

## CASE REPORT

PROF-1598

## FAMILIAL DUANE'S SYNDROME

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**ABSTRACT...Objectives:** To describe a family suffering from Duane's Syndrome. **Study Design:** Case series studies. **Setting:** Diagnosis and Research department of Ophthalmology, Allied Hospital, PMC, Faisalabad. **Period:** Oct 2009 (One month). **Presentation:** A Gentle man, 65 years old, shopkeeper, resident of Jaranwala, Faisalabad, presented with his four children two male and two female with complaint of asymmetry of their unilateral eyes, since birth. There was no history of maternal intake of drugs, or smoking during pregnancy. There was no history of trauma or surgery. On examination the unaided vision of all the study cases was 6/6, 6/6. The cornea, conjunctiva, lens, iris, pupil, vitreous and retina of all the subjects was normal. The IOP of all was also within normal range. The ocular motility and lid fissure narrowing was noted in different ocular gazes. The lymph nodes and Thyroid gland examination was done. The Hiss screen test was done to quantify individual muscle action. The Levator function test, cover tests, papillary reactions, Slit Lamp Examination, Fundus examination were performed. The lids fatigue ability tests and strength of hands grip was estimated at the last. All cranial nerves were examined. Urine, Blood and Thyroid tests were requested and documented. **Results;** The all the cases had lid fissure narrowing in one of their horizontal gazes. The all of them had under action of one of their horizontal muscles. So all the children had been proved to be suffering from unilateral Familial Duane's Syndrome. In our clinical practice the Familial DS is rarely found. None of the study patients showed amblyopia, diplopia, upshots or down shoots phenomenon. Similarly no associated systemic anomalies were seen.

**Key words:** MR= Medial rectus, LR= lateral rectus, IR= inferior rectus, SR= superior rectus, IO= inferior oblique, SO= superior oblique, IOP= intra ocular pressure, DS= Duane's syndrome, FDT= forced duction test, SLE=Slit lamp examination, EOM= extra ocular muscles, CNS= central nervous system, RA= rheumatoid arthritis.

## INTRODUCTION

The Duane's syndrome is a congenital condition caused by Congenital hypoplasia of the 6th nerve nucleus with misdirection of the medial rectus nerve (3<sup>rd</sup> nerve), splitting to innervate both the medial rectus and the lateral rectus muscles. It is usually unilateral or rarely bilateral.

As the medial rectus and lateral rectus both are innervated by the nerve to the medial rectus, so both muscles fire and contract simultaneously on the attempted adduction. This causes globe retraction with lid fissure narrowing on attempted adduction. On attempted abduction however the lid fissure widens because the lateral rectus is paretic and medial rectus tone is inhibited by the Sherrington's law<sup>1</sup>.

Both genetic and environmental factors are likely to play a role in the development of DS. Most cases of DS are sporadic with approximately 2-5% of patients having a

familial pattern. Both dominant and recessive forms of DS have been documented<sup>2</sup>.

The duane's Syndrome is rarely associated with amblyopia, diplopia, face or neck turn and binocular deficiency. Therefore it is not treated aggressively.

Indications for surgery have been mentioned as;

- I. Loss of binocular single vision
- II. Unacceptable up or down shoots
- III. Significant face turn

If DS is type 1 and is associated with an esotropia in primary position and face turn towards the duane's eye

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,then it requires ipsilateral MR. recession about 6 to 6.5 mm.

If DS is type 3 and is associated with an esotropia, face turn away from the Duane,s eye, then LR. recession should be performed. If these patients show up or down shoots then ,Y, Splitting of the LR in addition to recession should be performed<sup>3</sup>.

Depending on the clinical presentation<sup>4</sup> and electromyographic<sup>5</sup> studies

The Classification of Duane,s Syndrome is as under;

### 1. Type 1

Commonest: (70-80%);

- Limited or absent abduction
- Normal adduction
- In primary position straight eyes or slight esotropia.
- A or V pattern

### 2. Type 2

Least common (7%);

- Limited or absent adduction.
- Normal abduction
- Primary position straight eyes or slightly exotropia.

### 3. Type 3

(15%)

- Limited adduction and abduction
- In primary position straight eyes or slight esotropia.

### 4. Type 4

Is called ,Unusual Syndrome or DS type appearance. It is also called Synergistic divergence. On attempted adduction the Duane,s eye paradoxically abducts because nerve to MR supplies more fibers to LR.

