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Study of clinico-etiological profile of patients with paralytic and restrictive strabismus

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ABSTRACT

Introduction: Incomitant strabismus typically results in intractable diplopia, blepharoptosis, ocular motility disturbance, disturbance in pupil reaction and an unacceptable compensatory head or face posture. A complete clinico etiological profile is necessary for identifying the underlying risk factors and etiology pertaining to paralytic and restrictive strabismus which is imperative in prevention, early detection, managing the etiology and timely recovery in these patients.

Objective: To study the clinico-etiological profile of patients with paralytic and restrictive strabismus in a tertiary eye hospital.

Material and Methods: Patients reporting to a tertiary eye hospital in north India and diagnosed with paralytic and restrictive strabismus, over a period of one year , were enrolled in the present study . Patients were subjected to detailed ocular examination and evaluated for ocular motility or related disorders. This included detailed clinical history, ophthalmic examination, orthoptic workup , sensory evaluation, diplopia charting, Hess charting, slit lamp examination and fundus examination . In patients of suspected restricted strabismus, forced duction test was performed. Relevant systemic investigations including blood pressure , blood sugar and lipid profile were carried out . M.R.I orbit was done in every patient. Neuroimaging was advised in those patients where indicated.

Results: 48 patients were diagnosed with incomitant strabismus and enrolled in the study. 81.3%were of paralytic strabismus while 18.8 % were of restrictive strabismus. In the group of paralytic strabismus, 43.8 % patients had sixth nerve palsy , 27.1% patients had third nerve palsy , 6.3% patients had fourth nerve palsy and combined nerve palsy patients were 4.2%. Analysing the etiology, out of total patients of paralytic strabismus, 48.7% of cases were idiopathic, 25.6% were post traumatic, 15.4% were due to microvascular causes , 5.1% of cases were due to intracranial anomalies and rest 5.1% were due to inflammatory causes. In restrictive strabismus group, 66.7% were congenital and rest 33.3% were acquired.

Conclusion: Our study has reflected the prevalence, distribution, etiological and clinical profile of patients presenting with incomitant strabismus. It is helpful for ophthalmologists and other physicians in guiding diagnosis and evaluation and is useful for proper and timely intervention and timely recovery in these patients.

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1. Introduction

Strabismus is a frequent cause of ocular morbidity worldwide including India.

Population studies mostly from Western European and North American population suggest the mean prevalence of strabismus between 2% and 5% with predominance of

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esodeviation .However, Asian data suggests slightly lower prevalence of strabismus and predominance of exodeviation in the population. ^{1,2}

Worldwide, the prevalence of strabismus in children has been reported to vary from 1.3% to 5.7%, although a study from southern India has shown a lower prevalence of 0.7%. 3,4

In the past, strabismus has not been in spotlight of India's national program for control of blindness but now,

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after getting recognized as contributing factor in vision loss and impaired binocular function in children, it has been given due attention, in the recent thrust on paediatric ophthalmology in the Vision 2020 initiative.⁵

Strabismus causes grave ocular dysfunction including double vision, eyestrain, vision loss, poor depth perception as well as cosmetic stigma among patients. Psychosocial difficulties relating to socially noticeable strabismus persist into teenage and adult years. ⁶

Incomitant strabismus typically results in intractable diplopia, blepharoptosis, ocular motility disturbance, disturbance in pupil reaction and an unacceptable compensatory head or face posture.

The etiology of ocular motor nerve palsies remain unknown in more than 25% cases. Etiology varies in different types of paralytic strabismus. Numerous studies have shown that ocular motor cranial nerve palsies have a variety of causes including vascular disease, head trauma, intracranial tumor or aneurysm and inflammatory disorders. ^{7,8}

Rucker and Young have reported third cranial nerve palsy as the second most prevalent ocular motor palsy following sixth cranial nerve palsy. ⁸

For better management of incomitant strabismus, a complete clinico etiological profile is necessary. This helps in identifying the underlying risk factors and etiology pertaining to paralytic and restrictive strabismus which is imperative in prevention, early detection, managing the etiology and timely recovery in these patients.

The clinical presentation of paralytic and restrictive strabismus can be varied. The literature abounds in studies focusing on paralytic strabismus, many of which have discussed diagnostic imaging techniques and management.

