

Healthy Hearing Program



Medical Guidelines for Children born with a Permanent Hearing Loss

Table of Contents

1.0	Introduction		
	1.1	Background	5
	1.2	Aims	5
	1.3	Why investigate the cause of hearing loss	6
	1.4	Definition of terms	7
	1.5	Family centred care	7
2.0		Companion documents	8
3.0		Algorithm	9
4.0		Guidelines	10
	4.1	Audiology	10
	4.2	Queensland Hearing Loss Family Support Service	11
	4.3	Medical Evaluation	11
	4.4	ENT	14
	4.5	Paediatrician	16
	4.6	Clinical Geneticist	17
	4.7	Ophthalmologist	18
	4.8	Radiology	18
	4.9	Neurology	20
Appendices			
Appendix 1		Healthy Hearing Program Summary	
Appendix 2		Hearing Loss Management Summary	
Appendix 3		Medical Guidelines for the Assessment of Children with Permanent Hearing Loss - summary	
Appendix 4		Poster	
Appendix 5		Parent Handbook	
Appendix 6		Investigation pages for the Personal Held Record	

Bibliography

21

Document title: Medical Guidelines for children born with a Permanent Hearing Loss

Publication date: March 2010

Replaces document: New document

Author: Healthy Hearing Program

Audience: Health professionals in Queensland Health private children's health services

Exclusions: Nil

Review date: 2012

Endorsed by: Hearing Loss Medical Guidelines Advisory Group

Contact: Anne Geddes Project Officer, QHLFSS
Dr Alison Harris Paediatrician

Disclaimer

These guidelines have been prepared to promote and facilitate standardisation and consistency of practice, using a multidisciplinary approach.

Information in this guideline is current at time of publication.

Queensland Health does not accept liability to any person for loss or damage incurred as a result of reliance upon the material contained in this guideline.

Clinical material offered in this guideline does not replace or remove clinical judgement or the professional care and duty necessary for each specific patient case.

Clinical care carried out in accordance with this guideline should be provided within the context of locally available resources and expertise.

This Guideline does not address all elements of standard practice and assumes that individual clinicians are responsible to:

- **Discuss care with consumers in an environment that is culturally appropriate and which enables respectful confidential discussion. This includes the use of interpreter services where necessary.**
- **Advise consumers of their choice and ensure informed consent is obtained**
- **Provide care within scope of practice, meet all legislative requirements and maintain standards of professional conduct**
- **Apply standard precautions and additional precautions as necessary, when delivering care**
- **Document all care in accordance with mandatory and local requirements**

Abbreviations

CI	Cochlear implant
CMV	Cytomegalovirus
CT	Computerised tomography
DBS	Dried blood spot
EVA	enlarged vestibular aqueduct
FH	Family history
FSF	Family Support Facilitator
GA	General anaesthetic
HH	Healthy Hearing
HIE	Hypoxic ischemic encephalopathy
IAM	Internal auditory meati
IgG	Immunoglobulin G
IgM	Immunoglobulin M
IPPV	Intermittent positive pressure ventilation
JLNS	Jervell and Lange-Nielsen syndrome
MRI	Magnetic resonance imaging
PAP	positive airway pressure
PCHL	Permanent childhood hearing loss
PCR	Polymerase chain reaction
PHL	Permanent hearing loss
PHR	Personal health record
QHLFSS	Queensland Hearing Loss Family Support Service

New born screen -
 Guthrie card
 DBS

} interchangeable

Medical Guidelines for Children Born with a Permanent Hearing Loss

1. Introduction

1.1 Background

Children born with a permanent hearing loss run the risk of life long speech deficits and delays in the acquisition of language. Deficits in speech and language may result in poor academic performance, psycho-social difficulties, behaviour and emotional problems.

Technological developments in recent years have widened the scope of potential outcomes for the development of communication skills for children born with a permanent hearing loss. This together with early screening processes and research providing evidence of the value of prompt early intervention services (Yoshinaga-Itano, 2004) provide a window of time for making a real difference in long term outcomes for children with PHL and their families. To make the best use of these opportunities, it is necessary to develop and set in place efficient systems to guide and co-ordinate parents and practitioners through the complex medical assessments and investigations involved with this diagnosis.

