

Original article:

Role of high resolution magnetic resonance imaging in Duane's retraction syndrome

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Abstract

Introduction: Duane's retraction syndrome is one of the causes of loss of abduction, proper evaluation of this condition is important so as to rule out other causes before starting treatment. In this study we discuss the importance of high resolution magnetic resonance imaging in evaluating and diagnosing Duane's retraction syndrome.

Materials & Methods: Thin section gradient echo imaging was performed at level of brainstem on a 1.5 Tesla magnetic resonance imaging machine in seven paediatric patients of clinically diagnosed Duane's retraction syndrome. The same test was performed on 10 control subjects. Analysis of imaging findings was done focussed on presence or absence of abducens nerve.

Results: The abducens nerve on the affected side was absent in eight of eight affected eyes in seven patients. The right and left abducens nerve were well identified in all 10 control subjects.

Conclusion: High resolution magnetic resonance imaging is a useful robust tool for demonstration of absent abducens nerve in Duane's retraction syndrome. It is also useful for differential diagnosis and conformation of Duane's retraction syndrome.

Key words: Duane's retraction syndrome, Magnetic resonance imaging, abducens nerve

Introduction:

Duane's retraction syndrome is characterized by congenital abduction deficit, narrowing of palpebral fissure on adduction and globe retraction with up shoot or down shoot in adduction¹. Early electrophysiological studies suggested that Duane's retraction syndrome is a neurogenic disorder involving either a supranuclear lesion or a cranial nerve anomaly in which lateral rectus muscle is not innervated by abducens nerve; instead it is innervated by branch of oculomotor nerve². Early autopsy studies showed that abducens nuclei and nerves were absent from brainstem, and the lateral

rectus was partially innervated by inferior division of oculomotor nerves and fibrosis seen in areas of lateral rectus that were not innervated^{3, 4}. MRI (magnetic resonance imaging) was used to demonstrate absence of cisternal portion of abducens nerve in Duane's retraction syndrome by Parsa CF et al for the first time⁵.

Materials and methods:

A total of seven cases (eight eyes) including five cases with type I and two case with type III were included in the study. One case was found to be bilaterally affected type I. Six cases were unilaterally affected. In this study left eye was

involved in four cases and right in two cases. Five patients were female and two male. The mean age was 13 years. Ten children (mean age 13years) were included as control subjects to evaluate the accuracy of technique for visualization of abducens nerve. They were selected from the patients who underwent brain MRI for headache.

Test was performed with a 1.5 Tesla MRI system using 8- channel phased array neurovascular coil (HDc signa, General Electric, Milwaukee). Imaging of cisternal portion of cranial nerve was performed at brainstem level by taking continuous axial slices using heavily T2 weighed 3D FIESTA sequence, which provides good contrast against the surrounding cerebrospinal fluid. 3D FIESTA is a generic name for 3D steady state free process (SSFP) pulse sequence. The parameters used during scan were as follows. TR (repetition time) 10.6ms, TE (echo time) 2.6ms, slice thickness 0.8mm, field of view 19cm, matrix 320x192, NEX 2. In addition spin echo T1 weighted images (T1W), fast spin echo T2 weighted images (T2W) and fluid attenuation inversion recovery images (FLAIR) of entire brain was performed to rule out other intracranial abnormality.

Results:

Among seven cases of clinically diagnosed Duane's retraction syndrome, bilateral abducens nerves were absent in one affected patient. In four cases of left sided Duane's retraction syndrome the abducens nerve was absent on left side. The right abducens nerve was absent in two cases of right sided Duane's retraction syndrome. Among the 10 control subjects right and left abducens nerves were well identified in all cases. There was no other gross intracranial abnormality identified.

Discussion:

Duane's retraction syndrome is a congenital eye movement disorder characterized by marked limitation or absence of abduction, variable

limitation of adduction and palpebral fissure narrowing and globe retraction on attempted adduction. However these characteristic diagnostic signs of Duane's retraction syndrome may not be manifested in some patients, particularly in children and thus magnetic resonance imaging has been used for diagnosis of Duane's retraction syndrome⁶. MRI is very helpful for accurate visualization of normal abducens nerve and congenital abnormalities of abducens nerve⁷. The syndrome has been divided into three types depending on the amount of aberrant innervation to lateral rectus. Type-I is characterized by limited or absent abduction with relatively normal adduction. Type-II has limited or absent adduction with relatively normal abduction. Type-III has limited or absent abduction and adduction. It has been reported that females are more affected than males and left eye is more frequently involved than right eye⁸. New classification of Duane's retraction syndrome has subdivided type-I into three subgroups as type-I A, type-I B, type-I C and also described synergistic divergence type a variant of Duane's retraction syndrome type II. Among these types of Duane's retraction syndrome type-I is most common followed by type-III and type-II⁹. Some of the proposed etiologic factors for this syndrome are brainstem pathologies, orbital pathologies, trauma during labour, absent abducens nerve and nucleus¹⁰. However autopsy studies showed absent abducens nerve and nucleus and abnormal innervation of lateral rectus by inferior division of oculomotor nerve in this syndrome^{3,4}.