The current prospective study was conducted on patients of paralytic and restrictive strabismus attending paediatric ophthalmology and strabismus clinic in a tertiary eye hospital in North India to better understand the incidence, etiology ,clinical characteristics and risk factors in these patients.

2. Materials and Methods

The present prospective and analytical study was conducted on patients presenting to the paediatric ophthalmology and strabismus clinic in a tertiary eye hospital in north India over a period of one year. All patients diagnosed with paralytic and restrictive strabismus with limitation of ocular motility were enrolled in the study.

Patients with comitant strabismus, A and V pattern, myasthenia gravis and previous strabismus surgery were excluded from the study. Critically ill and mentally unstable patients were also excluded.

Patients were subjected to a detailed ocular examination and evaluated for ocular motility or related disorders. This included a detailed clinical history comprising of age of onset, duration and progression of symptoms (like diplopia, headache with vomiting, vertigo and asthenopia), history of trauma, systemic illness (like hypertension, diabetes mellitus, hypercholesterolemia), cigarette smoking, any previous treatment taken and family history, along with a detailed systemic examination including neurological examination, for sensory and motor deficits. The patients underwent ophthalmic examination which included best corrected visual acuity, age appropriate cycloplegic refraction, fixation pattern, ocular motility (version and duction) and orthoptic evaluation. Orthoptic workup included cover test, alternate cover- uncover test both at distance(6m) and at near (33cm), Krimsky test(for patients with low visual acuity) ,prism bar cover test (PBCT) in all cardinal positions and head tilt for measurement of ocular deviation. In patients of suspected restricted strabismus, forced duction test was performed. In sensory evaluation, Worth Four Dot test (WFDT) was used for determining distance and near stereopsis while subjective torsion was evaluated by Double Maddox rod test and objective torsion with indirect ophthalmoscopy.

Presence or absence of nystagmus and amblyopia was looked for. Diplopia charting, Hess charting, slit lamp examination and fundus examination were also done in each patient.

Relevant systemic investigations included blood pressure recording, blood sugar level and lipid profile . M.R.I orbit was done in every patient. Neuroimaging was advised in those patients where indicated .

3. Results and Observation

A total of 300 strabismus patients visited the paediatric ophthalmology and strabismus clinic of our eye institute during the year of study. Of these, 48 patients were diagnosed with incomitant strabismus and enrolled in the study.

The mean age of the study population was 33.4 +/- 17.6 years out of which the mean age of patients in paralytic strabismus group was 37.3+/-16.3 years with majority being in the age group of 31-45 years(33.3%). The mean age of patients in restrictive strabismus group was 16.6 +/- 13.4 years with the majority being in age group of 1-15 years (55.6%). No patients of restrictive strabismus belonged to age group 31 to 45 years. The ratio of male: female patients in study population was 2:1.[Figure 1]

Out of all the patients enrolled in the study, only 39.6% presented with a precipitating cause. The most common precipitating cause was trauma which was seen in 31%, fever in 6% inflammation in 2% and rest had no precipitating event.

In the total study population, neurological features were seen in 19 patients (39.6%) of the patients while they were absent in 29 patients (60.4%) patients. The presenting complaints among all subjects including paralytic and

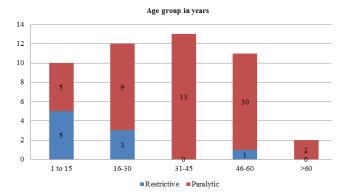


Fig. 1: Total distribution of cases according to age group in years

restrictive was diplopia in 52.1% patients , ptosis in 20.8% and 27.1% patients presented with deviation of eyes.[Figure 2]

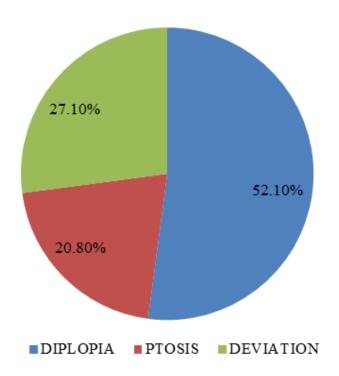


Fig. 2: Presenting complaints in the study population

Compensatory head posture was seen in 64.7% cases with chin lift in 14.6%, face turn in 43.8% and head tilt in 6.3%. Hence, most frequent head posture was face turn.

