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Deafness Management in Children with the Charge Association

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Case Report

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Abstract: The acronym CHARGE is used to describe specific congenital birth defects in children: Colobomata, Heart defect, Atresia of the choanae, Retarded growth or development, Genital hypoplasia, and Ear anomalies or deafness. To confirm a diagnosis of CHARGE association, the presence of at least four of these six abnormalities is sufficient.
Keywords: Charge-Inner ear malformation-Otology-hearing aid.

INTRODUCTION

Initially described by Hall in 1979 and identified by the acronym CHARGE by Pagon in 1981. Some of these children may have additional defects such as facial palsy and cleft lip and/or cleft palate. Abnormal vestibular structures that contribute to balancing difficulties are also found in some children with CHARGE association.

The incidence of CHARGE association varies between 0.54 and 1.13 per 10,000, with no difference between genders or races.

The etiology of CHARGE association is still not known, although it is typically sporadic. Autosomal dominant and, less commonly, autosomal recessive transmission have also been inferred from multigenerational family studies. There is a low recurrence risk in subsequent children for normal parents with a CHARGE child.

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.

CASE REPORT

We report the case of a male child of 3 years old. This full-term, clinically male child was born after an uneventful pregnancy

However, the child has been surgered for a cardiac malformation and also for cryptorchidism.

He has consulted in our service for language development disorders. The child's clinical examination has revealed the presence of frontal wrinkles and malformation of external ears (fig-1A).

The evoqued auditory potential has revealed a bilateral deafness (70 dB right and 50 dB left)

The radiological assessment (TDM and IRM) has objectified Cochlea-vestibular malformations (fig-1(B, C, D)).

After these clinical signs and paraclinical and the antecedents of the children, the diagnosis of the charge syndrome was confirmed.

Our patient has benefited from a bilateral hearing aid with orthophontic sessions regularly, which allowed the child to develop his language as well as correct schooling. Currently the child is in primary class without scholar difficulty.