Up to 40% (Moeller, 2007) of children with a hearing loss have additional health needs or developmental problems and 20% have more than 2 (Fortnum and Davis 1997, 1998). The complexity of referral pathways and the number of specialities and practitioners that may be involved, make medical guidelines essential in order to deliver well co-ordinated, quality service and to avoid any replication or unnecessary procedures (both from a patient safety point of view and financially). Accurate information and record keeping shared between parents and their medical contacts can lead to greater engagement, clearer understanding and enable informed decision making in the care pathway process.

Since the Queensland Healthy Hearing Statewide Screening programme commenced in 2004, children with hearing loss are now being identified in their first few months. Having identified these children, we need to have effective, efficient pathways for the management of their future health, development and family needs.

1.2 Aims of the medical guidelines for children born with permanent hearing loss

1. To ensure effective, efficient and accurate assessment, diagnosis and management of children born with permanent hearing loss.

2. To ensure consistency and equity of access to quality medical services for all children with PHL statewide.
3. To ensure that medical evaluation and diagnostic procedures are evidence based reflecting best practice.
4. To provide a basis for collection of accurate data for evaluation and reporting purposes.
5. Continued monitoring of current practice via consultation by the Medical Guidelines Advisory Group.

1.3 Why investigate the causes of hearing loss?

- To answer the parents question 'Why is my child deaf?'
- Identification of associated conditions, medical or developmental problems.
- The results of investigations can assist the health practitioners to make informed decisions about the most appropriate management plan in terms of frequency of follow up, cross referrals to other specialities, amplification needs, further investigations, educational programmes and family counselling etc as indicated.
- To give families advice about likely outcomes, progression of hearing loss, complications which may arise and prevention of complications eg. for those children with cardiac conduction defects.
- To inform genetic counselling.
- Epidemiological information for research and planning, surveillance and prevention of hearing impairment.

Identifying the cause and extent of the hearing loss can be a long ongoing process and in 30-40% of cases, the aetiology remains unknown. (Parving, 1984, Maki-Torkko 2003)

The timing of investigations will depend on the family's readiness to proceed with the investigations and how well the child can cooperate with the tests and other factors such as timing of anaesthetics. Investigation for causes of hearing loss may be delayed if the child has additional medical problems.

While we acknowledge the evidence supporting a sequential pattern of investigation, (Preciado, 2005) clinical reasoning needs to be applied to strike a balance between cost effectiveness and the least intrusive process of care for the child and their family.

Reasons for carrying out aetiological investigations early:

- CMV: to monitor development and progression of hearing loss.
- EVA: to educate parents with a view to minimising fluctuations/progression of hearing loss
- Long QT: to minimise risk of cardiac complications
- Early genetic opinion on recurrence risks and syndromal diagnosis
- Some investigations could be carried out under natural sleep
- Early intervention for both the hearing loss and all the developmental needs of the child can be planned and managed.

“Clinical Pathways never replace clinical judgement. Care outlined in this pathway must be altered if it is not clinically appropriate for the individual patient”.

1.4 **Definition of Terms**

Permanent hearing loss (PHL) – permanent hearing loss, which includes both sensorineural hearing loss and permanent conductive hearing loss, and mixed hearing loss (sensorineural and conductive), but excludes temporary conductive hearing loss such as associated with otitis media with effusion (OME) or “glue ear”. For the purpose of this document PHL includes both bilateral and unilateral HL, and any degree of severity (mild, moderate, severe and profound). The document will most commonly be utilised for children with congenital PHL, but also is relevant to children with late-onset PHL such as that due to congenital CMV infection or genetic progressive hearing loss.

PCHL – permanent childhood hearing loss

1.5 **Family Centred Care**

Medical investigations including aetiology needs to be available to all children diagnosed with HL with full parent involvement.

