

Medical Guidelines for the Assessment of Children with Permanent Hearing Loss (including unilateral loss)

Clinical Assessment

History- pre-natal, post- natal, family history - don't forget 3 generation family tree, audiology results
Examination – general, dysmorphology, head and neck, neurological, developmental



Referrals

Investigations

ALL children

Audiology
Family audiology testing (mother, father, siblings)

ENT
All children ASAP

QHLFSF
(family support facilitator)

Australian Hearing
All children ASAP

Paediatrician
All children ASAP then 4-6 mth, 12, 18, 24 months

Genetics
All children 6-12 months

Ophthalmologist

- All children approximately 6 months
- If not walking at 18 mths and aetiology unknown, review for Usher's Syndrome
- If no known aetiology by 6 yrs, ERG to assess for Usher's Syndrome

ALL children

Blood

- FBC
- U&Es
- thyroid function
- CMV, rubella, toxoplasmosis IgG and Igm, syphilis serology
- CMV PCR from Newborn Screening Card (DBS)
- Chromosomes if developmental delay or dysmorphic features
- Connexin 26 common mutation screen unless clear diagnosis of syndrome associated with HL

Urine

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

as indicated

Radiology

CT petrous bone, brain scan

- children with severe bilateral SNHL or greater
- progressive unilateral or bilateral SNHL
- auditory neuropathy
- structural renal abnormalities (Or as indicated)

MRI inner ear and internal auditory meatus, brain scan

- children with severe bilateral SNHL or greater
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Renal Ultrasound

- children with suspected branchio-oto-renal syndrome: auricular pits, branchial sinuses or cysts
- multiple or multi system abnormalities
- family history of structural renal problems
- Mondini defect on imaging

ECG (+/- holter tape)

- Children with severe bilateral SNHL or greater
- may need repeating when child is older
- interpretation by Paediatric Cardiologist
- if QT interval > expected for age, refer to Paediatric Cardiologist
- if QT interval > expected for age, refer other family members for ECG