Prevalence of anisometropia and amblyopia in duane retraction syndrome of patients attened ibn Al-Haitham Teaching Eye Hospital

Prevalencia de anisometropía y ambliopía en el síndrome de retracción de duane de pacientes atendidos en el Ibn Al-Haitham Teaching Eye Hospital

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Abstract

Duane's Retraction Syndrome (DRS) is a congenital oculomotor anomaly that presents as globe retraction with simultaneous narrowing of the palpebral fissure on attempted adduction. To determine the prevalence of anisometropia and amblyopia in Duane retraction syndrome in patients referred to tertiary eye center (Ibn Al-Haitham Teaching Eye Hospital squint department). All recorded files of patients attended to Ibn-Al Haitham Teaching Eye Hospital between 2009 and 2015, were retrospectively reviewed, a total number of 5000 files, 120 patients with the diagnosis of Duane retraction syndrome were identified. The prevalence of the Duane retraction syndrome in our study related to strabismus population was 2.4% (95% confidence interval [CI]: 1.4–1.9).Data collection regarding, clinical features , gender, age at diagnosis, laterality and type of Duane retraction syndrome, abnormal head posture, amblyopia, Anisometropia (refractive errors), corrected visual acuity, manifest and cycloplegic refraction, alternate cover test, ductions and version ocular movement, fundus examination, and slit lamp biomicroscopy. Amblyopia is a unilateral or, less commonly, bilateral reduction of best -corrected visual acuity that cannot be attributed directly to the effect of any structural abnormality of the eye or visual pathways. Anisometropia dissimilar refractive errors in the 2 eyes cause the image on one retina to be chronically defocused. Levels of anisometropia that can lead to amblyopia are greater than 1.50 D of hyperopia, 2.00 D of astigmatism, and 3.00 D of myopia. Higher levels are associated with greater risk. 59 males (49.2%) and 61 females (50.8%) presented with age groups, below 2yrs 48 cases (40.0%), 2-5yrs 54 cases (45.0%) and more than 5yrs 18 cases(15.0%). 120 Duane Retraction Syndrome, the unilateral cases were 104 (86.7%) compared to bilateral cases were 16 (13.3%), in unilateral cases, left eye was involved in 90 cases (75.0%) and the right eye 14 cases (11.7%) type I were 108 eyes (90.0%) type II were 7 eyes (5.8%)type III were 3 eyes (2.5%). In bilateral cases type I for right eye &II for left eye was 1 case (0.8%), type I for right eye &III for left eye was 1 case (0.8%), type I for right & left eyes were 14 cases (99.2%). 11 cases with abnormal Head Posture recorded (9.2%). Total number of eyes with higher refractive error (Anisometropia) in Duane retraction syndrome was found in 37 cases, unilateral Duane eyes 29 cases 78.4%and bilateral Duane eyes 8cases 21.6%. Total number of eyes with amblyopia in Duane retraction syndrome were 43 cases, unilateral Duane eye 33 cases 76.75% (29 of them due to anisometropia and 4 cases due to Duane per se) while in bilateral Duane eye 10cases 23.25% (8 of them due anisometropia and 2 cases due to Duane per se). Each of DRS and anisometropia independently causing amblyopia. But Amblyopia due to Anisometropia found to be more significant association.

Keywords: Anisometropia, Amblyopia, Duane Retraction Syndrome

Resumen

El síndrome de retracción de Duane (DRS) es una anomalía oculomotora congénita que se presenta como retracción del globo con estrechamiento simultáneo de la fisura palpebral en el intento de aducción. Determinar la prevalencia de anisometropía y ambliopía en el síndrome de retracción de Duane en pacientes remitidos al centro de atención terciaria del ojo (departamento de estrabismo del Hospital de Enseñanza Ibn Al-Haitham). Todos los archivos registrados de pacientes atendidos en el Ibn-Al Haitham Teaching Eye

