrument of emotional regulation, as well as having a critical impact on executive functions, cognition and the ability to understand the emotional states and viewpoints of others.

An average child has the potential to learn 10000–15000 words between 18 months and 5 five years of age. This rapid learning is the result of an in-built tendency to communicate and imitate (Tomasello 2000). These imitative abilities have been demonstrated by a large body of research and are supported by the discovery of the ‘mirror neuron system’ (Box 4) (Rizzolatti 1996). By the time children utter their first lexicogrammatical speech (words used within grammatical parameters) they have begun to master grammar, to use words to refer to and mean things, and have learnt how to realise their intentions communicatively (Bruner 1983).

Vygotsky (1978), Bruner (1983) and Tomasello (2008) highlight the importance of a shared human interest in the process of learning, over and above our natural tendency to learn. Bruner (1983) calls this social support to the process of acquiring language the ‘language acquisition support system’

**Box 2 Intersubjectivity**

- Intersubjectivity has been described as the sharing of subjective states by two or more individuals (Scheff 2006).
- The experience of one’s subjectivity must contain the anticipation of the other: if I observe myself while I am doing something, I am anticipating what I would see in others if they did it or what others would see while observing me (Merleau-Ponty 1945).
- Between 2 and 12 months of age, babies become capable of grasping other people’s intentions (Tomasello 1999). This occurs through shared attention, the ability to follow the other’s gaze, pointing and imitative learning.
- By 4 or 5 years of age, children are able to understand others as ‘mental agents’ with different beliefs from themselves.
- The affective coordination between the child’s gestures and facial expressions and those of the carer is a form of primary intersubjectivity (Trevarthen 1979). This stays with the individual for life and supports actions that require explanation and prediction of others’ mental states.
- When children start linking actions to pragmatic contexts they are accessing secondary intersubjectivity (Trevarthen 2001).
- The abilities linked to both primary and secondary intersubjectivity are embodied, being sensorimotor, emotion-led, perceptive abilities. Both allow us to comprehend others in practical situations within the context of everyday life.

**Box 3 Affect labelling and the amygdala response**

- The amygdala is the key player in an extensive neural network involved in the processing of fear and other basic emotions. It appears to play an important role in five major areas (LeDoux 2005):
  - implicit emotional learning and memory
  - emotional modulation of memory
  - emotional influences on attention and perception
  - emotion and social behaviour
  - emotion inhibition and regulation.
- A relationship between the prefrontal cortex and amygdala has been identified in fear extinction: neural activity in the ventromedial prefrontal cortex increases as extinction is learnt (Milad 2002; Rosenkranz 2003).
- Putting feelings into words may alleviate negative emotional response: studies on ‘affect labelling’ show that linguistic processing of the emotional aspect of an emotional image produces reduced amygdala activity compared with the perceptual processing of the emotional aspect of the same image (Lieberman 2005).
- These studies have also demonstrated greater activity in the right ventrolateral prefrontal cortex (RVLPC) during linguistic processing. This region is associated with the symbolic processing of emotional information (Nomura 2003). Owing to the inverse correlation of the activity of the RVLPC and the amygdala during affect labelling, it might be suggested that putting feelings into words may activate the RVLPC, which in turn reduces the amygdala response and, thus, the emotional distress.

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Old children improve their theory of mind through... (LASS). This theory is supported by a body of research showing how children's grammatical skills tend to be varied over the first 2.5 years of life and links this finding to environmental factors as well as innate ones. Some studies (Nelson 1977) have found that providing children with extra examples of complex syntactical constructions facilitates the acquisition of such constructions. Their mastery of complex construction depends on the extent to which these are used by both parents and teachers (Huttenlocher 2002). Thus, the learning of language relies not only on innateness but also on the environment, represented by families, teachers, media and other cultural inputs. Indeed, recent developments in neuroscience are highlighting how environment can have an effect on brain development itself.

The plastic brain

Plasticity is an intrinsic property of the brain, allowing the nervous system 'to escape the restriction of its own genome and thus adapt to environmental pressures, physiological changes and experiences' (Pascual-Leone 2005). Over the past few decades it has become clear that the anatomy of the brain is influenced by the physiology of the body, by emotions and cognitions (Damasio 1994) and also by environmental stimuli (Trevathan 2001). It is through this plasticity of the brain that we can see that executive functions are systematically linked to language acquisition. Research conducted on theory of mind has shed light on this important relationship (Hauser 2008). Lohman & Tomasello (2003) describe how 3-year-old children improve their theory of mind through the appropriate use of language skills. This supports a large study conducted by Slomkowski & Dunn, who observed that 40-month-old toddlers display differing abilities in understanding others' feelings and 'reading' others' minds based on their differing levels of verbal ability and articulateness (Slomkowski 1996).

