

A Case of Congenital Inverse Duane's Retraction Syndrome

Helen Lew¹, Jong-Bok Lee¹, Hee-Seon Kim², and Sueng-Han Han¹

Abstract

Inverse Duane's retraction syndrome is very uncommon. Congenital cases are even more unusual. A 6-year-old girl with convergent squint along with severe restriction on abduction is described. On attempted abduction, a narrowing of the palpebral fissure, upshoot and retraction of the eyeball were observed. Brain and orbit MRI demonstrated no intracranial or intraorbital mass, fracture, or entrapment of the medial rectus. Forced duction test was strongly positive. The primary lesion was found to be a tight medial rectus with shortening and soft tissue contracture. Surgical tenotomy of the medial rectus led to successful postoperative motility, but some limitation at full adduction and abduction persisted. This is a case reported with congenital medial rectus shortening, suggesting that this condition may be one of the etiologies of the rare inverse Duane's retraction syndrome.

Key Words: Forced duction test, inverse Duane's retraction syndrome, tenotomy, tight contracture

INTRODUCTION

Duane's retraction syndrome is a well-known congenital musculo-facial anomaly.¹ The etiology of the syndrome is found to be complex. The electromyographic data suggest that there is a paradoxical innervation in Duane's retraction syndrome.² Co-contraction of the medial and lateral rectus muscles in adduction was thought to be responsible for the retraction of the globe with the explanation that the oculomotor nerve may send fibers to the lateral rectus.³ There has been another suggestion that most of the innervation to the extraocular muscles reaches them via the gamma efferent fibers. These carry impulses to the muscle spindles. The spindles function as length setters or automatic load compensators. They act via the servo-loop through the spindle afferents and the main alpha motor neurons. There exists a direct anatomical link within the muscle by which the alpha motor neurons influence the discharge of the spindle afferents. Should this link be

disturbed by the muscle being abnormally stiff, then this servo-loop would be broken or drastically modified. As a result, much of the innervation destined for the alpha motor neurons and the main muscle fibers would never reach them.⁴

Inverse Duane's retraction syndrome is a condition with reverse clinical features.⁵ Abduction of the affected eye is possible to some extent and it is accompanied by retraction of the eyeball, narrowing of the palpebral fissure, and pseudoptosis. There may be some retraction of movement on adduction. The primary lesion is suspected to be the medial rectus muscle. Frequently, the muscle is of this sort following trauma to the medial wall of the orbit. In both conditions, with the limitation of abduction, the patient generally shows an esotropia in the primary position. There is only one case of congenital inverse Duane's retraction syndrome reported by Chatterjee et al., of which the cause was found to be extensive fibrous bands surrounding the medial rectus muscles extended to the medial orbital wall.⁶ However, we report a typical case of congenital inverse Duane's retraction syndrome whose medial rectus muscle showed no entrapment from trauma or medial orbital wall-related deformity.

CASE REPORT

A 6-year-old girl had complained of her squint

Received July 14, 1999

Accepted September 3, 1999

¹Department of Ophthalmology, Yonsei University College of Medicine, ²Department of Ophthalmology, Cha University College of Medicine, Sungnam, Korea.

Address reprint request to Dr. J. B. Lee, Department of Ophthalmology, Yonsei University College of Medicine, C.P.O. Box 8044, Seoul 120-752, Korea. Tel: 82-2-361-8450, Fax: 82-2-312-0541, E-mail: 491209@yumc.yonsei.ac.kr

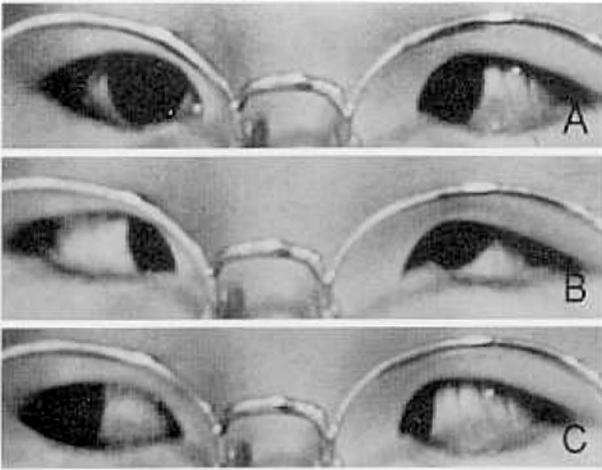


Fig. 1. (A) Primary position shows the marked esotropic deviation at primary position. (B) At left gaze, left eye has not moved past midline. Retraction of left eye and narrowing of the palpebral fissure with upshoot were noted. (C) At right gaze, limitation of adduction was not noted in left eye.

