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Non-routine paediatric case studies

With thanks to Kirsty Waite, Yorkshire Auditory Implant Service

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ANSD case 1 - history

- 11 Year old F
- Premature birth with ventilation required for 6 weeks
- Hearing loss first diagnosed at 16 weeks old following 12 week stay on SCBU
- Hearing aids issued at time of HL diagnosis
- Good use of HAs
- Previously assessed at 12 months old however discharged

- Recent diagnosis of ANSD
- Reports to communicate well when wearing HAs
- Struggles when in background/noisy environment
- Using radio aid at school and has teaching assistant to help when in class

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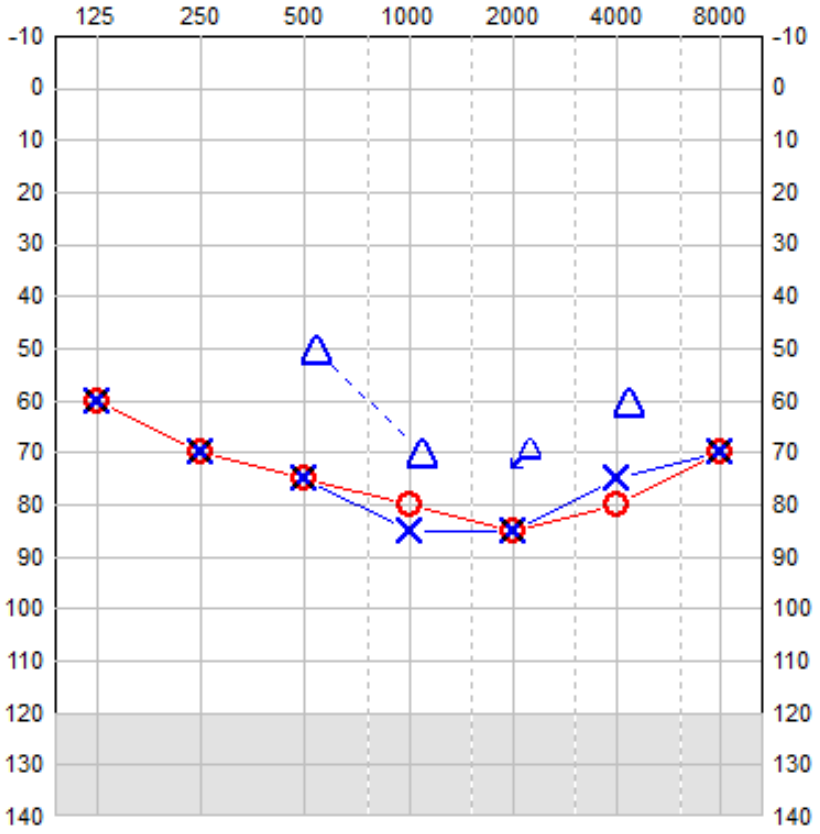
Assessment

- PTA results indicate bilateral severe hearing loss
- Speech test results (tested at 70dBSPL):

  BKB:
  - Bilateral HA(female) = 86%
  - Bilateral HA (male) = 94%
  - Right HA (female) = 86%
  - Left HA (female) = 88%



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Decision

- Decision to discharge at this time
- Although diagnosed with ANSD, patient managing well with HAs
- Likely would not gain any further significant benefit from a cochlear implant

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ANSD case 2 – history

- Referred for CI assessment following diagnosis of ANSD at 10 months
- Bilateral hearing aids issued at diagnosis
- Delay to assessment due to persistent MEE requiring grommets
- VRA assessment revealed profound SNHL
- Limited vocalisations consistent with profound HL
- Lack of responses to sound

Decision - Team decision to proceed with CI as not making progress with HAs

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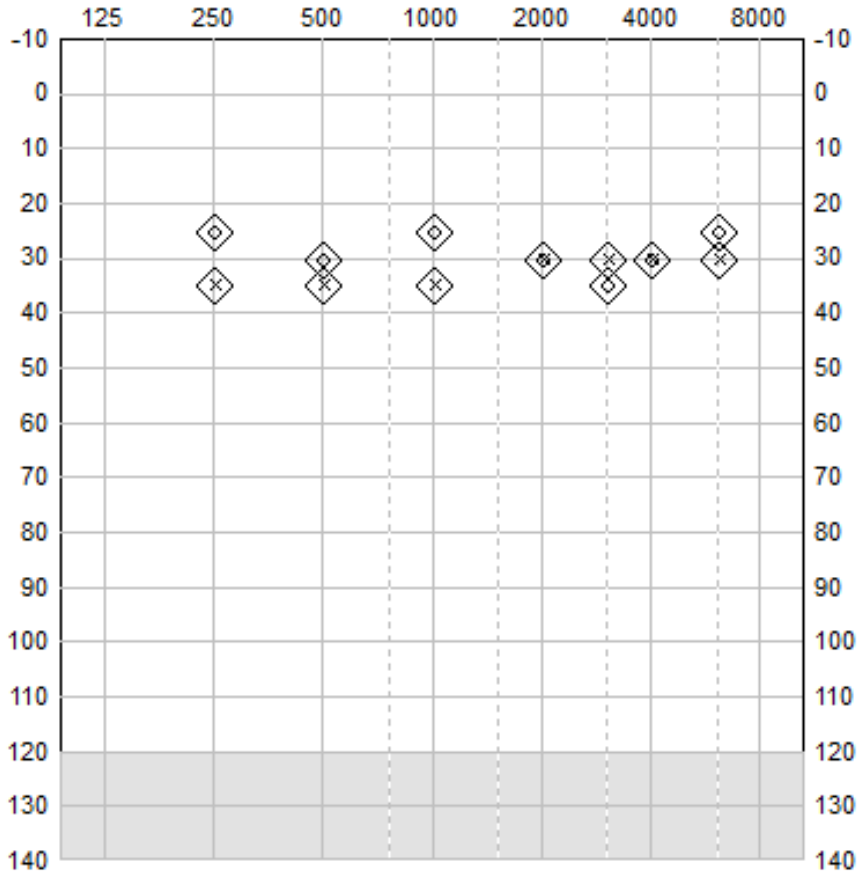
- Bilateral implantation at 20 months
- Excellent aided thresholds bilaterally

