

Is earlier always possible?

A review of infants diagnosed with Unilateral ANSD

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Permanent unilateral hearing loss in children

- Early identification through newborn hearing screening happens routinely now
- Presents in approximately 15-30% of infants diagnosed through newborn hearing screening (SWISH, 2010)
- Large incidence of temporal bone anomalies in children with unilateral hearing loss (UHL) (Purcell, Shinn, Coggeshall, Phillips, et al., 2017; Lieu, 2018)
- Incidence of cochlear nerve deficiency/aplasia is a significant cause of unilateral Auditory Neuropathy Spectrum Disorder (ANSD) (Lui et al., 2012)

Unilateral ANSD in children

- Diagnosis occurs early
- More hearing device options are available
- Early intervention can be accessed

- **BUT...** ANSD is a complex auditory condition
- Audiological and medical assessment results are critical to informing clinical management and hearing device options

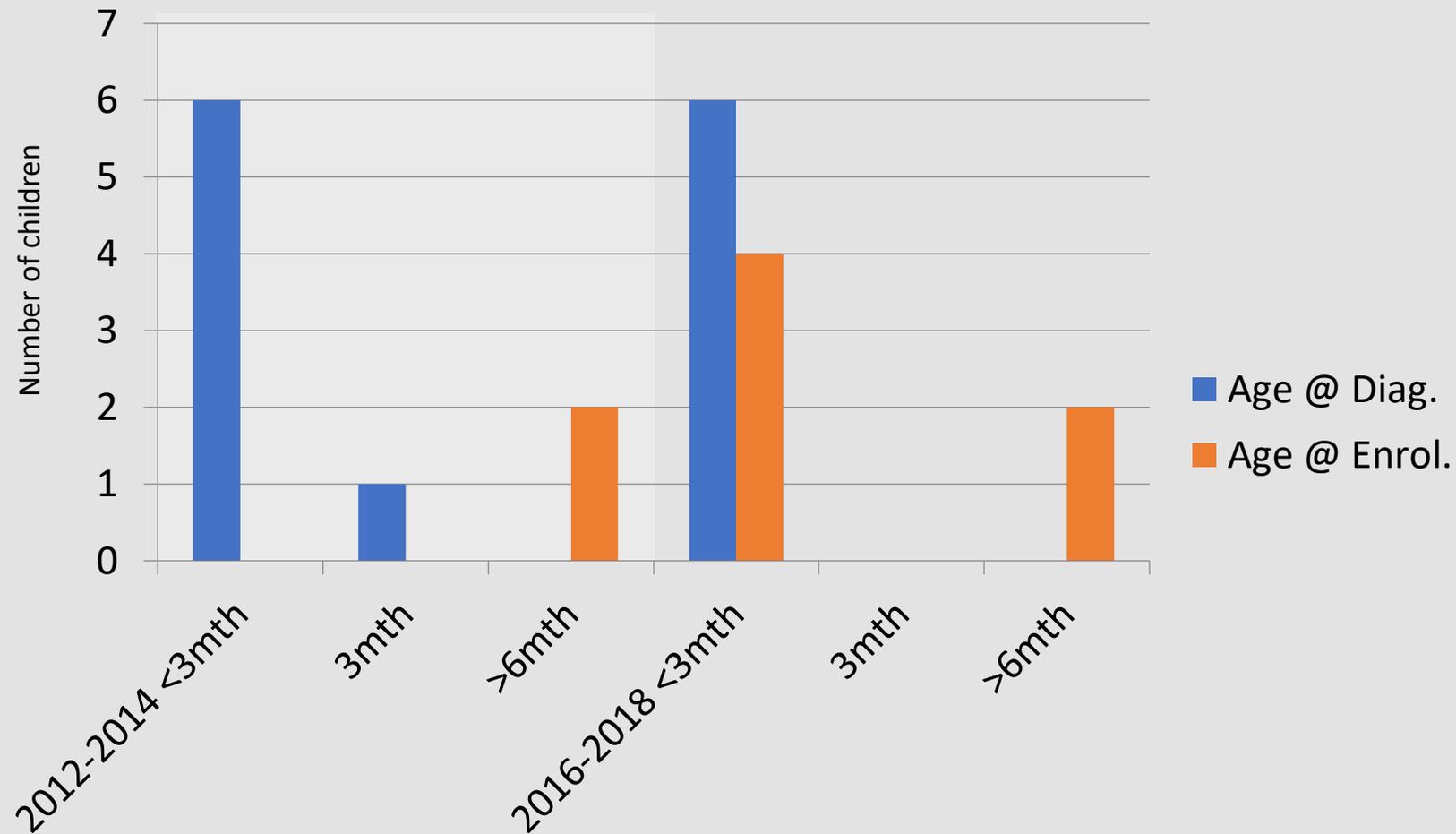
Seeking answers...