since birth. There was one member of the family having a squint on the maternal side. No history of forceps delivery or trauma was confirmed. Visual acuity was RE 20/30, 20/20 corrected with-sph 0.75 Diopters and LE 20/200, which was not improved with correction. The results of the anterior segment examination were normal. Fundus showed no specific signs in both eyes. The Krimsky method showed 45 prism diopter left esotropia. Abduction was markedly restricted and the eyeball could not cross the midline to the left side. Retraction of the eyeball and narrowing of the palpebral fissure with upshoot were noted on attempted abduction and slight widening of the palpebral fissures on adduction (Fig. 1). Forced duction test showed marked restriction of abduction. Magnetic resonance imaging (MRI) performed with axial and coronal images did not disclose any abnormalities of the brain and orbital structures. No routine lab study for general anesthesia were of value.

During the operation, extensive restriction and tight contracture of the medial rectus muscle were noted. The insertion site of the medial rectus was about 3.5 mm distant from the limbus. Tenotomy of the medial rectus muscle was carried out instead of medial rectus recession on the left side. After the tenotomy, forced duction tests were performed again. There was much alleviation of the restriction and the eye could be abducted fully by forced duction test (Fig. 2). The primary position of the globe was cos-

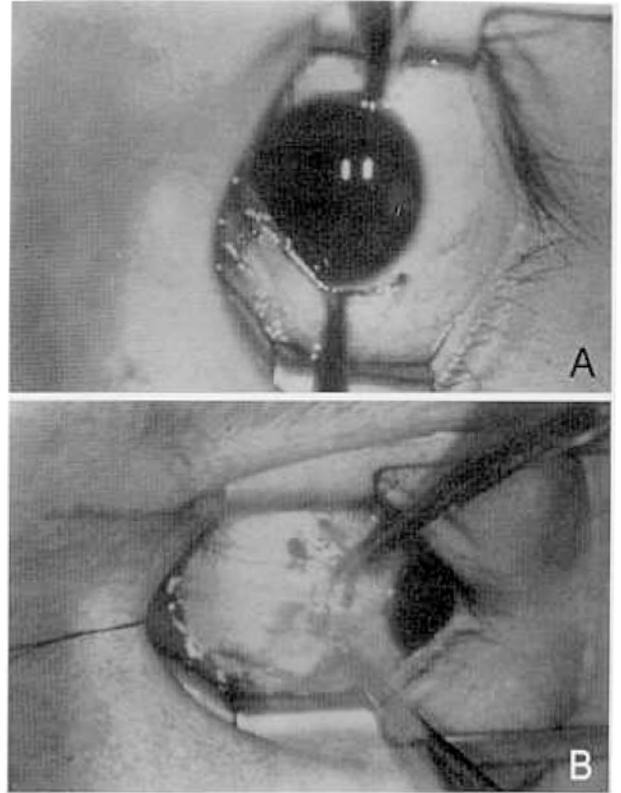


Fig. 2. (A) Intraoperatively forced duction test is performed. (B) After tenotomy, the result of forced duction test was much improved.



Fig. 3. (A) At primary position, left eye is almost straight. (B) At left gaze, slight widening of the palpebral fissure and improvement of limited abduction were noted. (C) At right gaze, slight limitation of adduction of left eye was noted.

metically acceptable. The medial rectus muscle shortening was suspected to be the main etiology of the condition. Postoperatively, esotropia was much im-

proved and the eyeball could cross the midline to the left side. On attempted abduction, slight widening of the palpebral fissures was noted and upshoot of the eyeball disappeared. However, partial limitation of abduction and adduction persisted (Fig. 3).

DISCUSSION

Duane's retraction syndrome is more common in females. It is usually thought to occur as an isolated sporadic entity although familial patterns have been observed in 5 to 10% of cases.⁷ Even though the involvement is more frequent on the left side, it is bilateral in 18% of cases.⁸ A number of associated findings have been reported with retraction syndrome.⁹ This has led some authors to propose that a teratogenic insult at about eight weeks gestational age may be the cause. The associated anomalies include spinal deformities, ear malformations with hearing defects, palatal changes, epibulbar dermoids, and other syndromes. Perceptive deafness is associated in 18% of cases.¹⁰

The abnormality in Duane's retraction syndrome is primarily related to paradoxical innervation of the lateral rectus muscle. A wide variety of innervation abnormalities of the lateral recti have been reported in many studies including co-firing in adduction, reduced activity on attempted abduction, inappropriate responses on upgaze and downgaze and abnormal oblique movements. Globe retraction and narrowing of the palpebral fissure in adduction is also thought to be related to this primary innervational anomaly of co-contraction of the horizontal rectus muscles, or additional involvement of the inferior and superior rectus muscles as well.¹¹