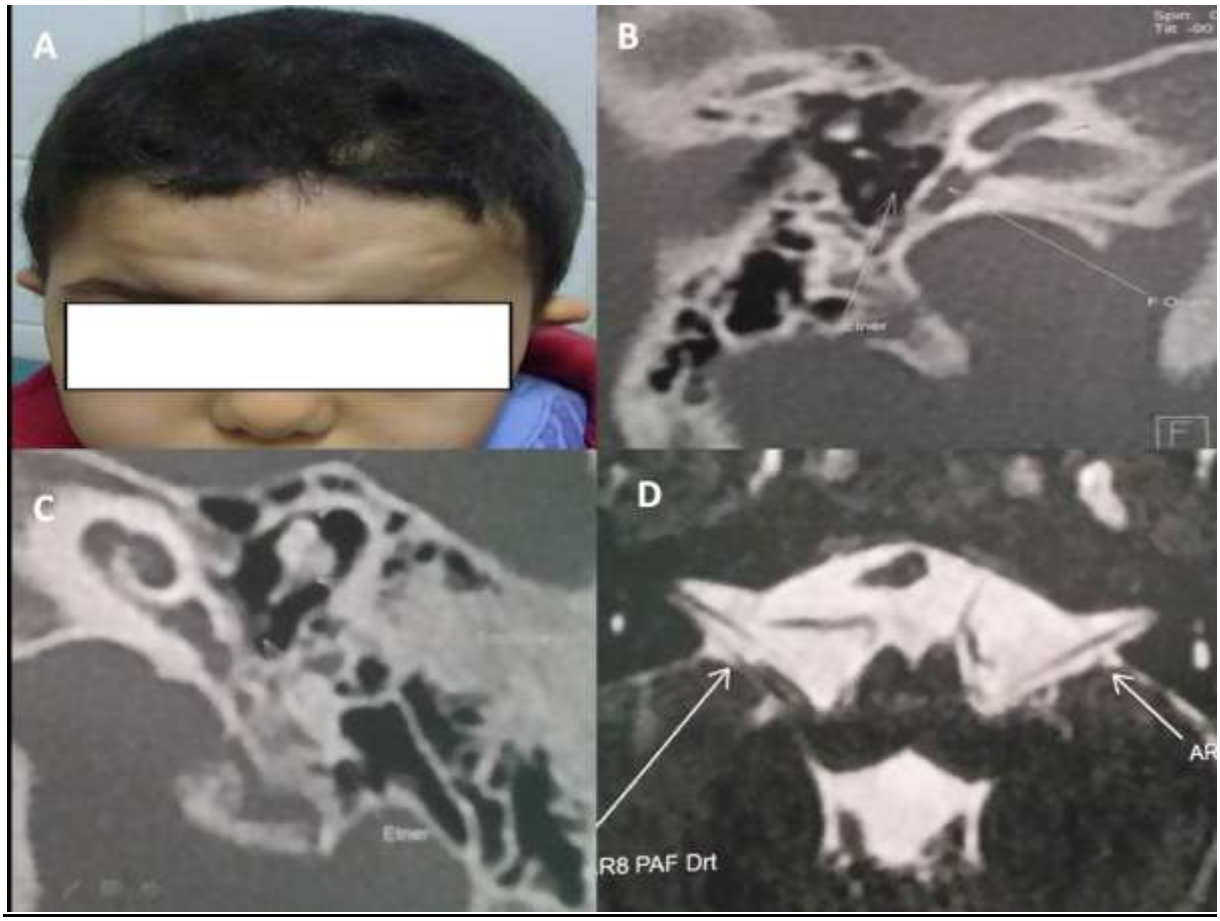

Fig-1: (A) The frontal wrinkles, (B) CT scan the semicircular canals are absent (Right ear), (C) CT scan the semicircular canals are absent (Left ear), (D) MRI the semicircular canals are absent. The facial and acoustic nerves are presents

DISCUSSION

Patients with the CHARGE association as first defined by Pagon are affected by ocular, cardiac, airway, central nervous system and developmental, genital and otologic anomalies. Facial nerve and renal anomalies are considered common adjuncts to this spectrum. The otorhinolaryngologist is often a critical participant in the successful management of these medically complicated children [1].

In our case, the CHARGE association was etablishled by the presence of major criteria (cardiac malformations, external and internal ear malformation, cryptorchidism) and minor criteria (characteristic face) [1].

Since the bilateral deafness was identified (70dB left 50dB right), a bilateral hearing aid was established associated with a regularly orthophonic sessions.

Cochlear implantation in patients with CHARGE syndrome represents one of the most technically challenging implantation scenarios. There is a higher rate of surgical complications including facial nerve injury secondary to the dysmorphic temporal bone anatomy seen in CHARGE syndrome [2].

The benefits of cochlear implantation in children with multiple congenital defects are discussed [3].

Auricular anomalies made hearing aid fit difficult but not impossible. The importance of early identification is emphasized by the long-term viability of these medically complex children [4].

Audiologists who followed hearing aid use and educational status reported complex issues requiring close attention and coordination by the otorhinolaryngologist and the audiologist [5].

Management of these children requires early evaluation by the Otorhinolaryngologist, Audiologist, Speech therapist, and the Developmental Pediatrics and Genetics Services.

Close otolaryngologic follow-up is essential for airway and swallowing issues, as well as for otologic care [4].

Early identification of hearing deficits does not necessarily translate into early aiding: life-threatening airway, cardiac, nutritional and neurologic disease often takes precedence over hearing and speech optimization [4].

CONCLUSION

CHARGE syndrome consists of a complex cluster of congenital abnormalities, Including external and internal ear malformations associated to deafness. The audiological management of children with charge syndrome requires early management in order to have a better outcome, for this collaboration between pediatrician and otorhinolaryngologists are necessary.

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