Duane Retraction Syndrome

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Abstract

Purpose of Study: To evaluate and to manage if management indicated for cases of Duane retraction syndrome.

Patients and Methods: 15 Duane retraction syndrome (DRS) patients seen in Pediatric clinic in Tripoli Eye Hospital in period from January 2006-December 2006. Complete ophthalmic examination including ortho-optic assessment for all cases.

Results: Patients age ranged 1 year to 20 years in this group of study, females were affected more than males with 2 to 1 ratio. Type 1 (esotropic) is the most common 80% of cases. Left eye was affected more than right eye. Bilateral in 13.3% of cases. DRS clinical picture varies widely, surgical intervention will not eliminate the abnormality but will lessen it. Two cases were operated upon to improve alignment in primary position. Medial rectus muscle was found too tight on forcedduction test surgery included bilateral medial rectus muscle recession.

Conclusion: DRS picture varies widely, no treatment is available to eliminate the abnormalities but only to lessen them. The limitations in treatment are due to poor correction of ductions and versions. Also, up shoot, down shoot and enophthalmos correction is not complete.


Introduction

DUANE Retraction Syndrome (DRS) is an abnormal pattern of ocular motility characterized by retraction of globe with narrowing of palpebral fissure on attempted adduction and a variety of other abnormal movement of the affected eye when the other eye fixates in various cardinal positions [1,2,3].

The abnormal pattern of ocular motility of DRS is the result of developmental adaptation that occurs in the embryo as consequence of the absence of 6th cranial nerve [4,5,6].

Electromyography studies have shown paradoxical innervations of Lateral rectus muscle and anomalous synergistic innervations of medial rectus, inferior rectus, superior rectus and oblique muscles [7,8].

In most cases of DRS the entire 6th nerve atrophy instead of post half of 6th nerve (without specific teratogenic stimulus) 95% of DRS cases this is the only initial abnormality. In about 5% of cases other abnormalities seen (e.g. nerve deafness). Most cases of DRS are sporadic [5,9].

Etiology:

Etiology of DRS has been proposed by several investigators to be fibrosis of lateral rectus (LR)-deficient abduction and retraction of the globe on adduction, due to the pull of medial rectus (MR) against the tight LR [7].

Abnormal innervation of LR-co-correction of LR and MR on adduction-retraction [10].

Co-contraction of vertical recti on adduction-retraction of the globe.

Protrusion of globe on attempted abduction, due to co-contraction of oblique muscles [11].

So, the etiology of DRS may be a mixture of anatomical and neurological anomalies. Therefore, Fibrosis of LR may be secondary to abnormal innervation. Electromyographic studies show co-contraction of horizontal recti, also show co-contraction of vertical recti on adduction [13].

Classification

HUBER Classification:

- Type (1):
  - Esotropia in PP, restrict abduction.
  - Most common type.
Differential diagnosis of Duane’s retraction syndrome:
1- Abducent nerve palsy: There is no retraction of the globe on attempted adduction. No widening of palpebral fissure nor protrusion of the globe on attempted abduction.
2- Pseudo-Duane: Due to fracture of the medial orbital wall-entrapment of medial rectus muscle. CT scan will reveal the problem [12,13].

Aim of study:
To evaluate the clinical findings in DRS cases seen in year 2006 in Pediatric Clinic in Tripoli Eye hospital and to manage cases if management indicated.

Patients and Methods
This study included 15 patients in period (January 2006-December 2006) in Tripoli Eye Hospital (Pediatric Clinic). Age range from 1 year to 20 years. All cases were subjected to the following:
1- History: About onset, face turn, progression and family history.
2- Ocular examination:
a- Vision and refraction under cycloplegia to reach best corrected vision.
b- Ortho-optic status.
c- Ocular motility.
d- Anterior segment.
e- Fundus examination.
f- Systemic examination for any associated anomaly.

Results
Positive history of deviation, abnormal motility in all patients.
No history of similar problem in the family their families.
DRS was seen more in female patients in this group of study 66.7% females, 33.3% males (Fig. 1).

Fig. (1): Gender distribution in DRS cases in Pediatric Clinic 2006.

DRS distribution according to type: (Fig. 2)
• Type (1) 80%.
• Type (2) 13%.
• Type (3) 7%.

Fig. (2): Distribution of Duane syndrome according to the type.
Type 1:
- In 80% of this group of study.
- Esotropia with head straight or no deviation in primary position.
- Face turn in 2 cases of this type.
- Limited abduction.
- Narrowing of palperal fissure on adduction.
- Enophthalmos on adduction.

Picture (1): No deviation in pp, limited abduction and retraction.

Picture (2): L (esotropic in pp, defective abduction, retraction).

Picture (3): L (esotropic in pp, limited abduction).

Picture (4): Marked retraction on adduction.
Type 2:
- 13% in this group of study exotropia.
- Limited adduction, full to slight limited abduction.
- Narrowing of palperal fissure & enophthalmos.

Type 3:
- In 7% in the group of study.
- Eyes are aligned in Primary position, with head straight, with fusion.
- Limited abduction and adduction.
- Severe up and down shoot.

- DRS seen more in left eye in this group of study 53.33%.
- DRS seen in right eye in 5 patients 33.33%.
- Bilateral in 2 patients 13.33% (Fig. 3).

- Good fusion in 2 patients 13.3%.
- Due to anisometropia amblyopia seen in 6 patients 40%.
- Occlusion therapy trial failed in 1 patient (7 years old, type 1) patient operated.
- Anterior segment and fundus normal in all patients.
- 2 patients: Pre-auricular skin tag (Picture 6).
- (Ear-Nose-Throat examination: normal).
- 1 patient has hand anomaly.
- No other abnormality detected.
**Discussion**

- The clinical picture of DRS varies widely [2].
- Careful assessment of ocular motility is important [1,3,5].
- Each case of DRS requires unique approach to treatment if treatment is even considered.
- The goals of treatment of DRS are:
  - Improve head posture.
  - Reduction of up and down shoot.
  - Reduction of enophthalmos.
  - Alignment of eyes in PP [6,7,10].
- No treatment available will eliminate the abnormality of DRS but will lessen them [10].
- The limitation of treatment:
  - Normal ductions and versions cannot be achieved.
Up shoot, down shoot & enophthalmos can be greatly reduced but not eliminated [10].

Two patients operated to improve alignment in primary position in this group of study (13.3%). Medial rectus muscle were too tight on forcedduction test. Both Medial rectus muscle were recessed with acceptable improvement of head posture, reduction in enophthalmos.

References

1- HELVESTON E.: Surgical management of strabismus 5: Wayenborgh, Belgium, Chapter 5: 149-150, 2005.