- Automated McCormick Toy Test = 39dB(A) bilaterally
- ASSE = 100% bilaterally

- Progressing well at school
- Uses radio aid in class
- Speech has improved – understood by familiar listeners



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Meningitis - Case study 3

- Well baby
- Healthy birth
- Passed NHSP assessment
- Admitted to hospital at 6 weeks old due to meningitis
- ABR testing while inpatient now indicated bilateral profound hearing loss
- MRI scans indicated that ossification was likely to progress
- Shunt required due to hydrocephalus

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Decision

- MDT discussion:
  - unilateral implant to be fitted on the contralateral ear to the side with the shunt
  - dummy implant to be implanted into the side with the shunt to try and keep option open for future bilateral cochlear implants

- Urgent cochlear implant surgery arranged at 11 weeks of age due to progression of ossification.

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Progress so far

- Attended for activation of CI at 6 months old
- Behavioural testing not possible at this time
- Intra-op NRT testing result used to create a MAP
- Initial problems with retention - now improved
- At last appointments - listening down to minimal levels at frequencies assessed so far

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Progress continued

- Vocalisations are now developing and improving
- No parental concerns
- Happy with listening
- Responding to wide range of loud and quiet sounds

- Recent discussion of proceeding to bilateral implants however now has programmable shunt in contralateral ear so not appropriate

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Learning point

Do not delay with meningitis cases

Cochlear ossification happens quickly (weeks and months after recovery)

Assessment of hearing should be priority after recovery

All cases of severe-profound HL should be referred for urgent CI assessment

CI needs to take place prior to ossification

(www.meningitis.org)

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West Syndrome – Case study 4

The National Organisation for Rare Disease defines this syndrome as:

"West syndrome is a constellation of symptoms characterised by epileptic/infantile spasms, abnormal brain wave patterns and intellectual disability"

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- **March 2019** - 4 month old F referred
- Failed newborn hearing screen
- ABR results at diagnosis:

	1kHz	2kHz	4kHz
Right AC	<=90dBeHL	=90dBeHL	>90dBeHL
Left AC	>80dBeHL	=90dBeHL	>90dBeHL
BC			>50dBeHL

(Tympanometry not performed at ABR appointments therefore unable to conclusively know if within current criteria as unable to discount any conductive overlay)

- No known health or developmental issues at referral therefore initially following well baby pathway
- First CI appointment at 7m old
- Not developmentally ready for VRA testing

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- **June 2019** - Diagnosed with West Syndrome after a period of repeated seizures.
- Further appointments to continue behavioural testing, due to regression/delay in development unable to obtain any reliable VRA results
- Medication prescribed for epilepsy management and also steroids for swelling on brain
- No responses seen to any sounds with/without HAs
- Parents wanted CI for HL but concerned about proceeding due to health issues associated with West Syndrome
- **March 2020** - Assessment put on hold
- Family felt they need to concentrate on getting West Syndrome symptoms stabilised
- Received COVID shielding letter
- Did not feel that hearing was the highest priority.

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- **October 2020** – assessment started again
- Seizures better controlled
- Delayed development – reported development age equivalent to around 6 months
- Still no behavioural tests
- ABR results inconsistent due to interference on traces

- MDT meeting – decision to offer CI as although results of ABR inconclusive, functionally consistent with profound HL
- Offered unilateral CI to minimise shadowing on any future MRI's
- Parents happy with this decision

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- **7th May 2021** - Implant surgery
- Switch on appointments 3 weeks later
- Intra-op measurements used for MAP
- Initially no responses seen
- Used slower rate for MAP to allow time for brain to process

- Current parental reports of improved responses
- Copying some sounds such as “ssss”
- Generally more engaged
- Initially retention issues but this has improved as now wearing an epilepsy helmet

Learning point – despite complex needs, children can still significantly benefit from CI. Do not delay referral.