- What did past, and now current clinical management pathways look like for children with Unilateral ANSD?
- Is earlier always possible?
- What hearing devices (if any), are fitted in children with Unilateral ANSD?
- What speech and language outcomes are seen in children with Unilateral ANSD?
- What type of early intervention services are children with Unilateral ANSD accessing?

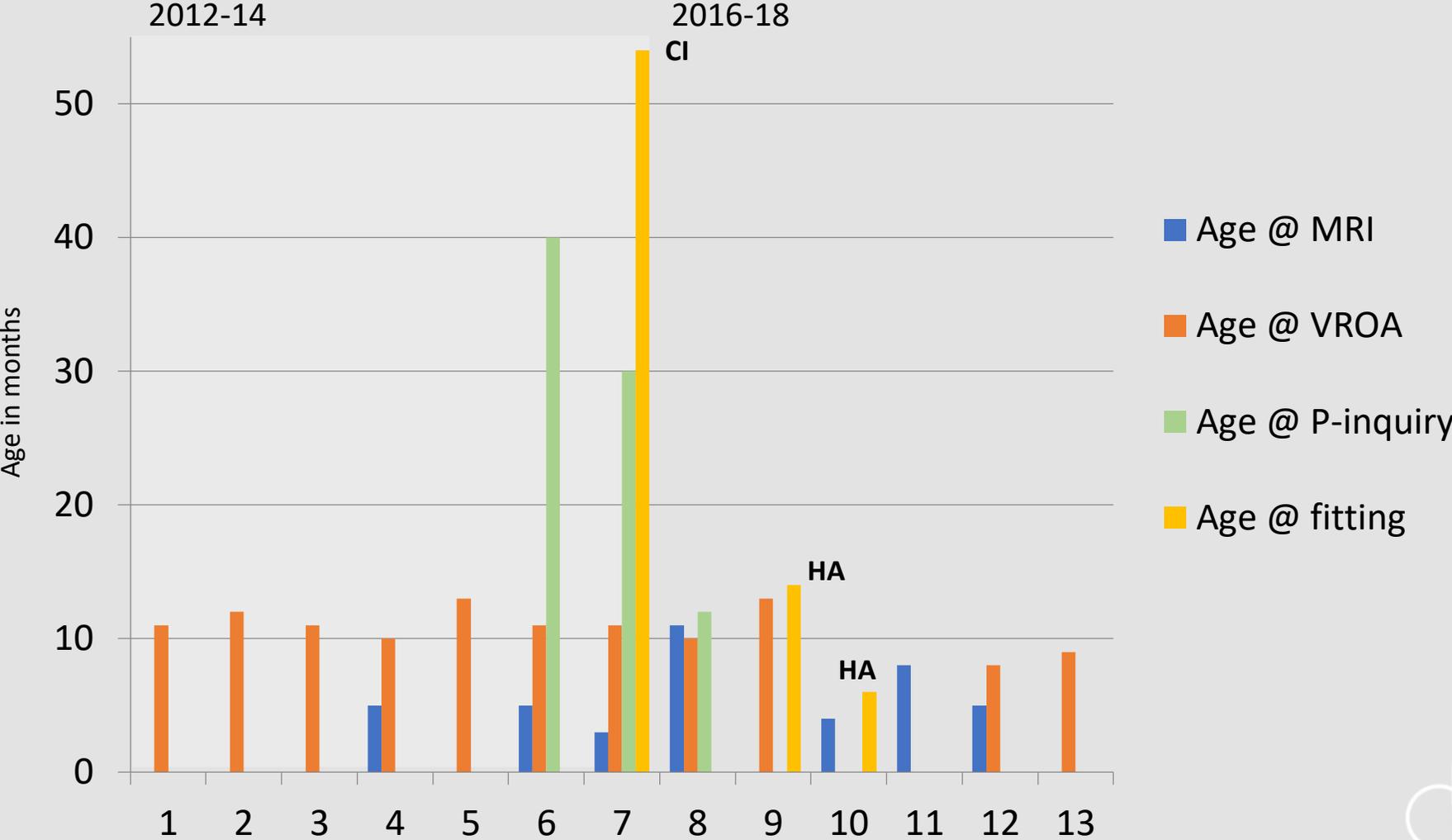
Who?

