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Audio-verbal outcomes following cochlear implantation in USH1C patients

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Introduction

BACKGROUND: Usher syndrome (Usher) is the most common genetic cause of concurrent deafness and blindness. Currently, 3 clinical types and 10 genes are associated with the disease. Approximately 2.5% of Usher is caused by mutations in the USH1C gene. At least 50 mutations in the USH1C gene have been reported to cause USH1C, however the c.216G>A mutation accounts for nearly all Usher 1 cases in Acadian patients of Louisiana, USA, and Canada. Progress over the past two decades in access to genetic testing, early diagnosis, and cochlear implantation has significantly improved clinical management for patients with hearing loss, including Usher. Today, USH1C patients typically receive cochlear implants with follow-up care from otolaryngologists, audiologists, and speech language pathologists, and use oral communication. However, long-term audio-verbal outcomes following implantation in USH1C patients are not known.

OBJECTIVES: The objective of this study is to determine cochlear implant success in USH1C patients via retrospective chart review.

Demographics

Total participants enrolled	109
# of participants with cochlear implants	28/109 (25.7%)
Sex	
Male	13/28 (46%)
Diagnosis	USH1C (27/28)
	USH2A (1/28)
Genetic confirmation	100% (28/28)
Number of participants with meaningful retrospective data	10/28 (35.7%)
Average age of implantation (years)	1.1 (n=9, 1 outlier at 4 years)
Lateralization	10/28
R	1
L	0
B	9

Fig 1. Natural history study demographics including USH1C patients with cochlear implants.

Timeline & List of Follow-up Procedures

Subject ID	Age & Device at Implantation	Post-Implant Follow-up Procedures
896-01	• 1 year old • Cochlear™ Nucleus® CP810 Sound Processor	6 months: • L6D • SDT 8 months: • L6D • SDT 10 months: • PTA • SRT • WD
896-03	• 1 year old • **R** - Cochlear™ Nucleus® CI422 cochlear implant with straight electrode • **L** - Cochlear™ Nucleus® CI512 cochlear implant with Contour Advance® electrode	2 months: • PTA • SDT • L6D 4 months: • SAT 7 months: • L6D • SDT 11 months: • L6D • SDT 13 months: • L6D 20 months: • PTA • L6D • WD • SRT 27 months: • PTA • SRT • SDT • WD 30 months: • L6D • SRT • WD 34 Months: • PTA • SRT • WD 39 months: • PTA • SRT • WD
896-13	• 2 years old • **B** -Advanced Bionics® HiRes 90K® with Mid-Scala electrode	1 month: • L6D • SDT 4 months: • PTA • L6D • SDT 9 months: • L6D • PTA 19 months: • PTA • SDT • L6D
896-14	• 2 years old • **R** - Cochlear™ Nucleus® CP910 Sound Processor	38 months: • PTA • L6D • SRT 50 months: • PTA • SRT 54 Months: • PTA • SRT • WD 111 months: • PTA • SRT • WD 112 months: • PTA • SRT
896-15	• 2 years old • **R** - Cochlear™ Nucleus® CI422 cochlear implant with straight electrode • **L** - Cochlear™ Nucleus® 24 cochlear implant CI24M	12.6 years: • PTA • L6D • SRT 14 years: • PTA • DPOAES • Tymp 14.7 years: • PTA • SRT • SDT • WD • L6D 18 years: • PTA • SRT • SDT • WD 19.3 years: • PTA • SRT • DPOAES • Tymp
896-17	• 1 year old • **B** - Cochlear™ Nucleus® CI24RE Contour Advance ™ with N7 processor	14 months: • SRT • PTA 26 months: • PTA • SRT 38 months: • PTA • SRT 48 months: • PTA • SRT 81 months: • PTA • SRT • HINT 104 months: • PTA • SRT • HINT • WD
896-18	• 4 years old with re-implantation at 14 years old • **R** - Cochlear™ Nucleus® CI24RE Contour Advance ™ with N7 processor • **L** -Cochlear™ Nucleus® CI24R Contour Advance ™ with N7 processor	19 months: • SRT • PTA • WD 30 months: • PTA • SRT • WD 44 months: • PTA • SRT • WD 45 months: • PTA • SDT • Tymp 58 months: • PTA • SRT • WD 113 months: • PTA • SRT • WD
896-21	• 1 year old • **R** - Cochlear™ Nucleus® CI512 cochlear implant with straight electrode with N6 processor	7 months: • PTA • SRT 12 months: • PTA • SRT • L6D • Speech therapy report 19 months: • PTA • SRT 32 months: • Speech therapy report 43 months: • PTA • SRT • WD 49 months: • PTA • SRT • WD
896-42	• 1 year old • **B** - Cochlear™ Nucleus® CI512 cochlear implant with straight electrode with N7 processor	6 years: • PTA • SRT • WD 8.7 years: • PTA • SRT • WD 10 years: • PTA • SRT • WD 11.5 years: • PTA • SRT • WD
896-89	• 0 years old • **B** - Cochlear™ Nucleus® CI512 cochlear implant with straight electrode with N7 processor	3 months: • PTA • SAT 6 months: • PTA • SAT 9 months: • PTA • SAT 11 months: • PTA • SAT

Key	
PTA = Pure Tone Audiogram	DPOAES = Distortion Product Otoacoustic Emissions
SDT = Speech Detection Threshold	HINT = Hearing in Noise Test
SRT = Speech Reception Threshold	L6D = Ling 6 Detection
WD = Word Discrimination	Tymp = Tympanogram
SAT = Speech Awareness Threshold	

Fig 2. Post-implantation audio-verbal records. Age of implantation is indicated along with follow-up audio-verbal records at specified interval.

Methods

Demographics, genetic diagnosis, cochlear implant device, and post-implantation hearing and audio-verbal therapy will be evaluated by retrospective chart review. Recruitment of patients will occur through multiple means, including deaf-blind support groups, physicians, and Usher syndrome symposiums throughout Louisiana. We defined patients as having meaningful data if we were able to collect the DOB, gender, genetic confirmation, DOS, device information, and audio-verbal therapy records.

Results

- **109 participants are currently enrolled in our retrospective natural history of Usher syndrome in Louisiana study.**
- **Of these, 28/109 (25.7%) use cochlear implants and have genetic confirmation of USH1C disease.**
- **10/28 (35.7%) participants range in age from 1 – 24 years and have meaningful retrospective data, including cochlear implant device manufacturer and electrode type, age at implantation, pure tone audiograms, speech awareness and reception thresholds, word discrimination tests, and speech therapy metrics.**
- **Retrospective data collected shows the variability in type and timing of follow-up care.**
- **Collection of the remaining charts and preliminary assessments of longitudinal hearing and audio-verbal measures are underway.**

Preliminary Conclusions

- **Currently, there is no standard of care regarding cochlear implantation follow-up care, which creates a challenge to evaluate success.**
- **A careful understanding of audio-verbal outcomes, implant device, and USH1C genotype will improve clinical knowledge and care for patients with Usher syndrome.**

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