

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

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

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

as indicated

ALL children

