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A Rare Case of Bilateral Duane Syndrome Type I with Inner Ear Developmental Arrest

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Abstract

Background: Duane Syndrome is a congenital cranial dysinnervation disorder that primarily restricts eye movements. The majority of patients have isolated Duane syndrome with no other congenital abnormalities, however in certain cases, there are non-ocular symptoms which do not fit aspecific syndrome. In this case we describe bilateral Duane syndrome type 1(limited eye abduction) with findings of developmental arrest in the inner ear and cochlear aplasia which to our knowledge has not been reported yet.

Case presentation: This case reports a 3 year old boy who presented with watery eyes during feeding and speech delay. He had a right sided sensorineural hearing loss, with ABR being inconclusive and ASSR showed severe hearing loss. CT mastoids and MRI were performed and a diagnosis of cochlear aplasia with inner ear developmental arrest was made. The patient showed good response and improvement in speech with a soft band bone anchored hearing aid.

Conclusion: The early recognition and diagnosis of other organ anomalies associated with Duane's syndrome is of utmost importance for the prognosis of the patient. Radiological imaging, most importantly MRI is the gold-standard for identification of inner ear abnormalities.

Keywords: Inner ear • Congenital abnormalities • Duane syndrome • Cochlear aplasia

Abbreviations: ABR: Auditory Brainstem Response • ASSR: Auditory Steady State Response • BAHA: Bone Anchored Hearing Aid

Introduction

Duane syndrome is an inherited autosomal dominant strabismus condition which is clinically seen as congenital, non-progressive limited horizontal eye movement together with global retraction which causes the palpebral fissures to narrow. Three different types of Duane Syndrome have been described in the literature and classified by Huber; type 1; (limited abduction with or without esotropia), type 2; (limited adduction, with or without exotropia) and type 3; (limitation of both abduction and adduction and any form of horizontal strabismus) [1]. More than 70% of individuals with Duane's syndrome do not have other congenital anomalies, however in the rest there have been reports of deafness, skeletal abnormalities, other ocular anomalies and cranial nerve palsies.Interestingly, one case has been reported in 1984 of a patient with bilateral Duane syndrome and crocodile tears associated with external, middle and inner ear abnormalities. In this case, an exploratory tympanotomy showed a rudimental stapes superstructureconnected to the abbreviated incudal lenticular process with connective tissue and absence of the oval window [2].

Case Presentation

A 3 year old boy who had an uneventful perinatal period and birth was referred to our audiology department in view of delay in attaining specific milestones including speech delay. His mother noticed that he had a long-standing watery left eye during feeding, especially on chewing solid foods. An ophthalmic review revealed that both palpebral apertures narrowed on adduction and widened on attempted abduction. It was also noted that the palpebral apertures straighten on adopting a 'chin-up' posture. This confirmed bilateral Duane's syndrome type 1 along with 'crocodile tears'. An MRI of the brain and orbits showed normal examination. The patient was fitted with glasses and did well, thus surgery was not needed.

An echocardiogram showed a small apical atrial septal defect, a small patent ductus arteriosus and a patent foramen ovale. Since the patient was asymptomatic cardiology follow-up was organised without the need for further interventions.

Visual reinforcement audiometry revealed bilateral borderline hearing thresholds. Left tympanometry was noted to be type B and grommet was inserted. Repeat audiometry post-operatively still showed moderate loss so an auditory brainstem response (ABR) was performed.

ABR was inconclusive, an auditory steady state response (ASSR) was performed, and a response was detected at 90dB at 2kHz and 100 dB at 4 kHz on the right, whilst the left showed a response of 25dB at 2kHz and 4kHz and 40dB at 1 kHz. This test was done using masking difference of 25dB. A CT mastoids and MRI of the inner acoustic meatus showed an absent right cochlea. There was also no separation between the vestibule and the lateral and posterior semicircular canals (Figure 1-3). The superior semicircular canal was noted to be dilated and the vestibular aqueduct was not visible. These findings represent developmental arrest of the inner ear.

The patient showed good response with the soft band bone anchored hearing aid and during the last otology clinic follow-up, a bone anchored hearing aid (BAHA) insertion was discussed with the parents, however since the boy is doing well a watch and wait approach was taken.

Results and Discussion

Duane's syndrome was first described by Jakob Stilling (1887) and SiegmundTürk (1896) and subsequently named after Alexander Duane who discussed the disorder in more detail in 1905 [3]. It is a rare, congenital disease, where commonly, the abducent nucleus and nerve are absent or

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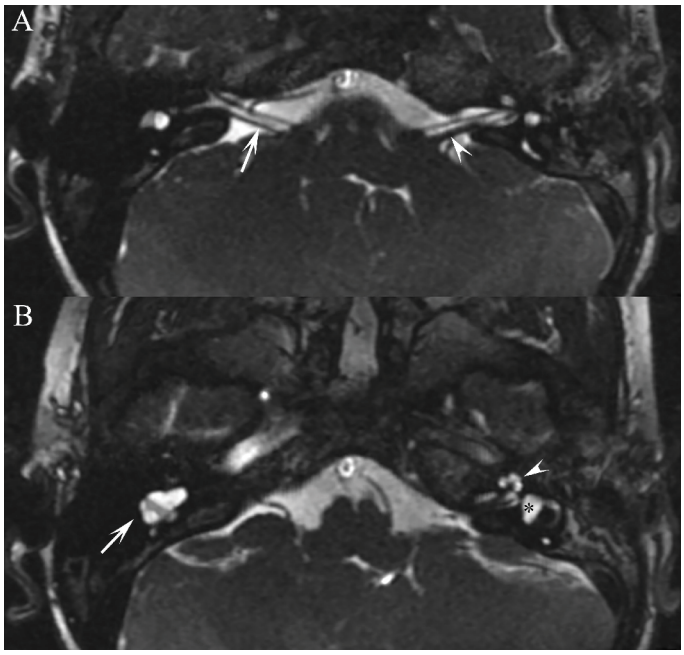

Figure 1. High resolution T2 weighted MRI (FIESTA) of the inner ears. A. The right vestibulocochlear nerve (arrow) is hypoplastic when compared to its left counterpart (arrowhead). B. The right cochlea is absent and the vestibule is inseparable from the horizontal semicircular canal (arrow). Normal left cochlea (arrowhead) and vestibule (black asterix).

