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