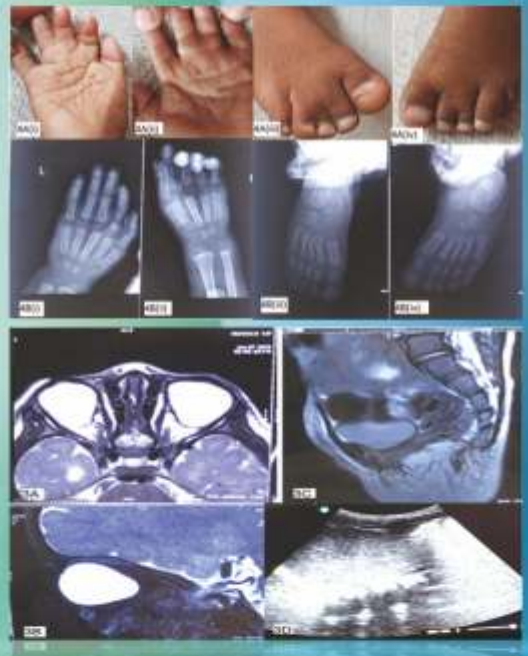
Indian Journal of
Strabismus and Paediatric Ophthalmology
Volume 8 • Issue 1 • December 2020



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Acknowledgement

We have been passing through most catastrophic phase of this century with Global SARS-COV-2 pandemic sweeping the planet for over a year now, leaving it's own trail of death and human misery all around. The pandemic has brought in it's own cascade of financial misery, dislocation of work and loss of employment for large swathes of population. We are passing through the second wave which has been far more devastating than the first one, we have lost many of our colleagues in the medical fraternity and near and dear ones. Under such extenuating circumstances, there has been some delay in bringing out this issue of the IJSPO which is deeply regretted.

Despite suchall pervasive gloom, fear of the known and the unknown and unebbing human misery, I have received overwhelming support from every quarter. The Inputs, suggestions and criticism have all contributed immenselyin bringing out this issue of IJSPO. I thank president and secretary SPOSI for their valuable suggestions and inputs. My special thanks are to Prof. Subhash Dadeya, our past president who has been an unwavering pillar of support. His valuable suggestions, inputs and spirit of camaraderie has been a source of constant inspiration. My thanks are also due to Prof. A.K. Khurana, our past president, who has always given sagely advice from his vast experience. I would also like to thank Dr Rupak Brahma Chaudhary and Priya Saraf for their hard work and tireless effort put- in to bring out this issue. I am extremely grateful to our printers, Shri Ashish Jiand Girish ji for theirimmense help and cooperation in bringing out this issue.

Publication of a peer reviewed journal / periodical is the soul and spirit of any scientific body, lest we be intellectual orphans. It's our endeavor to improve the scientific content, introduce peer review process and publish the journal on quarterly basis, however at present costs are a big deterrent. Views, opinions, suggestions and healthy criticism are solicited from all to make the journal vibrant and scale new unchartered heights. I am sure my successor, Dr. Urmil Chawla will do an outstanding job in her tenure. As secretary SPOSI, I wish her all the best and promise her unebbing support in all her endeavors. In this Corona infested period, we have to explore the possibility of bringing out only online issues.

Long live SPOSI and IJSPO.

Pramod K. Pandey
Editor,
Indian Journal of Strabismus and Pediatric Ophthalmology

New Delhi
12.12.20

Editorial

The wages of denial of insurance claims for Strabismus Surgery; time to ponder and reflect.

'Those who will not reason are bigots, those who cannot, are fools, and those who dare not, are slaves'- Lord Byron.

Socrates used to say that 'I am wise because I know that I do not know', implying that all we know is infinitely far less than what we don't know. But when it comes to strabismus and amblyopia, some of us adopt an ostrich like approach and stoutly deny the undeniable.In order to short- change the insured, the insurance firms turn a Nelson's Eye and just to save some pennies, turn down legitimate insurance claims and what isalready known,gets inextricablysubsumed and consumed in the troughs of the unknown. Surgeries are frequently deferred, the strabismus and amblyopia fester with totally avoidable consequences.

It's like missing 'the woods for the trees' or being 'penny wise and pound foolish'. The economic consequences of loss of binocularity and attendant visual loss in one eye due to resultant amblyopia are far more damning and devastating than the money saved by the insurance firms or Govt. agencies by denying insurance claims or financing the treatment. The tenuous alibi that strabismus is cosmetic in nature andit's surgical correction has no functional, psycho-social or quality of life issues, notwithstanding a plethora of body of evidence to the contrary, is absurd to say the least. The burden of strabismus and amblyopia is further compounded by inevitable and often inexorable deterioration with passage of time. The wages of this intransigence are borne by both by the individual and the society at large. Looking at the magnitude of the problem, a course correction is in order and long overdue.

Strabismus may affect upto 4 to 5% of adult population withtremendous functional, cosmetic, psycho-social and quality of life implications. The additional burden of amblyopia and uncorrected refractive errors, frequent fellow travelers compound the problem manifold with a battery of downstream effects rendering the person almost one eyed, cosmetic disfigurement from the strabismusbeingthe last Act in the Play and just the tip of the iceberg. Ironically most of those afflictedare in pediatric age group and migrate to adulthood carrying the unmitigatedmalady with all it's attendant temporal and spatial consequences. By sheer numbers, the problem assumes public health proportions.

As compared to western societies, the share of adult strabismus in our population is likely exponentially much higher as it is not treated at the appropriate time and left to fester into adulthood in all it'spristine glory. There is more to it,many among us hold the considered view that strabismus can't be fixed and keep vacillating fromputting them under the knife to watchful neglect, with all it's attendant phenomena and epiphenomena. In a society embellished by poverty, ignorance, poor communications, lack of proper health care infra structure with poor access, social dogmas are all pivotal in derailing the health delivery system for strabismus. For any activity to survive and prosper in present times, money is like life- blood, staunch the money trail and the activity will get asphyxiated in no time. Due to astronomical figures, witnessing a further escalation due rising graph of myopia, strabismus is a problem of the community as well, besides being an individual affliction. It is thus highly desirable that lack of finances should not be a hurdle for timely management of strabismus and amblyopia.

In great majority of cases, insurance claims are being shot down under the speciousalibi that strabismus is either cosmetic condition or it was present since birth, hence congenital. Sui generis, both these premises are totally skewed, misplaced and are counter- productive. The truth is that strabismus primarily turns cosmetic if not treated and most of what is presumed to be congenital is not actually congenital but acquired in early months of life as the two parallel monocular visual systems from the lower lateral eyed animals proselytize into binocular single vision of frontal eyed animals including man.

Thus most concomitant strabismus is akin to a language ill learnt in early childhood. The binocular wiring, a pre-requisite for binocular single vision has to take place in the occipital cortex, if it's stalled or goes haywire, strabismus and amblyopia will invariably result. Strabismus and amblyopia are thus cause and effect of disordered wiring in the cerebral visual cortex and other association areas of the brain. The strategy should be to catch it young and remove the obstacle in the path of orderly development of binocular single vision as well as contain and repair whatever damage has already been done. It's a travesty that we move in reverse gear and let both strabismus and amblyopia take roots, get inexorably entrenched and once the fire has raged, we

brand it cosmetic.

N most developedcountries, the insurance firms are reimbursing squint surgeries at par with say cataract surgeries A case in point is Canada where a cataract is reimbursed at 395 Canadian Dollars vis a vis strabismus-one muscle at 360 and 2 muscles at 460 Dollars but in India strabismus is a great asset and it's treatment an encumbrance.

Such misplaced precepts of cosmetic versus functional/restorativeipso facto imply that the 2 eyes in humans are like 2 engines in the plane, one is kept in reserve for emergency purposes, absurdly negating the very foundation of frontal eyed binocular single vision in primates and other animals. We owe it to the basic plant of 2 parallel fully crossed monocular visual systems in lateral eyed animals, providing a panoramic view of their environment, with right optic nerve going to left side of the brain and vice versa. For example, a rabbit can see a tiger with one eye,a snake with the other and his burrow in front with both eyes. By extrapolation, one may inanelyinferthat a rabbit who has lost one eye suffers of only cosmetic blemish with no functional overtones for loss of entire hemifield of theaffected side, can only be termed ridiculous and insane. Not only the frontal placement of the eyes but the entire afferent and efferent visual system has undergone tremendous transformation, half crossed chiasma, distinctive representation in the lateral geniculate body, wiring of binocular neurons in the occipital visual cortex, the dorsal and ventral streams of visual processing, motion vision processing, the evolution of smooth pursuit and vergence systems of eye movements along with Donder's and Listing's laws of ocular motility speak volumes about complex evolutionary underpinnings of frontal eyed binocular single vision. How fallacious and absurd we can be by branding loss of primate binocular vision into mere cosmetic loss of an eye.

Evolution, adaptations and exaptations, be it convergent or divergent is never purposeless, wasteful and irrational, if there were no / less gains and more losses, binocular frontal eyed vision would have never evolved or wouldquickly have turned atavistic. The fact that it got firmly entrenched, not only in primates but also in birds and carnivores, may still be upgrading itself, speaks volumesabout the proof of the pudding.Strabismus and amblyopia thus evolved as an epiphenomenon of frontal eyed binocular vision andare a great hindrance in the path of achieving and maintaining binocular single vision andhave to be eliminated with all our might.

Apart from cosmesis,sub-normal / loss of binocularity, amblyopia, impact on quality of life and psychosocial effects, the other potentially detrimental effect of strabismus on visual function relates to overlooked decreased binocular summation (BiS) and binocular inhibition. Binocular summationis characterized by enhancement in certain aspects of visual function such as low contrast sensitivity when viewing under binocular conditions as compared to monocular conditions. Binocular inhibition may bring down the visual function while viewing binocularly especially when visual acuity between 2 eyes is vastly different.

Despite effective treatment modalities for strabismusincluding newersurgical modalitiesthat have evolved over time, our understanding of the functional binocular visual deficits still lags far behind. Most tests for binocularity involve tests for fusion and stereopsis, however some such tests have questionable validity due to monocular cues or dissociative testing protocols. Besides all such tests require minimum level of visual acuity in each eye. To cap it all, patients with early onset or longstanding strabismus perform poorly on such tests and show little improvement after surgical treatment. It follows that fusion and stereopsis, used almost exclusively for sensory evaluation are not foolproof as clinical trial outcomes in such patient populations, a case in point is stereopsis in intermittent exotropia.

Although BiS has been studied in laboratory settings for more than 50 years, its poorly studied in patients with strabismus. Most such studies have used a cohort of less than 20 patients andoften visually evoked potentials alone. Contrary to stereoacuity, BiS is not affected by monocular cues and can be assessed in patients with poor vision in one eye or those with childhood strabismus with sub-optimal fusional potential. It's also known that advanced age and significant difference in interocular visual acuity, multiple sclerosis and age-related macular degeneration, unilateral cataract may all impair BiS. When interocular difference in VA is large, a destructive neuronal interaction can also occur, known as binocular inhibition,ie diminished binocular visual acuity over uniocular acuity.

It has been conclusively shown that strabismic patients often exhibit deceased BiS or binocular inhibition particularly for very low contrast tasks. Thereby, measures of binocularity like BiS may have more validity that stereotyped stereopsis and fusion. The exact mechanism for BiS is in the realm of speculation but may lie in probability summation which assume complete independence of the two eyes like in lateral eyed animals with 2 independent fully crossed visual systems. This precept assumes that binocular observer has 2 opportunities to detect weak signals. The other concept is of neural summation, the result of binocularly enhanced performance that will exceed that by probability summation. Neural BiS for various electrophysiological and psychophysical tasks has been shown to reside at the level of layer IV in the visual cortex whereas that for binocular inhibition may rest in thelayer VI.

Binocular summation has also been studied in amblyopic patients in several independent studies, earlier studies showing decreased BiS or even binocular inhibition, the degree of loss of BiS and binocular inhibition seem to be directly linked to difference in VA between the two eyes. Recent studies have shown that decreased BiS in amblyopic patients can be improved by normalizing inter-ocular difference in VA by using neutral density filters, betraying that such amblyopic patients retain their neural mechanism for BiS but are disadvantaged due to difference in VA. Sloan low contrast acuity letter charts both at 2.5 %and 1.25 % contrast levels have been found to be especially useful in differentiating strabismic from control populations.

An improvement in BiS scores especially in Sloan

low contrasts at 2.5% and 1.5% has been shown in most patients undergoing strabismus surgery.

It has also been shown that diminished BIS and presence of binocular inhibition negatively impact QOL in strabismic patients. It has been documented that BiS can be remedied by surgical repair of strabismus.BiS is not subject to monocular clues that often contaminate tests of stereoacuity. In studies on BiS, patients with exotropia favored better than patients with esotropia. infantile and childhood onset esotropia had worse outcomes in the improvement of scores as compared to those with congenital superior oblique palsy and intermittent exotropia, Patients with acquired hypertropia or exotropia had much better improvement in BiS. Age at onset, age at surgery, presence or absence of diplopia may also be impacting gain in BiS and need to be studied further.

There is thus robust emerging evidence that there may be functional benefits in strabismus surgery beyond stereopsis through many other aspects of binocularity including BiS and binocular inhibition.

Contraction of peripheral binocular visual fields (BVF) in esotropia is well documented however the relationship between extent of BVF and angle of esotropia is less well understood. In small angle esotropia, there is a suppression scotoma confined to the central region of the deviated eye in a region corresponding to the fovea of the fixing eye. In large angle ET, marked restriction of nasal and temporal fields has been reported. It has been speculated that developmental gains in infantile ET often relate to expansion in BVF. Similarly, adults undergoing surgery for large angle ET have a consistent increase in the post- operative BVF.

In patients with hemianopic visual field defects, surgery may have pivotal role in visual fields expansion / centration and diplopia -management with great functional consequences.

Stereotyped acquired adult strabismus patients experiencing diplopia from road traffic accidents could be the only exceptions where Insurance firms are likely in a tight spot and are unable to refuse the claims. For adult patients not experiencing diplopia and with long standing childhood strabismus, function related aspects of 'Healthrelated quality of life' (HRQOL) can be significantly improved following strabismus surgery, particularly in the areas of reading, stress, hobbies, strain, worry, concentration and depth perception. These specific function related benefits should be taken into account while contemplating strabismus surgery in non- diplopic adult patients and insurance firms should be liable to honor claims.

In this regard, the 25 item National Eye Institute Visual functioning Questionnaire (NEI VFQ25), a valid and reliable scale that captures the effect of vision on multiple dimensions of quality of life including emotional well-being and social functioning has reported scores for diabetic retinopathy, age related macular degeneration, cataract and glaucoma. By comparing these scores to those in adult strabismus, similar / worse scores have been observed showcasing similar / worse impact of strabismus on quality of life but when it comes to reimbursement for strabismus, hypocrisy continues to rule the roost.

Decreased quality of life in strabismic patients may be related to both functional and psychosocial factors. Diplopia has been associated with markedly decreased QOL using both NEIVFQ-25 and adult strabismus-20 (AS-20) scales.

Psychosocial factors may also be a culprit in matters of quality of life instrabismic patients. Strabismic patients who experienced more appearance related distress were also found to have worse quality of life. Strabismus negatively affects self- image, job prospects, relationships, education and sports. Over and above young adults with a history of childhood strabismus have a higher rate of mental health problems, increased depressive symptoms and high rate of social phobia. The role of asthenopia on QOL in strabismic patientsneeds further study.

On peripheral vision subscale, strabismic patients are known to perform significantly worse that patients with cataracts and similar to glaucoma, this may be related to compromise of binocular visual field in some forms of strabismus. Age is also an important denominator, some studies finding worse QOL in younger strabismic patients while others report no effect of age. These findings underscore the importance of QOL in strabismic patients and justify reimbursement claims.

To cap it all, an impression has been fostered by peers that strabismus can not be cured and results are invariably sub- optimum, in most cases surgery must be deferred till adulthood with all the attendant consequences of a long standing deviationand in the process jettisoning strabismus from pediatric ophthalmology. Ensnaring medial and lateral recti with more or less violence is anyway considered amundane affair which ophthalmologist of any hue can accomplishand patient mentally programmed for improvement, not cure. The attendant burden of inappropriate outcomes passed on as appropriate, inexorable amblyopia, increase in the residual deviation angle, recurrence, need for multiple procedures, loss of binocularity and stereopsis are subsumed further with economic constraints due to denial of claims.

That most of strabismus is a congenital disorder present since birth thus being beyond the pale of reimbursement has it's own temporal trajectory . Congenital conditions like Duane Syndrome, oculomotor nerve palsies may get worse with time with development of secondary muscle sequelae and deep amblyopia may set in, punctuating an additional baggage on a congenital disorder. Most of the concomitant strabismus, including infantile is not congenital and only develops subsequently. Except a small minority most such infants are developmentally normal, further negating congenital precepts.

There isthus an urgent need for creating awareness and course correction in our approach to strabismus and amblyopia, that both can be treated and need to be treated early in lifein order to avoid dreadful sequelae that invariably follow with passage of time and become burden to the individual and to the society at large. Early detection, timely, appropriate intervention untrammeled by financial red flags is the roadmap. There is also an overriding need to create a workforce of trained manpower, scaled up

educational efforts regarding strabismus and amblyopia, creating institutions of excellence to train manpower, to generate epidemiological data about the prevalence and incidence of these curable disorders and to undertake applied research in the right perspective.

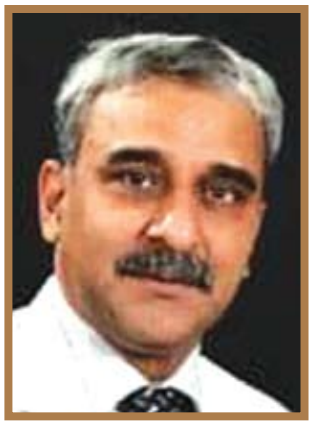
A comprehensive program to control visual disability and blindness from strabismus and amblyopia is need of the hour. To circumvent financial red flags, its imperative to engage with institutions like 'Insurance Regulatory and Development Authority of India' and other appropriate governmental agencies to bring about a radical change in their attitude regarding strabismus as a purely cosmetic problem and their profound amblyopia to amblyopia. The functional aspects as highlighted above need to be stressed and hammered in. The onus is cast upon Strabismus and Pediatric Ophthalmology Society of India to take up the issue of denial of insurance claims for strabismus, to create awareness about childhood blindness and devise comprehensive strategyregarding curative and preventive aspects.

According to Arthur Schopenhauer, a nineteenth century German philosopher- 'All truth passes through three stages. First,it is ridiculed. Second, it is violently opposed. Third it is accepted as being self- evident'. The sooner we pass to stage three in matters of strabismus and amblyopia, better it is. It is imperative to bring a sea change in the attitude of insurance firms, government agencies, let us jointly and unitedly usher in the much- needed change.

Together, we can.

