



# PREVALENCE OF DUANE RETRACTION SYNDROME OF INDIGENOUS STRABISMIC PATIENTS PRESENTING IN MAYO HOSPITAL LAHORE

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**Purpose:** To find out the prevalence of Duane's retraction syndrome in strabismic patients and its types

**Methodology:** In this descriptive cross sectional study, 900 strabismic patients presenting in orthoptic clinic of Mayo Hospital from January 2010 to October 2011 underwent orthoptic assessment. Visual Acuity was recorded with VLC monitor. Orthoptic assessment and ocular motility testing were done. Diagnosis was then made on the basis of horizontal eye movement limitation, Globe Retraction with Palpebral Aperture Narrowing on attempted adduction, enophthalmos and Upshoot/Downshoot.

**Results:** Out of 900 strabismic patients, 29(3.2%) patients had Duane Syndrome. 25(2.7%) patients had Duane type I, 3(0.3%) patients had Duane Type III and 1(0.1%) patient had Duane Type II.

**Conclusion:** The prevalence of Duane Retraction Syndrome was found to be 3.2% which is significantly high.

**Keywords:** *Duane retraction syndrome, globe retraction, peplpebral aperture narrowing*



## INTRODUCTION

Duane syndrome, also called Duane retraction syndrome (DRS), is a group of ocular motility disorders characterized by difficult or restricted rotation of one or both eyes. This restriction can both be outward (abduction) or inward (adduction). It is usually associated with a narrowing of palpebral aperture and enophthalmos (retraction of the eyeball) on adduction.

Other associated ocular anomalies include heterochromia, iris dysplasia, ptosis, keratoconus, nystagmus, papillary anomalies, choroidal colobomas, dystichiasis, microphthalmos, optic nerve hypoplasia, Morning Glory syndrome and Marcus Gunn phenomenon.

Systemic associated anomalies include Goldenhar's syndrome, Klippel-Feil syndrome, vertebral column anomalies, facial anomalies, rib anomalies, sensory neural hearing loss, malformation of the external ear, Rubinstein-Taybi Syndrome, Dandi-Walker syndrome, genitourinary anomalies and anomalies of the limb, feet and hands.<sup>1</sup>

Duane's retraction syndrome (DRS) occurs due to abnormal development or absence of sixth (Abducent) nerve and its nucleus, usually associated with an abnormal splitting of nerve to the medial rectus muscle (i.e. inferior branch of 3<sup>rd</sup> nerve) so that it innervates both the medial and lateral rectus muscle simultaneously. Because both these recti are innervated by the nerve to the medial rectus muscle, any attempt at adduction causes the globe to retract, resulting in mild enophthalmos and diminishing of the palpebral fissure.<sup>2</sup>

Theories have been put forward to explain this mysterious disorder citing disorders at cellular level with cytopathological, neurological and embryological abnormal findings. These findings were derived from Electromyography (EMG), electro-oculography (EOG), auditory evoked potentials, and even autopsy.<sup>3</sup>

Some of these theories postulate that abnormal development of the neuronal cells in the sixth nerve nucleus results non functional abducent nerve with restricted or absent abduction. The lateral rectus is compensated by abnormally split branches originating from oculomotor nuclei through the third nerve. Some neuropathologists have reported an altogether lack of sixth nerve nucleus and nerve while others say that dual innervation of lateral rectus is present at least in the type I and II variants. Whatever the pathophysiology, DRS mechanically is characterized by the development of an abnormal communication between the innervation of the lateral & medial recti via the inferior division of cranial nerve III, producing a dual electrical firing of both these muscles during adduction, causing enophthalmos, and

palpebral fissure narrowing.<sup>4</sup>

Recent electromyographic studies have suggested that most cases of the Duane's retraction syndrome may indeed be due to an abnormal innervational mechanism rather than due to structural abnormalities.<sup>5</sup>

## Classification of Duane's Syndrome

### 1. Brown's Classification<sup>6</sup>

Based upon clinical observations, Brown has classified DRS into the following three subtypes,

- Type A: Limited abduction and less limited adduction (classical Duane syndrome).
- Type B: Limited abduction but normal adduction.
- Type C: Restriction of adduction being more than that of abduction, resulting in a divergent squint and a head posture with the face turned away from the side of the affected eye.

### 2. Huber Classification<sup>5</sup>

Huber based his classification on clinical variation:

- Type I: abduction is markedly restricted or completely absent; adduction may be normal or slightly defective; adduction results in narrowing of palpebral fissure and eyeball retraction; abduction results in increase in palpebral fissure width.