Out of 48 subjects enrolled in study, 39 cases (81.3%) were of paralytic strabismus while 9 cases (18.8%) were of restrictive strabismus.

Clinically in this group, 43.8 % patients had sixth nerve palsy, 27.1% patients had third nerve palsy, 6.3% patients had fourth nerve palsy and patients of combined nerve palsy were 4.2%. [Table 1]

Analysing the etiology, out of total patients of paralytic strabismus, 48.7% of cases were idiopathic, 25.6% were post traumatic, 15.4% were due to microvascular causes, 5.1% of cases were due to intracranial anomalies and rest 5.1% were due to inflammatory causes.[Table 2], [Figure 3]

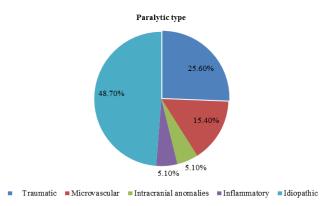


Fig. 3: Paralytic etiology among the patients

Out of 13 patients of third nerve palsy, idiopathic accounted for 38.5% of cases, 38.5% were post traumatic, 15.4% were microvascular causes and inflammatory causes accounted for another 7.7% cases.[Table 3]

Out of total patients of paralytic strabismus, fourth nerve palsy was seen in 6.3% of cases of which, traumatic were 66.7% while idiopathic accounted for 33.3% of cases.[Table 4]

Out of 48 patients of paralytic strabismus sixth nerve palsy was seen in 21 cases, idiopathic accounted for 52.4% of cases, 14.3% were post traumatic, 19% were due to microvascular causes, intracranial anomalies accounted for 9.5% and inflammatory causes were involved in another 5.1%.[Table 5]

On analysis of restrictive strabismus, 66.7% were congenital (majorly Duane's retraction syndrome and fibrosis of extrocular muscles) and rest 33.3% were acquired (mainly blow out fracture, inferior rectus avulsion, acquired case of MED due to inferior rectus fibrosis). The ratio of male: female patients of restrictive strabismus is 2:1.[Figure 4]

4. Discussion

The restrictive and paralytic strabismus cause more ocular morbidity as compared to comitant strabismus because of their various associations and sequelae. Our study was aimed to elicit the clinic-etiological profile of all patients of paralytic and restrictive strabismus reporting to a tertiary care hospital. A total of 300 strabismus patients were seen over one year period. Of these, 48 patients were diagnosed with incomitant strabismus and enrolled in the study and underwent a detailed evaluation for ocular motility and/or related systemic disorders.

Table 1: Distribution of nerve palsies in Paralytic Strabismus

Types of paralytic strabismus			Se	Total	
			\mathbf{F}	M	Total
	Third nerve palsy	Number of cases	5	8	13
		% of cases	20.0%	9.1%	33.3%
	Fourth nerve palsy	Number of cases	2	1	3
Diamosis		% of cases	13.3%	3.0%	7.7%
Diagnosis	Sixth nerve palsy	Number of cases	5	16	21
		% of cases	33.3%	48.5%	53.8%
	Combined nerve	Number of cases	0	2	2
	palsy	% of cases	0.0%	6.1%	5.1%

Table 2: Paralytic etiology among the patients:

			SEX		Total
			F	M	
	Traumatic	Number of cases	3	7	10
		% of cases	25.0%	25.9%	25.6%
	Microvascular	Number of cases	2	4	6
		% of cases	16.7%	14.8%	15.4%
Donalystia tyma	Intracranial anomalies	Number of cases	1	1	2
Paralytic type		% of cases	8.3%	3.7%	5.1%
	Inflammatory	Number of cases	0	2	2
		% of cases	0.0%	7.4%	5.1%
	Idiopathic	Number of cases	6	13	19
		% of cases	50.0%	48.1%	48.7%
Total		Count	12	27	39
10141		% within SEX	100.0%	100.0%	100.0%
P value		0.868			