Hospital entre 2009 y 2015 se revisaron retrospectivamente, se identificaron un total de 5000 archivos, 120 pacientes con el diagnóstico de síndrome de retracción de Duane. La prevalencia del síndrome de retracción de Duane en nuestro estudio relacionado con la población con estrabismo fue del 2,4% (intervalo de confianza [IC] del 95%: 1,4–1,9). Recopilación de datos, características clínicas, sexo, edad en el momento del diagnóstico, lateralidad y tipo de síndrome de retracción de Duane, postura anormal de la cabeza, ambliopía, anisometropía (errores refractivos), agudeza visual corregida, refracción manifiesta y cicloplejica, prueba de cobertura alternativa, deducciones y versión ocular Movimiento, examen del fondo de ojo, y biomicroscopia con lámpara de hendidura. La ambliopía es una reducción unilateral o, menos frecuentemente, bilateral de la mejor agudeza visual corregida que no se puede atribuir directamente al efecto de cualquier anomalía estructural del ojo o vías visuales. Los errores refractivos disímiles de la anisometropía en los 2 ojos hacen que la imagen en una retina esté desenfocada crónicamente. Los niveles de anisometropía que pueden llevar a la ambliopía son mayores que 1.50 D de hipermetropía, 2.00 D de astigmatismo y 3.00 D de miopía. Los niveles más altos están asociados con un mayor riesgo. 59 hombres (49.2%) y 61 mujeres (50.8%) presentaron grupos de edad, menores de 2 años, 48 casos (40.0%), 2-5 años, 54 casos (45.0%) y más de 5 años, 18 casos (15.0%). 120 Síndrome de retracción de Duane, los casos unilaterales fueron 104 (86.7%) en comparación con los casos bilaterales 16 (13.3%), en casos unilaterales, el ojo izquierdo estuvo involucrado en 90 casos (75.0%) y el ojo derecho 14 casos (11.7%) tipo I fueron 108 ojos (90.0%) tipo II fueron 7 ojos (5.8%) tipo III fueron 3 ojos (2.5%). En los casos bilaterales, el tipo I para el ojo derecho y II para el ojo izquierdo fue de 1 caso (0,8%), el tipo I para el ojo derecho y III para el ojo izquierdo fue de 1 caso (0,8%), el tipo I para los ojos derecho e izquierdo fue de 14 casos (99,2%).11 casos con Postura de Cabeza anormal registrada (9.2%). El número total de ojos con mayor error refractivo (anisometropía) en el síndrome de retracción de Duane se encontró en 37 casos, los ojos unilaterales de Duane 29 casos de 78,4% y los ojos bilaterales de Duane de 8 casos en el 21,6%. El número total de ojos con ambliopía en el síndrome de retracción de Duane fue de 43 casos. Ojo de Duane unilateral 33 casos 76.75% (29 de ellos debido a anisometropía y 4 casos debido a Duane per se), mientras que en ojo de Duane bilateral 10 casos 23.25% (8 de ellos debido a anisometropía y 2 casos debido a Duane per se). Cada uno de DRS y anisometropía causan independientemente ambliopía. Pero la ambliopía debida a la anisometropía resultó ser una asociación más significativa.

Palabras clave: Anisometropía, Ambliopía, Síndrome de Retracción de Duane

Introduction

Duane's Retraction Syndrome (DRS), also known as the Stilling-Turk-Duane syndrome, is a congenital oculomotor anomaly that presents as globe retraction with simultaneous narrowing of the palpebral fissure on attempted adduction. It is caused by an absence or hypoplasia of both the abducens nucleus and the nerve with an anomalous innervation of its target, the lateral rectus muscle, by a branch of the oculomotor nerve[1,18].

The incidence of DRS is approximately 1% of the total cases of strabismus. It commonly occurs unilaterally and sporadically, with a predominant tendency to affect females and the left eye. 10% of the Duane syndrome cases are inherited in an autosomal dominant pattern and are usually bilateral. Disorders which are similar in presentation to the Duane syndrome can be acquired as a result of trauma, or following a localized infection of the orbit, leading to inflammation and consequent mechanical restrictions of the eye movement. A complete case history will usually help in distinguishing between these conditions. Sixth nerve paralysis, a tight medial rectus, crossed fixation, pseudo duanes or inverse duanes are some of the other differential diagnosis for this condition. The primary lesion in pseudo-Duane's retraction syndrome is suspected to be due to entrapment of the medial rectus muscle within the medial orbital wall due to trauma[2,19].