Dr. Pramod K. Pandey
Editor,
Indian Journal of Strabismus and
Pediatric Ophthalmology

New Delhi
12.12.20

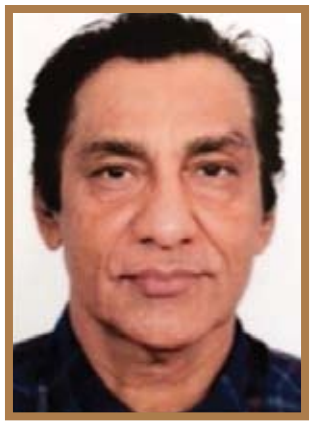


From the desk of President

It gives me immense pleasure to learn that Volume 8 issue 1 of the Indian Journal of Strabismus and Pediatric Ophthalmology is being brought out albeit some delay due to ongoing Covid pandemic. This has been possible due to sheer hard work put in by our editor, Dr. P.K. Pandey, despite raging pandemic. It has been a difficult proposition to bring out the Journal despite lockdown with almost everything disrupted and shut. Publication of a periodical/ journal is like soul of an organization and truly reflects not onlyit's aspirations but also grit and determination to forge ahead against all odds. It also mirrors organizational commitment to exchange our views and find solutions to the issue confronting us.

I congratulate Dr. Pandey in bringing out this issue of the IJSPO and also thank the contributors for having contributed manuscripts to the Journal. I wish IJSPO scale new heights in future and continue to be the official mouthpiece of the organization.

Professor Sudershan Khokkhar



From the desk of Secretary, SPOSI

I am extremely pleased to see the several months of effort finally bearing fruit.This journal of the Strabismus & Pediatric Ophthalmology Society of India(SPOSI), is the mirror of the scientific activities of the society and represents a mirrage of thoughts of the members of the society.

Dr P.K. Pandey, the Editor of this journal, has taken great pains in bringing out this edition. I congratulate him for the efforts he has taken and thank the contributers for this journal and wish that the future editions would be more informative.

with best of wishes,

Prof. (Dr). Yogesh Shukla

Altered Pattern of Ocular Trauma Incidence and Outcome During COVID-19 Lockdown Period

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Abstract :

Introduction: *This study investigated the effect of coronavirus disease 2019 (COVID-19) lockdown on the incidence and management patterns of ocular trauma.*

Methods: *In this retrospective cohort study, the data of patients with ocular trauma in the emergency department during the lockdown period (from 22nd March 2020 to 1st June 2020) were collected. Also, the data of patients presenting ocular trauma during the same period in the previous year were collected. The different variables during these two periods were compared.*

Results: *The cohort consisted of 353 patients, of which, 243 were from 2019 and 110 from 2020. A significant difference was detected in the total number of cases, total ocular trauma cases, total cases needing surgery, paediatric cases with ocular trauma, the ratio of open and closed globe injuries, common objects causing injuries and common activity during injury during the two periods.*

Conclusions: *COVID-19 lockdown exerted a significant effect on the incidence and outcome of ocular trauma.*

Keywords: *lockdown; incidence of ocular trauma; visual outcome; COVID-19*

INTRODUCTION:

Coronavirus disease 2019 (COVID-19) pandemic is disrupting the world and representing the most significant stress for several national healthcare systems and services since their foundation. The supply-chain disruption and the unprecedented request for intensive care unit beds have created conditions, which are typical of low-resource settings in Europe. The COVID-19 epidemic, which originated in Wuhan, Hubei Province, China, and rapidly spread to other provinces in China and 190 countries worldwide. It was declared a global pandemic by the World Health Organization (WHO) on 9th March 2020, thereby becoming a 'public health emergency of international concern'. Patients who are positive for COVID-19 infection are the main source of infection. Nonetheless, asymptomatic COVID-19 patients are also extremely contagious with strong infectivity in the incubation period of 1–14 days. The person-to-person transmission routes of the 2019 novel coronavirus (2019-nCoV) include direct transmission through coughing, sneezing and droplet inhalation transmission and contact transmission through oral, nasal, and eye mucous membrane contact. However, whether COVID-19 transmission occurs through the faecal-oral route remains to be determined. Thus, infection control measures are mandatory to prevent the virus from spreading and control the epidemic situation.[1-4] Because of the rapid identification of the infection during the diagnosis and treatment of ocular diseases, non-urgent outpatient ocular treatments were suspended, and only crucial emergency situations were handled.[5-6]

COVID-19 pandemic is constantly remodelling the healthcare access and practice patterns across all fields of medicine.[7-10] In an effort to decrease the transmission of 2019-nCoV, various governments have imposed social distancing measures and lockdowns. India went into lockdown on March 23rd 2020[1]. Although effective, these social distancing measures have posed new challenges, such as restricted access to healthcare as a result of travel restriction and lack of transport facilities.[11-14]

Ocular trauma is a leading cause of monocular blindness[3]. The problem of treatment of this ocular emergency has been elevated manifold during this pandemic situation.[15-17] Several studies have reported a change in the trend of ophthalmic practices due to the lockdown.[5-7] However, only a few have shown the impact of these measures on the incidence of ocular trauma with respect to limited healthcare access and the subsequent outcomes.[6-9]

In this study, we provide an insight on the demographic patterns and outcomes of ocular trauma during the lockdown and suggest alternatives or change in policies for the prevention of such injuries and their associated comorbidities.

METHODS:

This retrospective cohort study was approved by the

Hospital Ethics Committee. In WESTERN CENTRAL India, since the imposition of COVID-19 lockdown from 22nd March, 2020, travel restrictions have been implemented and inter-district transportation has been stopped. Thus, for this study, we retrieved data of patients who presented ocular trauma to our Emergency Department between 22nd March and 1st June 2020, from the electronic medical record system of the hospital.

Informed standard consent for electronic data privacy was provided by the patient or the parents or guardians of the patient at the time of registration at the hospital. None of the identifiable parameters of the patient information was used for the analysis of the data. The study adhered to the Declaration of Helsinki.

All patients underwent a thorough ophthalmic examination, including visual acuity testing using Snellen's chart and slit lamp biomicroscope. The fundus was evaluated using an indirect ophthalmoscope and +20D lens. However, when the optical medium was not transparent, USG-B scan was used to evaluate the posterior segment. In addition, we collected data of patients presenting ocular trauma during the same period in the previous year. Subsequently, the number of patients who presented ocular trauma, the number of patients who required surgery, and the number of paediatric patients was determined; these data were compared between the two study periods.

During the examination, the data were entered online, using a specified pretested format designed by the International Society of Ocular Trauma (initial and follow-up forms), by trained ophthalmic personnel and supervised by an ophthalmologist. Then, the data were exported to a Microsoft Excel spreadsheet and analysed using SPSS[22].

Frequency distribution and cross-tabulation were analysed and 95% 95% confidence intervals (CIs) calculated. P<0.05 was considered as statistically significant.

RESULTS:

The current cohort consisted of 353 eyes, of which 110 were from 2020 and 243 were collected during 2019. The patterns of ocular trauma were different for both years. Also, the total outdoor injuries were more during 2020 in the paediatric age group. Surgical management addressed a similar incidence of open globe injuries in both adult as well as paediatric age group during 2020 (Table 1).

Table-1 Comparative study between Lockdown and non lockdown period

	2019 Non Lockdown Period	2020 Lockdown Period	P Value	Total
Total Cases	3106	483	<0.05	3589
Ocular Trauma cases (%)	243(7.8%)	110(22.7)	<0.05	454(12.6%)
Ocular Trauma cases needed Surgeries (%)	62(25.5%)	52(46.4%)	<0.05	122(34.6%)
M:F Ration	74.9:25.1	78.6:21.4	NS	75.9:24.1
Mean Age	34.60	27.68	NS	32.42
Pediatric Cases with Ocular Trauma (%)	57(23.5%)	40(36.4%)	<0.05	97(27.5%)
Pediatric Cases with Ocular Trauma requiring surgical intervention (%)	27(47.3%)	24(60.5%)	NS	52(53.6%)
Mean Interval between Injury and Presentation in days	7.03	6.09	NS	6.71
Mean Follow up in days	25.5	18.9	NS	23.44
Open: Closed globe Ratio	88.2:11.8	60.9:39.1	<0.05	20.4:79.6
Common Activity during injury Play	58(23.9%)	31(28.5%)	<0.05	89(25.2)
Common Object of injury Wooden stick	69(28.4%)	45 (40.9%)	<0.05	114(32.2)
Average number of surgeries for eye	0.38	0.82	<0.05	0.52

The comparative study on the pre- and post-intervention visual acuity showed a significant impact of the treatment and improvement in the vision with better outcomes (Table 2).

Table-2 Comparative Studies of Visual Outcome of Total cases

Post treatment vision	Pre treatment vision								
	NOPL	<1/60	1/60-3/60	6/60-6/36	6/24-6/18	6/12-6/9	6/6-6/5	NTV	Total
NOPL	14	4	1	1	0	0	0	3	23
<1/60	3	69	6	1	0	0	0	1	80
1/60-3/60	1	9	13	2	1	0	2	0	28

6/60-6/36	1	5	4	18	1	4	0	0	33
6/24-6/18	1	6	2	3	18	0	0	0	30
6/12-6/9	0	9	7	3	6	62	2	0	89
6/6-6/5	0	6	1	1	1	9	42	0	60
LF	1	3	1	1	2	1	0	0	9
NTV	0	0	0	0	0	0	0	1	1
Total	21	111	35	30	29	76	46	5	353

P-0.000

Next, we compared the year-wise outcome between 2019 and 2020 (Table 3).

Table-3 Comparative studies of visual outcome between Loctdown and non lockdown period

Vision Category	Year		Total
	2019	2020	
NOPL	18	5	23
<1/60	59	21	80
1/60-3/60	18	10	28
6/60-6/36	19	14	33
6/24-6/18	23	7	30
6/12-6/9	64	25	89
6/6-6/5	33	27	60
LF	9	0	9
NTV	0	1	1
Total	243	110	353

P=0.031

The comparative study on the open and closed globe injuries pre- and post-intervention visual acuity showed that treatment had a significant impact, and closed globe injury had a better outcome (Table 4).

Table-4 Comparative study of visualvisual outcome according to BETTS :

Post treatment vision	BETTS CATEGORY		Total
	CLOSED GLOBE	OPEN GLOBE	
NOPL	16	7	23
<1/60	47	33	80
1/60-3/60	21	7	28
6/60-6/36	25	8	33
6/24-6/18	26	4	30
6/12-6/9	83	6	89
6/6-6/5	54	6	60
LF	9	0	9
NTV	0	1	1
Total	281	72	353

P=0.000

Wooden stick is the most common object of injury (32.2%) in our set-up, followed by play as the most common activity (25%) and job-related activities (19%) causing injury.

The proportion of cases with traumatic endophthalmitis increased from 0.85% (3/353) to 6.25% (7/110) during the lockdown period; all were open globe and only one was presented after 24 h post-injury. Primary wound repair (24%) is the most common surgery, followed by corneal foreign body removal (16.1%) and vitrectomy that was required in 11.6% and 5.4% of our cases.

The comparative analysis of the number of structures involved, the type of injury as per Birmingham Eye Trauma Terminology system, and the number of surgeries required post-intervention visual acuity revealed that these factors had a significant impact on the outcomes. In addition, injuries that were associated with ≥3 ocular structure, those that required >2 surgeries, and open globe injuries had poor outcomes.

The most common probable cause of no improvement in the vision was corneal opacification and traumatic endophthalmitis.

DISCUSSION:

The goal of the current study was to investigate the changing patterns of ocular trauma demographics and outcomes. During the lockdown, the imposition of travel restriction increased the overall incidence of ocular trauma (this could be due to the decrease in the total number of patients at our centre since the routine outpatient department was closed). Das and Pellegrini reported altered patterns of ophthalmic trauma in the Emergency Department.[18,19] The proportion of cases amongst the paediatric age group increased from 23.5% to 36.4% (Table 1). Similar findings have been reported by Hamraoush and Ahmed[20] and Shah et al.[218] This phenomenon could be attributed to the lack of activity and limited mobility since schools are shut, leading to boredom and excess pent-up energy in children who are more likely to be engaged in play activities However, Pellegrini et al. reported[187] a decline in the number of children getting injured, which might be due to the regional differences.

The mean interval of the presentation was 6.08 days less as compared to the previous year because only severe cases were presented during this lockdown period. To the best of our knowledge, a comparison

between lockdown and non-lockdown periods has not been carried out previously.

Open globe injuries were common as only severe cases were presented because of lockdown and no transport. Furthermore, play activity was increased during the lockdown period as children were free and not going to school and playing with a wooden stick. However, injuries due to household activities/domestic chores increased from 6.9% to 24.1% and those due to job-related activities decreased from 18.7% to 12.5%.

In our cohort, 52.7% showed a delay in the presentation of the injury by more than 24 h; nonetheless, this did not have a significant bearing on the post-intervention visual acuity. Singh and Sharma[17] reported that 70% of their cases had delayed presentation and were associated with poor prognosis. Strikingly, delayed presentation in ocular trauma has been described before in the rural population.[15-17]

Bapaye et al. warned about the possibility of increased penetrating injuries due to bow and arrows following the broadcast of the popular TV show Ramayana.[22] Herein, the penetrating injuries increased from 6.6% last year to 19.6% during the lockdown, but none were due to the use of bow and arrows. This phenomenon could be attributed to the fact that most of the injuries due to wooden sticks, which have a diverse morphology, i.e., some parts are sharp, while others are blunt and hard that might cause penetrating injuries.

The proportion of cases with traumatic end ophthalmitis increased due to late presentation during the lockdown period. Singh et al. 11 reported 1.81% cases in their cohort with traumatic endophthalmitis, while Das et al[(19)] reported that 2.97% of patients in their cohort presented acute endophthalmitis during the lockdown period.

In the current study, 17% of our patients showed and involvement of >3 ocular structures, while 42.9% presented a visual acuity of ≤3/60, whereas Pellegrini et al.[18] reported that only 5.6% of the patients in their cohort had significant injuries that require monitoring.

The comparison of the effects of lockdown revealed a significant effect of the lockdown in the BETTS subtype of injury and on age; however, it was not associated with visual acuity or time interval between the injury to the presentation. This phenomenon reflects the healthcare-seeking behaviour of our cohort.

CONCLUSION:

It may be prudent to develop better preventive strategies and educate the parents on the risks of ocular trauma during such lockdown times. Maintaining a routine, structuring the playtime, and assigning creative tasks/chores to children might reduce the risk of such injuries. The maintenance of transport and availability of the emergency facility for treatment improved the visual outcomes. Poor prognosis was associated with open globe injuries, the involvement of >3 structures, the requirement of >2 surgeries, and presence of infection.

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OCT Study or Pediatric Normative Database in Indian Pediatric Population

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Abstract : *Purpose: To form normative database of macular and optic disc parameters in pediatric Indian population using spectral domain (SD) OCT (Cirrus 4000 HD-OCT, Carl Zeiss Meditec, Germany).*

Methods: *This was an observational, cross-sectional study. The eyes of healthy subjects aged between 7-18 years, belonging to western part of India, with no ocular disease and best corrected visual acuity of 20/20 were included in the study. Mean macular and optic disc parameters were measured for this group. These measurements were compared with those in the published literature. Sex and laterality related variations were also studied for this study group.*

Results: *Two hundred and eighty six eyes of 155 pediatric subjects were included. Mean age of our study group was 13.38 ± 3.1 years. There were 148 male and 138 female eyes. The mean central subfoveal thickness measured using SD-OCT was 237.35 ± 20.38 µm. The thickness measurement of inner nasal quadrant was greatest and it was lowest for outer temporal quadrant. We found males to have significantly thicker inner central macular thickness compared to females ($p<0.01$). The mean RNFL thickness was 95.07 ± 10.46 µm. Mean of average cup: disc (C:D) ratio was 0.45 and of vertical C:D ratio was 0.42. Average disc area was 2.1 mm^2 and average rim area was 1.55 mm^2. There was no significant difference in average RNFL thickness or any other optic disc parameters with respect to sex and laterality.*

Conclusions: *This study establishes normative database for pediatric Indian population on Cirrus HD-OCT.*

Keywords: *Normative database, OCT, macula, ONH, pediatric population.*

INTRODUCTION:

OCT is an established, reproducible tool to objectively measure macular thickness and RNFL analysis in various conditions of the eye in all age groups. The various models of OCT differ in their measurement of macular thickness due to different retinal segmentation algorithms used (Fourier-domain OCT shows higher values than Time-domain OCT).[1,2] Stratus OCT, Cirrus HD-OCT and Spectralis OCT measure the macular thickness in different manner and so their measurements are not uniform.[3]

There have been plenty of reports highlighting racial differences in optic disc area, average cup-disc ratio, vertical cup-disc ratio, cup volume, and RNFL thickness as measured by Cirrus HD-OCT. Ethnic variations have also been well documented in normal macular thickness analysis.[1,4]

All the mainline Optical Coherence Tomographers except for the Zeiss Cirrus HD-OCT, which has Asian normative database, have minority Asian representation in their normative database creation and next to no Indian representation in the same. Also there is no existent normative database for population below 18 years of age in any of the mainline OCT machinery softwares.

There is an increasing recognition of importance of OCT in paediatric population and it is evident from existing literature that adult normative parameters cannot be extrapolated to pediatric population. Aim of this study is to form a normative database for macular and disc measurements on Cirrus HD-OCT for pediatric Indian population.

MATERIALS AND METHODS:

Procedures followed in this study were in accordance with the ethical standards of the institutional committee and with the Helsinki Declaration of 1975, as revised in 2000.

Study design: This is a descriptive, cross-sectional, observational study.

We enrolled healthy pediatric (7-18 years) individuals who were carefully screened and evaluated for eligibility. Medical and ophthalmic histories were taken prior to enrolling the subjects in the study. All subjects had been born at full-term (>/= 37 weeks of gestational age) which was verbally confirmed with their parents/guardian. After undergoing a general ophthalmic examination which included measuring best corrected visual acuity (BCVA), applanation tonometry (GAT), keratometry, cycloplegic refraction, axial length measurement using IOL Master, slit lamp and dilated ophthalmic examination, the qualifying and consenting subjects underwent retinal scanning with the Cirrus HD-OCT (Cirrus 4000 HD-OCT, Carl Zeiss Meditec, Germany) instrument.

Informed consent was taken from parent or guardian of patient for conducting the test.

Males or females </= 18 years of age whose parent or guardian had given a valid consent were included in the study. Subjects with BCVA in either eye worse than 20/20, refractive error (spherical equivalent) outside -3.00 to +3.00D range, presence of amblyopia, previous laser or intraocular surgery, any active infection of anterior or posterior segments or any optic disc or retinal abnormalities were excluded from the study. Any systemic condition that precluded detailed patient evaluation and performing above investigations were also excluded.

Normal subjects were defined by Principal Investigators after review of clinical data, and considering inclusion and exclusion criteria.

The Cirrus instrument was not used in determining the normalcy of the subjects.