“Parents must be given comprehensive and unbiased information about the medical investigation which is carried out to identify the cause of the hearing impairment and the diagnosis and treatment of any co-existing conditions, including both the benefits and the disadvantages, and be given every opportunity to further discuss their views and concerns with the doctor so that they can make an informed decision about whether they want their child to have these investigations and if so, the nature and timing..... Parents must be kept informed at every stage of the investigations and should have reasonable access to the doctor for further information and explanation as required.”*

* Medical management of infants with significant congenital hearing loss identified through the national newborn hearing screening programme- Best Practice Guidelines (NHSP in conjunction with BAAP and BACADA)2004

- **Dignity and respect** needs to be shown for the family and their part in the decision making process and their right to decline an investigation if they feel it is not right for them. Opportunities should be provided to revisit these decisions at a later stage, being mindful that the adjustment stage may influence readiness/ openness to investigations. There needs to be respect and understanding for the adjustment stages the families of children with this diagnosis will be going through.
- **Information sharing-** The parent booklet that forms part of the Personal Health Record contains information on the roles of the various professionals that may be involved after a diagnosis of hearing loss. It also contains details of the types of investigations and procedures that may be required as well as information on developmental milestones etc.
- **Participation-** Providing detailed information to parents enables them to make informed decisions around management planning of the hearing loss and future investigations. It gives them the rationale for the importance of

early intervention for optimum outcomes in the development of their child’s speech and language, promoting the need for timely medical investigation.

- **Collaboration** - The personal health record provides opportunities for a collaborative approach between families and professionals with the family holding the child’s information and sharing it across sites. It relies on the clinician to be an active participant in maintaining this record.

2. Companion Documents

- Hearing Loss Management Summary
- Algorithm
- Summary sheet
- Poster
- Parent booklet
- Investigation results

} Part of the Personal Health Record

Resource	Application
Medical Guidelines Forms	<p>Hearing loss Management Summary</p> <ul style="list-style-type: none"> • Filed in the child’s medical chart when children are first diagnosed with hearing loss. • Investigation results and referrals to be entered by the initiator. <p>Algorithm</p> <ul style="list-style-type: none"> • Used as a clinical guide and aide-memoire <p>Summary Sheet</p> <ul style="list-style-type: none"> • Used as a clinical guide with greater detail and text form
Medical Guidelines Parent information	<p>Poster</p> <ul style="list-style-type: none"> • A flow chart of the medical assessment pathway <p>Parent Booklet</p> <ul style="list-style-type: none"> • Information for parents on medical investigations, referrals and the roles of the different professionals that may be involved. Timelines, developmental stages etc. <p>Investigation pages</p> <ul style="list-style-type: none"> • Used to record information and test results. To be taken to all medical appointments and updated. • These pages and the booklet are additions to be added to the child’s Personal Held Record. They form part of the Possibilities and Pathways kit that is given to families of children with hearing loss (by QHLFSS or Audiology).

Available to download from the Healthy Hearing website www.health.qld.gov.au/healthyhearing

4. Guidelines

4.1

Audiology

- Confirm permanent sensorineural or conductive hearing loss
- Progress referral to ENT
- Send hearing assessments results to relevant professionals
- Progress referral of parents and siblings for hearing assessment

On the completion of the diagnostic process, the diagnostic audiologist is responsible for:

- Progressing a referral to an Ear Nose and Throat Specialist (ENT)
- Sending the assessment results to relevant professionals, including:
 - Ear Nose and Throat Specialist
 - Queensland Hearing Loss Family Support Facilitator –if accepted
 - General Practitioner/Family Doctor
 - Paediatrician present at child's birth
 - Paediatrician child will visit for post-diagnostic assessment if different from original Paediatrician present at the child's birth
 - Neonatologist (if applicable)
 - Australian Hearing
 - Habilitation centre/s once parents have made these decisions
 - Other relevant professionals
- Facilitating parent and sibling hearing assessment
 Except in circumstances it can be confidently ruled out that the child's hearing loss was acquired, it is recommended that hearing assessment of both the infant's parents and all of the infant's siblings should be undertaken even where newborn hearing screen was negative.