Pseudo-Duane's retraction syndrome is most often due to trauma of the medial wall with entrapment of the medial rectus extending through the fractured lamina papyracea. It may occur secondarily to other changes in the orbit, especially in the medial wall as Chatterjee et al reported.⁶ With medial rectus entrapment, just the opposite of Duane's retraction syndrome is present, namely the inability to abduct because of the involvement of the medial rectus. In both conditions, a narrowing of the palpebral fissure appears with a retraction enophthalmos and pseudoptosis occurring secondarily to the attempted movement. However, the etiology and pathogenesis of the

syndromes are entirely different as mentioned above. Duane et al concluded that the terminology most appropriate for the entrapped medial rectus features to be: pseudo-Duane's retraction syndrome and carefully avoided such words as reverse, inverse, and mirror image because they found them either inaccurate or conflicting with their observations.⁵ However, the condition with reverse clinical features mentioned above is usually described to be inverse Duane's retraction syndrome on the basis of clinical manifestations in textbooks and many articles.⁶

Inverse or pseudo-Duane's retraction syndrome is very uncommon. The fact that other congenital anomalies can be associated with this condition could not be justified in this patient. One feature of this particular case is worth noticing. Most cases of the inverse retraction syndrome result from the entrapment of the medial rectus muscle in the medial orbital wall due to trauma.¹² However, there was neither a history of injury nor any intraorbital mass in this case. Only the medial rectus shortening with soft tissue contracture was found. Tenotomy was the dramatic method of treatment for motility recovery. However, the partially remaining limitation of full abduction may be due to soft tissue contracture and fibrosis around the medial rectus, which could not fully be dissected in the deep retrobulbar space. Furthermore, the limitation of adduction may result from the medial rectus disinsertion.

There are some cardinal features of the inverse or pseudo-Duane's retraction syndrome which should be highlighted. Radiological studies such as CT or MRI scans are of maximum help and are essential to the diagnosis when medial rectus entrapment is suspected. Forced duction tests, performed at the time of surgery, not only help establish the degree of entrapment of the medial rectus, but also serve as indicators as to whether the malady has been alleviated with attempted surgical intervention. Surgical prognosis ranges from fair to excellent in pseudo-Duane's retraction syndrome except in such entities as metastatic orbital disease. In general, the longer the condition has been present, the poorer the prognosis will be for full recovery of lateral motion with complete elimination of lateral gaze diplopia. Unlike the classic Duane's retraction syndrome, amblyopia is not a problem in inverse Duane's retraction syndrome resulting from traumatic entrapment of the medial rectus.

However, this congenital case showed amblyopia suggesting other characteristics of congenital inverse Duane's retraction syndrome. Shortening of the medial rectus muscle is suspected to be the main etiology of the congenital inverse Duane's retraction syndrome.

REFERENCES

1. Duane A. Congenital deficiency of abduction with impairment of abduction, retraction movements, contraction of the palpebral fissure, and oblique movements of the eye. *Arch Ophthalmol* 1996;114:1255-6.
2. Sato S. Electromyographic studies on Duane's retraction syndrome. Qualitative changes of motion currents of extraocular muscles in ocular movements. *Acta Soc Ophthalmol Jpn* 1959;63:2028-30.
3. Hoyt WF, Nachllgaller H. Anomalies of ocular motor nerves: Neuroanatomic correlates of paradoxical innervation in Duane's syndrome and related congenital ocular motor disorders. *Am J Ophthalmol* 1965;60:443-8.
4. Hughes WF. The year book of ophthalmology. Chicago: Year Book Medical publishers; 1978. p.62-3.
5. Duane TD, Schatz NJ, Caputo AR. Pseudo-Duane's retraction syndrome. *Trans Am Ophthalmol Soc* 1976;74:122-32.
6. Chatterjee PK, Bhunia J, Bhahhacharyya I. Bilateral inverse Duane's retraction syndrome-a case report. *Indian J Ophthalmol* 1991;39:183-5.
7. Kirkham TH. Inheritance of Duane's syndrome. *Br J Ophthalmol* 1970;54:323-9.
8. Aebh R. Retraction syndrome. *Arch Ophthalmol* 1933;10:602-5.
9. Cross HE, Pfaffenbach DD. Duane's retraction syndrome and associated congenital malformations. *Am J Ophthalmol* 1972;73:442-50.
10. Pfaffenbach DD, Cross HE, Kearns TP. Congenital anomalies in Duane's retraction syndrome. *Arch Ophthalmol* 1972;88:635-9.
11. DeRespinis PA, Caputo AR, Wagner RS, Guo S. Duane's retraction syndrome. *Surv Ophthalmol* 1993;38:257-88.
12. Gittinger JW Jr, Hughes JP, Suran EL. Medial orbital wall blow-out fracture producing an acquired retraction syndrome. *J Clin Neuroophthalmol* 1986;6:153-6.