DATA COLLECTION:

For the RNFL normative database, each eye was scanned three times with the Optic Disc Cube 200x200 scan.

For the macula normative database, each eye was scanned three times with the Macular Cube 512x128 scan and the macula was divided into 3 concentric circles with fovea at the centre. This was superimposition of the Early Treatment Diabetic Retinopathy Study (ETDRS) circles on the OCT macular map.

We further studied sex and laterality-wise distribution of inner central macular quadrant thickness and average RNFL thickness.

SCAN SELECTION CRITERIA:

The scans were reviewed for image quality. One best quality scan for each scan type was chosen for each subject per eye.

Scans withsignal strength of 6 or lower, large eye motion during image acquisition, resulting in a saccade that was within the central 80% of the scan area and area of data loss greater than 10% at the edge of the scan area were excluded from the database.

STATISTICAL ANALYSIS:

All the data was entered and analysed with help of the SPSS 17 software package. Appropriate statistical tests like mean, SD, range will be applied for descriptive purpose and for drawing inference of collected data tests of significance like paired and unpaired t test, Mann-Whitney test, bland altmann analysis, correlation coefficient, linear regression analysis will be applied with the help of biostatistician.

RESULTS AND ANALYSIS:

Demography:

Overall, 286 eyes of 155 healthy pediatric subjects were included in the study. Their mean age was 13.38 years; the range being from 7 to 18 years. There were 81 (148 eyes) male and 74 (138 eyes) female subjects. We had 146 right eyes and 140 left eyes. (Table 1).

There was no statistically significant difference in refractive error between male and female subjects. (p=0.716)

TABLE 1: PATIENT DEMOGRAPHY

PARAMETER	MEASURE
TOTAL NUMBER OF EYES	286
MALE:FEMALE	81:74 (148:138)
LATERALITY	146:140

Macular parameters:

Macular parameters for pediatric population are as shown (Table 2).

We found macular thickness to be greatest in inner nasal quadrant and lowest in outer temporal quadrant. The inner macular quadrants had greater thickness compared to the outer ones.

The sex and laterality-wise distributions of average inner central macular thickness were studied.We found that males had significantly higher inner central macular thickness compared to females (p<0.01) but we found no statistically significant difference in inner central macular thickness between right and left eyes in the pediatric age group. (p=0.936)

TABLE 2: PEDIATRIC NORMATIVE MACULAR MEASUREMENTS

MACULAR PARAMETERS	MEAN (STD DEVIATION) in microns (N=286)
INNER CENTRAL MACULAR THICKNESS	237.4 (20.4)
INNER SUPERIOR MACULAR THICKNESS	313.6 (16.4)
INNER INFERIOR MACULAR THICKNESS	309.9 (16.9)
INNER NASAL MACULAR THICKNESS	314.6 (16.8)
INNER TEMPORAL MACULAR THICKNESS	298.9 (15.9)
OUTER SUPERIOR MACULAR THICKNESS	278.1 (15.7)
OUTER INFERIOR MACULAR THICKNESS	265.1 (15.6)
OUTER NASAL MACULAR THICKNESS	295.9 (15.7)
OUTER TEMPORAL MACULAR THICKNESS	257 (13.7)
VOLUME CUBE	9.75 (0.49)
AVERAGE CUBE THICKNESS	273.1 (13.8)

RNFL thickness:

RNFL and optic nerve head parameters: (Table 3)

The sex and laterality-wise distributions of average RNFL thickness were studied. We found no significant difference in inner central macular thickness between males and females (p=0.242). There was no statistically

significant difference in average RNFL thickness between right and left eyes. (p=0.696)

TABLE 3: PEDIATRIC NORMATIVE OPTIC NERVE HEAD MEASUREMENTS

RNFL & OPTIC NERVE HEAD PARAMETERS	MEAN (STD DEVIATION) (n=286)
Retinal nerve fibre layer thickness	95.1 (10.5)
AVERAGE C/D RATIO	0.45 (0.17)
VERTICAL C/D RATIO	0.42 (0.17)
DISC AREA (mm2)	2.1 (0.43)
RIM AREA (mm2)	1.55 (0.25)
CUP VOLUME (mm3)	0.15 (0.15)

Optic Nerve Head parameters:

40% of children had disc area less than 2.6mm^2. Approximately 40% of children had rim area more than 1.6mm^2. We did not find statistically significant difference based on sex or laterality for any of the optic nerve head parameters: Average C/D ratio (sex, p=0.912; laterality, p=0.529), vertical C/D ratio (sex, p=0.810; laterality, p=0.246), disc area (sex, p=0.441; laterality, p=0.379), rim area (sex, p=0.352; laterality, p=0.674) and cup volume (sex, p=0.503; laterality, p=0.589).

DISCUSSION:

We studied a total of 286 pediatric (7 to 18 years) patients. The macular, RNFL and optic nerve head parameters have been described in Table 1. We calculated and compared the age-wise, sex-wise and laterality-wise distribution of inner central macular thickness and RNFL thickness.The inner central macular thickness was found to be significantly higher in males compared to females in the pediatric age group. We found no significant difference in average RNFL thickness based on sex or laterality.

Different normative databases are available for different OCTs due to the difference in their measurement techniques and their resolution powers. On the basis of various studies as mentioned earlier and further elaborated later, there are substantial evidences to say that macular thickness as well as RNFL thickness varies with ethnicity, age and sex. This builds up the need for an indigenous pediatricnormative database.

For Cirrus HD OCT 4000[5], the available normative databases provided by the manufacturing company are:Diversified- including participants of age group 19 to 84 years from different centres of varied ethnicity and Asian- including participants of age group 19 to 79 years from centres located in Japan, China and India. Ethnicity breakdown of the Cirrus Asian RNFL and Asian Macula normative databases is as follows: 44% Japanese, 44% Chinese, and 12% Indian.

Thus, due to the lack of pure Indian ethnicity and specifically lack of pediatric population in the available normative databases for Cirrus HD OCT 4000; we decided to create a normative database to fill-up these lacunae.

There has been an increase in knowledge of pediatric retinal and optic nerve disorders which has led to the need of pediatric OCT normative database. All the above mentioned databases are for population above 18 years of age.

Macular thickness:

Different OCT machines measure the central macular thickness in different manners. Stratus OCT measures the thickness of the retina as the distance between the inner limiting membrane (ILM) and interdigitation zone (IZ), Cirrus SD-OCT reports it as the distance from the anterior border of the retinal pigment epithelium (RPE) to the ILM, while Spectralis OCT measures the distance from the posterior border of the RPE to the ILM. We reviewed various other articles that reported pediatric macular parameters and have listed them along with our measurement in table 4. The measurement from our study was higher than that reported by Huynh et al.[6] and El Dairi et al.[7] who measured the macular thickness on time-domain OCTs. Mean central macular thickness of our subjects was similar to that found by Katiyar et al.[8] and Nigam et al.[9] but less than that reported by Barrio Barrio et al.[10] and Al Haddad et al.[11], even when measurements in all of these studies were taken with Cirrus OCT, signifying the role of ethnicity in the variations in macular thickness.

In our study, we found the inner nasal macular quadrant to be the thickest, this was followed by inner superior, inner inner inferior and inner temporal quadrants. The outer macular quadrants showed a similar pattern. The inner central macular quadrant was the thinnest. Our findings were similar to those reported by Nigam et al.[9], Katiyar et al.[8] and Eriksson et al.[12]Mean central volume of macula in our study (9.75 +/- 0.49 mm^3) was slightly lower than that reported by Nigam et al. (9.85 +/- 0.57 mm^3).

Previous reports by Barrio-Barrio et al,[10] Huynh et al,[6] and Al-Haddad et al[11] reported that gender differences applied only to central macular thickness measurements, which were significantly increased in malesand we found similar results in our study. On the other hand, Katiyar et al.[8] found no statistically significant difference in the central macular thickness between males and females in the pediatric age group.

TABLE 4: COMPARISON WITH STUDIES WHICH MEASURED MACULAR THICKNESS IN NORMAL CHILDREN

OCT	TYPE OF OCT	SOURCE POPULATION STUDIED (COUNTRY)	AGE (YEARS) (NUMBER OF EYES)	CENTRAL THICKNESS(MICROM)
STRATUS OCT	TIME DOMAIN (TD)	HUYNH et al. [6] MULTIETHNIC (AUSTRALIA)	6.7 +/- 0.4 (1543)	193.6 +/- 17.9

STRATUS OCT	TD	EL-DAIRI et al.[7] MULTIETHNIC (USA)	8.6 +/- 3.1 (286)	188.8 +/- 25.0
STRATUS OCT	TD	ERIKSSON et al.[12] CAUCASIAN (SWEDEN)	5-16 (10.1) (56)	204 +/- 19
SPECTRALIS OCT	SPECTRAL DOMAIN (SD)	TURK et al. [19] MIDDLE EASTERN (TURKEY)	10.5 +/- 2.9 (107)	211.4 +/- 12.2
CIRRUS OCT	SD	BARRIO-BARRIO et al.[10] CAUCASIAN (SPAIN)	9.6 +/- 3.12 (281)	253.9 +/- 19.8
CIRRUS OCT	SD	AL-HADDAD et al. [11] MIDDLE EASTERN (LEBANON)	10.7 +/- 3.14 (108)	249.1 +/- 20.2
CIRRUS HD-OCT	SD	KATIYAR et al.[8] SOUTH ASIAN (INDIA)	12.59 +/- 3.5 (157)	234.94 +/- 18.62
CIRRUS HD-OCT	SD	NIGAM B. et al.[9] SOUTH ASIAN (INDIA)	5-17 (10.4 +/- 2.7) (340)	235.51 (5-9 years), 237.11 (10-13 years), 240.10 (14-17 years)
CIRRUS HD-OCT	SD	OUR STUDY SOUTH ASIAN (INDIA)	7-18 (13.38 +/- 3.1) (286)	237.35 +/- 20.38

Average RNFL thickness and other optic disc parameters:

Different OCT machines have different software algorithms for measurement of average RNFL thickness. We reviewed various other articles that reported pediatric optic nerve head parameters and have listed them along with our measurement in table 5. We found that groups which used Stratus Oct for measurement like Ahn et al.,[13] N. Pawar et al.[14] and Salchow et al.[15] found higher average RNFL thickness compared to that in our study which was measured using Cirrus HD-OCT. Our results were quite similar to that reported by Aparna Rao et al.[16] and Al Haddad et al.[11] but the measurement was lower than that found by Elia et al.[17] and Barrio Barrio et al.[10]

We found no significant correlation of RNFL thickness with sex (P<0.155) and laterality (P<0.284)which was similar to that found by N. Pawar et al.[14] and Aparna Rao et al.[16]

Huynh et al.[6] reported cup-to-disc diameter ratios of 0.46 +/- 0.16 horizontally and 0.42 +/- 0.15 vertically in 6 years old Australian children, this was similar to what we found in our study, although the age range of subjects in our study was from 7 through 18 years. N. Pawar et al.[18] in their study of 70 subjects found an average C/D ratio of 0.64 in right eye and 0.63 in left eye. They reported a vertical C/D ratio of 0.61 in right eye and 0.59 in left eye. This was much higher than that we reported in our study. They reported average disc area of 2.45 mm^2 in right eye and 2.46 mm^2 in left eye and an average rim area of 1.31 mm^2 in right eye and 1.34 mm^2 in the left eye. The average disc area in their study was higher but the average rim area was lower compared to that in our participants. El Dairi et al.[7] in their study of 286 healthy children conducted at Duke Eye Centre, Durham, USA, reported a much average higher disc area of 2.42 mm^2 and rim area of 1.96 mm^2 compared to our study.

TABLE 5: COMPARISON WITH STUDIES WHICH MEASURED RETINAL NERVE FIBER LAYER THICKNESS IN NORMAL CHILDREN

OCT	TYPE OF OCT	SOURCE POPULATION STUDIED (COUNTRY)	AGE (YEARS) (NUMBER OF EYES)	AVERAGE RNFL (MICROM) MEAN +/- SD
STRATUS OCT	TIME DOMAIN (TD)	AHN et al. [13] KOREAN (SOUTH KOREA)	9-18 (72)	105.53 +/- 0.33
STRATUS OCT	TD	PARIKH RS et al.[20] SOUTH ASIAN (INDIA)	5-20 (59)	100.15 +/- 10.8
STRATUS OCT	TD	PAWAR N et al. [14] SOUTH ASIAN (INDIA)	5-17 (120)	106.11 +/- 9.5
STRATUS OCT	TD	SALCHOW et al.[15] MULTIETHNIC (USA)	4-17 (92)	107 +/- 11.1
STRATUS OCT	TD	EL DAIRI et al. [7] MULTIETHNIC (USA)	3-17 (286)	108.27 +/- 9.8
SPECTRALIS OCT	SPECTRAL DOMAIN (SD)	TURK et al. [19] MIDDLE EASTERN (TURKEY)	6-16 (107)	106.45 +/- 9.41
SPECTRALIS OCT	SD	YANNI et al. [21] MULTIETHNIC (USA)	5-15 (8.9) (83)	107.6 +/- 1.2

RTVue-100 OCT	SD	TSAI et al. [22] CHINESE (TAIWAN, ROC)	6.5-12.5 (92) (470)	109.4 +/- 10.0
CIRRUS OCT	SD	ELIA et al. [17] CAUCASIAN (SPAIN)	9.2 +/-1.7 (344)	98.5 +/- 10.8
CIRRUS OCT	SD	BARRIO-BARRIO et al. [10] CAUCASIAN (SPAIN)	9.6 +/- 3.1 (283)	97.4 +/- 9.0
CIRRUS OCT	SD	AL-HADDAD et al.[11] MIDDLE EASTERN (LEBANON)	10.7 +/- 3.1 (108)	95.6 +/- 8.7
CIRRUS SD-OCT	SD	APARNA RAO et al. [16] SOUTH ASIAN (INDIA)	4-17 (148)	OD=94 +/- 10.9 OS=93 +/- 10.6
CIRRUS HD-OCT	SD	OUR STUDY SOUTH ASIAN (INDIA)	**7-18 (13.77 +/- 3.1) (286)**	95.07 +/- 10.46

Limitations of the study:

This study was conducted at a single centre in western India and so the study group might not be a true representation of the Indian population which is so diverse. The measurements with Cirrus HD-OCT cannot be used for the other OCT machines due to the segmentation differences. We have not studied average RNFL thickness in subjects with refractive error > +/-3.00 D spherical equivalent or for BCVA<6/6, N6.

CONCLUSIONS:

There is a significant difference in the macular and retinal nerve fibre layer/optic nerve head parameters based on the variables of age, sex and ethnicity as measured by Optical Coherence Tomography. Indigenous pediatric normative database in OCT is fast becoming a necessity due to increasing understanding of disease processes of the pediatric age group and the reliability of and reliance upon OCT as an accurate measuring device for retinal and optic nerve parameters.Inner central macular thickness is significantly higher in male children compared to age-matched female children. The optic nerve head parameters did not differ significantly with respect to sex and laterality.

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Congenital Cranial Dysinnervation Disorders - Still A Dilemma for Strabismologists

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PGIMS, Rohtak

Abstract : *A group of congenital disorders that had been assumed to result from restrictive limitation of eye movements in reality results from agenesis of ocular motorneurons in the brain stem. Recent advancements in genetics and neuro radiology have made the strabismologists to regroup the congenital neuro muscular disorders into one unified group which has a neuropathic origin mainly due to primary or secondary dysinnervation and redefined thewm as congenital cranial dysinnervation disorders. These are a challenge for strabismologists to treat due to their variable clinical presentation and unpredictable surgical results. We here present a few cases of different variants of congenital cranial dysinnervation disorders and their best possible managements. This article helps us to summarise a few important clinical aspects which help us in diagnosing, grouping and treating these challenging cases.*

INTRODUCTION:

Congenital cranial dysinnervation disorders are defined as Congenital, non-progressive,Sporadic or familial abnormalities of cranial musculature which results from developmental abnormalities like Complete absence of, one or more cranial nerves with primary or Secondary muscle dysinnervation.

EVOLUTION:

Earlier, congenital fibrosis syndrome of the extra-ocular muscles (CFEOMs) was considered to be of primary muscle pathology. However, evidence from multiple sources suggests that it is a neurological disorder rather than primary muscle pathology. Recent advancements in genetics and neuro-radiology have now determined the initial observation of fibrotic muscles is secondary to a primary lack of innervation from deficient, absent, or misguided cranial nerves. This presentation provides an overview of the known genes and phenotypes currently recognized within the CCDD domain. This puts CFEOM in the category of other congenital ocular muscle disorders, which have a neurological basis, such as Duane's retraction syndrome (DRS), monocular elevation deficiency (MED) and Möbius syndrome. All these syndromes have Neuropathic origin which can be explained by the findings such as - anatomic absence of abducens nerve, EMG revealed anomalous innervation of lateral rectus by oculomotor nerve, Neuropathologic studies support neurogenic hypothesis for variants of CFEOM and developmental dysinnervation as the causative pathology in Horizontal gaze palsy with progressive scoliosisand Mobius syndrome.

Nowadays, many investigators suggest unifying all these various congenital oculo-motor disorders into one entity. This entity may include abnormalities in innervation affecting cranial nerves and beyond, motor and may be sensory. For such a collection of congenital innervation disorders, cranial or otherwise, the term congenital innervation dysgenesis syndrome (CID) seems appropriate. They result from developmental errors in innervation of the ocular and facial muscles .

CLASSIFICATION:

CCDD's results from developmental errors in innervation of the ocular and facial muscles and not from primary dysfunction of the muscles. They can be classified as follows:

Classification:

1. CCDDs primarily affecting horizontal ocular motility
   - Duanes Retraction Syndrome
   - Horizontal gaze palsy with progressive scoliosis
2. CCDDs primarily affecting vertical ocular motility
   - CFEOMs
   - Congenital third nerve palsy
   - Monocular elevation deficiency (MED)
3. CCDDs primarily affecting facial muscles with associated ocular motility defects
   - Congenital facial weakness
   - Mobius syndrome

Congenital fibrosis of the extraocular muscles:

(CFEOM) is a rare clinical syndrome characterisedby variable impairment of horizontal and/or vertical eye movements and ptosis. It can be Unilateral or bilateral, with bilateral presentation more commonly seen. Eyesare partially or completely fixed in a downward and strabismic position usually divergent, and additional features like aberrant residual eye movements and Variable abnormalities of eyelids and globes such as –lid

retraction, proptosis, lagophthalmos.

Histo-Pathogenesis- It results from neuromyopathic fibrosis of the extraocular muscles.

Attempts at defining subgroups for CFEOM have been made but considerable overlap and intrafamilial variability exists. Three Phenotypes and Four genetic loci of CFEOM have been described:

CFEOM-Type 1 (Autosomal Dominant): The prototype is Congenital Fibrosis of extraocular muscle type 1, which is autosomal dominant disorder, characterized by bilateral ptosis and Infraduction of both eyes with positive forced ductions. Bilateral restrictions of upgaze and Variable limitations of horizontal gaze are seen with a few additional features like Synergistic convergence and there is absence of Globe retraction. patients usually have an anomalous head posture especially chin up as they attempt to look under their severely ptotic eyelids.

