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Children with congenital hearing loss – a vulnerable group

PERSPECTIVES

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Children with congenital or early-onset sensorineural hearing loss are at risk of additional neurodevelopmental disorders, including visual, cognitive and motor impairments. Systematic assessment and follow-up of children with diagnosed sensorineural hearing loss is important to detect such problems at an early age.

Illustration: Lisbeth Moen

Congenital or early-onset sensorineural hearing loss may be the result of genetic or environmental factors, or a combination of the two. A genetic cause is identified in up to 60 % of cases, whereof about 30 % are syndromic and 70 % non-syndromic, i.e. hearing loss is the only disability (1). About 40 % are caused by environmental factors such as asphyxia, hyperbilirubinaemia, prematurity, meningitis, ototoxic drugs and congenital infections (with cytomegalovirus infection the most common).

A large proportion of cases of sensorineural hearing loss due to congenital cytomegalovirus infection have late onset and are not detected during neonatal screening (2). All prenatal and neonatal infections in children (so-called TORCH infections: toxoplasmosis, rubella, cytomegalovirus infection, herpes simplex) may cause sensorineural hearing loss and brain pathology (3). The Norwegian national guidelines for the assessment and follow-up of hearing in young children (0–3 years) recommend testing for cytomegalovirus infection only when the cause of the hearing loss is unknown. Focus on cytomegalovirus infection means that testing for other agents is not always performed. This may lead to other causes of hearing loss being underdiagnosed.

The known environmental causes of sensorineural hearing loss may also cause neurodevelopmental impairments in children, and findings on brain MRI are often non-specific. The authors of a systematic review concluded that 31 % of children who experience a preterm or neonatal insult develop at least one long-term neurodevelopmental sequela (4). The most common sequelae are cognitive impairments, developmental delay or learning difficulties (59 %), cerebral palsy (21 %), hearing loss (20 %), visual impairment (18 %) and gross motor and coordination problems (8 %).

Undiagnosed coexisting impairments

Each year, the Norwegian National Unit for Hearing Impairment and Mental Health at Oslo University Hospital treats about 90 children and adolescents aged up to 18 years with sensorineural hearing loss and psychopathology.

To our knowledge, a number of the children referred have coexisting impairments that have not previously been diagnosed. These may include difficulties with motor coordination, impaired executive or social functioning, learning difficulties, binocular visual problems, and strabismus. The children's health problems have often been regarded as a consequence of their hearing loss and no attempts have been made to treat or accommodate them.

Brain MRI has been performed on a number of children with such impairments, showing periventricular leukomalacia (PVL), enlarged ventricles and white matter injury. Children with findings on brain MRI undergo examination by a paediatrician, either in connection with admission to a neonatal intensive care unit or as part of an assessment for cochlear implantation. However, when neuromotor development is normal, paediatric follow-up is often discontinued because the MRI findings are assumed to be non-specific and without predictive value.

Assessment of the aetiology of hearing loss in children referred to the Norwegian National Unit for Hearing Impairment and Mental Health varies from minimal, to extensive genetic and physical examinations, including brain MRI. The follow-up of conditions other than hearing loss is also variable, despite the high frequency of comorbid neurodevelopmental disorders.

Brain MRI and hearing loss

Few studies have examined the relationship between non-specific findings on brain MRI and the risk of long-term sequelae in children with sensorineural hearing loss. Chilosi et al. found that 48 out of 100 children with sensorineural hearing loss had neurodevelopmental disorders (5). Additionally, 19 children had language disorders, which were not classified as coexisting impairments in this particular study. The risk of coexisting impairments was twice as high when changes in the white matter were detected on MRI. It was also somewhat higher in the presence of migration disorders (5).

Independent of hearing status, several studies have examined the relationship between non-specific findings on brain MRI in children and the risk of long-term sequelae. The results are highly heterogeneous: 7.9–39.4 % (6–10). Children with known risk for brain injury, but without pathological findings on brain MRI, may also have neurodevelopmental impairments (11).

The divergent results may explain why pathological MRI findings alone do not result in children receiving further follow-up. However, methodological differences in image acquisition protocols, and differences in outcome parameters and duration of follow-up make it difficult to compare studies. Technological developments have led to

improvements in the diagnostic capabilities of brain MRI compared to 20–30 years ago. There is a need for studies with longer follow-up (to school age, possibly adolescence), and which examine balance, coordination, vision, cognition and psychopathology.