Executive functions

The executive system of the brain is involved in a number of psychiatric disorders seen in children and young people, including intellectual disability (Siegel 1989), ADHD (Pratt 2000) and Tourette syndrome (Singer 1991). It has therefore become a focus for neuropsychologists and researchers in the field of child development. Executive functions are cognitive abilities that control and regulate other abilities and behaviours and they are needed for planning ahead, goal-directed behaviours, abstract thinking, inhibiting actions and forming new concepts. They are mostly localised in the frontal area of the brain. The dorsolateral prefrontal area has been specifically identified with executive functions and working memory (MacPherson 2002). Executive functions allow us to organise our behaviour temporally and to override immediate demands in favour of longer-term goals (Dawson 2004). This skill requires self-control and it develops as the brain matures. Studies illustrate how infants’ and children's cognitive achievements seem to depend on the development of the prefrontal cortex (Posner 1998). Hence, self-regulatory skills, for both attention and emotions, appear to begin in infancy (Haith 1988) and continue into late adolescence (Steinberg 2003). Self-regulation of attention develops particularly rapidly between 2 and 7 years of age (Rothbard 2003). The prefrontal cortex does not complete its development until early adulthood (Giedd 2004).

The human prefrontal cortex is the brain structure exposed the longest to the shaping influence of the environment and so the environment will have a major influence on an individual's developmental pathway.

In summary, poor executive functions lead to psychiatric problems and poor language acquisition has a negative impact on executive functions. There is therefore a potential link between the quality of language acquisition and some mental health difficulties in childhood.

Essential and intermediary functions of language

It is essential to differentiate between functions for which language development is crucial and those for which language has only a mediating effect (Box 5). Functions such as communication,
The role of language

Language is crucial in:
- communication
- meaning-making and the sharing of experience
- hypothesis articulation
- thought abstraction
- emotion labelling and identification

Language has only a mediating effect in:
- attachment
- emotional attunement with carers
- intersubjective relationships

Studies have shown a different pattern of interaction between deaf children and hearing mothers compared with hearing children and hearing mothers. Although deafness does not prevent the development of a healthy attachment between a hearing mother and a deaf child, these dyads tend to spend significantly less time interacting, the mother initiates interaction more frequently but the child tends to terminate such interaction sooner (Lederberg 1990). This may be because there is a lack of attunement. In clinical practice, mothers have often stated their preoccupation with their child’s sensory impairment, which could distract them from the child’s non-linguistic cues, causing the intersubjective rhythm to be lost. Hence, the interaction between a hearing mother and a deaf child is more susceptible to poorer emotional regulation, linked to the mother’s potential growing sense of incompetence at communication and inability to tune in to the child in a ‘deaf-aware’ manner.

Language and emotional regulation

The intricate way in which language development has an impact on brain development is now clearer. It affects both the brain and the developing child’s sense of self. Lack of language will reduce possible interactions with hearing carers. This will affect the child’s attachment style and the parent’s ability to ‘tune in’ to their child. Concurrently, lack of effective communication will prevent the developing deaf child from elaborating, through abstract thinking, events that occur in their life. This in turn will stop the child integrating the experience of those events into their developing sense of self. At the same time, the developing executive system will be affected: the child will progressively become more unable to control their actions, have great difficulty in learning to think abstractly, be unable to adjust to new situations and be less able to understand others’ points of view and mental states, or will tend to develop this ability much later than hearing peers. This will lead to a greater tendency to develop emotional and behavioural difficulties, for instance being more likely to ‘explode’ or show ADHD-like symptoms. Furthermore, these difficulties may not be well managed by the parents when they first occur, because of problems with communication and a tendency to make more allowance for the ‘disabled’ child.

Of course, the degree to which all of this occurs varies depending not only on the degree of interaction with the parents but also on life circumstances (e.g. abuse of the child or illness of another family member), parental education, the information received at the time of diagnosis and the support provided by local services.
Language and theory of mind

Autism-spectrum disorders are neurodevelopmental disorders characterised by repetitive behaviour, delays in language development and difficulties in social communication. Research indicates that genes are a significant causative factor in these disorders (Rutter 2000), but the picture is complex, with multiple candidate genes implicated (Risch 1999).

Difficulties of diagnosis

Deafness and autism-spectrum disorders both have a considerable impact on communication and, when concurrent, are liable to have a cumulative effect. The combination of their features may confuse the diagnostic picture, resulting in early misdiagnosis and inappropriate treatment, or diagnostic overshadowing and subsequent delays in recognition and intervention. Although deaf children with autism-spectrum disorders demonstrate the same symptomatology as similarly diagnosed hearing children (Roper 2003) deaf children have been found to be diagnosed on average 12 months later than their hearing peers (Mandel 2005). The lack of diagnostic tools for assessment of autism-spectrum disorders in deaf children has been posited as a contributory factor (Szymanski 2008).

Our clinical experience points to the potential for false-positive diagnosis by people unaccustomed to working with deaf children: in an environment with limited access to language, deaf children are liable to have impaired communication strategies and may show a strong preference for predictability, routine and inflexibility that has the appearance of stereotyped behaviour (which resolves when successful communication strategies are employed). Children in a mainstream setting who appear isolated from hearing peers and relate better to the adults around them providing communication support may present differently in the company of age-appropriate deaf peers.