CFEOM- Type 2: This is usually Autosomal Recessive disorder which is characterized by Bilateral Ptosis and fixed Exotropia. There is severe limitation of vertical and horizontal eye movements and this is usually seen in consanguinous families.

CFEOM Type 3: This is usually Autosomal dominant with incomplete inheritance. It is an Unilateral disease with unilateral or absent ptosis. There is Orthotropia or hypertropia in primary gaze. There is near normal elevation and positive force ductions and misdirected movements are rare.

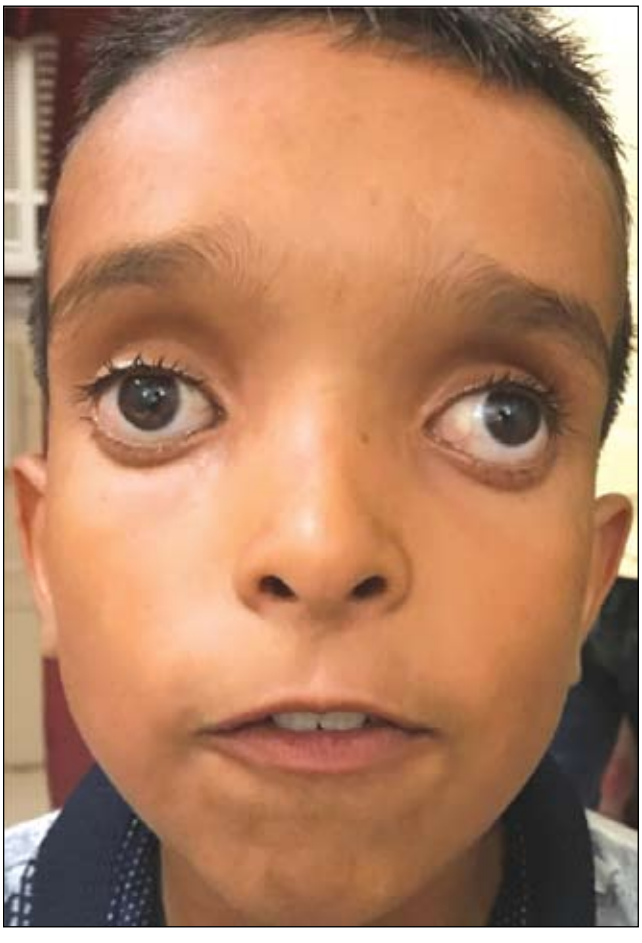


Figure1- chin up head posture

Case Report: 10 years old male child reported on 6-7-2017 to the paediatric ophthalmologic clinic with chief complaint of outward deviation of both eyes since birth. There is downward and outward rotation of fixing eye alternately. There is Limitation of ocular movements in lateral gazes in both eyes and also limitation of elevation in adduction and depression in all gazes in both eyes. There was no complaint of decreased visual acuity and there was no significant birth history, no developmental delay and no prior history of use of glasses, occlusion therapy, surgery or trauma and no significant family history.

CLINICAL PRESENTATION:

His unaided visual acuity R/E 6/6 and L/E 6/9. The patient had chin up head posture for Distance. Abducting nystagmus was present in left eyeand no Gross stereopsis was present (tested on Titmus fly test). Anterior segment examination was normal and Fundus examination was normal.

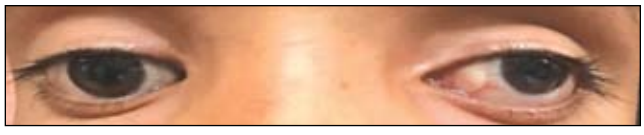


Figure 2- Inferior lid retraction present in both eyes

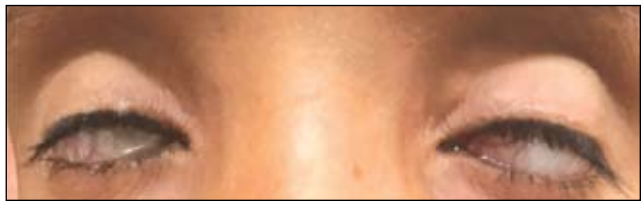


Figure 3- Presence of LAGOPHTHALMOS in both eyes

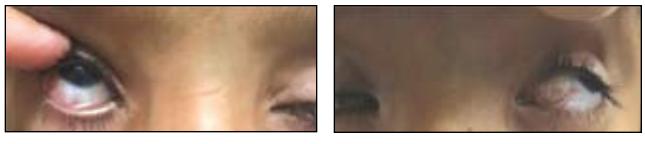


Figure 4- Presence of Bell's phenomenon

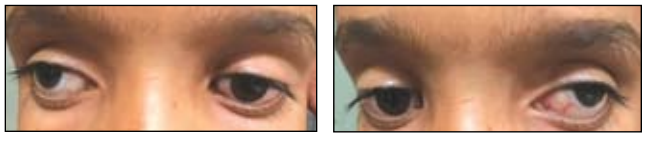


Figure 5- ABSENCE OF LID CREASE in both eyes.

ORTHOPTICS:

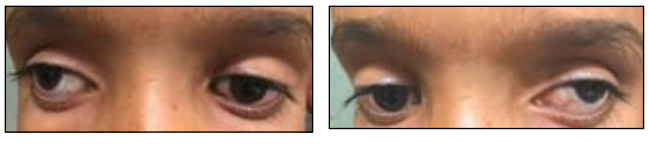
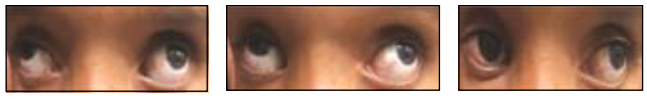
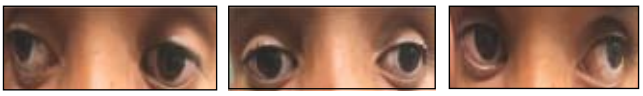


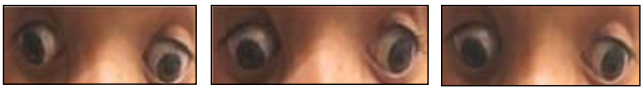
Figure 6- There was presence of exotropia with hypotropia.



45 BI with 7L/R



35 BI with 6L/R



20 BI with 6 L/R

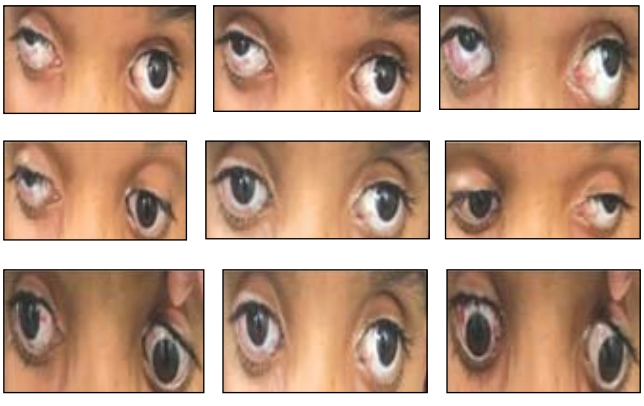
Figure 7 - Nine diagnostic positions of gaze showing exotropia with hypertropia left eye when fixing with right eye in hypotropic position.

Orthoptic evaluation revealed exotropia withalternating hypotropia when fixing with eye in hypotropic position. There was 35 BI of exotropia with 6 Prism Dioptres of hypertropia in primary position. V pattern exotropia of 25 prism diopter was present increasing in upgaze. Limitation of movements in lateral gazes and in downgaze, deficit of elevation in adduction in both eyes was present.

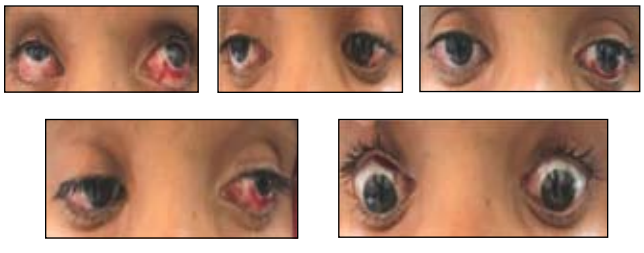
Diagnosis- The case was diagnosed as Congenital fibrosis of extraocular muscles.

Management: Surgical treatment was planned and Intraoperative FDT was done which showed moderately tight lateral rectus muscle on adduction and superior rectus on depression. Intraoperatively fibrous strands present inplace of muscle fibres, no insertion line visible, postequatorial fibrotic strand present behind superior rectus muscle.

First Surgery done-B/L LR RC 14mm from limbus, B/L SR RC 11 mm



At 4 weeks follow up residual exotropia of 35 BI still seen, so second surgery done after 2 months. B/L MR resection 6.5mm and IR tenotomy B/E was done. Intraoperatively the FDT was tight in upgaze. Strands of fibrous tissue seen in place of medial rectus and inferior rectus mainly.



ON follow up after second surgery the patient was well aligned with almost ortho for distance and, small residual for near 12BI with 5R/L.

RESULTS

Cosmetically eyes were well aligned with restoration of stereopsis of 200 sec of arc, improvement in binocular visual field though not much improvement in movements.

DISCUSSION

CFEOM refers to complex strabismus syndromes characterised by congenital non progressive ophthalmoplegia with or without ptosis affecting part or all of the oculomotor nerve and its innervated muscles or the trochlear nerve and its innervated muscle or both. CFEOM haas been found to run in families and is believed to have a genetic basis.

Management of patients with CFEOM has always been challenging. In general affected children have variable limitation of horizontal n vertical gazes, and frequently compensate by turning their heads rather than eyes to track objects. Surgery can correct the deviation of eyes, although restoration of full ocular movements not possible

CONCLUSION

For most patients suspicion of CFEOM is first aroused based on clinical presentation, mainly ophthalmological findings, and some subtypes depend on identification of associated findings. Adequate knowledge of variable clinical presentations, detailed examination and confirmation by means of intraoperative findings and histopathological reinforcement goes a long way in treating such challenging rare cases of complex strabismus.

MOBIUS SYNDROME:

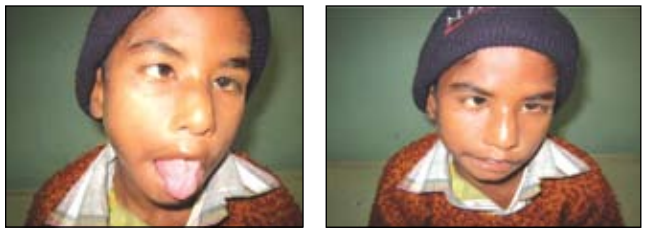
Mobius syndrome is a rare congenital non-progressive neurological disorder caused by the absence or underdevelopment of the 6th and 7th cranial nerves which control eye movements and facial expression.[2] Complete or partial bilateral facial paralysis with bilateral or unilateral abducens paralysis is considered minimal essential criteria for diagnosis. Clinically patient presents with esotropia and limitation of abduction with incomplete closure of eyes. It is named after Paul Julius Möbius, a neurologist who first described the syndrome in 1888.[2] Although its rarity often leads to late diagnosis, infants with this disorder can be identified at birth by a "mask-like" face withlack of expression that is detectable during crying or laughing and by an inability to suck while nursing because of paresis(palsy) of the seventh cranial nerve[3,4]. Additionally, patients with Möbius syndrome cannot follow objects by moving their eyes from side to sideand turn their head instead.[5]

Hypoplasia of the tongue owing to hypoglossal (XII) nerveparalysis,swallowing and speech difficulties owing to trigeminal(V), glossopharyngeal(IX), and vagus(X) nerve palsies; malformations of the orofacial structures

(bifid uvula, micrognathia, and ear deformities); anomalies of the musculoskeletal system , rib defects, and brachial muscle defects may be some other associated features.[6,7]

Moebius syndrome is unlikely to be a single entity as a variety of pathological disturbances may produce a phenotype recognisable as Moebius syndrome. The wide spectrum congenital malformations points to some disturbance during early fetal development. Moebius, in his original description of the conditionpostulated that the anomaly resulted fromdegeneration of the nuclei of the sixth and seventh cranial nerves.[8] The pathogenesis of cranial nerve palsies associatedwith limb anomalies is difficult to explain. An ischaemic process resulting from an interruption inthe vascular supply during early fetaldevelopment,probably around four to six weeks of gestation, may result in facial and limb anomalies characteristic ofMoebius syndrome.[9,10]

A seven year old male child visited OUT PATIENT DEPARTMENT for ophthalmic evaluation. The child was born of full-term uncomplicated pregnancy. There was no history of maternal alcohol or drug ingestion during pregnancy. Systemic evaluation by paediatrician showed no limb or skeletal abnormalities. His best corrected visual acuity was 6/24 with +1.5DS in the right eye and 6/36 with +2DS/-1.75 DC 180° in the left eye. He adopts anomalous head posture while fixing on 6/12 snellen's letter at a distance of 6m (head tilt towards left and right face turn). Anomalous head posture was measured by means of Goniometer. It was 25° right face turn and 5-10° of left head tilt. Ophthalmic examination showed expressionless face, loss of nasolabial folds, entropion of upper lids of both eyes with misdirected lashes. Krimskytest showed large esotropia (60 PD) with left hypertropia (12PD) with fixation preference for right eye.Ocular motility test showedbilateral abduction limitation of -5 (not reaching up to midline) and under action of (-2) in adduction of both eyes. Right eye had -1 limitation in elevation. He had to move his head for viewing objects laterally. He showed increased convergence and upshoots on attempting lateral gaze. Anterior and posterior segment examination was within normal limits in both eyes. Forced duction test under anaesthesia revealed restriction in abduction due to very tight (forced abduction possible just beyond midline) medial rectus muscles in botheyes. The child was operated for entropion of upper lids in both eyes using the technique of tarsal fracture with everting sutures about a month before strabismus surgery. Bilateral large semi hangback recession of medial rectus of 7mm was done along with unilateral augmented superior rectus transposition in the left eye. The child was followed post operatively on day 1, after 1 week and 4 weeks. On his last follow up, the child showed 20 PD of residual esotropia in primary gaze, with mild improvement of abduction (improving to -4) in left eye. With anomalous head posture (20° right face turn, 5° of left head tilt) the cover test showed right hypertropia of 10 PD. He still has to move his head for viewing objects more so in right lateral gaze. Best corrected visual acuity improved to 6/9 part in right eye and 6/12 part in left eye due to increase in his binocular field. The cilia of both eyes were not touching the corneas.



AHP- 25° right face turn and 5-10° of left head tilt with bifid tongue

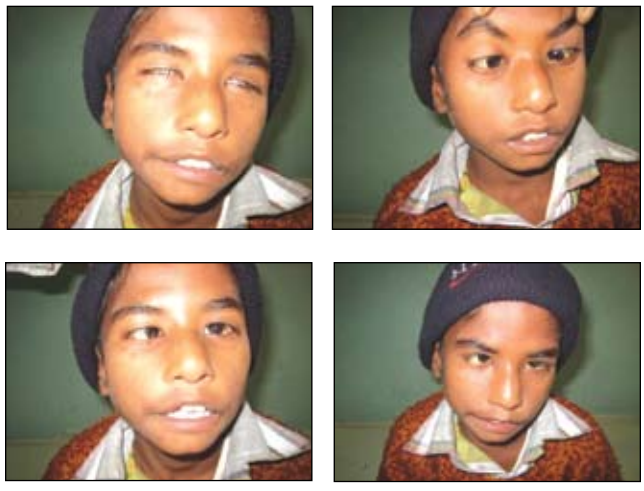


Figure showing, Lagophthalmos(A), Large angle esotropia in primary gaze(B), Bilateral abduction limitation (B,C), Limitation of elevation in right eye(D)

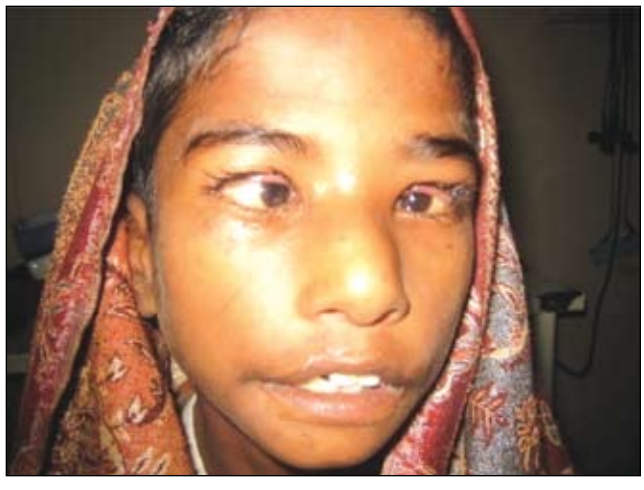


FIG 1st Day Post operative photograph, of B/L entropion correction of upper lids.

FIG 1ST Day Post operative photograph after B/L Medial rectus recession and Augmented Superior rectus transposition in left eye was done.

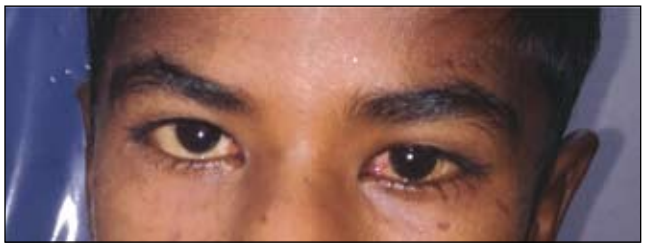
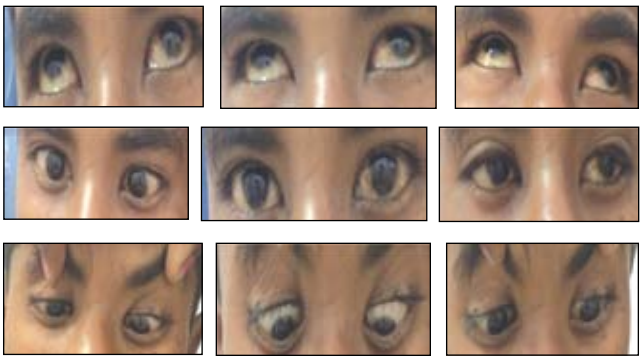


FIG 4 Weeks Post op, photograph shows good alignment in primary position

Figure showing correction of head posture and entropion



Post operative photos taken 4 weeks after B/L Medial rectus recession with augmented superior rectus transposition procedure in left eye was done.The patient still showed 20 PD of Esotropia with right hypertropia of 10 PD with mild improvement in abduction in left eye.

DISCUSSION :

Mobius syndrome is characterized by bilateral congenital abducens paralysis and facial diplegia.