Type II: Adduction limited or absent, with divergent squint of the affected eye; abduction is normal or limited slightly; narrowing of palpebral fissure and retraction of the globe on attempted adduction.

Type III: Combination or absence of both abduction and adduction; retraction of the globe and narrowing of the palpebral fissure on attempted adduction.

### Clinical Findings and Diagnosis<sup>5</sup>

The classical Duane retraction syndrome has the following features:

1. Congenital with acquired forms being extremely rare.
2. Marked restriction of abduction
3. Mild limitation of adduction
4. Retraction of eyeball with narrowing of palpebral fissure on adduction
5. Often associated with variable degree of elevation





or depression in adduction

### Aims & Objectives

1. To find out the proportion of Duane's retraction syndrome in general population with strabismus and motility problems presenting in orthoptic clinic of Mayo Hospital of Lahore.
2. To find out the proportion of different types of Duane's syndrome as well.

### Method

Total of 900 strabismic patients of both genders presenting in orthoptic clinic of Mayo Hospital from January 2010 to October 2011 underwent orthoptic assessment. Visual Acuity was recorded with VLC monitor using decimal notation. Detailed orthoptic assessment and ocular motility testing were done. Diagnosis was then made on the basis different clinical features including type of deviation, limitation in horizontal eye movement, globe retraction with narrowing of palpebral aperture during adduction, enophthalmos and upshoot/ downshoot. Type of Duane's syndrome was also included in the diagnosis.

#### Inclusion Criteria

- Strabismic patients or patients with motility problems
- Patients of all age
- Best corrected visual acuity

#### Exclusion Criteria

- Patients diagnosed with paresis
- Any previous ocular surgery
- Any trauma
- Mentally handicapped

### Statistics

Data was collected. The analysis of the data was done by making graphs and tables in Microsoft word and excel.

### Results

Out of 900 strabismic patients, 29(3.2%) patients had Duane Syndrome. (Table 1)

25(2.7%) patients had Duane type I, 3(0.3%) patients had Duane Type III and 1(0.1%) patient had Duane Type II of all the 900 strabismic patients.

In this study Type I was found to be predominant of all type almost 86%, Type II was 4% and Type III was 10% of all the patients having DRS. (Table 2)

Out of 29 patients with DRS 16 (55.17%) were female and 13 (45%) were male. (Table 3)

Left eye was more affected about 48.3% than right eye 37.4%. Bilateral cases were found to be 13.8% of all the patients having DRS. (Table 4)

### Conclusion

In this study the prevalence of Duane's Retraction Syndrome of all the strabismic patients was found to be 3.2%. Type I proportion of all the duane's found to be 86%, females were more affected 55.1% and left eye 48.3 % was predominantly involved than right eye.

### Discussion:

DRS is described as having a sporadic occurrence with predominance for the left side and having a slight female preponderance.

The prevalence of Duane's Retraction syndrome in our study was 3.2%. According to Rhee DJ, DRS exist in 0.1 % of the general population and accounts for 1-5 % of all strabismus cases which could be correlated with our study.<sup>7</sup>

The relative proportion of Duane's r syndrome in a population of strabismus cases in Iran was found to be 1.7%.<sup>8</sup> In another study conducted, the prevalence of Duane's Syndrome is found to be 1-5/ 10,000 patients.<sup>9</sup>

Females (55.17%) were more affected with DRS than the males (45%). According to Gauri Shankar Duane's retraction syndrome is more common in female patients (58.8%) than in male patients.<sup>10</sup>

In the current study the proportion of DRS was more in left eye than right eye and bilateral which was 48.3%, 37.4% and 13.8% respectively and Type I was found to be 86% of all the DRS patients, Type II was 4% and Type III was 10 %. These can be correlated with other studies which also showed similar results. According to DeRespinis, DRS is more in females (58%) than males (42%) and Left is affected more (59%) than right eye (23 %) and bilateral (18%) respectively.<sup>5</sup>

In a study conducted by Zhang F, 65.88% of DRS cases had left eye involvement with Type I being the most common form. (184 patients, 91.54%).<sup>11</sup>


**Tables**
**Table 1: Prevalence of Duane's Retraction Syndrome**

Total no. of Patients	Patients with DRS (n)	Prevalence (%)
900	29	3.2%

**Table 2: Types of Duane's Retraction Syndrome**

Type I	Type II	Type III
86%	4%	10%

**Table 3: Gender Distribution of Duane's Retraction Syndrome**

Total Patients	Male(%)	Female(%)
29	45%	55.2%

**Table 4: Laterality of Duane's Retraction Syndrome**

Total Patients	Right Eye%	Left Eye%	Bilateral %
29	37.4%	48.3%	13.8%

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