Classified DRS into the three types: Duane 1, Duane 2, and Duane 3. Type 1 is characterized by marked limitation or complete absence of abduction with associated esotropia, and is the most common type. Type 2 is characterized by marked limitation of adduction and exotropia of the involved eye, and is the least common; while type 3 is characterized by marked limitation of both adduction and abduction and is often associated with a straight eye in primary position of gaze.

The aetiopathogenesis of DRS has been extensively studied. Huber[3] suggested, based on electromyography (EMG) findings, that a paradoxical anomalous innervation of the lateral rectus muscle from the oculomotor (third cranial nerve) nucleus is the underlying cause in all three types of DRS. This causes the lateral rectus muscle to co-contract with the medial rectus on attempted adduction with resultant retraction of the globe and narrowing of the palpebral aperture[3].

Duane retraction syndrome (DRS) patients can be orthotropic, esotropic, or exotropic in the primary position. Indications for surgery include ocular misalignment in the primary position, noticeable abnormal head position, severe globe retraction in side gaze, and abnormal vertical movements[4]. Surgical management of esotropic DRS includes options such as medial rectus muscle recession[5] or

vertical rectus transposition[6]. Amblyopia is a unilateral or, less commonly, bilateral reduction of best -corrected visual acuity (also referred to as corrected distance visual acuity) that cannot be attributed directly to the effect of any structural abnormality of the eye or visual pathways. Amblyopia signifies a failure of normal neural development in the immature visual system and is caused by abnormal visual experience early in life resulting from one of the following:

• strabismus

• refractive error: anisometropia or high bilateral refractive errors (isoametropia)

• visual deprivation

Anisometropia dissimilar refractive errors in the 2 eyes cause the image on 1 retina to be chronically defocused. Considered more prevalent than strabismic amblyopia in some recent studies, this condition is thought to result partly from the direct effect of image blur and partly from interocular competition or inhibition similar (but not identical) to that responsible for strabismic amblyopia. Levels of anisometropia that can lead to amblyopia are greater than 1.50 D of anisohyperopia, 2.00 D of anisoastigmatism, and 3.00 D of anisomyopia. Higher levels are associated with greater risk. DRS can be associated with refractive errors. In particular, hyperopia is present in 57-82% of patients[7,8,9] High hyperopia, defined as a refractive error greater than 4 D is present in less than 25% of patients with DRS[7]

Patients and methods

All files of patients attended Ibn-Al Haitham Teaching Eye Hospital squint department between 2009 and 2015, were retrospectively reviewed. Total of 5000 files were included, 120 patients with the diagnosis of DRS were identified.

Hubers classification was used for classifying clinical types of DRS. Amblyopia defined as decrease best corrected visual acuity 6/9 or less in either eyes Anisometropia defined as difference in cycloplegic spherical equivalent greater than 2.0 dioptor(D) between two eyes.

Data regarding gender, age at diagnosis, laterality and type of DRS, abnormal head posture, amblyopia, anisometropia, and refractive errors, recorded in all cases, including patients complaints, past history medical and surgical, best corrected visual acuity, manifest and cycloplegic refraction, alternate cover test, duction and version ocular movement, fundus examination ,and slit lamp bio microscopy. Positive family history taken in consideration.

In unilateral DRS, anisometropic eye reffered to an eye with higher refractive errors, whether it is Duane eye or non Duane eye or (normal eye).

In bilateral DRS, anisometropic eye reffered to an eye with higher refrective errors.

Exclusion Criteria: ptosis, facial anomalies, ocular trauma, media opacities, history of orbital fracture, less informative files, and child with cerebral palsy.

