British Association of Audiological Physicians (BAAP)
British Association of Community Doctors in Audiology (BACDA)

Guidelines for Good Practice

Investigation of new cases of severe and profound bilateral sensorineural hearing loss in children

These guidelines have been written by a working party mandated by the BAAP Evidence Based Clinical Practice and Audit Group. This paper presents evidence-based recommendations for the investigation of newly diagnosed children with bilateral severe and profound sensorineural deafness. The recommendations in this guideline are of relevance to all professionals and parents of deaf children.
The guidelines will be reviewed again in September 2002.

Aims

The aim of these guidelines to propose a rational approach to the investigation of the cause of deafness in children. Guidelines are ' systematically developed statements to assist decisions about appropriate care for specific clinical circumstances' based on systematic reviews of research literature.
Guidelines are not intended to restrict clinical freedom, but practitioners are expected to use the recommendations as a basis for their practice. Local resources and the circumstances and preferences of individual patients need to be taken into account. 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.

Grades of evidence

Grade A: Requires at least one randomised controlled trial as part of the body of overall good quality and consistency addressing the specific recommendation.

Grade B: Requires availability of well coordinated clinical trials but no randomised clinical trials on the topic of the recommendation.

Grade C : Requires evidence from expert committee reports or opinions and/or clinical experience of respected authorities. Indicates absence of directly applicable studies of good quality.

Why investigate hearing loss

There are several reasons why it is important to investigate deafness:

1. To try and answer parents who ask "why is my child deaf "?
2. To identify and treat medical conditions e.g. 8th nerve aplasia congenital infection, Jervell and Lange-Nielsen syndrome, Alport's syndrome, Neurofibromatosis type 2, Ushers Syndrome, and vestibular hypofunction.
3. The results of investigations can assist the family in making decisions about the most appropriate communication mode, educational placement and counselling on cochlear implantation e.g. in 8th nerve aplasia, Ushers syndrome.
4. To inform genetic counselling
5. The information from investigation of childhood deafness informs epidemiological research

The yield and cost benefits of tests are considered. It is important to use the least invasive, most cost effective and most readily available investigations. This has to be counterbalanced by the need/cost to identify rare conditions that can be life threatening and require prompt medical treatment.

Subjects

All children with bilateral sensorineural hearing loss and thresholds over 70 dB in the better ear averaged across 500, 1,000, 2,000 and 4000Hz.

Guidelines for Good Practice

Level 1 investigations for confirmed severe to profound bilateral sensorineural hearing loss

Investigation	Grade
Paediatric history: Detailed history of pregnancy delivery and postnatal period. Developmental milestones including Speech and Language and motor milestones, pre and post natal noise exposure, history of ototoxic medications, head injuries, ear disease, meningitis, viral illness and immunisation status	C
Family History of Deafness: or risk factors associated with hearing loss in first and second degree relatives	C
Clinical Examination: inspection and physical measurement of craniofacial region, assessment of the neck, skin and nails, limbs, chest and abdomen. Developmental examination.	C
Family Audiograms : On 1st degree relatives (1)	C
Electrocardiography (ECG) : An ECG should be performed to exclude prolongation of the QT interval (2)	C
Referral to Ophthalmologist : Every child should be referred for assessment of visual acuity and dilated fundoscopy if delayed motor milestones discuss with Ophthalmologist re: consideration for electro-retinography (3)(4)(5)(6)	
Urine for microscopic haematuria (labstix) (7)(8)(9)	
Blood for Connexin 26 mutation testing (10)(11)(12)(13)	
MRI of Internal Auditory Meati or CT Scan of Petrous Temporal Bone (14)(15)(16)	C

The above investigations should be performed on each child - the timing will depend on several factors, which may include when the family accept and agree that testing is important, the availability of local test facilities, how well the child can cooperate with the testing.

Level 2 investigations

Investigation	Grade
Serology : should be performed to exclude congenital infection , maternal stored serum should be tested	C
Haematology and Biochemistry : where clinically indicated	C
Thyroid Tests : where family history of thyroid disease or goitre present	C
Immunology Tests : where clinically indicated	C
Metabolic Screen Blood and Urine : where clinically indicated	C
Renal ultrasound - consider if child has preauricular pits or sinuses, branchial cleft or cysts, or Mondini cochlea on imaging *	C
Clinical photography	
Chromosomal studies : if history of developmental delay or dysmorphic features	C
Referral to Clinical Geneticist *	C
Vestibular investigations *	C

* consider in all cases

In Level 2 investigation it is recommended that vestibular assessment is performed on all children and that a referral to a Clinical Geneticist is considered especially if there is a family history of consanguinity. The remaining investigations in level 2 will be indicated from history and clinical findings.

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