The patients of Moebius syndrome usually are referred to ophthalmologists for esotropia. The few published results of strabismus surgery in patients with moebius syndrome advocate surgical options in the form of bilateral medial rectus recession, medial rectus recession with lateral rectus resection, transposition of vertical rectus muscles. The large esotropia in our case was corrected using large bilateral medial rectus recession (7mm) along with augmented superior rectus transposition to improve abduction.Sunetal in 2011 describe the surgical outcome in 3 patients of moebiussyndrome who presented to them with moderate to large angle esotropia with the augmented vertical rectus muscles transposition[14]. In the absence of medial rectus muscle contracture, the augmented VRT procedure alone appears to be more efficacious in correcting moderate- to large-angle esotropia in patients with Möbius syndrome compared with medial recession alone. However, when significant medial rectus muscle contracture is present, medial rectus muscle weakening by either chemodenervation or surgical recession is likely to be required[14].

Till date there is no published literature on advocating superior rectus transposition, as equally efficacious but more safer strategy in terms of preventing anterior segment ischaemia in treating esotropia in case of moebius syndrome. In our case there was medial rectus contracture present, so we did bilateral medial rectus recession along with unilateral augmented superior rectus transposition in the hypertropiceye. Our decision of preferring augmented superior rectus transposition over both vertical recti transposition was that Superior rectus transposition allows the option for simultaneous medial rectus recession in those patients presenting with severe abduction limitation, and require transposition surgery. Combining superior rectus transposition with medial rectus recession improved esotropia, abduction limitation, stereopsis, compensatory head posture in many of the cases of sixth neve palsy or duanes' syndrome reported by authors.[17,18]

DUANE'S RETRACTION SYNDROME:

This syndrome was first described by Stilling (1887) and Turk (1896) and is also known as the Stilling-Turk-Duane's syndrome. Duane (1905) discussed the disorder in more detail and it became generally known as Duane's retraction syndrome (DRS).

Duane's syndrome is a congenital eye movement disorder due to misdirection of the nerve fiber on eye muscle causing some eye muscle to contract when they shouldn't, vice versa can be unilateral or bilateral. It is usually congenital but may be acquired. DRS affect females to males in ratio of 3:2.

Exact etiology remains controversial but certain factors are thought to be responsible for its causation. Mechanical factors include fibrosed lateral rectus, abnormally inserted medial rectus and binding of medial rectus sheath to wall. Embryonic factors such as disturbance in normal embryonic development of 3rd, 4th, and 6th cranial nerves during second month of gestation also play a role. Due to this, there is paradoxical innervation i.e. increase innervation to both medial rectus and lateral rectus during adduction and relaxation of both medial and lateral rectus during abduction. Some studies have reported history of trauma associated with DRS.

Clinical features of DRS include limitation of abduction with or without limitation of adduction, retraction of the globe with narrowing of fissure on attempting adduction, protrusion & widening of the palpebral fissure on abduction. The patient may or may not have abnormal head posture. Possible defect of convergence is there. Strabismus may have updrift or downdrift on adduction with A or V pattern. Amblyopia may be there, if child has not adopted a head posture. Generally forced duction test (done by ophthalmologist) comes positive.

CLASSIFICATION:

Huber's Classification:

Clinically, it is often subdivided into 4 types with

associated symptoms:

- 1. Type 1
- 2. Type 2
- 3. Type 3
- 4. Type 4

Each of the group can be further classified into 3 subgroups depends on where the eyes are when on the primary gaze:

- Subgroup A (Effected eye looks esotropia)
- Subgroup B (Effected eye looks exotropia)
- Subgroup C (Effected eye looks almost orthophoria)

Type I the most common, is characterized by:

- Limited or absent ABDUCTION
- Normal or mildly limited adduction.
- In the primary position, straight or slight esotropia.

Type II, the least common, is characterized by:

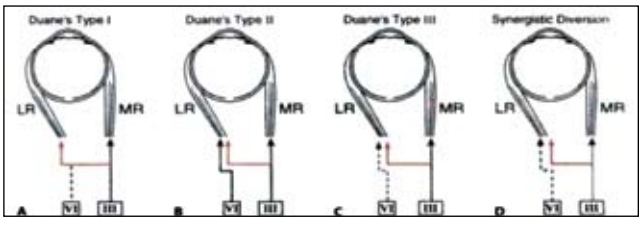
- Limited ADDUCTION.
- Normal or mildly limited abduction.
- In primary position, straight or slight exotropia.

Type III is characterized by:

- Limited ADDUCTION AND ABDUCTION.
- In the primary position, straight or slight esotropia

Type IV is very rare, characterized by:

- Synergistic diversion
- Eye paradoxically abducts on attempt adduction
- In primary position, slight exotropia.



TYPE 1	TYPE 2	TYPE 3	TYPE 4
-Poor abduction, good adduction -agenesis of 6th nerve -3rd nerve split innervate LR, MR -adduction intact as most nerve goes to MR	-Poor adduction, good abduction -6th nerve intact -3rd nerve split innervate LR, MR -Poor adduction as LR contract against MR	-Poor adduction, poor abduction -6th nerve agenesis -3rd nerve split innervate LR, MR -The split is equal -Eye not moves in/out	-Paradoxical abduction on attempt adduction -6th nerve agenesis -3rd nerve split innervate LR, MR -most innervate LR -when ADD it ABD

The prevalence of DRS is about 1/1000 in general population and it accounts for up to 4% of all strabismus cases. In fact, it is the most common type of congenital aberrant ocular innervation. 80% of cases occur unilaterally, with a left eye predominance.

Many syndromes are associated with DRS such as:

Okihiro's syndrome: Duane syndrome and radial ray defects.

Goldenhar syndrome: malformation of the jaw, cheek and ear, usually on one side of the face.

Wildervanck syndrome: Duane syndrome, Klippel-Feil anomaly, and deafness.

Moebius syndrome: congenital paresis of facial and abducens cranial nerves.

Holt-Oram syndrome: abnormalities of the upper limbs and heart.

Morning Glory syndrome: abnormalities of the optic disc.

CASE SERIES:

CASE 1:

A 10 year old boy presented to our OPD with complaints of limitation of abduction in left eye and eye becomes smaller on abduction. On examination, there was small face turn of 15 with slight esotropia of 18PD on distance and 10PD on near and limitation of abduction (-4) in left eye. There was moderate globe retraction of 50%. On adduction, there was upshoot. (Fig 1). On orthoptics esotropia for distance was 18 BO and near was 10 BO without head posture.

Esotro Duanes syndrome:

This syndrome was first described by Stilling (1887) and Turk (1896) and is also known as the Stilling-Turk-Duane's.

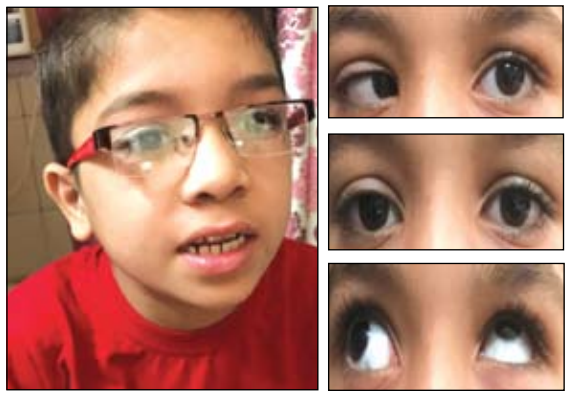


Fig1 – limitation of abduction in left eye with typical upshoot on adduction .

Left eye MR recession with Y-split of LR was done. Post-operatively the patient was orthotropic with reduced upshoot. (Fig 2).

Postop

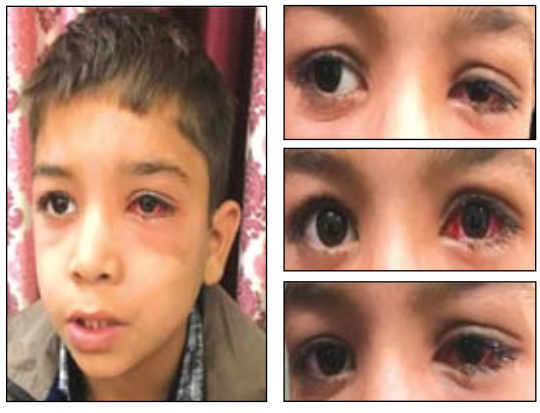


Figure 2- shows no upshot on adduction and orthotropia in primary position.

CASE 2:

A 4 year old male child had right eye mild abduction limitation and severe globe retraction (60%) on adduction. There was typical upshoot on adduction with severe globe retraction of 60%. His BCVA was 6/12 in right eye and 6/9 in left eye. On PBCT, there was slight right exotropia of 20 BI on distance and 16 BI on near. (Fig 3).

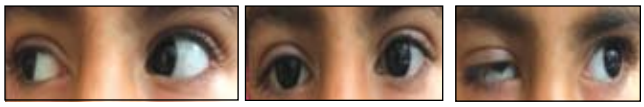
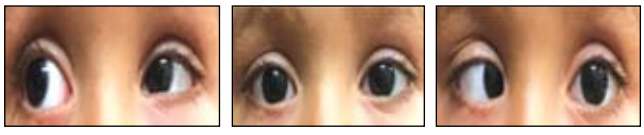


Figure 3- presence of exoduanes in right eye with upshoot and severe globe retraction with mild limitation of adduction

CASE 3:

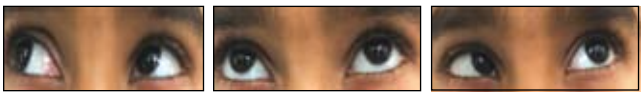
A 4 year old boy presented with Eso-DRS in left eye. There was limitation on abduction in left eye with globe retraction on adduction. On PBCT, there was esotropia of 25 PD. (Fig 4).



(Fig 4)

CASE 4:

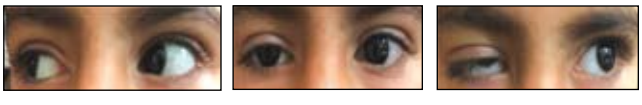
6 year old boy had eso-DRS. There was limitation of abduction in left eye, but no globe retraction. The esotropia came out to be 16 PD on measurement. BCVA in both the eyes was 6/6. (Fig 5).



(Fig 5)

CASE 5:

A 23 year old female patient presented with right convergent squint since birth. On examination, right eye had strabismic amblyopia with esotropia of 40 PD. There was limitation of abduction (-2) in right eye with globe retraction of 60% on adduction. (Fig 6).



(Fig 6)

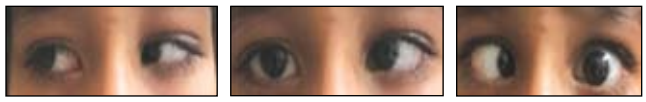
CASE 6:

A 24 year old boy had bilateral DRS. In primary position, the eyes were orthotropic with limitation of abduction (-4) in both the eyes. There was no globe retraction or upshoots on adduction. (Fig 7).

(Fig 7)

CASE 7:

A 11 year old girl presented with eso-DRS in left eye. BCVA in right eye was 6/12 and in left eye 6/36 with horizontal nystagmus. There was abduction limitation (-4) with mild globe retraction on adduction. (Fig 8).



(Fig 8)

DISCUSSION

Although DRS is a complicated squint, simple horizontal rectus muscle recession may have beneficial effect. Sethi and associates reported that unilateral or bilateral horizontal rectus recession is effective for correction of abnormal head posture. For vertical shooting of eye, usually Y-splitting of lateral rectus muscle is performed. Recently, novel techniques such as superior rectus transposition and orbital fixation of the lateral rectus have been introduced with good acceptance.

CONCLUSION

Duane's retraction syndrome like any other type of strabismus, is associated with social stigma and cosmetic blemish. By doing detailed work-up in such case, DRS can be timely diagnosed and can be properly corrected. In this way , we can overcome the adverse situations associated with DRS like ductions limitation, upshoots, downshoots, globe retraction and amblyopia.

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Editor's note :-

The CCDDs share vast heterogeneity in their phenotypes.The patient described under CFEOM is likely not a typical presentation. When confronted with atypical presentations, it's imperative to consider differential diagnoses like congenital bilateral 3rd nerve palsies, bilateral congenital Brown's syndrome, Craniofacial disorders, chronic progressive external ophthalmoplegia, and bilateral Duane's syndrome among other possibilities.

Demystifying Dissociated Vertical Deviation

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It is an independent dissociated movement of one eye with respect to another. Hering's law of equal innervation is violated here. The other eye doesn't move when the deviated eye returns for refixation. Other terms used to describe DVD are alternating hyperphoria/hypertropia, anatopia, double hypertropia, occlusion hypertropia, alternating sursumduction, dissociated double hypertropia, dissociated alternating hyperphoria, and dissociated vertical divergence.[1,2] The term dissociated vertical divergence was given by Bielschowsky (1938).

There are three components of this complex that Wilson has termed as the dissociated strabismus complex.[3] (Figure 1)

- Upward excursion - Dissociated vertical deviation
- Excyclotorsion - Dissociated torsional deviation
- Lateral movement- Dissociated horizontal deviation

All these three components can occur independently or coexist.

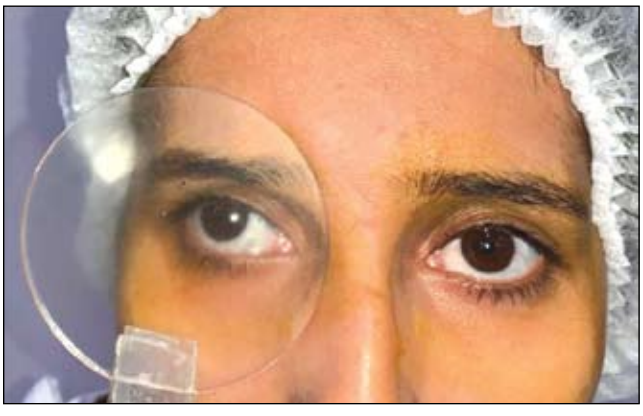


Figure 1 : the eye under cover shows upward deviation and excycloduction Picture credits: Dr. Sumant Vinayak Sharma

ETIOPATHOGENESIS

Failure to develop binocularity can lead to abnormal persistent immature monocular circuits. These can be either visual or vestibular. Vertical pursuit asymmetry can be noted in DVD patients with better pursuits directed when down to up than vice-versa. Variability of DVD with postural changes suggest involvement of vestibular or otolithic pathways. In a patient with strabismus, where binocularity is disturbed these biases become manifest. Imbalance of binocular stimulation is postulated by Spielmann[4] whereas alternate and intermittent excitation of subcortical centres governing vertical divergence was suggested by Bielschowsky.[5]

According to Brodsky[6], unequal visual stimulus causes DVD, a manifestation of primitive dorsal light reflex.

Dorsal light reflex: When light stimulus falls from one side of fish, it mis-registers it as its subjective vertical meridian with a counterclockwise tilt. Therefore occurs a clockwise tilt for equal binocular stimulation. This righting reflex causes vertical divergence of eyes to prevent the corrective body tilt. Therefore downward movement of ipsilateral eye and upward contralateral eye occurs (Figure 2).

Other theories are manifestation of atavistic oculomotor reflexes present in birds and fishes, elastic preponderance[7] and paresis of elevators[8] or depressors(bilateral paresis), oblique muscle induced cycloversion as a nystagmus dampening mechanism and abnormal visual pathway routing as seen in albinism[9].

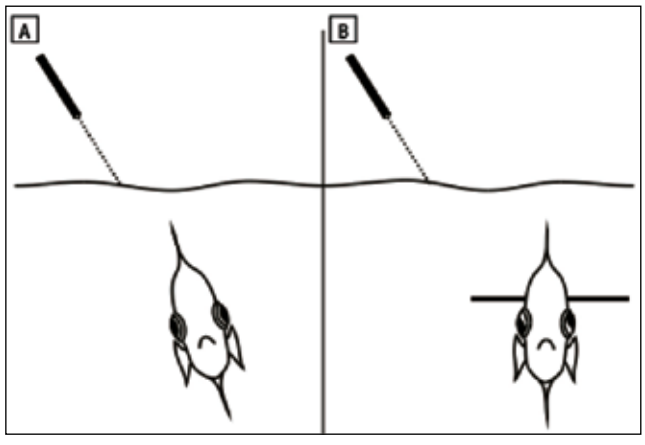


Figure 2 : Schematic diagram eliciting dorsal light reflex.

Picture source: Brodsky MC. Dissociated Vertical Divergence: Perceptual Correlates of the Human Dorsal Light Reflex. Arch Ophthalmol. 2002;120(9):1174–1178. doi:10.1001/archopht.120.9.1174

CLINICAL PRESENTATION

It is seen as a spontaneous upward drifting of non-fixing eye, especially during day dreaming or disruption of fusion as seen in eye under cover. The eye under cover drifts up and out (elevation and excycloduction) with characteristic slow recovery on uncovering.

It is seen in association with essential infantile esotropia more often and less often with acquired accommodative esotropia, sensory heterotropia, exotropia and Duane's syndrome.[10,11] It is generally a bilateral and asymmetrical phenomenon but can also be unilateral in deep amblyopes.

Abnormal head posture is noted in some patients with DVD. It can be seen as contralateral head tilt or chin depression in bilateral cases.[12]

At times, elevation of eye can be more in adduction as nose can occlude visual input in that position. It can resemble inferior oblique overaction and needs to be differentiated from the same. In children under 2-3 years of age, underdevelopment of nose can be a reason for less frequency of upshoot in adduction.

DVD needs to be differentiated from inferior oblique overaction. Presence of anatomical torsion in primary position can direct the diagnosis towards inferior oblique overaction (IOOA). Few other differentiating points are summarized in the table below.

IOOA VS DVD

	DVD	IOOA
Elevation	From primary position, adduction and abduction	Maximal in adduction, never in abduction
Superior oblique action	May overact	Usually underaction
V pattern	Absent	Often present
Pseudoparesis of contralateral superior rectus	Absent	Present
Incycloduction on refixation	present	Absent
Saccadic velocity of refixation movement	10 deg–200 deg/s	200 deg –400 deg/s
Latent nystagmus	Often present	Absent
Bielschowsky phenomenon	Often present	Absent

In comitant DVD the vertical deviation measures the same in abduction, primary position and adduction, on the contrary in incomitant DVD there is a difference in the vertical deviation in primary position, abduction and adduction. On the basis of vertical deviation, DVD can be divided into mild (0-9PD), moderate (10-19PD) and severe (>20PD).

MEASUREMENT OF DVD

Accurate measurements of DVD is a very difficult task in view of variable nature of the same.

Spielmann's translucent occluder : upward and outward movement of the eye under cover helps in the diagnosis and demonstration of this condition. +4D lens also serves the same purpose.[13]

Prism Bar Under Cover Test: Base down prisms are held infront of nonfixing eye under occlude. Alternate occlusion is done till the recovery movement is neutralized. Same procedure is repeated in the other eye as well. In cases with associated inferior oblique overaction, the difference between total upward drift by PBUCT and hypotropic refixation movement by PBCT is contributed by DVD.