Statistical Analysis: Database was first entered into an excel file then transferred for analysis on Statistical Package for Social Sciences (SPSS) file (version 22).

Data presented as numbers and percentages. Chi-square test for independence was used to test the significance of observed findings.

Adjusted effect of each Duane disorder and anisometropia upon developing amblyopia studied with Odd's Ratio using binary logistic regression analysis which is a type of multivariate analysis.

Findings with P value less than 0.05 was considered to be significant.

Results

The prevalence of the DRS in our study related to strabismus population was 2.4% (95% confidence interval [CI]: 1.4–1.9). 59 males (49.2%) and 61 females (50.8%) presented with age groups, below 2yrs 48 cases (40.0%), 2-5yrs 54 cases (45.0%) and more than 5yrs 18 cases (15.0%). 120 Duane Retraction Syndrome, the unilateral cases were 104 (86.7%) compared to bilateral cases were 16 (13.3%), in unilateral cases, left eye was involved in 90 cases (75.0%) and the right eye 14 cases (11.7%) type I were 108 eyes (90.0%) type II were 7 eyes (5.8%) type III were 3 eyes (2.5%). In bilateral cases type I for right eye &II for left eye was 1 case (0.8%) type I for right eye & III for left eye was 1 case (0.8%), type I for right & left eyes were 14cases (99.2%). 11 cases with abnormal head Posture recorded (9.2%). 109 of DRS 90.8% with no abnormal head posture, six of them were found to have strabismic ambyliopia due to esotropia. (Table 1)

Table 1.Characteristics of sampled DRS patients

Variables	N=120	100.0%
Gender		
• Male	59	49.2%
• Female	61	50.8%
Age Group		
• below 2 y	48	40.0%
• 2.1- 5 y	54	45.0%
• > 5 y	18	15.0%
Laterality of Duane		
• Unilateral	104	86.7%
• Bilateral*	16	13.3%
Duane Eye		
• Right Eye	14	11.7%
• Left Eye	90	75.0%
• Both	16	13.3%
Type of DRS		
• I	108	90.0%
• II	7	5.8%
• III	3	2.5%
• I for Rt eye & II for Lt eye	1	0.8%
• I for Rt eye & III for Lt eye	1	0.8%
Abnormal Head Posture		
• Yes	11	9.2%
• No	109	90.8%

*All eyes in bilateral cases are type I DRS for right and left eyes except two cases (right eye is type I and the left is type II on one case and type III in the other case).

Table 2.Characteristics of unilateral DRS according to the side of the eye

	Right Eye		Left Eye		
Variables	**N=104**	**100.0%**	**N=104**	**100.0%**	**P value**
Duane Eye					**< 0.001**
• Duane Eye	14	25.0%	90	88.3%	
• Non Duane Eye	90	75.0%	14	11.7%	

Duane disorder is significantly associated with left eyes (P < 0.05, table 2)

Table 3. Characteristics of unilateral DRS according to the Anisometropia and Amblyopia

	Duane Eye		Non Duane Eye		
Variables	**N=104**	**100.0%**	**N=104**	**100.0%**	**P value**
Anisometropia					**< 0.001**
• Yes	29	27.9%	7	6.7%	
• No	75	72.1%	97	93.3%	
Amblyopia					**< 0.001**
	33	31.7%	8	7.7%	
• No	71	68.3%	96	92.3%	

Duane status is significantly associated with anisometropia & amblyopia of the Duane eye (P < 0.05, table 3)

Table 4.Distribution of Duane eyes according to presence of amblyopia and anisometropia

Anisometropia	Amblyopia				Total		
	Yes		No				
	N	%	N	%	N	%	P value
• Yes	**29**	**87.8%**	0	0%	29	87.8%	**< 0.001**
• No	4	12.2%	71	100%	75	12.2%	
Total	*33*	*100.0%*	*71*	*100.0%*	***104***	*100.0%*	

Table 5. Prevalence of anisometropia and amblyopia in bilateral DRS patients

Variables	N=16	100.0%
Anisometropia		
• Yes	8	50%
Amblyopia		
• Yes	10	62.5%

Table 6. Prevalence of amblyopia and anisometropia related to laterality of DRS

	Amblyopia		Anisometropia	
Variables	**N=43**	**100.0%**	**N=37**	
Laterality of Duane				
• Unilateral	33	76.75%	29	78.4%
• Bilateral	10	23.25%	8	21.6%

The probability to develop amblyopia in presence of DRS is around 5 times if compared to cases not having DRS (table 7).