Our clinical observations have indicated that the increasing use of cochlear implantation has resulted in a growing awareness of the higher incidence of specific language disorders in deaf children which, in combination with other factors, may present similarly to autism-spectrum disorders.

Theory of mind

Just as children with autism-spectrum disorders are significantly delayed in their development of theory of mind (Baron-Cohen 1985), so are deaf children of hearing parents (de Villiers 2000). This delay may further contribute to diagnostic uncertainty. It has been attributed to the children’s restricted exposure to and participation in talk about the mind – mental state discourse (Peterson 2000) – and is irrespective of communication modality (Peterson 2004). In contrast, the theory of mind performance of deaf children of deaf parents equates to that of their hearing peers (Schick 2007), supporting the view that the delay relates to deficits in communicative experience. Mothers’ mental state discourses have been related to their deaf children’s theory of mind skills and strongly correlated with the mothers’ greater proficiency at signing. The effect of this maternal discourse remained significant even when the deaf children started school (Moeller 2006). Consequently, poor theory of mind is not a reliable indicator of autism-spectrum disorders in deaf children.

Diagnostic markers

So what are the reliable diagnostic clues? Box 6 lists a number of ‘red flags’ (Szymanski 2008) and indicators (Edwards 2008) of autism-spectrum disorders in children who are deaf.

An interesting finding is that even where correct diagnoses of autism-spectrum disorders have been made, communication within the family unit may have an influence on the perception of the symptomatology and, consequently, the focus of therapy. This is evidenced by a small study which found that hearing parents of deaf children with autism-spectrum disorders reported difficulties in the areas of socialisation, communication and behaviour (e.g. repetitive or self-injurious behaviour), whereas deaf parents reported that socialisation problems were the most problematic factors (Szymanski 2008).

However, outcome research into effective interventions has been shown to have limitations...
(Diggle 2007) and the development of effective interventions for deaf children with autism-spectrum disorders is still work in progress.

Conclusions

Children with severe hearing impairment are at greater risk of developing psychiatric disorders and of poor psychosocial adjustment compared with their hearing peers. The reason for this is the barrier to efficient language acquisition. It is therefore essential to minimise the damage that lack of language or delayed language development can cause and this can be achieved through two strategies.

First, services must ensure early diagnosis of hearing impairment, followed by provision of the right information to parents on the use of language. This is the aim of the National Health Service’s newborn hearing screening programmes in the four countries of the UK. At a very specific level, for instance, it has been suggested that children with autism-spectrum disorders have auditory processing deficits and may struggle with conversion of auditory information to vocal utterances (Bonvillian 1981). However, the incorporation of facial expression and gesture into signing can help communication (Chamberlain 2000) and it is important for the hearing parent to be aware of this at diagnosis.

Facilitating better interaction between parents and their deaf child minimises the possibility that the child will develop an attachment disorder or emotional dysregulation. This is achieved through post-diagnosis counselling for parents and through helping parents to use communication strategies focused on the child’s specific needs. It is important to ensure that the child is fully integrated into a school that is aware of and respects the child’s communication needs.

Second, if necessary, families must have access to specialised child and adolescent mental health services, so that the right assessment and treatment is offered to children and young people with hearing impairment. Clinicians without experience of hearing-impaired children often misinterpret the ‘signs’ and wrongly diagnose and treat them. This failure has several causes: their inadequate skills in communicating with deaf children; their lack of knowledge of the complex factors implicated in the development of psychiatric problems in this group; and the paucity of tools validated for the use with deaf children and young people.

Symptoms seen in ADHD are common to a number of disorders (such as anxiety, oppositional defiant disorder, reactive attachment disorder, and communication/language disorders) and we have discussed how deaf children from hearing families are more vulnerable to these. Lack of language can also mimic some of the symptoms of an autism-spectrum disorder, such as lack of empathy, echolalia and isolation. Hence, assessing a deaf child with emotional/behavioural difficulties can be a complex task, as a painstaking unpicking of variables is needed to ensure a correct diagnosis and an effective treatment plan. This may be achieved by offering a careful, multidisciplinary assessment by skilled, specially trained professionals. Assessors should be supported by sign language interpreters with training and experience in child mental health and developmental psychology.

References


MCQ answers

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MCQs
Select the single best option for each question stem

1 What percentage of deaf children are born to deaf parents?
   a 90%
   b 20%
   c 5%
   d 95%
   e 45%.

2 What is the most significant contributory factor to the additional mental health needs of deaf children?
   a genetic factors
   b infective agents
   c prematurity
   d impaired communication
   e idiopathic conditions.

3 How does the incidence of mental health problems in deaf children compare to their hearing peers?
   a half
   b same incidence
   c double
   d 10% higher
   e ten times higher.

4 Which is a reliable indicator for making a diagnosis of autism-spectrum disorders in deaf children?
   a delay in acquisition of speech
   b delay in developing theory of mind
   c the child does not turn when called by name
   d inflexible patterns of behaviour
   e absence of joint attention skills.

5 Which of the functions below occur independently of language?
   a hypothesis articulation
   b emotional labelling
   c attachment
   d thought abstraction
   e meaning making.

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