It is further subdivided in to 3 subtypes A, B, and C

- A, affected eye is esotropic.
- B, affected eye is exotropic

- C, eyes are straight in primary position

Mostly DS patients are unilateral (80%). The left eye is more often affected (60-70%)<sup>6</sup>.

There are other conditions that may be associated with DS<sup>7</sup>.

- Skeleton
- Ears
- Eyes
- Kidneys
- CNS
- Heart Defects
- Morning glory syndrome
- Golden har Syndrome

**The other signs of DS may include as follows<sup>8</sup>.**

- I. Head turn (66%)
- II. Strabismus (77%)
- III. Globe retraction
- IV. Palpebral fissure narrowing
- V. Anisometropia and amblyopia
- VI. Heterochromia
- VII. Iris dysplasia
- VIII. Ptosis
- IX. Nystagmus
- X. Choroidal coloboma
- XI. Optic nerve hypoplasia

## METHODS

A through family and birth history was enquired. The history of trauma and surgery was taken. Ocular examination was conducted with special attention to the other ocular and systematic malformations. The measurements of visual acuity, ocular misalignment, ocular motility, head turn, globe retraction, size of the palpebral fissure in different gazes, up and down shoots phenomenon were noted. Examination of conjunctiva, iris, pupil, lens, anterior vitreous and binocular fundus examination was done. Forced abduction test and Herschberg tests, cover, uncover and alternate cover tests were done. Refraction of all patients was done. IOP was determined by goldmann applanation tonometer. The Lid crease, Marcus gun Jaw winking phenomena

was noted. The Pupillary reactions were also elicited and noted. The lids fatigue ability and thyroid gland functions were done and noted. Hiss screen charting was plotted for all the four study cases. All cranial nerves were examined.

## RESULTS

### Male 1

Left was Duane's eye with

- Limited abduction
- Normal abduction
- In Primary position, no esotropia or exotropia.
- No face or head turn
- On Hiss screen charting the lateral rectus was under acting, while all other extra ocular muscles were normal.
- No up or down shoots seen.
- Forced duction test was negative
- (Duane,s type 1 syndrome)

### Male 2

Right, Duane's eye with

- Limited adduction
- Normal abduction
- Normal other motility
- Mild exotropia in primary position.
- On Hiss screen charting the medial rectus was under acting
- No up or down shoots noted
- Forced duction test was negative
- (Duane,s type 2 syndrome)

### Female 1

Right was duane,s eye

Limited Abduction

Normal adduction

No eso/exo tropia in primary position

No face or neck turn

On Hiss charting lateral rectus was under acting.

No up or down shoots noted

Forced duction test was negative

(Duane,s type 1 syndrome)

### Female 2

Right Duane,s eye

Limited abduction

Normal adduction

No other motility deficit

No eso / exo in primary position

On Hiss charting Lateral retus under acting

No up or down shoots noted

Forced duction test was negative

(Duane,s type 1 syndrome)

## DISCUSSION

The DS is a miswiring of the medial and lateral eye muscles or the muscles that move the eyes .Also, the patients with DS lack the abducens nerve which is involved in eye movement<sup>9</sup>. However the etiology and origin of these malformation remains mystery. Many researches believe that DS results from a disturbance due to either genetic factors or environmental factors during embryonic development. Because the cranial nerves and ocular muscles are developing between the third and eighth weeks of pregnancy, this is most likely when the disturbance happens. So DS causes some eye muscles to contract when they should not and others not to contract when they should. Similarly 1895 Bahr<sup>10</sup> (in 1896), Stilling<sup>11</sup> (in 1887), Turk<sup>12</sup> (in 1899) and Wolff<sup>13</sup> (in 1900) first described Duane's retraction syndrome (DRS) in 1905, Dave reported 54 cases with clinical findings, theories, pathogenesis and treatment of the disease.

None of the cases in our study should abnormal head posture, or up and down shoot phenomenon. None of the patients was esotropic while one of male patient was exotropic in primary position. None of the patients showed any associated systemic abnormalities. On the basis of clinical findings and literature review following differential diagnosis was considered;

- Duane's syndrome
- Congenital fibrosis syndrome
- Congenital (6<sup>th</sup> Nerve Palsy) (Mobius syndrome)
- Infantile myasthenia gravis
- Infantile esotropia
- Orbital floor fracture
- Brown syndrome



Table showing involvement of eyes and type of Duane,s Syndrome

Muscle involved	Patients Sex	Eye involved	Adduction	Abduction	Primary positions Exo/Iso	Face/neck turn	Duane's Type
LR	Male	Left	Normal	Abnormal	No	No	1
MR	Male	Right	Abnormal	Normal	Exo	No	2
LR	Female	Right	Normal	Abnormal	No	No	1
LR	Female	Right	Normal	Abnormal	No	No	1