Red filter test: Red image is always lower in the eye occluded by red filter in case of DVD as the eye drifts upwards after dissociation with red filter. This also helps differentiate DVD from hypertropia.

Graded Neutral Density Filter test: Filters of increasing density are held infront of fixing eye. This reduces the visual input to the fixating eye which triggers an abnormal innervation to elevators to maintain fixation. This elicits compensatory innervation in depressors in contralateral eye and subsequently shifts downwards, sometimes even below the primary position.

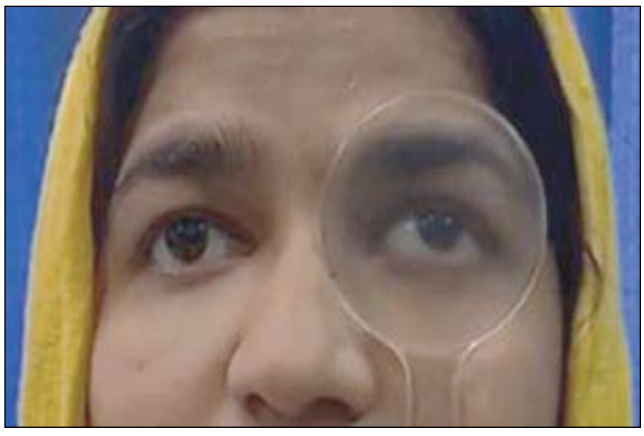


Figure 3a : DVD seen as upward deviation of non fixing eye (left) under cover (Spielmann's translucent occluder.

Figure 3b : On holding graded density filter infront of fixing eye (right), downward shifting of nonfixing (left) can be seen.
Picture credits: Dr. Anju Bhari

TREATMENT

Nonsurgical treatment is generally advised in asymptomatic patients, latent DVD and small deviations with infrequent manifestation.[1,2]

Non Surgical	Surgical
Observation No significant decrease in horizontal deviation for as long as 7.3 years	Superior recti-Large recession Retro equatorial myopexy Y Split
Encourage Fusion – Optimal Refractive correction Correcting associated horizontal deviation	Inferior recti – Resection Plication

Switching fixation Occlusion of fixing eye Fogging good eye with plus lens	Inferior Oblique – Total antero-positioning

Indications of Surgical management

- Increasing frequency of manifest phase of DVD in patient with peripheral fusion especially in patients with a high risk of amblyopia
- Patients with anomalous head posture
- Large and frequent DVD

SURGICAL MANAGEMENT

Superior rectus recession

Superior rectus recession is the most commonly used surgical technique, the superior rectus muscle is hooked and dis-inserted from the insertion. The muscle is then re-inserted at the desired site. A 7mm recession of superior rectus is usually performed for the correction of DVD. To further enhance the effect of superior rectus posterior fixation sutures can be added.

Inferior rectus resection

Inferior rectus resection involves the strengthening and shortening of inferior rectus after disinserting the muscle. The procedure is not commonly performed.

Inferior rectus plication or tuck is another example of a muscle strengthening procedure in which the muscle is not disinserted, thus avoiding the disruption of blood supply to the anterior segment, and reducing the risk of anterior segment ischemia. The procedure is also reversible adding an extra advantage over resection.

Inferior oblique total anterior positioning

Inferior oblique total anterior positioning is usually performed in patients of DVD with inferior oblique overaction. Total anterior positioning involves anterior transposition of inferior oblique near the insertion of inferior rectus. Inferior oblique muscle is converted from an elevator into a depressor after the anterior transposition procedure. Thus, bringing the eyes down.

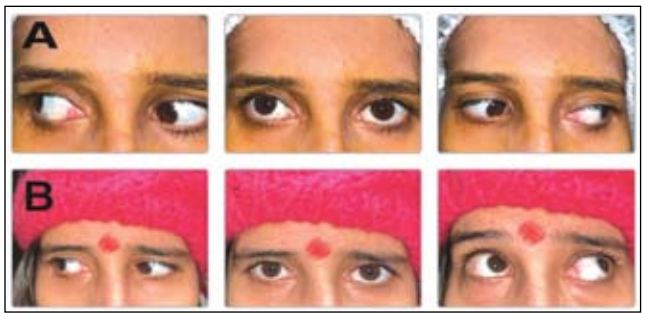
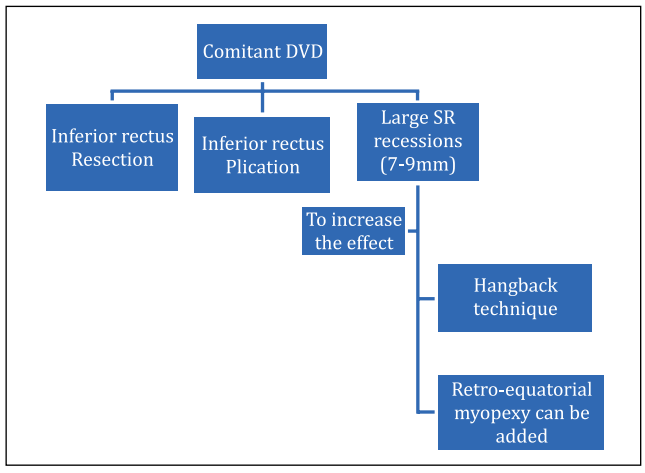


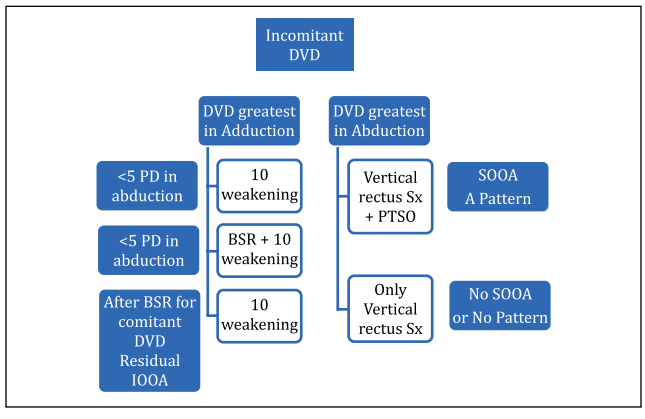
Figure 4: Manifest DVD. A) Hypertropia of right eye more in abduction B) 6 weeks post after IR plication

Complications of Surgery

- Recurrence of DVD.
- Residual DVD.
- Large asymmetric SR Recessions may lead to vertical imbalance, resulting in a hypotropia.
- Up gaze limitation.
- Pseudo IO overaction can be seen after SR recession due to fixation duress of contralateral IO.
- Palpebral fissure changes
  - Widened PF, ptosis with SR surgery.
  - Narrow PF with IR resections.



In case of unilateral surgery, latent DVD in other eye can become manifest, in which we may need to operate other eye as well. Asymmetric SR recessions (7-10 mm) are done in case of bilateral DVD with greater amounts of recessions in manifest DVD.



Helveston syndrome

- It is a triad of A- pattern exotropia, superior oblique overaction and DVD.[15]
- Four oblique surgery (Bilateral PTSO with IO ANT) along with horizontal muscle recession/resections for exotropia.[16]

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Role of Omega Fatty Acids in Modifying Visual Plasticity as an Emerging Treatment in Amblyopia

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INTRODUCTION:

Nowadays amblyopia is considered as the result of a lack of normal plasticity. By gaining some knowledge of neuroplasticity, the factors that modify the opening and closure of critical periods will lead to new therapeutic modifications which can lead to recovery of visual functions in both children and adults with amblyopia. Various treatment modalities (Refractive correction[1], Occlusion therapy[2], Penalization[3], Drug therapy[4-5], Home vision therapy[6], Refractive Surgery[7], pleoptics, CAM stimulator, red filter[8-9], acupuncture[10-11], transcranial magnetic brain stimulation[12], Perpetual learning[13], Near visual activities in the form of Television Games, Smart glasses and mobile games[14-15] have been tried, but none of them is fool proof. The ratio between excitation (glutamate receptors) and inhibition (GABA receptors) must be increased in order to recover from amblyopia by reducing intracortical inhibition and to restore visual plasticity which is considered as a critical factor. Omega fatty acids act on hippocampal neurons by stimulating glutamatergic synaptic activities and inhibiting GABA receptor-mediated responses.[31-32] Omega fatty acids have been tried & found useful in treatment of dry eye, glaucoma & ARMD[16-18]. The belief that there are specific "brain foods" dates back quite a long way––or at least as far back as the lifetimes of our grandmothers who fed us cod liver oil to "cure all ills and make us smart." Today, we see an increasing abundance of research about the positive systemic and ocular effects of omega-3 fatty acids.[19-22] This study of omega fatty acids in treatment of amblyopia based on antioxidant, anti-inflammatory, neuro protector, anti apoptotic, trophic stimulus and neuronal differentiation effects of omega fatty acids in treatment of amblyopia.

METHODS

A prospective, randomised, interventional study included thirty two patients aged 5- 12 years of age with unilateral amblyopia or bilateral amblyopia. One group (A) of sixteen patients were prescribed occlusion therapy and the other group (B) received 1000 mg per day of omega fatty acids along with occlusion for a period of three months. Follow-up assessments included best corrected visual acuity (BCVA) (both distance and near) and stereoacuity measurements at 1 week and end of three months. All improvements in visual acuity were assessed from best corrected visual acuity i.e. visual acuity after 6 weeks of initial spectacle use was compared using unpaired t-test between groups and paired t-test within each group at various follow-ups. The qualitative variables were expressed as frequencies/percentages and compared using Chi-square test. A p-value < 0.05 was considered statistically significant.

RESULTS:

Mean age of the patients was 8.68 ± 1.55 years. The mean baseline visual acuity for distance was 0.85 ± 0.071 Log MAR e.q. for Group A and 0.81 ± 0.073 Log MAR e.q. for Group B. At the end of therapy, the mean visual acuity was 0.48 ± 0.91 Log MAR e.q. and 0.40 ± 0.20 Log MAR e.q. for Group A and B respectively. Within the Groups there was statistically significant improvement in vision Group A (P value-0.0008) and Group B (P value-0.0001). When both groups were compared results were insignificant (P value-0.373).

DISCUSSION

Visual cortical dominance by the better eye leads to correspondent visual deprivation of the representations related to the eye with worse acuity. Knowledge of neuroplasticity and the factors that control the opening and closure of critical periods will lead to new therapeutic strategies which may allow for greater recovery of visual functions in both children and adults with amblyopia.

Omega fatty acid has proven effects for retinal maturation, visual acuity development through several randomized trials.

Omega-3 fatty acids were provided to a group of students in a school in England and Australia, has shown improved school performance in the form of verbal intelligence, reading, learning and memory in comparison to students who didn't receive supplementation in the form of omega fatty acids.[19]

Neuroprotection is provided by DHA due to their anti-oxidative and anti inflammatory actions as a result, of these properties they reduce levels of reactive species(nitric oxide), pro-inflammatory mediators and by maintaining higher levels of GSH and anti-oxidant enzymes (i.e. glutathione peroxidase and glutathione reductase). These above mentioned effects of DHA leads to reduction in glutamate induced cytotoxicity.

James R drover et al studied the effects of DHA on language development and school readiness. School

readiness and receptive vocabulary was assessed at 2 and 3.5 years respectively. Dietary DHA during the first year of life did not enhance school readiness or language development.[23]

Birch et al determined whether the dietary supply of LCPs after weaning influenced the maturation of visual acuity (measured through Sweep VEP) and stereoacuity (randot stereoacuity). Infants who were provided with the formula that contained LCP's had significantly better visual acuity and stereoacuity at 17, 26, and 52 weeks of age, as compared to those who did not receive the formula. In their study better acuity and stereoacuity at 17 wk was correlated with higher concentrations of docosahexaenoic acid in plasma. Better acuity at 52 wk was correlated with higher concentrations of docosahexaenoic acid in plasma and red blood cells.[24]

In another study, Birch et al tested the hypothesis that infant formula lacking LCPs during the first 17 weeks of life would result in visual acuity and IQ at 4 years of age that was significantly poorer than those who had dietary supply of LCPs during the first 17 weeks of life .

Several randomized trials are conducted and found specific benefit of omega fatty acids supplementation for retinal maturation, visual acuity development, or cognitive development.

POSSIBLE MECHANISMS:

Neuroplasticity is the ability of the brain to reorganize and modulate the structure and function of its connections in response to any change in the environment .The brain is considered plastic and neural networks are initially modified by certain experiences during the sensitive period and subsequently stabilized during normal development. The beginning of the critical period for plasticity in the visual system is established by the experience-dependent maturation of GABA mediated inhibition. Hence, a reduction of inhibitory transmission halts the onset of the critical period for visual cortex plasticity during early period of life. In the adult visual cortex, the limited neuroplasticity can be enhanced by previous visual deprivation, which is associated with a loss of GABA receptors, and reduced by GABAergic modulators. The major modulatory systems in the brain, that is, adrenaline, noradrenaline, dopamine, acetylcholine, and serotonin also play significant role in enhancing the visual plasticity.[26-28]

Certain morphological manipulations that reduce dendritic mitochondria lead to loss of synapses and dendritic spines. In contrast, increasing dendritic mitochondrial content as well as activity enhances the number and plasticity of synapses. Therefore, the dendritic distribution of mitochondria can be both essential and limiting for the support and further growth of synapses. Black et al concluded that following treatment of omega 3 fatty acids for 24 and 48 hours, it significantly induced PGC-1α, an essential precursor for mitochondrial biosynthesis. This finding is supported by the increase in total mitochondrial content observed by both flow cytometry and microscopy. This suggests that they are effective at increasing mitochondrial number, density and networking without altering mitochondrial activity.[29-30]

It has been demonstrated that GABAergic signaling is a rate limiting step in development of visual cortex plasticity and was observed that a pharmacological decrease of inhibitory transmission effectively restores ocular dominance plasticity even in adulthood. It has thus been hypothesized that a critical factor in restoring plasticity and inducting recovery from amblyopia is to increase the ratio between excitation (glutamate receptors) and inhibition (GABA receptors) by reducing intracortical inhibition.

Kevela et al demonstrated that DHA enhances glutamatergic synaptic activities with simultaneous increase in synapsin and glutamate receptor subunit expression in hippocampal neurons. On the other hand, lack of DHA results in inhibition of synaptogenesis, decreases in synapsins and glutamate receptor subunits, and impairment of long-term potentiation in hippocampal neurons.[31]

Nabekura et al concluded that DHA inhibits GABA receptor-mediated responses in cultured neural cells in a concentration and time-dependent manner. The effect of DHA on GABA receptor is linked to the effect on the lipid microenvironment for the GABA receptors.[32]

An altered internalization of dopamine (DA) in the storage pool of the frontal cortex has been seen in chronic n-PUFA deficiency. These alterations in synaptic membrane DHA have an impact on DA synaptic neurotransmission and plasticity.[33]

It has been demonstrated that brain plasticity is modulated by cholinergic transmission, and that it may be possible to modulate and stimulate plasticity by manipulating cholinergic pathways, potentially reopening the window for effective amblyopia therapy after the ''critical period'' has passed. DHA deficiency results in decrease of muscarinic receptor binding, although acetylcholinesterase activity and the vesicular acetylcholine transporter are not affected.[34]

DHA supplementation normalizes levels of brain-derived neurotrophic factor (BDNF) by elevating brain DHA concentration. BDNF is one of the main factors contributing to the onset of the sensitive period by modifying the development of inhibitory innervation. More recently, it was found that insulin growth factor-1 (IGF-1) has similar effects as BDNF, accelerating the development of inhibitory synapses, the onset of the sensitive period and the increase of visual acuity.[35]

Docosahexaenoic acid (DHA) is an integral component of neural membranes and is present in 30-40% of the phospholipids in the gray matter of cerebral cortex and photoreceptor cells in the retina. It mediates its molecular and cellular effects not only through regulation of physicochemical properties such as membrane fluidity, permeability and viscosity in synaptic membranes, but also via modulation and stimulation or inhibition of neurotransmission, gene expression, and activities of enzymes, receptors and ion channels.

These processes are closely associated with activation of signaling pathways thus sustaining synaptic function and neuronal survival.[36-37]

CONCLUSION:

We did not find any beneficial effects of use of omega fatty acids in treatment of amblyopia in terms of distance visual acuity, near visual acuity and stereopsis. As more reports give an encouraging trend towards role of pharmacological therapy, it is an area that needs to be further explored. We recommend further studies on the subject matter.

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Clinico-Demographical Profile of Strabismus Cases at Department of Ophthalmology of A Rural Medical College

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Abstract :

Introduction: *Strabismus, also commonly known as squint or crossed eye or walled eye is a condition where a person cannot align both eyes simultaneously under normal conditions. Clinico-Demographical profileis likely to vary from area to area in rural and urban population and so need to be studied.*

Aim of study: *To evaluate the Clinico-Demographical profile of strabismus patients presenting to Department of Ophthalmology of a Rural Medical College.*

Material and method: *This is a combined retrospective, prospective hospital-based observationalstudy that evaluates all patients who visited a tertiary care center from March 2019 to March 2021, to define the clinical and demographic pattern of strabismus across all ages.Visual acuity, cycloplegic retinoscopy, stereopsis assessment were done in all patients. Strabismus was objectively measured using Prism Cover Test / Modified Krimsky method.*

Observations: *Out of a total of 14500 patients that visited the Dept. of Ophthalmology during the specified period, 120 patients were diagnosed with strabismus, 75 were males and 45 were females. Exotropia was our most frequently encountered strabismus entity.Refractive assessment of the patients revealed hypermetropia to be the most common refractive error and 35.8% patients were diagnosed with amblyopia.*

Conclusion: *This study helps the Ophthalmologists to know the incidence, various prevailing causes, laterality, socioeconomic status of the affected patients and about the age and sex distribution of strabismus, so that they can plan for a well equipped strabismus clinic with trained persons to lessen the menance of this socially cursed disease for which many people are suffering since a long time in this part of the country*

Keywords: *Strabismus, Farsightedness, Ocular motility, Binocular Single Vision*

INTRODUCTION:

Strabismus (squint) is not the infrequent cause of ocular morbidity. It typically affects children in the early years of life and subsequently may result in vision loss and impaired binocular function. This has been given due attention in the recent thrust on pediatric ophthalmology in the Vision 2020 initiative.[1] Society is prejudiced against strabismic individuals, often underestimating their Intelligent Quotient and stereotyping them.[2] It is of utmost importance to understand that the general societal standards classify strabismus as a cosmetic disadvantage only, failing to realize the functional aspect of this disease.[3,4] The patients are viewed as inferior to the peers with less appeal as potential friends and partners. The main goal of treatment in strabismus is restoration of eye alignment together with fusion and stereo acuity. Additional favorable outcomes include improvement or elimination of an anomalous head posture, expansion of binocular visual fields and elimination of double vision.[5] In adulthood, presence of strabismus may lead to psychological problems such as poor self-esteem, depression, poor interpersonal relationship and poor job opportunities because of cosmetically undesirable appearance.[6] So the patients with strabismus are suggested to have their eyes aligned optically or surgically to maximize their binocular vision outcome.[7] Strabismus does not get cured on its own. Therefore, treatment should be initiated as soon as it is diagnosed or at least as early as possible. Early diagnosis and treatment help us achieve the best binocular vision possible and diminishes psychological problems. A neglected case of strabismus may lead to a condition of loss of binocular single vision. While the literature abounds in studies discussing the prevalence of strabismus, both globally and in India, there are few hospital-based studies that provide the complete clinical- demographic profile of strabismus, including subtypes.Therefore, an effort has been made to study the clinico-demographical profile of strabismus patients presenting to Department of Ophthalmology of a rural Medical College.[8]

MATERIALS AND METHODS

Strabismus patients visiting rural tertiary eye care referral center in SGT Medical College, Hospital and Research Institute from March 2019 to March 2021, across all age groups were included in this study. Thorough clinical workup including visual acuity, cycloplegic retinoscopy, stereopsis assessment was done

in all patients. For measurement of squint, the prism bar cover test was applied in patients with both eye vision ≥20/30 having central fixation; the Krimsky test was used in patients with low vision in one eye (<20/200); and the Hirschberg test was used in young children, patients with bilateral poor vision, or uncooperative patients. Manifest strabismus was divided into comitant (labeled as esotropia or exotropia) and incomitant (labeled as paralytic or restrictive squint). Manifest horizontal strabismus was also classified as intermittent and constant. The presence of nystagmus was noted, as was its association with squint.