If there is any doubt about whether or not the child's hearing loss was acquired, family hearing assessment should be undertaken.

In order to reduce the travel demands on families living in regional, rural or remote areas, parent and sibling hearing assessment may be able to be undertaken at a location other than the original diagnosing centre (e.g. other Queensland Health facility, private audiologist, Australia Hearing). The diagnosing audiologist will facilitate the referral process to alternate services where this is required.

4.2 Queensland Hearing Loss Family Support Service- QHLFSS

QHLFSS offer support to families following newborn screening 'double refer' results. Typically Family Support Facilitators (FSF) contact families from the time of audiology confirmation of diagnosis of hearing loss. If a parent chooses to include a FSF in their ongoing support, the FSF will be available to assist them to co-ordinate their access to the post-diagnostic aetiological, developmental and health assessment processes.

Examples of the roles FSF may fulfil, in collaboration with other professionals and services, include assisting families:

- Through the delivery of information and guidance to support parents in planning and making informed decisions
- Through the delivery of information that is timely and supports understanding.
- Through promoting adjustment and positive emotional and health outcomes for families and children.
- Through support and advocacy, that includes negotiating and assisting co-ordinated approaches to the provision of services.
- To explore early intervention and communication options that are appropriate to needs.
- To understand the outcomes and implications of the integrated picture of the different assessments undertaken.

4.3

Medical Evaluation

This may be carried out by the ENT and/or the Paediatrician

All children

- Child and family history, physical examination
- Order primary blood tests
- Order CMV investigation from Guthrie Cards (DBS)

Child and Family History

- **Antenatal history**
 - immunization for rubella- IgG>10 IU/ml
 - screening for syphilis
 - drug use
 - alcohol use
 - spontaneous abortions
 - exposure to ototoxic medications (aminoglycosides, platinum derivatives, loop diuretics, quinine derivatives, salicylates)
 - 👉 risk factors for PHL
 - in-utero infection
 - 👉 risk factor for PHL- toxoplasmosis, CMV, herpes, rubella, syphilis
 - medications taken
 - antenatal ultrasound
 - other interventions

- other complications
- **Birth**
 - weeks gestation (X/40)
 - spontaneous/induced/reason for induction
 - mode
 - duration
 - delivery complications
 - interventions
 - instrumentation used
 - post partum complications (mother)
 - post partum interventions (mother)
- **Post-natal history**
 - Apgar 1 minute, Apgar 5 minutes
 - Birth weight 🔗 risk factor for PHL: $\leq 1500\text{g}$
 - Length, head circumference
 - Complications on delivery
 - Interventions on delivery
 - Post-partum complications (mother)
 - Post-partum interventions (mother)
 - NICU admission and duration
 - SCN admission and duration

 - Respiratory status at birth/ resuscitation 🔗 risk factor for PHL
Severe asphyxia
(Convulsions/HIE)
 - Ventilation- number of days 🔗 risk factor for PHL
 ≥ 5 days (IPPV/PAP)
 - Hypoxic/anoxic episodes in post-natal period
 - Maximum serum bilirubin level 🔗 risk factor for PHL
Term: $\geq 450\mu\text{mol/l}$,
Preterm: $\geq 340\mu\text{mol/l}$
 - Phototherapy
 - Abnormalities noted at birth 🔗 risk factor for PHL
Craniofacial anomalies
(excluding cleft lip and
skin tags)
 - Diagnosed/ suspected syndrome 🔗 risk factor for PHL:
syndrome
 - Other investigations
 - Immunizations
 - Early feeding: breast, bottle, tube fed, issues?
- **History of exposure to risk factors** 🔗
 - Bacterial meningitis (proven/suspected)
 - Head injuries
 - Proven infections toxoplasmosis, CMV, herpes, rubella
 - Suspected infections toxoplasmosis, CMV, herpes, rubella
 - Antibiotics
 - Ototoxic medications
 - Other medications

○ **Three generation family tree**

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

- Grandparents
- Parents
- Siblings

🔗 Risk factors associated with hearing loss in first and second degree relatives
 -structural renal problems (Alport Syndrome), goitre in Pendred's syndrome

○ **Family audiograms**

A hearing assessment is recommended for first degree relatives. Previously unsuspected audiometric abnormalities may be identified. In families where there is a history of hearing loss, the degree of hearing loss may be variable within the same condition (eg, Waardenburg syndrome). Siblings need to be retested even if their newborn screen was negative.