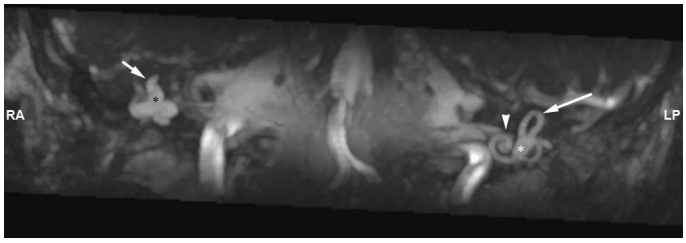

Figure 2. Volume rendered coronal reconstruction from a heavily weighted T2 sequence (FIESTA). The left superior semicircular canal is dilated and malformed (short arrow). There are no cleavage planes separating the vestibule from the semicircular canals (black asterix). Normal left cochlea (arrowhead), vestibule (white asterix) and superior semicircular canal (longarrow).

hypoplastic and the lateral rectus is innervated by the oculomotor nerve [4]. This results in opposing muscles being innervated by the same nerve, so on attempted abduction, both the lateral rectus and the medical rectus will be stimulated resulting in co-contraction of these muscles, limiting the amount of movement achievable and eye retraction in the socket. Patients are usually diagnosed within the 1st decade of life.The incidence is higher in girls (60% of cases) and on the left eye [5]. In our case, the patient could compensate for the limited eye movement with face turning and, so no surgery was suggested at present. With regards to the lacrimation during eating, it was explained to the parents that it is not a true nasolacrimal duct obstruction and did not need surgical management.

Cochlear aplasia is characterised by complete absence of the cochlea, together with absence of the cochlear nerve canal and the cochlear nerve. This happens during the 3rd week of pregnancy and accounts for only 3% of cochlear malformations making it a rare occurrence [6]. Two subtypes are identified in the literature; cochlear aplasia with and without dilated vestibule [7]. Radiological features include complete cochlear absence and dense otic bone at its site, absence of cochlear nerve and its canal and a hypoplastic internal auditory canal. Hearing loss in unilateral cochlear aplasia can be aided with implantation of a BAHA. If the patient decides against it, the only other option is a soft-band bone conduction aid, as was being used in this case and advising the patient and relatives to take care of the only good hearing ear. Limitations of bone conduction aid include difficulty in localisation of sound and discrimination of speech in noisy backgrounds.

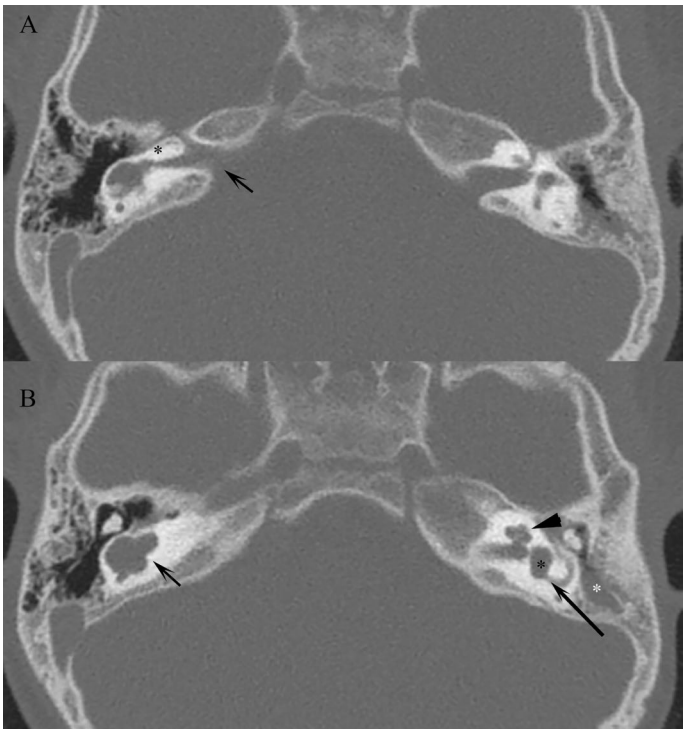

Figure 3. High-resolution CT scan of the temporal bones. A. The right internal auditory canal (arrow) is slightly narrower than the left one and the right cochlea is absent (asterix). B. There is a globular vestibular-horizontal semicircular canal anomaly on the right (arrow). Normal cochlea (arrowhead), vestibule (black asterix) and lateral semicircular canal (long arrow) on the left. Soft tissue fills the epitympanum and mastoid air cells on the left (white asterix).

Conclusion

In young patients presenting with ophthalmic features of Duane's syndrome, evaluation with CT and MRI are necessary to exclude any abnormalities in the inner ear. An early diagnosis limits speech delays and allows for proper audiological assessment and management.

Ethics Approval and Consent to Participate

N/A.

Consent for Publication

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Competing Interests

The authors declare that they have no competing interests.

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Nil.

Author Contributions

CV analysed the available history and diagnostic information and drafted the article. JC critically revised the article. EV provided the medical images and interpretation of the pathological abnormalities, together with their diagnostic importance. All authors approve the final version of the manuscript for publication.

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