DOB Cohort	Case No.#	Gender	Degree HL	Ear (L/R)
2012-2014	1	Female	Mild-Mod	L
	2	Male	Sev-Prof	R
	3	Female	Sev-Prof	R
	4	Male	Sev-Prof	L
	5	Female		R
	6	Female		R
	7	Male	Severe	L
2016-2018	8	Male	Mod-Sev	R
	9	Male	Profound	R
	10	Male	Sev-Prof	L
	11	Female	Sev-Prof	R
	12	Male	Profound	R
	13	Male		R

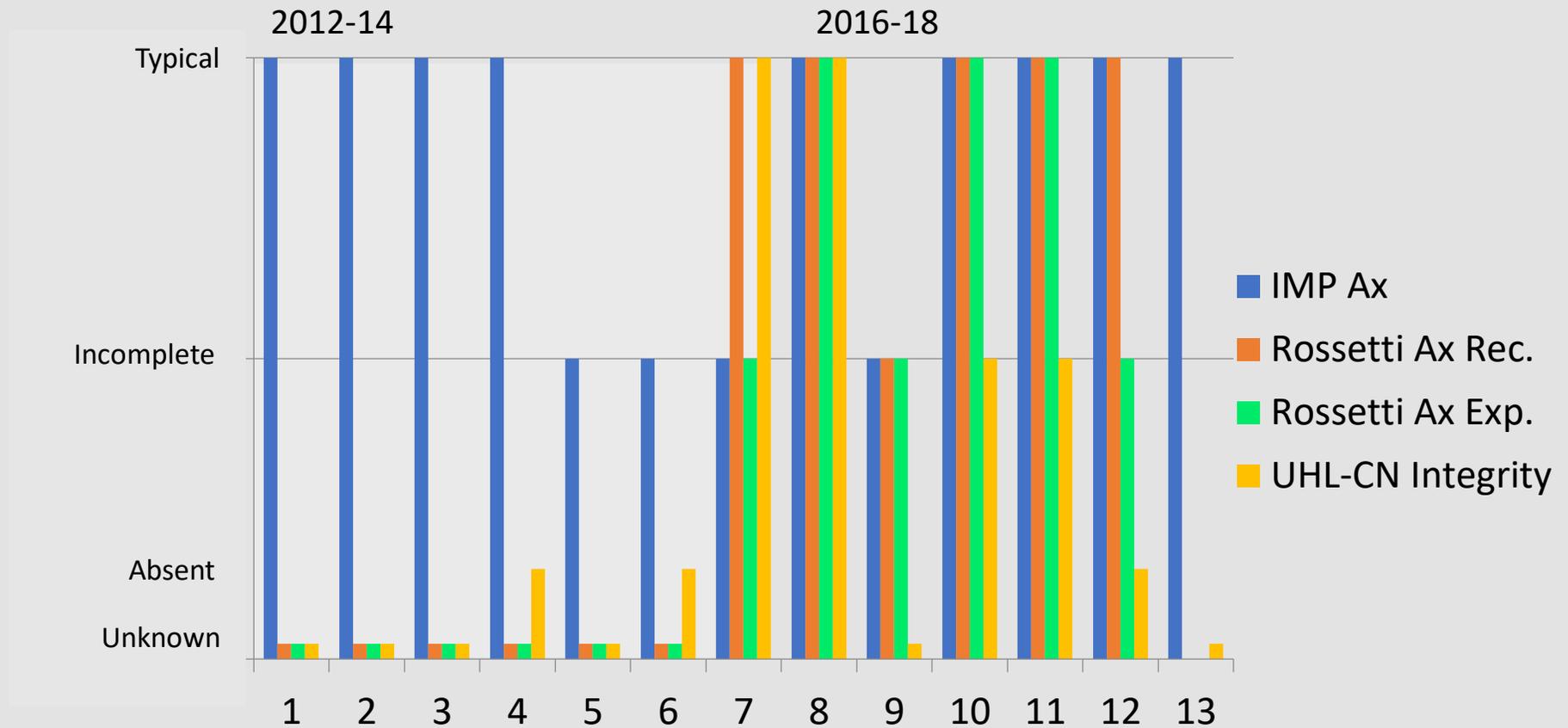
When? Age of diagnosis and enrolment in early intervention