Table 3: Etiology of third nerve palsy in paralytic strabismus

			Gender		m . 1	
			\mathbf{F}	\mathbf{M}		Total
	Traumatic	Number of cases	2	3	5	
		% of cases	40.0%	37.5%	38.5%	
Etiology Total	Microvascular	Number of cases	1	1	2	
		% of cases	20.0%	12.5%	15.4%	
	Inflammatory	Number of cases	0	1	1	
		% of cases	0.0%	12.5%	7.7%	
	Idiopathic	Number of cases	2	3	5	
	Idiopatific	% of cases	40.0%	37.5%	38.5%	
		Number of cases	5	8	13	
		% of cases	100.0%	100.0%	100.0%	
	P value	0.862				

Table 4: Etiology of fourth nerve palsy in paralytic strabismus

			Gen	Total		
			\mathbf{F}	M	Total	
Etiology	Traumatic	Number of cases	1	1	2	
		% of cases	50.0%	100.0%	66.7%	
	Idiopathic	Number of cases	1	0	1	
		% of cases	50.0%	0.0%	33.3%	
Total		Number of cases	2	1	3	
		% of cases	100.0%	100.0%	100.0%	
	P value	1.000				

Table 5: Etiology of sixth nerve palsy in paralytic strabismus

			G	Total	,		
			\mathbf{F}	M	10tai	iotai	
	Traumatic	Number of cases	0	3	3		
	Traumatic	% of cases	0.0%	18.8%	14.3%		
Etiology	Microvascular	Number of cases	1	3	4		
		% of cases	20.0%	18.8%	19.0%		
	Intracranial anomalies	Number of cases	1	1	2		
		% of cases	20.0%	6.3%	9.5%		
	I., A.,	Number of cases	0	1	1		
	Inflammatory	% of cases	0.0%	6.3%	4.8%		
Total	Idiamathia	Number of cases	3	8	11		
	Idiopathic	% of cases	60.0%	50.0%	52.4%		
		Number of cases	5	16	21		
		% of cases	100.0%	100.0%	100.0%		
P value		0.721					

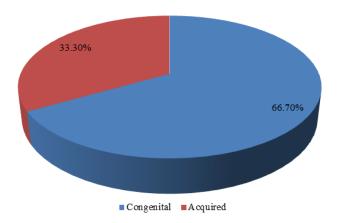


Fig. 4: Restrictive paralysis etiology

The mean age of the study population was 33.4 +/- 17.6 years. The mean age of patients in paralytic strabismus group was 37.3+/-16.3 years with majority being in the age group of 31-45 years(33.3%). The mean age of patients in restrictive strabismus group was 16.6 +/- 13.4 years with the majority being in age group of 1-15 years (55.6%). In a study conducted by Jung et al in 2015,the mean age of patients was 64.5 years. This could be due to the study being conducted on ischemic etiology of cranial nerve palsies which have a higher incidence in middle and late age groups. 9

In a study conducted by Kim et al in 2018, on patients with acquired cranial nerve palsies ,mean age was 59.8+/-14.5 years which can be explained due to different referral pattern in their institute(frequent referral of patients with vasculopathic causes in that institute, which are mainly seen in 4-5 decades of life). ¹⁰

In our study the higher incidence of paralytic and restrictive strabismus in second and third decade can be explained by the higher incidence of trauma in young population, our institute being tertiary level eye care referral

institute.

Out of 48 subjects enrolled in our study, 39 cases (81.3%) were of paralytic strabismus while 9 cases (18.8%) were of restrictive strabismus. Out of total 39 patients of paralytic strabismus, 27 (69.2%) were males and 12 (30.8%) were females. The ratio of male: female patients in our study was 2:1. This can be justified by higher incidence of trauma in males, who are involved more in field work and road side accidents. Also, the incidence of microvascular accidents is more in males due to higher prevalence of cigarette smoking and alcohol intake in male population.

