Medical Evaluation of children with permanent unilateral hearing loss.

Produced by the British Association of Audiovestibular Physicians and British Association of Paediatricians in Audiology

August 2009

Aim

The aim of these guidelines is to provide an evidence based approach to investigation of the cause of unilateral hearing loss in children.

This new guideline was developed by the working group at the request of the BAAP/BAPA federation.

Why investigate hearing loss

There are several reasons why it is important to establish the cause of the deafness:

1. To try and answer parents who ask "Why is my child deaf?"
2. To identify syndromes, gene mutations and where appropriate manage medical conditions e.g. Waardenburg syndrome, Goldenhar syndrome because of oculo-auriculo-vertebral anomalies, Branchio-oto-renal syndrome because of renal dysplasia, space occupying lesions, congenital infection, Neurofibromatosis type 2.
3. The results of investigations can assist the professionals in making decisions about the most appropriate management plan e.g. further investigations, intervention with amplification, frequency of follow ups and Educational placement e.g. if likely to be progressive and counselling the family appropriately.
4. To inform genetic counselling
5. The information from investigation of childhood deafness informs epidemiological research

The timing of investigations will depend on the family's readiness to proceed with tests and how well the child can cooperate with tests.

Search methodology : Medline, Embase, Cochrane
By NLH Specialist Library for ENT and Audiology , and hand search of journals.

Subjects

All children with unilateral

- Permanent sensorineural hearing loss of prelingual or late onset including fluctuating and progressive losses with two or more thresholds greater than 40dbHL.
- Permanent conductive hearing loss

Guidelines for Good Practice

The following investigations should be completed for every child.

1) <u>Paediatric history</u>: Strength of evidence - D

Detailed history of:

- onset of audiovestibular symptoms and progression of symptoms

History of exposure to risk factors e.g.

- noise
- ototoxic medications/ radiation
- head injury
- ear disease
- meningitis
- bacterial and viral illness
- immunisation status

Pregnancy, delivery and postnatal period

Developmental milestones including speech, language, motor milestones as well as social development

Family history of deafness or risk factors associated with hearing loss in first and second degree relatives.

History of consanguinity and ethnic origin

2) <u>Clinical Examination:</u> Strength of evidence - D

Should include

a) Measurement of height, weight and head circumference

b) Inspection of craniofacial region.

c) Examination of the ears, neck, skin and nails, limbs, chest, abdomen and gait.

3) <u>Family audiograms</u>: Strength of evidence - D

Parents and first degree relatives because many people can be unaware of a unilateral hearing loss.. (1,2,,3,4)

4) <u>Ophthalmological assessment</u>: Strength of evidence - D

Assessment of visual acuity and fundoscopy (5,6)

5) <u>Cytomegalovirus DNA Testing</u> - Strength of evidence – D

This is essential because unilateral hearing loss is well described as well as bilateral progressive hearing loss (7, 8,9).
< 1 yr : urine and/or saliva x 2
>1yr: if taking blood: IgG
if not taking blood: urine and/or saliva (if they are negative, test IgG)

If either are positive, request Dried Blood Spot for CMV testing:
Requires:

1. mother's address during first weeks of baby's life
2. signed parental consent
3. newborn screening laboratory address:

see http://www.newbornscreening.org/laboratories.asp
N.B. consider maternal IgG: if negative, exclude congenital CMV

6. <u>Other investigations</u>:

These will be indicated from history and clinical findings. The timing will depend on the family's readiness to proceed with tests, and how well the child can cooperate with tests.

<u>Imaging</u> (10,11,12,13). **Strength of evidence - D**

For children with severe to profound unilateral sensorineural deafness – MRI is the imaging of choice to ensure excellent visualisation of the vestibulocochlear nerve trunk, inner ear structures and posterior fossa. It is important also to be aware if the normally hearing ear is structurally normal.

Consider MRI in children with mild to moderate unilateral sensorineural hearing loss.

