Journal of Clinical & Experimental Ophthalmology

Taisan and Owaifeer, J Clin Exp Ophthalmol 2018, 9:1
DOI: 10.4172/2155-9570.1000708

Case Series Open Access

# Case Series: Botulinum Toxin in the Management of Duane Retraction Syndrome type I

**Abdulaziz A Al Taisan*** **and Adi M Al Owaifeer**

*Depatment of Ophthalmology, King Faisal University, Saudi Arabia*

***Corresponding author:** Abdulaziz A Al Taisan, Depatment of Ophthalmology, King Faisal University, Saudi Arabia, Tel: + 966500188442; E-mail: dr.altaisan@gmail.com

**Received date:** January 15, 2018; **Accepted date:** January 25, 2018; **Published date:** January 31, 2018



## Abstract

**Introduction:** Duane retraction syndrome (DRS) is a form of congenital strabismus related to impairment of extraocular muscle innervation. The condition was first described by Mr. Duane as "the absence or limitation of abduction with narrowing of palpebral fissure in adduction and globe retraction". DRS is thought to be caused primarily by agenesis of the sixth nerve nucleus, leading to fibrosis of the lateral rectus muscle. Horizontal rectus muscle recession is still considered the preferable surgical intervention. Indications include large deviation in primary position, abnormal head position or significant upshoot or downshoot. The rule of botox in DRS is still not fully understood. In this short series we aim to explore the efficacy of botulinum toxin as an alternative to surgical intervention in patients with DRS type 1.

**Cases:** The series include four patients of DRS type 1. Three patients underwent botox injection as a primary mode of treatment and one patient had a history of strabismus surgery two years prior to botox injection. Two patients maintained orthophoria over a follow-up period of 24 months; one patient didn't benefit from botox injection with similar angle of deviation pre and post injection. The fourth patient with history of strabismus surgery responded with reduction of the angle of deviation in the first and second visits, with return to the pre-injection angle on the six-month follow-up visit.

**Conclusion:** Botulinum toxin can serve as an alternative to surgery in DRS type I especially in young age groups where surgical intervention is not preferable. Further large-scale studies are needed to evaluate its use in other types of the syndrome and to explore its long-term effect.

**Keywords:** Duane; Strabismus; Botox; Botulinum toxin

## Introduction

Duane retraction syndrome (DRS) is a form of congenital strabismus related to impairment of extraocular muscle innervation [1]. The condition was first described by Mr. Duane as "the absence or limitation of abduction with narrowing of palpebral fissure in adduction and globe retraction" [2]. DRS is thought to be caused primarily by agenesis of the sixth nerve nucleus, leading to fibrosis of the lateral rectus muscle [3,4].

Huber classified DRS into three types based on Electromyography (EMG) findings. Type I is characterized by limitation of abduction with globed retraction and palpebral fissure narrowing in adduction. EMG shows paradoxical innervation of lateral rectus on adduction, it is considered the most common type of DRS (70-80%). Type II (7%) is characterized by limitation of adduction with exotropia in primary position, globe retraction and palpebral fissure narrowing in adduction. EMG shows maximal innervation of lateral rectus on abduction and a paradoxical lower impulse in adduction. Type III (15%) shows limitation of adduction and abduction with globe retraction and palpebral fissure narrowing on attempted abduction. EMG shows simultaneous innervation of medial and lateral recti in primary gaze, adduction and abduction [5].

Horizontal rectus muscle recession is still considered the preferable surgical intervention [6]. Indications include large deviation in primary position, abnormal head position or significant upshoot or downshoot [7]. The primary aim of surgery is to tackle the deviation in primary position and the abnormal head position, secondary aim is to eliminate upshot, downshoot and globe retraction. Reports show a success rate of 50-80% [8,9].

Botulinum toxin was introduced to the medical field in 1973 by Dr. Scott where he studied its effect on ocular alignment [10]. Since then, its uses have expanded exponentially to include infantile esotropia, partially accommodative esotropia and many more [11,12].

The rule of botox in DRS is still not fully understood. In this short series, we aim to explore the efficacy of botulinum toxin as an alternative to surgical intervention in patients with DRS type 1.