Dettmann et al. found that children with sensorineural hearing loss attributable to environmental factors have poorer receptive language standard scores after cochlear implant surgery than children with genetic non-syndromic hearing loss, as well as a higher degree of comorbidity (59 % vs. 8 %) (12).

There is international consensus that early diagnosis and follow-up of hearing loss in children is important for the children to achieve good language outcomes (13, 14). Other factors shown to have a positive impact on language acquisition include high maternal educational level and the absence of coexisting impairments in the child (14). However, many of the children have poor language acquisition of unknown aetiology. Examining brain MRI and aetiological factors might contribute to a better understanding.

Hearing loss and neurodevelopmental impairments

In all, 30–60 % of children with sensorineural hearing loss have coexisting visual impairments or cognitive, motor, emotional, behavioural or other difficulties (15–18). Visual impairments in particular are associated with hearing loss (40–60 %). These are often undiagnosed and may include correctable binocular visual problems, strabismus and refractive errors, or permanent retinopathy or optic nerve injury (16).

Few studies have examined potential associations between child development and causes of sensorineural hearing loss, but studies of individual environmental causal factors such as prematurity, asphyxia and congenital cytomegalovirus infection have found high levels of comorbid neurodevelopmental impairments, which may *also* have a progressive course. In our opinion, too little attention has been paid to this fact. Follow-up of preterm children shows that examination and assessment at two years of age has limited value for predicting cognitive and motor function at the age of five (19).

> ***«We believe the National guidelines should be changed so any coexisting impairments can be detected as early as possible»***

If cerebral palsy, retinopathy of prematurity and intellectual developmental delay are excluded, the child is often assumed to have no coexisting impairments, despite showing clumsiness, binocular visual problems, learning difficulties, etc. The child's difficulties are instead attributed to his or her hearing loss (diagnostic overshadowing). When the hearing loss is detected in infancy, it may involve a progressive course that may also apply to other functions. Extra vigilance is required to ensure that the development of any coexisting impairments is detected.

Deaf children of deaf parents are more likely to have non-syndromic genetic hearing loss. These children are thus less at risk of neurodevelopmental disorders than the children of hearing parents (20).

Clumsiness and psychopathology

Many children with sensorineural hearing loss have motor difficulties without having cerebral palsy (21). These are often not considered to be coexisting disorders, but are associated with difficulties in social relationships (21). Motor clumsiness, irrespective of hearing status, is often associated with psychopathology, including autism spectrum disorder and ADHD (22); anxiety disorders (23), as well as reduced quality of life (24). Early detection of such difficulties and targeted training can help improve the child's coping skills and quality of life.

Mental health

The prevalence of psychopathology in children with sensorineural hearing loss – 20–50 % – is higher than in children with normal hearing (25). Moreover, the prevalence is three times higher among children with sensorineural hearing loss and coexisting impairments than in hearing-impaired children without coexisting impairments (26). Children with hearing loss are referred to mental health services for treatment later than children with normal hearing (18). It is not the deafness per se that contributes to psychiatric problems. Low cognitive ability, poor language comprehension (in the child's preferred language, whether sign language or spoken language) and coexisting impairments are all associated with psychosocial problems (17, 18, 26).

Recommendations for follow-up

We believe that the Norwegian national guidelines for the assessment and follow-up of hearing in young children (0–3 years) should be changed so that any impairment in addition to the hearing loss can be detected as early as possible. A multidisciplinary approach should be provided, including contributions from paediatricians, otolaryngologists, radiologists, educational audiologists, ophthalmologists, orthoptists, physiotherapists, social workers, psychologists and child and adolescent psychiatrists, to ensure a comprehensive assessment and follow-up after diagnosis of sensorineural hearing loss. There should be a particular focus on vision, including binocular vision and strabismus, as well as on balance and coordination, cognition and mental health with systematic assessment and follow-up through to adolescence.

> ***«A thorough aetiological work-up, including assessment of cerebral MRI, can help reveal which children are at risk of comorbid impairments»***

A thorough aetiological work-up, including assessment of cerebral MRI, can help reveal which children are at risk of comorbid impairments. In cases of 'unknown' aetiology, but with pathology on cerebral MRI, a pre/perinatal cause of both the brain

injury and the hearing loss cannot be excluded. The child should then be followed up closely to ensure that any coexisting impairments are detected.

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