The probability to develop amblyopia in presence of anisometropia is around 27 times compared to cases not having anisometropia (table 7).

Each of DRS and anisometropia independently causing amblyopia, though in variable strength of association (five times for DRS and 27 times for anisometropia)

Table 7.Odd's Ratios for Duane and anisometropia for developing amblyopia

		Odd's Ratio		
Factor Under Study	**P value**	**Value**	**95% CI**	
			Lower	**Upper**
Duane Disorder[1]	**0.002**	5.076	1.795	14.354
Anisometropia[2]	**< 0.001**	27.206	11.221	65.961

[1]backgroupnd level is no Duane

[2]Background level is no Anisometropia

Discussion

In this study, the prevalence of DRS among the strabismus population (2.4%) which is comparable to the previously reported studies[10–13].

The age at presentation at 2-5 years was 45% of our patients, while Chau et al.[13] have stated that 85% of their cases had been identified by the age of 12 months. This because difference in educational level, positive family history, availability of health care facilities and socioeconomic levels.

There is no gender predilection, male 49.2% and female 50.8% which is opposite to most of previous studies said female most common[10,11-13].

This study found, DRS mostly unilateral (86.7%), left eye 75.0%.while type I 90% the commonest and it is comparable to other studies[10-12], type III 2.5% least common and was also similar to most previously reported studies[10].

In previous studies, amblyopia has been attributed to strabismus[10,14-17], and still the anisometropia major amblyopiogenic factor[10]. In our study, total number of amblyopia in DRS were 43 cases, unilateral Duane eye 33 cases 76.75% (29 of them anisometropia and 4 cases due to Duane per se) [table 4 & 6] while in bilateral Duane eye 10cases 23.25% (8 of them anisometropia and 2 cases due to Duane per se) [table 5 & 6]. Amblyopia is considered to be relatively uncommon in DRS by Von Noorden and Tredic[15]. In our study, odds ratio shows the probability to develop amblyopia in presence of DRS is around 5 times [table 7], but the probability to develop amblyopia in presence of anisometropia is around 27 times [table 7]. So amblyopia is significantly associated with DRS in presence of anisometropia [P <0.05] [table 4]

The percentage of anisometropia varies from 14% to 40% in previous studies[10]. In our study,total number of anisometropia in DRS were 37 cases, unilateral Duane eyes 29 cases 78.4% [table 4 & 6] and bilateral Duane eyes 8 cases 21.6% [table 5 & 6] .The hypermetropia was the most common refractive errors in all cases, which is significantly associated with each of Duane and amblyopia of the same eye (P <0.05) [table 4]

Tredici and Von Noorden[15], explained that it is the fibrotic lateral rectus muscle which causes the anisometropia, this could be due to anatomical fact, that the lateral rectus has largest contact arm with the globe, so fibrosis cause hardness and distortion of the globe that may affect the axial length and refractive power of the eye which lead to anisometropia.

Abnormal head posture in primary position is major determinants of the cosmetic and/or functional status of DRS cases, but in our study was 9.2% of all DRS cases, which was less than that reported by Anvari et al.[7] this discrepancy could, at least partly, be attributed to the higher degree of deviation in Chung et al.[13] AHP was more common in patients with unilateral DRS than bilateral DRS, which can be expected on the basis of both ocular motility and ocular alignment pattern.

The limitations in this study was short time for data collection, DRS files where not categorized as separate category were we depend on manual search for these files.

Conclusions

Each of DRS and anisometropia independently causing amblyopia. But amblyopia due to anisometropia found to be more significant association.

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