

Competencies for investigating the cause of hearing impairment in babies identified through the newborn hearing screening programme

Why investigate the cause of hearing impairment?

The identification of significant hearing impairment in a newborn baby can be a traumatic experience for families. No matter how sensitively the news of hearing impairment is shared by professionals, the family will have questions, the first of which is often, 'Why did it happen?'

Medical investigations are also carried out to identify and manage other conditions which may coexist with or add to difficulties caused by hearing impairment. Sometimes parents may wish to discuss the likelihood of a further child being born with hearing impairment.

Paediatric audiology services across the UK are generally multidisciplinary and teams consisting of a variety of professionals deliver services. It is important to ensure that appropriately trained, competent doctors carry out this aspect of the service. (we need to add, once agreed, who signed up to this document, and have to make sure all those professional groups that may be involved agree to it)

What is competence?

Competence incorporates an integrated approach to the idea of professional judgement needed for intelligent performance in a specific situation. It allows the incorporation of ethics and values as elements in competent performance. Evidence based medical knowledge and/or skills need to mesh with ethical standards and the capacity to communicate with families of hearing impaired children.

General Competencies

Attitude/Family Friendly Service

- Provide a child orientated environment for the family
- Avoid test battery approach
- Work at parents' pace, frequently checking understanding
- Be sensitive to parental wishes
- Respect parental views
- Encourage parents' full involvement
- Select tests appropriate for level of hearing loss
- Discuss risks and benefits with parents
- Recognise unique needs of the family
- Be willing to listen to families
- Adopt a flexible approach
- Be aware of the role of extended family
- Practise deaf awareness
- Allow parents time to digest the information
- Be ready to involve others on the team
- Have undisrupted time for parents to discuss issues**

How is competence assessed?

The elements of competence may be assessed in different ways. Attitude may be assessed by feedback questionnaires from service users or colleagues, knowledge by demonstration of completion of CPD requirements, skills by peer review and analysis of critical incident reporting and integration by meeting quality assurance targets and clinical audit.

How to use the document

The Best Practice Guidelines (Medical management of infants with significant congenital hearing loss identified through the National Newborn Hearing Screening Programme, 2004) describe in detail the procedure for carrying out aetiological investigations. This document identifies the attitude, skills, knowledge, and integration of these attributes in which the doctor needs to be competent, in order to address the unique needs of the child and family as set out in the Best Practice

Guidelines.

Family Friendly Hearing Services is a term used in Audiology to describe a paediatric audiology service that works in partnership with the family and other services i.e. health, education, social, voluntary and private sectors to provide support for hearing impaired children and their families. This document endeavours to uphold the principles of a Family Friendly Hearing Service as described by Baguley, Davis and Bamford; British Society of Audiology News, 29, (2000) p35-39.

Knowledge

Knowledge of all possible causes of hearing loss including the following:

Prenatal:

- Congenital infections
- Syndromes
- Genetic deafness (Genetic conditions may present post nately)
- intra-uterine growth retardation

Perinatal

- Prematurity
- Hyperbilirubinaemia
- Metabolic acidosis
- Hypoxia

Postnatal:

- Meningitis
- Ototoxicity/susceptibility to ototoxicity

Knowledge and causes of auditory neuropathy/dys-synchrony

Know investigations required to eliminate different causes

Know limitations of each test

Understand risk/benefit analysis of each test

Be familiar with technology used for confirming hearing loss

Skills

Have good communication skills (including sharing information)

Have good organisational skills. Be able to work in a team

Integration

Maintain a system for receiving, documenting, collating results and informing parents

Recognise the need for consultation with other specialists e.g. Ophthalmologist/radiologist/cardiologist/geneticist/virologist/other

Provide written information in an accessible form which is appropriate to local needs

Contribute to care plan with information about planned investigations

Be able to access services for investigations

Copy all professional correspondence to parents where appropriate and requested

Core Investigations

a) History

Attitude/ Family Friendly Service

Be sensitive to parents' feelings in the wake of what may be a devastating diagnosis

Allow child to be present

Be aware that history taking may be an ongoing process

Be aware of cultural issues

Make child central to discussion

Knowledge

Causes of deafness (refer to general section)

Developmental milestones

Cultural issues

Skills

Ability to take a thorough paediatric medical history to include the following

Pregnancy

Peri natal

Post natal

Take detailed family history (not just deafness)

May require a summary from Obstetrician and the Neonatal Paediatrician.

