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To determine the diagnostic utility of various tests and treatment option for pediatric patients presenting with a unilateral sensorineural hearing loss (SNHL)

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Aim: To determine the diagnostic utility of various tests and treatment option for pediatric patients presenting with a unilateral sensorineural hearing loss (SNHL).

Study Design: Retrospective chart review, uncontrolled case series.

Methods: Study was conducted at tertiary academic center; pediatric hospital. Pediatric patients (<18 years old) were evaluated for unilateral SNHL. Upon IRB approval patients with single sided SNHL were identified using an internal database. Audiologic data, genetic testing, imaging results and pertinent clinical information were analyzed to determine the most efficacious method of evaluating these patients.

Results: 407 patients were identified with average age (SD) of 87.4 months (53.9). 233 patients were males (53%). 37 patients had high frequency (HF) SNHL alone, 51 patients with normal HF PTA (abnormal low frequency PTA) and 319 with a flat audiometric configuration. Hearing loss was more common on the left side and in males. More than 50% of patients had mild to moderate SNHL. GJB2 was a rarely seen (0.7%), while mutations in the *SLC26A4* gene were seen in 18% of patients. Enlarged vestibular aqueduct (EVA) was found in 17% of patients. In patients with unilateral EVA, 19% had contralateral ear involvement. Overall, 19% of patients developed bilateral hearing loss. Treatments varied from observation, traditional hearing aids, CROS hearing aids and cochlear implantation. Seven patients with single sided deafness were implanted. All patients continue wearing their implants and getting appropriate aided benefit with speech reception thresholds at 30dB or better with a minimum of 2 years of follow up.

Conclusion: USNHL is a relatively common problem affecting children. In our population, progression occurred relatively frequently. Temporal bone anomalies such as EVA were common. Cochlear nerve deficiencies are also demonstrated in our study and are best demonstrated on MRI. Pendred syndrome and DFNB1 are rare. Based on our data, an algorithm for children with USNHL shows that imaging should be the primary diagnostic study. Magnetic resonance imaging may be the best imaging modality due to its ability to diagnose deficient cochlear nerves in addition to common temporal bone anomalies. Treatment should be individualized based on the medical needs of the child. Cochlear implantation is now an option for patients with single sided deafness.

Biography

John H Greinwald is a tenured Professor of Otolaryngology and Pediatrics with over 20 years of experience with a focus on the genetic causes and treatment of deafness. He co-founded the Ear and Hearing Center at Cincinnati Children's Hospital Medical Center. He has pioneered the establishment of diagnostic evaluation algorithms for children with sensorineural hearing loss and developed a next generation sequencing platform to determine the genetic causes of hearing loss in children. He has 81 peer review articles published with the majority related to hearing loss. His research interests concentrate on identifying novel causes of genetic diseases, mitigating barriers to genetic counseling in underserved populations, developing innovative methods of providing complex genetic information to patients and physicians and helped pioneer minimal access cochlear implant surgery. Clinically, he is the Medical Director of the Cochlear Implant Team and faculty in the Auditory Genetics Laboratory of the Ear and Hearing Center. He received his BS degree from Wofford College and MD degree from the Medical University of South Carolina. His Otolaryngology training was at the Naval Medical Center Portsmouth Virginia and his pediatric otolaryngology fellowship at University of Iowa. He is Board Certified in Otolaryngology Head and Neck Surgery.

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