Duane's retraction syndrome is sporadic in 90% of cases and upto 10% cases are familial. Most cases of familial Duane's syndrome are autosomal dominant. This can be associated with dysplasia of radial bone, radial artery and thumb and it is known as Duane-radial-ray syndrome. Genetic studies

showed the location of a Duane's retraction syndrome on chromosome 2.^{11,12}

In this study, we aimed to visualize the cisternal portion of abducens nerve by high resolution MRI to investigate the role of aplasia of abducens nerve on the affected side in the etiology of Duane's retraction syndrome. The abducens nerve nucleus is situated in pontine tegmentum near midline just anterior to fourth ventricle. Anatomically five segments of nerves can be defined and they are intra-axial, cisternal, intra-dural, cavernous and intra-orbital. The axons of abducens nerve, course antero-inferiorly through pontine tegmen and exit the brainstem at pontomedullary sulcus level. The nerve runs antero-superiorly and laterally in prepontine cistern and then penetrates dural matter of basisphenoid to enter Dorello canal, then the nerve penetrates the cavernous sinus and runs along infero-lateral aspect of cavernous internal carotid artery, finally it enters orbit through superior orbital fissure and passing through annulus of Zinn, it supplies lateral rectus muscle. Among these segments of nerves cisternal segment of nerve is

reliably visualized on high resolution MRI. The nerve entrance into Dorello canal may be visualized due to invagination of cerebrospinal fluid into proximal canal¹³.

Diagnosis of Duane's retraction syndrome in young children is more challenging clinically, because they may present with only abduction deficit without globe retraction and upward or downward shooting. In this situation high resolution MRI is very helpful to demonstrate absence of abducens nerve to correctly diagnose Duane's retraction syndrome from other diseases such as abducens palsy, congenital esotropia.

The correct differentiation is important for management⁶.

Conclusion:

High resolution MRI is a very useful non invasive investigation for demonstration of absent abducens nerve in Duane's retraction syndrome, this helps in conforming the diagnosis. It is also useful for differentiating from other causes of loss of abduction, which helps in employing appropriate management strategy which yields good results.

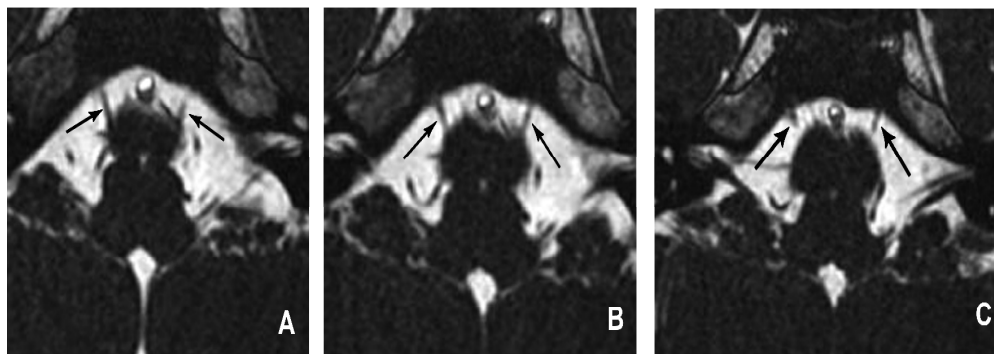


Figure .1: Axial 3D FIESTA MR images at level of pons show the entire course of right and left abducens nerve (arrows) as linear dark structures emerging from the pontomedullary sulcus (a), coursing in superior direction in (b) and finally entering the Dorello canal in (c).

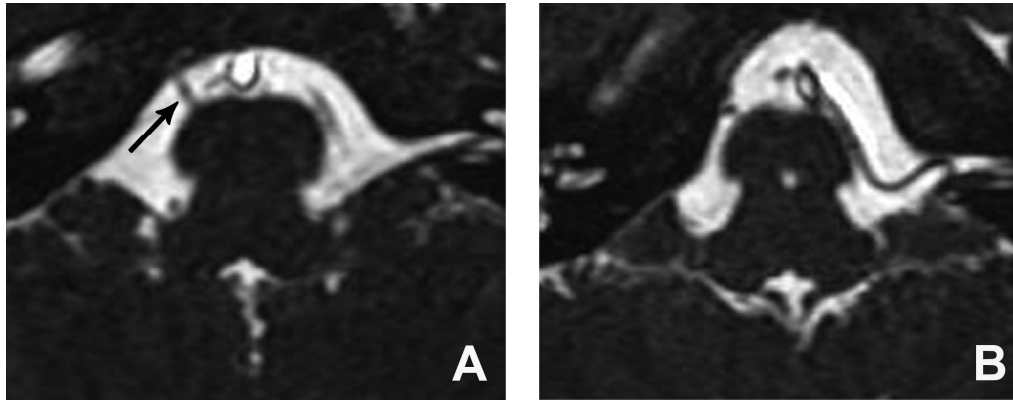


Fig 2. (a) Axial 3D FIESTA MR image at the level of pons, show the right abducent nerve(arrow) and absent left abducent nerve. (b) shows absent bilateral abducent nerve at expected location in another patient.

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