

Queensland Health 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

- Audiology**
Family audiology testing (mother, father, siblings)
- ENT**
All children ASAP
- QHLFSS**
(Queensland Hearing Loss Family Support Service)
- 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

Investigations

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 screen**
- children with severe bilateral SNHL or greater
 - children with moderate unilateral SNHL or greater
 - progressive SNHL
 - auditory neuropathy
 - structural renal abnormalities
 - congenital CMV infection (or as indicated)
- 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

ALL children

