WORLD HEARING CENTER
OF THE INSTITUTE OF PHYSIOLOGY AND PATHOLOGY OF HEARING
KAJETANY / WARSAW

18TH INTERNATIONAL CONFERENCE
ON COCHLEAR IMPLANTS AND OTHER IMPLANTABLE TECHNOLOGIES
10-13 MAY 2026, WARSAW

Evaluation of auditory outcomes after Cochlear Implantation in children with CHARGE syndrome

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INTRODUCTION

CHARGE syndrome consists of a complex cluster of congenital abnormalities. CHARGE stands for Coloboma, Heart defects, Atresia of the choanae, Retardation of growth and development, Genital hypoplasia and Ear abnormalities. In 1998 Blake refined the clinical diagnostic criteria by grouping features into major and minor. The major criteria are coloboma, choanal atresia, characteristic ear abnormalities, and cranial nerve abnormalities. Minor criteria are Genital hypoplasia, developmental delay, cardiovascular malformations, orofacial clefts, trachea-oesophagel fistulae and a distinctive face. The presence of these clinical features is variable. Ear abnormalities and hearing loss are common in children with CHARGE and both conductive hearing loss (due to glue ear, ossicular abnormalities or ossicular fixation) and sensorineural hearing loss (due to inner ear abnormalities) may occur A small number of these children have profound hearing loss and are considered for cochlear implantation. CHARGE is also associated with cranial nerve abnormalities , particularly involving the olfactory, vestibular and facial nerves, but including the cochlea nerve. The aim of our study was to evaluate the audiometric outcomes of children with hearing loss.

MATERIAL AND METHODS

The study group consisted 8 patients aged 0-13 years-old (M= 4,) operated on between 2013 and 2021. n. All patients had an auditory brainstem potentials (ABR) test before surgery. Postoperatively, the patients had two tests: Adaptive Auditory Speech Test (AAST) and free-field threshold audiometry. All were using hearing aids prior to implantation and had computed tomography performed before operation. The study group consisted of 8 children—5 boys and 3 girls. All children had additional comorbidities: heart defects, visual defects, delayed motor development or gastrointestinal problems.

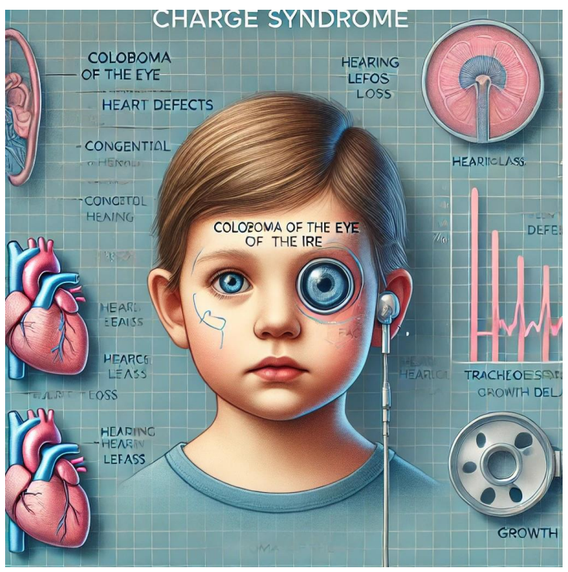

Fig 1. CHARGE accompanying symptomes. Created by chatGPT

RESULTS

In patients, auditory brainstem response (ABR) testing was performed preoperatively to determine their hearing thresholds. All children suffered from profound, bilateral hearing loss. Postoperatively, free-field audiometric testing was conducted. Depending on the child's abilities and level of cooperation, the test was performed using appropriate methods: independent work, VRA (Visual Reinforcement Audiometry), BOA (Behavioral Observation Audiometry), or play audiometry.Before surgery hearing thresholds in each patient, across 0.5-4 kHz frequencies, were > 90 dB. All patients had anatomical abnormalities of the middle and inner ears. Mean hearing threshold in free-field audiometry test (after operation) was 49 dB. Average AAST results were: (1) in quiet: 43 ; (2) in noise: - 6.125.

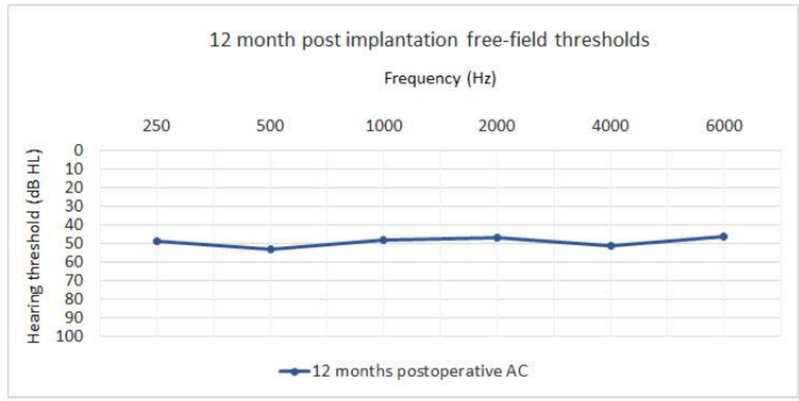

Fig. 2. 12months post-operative free-field pure-tone audiometry thresholds.

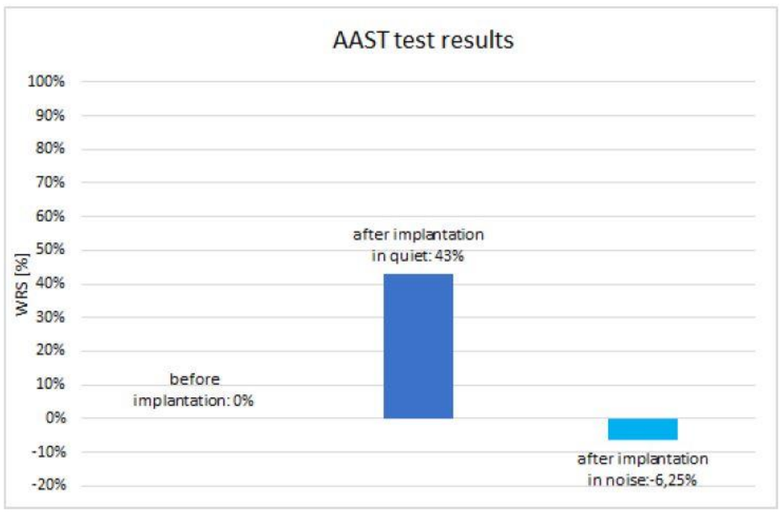

Fig. 3. 12 months post-operative AAST average test results.

CONCLUSION

Cochlear implantation in children with CHARGE syndrome presents unique challenges due to the complex nature of the condition. However, the results demonstrate that it can significantly improve auditory outcomes, fostering better communication abilities and quality of life. Early intervention and a multidisciplinary approach are critical for achieving optimal results.

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