Draw family tree

Ask about

- Head injury
- Medication
- Ear disease
- Neonatal meningitis
- Viral illness
- Immunisation status
- parental view of hearing

Take social history

Integration

Check Maternity/SCBU/NICU notes

b) Examination

Attitude/ Family Friendly Service

Be sensitive to parents and child

Refer to child by name

Knowledge

Know syndromes involving head and neck which are associated with deafness

Know ocular syndromes associated with deafness

Calculate W index (Waardenburg syndrome)

Recognise the range of normal development

Recognise physical signs in syndromes associated with deafness

Skills

Carry out full paediatric examination including special reference to primitive reflexes

Look for dysmorphism

Examine head and neck, looking for

- eye abnormalities
- external ear abnormalities
- abnormalities of outer ear
- abnormalities of tympanic membrane
- ear pits and tags
- neck abnormalities including branchial sinuses
- abnormalities of palate
- lip pits

Cardio vascular examination

Full age appropriate developmental examination

Integration

Refer to developmental paediatrician if there is developmental delay

Confer with geneticist if syndromal diagnosis suspected

c) Family Audiograms

Attitude/ Family Friendly Service

Be able to explain the reasons for carrying out family audiograms

Be sensitive to the possibility of finding a previously unsuspected hearing loss in parents/siblings

Knowledge

Be aware of variability of phenotypic expression of the same gene

Knowledge of different types of inheritance

Skills

Obtain audiometry on all first degree relatives including older siblings (who may be too young for testing with headphones)

Integration

Ensure age appropriate audiology assessment is done for children aged under 3 years

Refer parent to adult audiology service

Consider further investigation of parent/sibling with hearing loss

Avoid duplication

d). Specific investigations

1. Imaging

Attitude/ Family Friendly Service

Be sensitive to parents' wishes re. ionising radiation and possible need for GA

Knowledge

Know the anatomical abnormalities associated with hearing losses, involving

- cochlea
- middle ear
- IAMs
- associated structures

Recognise common conditions seen on MRI brain, which affect hearing e.g. toxoplasmosis

Choose appropriate timing for investigations

Know when to request renal ultrasound

Skills

Explain to parents

- diagnostic possibilities
- radiation risk
- sedation/anaesthetic risk
- findings — if no abnormality found
— if abnormality found

Be able to give relevant clinical details when requesting imaging

Be able to interpret CT/MRI of skull base

Be able to demonstrate abnormalities to parents

Integration

Discuss with radiologist, ENT, neurologist

2. ECG

Attitude/ Family Friendly Service

Be sensitive to parents concerns about this investigation

Knowledge

Have knowledge of long QT syndromes, in terms of.

- prevalence
- association with HL
- association with vestibular abnormalities

Understand changes in QT interval with age,

Skills

Be able to calculate QTc

Integration

Explain findings to parents

Involve cardiologist to check ECG

Refer to paediatric cardiologist if long QTc found

3. Genetics

Attitude/ Family Friendly Service

Be in tune with parents' beliefs, culture and value systems

Avoid negative language

Use simple language, with careful explanation of technical terms

Be sensitive to parents' feelings with regards to blame

Be sensitive to parents' anxiety about blood tests

Knowledge

Know the common syndromes associated with hearing loss including chromosomal abnormalities

Know where to obtain correct blood request forms –

Know which sample bottles to use

Know how to store specimens

Know which test to request: e.g.

- Connexin 26 and 30 mutations (and other connexin mutations)

- Mitochondrial mutations causing hearing loss including AI 555G

- Pendred gene

- Waardenburg genes

- Other tests which may be available on a research basis

Understand that tests for further genes may become more readily available in future and that bloods samples may be stored for this purpose.

Understand the relevance of each test to other positive findings

Know the method used for each test

Understand the feasibility of the test

Know which laboratory performs the test

Know whether it is being done as a service or as research

Know the cost / funding implications

Know how to access written information on specific conditions and basic genetic principles

Skills

Be able to interpret the result with reference to:

- mendelian genetics
- chromosomal abnormalities
- mitochondrial abnormalities

Perform basic genetic counselling with regard to recurrence risks

Where there is no written information available, be able to signpost suitable material.