Similar results were found in a study conducted by Richards et al in 1992, on patients of cranial nerve palsies . In their study on 1278 patients of paralytic strabismus 694 were males and 584 were females. Similar results were reported in two separate studies conducted by Bagheri et al in 2010 and 2014 on patients of paralytic strabismus with a male preponderance. Similarly, Kim et al 2018 conducted a study which comprised of total 153 patients of new onset paralytic strabismus. There were 87 (56.86%) male and 66 (44.14%) female. ^{11–14}

In our study, out of the total patients of paralytic strabismus (81.3%), sixth nerve palsy was seen in 43.8% patients, third nerve palsy in 27.1%, fourth nerve palsy in 6.3% and combined nerve palsy in 4.2%. Hence, out of all paralytic strabismus patients, sixth nerve palsy (43.8%) was the most common, followed by third nerve involvement (27.1%). This can be explained due to long intradural course of sixth nerve making it particularly vulnerable to head trauma/idiopathic cause.

The distribution pattern of various cranial nerve palsies in our study, with sixth nerve palsy being the commonest, is consistent with majority of previous large studies which also reported that sixth nerve palsy had higher proportion of traumatic causes as compared to third and fourth cranial nerves. Rucker C W, in 1966 reported 515 cases of sixth nerve palsy and 274 cases of isolated third nerve palsy, out of a total of 1,000 cases of paralytic strabismus .Study conducted by Richards et al in 1992 also a had similar outcome. In their study sixth nerve was the most affected followed by third nerve while combination palsies were least common. Out of 4,278 cases of paralytic strabismus,1918 were sixth nerve palsy, 1225 were third nerve palsy, 657 were fourth nerve palsy and multiple nerve palsy were 573. Hence results are similar to our study.Study conducted by Tiffin et al in 1996,had total 165 patients of paralytic strabismus, out of which sixth nerve palsy were 93(57%), third nerve palsy were 28(17%) and fourth nerve were 35(21%). According to this study the most common paralytic strabismus is sixth nerve, which is similar to our study. ^{15,16}

In contrast, in a study conducted by Holmes et al in 1999, the most commonly affected nerve was fourth, followed by sixth and third cranial nerve. This can be explained as this study provided data on cranial nerve palsy in pediatric population in which common cause is congenital. ¹⁷

In all patients enrolled in our study, MRI orbit was conducted. Ischemic causes could readily be identified and thorough systemic examination along with relevant investigation performed on all patients helped to rule out / elicit various causes. Patients in which no cause could be elicited were labeled as idiopathic.

The etiological profile showed that out of total patients of paralytic strabismus, most common etiology was idiopathic which accounted for 48.7% of cases. Second most common etiology in paralytic strabismus was trauma (25.6%), followed by microvascular causes (15.4%). Intracranial anomalies accounted for 5.1% and inflammatory causes were detected in another 5.1%.

Literature shows that the etiological profile of paralytical strabismus is varied, mainly dependant on profile of study group. This can be explained by different referral patterns of the population under study. Our institute being referral centre mainly for trauma patients, our study showed higher incidence of sixth nerve palsy.

In a study conducted by Richards et al in 1992, similar result was obtained, with 25% cases of unknown etiology and 15% of vascular causes. Similarly, a study conducted by Tiffin et al in 1996, 35% patients were of unknown origin while 33% had vascular causes. Both these studies are in consensus with the results of our study. 16,18

While in contrast, Park et al in 2005, reported vasculopathy as the most common etiology and Ho et al in 2013, also reported vasculopathy as the most common cause. ¹⁹

Such differences in the distribution can be attributed to difference in patient characteristics, diagnostic tests and classification criteria used in different studies. In our study neurological features were associated with 39.6% of the patients while they were absent in 60.4% patients. Out of 39.6% patients who had associated neurological features,

89.5% were patients of paralytic strabismus while 10.5% were patients of restrictive strabismus.

This study has adequately pointed out the risk factors and significance of detailed documentation of the demographic features, clinical examination and necessary investigations in understanding the disease and its associated factors. Our study has reflected the prevalence, distribution, etiological and clinical profile of patients presenting with incomitant strabismus including paralytic and restrictive cases.

5. Conclusion

Data on the incidence and etiology of different types of paralytic and restrictive strabismus is helpful for ophthalmologists and other physicians in guiding diagnosis and evaluation and is useful for proper and timely intervention and timely recovery in these patients. A larger sample size specially for cases of restrictive strabismus and longer follow up is however required

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7. Source of funding

None.

8. Conflicts of interest

None.

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