RESULTS

The present study was aimed to find out the occurrence of strabismus in a limited population visiting at SGT Medical College, Hospital and Research Institute, Budhera, Gurugram. Out of a total of 14500 patients that visited during the specified period, 120 patients were diagnosed with strabismus, 75 were males and 45 were females. Most of the patients were in between 10 to 30 years of age. Only 10 patients were above the age of 30. The numbers of patients across different age groups are shown in Table 1. Our 4 patients were born out of consanguineous marriages, 22 patients were born prematurely, and 60 patients had turbulent perinatal history including Neonatal Intensive Care Unit admission. Thirty four patients gave family history of strabismus. Table 2 depicts the distribution of different types of strabismus cases visited our tertiary eye care centre.

Table 1. Age wise distribution of patients

Sr No.	Age Distribution	No. of Patients	Percentage
1.	<10 years	14	11.7%
2.	10-20 years	39	32.5%
3.	20-30 years	57	47.5%
4.	>30 years	10	8.3%

Table 2. Distribution of different types of strabismus.

Sr No.	Attributes	No. of patients	Percentage
1.	Exotropia	63	52.5%
2.	Esotropia	25	20.8%
3.	Dissociated Vertical Deviation	09	7.5%
4.	Inferior oblique over action	07	5.8%
5.	Superior oblique palsy	06	5%
6.	Duane's retraction syndrome	04	3.4%
7.	Lateral rectus palsy	04	3.4%
8.	Third nerve palsy	02	1.6%
	Total	**120**	**100%**

Refractive assessment of the patients revealed hypermetropia to be the most common refractive error with 63% of esotropes and 2% of exotropes being hyperopic [S.E>+2 Dioptres]. Out of the total number, 19.6% of exotropes and 3% esotropes respectively were myopic [S.E<-0.5 D]. Simple and compound hyperopic astigmatism was noted in 19.5% of esotropes and 11.3% of exotropes. Amblyopia was diagnosed in 35.8% patients. Prism cover test was employed to measure the deviation objectively. In amblyopic or sensory strabismus patients, Modified Krimsky method was used. Most of our strabismus patients had large angle squints. The composite variation of the measured deviations is summarized in Figure 1.

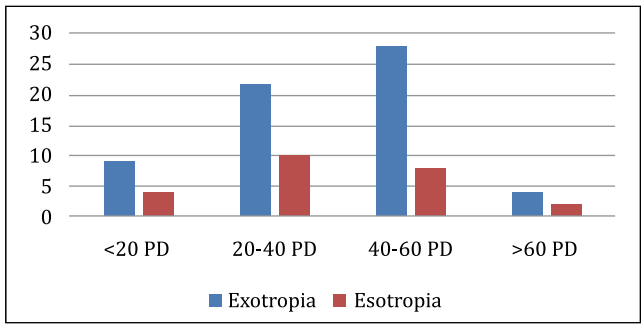

Figure 1: Showing the composite variation of the deviations assessed with prism bar cover test in patients visiting our eye care centre.

Exotropia was the most frequently encountered strabismus entity in this study. Most of our exotropia patients were diagnosed with intermittent exotropia. Stereopsis was seen in 91% of intermittent exotropes and 31.1% of accommodative esotropia patients. Out of the total 120 patients, 25 were found to have Esotropia. When the sub type analysis was undertaken, we found 27.7% patients had accommodative esotropia and 36.17% had Partially Accommodative Esotropia. Congenital Esotropia formed only 19% of our study cohort (Table 4).

Photographic record of each patient was kept, as an example, nine gazes cardinal photographs of a case having left exotropia are shown in Figure 2 and Preoperative and postoperative photographs of a patient operated for exotropiaare shown in Figure 3.

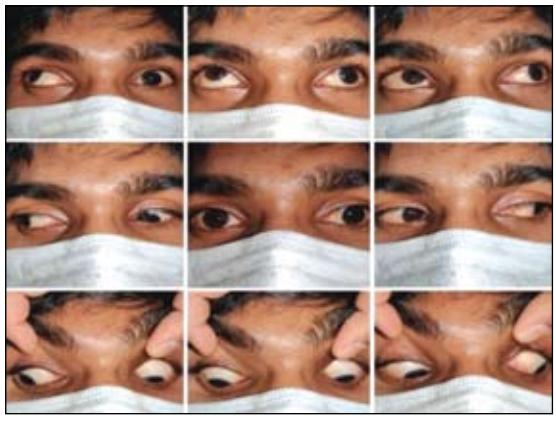

Figure 2 : Showing nine cardinal photographs of a patient visited at our tertiary eye care center showing outward deviation of left eye since birth with L/E 15-degree exotropia with hypotropia and down shoot in adduction. (V-pattern)

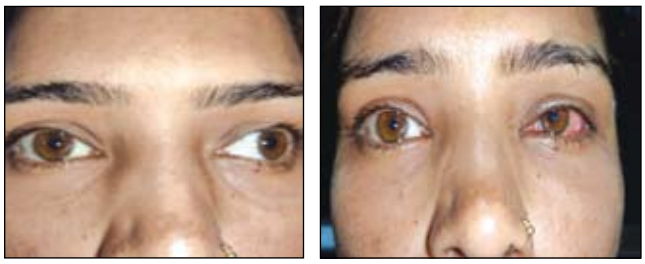

Figure 3. Showing the pre-operative and postoperative photographs of the strabismus surgery of a patient with exotropia.

Table 3. Distribution of exotropic patients

Characteristics of exotropia patients (63)	Percentage
Intermittent exotropia	57.4%
Constant exotropia	25.8%
Sensory exotropia	11.5%
Convergence insufficiency	5.3%

Table 4. Distribution of esotropic patients

Characteristics of esotropia patients (25)	Percentage
Accommodative esotropia	27.7%
Partially accommodative esotropia	36.15%
Essential infantile esotropia	19.2%
Non refractive late onset esotropia	10.2%
Sensory esotropia	6.9%

DISCUSSION

The study aimed to note the clinical and demographic profile of patients visiting the outpatient department and squint clinic of a Rural Tertiary Level Healthcare Institution in India.In our study, exotropia was more common than esotropia. This finding was similar to others where exotropia cases were greater than esotropia.[11] According to the present study, paralytic and intermittent type of strabismus was less frequently seen, so they are not common types. In a hospital-based study in North India, 2% had intermittent exotropia and 0.8% had paralytic strabismus. The magnitude of squints among all patients seeking an Ophthalmology consultation at our institution was 6% and that for children was 26.6%. While there are no previous hospital-based studies that may be compared, population-based surveys among children demonstrate a wide range of prevalence of strabismus varying from 0.29% to 5%. This again indicates the excessive referral of paediatric strabismus even to a rural tertiary care institution due to lack of adequate facilities for management at a primary or secondary level. Many patients reach this rural centre even from surrounding big cities. So, such services in rural area are more prudent as a patient from rural area who is usually afraid of visiting hospitals in big cities can easily attend such centres. The need for developing more paediatric Ophthalmology setups and training ophthalmologists for handling such cases in developing countries including India is further emphasized by the fact that children account for over half of the total patients seen in our squint clinic. This study will help the Ophthalmologists to know the incidence, various prevailing causes, laterality, socioeconomic status of the affected patients and about the age and sex distribution of strabismus, so that they can plan for a well equipped strabismus clinic with trained persons to lessen the burden of this socially cursed disease for which many people are suffering since a long time in this part of the country. There is a significant financial and socioeconomic burden on society for developing and managing tertiary health care infrastructures. So, wide publicity should be given to such centres, so that public at large, especially from rural areas could get the best quality care.

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Retinopathy of Prematurity Aphorisms on Epidemiology and Statistics

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Abstract : *Retinopathy of Prematurity is a disorder that occurs in the non-vascularized retina of premature neonates. Blindness related to Retinopathy of Prematurity is on the rise due to improved neonatal care and increased survival of pre-term neonates. So far, the world has encountered three epidemics of Retinopathy of Prematurity- related blindness. The "first epidemic" in the 1940s and 1950s principally affected premature babies in the United States of America and Western Europe. A "second epidemic" in industrialized countries in the 1970s was a consequence of higher survival rates in premature babies. The "third epidemic" occurred in middle income countries like India and China as the rates of preterm births tend to be higher in these countries. Retinopathy of Prematurity-related blindness can be prevented by a multidisciplinary team approach, which includes obstetricians, neonatologists/pediatricians, nurses, ophthalmologists, health care workers, and parents. Recent data from Canada, the United States and the United Kingdom regarding the mean birth weights of babies needing treatment for threshold disease have continued to guide screening protocols. Through this review article, we aim to amalgamate the data on worldwide statistics and epidemiology of Retinopathy of Prematurity, and the ever-changing trends in the screening and management of the same. It is of paramount importance to create awareness amongst Pediatricians and Neonatologists, as well as Ophthalmologists so as to reduce the burden of avoidable blindness.*

Keywords: Retinopathy of Prematurity, epidemics, statistics, epidemiology, screening, blindness.

INTRODUCTION:

Retinopathy of prematurity (ROP) is an important avoidable cause of childhood blindness. Currently, middle income countries like India and China are experiencing a third epidemic of ROP blindness due to increased survival of preterm babies with presence of variable neonatal intensive care and ophthalmic coverage for screening.

- The "first epidemic" (of blindness) occurred in the 1940s and 1950s and principally affected premature babies in the US and to a lesser extent, Western Europe. At that time unmonitored supplemental oxygen wasthe principal riskfactor.[1]
- A "second epidemic" (of acute ROP) in industrialized countries started in the 1970s, as a consequence of higher survival rates in extremely premature babies[1]—larger, more mature babies were surviving usuallywithout developing severe, acutedisease.[2]
- The "third epidemic" of ROP blindness has several explanations. Firstly, rates of preterm birth tend to be higher in middle income countries thanin high income countries, particularly in Latin America where teenage pregnancies arecommon.[1]

The first epidemic of ROP occurred due to unrestricted oxygen use and second epidemic due to increased survival of very preterm babies in high-income countries. India and other middle-income countries are facing the third epidemic of ROP due to various factors such as increased survival of preterm babies, inadequate quality of neonatal care, and low coverage of screening and treatment services for ROP.[1]

REVIEW OF LITERATURE:

In 2010, the annual incidence of blindness and visual impairment from ROP was estimated to be 32,200 cases worldwide. India accounted for nearly 10% of all estimated worldwide visual impairment following ROP in 2010 with at least 5,000 children developing severe disease and 2,900 children surviving with visual impairment related to ROP.[3,4]

Systematic reviews and meta-analyses were undertaken by Blencowe et al. to estimate the risk of ROP and subsequent visual impairment for surviving preterm babies by level of neonatal care, access to ROP screening, and treatment. They estimated 32,300 (24,800–44,500) preterm survivors in 2010 suffered from long- term visual impairment secondary to ROP: 20,000 (15,500–27,200) with severe visual impairment or blindness and a further 12,300 (8,300–18,400) with mild or moderate visual impairment. Worldwide, only 6.2% (2,000 (1,400–2,900)) of those with visual impairment were born at 32–36 wk gestational age.[5]

In industrialized countries, two epidemics of retinopathy of prematurity (ROP) have been described. In the UK, the mean birth weight (BW) of affected babies was 1370 g (range 936–1843 g)[1], and 1354 g (range 770–3421 g) in the USA.[6]

In middle income countries, the proportion of women who deliver in health care facilities is high, and

premature babies are, therefore, likely to be admitted to Neonatal Intensive Care Unit (NICU).

Recent data from Canada, the USA and the UK show that the mean BWs of babies needing treatment for threshold disease are 759 g (range 440–1785 g), 763 g (range 415–1255 g) and 737 g (range 450–1260 g), respectively. The gestational ages of the same babies being 25.6 (range 22–32), 25.4 (range 23–29) and 25.3 weeks (range 23–32), respectively.[7] Programs for detecting ROP are well established in most countries in North America, Western Europe and in the industrialized countries of the Pacific Basin providing information on the population of babies needing treatment, and how this population is changing over time.

This information has and continues to be used to refine screening criteria, to ensure that programs are as cost effective and efficient as possible. However, these are not "screening" programs in the true sense of the word, but are "casedetection" initiatives. Screening would entail the use of a simple, safe, non-invasive and valid test which identified babies needing a "gold standard"diagnosis.[7]

Thirdly, rates of severe ROP are higher in premature babies in low and middle income countries[8,9,10] even when wider screening criteria have been used, suggesting that babies are being exposed to risk factors which are now largely controlled in industrialized countries.

Sanghi et al. studied demographic profile of infants presenting as stage 5 retinopathy of prematurity (ROP) at a tertiary referral center of North India. According to the study, mean birth weight of 66 infants was 1250.23 ± 486.45 g and gestational age at birth was 28.5 ± 2.2 weeks. Ten (15.1%) infants were above 1500 g birth weight. Median age at presentation was 7 month (range, 2-84 months).[11]

Approximately 15 million babies are born preterm worldwide each year and India has the highest number of preterm births.[11] In 2010, there was an estimate of 3,519,100 preterm births in India. If 30% of these babies have access to neonatal care, about one lakh babies are found to survive each year who are at risk of developing ROP and requiring screening.[13]

To address this vast disparity, newborn health had captured the attention of policy- makers in India. This resulted in strong political commitment to end preventable newborn stillbirths and deaths and also to recognize newborn health as a national development necessity.

As a result, a nationwide network of facility-based newborn care was established at various levels: 14,135 newborn care corners at the point of child birth; 1810 Newborn Stabilization Units; 548 Special Newborn Care Units (SNCUs) for sick and small newborns, with care to more than 6 lakhs newborns being provided in SNCUs each year. The number of these units has increased over a period of time.[14]

This led to increased survival of babies who are at risk to develop blindness due to ROP.[11]

India accounts for nearly 10% of the worldwide estimate of blindness and visual impairment due to ROP. It has been reported that majority of babies presentwith stage 5 disease due to lack ofscreening.[12]

The 11-City 9-State study done to assess the services for ROP in India recommended that eye care services for ROP need to expand. Low screening rates are also due to lack of awareness among neonatologist/pediatricians and non- availability of trained ophthalmologists.[13]

ROP-related blindness can be prevented by a multidisciplinary team approach, which includes obstetricians, neonatologists/pediatricians, nurses, ophthalmologists, health care workers, and parents.[12]

A cross-sectional study was undertaken by Azad, Chandra, Gangwe and Kumar to study the barriers to effective screening, early detection and treatment of Retinopathy of Prematurity leading to advanced disease. According to them, while 99 babies (86.1%) were referred by ophthalmologists, only 10 babies (8.7%) were referred by pediatricians.[16]

Fifty-seven (86.4 %) infants were never screened for ROP and 2(3%) were lost to follow-up after single screening. Seven (10.6%) infants had screening and treatment.[11]

Neonatologists play a central role in identifying infants who require screening for retinopathy of prematurity (ROP) and in coordinating screening and treatment when necessary.[13] While under-referral could lead to missed cases of severe ROP and blindness, over-referral could exacerbate any workforce shortage and crowd out those children at higher risk for the development of ROP.[17]

Although there is increased awareness among the neonatologists/pediatricians in the recent past, ROP screening and treatment programs are not in place in many neonatal care units. Continuous medical education programs and workshops on ROP should be conducted.[12]

A study conducted by Sathiamohanraj et al. found that only 65.1% of the pediatricians were aware about ROP. Only 39.8% thought it was preventable and 42.2% had no idea on risk factors of ROP. 41% were not aware of which part of the eye is to be examined to identify ROP and 47% did not know who needs to perform the test. 45.8% of the pediatricians were not aware regarding the period of first eye test for ROP screening. In fact, 14.5% thought that first ROP screening should be done between 6 and 12 months of age. 44.6% of the pediatricians had no idea about the treatment modalities of ROP, while 18.1% thought that ROP isnot treatable.[18]

DISCUSSION:

Major deterrent in ROP screening was perceived as non-availability of trained ophthalmologists according to a pilot survey conducted by Patwardhan et al., published in the Indian Journal of Ophthalmology in 2011.[19]

Training and awareness programs may be conducted for pediatricians/neonatologists, and ophthalmologists

together, to improve rapport among them. An effort was made by the World Health Organization (WHO) and the Ministry of Health and Family Welfare, by conducting ROP workshops initiated by R.P. Center, All India Institute of Medical Sciences, Delhi, India across thecountry.[12]

They were the first to bring neonatologists and ophthalmologists onto one platform. Training programs have also been carried out in partnership with Non-Governmental Organizations (NGOs) such as Sight savers, Orbis International, and the Queen Elizabeth Diamond JubileeTrust.[20]

Although the number of blind children is relatively low, they have a lifetime of blindness ahead, with an estimated 75 million blind years (number blind × length of life). It is estimated that at least 2–3 lakhs children in India have severe visual impairment or blindness and more than 3000 infants become blind or visually impaired each year due to lack of screening and treatment for ROP.[12]

Lee and colleagues addressed the controversy related to the guidelines recommended by the Canadian Pediatric Society (CPS) and American Academy of Pediatrics (AAP) for routine screening for retinopathy of prematurity (ROP) in 2001. They concluded that screening only infants having a BW of 1200 g or less is the most cost-effective strategy for routine ROP screening.[21]

A study by Vinekar et al. showed that the fiscal quantum of blind person-years saved is 108.4 million USD, annually by expansion of telemedicine screening for ROP to 10 states in India.[22]

CONCLUSION:

ROP awareness stands as a good example of all the strategies for prevention which includes:

i. Primary prevention: prevention of the disease by improving obstetricand neonatal care.

ii. Secondary prevention: screening and treatment programs forROP.

iii. Tertiary prevention: treating complications and rehabilitation to reduce disability.[11]

There is a necessity to create more awareness about the condition and to highlight the importance of timely screening and early detection of changes associated with it in order to undertake early treatment aimed at retarding or halting progression of the disease thus, preventing the resulting blindness.