Integration

Refer to geneticist according to local protocols

4. Infection screen

Attitude/ Family Friendly Service

Recognise the urgency for doing these investigations and explain this to parents

Skills

Be able to recognise signs and symptoms of congenital infections

Be able to interpret the results

Knowledge

Know where to obtain correct blood request forms

Know which sample bottles to use

Know how to store specimens

Select test depending on age of baby (see Best Practice guidelines)

Integration

Discuss results with virologist where necessary

Refer to paediatrician where necessary

5. Ophthalmology

Attitude/ Family Friendly Service

Be sensitive to parents' feelings about another possible problem for their child

Knowledge

Be aware of syndromes/conditions which affect vision and are associated with hearing loss

Understand pattern of presentation of different syndromes

Understand the limitations of ophthalmic investigation and examination in babies

Skills

Examine for red reflex

Examine for strabismus

Inform parents about the need for continued monitoring of vision

Integration

Understand the implications of dual sensory impairment

Refer to ophthalmologist (see NDCS/Sense Quality Standards 2004)

Liaise with orthoptist

Refer to Sensory Impairment service where necessary

Additional Investigations

6. Haematology

Attitude/ Family Friendly Service

Be aware that blood tests are an invasive procedure

Be sensitive to parents' feelings about blood tests

Knowledge

Know the conditions where tests are more likely to give positive result

Skills

Be selective about which tests to use

Integration

Discuss with haematologist

Refer to paediatrician if required

7. Biochemistry

Attitude/ Family Friendly Service

Be sensitive to parents' feelings about blood tests

Skills

Be selective about which tests to use

Discuss predictive value of tests

Knowledge

Be familiar with signs and symptoms of conditions having biochemical basis which are associated with hearing loss Understand the natural history of conditions associated with biochemical abnormalities

Be aware of the association of Pendred syndrome with abnormalities within the inner ear

Know the other tests available to help confirm the diagnosis e.g. perchlorate test, renal scan

Integration

Refer to paediatric endocrinologist if required

Refer to geneticist if required

8. Urine examination

Attitude

Be sensitive to parents' feelings if metabolic/life threatening disease is suspected

Skills

Practicalities of urine collection in small babies

Knowledge

Know which tests to request

Understand the natural history of the condition

Integration

Refer to paediatrician

The National Committee of Professionals in Audiology (NCPA)

The NCPA has now been in existence for 14 years and had its 60th meeting on October 19th 2004.

It was set up in 1990 as an independent committee representing the interests of the entire range of professional groups active in the field of Audiology. Thus it has a unique status in that it can be said to be a voice for the whole Audiology profession.

Each of the member organisations is represented by a single committee member who acts as link between the NCPA and their own professional organisation and provides a briefing about the activities of their own professional group at each meeting.

Clerical support is provided by the BSA entirely free of charge and meetings are arranged on a quarterly basis at the National Hospital, Queens Square, London. Our main point of contact is through Ellen Godden at the *British Society of Audiology*, 80 Brighton Rd, Reading RG6 1PS. Tel. 0118 966 0002 or by email to ellen@thebsa.org.uk

The following organisations have a seat on the committee :

- BAA — British Academy of Audiology
- BAAP— British Association of Audiological Physicians
- BACDA—British Association of Community Doctors in Audiology
- BAO-HNS - British Association of Otolaryngologists – Head & Neck Surgeons
- BATOD— British Association of Teachers of the Deaf
- BAEA— British Association of Educational Audiologists
- RCSLT — Royal College of Speech and Language Therapists
- BSHAA— British Society of Hearing Aid Audiologists.

In addition to the above organisations five “observer” organisations sit on the committee :

- BSA — British Society of Audiology
- DOH— Department of Health
- NDCS — National Deaf Children’s Society
- PASA— NHS Purchasing and Supply Agency
- RNID — Royal National Institute for Deaf People

The National Committee of Professionals in Audiology is thus unusual in that it is very broad-based, consisting of representatives from the field of education as well as healthcare, from charities as well as professional bodies, from the private as well as the public sector.

The primary aims of the NCPA are to provide a national forum for debate of professional issues, for the sharing of information and as a sounding board for new ideas. As NCPA represents all the associations it is able to view audiological issues from a wider perspective than may be possible in totally profession based organisations. It is therefore in a very good position to act as a channel of access between professional associations and the relevant government departments.

NCPA will also, via working parties, produce position statements, guidelines and statements of best practice on a very wide range of issues, for example UNHS, lost/damaged hearing aids, classroom acoustics and service provision. Currently we intend to look at issues surrounding Cochlear Implants such as funding and the demands of an ever increasing client base faced by many teams across the U.K..

NCPA is the only place where all professionals groups in Audiology meet regularly. Its strength lies in its wide membership (approximately 10,000 members represented across all the organisations) with a common interest in the provision of a high quality service for the hearing-impaired including generic/specialist speech and language therapists who support deaf children and adults.

There is no other single representation of the UK Audiology Profession and without it there is no mechanism for making profession wide decisions.

Simon Blake (Chair NCPA)

Acknowledgements to Lucy Handscombe and Roger Lewin (past chairs of NCPA) for their help in compiling this article.