Consider CT scan of the Petrous temporal bones in children with a unilateral fixed conductive hearing loss.

Decisions about imaging should be based on the clinical findings.

<u>Gene mutation testing</u>: **Strength of evidence - D**

Consider mutation testing in cases where a syndrome is suspected e.g. Waardenburg, Branchio -oto- renal, hemifacial microsomia or facial weakness or where the result of the investigation will affect the management of the child or family. Blood tests for mutation analysis requires consent from parents, an explanation that DNA is stored afterwards in the lab, that genetic testing can take a considerable time, and for permission to share results with other family members and professionals, and specify which professionals should be copied into correspondence (see guidelines for consent for genetic testing (14)

<u>There is no convincing evidence that mutations in the Connexin 26 gene cause unilateral hearing loss.</u>

<u>Other blood investigations</u> - Strength of evidence - D

These will be indicated from history and clinical findings (15, 16)

<u>Screen for Autoimmune conditions:</u> where there is evidence of progression of hearing loss and or other systemic symptoms (15,17)

Serology for congenital infection: where there is a need to exclude congenital rubella, toxoplasmosis, syphilis. Investigation may involve testing maternal stored (booking) serum if available.

Biochemistry:

Where clinically indicated

Metabolic Screen and Chromosomal studies:

May be indicated if there is a history of developmental delay or Dysmorphic features

(Follow Local Child Development Team protocol)

Renal ultrasound:

- If child has preauricular pits or sinuses, deformity of ear, branchial cleft or cysts
- Mondini defect on imaging.
- permanent conductive or mixed hearing loss
- multiple congenital abnormalities (18)

Vestibular investigations.

Consider in cases where there are vestibular symptoms, motor milestone delay or where there is fluctuating and/or progressive deafness.

Consider referral to Clinical Geneticist especially

- if a syndrome is suspected,
- child has multiple system abnormalities
- parental request
- opinion required on interpretation of genetic mutation testing
- After completion of investigations if a genetic disorder is diagnosed.

In some cases following medical evaluation no cause for the deafness is found. These cases require periodic review for arranging further investigations as needed and to repeat some previously normal investigations.

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Appendix

Guidelines are ' systematically developed statements to assist decisions about appropriate care for specific clinical circumstances' based on systematic reviews of research literature (1, 2, 3).
Guidelines are not intended to restrict clinical freedom, but practitioners are expected to use the recommendations as a basis for their practice. Where possible recommendations are based on, and linked to the evidence that supports them. Areas lacking in evidence are highlighted and may form a basis for future research

Categories of evidence:

Ia Evidence from meta-analysis of randomised controlled trials
Ib Evidence from at least one randomised controlled trial
IIa Evidence from at least one controlled study without randomisation
IIb Evidence from at least one other type of quasi-experimental study
lII Evidence from non-experimental descriptive studies, such as comparative studies, correlation studies, and case-control studies
IV Evidence from expert committee reports, or opinions or clinical experience of respected authorities, or both

- strength of recommendations is expressed thus:
    - A directly based on category I evidence
    - B directly based on category II evidence or extrapolated recommendation from category I evidence
    - C directly based on category III evidence or extrapolated recommendation from category I or II evidence
    - D directly based on category IV evidence or extrapolated recommendation from category I, II, or III evidence

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Glossary

Permanent Conductive hearing loss
A type of hearing loss due to permanent defect in the structure of the outer and/or middle ear. The defect interferes with the transmission of sound to the inner ear resulting in a hearing loss which is usually of mild to moderate degree. It may co-exist with conductive hearing loss due to otitis media with effusion or with sensorineural hearing loss.

Sensorineural hearing loss
A type of hearing loss due to a defect or damage in the inner ear (cochlea), 8th cranial nerve , the auditory pathway or auditory cortex. The hearing loss is permanent and may be of variable degree - mild, moderate, severe, or profound. It may co-exist with otitis media with effusion.

August 2009 (Edited by Dr Tony Sirimanna, to be reviewed in August 2011)