## Cases

### Case 1

A nine-month-old male presented to the clinic with a left eye deviation since birth. On examination, the patient was following and fixating, left esotropia of 20 PD, limitation of abduction of both eyes (-1 right eye and -3 left eye), otherwise the ocular exam was normal. He was diagnosed to have bilateral DRS type 1. He underwent a part-time occlusion (2 h/day, right eye). At the age of twelve months, he

underwent bilateral botulinum toxin injection 7.5 IU to each medial rectus. In the first post-injection visit (two weeks) he was having ptosis and exotropia. The patient was then followed up at 3, 6, 12, 18 and 24 months with maintained orthophoria for both near and distant fixation.

### Case 2

A twelve-year-old male patient presented with esotropia, he was having 10 PD right esotropia at near and 18 PD esotropia at distance, left medial rectus restriction (-1). Cycloplegic refraction was plano in both eyes and visual acuity was 20/25 in the right eye and 20/22 in the left eye. A diagnosis of DRS type 1 was established. The patient underwent right medial rectus botox injection (5 IU). In the first post-injection visit, the patient was having right esotropia measuring 10 PD that persisted over three months. Botox injection was not repeated and the patient underwent right medial rectus recession with resultant orthophoria over a follow-up period of twelve months.

### Case 3

A sixteen-month-old male patient with esotropia noticed since birth, he was having a large angle esotropia and bilateral restriction of abduction, otherwise his ocular exam was normal. Cycloplegic refraction was done (+4.50 -0.50 × 180 in both eyes) and spectacles were prescribed. reevaluation 3 months later confirmed the diagnosis of bilateral DRS type 1. The patient was planned for bilateral medial rectus recession but the parents were not motivated for surgery. As a result, an alternate plan was taken and botulinum toxin was injected into both medial recti (5 IU). In the first post-injection visit, the patient was having bilateral ptosis and orthophoria. Subsequent visits revealed maintained orthophoria over eighteen months.

### Case 4

A 7-year-old male diagnosed with right DRS type 1.2 years prior to presentation. He underwent right medial rectus recession (6 mm) in another hospital, 1 year prior to presentation. On examination, visual acuity (VA) was 20/22 in the right eye and 20/20 in the left eye, cycloplegic refraction was plano in both eyes. He was having limitation of abduction of the right eye (-1) and a right face turn, in primary position: right esotropia (ET) of 10 prism diopters (PD) at distance and 18 PD at near, otherwise his ocular examination was normal. The patient underwent left medial rectus recession 5.5 mm. Postoperatively the deviation persisted as 10 and 20 PD for distance and near consecutively and persistent right head turn. The patient was observed for multiple visits with repeated cycloplegic refraction, about two years later he underwent botulinum toxin injection bilaterally to the medial rectus, the dose was 7.5 IU. Post-injection, the patient was orthophoric at 2 weeks and 3 months, but at the 6-month visit, he was esotropic 12 PD at distance and 18 PD at near. He was given a base out prism over his spectacles.

## Discussion

DRS is a congenital abnormality in extraocular motility where horizontal muscle recession surgery is considered the preferred intervention. The main aim of surgery is to improve the primary deviation and to address the head turn [13]. Reports show that surgical intervention has a success rate of 50-80% [8,9]. Botulinum toxin injection is a rising technique that can be utilized in the management of DRS as an alternative to surgical intervention. The main advantage of botulinum toxin injection is that it is characterized by a low rate of complications when compared to muscle surgery [14]. Moreover, in cases of failure the injections can be repeated without producing permanent muscle damage. However, its efficacy is yet to be established in DRS.

In our series, two patients maintained orthophoria over a follow-up period of 18-24 months with a single injection. However, the third patient did not respond and maintained the same pre-injection angle. The fourth patient maintained orthophoria for six months only, followed by reappearance of esotropia. This patient had a history of failed strabismus surgery.

In two series, the success rate of botox injection in DRS ranged from 37-50%, with forced duction test as a possible predictive factor for response to botox [9,13].

In the series of Merino et al. [8], one case underwent botox injection post horizontal muscle recession surgery due to under correction, with resultant resolution of the deviation. However, botox was injected in the early post-operative period, in contrast to our case where botox injection was given two years after the recession surgery. Possible explanations of such outcome might be post-operative tissue-adhesion, scar formation or inadvertent muscle capture [14].