Physical Examination

Including:

- head
- face
- neck
- eyes
- ears
- hands
- feet

Investigations

○ **Order Primary Blood Tests**

- Full blood count
 - Urea
 - Electrolytes
 - Serum creatinine
 - Glucose
- } to exclude renal function abnormalities and where conditions such as Alport or Alstrom's syndromes are suspected

○ **CMV PCR from newborn screening card**

○ **Order CMV, Toxoplasma, rubella and syphilis serology**

CMV

- Detection of CMV by PCR in saliva, blood or urine or positive IgM prior to 3 weeks of age suggests congenital CMV infection.
- After 3 weeks of age, viral detection or IgM could be due to either congenital or postnatally acquired infection. Postnatally acquired infection would not usually be associated with adverse outcome.
- Detection of CMV by PCR from newborn screening card suggests congenital CMV infection.

Rubella

- IgM positivity prior to 6 months suggests congenital rubella infection
- Falling or absent IgG after 9 months of age suggests absence of infection

- Between 6 months and 3 years, absence of rubella specific T cell response (heparinised blood) excludes rubella.

Toxoplasmosis

- IgM or blood/CSF PCR positivity prior to 6 months of age suggests congenital infection.
- IgG titre significantly higher than mother's IgG titre suggests congenital infection
- IgG positivity after 6 months of age suggests congenital infection.

Syphilis

- Positive serology in baby warrants further investigation eg IgM testing and discussion with ID specialist

o **Genetics**

- Chromosomes - if there is developmental delay or dysmorphic features
- Connexin 26- the 30delG (35delG) mutation screen - unless clear diagnosis of a syndrome associated with hearing loss. Expanded genetic testing by Genetic Health Queensland as needed.

o **Urine**

- All children –protein, microscopy
- CMV PCR if DBS positive
- Urine metabolic screen if there is developmental delay or failure to thrive

4.4

ENT

All children

- o Physical examination of head, neck
- o Refer to Australian Hearing, Paediatrician, Ophthalmologist, Clinical Geneticist*
- o Order primary blood tests
- o Discuss CT and MRI with all parents
- o Order CMV investigation from Guthrie Cards

Where indicated

- o Refer for CT
- o Refer for MRI
- o Refer for renal ultrasound
- o Refer for ECG

*except if syndromic or non-syndromic genetic aetiology can be confidently ruled out

CT and MR Imaging

Discuss the purpose of CT (Computerised Tomography) and MR (Magnetic Resonance) imaging with ALL families. CT and MR imaging are known to provide complementary information rather than one or the other procedure providing complete information alone.

CT scan involving radiation and showing the bony structures including middle ear ossicles.

Recommend CT petrous temporal bone, brain scan imaging for all children with:

- severe bilateral sensorineural hearing loss or greater
- progressive unilateral or bilateral hearing loss
- auditory neuropathy
- structural renal abnormalities (or as indicated)

MRI showing the soft tissue- brain, VIIth and VIIIth nerves and membranous labyrinth including the endolymphatic sac.

Recommend MRI inner ear and internal auditory meatus (IAMs), brain scan imaging for all children with:

- severe bilateral sensorineural hearing loss or greater
- progressive unilateral or bilateral hearing loss
- auditory neuropathy
- structural renal abnormalities (or as indicated)

To ensure the quality of images, a GA is required for all children up to approximately 4 years of age.

Unless clinically indicated (eg. Assessment for early cochlear implantation), it is recommended that imaging does not occur on children under 6 months in order to minimise GA risks.