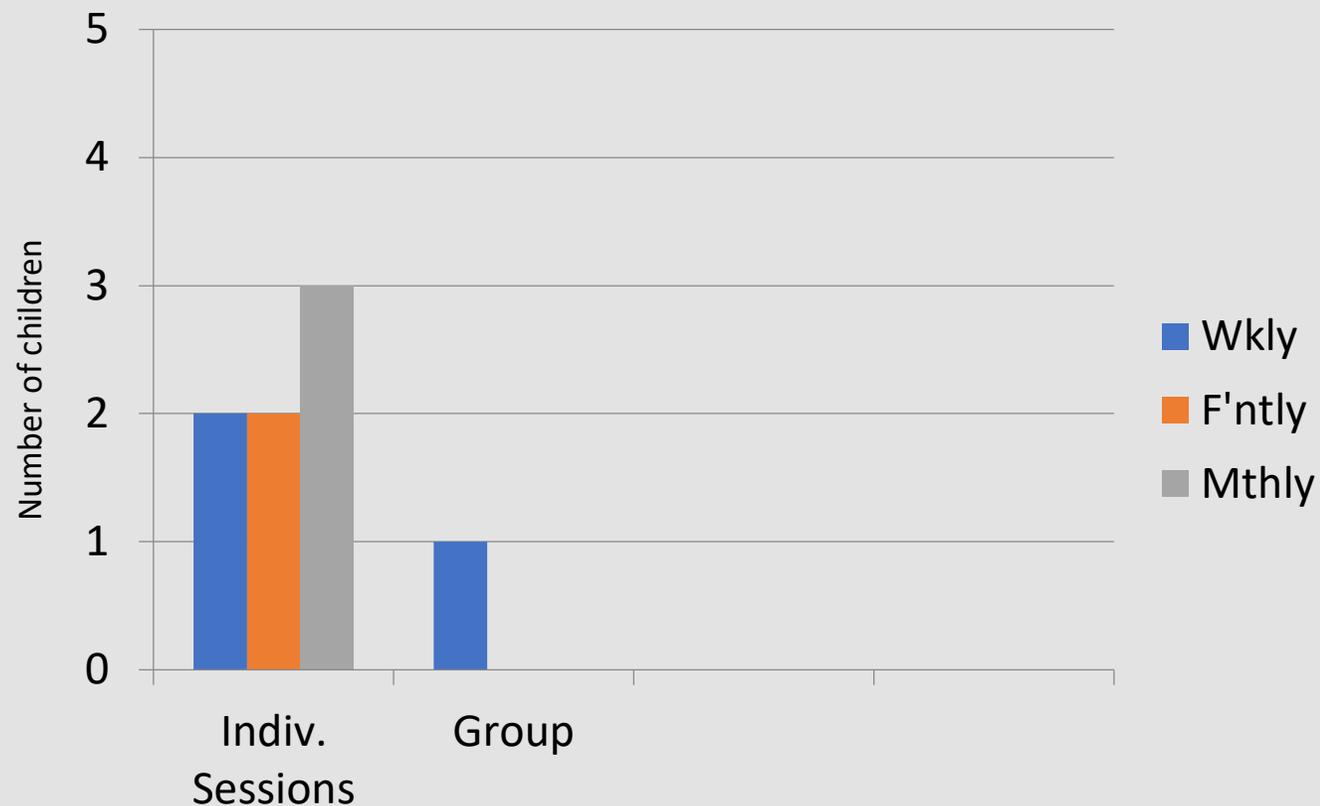
When? Age at MRI – device fitting - VROA