Surgical intervention in DRS is usually carried out at the age of 6-13 years to allow for stabilization of angle of deviation and face turn, and due to the fact that patients under the age of 4 years neither have large deviation nor significant face turn [15,16]. However, in our two successful cases, botox was injected at a younger age (12 and 20-month-old), due to the fact that accurate measurement of the angle of deviation is not required in botox injection in contrast to strabismus surgery. The ideal age for botox injection in strabismus is debatable, some authors advice for early injection to reduce tonic muscle contracture [17] while others prefer to delay the injection beyond two years of age to reduce the rate of reoperation and abnormal binocularity due to the improper alignment of eyes in the early post-operative period [18].

Two out of the three patients had transient ptosis and one patient had transient exotropia, however these complications resolved in the second post-injection visits. No other complications such as scleral perforation, sub conjunctival hemorrhage were reported.

A small number of patients, variable botox doses, no objective measurement of face turn, and lack of specific follow up protocol were among the limitation of this short series.

## Conclusion

Botulinum toxin can serve as an alternative to surgery in DRS type I especially in young age groups where surgical intervention is not preferable. Further large-scale studies are needed to evaluate its use in other types of the syndrome and to explore its long-term effect.

## References

1. Duane A (1905) Congenital deficiency of abduction associated with impairment of adduction, retraction movement, contraction of the palpebral fissure and oblique movements of the eye. Arch Ophthalmol 34: 133-145.
2. Wilson ME, Buckley EG, Kivlin JD (2004-2005) Special forms of strabismus. In: Pediatric Ophthalmology and Strabismus, Basic and clinical science course, AAO, San Francisco, 6: 11.

3. Tomi A, Preda C, Poenaru O, Zamfiroiu F (2005) Duane's syndrome – Etiopathogenesis, clinical features and diagnosis. Ophthalmologia 49: 10-14.
4. Kim JH, Hwang JM (2005) Presence of the abducens nerve according to the type of Duane's retraction syndrome. Ophthalmology 112: 109-113.
5. Huber A (1974) Electrophysiology of the retraction syndromes. Br J Ophthalmol 58: 293-300.
6. Farvardin M, Rad AH, Ashrafzadeh A (2009) Results of bilateral medial rectus muscle recession in unilateral esotropic Duane syndrome. J AAPOS 13: 339-342.
7. BCSC (2010-2011) 6: 127-130.
8. Merino P, Merino M, Gómez De Liaño P, Blanco N (2012) Horizontal rectus surgery in Duane syndrome. Eur J Ophthalmol 22: 125-130.
9. Maya JF, de Liaño RG, Catalán MR, Rayward O (2013) Botulinum toxin treatment in patients up to 3 years of age who have esotropic Duane retraction syndrome. Strabismus 21: 4-7.
10. Scott AB, Rosenbaum A, Collins C (1973) Pharmacologic weakening of extraocular muscles. Invest Ophthalmol 12: 924-927.
11. Dutton J, Fowler A (2007) Botulinum toxin in ophthalmology. Surv Ophthalmol 52: 13-31.
12. Emel B, Ceyhun A (2016) Use of botulinum neurotoxin in ophthalmology. Turk J Ophthalmol 46: 282-290.
13. Ahmad A, Farzad F, Fatemeh B, Arash M, Akbari M, et al. (2017) Botulinum toxin injection in the patients with Duane syndrome type 1. J Curr Ophthalmol 29: 50-53.
14. Seyhan BO (2016) Restrictive problems related to strabismus surgery. Taiwan J Ophthalmol 6: 102-107.
15. Fricke J, Neugebauer A, Russmann W (2006) Surgical options in retraction syndrome. Klin Monatsbl Augenheilkd 223: 42-47.
16. Rowe F, Noonan C (2009) Complications of botulinum toxin and their adverse effects. Strabismus 17: 139-142.
17. Campos EC (2008) Why do the eyes cross? A review and discussion of the nature and origin of essential infantile esotropia, microstrabismus, accommodative esotropia, and acute comitant esotropia. J AAPOS 12: 326-331.
18. Leffler CT, Vaziri K, Schwartz SG, Cavuoto KM, McKeown CA, et al. (2016) Rates of reoperation and abnormal binocularity following strabismus surgery in children. Am J Ophthalmol 162: 159-166.