CT and MR imaging are only able to be completed with infants under GA at:

Royal Children's Hospital
Mater Children's Hospital
Mater Private Hospital
The Townsville hospital
Gold Coast hospital

Renal Ultrasound

- Children with suspected branchio-oto-renal syndrome- where pre-auricular pits, branchial sinuses or cysts are present
- Multiple or multi system abnormalities
- Family history of structural renal problems
- Mondini dysplasia on imaging

Electrocardiography ECG (with holter tape)

- Children with severe bilateral sensorineural hearing loss or greater
- May need repeating when the child is older
- ECG needs to be interpreted by a Paediatric Cardiologist
- If QT interval > expected for age, refer to a Paediatric Cardiologist
- If QT interval > expected for age, refer other family members for an ECG

Jervell and Lange-Nielsen syndrome (JLNS) is a rare autosomal recessive syndrome with long QT interval and sensorineural hearing loss due to the homozygosity for mutations in the *KVLQT1* or *KCNE1* genes.

Referrals

- **Hearing Aid Clearance and referral to Australian Hearing** (where appropriate)

○ **Referral to Paediatrician**

A number of paediatricians who have a specific interest in working with children identified with permanent hearing loss will be identified at each of the following hospitals:

- Royal Children’s Hospital
- Mater Children’s Hospital
- The Townsville Hospital
- Gold Coast Hospital

All children will be offered an assessment by a paediatrician trained in the guidelines for developmental follow-up of children diagnosed with PHL at one of these hospitals. Children diagnosed, will be engaged with a paediatrician and they may wish to maintain this relationship or can be referred for routine developmental follow-up at 4 to 6 months, 12, 18 and 24 months.

○ **Referral to a Clinical Geneticist – (Genetic Health Queensland)**

-request for assessment at 6 months.

○ **Referral to Ophthalmologist**

-request for assessment at 6 -12 months. Assessment of visual acuity and fundoscopy.

40% of children with sensorineural hearing loss have additional needs and/or ophthalmic conditions. Problems may be non-specific to any underlying condition such as a squint or refractive errors or other findings on examination may help to clarify diagnosis such as Usher Syndrome, CHARGE, congenital CMV or rubella.

4.5

Paediatrician

All children

- Child and family history
- Physical, developmental, health examination
- Blood, urine and CMV if not already requested

Where indicated:

- Order chromosome test
- Order metabolic screen

The assessment by the Paediatrician will include:

- Child and family history
 - Examination
- } If not already done by ENT
- Developmental history
 - Examination
 - Growth parameters
 - Development

- Neurological assessment
 - Cardiovascular system
 - Head
 - Face
 - Neck
 - Skin
 - Chest
 - Abdomen
 - Limbs
 - Nails
- Developmental Assessment
 - Order Chromosome test when:
 - Developmental delay evident
 - Dysmorphic features evident
 - Order metabolic screen when:
 - Child suspected of failure to thrive
 - Developmental delay noted

4.6

Clinical Geneticist- Genetic Health Queensland (GHQ)

- Assessment at 6 months

Not required if syndromic or non-syndromic genetic aetiology can be confidently ruled out

To ensure families have the most timely access to information relevant to their future family planning, it is recommended that assessment by a Clinical Geneticist occurs when the child is approximately 6 months of age, and preferably no later than 9 months.

GHQ provides services in Brisbane, Gold Coast, Nambour, Toowoomba, Ipswich, Bundaberg, Hervey Bay, Mackay, Mt Isa, Rockhampton, Townsville and Cairns.