How? Progress: Stage-for-age



What? Early intervention services accessed in 2018



Strengths of current practice

- Diagnosis of unilateral ANSD confirmed before 3 months of age in all cases
- More children enrolled in early intervention shortly after diagnosis
- Speech and language delay can be identified early
- Monitoring speech and language is important as delays may not become apparent until later
- Delivery of individualised, tailored early intervention support based on child and family needs

Challenges with current practice

- Gaps in audiological records, e.g. timing and results of VROA; CAEP; hearing device fitting; ongoing clinical management plan

Earlier is not always possible

- Variability in parental choice to access Australian Hearing Services
- Issue of audiological follow through of children not fitted with a hearing device
- Variability/delay in time frames for MRI
- Variability in timeframes of decision-making about suitable hearing device options
- Variability in parent decisions to proceed with hearing device fitting
- Variability in age at first hearing device fitting
- Variability in families choosing to access early intervention services

Challenges with current practice

One size does not fit all

- Individual variability in speech and language outcomes
- Individual variability in early intervention needs- type and frequency of services

Questions remain...

- What are parents' experiences of the current management pathways for children with unilateral ANSD?
- How can we best support families who learn early about unilateral ANSD?
- What hearing devices (if any), should be recommended for children with unilateral cochlea nerve aplasia or hypoplasia? (Lieu, 2018)
- If a cochlear implant is recommended, when is it too late? (Lieu, 2018)

Questions remain...

- Who monitors the hearing in the *normal* hearing ear?
- What early intervention services should we provide to children with Unilateral Hearing Loss (UHL)? (Fitzpatrick, Grandpierre, Durieux-Smith, Gaboury, Coyle, Na & Sallam, 2016)
- Which predictors will predict risk for communication and/or other developmental delay in children with unilateral hearing loss? (Lieu, 2018)
- Is early intervention effective for children with UHL (including children with ANSD)? (Lieu, 2018)

Enhancing **audiological** support and future management pathways

- Consistent audiological management practices
- Development and sharing of follow-up and management protocols
- Documenting and reporting on clinical management
- Increased sharing of information between internal and external stakeholders

Enhancing **early intervention** support and **future management pathways**

- Development of speech and language assessment protocols that are sensitive to identify delay early
- Consistent collection and documenting of client outcomes
- Development of evidence-based individual and group programs to address current and future needs
- Provision of flexible early intervention programs to address individual needs
- Increase parent-to-parent support
- Liaise regularly with audiological service provider to stay up to date with clinical management plan; contribute to management plan, and ensure monitoring of hearing (including normal hearing ear)

Thank you for listening

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References

- Fitzpatrick, E., Grandpierre, V., Durieux-Smith, A., Gaboury, I., Coyle, D., Na, E., & Sallam, N. (2016). Children with mild bilateral and unilateral hearing loss: parents' reflections on experiences and outcomes. *Journal of Deaf Studies and Deaf Education*, 34-43.doi:10.1093/deafed/env047
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