- Double elevator palsy
- Thyroid eye disease

All the patients were emmetropic. The amblyopia was also absent in our study cases while it was present in 21% and anisometropia in 50% of cases of Kirkhan,s<sup>14</sup> study. Maruo and Coworkers<sup>15</sup> found in 3.6% prevalence of amblyopia while tredici<sup>16</sup> and von Noorden<sup>17</sup> reported 3% incidence of amblyopia. Hoyt and Nachtigaller<sup>18</sup> have proposed that ocular retraction during adduction as well as the EMG findings of synergistic innervations of medial and lateral rectus muscles can be explained on the basis of substitute of the paretic lateral rectus muscles by an extra branch of the oculomotor nerve.

Involvement of left eye was seen in only one male patient in our study in contrary to other studies in which left eye was predominantly affected in Duane,s syndrome. Our all cases were proved to be unilateral Duane,s syndrome and this is in well according to studies of Goldstein J H and Sacks D B.(6) who also found predominantly unilateral Duan,s syndrome.

The Duane,s Syndrome 1 was most common in our study as is mentioned in other studies (4).

None of the our cases required any surgical intervention as having no diplopia ,no face turn, no up or down shoots and no defect binocularity.

## CONCLUSION

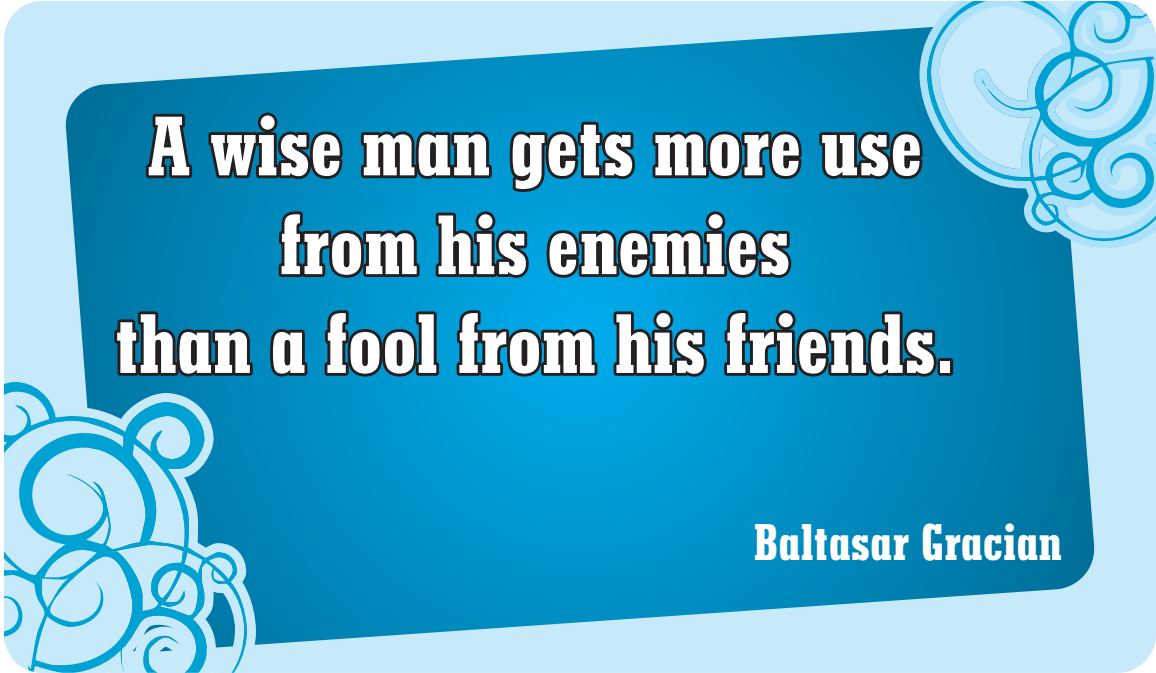
The Familial Duane,s Syndrome is rarely seen in clinical practice. Type 1 is more common than the others forms of the Duane,s Syndrome. Associated systemic abnormalities are not seen. .No surgical intervention was required as none of cases had defect in face position, up or down shoots problems nor defect in binocular potential.

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**A wise man gets more use  
from his enemies  
than a fool from his friends.**

**Baltasar Gracian**