The assessment by the Clinical Geneticist may include:

- Family History
- Physical Examination
- Examination and Measurement of craniofacial region, +/- W index
- Assessment as indicated for:
 - **Connexin 26** – common mutations, followed by full sequencing if necessary
 - not indicated in context of clear syndromic aetiology
 - not indicated where 3 generation family history indicates obvious dominant inheritance

- if one mutation found on Connexin 26, then Connexin 30 should be completed
- **Connexin 30** – common mutations, followed by full sequencing if necessary
 - not indicated in context of clear syndromic aetiology
 - if one mutation found on Connexin 26, then Connexin 30 should be completed
- **Pendred**
 - all children with large vestibular aqueduct syndrome (LVA)
 - all children with cochlear dysplasia
 - all children with hypothyroidism
 - not indicated in context of clear syndromic aetiology
 - not indicated where 3 generation family history indicates obvious dominant inheritance
- **A1555G (mitochondrial DNA mutation)**
 - All children with exposure to aminoglycosides
 - All children with a pedigree indicative of mitochondrial inheritance/ maternal inheritance
- **Biochemical genetic assessment**
 - as determined by Geneticist
- **Chromosome assessment**
 - all children with developmental delay
 - all children with dysmorphic features

4.7

Ophthalmologist

- Assessment at 6 months
- Assessment at 18 months
- ERG at 6 years if cause of hearing loss not identified through other means

Approximately half of children with severe to profound hearing loss will also have ocular abnormalities. The majority of these will be refractive errors but other ocular pathologies may contribute to aetiology and confirm a syndrome (for example, Usher) or suggest a congenital infection (for example, congenital rubella).

Initial Assessment – 6 months

- It is recommended that all children are referred for assessment by an ophthalmologist for indicators of Usher Syndrome Type 1
- Children who show signs of night blindness should be assessed by an Ophthalmologist for indicators of Usher Syndrome
- Children who have had no known aetiology for their hearing loss identified by 6 years of age should be assessed using electroretinography to identify indicators of Usher Syndrome.

4.8

**Radiology
CT and MRI**

- CT and MRI after 6 months (earlier if indicated)

▪ **CT and MR Imaging**

- CT and MRI imaging are known to provide complementary information rather than one or the other procedure providing complete information alone. However, for a child who is known to have been totally deaf from birth, it is recommended that MRI is completed first to determine the presence or absence of the cochlear nerve and identification of other congenital abnormalities of the inner ear.
- Recommend CT and MR imaging for all children with:
 - All children with severe bilateral sensorineural hearing loss or greater
 - All children with progressive unilateral or bilateral hearing loss
 - All children with auditory neuropathy
 - All children with structural renal abnormalities
- To ensure high quality images, a GA is required for all children up to approximately 4 years of age
- Unless clinically indicated (e.g. assessment for early cochlear implantation), it is recommended that imaging does not occur on children under 6 months in order to minimise GA risks.
- CT and MR imaging are only able to be completed with infants under GA at:
 - Royal Children’s Hospital
 - Mater Children’s Hospital
 - Mater Private Hospital
 - The Townsville Hospital
 - Gold Coast Hospital

Electrocardiogram and Cardiology Referral

- Infants with severe bilateral sensorineural hearing loss or greater are to be referred for an ECG
- The ECG is to be read by a paediatric cardiologist
- Where ECG results indicate that an infant’s QT interval is greater than expected for their age, referral to a Cardiologist for further management is recommended.

Renal Ultrasound and Nephrology Referral

- Renal ultrasound is recommended for all children with:
 - Suspected branchio-oto-renal syndrome (i.e. pre-auricular pits, branchial sinuses or cysts)
 - Multiple or multisystem abnormalities
 - Family history of structural renal problems
 - Mondini dysplasia on imaging
- For some infants, adequate information may be available from pre-natal ultrasounds.

4.9

Neurology

- The need for referral to a Neurologist will be determined by a relevant medical practitioner (e.g. Paediatrician, ENT or Geneticist)

Bibliography:

- BAAP/BAPA 'Guidelines for aetiological investigation of infants with congenital hearing loss identified through newborn hearing screening' *NHSP Clinical Group Jan 2009*
- BAAP/BAPA 'Medical management of infants with significant congenital hearing loss Identified through the national newborn hearing screening programme- best practice guidelines' 2004
- Mapofmedicine Institute for Innovation NHS 'Permanent childhood hearing impairment management' 2009 healthguidelines-mapofmedicine.com/choices/map-open/index.html
- Joint Committee on Infant Hearing 'Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs' *Pediatrics* 2007; 120: 898-921
- National Deaf Children's Society 'Medical assessments for children with permanent hearing loss' www.aussiedeafkids.org.au/medical-assessments.html
- Bamiou D E, Macardle B, Bitner-Glindzicz M, Sirimanna T 'Aetiological investigations of hearing loss in childhood: a review' *Clinical Otolaryngology*, 1999; 25: 98-106
- Barclay L 'Guidelines issued for hearing assessment in infants, children beyond neonatal screening' www.medscape.com/viewarticle/709855
- Buz Harlor A D, Bower C 'Hearing Assessment in Infants and Children: Recommendations beyond Neonatal Screening' *Pediatrics* 2009; 124:1252-1263
- Espeso A, Owens D, Williams G 'The diagnosis of hearing loss in children: common presentations and investigations' *Current Paediatrics* 2006; 16:484-488
- Declau F, Boudewyns A, Van den Ende J, Peeters A, van den Heyning P 'Etiologic and audiologic evaluations after universal neonatal hearing screening: Analysis of 170 referred neonates' *Pediatrics* 2008; 121: 1119-1126
- MacArdle B, Bitner-Glindzicz M 'Investigation of the child with permanent hearing impairment' *Arch Dis Child Educ Pract Ed* 2010; 95:14-23
- Maki-Torkko E 'Current issues on aetiological evaluation of hearing-impaired infants' *Audiological Medicine* 2003; 1: 3, 185-190
- Nokolopoulo T, Lioumi D, Stamataki S, Donoghue G 2006 'Evidence-based overview of Ophthalmic disorders in deaf children: a literature update' *Otology and Neurology* vol 27 S1-S24
- Preciado D, Lawson L 2005 'Improved diagnostic effectiveness with a sequential diagnostic paradigm in idiopathic pediatric sensorineural hearing loss' *Otol Neurotol* 26(4): 610-615
- Pickett B P, Ahlstrom K 'Clinical evaluation of the hearing impaired infant' *Otolaryngologic Clinics of North America* 1999; vol 32 no 6 1019-1035
- Sanjay M, Westerberg B, Kozak F 'Systematic review of the etiology of bilateral sensorineural hearing loss in children' *International Journal of Pediatric Otorhinolaryngology* 2004; 68: 1193-1198
- Wilson C, Roberts A, Stephens D 'Aetiological investigations of sensorineural hearing loss in children' *Arch Dis Child* 2005 90: 307-309
- Wilson C, Roberts A, Stephens D 'Improving Aetiological Investigation of Permanent Hearing Impairment in Children' *Audiological Medicine* 2004; 2: 241-246
- Quality Standards in Vision Care for Deaf Children and young People (2004) www.sense.org.uk

Members of the Advisory Group

Medical guidelines for children born with a permanent Hearing loss

Ven-nice Ryan	Director, Child Development Program
Dr Alison Harris	Paediatrician
Anne Geddes	Project Officer
Dr Susan Moloney	Paediatrician, Gold Coast Hospital
Dr Fiona Panizza	ENT
Dr David Bell-Allen	ENT
Shirley Glennon	Healthy Hearing, Team Leader
Jackie Moon	Audiology, Mater
Lia Traves	Audiology, CI, RCH
Tim Wood	Team Leader, QHLFSS
Sreedevi Aithal	Paediatric audiologist, Townsville
Nuala Beahan	Audiology, Healthy Hearing
Dr Alan Sive	Paediatrician, Townsville
Heather Price	FSF, Townsville
John Gavranich	Director of Paediatrics
Katrina Roberts	Healthy Hearing, Townsville
Julie McGaughran	Genetic Health Queensland
Dr Anne Kynaston	Paediatrician
Dr Brian Morris	Director of Paediatrics
Richard Heazlewood	Paediatric Outreach
Luke Jardine	Neonatologist
Rachel Susman	Clinical Geneticist

Link to Appendices