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
- 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