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Tackling Problem Behaviour During Eye Examination with Autism and Autism Spectrum Disorders

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Abstract : *Autism , a neurodevelopmental disorder is characterized by impairment in social interaction and communication with restricted, repetitive, stereotyped patterns of behavior, interests, and activities. Through this review article, we have summarized the ophthalmic conditions associated with Autism Spectrum Disorder, and the evaluation and management of children affected by autism. Various challenges associated with the evaluation of autism must be addressed while simultaneously proceeding with the examination.The most common and obvious ocular abnormality is refractive error and children with autism are significantly more likely to have ocular morbidity than normally developing children. Early detection of ocular defects in autism is crucial. Pediatricians treating these patients should be encouraged to refer them for ophthalmic evaluation upon diagnosis. Physical and visual rehabilitation are important for the improvement of visual function, along with augmentative and alternative communication devices. With little extra time and resources, the examiner is able to successfully test these patients in most cases, thus easing the implementation of appropriate protocols.*

Keywords: *Autism Spectrum Disorder, Refractive error, Rehabilitation, Communication devices.*

INTRODUCTION:

Autism was first marked out by child psychiatrist, Leo Kanner in 1943.[1]] Since the 1980s, autism has been conceded as a neuro developmental disorder defined by impairment in social interaction and communication with restricted, repetitive, stereotyped patterns of behavior, interests, and activities.[2]

Many systemic conditions are bracketed with autism, including associations with intellectual disability, epilepsy, disruptive behavior, and learning difficulty.[1]

Although the strong association allying autism and genetic factors has been overlong established, the complete genetic context of autism is still unclear.[3]

Clinicians evaluate three main classes of behavior:[4,5]

1) Impairment in social interaction,
2) Impairment in communication,
3) Repetitive, stereotyped behavior patterns.

A patient can only be contemplated to have one of the Autism Spectrum Disorders (ASD) if abnormal behaviour is observed in at least one of these three areas prior to age three.

Difficulties arise to examine children with autism due to constraints in communication skills and changeable behavior.

DISORDER

Autism is characterized by:[6]

1) Persistent differences in communication, interpersonal relationships, and social interaction across different environments, characterized by being non-verbal or having atypical speech patterns, having trouble understanding non-verbal communication, difficulty making and keeping friends, difficulty maintaining typical back-and-forth conversationalstyle
2) Restricted and repetitive behavior, patterns, activities and interests, characterized by repeating sounds or phrases (echolalia), repetitive movements, preference for sameness and difficulty with transition or routine, rigid or highly restricted and intense interests, extreme sensitivity to or significantly lower sensitivity to various sensorystimuli Children with Autistic Disorder often exhibita typical gazeorgaze avoidance and manifest stereotypical behavior such as eye pressing, hand flicking, and light gazing.[1]

   Refractive error and visual acuityare highly fickle in these patients. Strabismus is often present, and pursuits and saccades have been found to be inadequate.[1]

Pointers of atypical visual development:[7]

- Absence of eyecontact,
- Benumbed and unresponsive to facialgesture,
- Difficulty in sharing joint visualattention.

A comparative study with autistic children and a normal cohort showed that looking at the eyes of others was significantly decreased in children with autism, and fixation time on eyes by the children with autism correlated with their level of social disability.[8]

Individuals with ASD have difficulties in reciprocating during social interaction, verbal and non-verbal

communication and fine processing sensory stimuli. Because of these difficulties, they may find it difficult to respond to subjective vision testing.[9]

TESTABILITY

Testability is defined as the ability of a patient to complete a vision test.

Clinically, it is determined as the proportion of patients within a defined population who are able to complete a test or procedure.

For the pre-school population, studies of vision testing have reported testability of in visual acuity[10-13], stereoacuity,[10,13,14] and refraction tests.[10,15]

SOME OF THE OBSTACLES THAT SHOULD BE ADDRESSED WITH ASD INCLUDE:

1) Communication:

- Short and concise communicating statements should be used.
- The pace of speech should allow the patient to hear, absorb, and answer.
- Augmentative and alternative communication (AAC) is any tool, strategy, or technology that compensates for, enhances, expands, or helps develop communication skills. AAC canbe unaided or aided. Unaided manual signs and gestures. Aided include communication boards, speech generating devices, keyboards, email, and instant messaging.[16,17]

2) Visual Supports:

- Visual supports can be used to enable patients to understand test instructions of the eye examination.[9]
- Yes/No Application presented on a smartphone, tablet or computer.
- Verbal directions with pictures and gestures could help patients better understand directions in eye examination.[10,11,12]

3) Social Stories:

- Social stories should be used to prepare patients for visits.
- Social stories decrease tantrums and inappropriate behaviors and increase positive behaviors such as initiating social interactions andresponding.[9]

4) Visual Schedule:

- Visual schedules use photographs or pictures to represent theactivities and procedures in clinic.
- Visual schedules help patients to make transitions, respond and reduce anxiety due to hesitancy.[18]

5) Motor Activities:[9]

- Patients have difficulties in bringing forth a motor movement to accomplishing it.
- Motor approximations can beaccepted if patients respond to a gesture,
- For example, patients responding to visual acuity testing on a picture on a response card are allowed to respond by touching the picture with their whole hand rather than required to point to thepictures.

6) Sensory activities:[9]

- Bright lights such as indirect ophthalmoscopy and tests that involve touching the patient's face and eye area such as slit lamp examination and eye dropinstillation can be challenging.
- Distraction techniques such as using colourful toys and having the examiner sing or recite the alphabet with patients while performing the procedure.

7) Behavioral tactics:

- Eliciting tolerance to a desired behavior.
- For example, a patient tolerates a indirect ophthalmoscope light, first on his legs, then his shoulders, then his face, and finally hiseyes or demonstrating the same on the attendant of the patient.
- The brightness of the light should be ambient and not too high.

Coulter et al. conducted a prospective pilot study to compare testability of vision and eye tests in an examination protocol of 9- to 17-year-old patients with ASD to typically developing peers.[9]

Ocular Health Testing must be sequenced as follows:

1. Distance Visual Acuity
2. Retinoscopy
3. Cover test at distance and near and alternate covertest
4. Near Point of Convergence(NPC)
5. Evaluation offixation
6. Negative fusional vergence atnear
7. Positive fusional vergence (Convergenceamplitudes)
8. Monocular Estimation MethodRetinoscopy[9]
9. Near Visual Acuity[9]
10. Stereoacuity[9]
11. Extraocular muscle movements andpupils[9]
12. Anterior segment evaluation andtonometry[9]
13. Instillation ofmydriatic/cycloplegic[9]
14. Posterior segment evaluation and cycloplegicretinoscopy[9]
15. Binocular Indirect Ophthalmoscopy[9]

NEED FOR PROTOCOL FOR EYE EXAMINATION:

Challenges associated with ASD include difficulties with communication, motor and sensory activities, transitioning from one task to another and maintaining attention.

An awareness of the similarity in traits means that autism-support practitioners must be wary of attributing all impaired communication to an individual's autism diagnosis without considering the possibility of

coincidental visual impairment a phenomenon known as diagnostic overshadowing.[19]

TAKE HOME MESSAGE:

1. Testability of near binocular visual acuity and IOP varied for ASD patients by the level of verbal communication reported by theparent.
2. Visual and ocular findings in autistic children have high prevalence of uncorrected refractive error strabismus and binocular visionproblems with high ocular morbidity.
2. Early visual evaluation and rehabilitation as an intervention are important for the improvement of visualfunction.
5. Pediatricians and Family Physicians involved in the care and management of autistic children should be encouraged to refer them for thorough ophthalmic evaluation.

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Pediatric Eye Examination

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INTRODUCTION:

In a general patients outdoor, the presence of a small childas patient, is viewed with some distaste, as most of cliniciansare not conversant with the techniques of eye examination of a small child. In essence, ocular examination of a small childrequires patience, skill, and some talent. If one has to becomeproficient as a pediatric ophthalmologist, the person has to train himself to learn the tricks for a smooth and efficient examinationin children. We will focus most of our attention on learning examination of infants and preschool children, where the examination is difficult and requires special skills.

Sophisticated technological advances in medicine have proved to be remarkably useful in the diagnostic process, yet the well observed history and physical examination remain a clinicians most important tools. They are venerated elements of the art of medicine, the best series of tests we have. Numerous medical anecdotes relate instances in which the examination revealed findings unrelated and unexpected from the patients complaints and concerns.

Timely eye examinations and visual assessment are critical for detection of conditions that may result in irreversible visual impairment and in some cases threaten a child's life.

There is a difference in your approach when taking history/examining a small child's eyes than an older child, with only vague complaints by parents. Many a times parents will bring children for just routine examination as their other sibling already has an eye problem. The approach to examination will depend upon the age, level of development, and level of understanding of the child. Inspection and observation are probably the most important part of examination.

To architect a pediatric clinic, one has to make certainchanges in the environment of the clinic. For example, thewaiting area should be different than a general patients area,with comfortable sitting and some toys, big and small, whichmakes the child feel at home rather than a hostile hospitalatmosphere. Some institutions arrange a 'play area' where thechild can play during the waiting period. The waiting period should not be too long, as the child may becomesleepy or hungry and would be uncooperative duringexamination. It is imperative that the child must be accompanied by theparents, preferably mother. The examination room shouldbe well lighted at the beginning so that the child is notapprehensive to enter. This has an additional benefit, that is, the clinician can 'observe' the child as he/she comes to thedoctor.

THE PROCESS OF EXAMINATION

Observation: Immense amount of information can be hadduring simple observation of a child. Do not rush to examinethe child. Let the child sit comfortably on parents lap forsome time. This will give time for the child to adjust to thenew sorroundings and more importantly, gives time to theclinician to observe the child. Simple observation will reveala lot of information, viz. thefixation of eyes, any obviousmisalignment, nystagmus, etc. The child's behavior can alsobe ascertained at this juncture. An irritable child would notco-operate, may not even open eyes and therefore the examinationmay be deferred for some other time, or the child may berecalled once he/she has settled. It must be remembered thata sleepy or hungry child will not cooperate and therefore itis prudent to let the child have its timely feed and thenrecalled after an hour or so.

History: This is most crucial and should be listened carefully from theparents, since no information can be had from a small child. A very detailed history is not needed as you may looseprecious time till the child is cooperative. Relevant and 'focussed'history is the key and unnecessary questions should beavoided. An old adage that "the patient is always right" doesnot necessarily apply here. Many a times the mother/parentare ignorant and may not understand the illness; the socialcircle around them may not perceive subtle strabismus ornystagmus as a threat to vision. The general thinking thatsuch problems occur frequently in infants and small childrenand will gradually outgrow with time is deeply prevalentand the parents may not record the time when it was noticed.Sometimes, the parent will casually declare that this problemis present since 'beginning', even it may have arisen just acouple of weeks back.

History relating to perinatal birthtrauma, hypoxia, febrile illness, or any such birth problemshould be specifically asked. At times it is a good idea toask direct questions if unnecessarily time is being wasted. A slightly older child may himself narrate certain symptomsand this should be carefully noted. Manychildren may notcomplain of blurry vision—as they have learned to live with it-and appropriate visual test is mandatory. In an olderchild the head posture can be observed while he/she isspeaking, and other facial anomalies can also be observed atthis time.Any rubbing of eyes and photophobia should be enquired.

The problem for which the child has been brought

forshould be asked from the parent or the older childhimself (achild of 4 - 6 years are verbal and should be well communicative), and further querries in that direction should befocused. Whether the ailment is congenital or acquired shouldbe asked and if acquired, then the age of onset. This has abearing on the prognosis of treatment. If strabismus ornystagmus is observed, the time of onset and frequency should be asked; but if vision defect is the chief complaint, thenparents need to specify whether the baby can see lights, respond to gestures, catches small toys, or very small objects. In small child with strabismus, most of times, a vague answer that strabismus is present from beginning is given. As a rule, the parents should be asked to show photographs of child since birth, which will reveal the time of onset. This will greatly help in deciding management and prognosis of vision.

In older children, the child's behaviour in school should beasked, speciallyfor fatigues, headaches, vertigo, sleepiness andregarding any specific complaint which has come from school management. Any neurological deficit should be enquired andthe referring physicians notes be seen. Whether the child is onany medication, should be specifically asked and the type of medication enquired into. Many medications for any neurologicproblem or GI problem contain salts that may cause drowsinessand create unattentiveness which may be perceived as vision defect. Sudden occurance of strabismus, diminution of vision or diplopia, needs a thourough neurologic checkup and appropriate referral should be done.

The sequence of developmental mile stones should be askedand any discrepancy noted.

It is important to remember that all general and visual milestones in a premature infant are delayed, and thus all parameters of visual functions are extended.

Lastly, the family history is alsoimportant . Enquiry should be done of any similar problem in other sibling, or parents or other direct relatives. Previousmiscarriages should be enquired .

In short, as much of history possible should be collected inthe shortest time possible; as the child may not cooperate forlonger period and the actual physical examination would become difficult.

Older children are better to deal with. They can answerregarding their problems and whatever they narrate should betaken note of it. It is again prudent to develop some kind offriendship by asking about their hobbies, their school program, and about their likes and dislikes, before commencing thephysical exam.

PHYSICAL EXAMINATION

Children can be unpredictable, uncooperative, and non communicative.

Children between 1–2 years are most difficult toexamine. In later ages they become more playful and communicative. They start knowing toys and listen to your requests.

For infants, the only source of information is their parentsand examination during feeding with a bottle makes thingseasier. Larger, brightly colored toys are usually used to attracttheir attention. Noise making toys are not recommended, asmovement of eyes due to 'sound' gives false information.

Infants and small children should always be examined whenseated on mothers lap, where they feel most comfortable andsecure. There should be no hurry to finish the examination,and it is advisable to keep as much distance away as possible.

Examination should be done with subdued light, as bright light can be intimidating and irritate thechild and may close the lids making examination difficult.

Children over 2 years are more responsive, and therefore, calling by their name or nickname, makes them feel better. Afriendly rapport should be first established with the child before embarking on any manuvre. Always begin with 'non-contact'things: cover test, fixation pattern, red glow, pupillary examination, etc. Many small children get afraid by touch of a stranger, and once they get upset, it is the end of the examination. Allow the parent to show them toys of 'appropriatesize' ,while you watch the eye movements for fixation.Appropriate size means the size of an object recommended forthat particular age for testing purpose.

External examination: The child's overall appearance and levelof alertness can be judged during history taking from parentsor child himself. Ocular alignment and position of head shouldbe the first thing noted. The history will guide in whichdirection the physicalexamination should proceed includingany specific tests required. The position of lids and lid aperturecan be evaluated at this time.

Visual acuity assessment: infants :After a generalidea, the first and foremost step is to assess their visual acuity.

Gross visual acuity in infants is mostly tested for fixation and following movements, monocularly. The examiner must knowthe appropriate size to which the infant may hold attention.For a one to three month old , the 'human face' is the best target, whilea toy of size of 'thumb' suffices for one year infant. Objects(toys) of variable sizes fall in between these two ages. Usuallyin infants, slow pursuit movement arises around 4–6 months,but saccadic pursuit is even present before this age. Thereforeduring evaluation this has to be kept in mind.(see photo1 & 2)

Photo 1: Recommended Toy photos for infants around 6 months.

Photo 2: Fixation toys for infants around 1 year.

Preverbal children above 1 year of age, respond to differentvarieties of vision testing, which has been described inchapter on vision evaluation.

Fixation: Fixation is tested monocularly and binocularly. Inmonocular fixation one assesses whether the patient fixes withthe fovea (central) and the quality of fixation. Each eye should be occluded in turn and the smallest possible target, appropriatefor that age, that elicits the response should be used. Fixationis assessed for three different functions: location (central versus (Eccentric); quality (good versus poor); and duration (maintainingfixation). In day to day practice the dictum CSMisusedwhichdenotes'central,steadyandmaintained'. 'Central' denotes 'foveal fixation';'steady'denotes 'quality' (no nystagmus or any unsteady movements); and 'maintained' proves that the fixation is maintained when the patient 'follows' the movement of light across from one side to another.

Sometimes theword FF is also used for quality maintenance which means 'fixand follow'. Steady, central fixation is a good sign and the visionfor that age seems to be normal. Eccentric fixation is an ominoussign and the vision is assumed to be 6/200 or less, Snellen. The target should be moved slowly across the visual field toassess the 'quality' of fixation The target size and distanceshould be documented. The 'fix & follow' movements will also simultaneously show the range of both monocular and binocular eye movementsThe examiner should be aware of visualmilestones in an infant. Newborns have only 'sporadic saccadic'movements with very poor fix and follow pattern. By 6 weeks, infants show some smooth pursuit movements with centralfixation and by 8 weeks they have well developed central andsteady fixation with good fix and follow movements. It shouldbe remembered that upto 3-4 months the smooth pursuit movement (as demonstrated by Optokinetic testing) ispredominantly temporal to nasal, and this has to be kept inmind when testing for fix and follow movements. One shouldremember that there is a small subset of patients who havedelayed maturation and may not comply to the normal testing; in these cases it is better to recall after some months, but shouldshow definite CSM by one year of age.

Binocular testing compares the vision of one eye to the other. This test shows 'fixation'preference' of one eye and predictsdiminished vision or amblyopia in the non –prefered eye. Thistest has the advantage over monocular testing as even smalldeficiency of vision can be brought forth as the non-preferredeye may deviate or may not follow coordinated movementalongwith preferred eye during'maintenance' of fixation testing. Binocular testing also hasthe advantage that the vision of one eye may be very low, stillthe eye may fix monocularly if the target is very attractive; butthe discrepancy will be elicited binoocular testing.

It is important to do monocular testing prior to binocular testing to rule out possibility of bilateral symmetric visual loss.In patients with straight eyes or microtropia (strabismusless than 10 pd) the fixation preference can be tested using thevertical prism test. In straight eyes, it is impossible to say whicheye is fixing. The vertical prism test induces a vertical deviationandthereforeallows us to examine fixation pattern. Fixationpreference testing is a quick and accurate way of knowingfixation preference in cases of amblyopia due to anisometropia,unilateral ptosis, post operative residual tropias, and otherconditions that could cause unilateral amblyopia.

Children who demonstrate poor fixation to above mentionedtechniques, can be assessed by OKN drumor the Catforddrum. OKN (Opto kinetic nystagmus) is an involuntary pursuit response to a moving target of high contrast. Since the OKNdrum consists of stripes of high contrast, the child is attractedto them even who are disinterested in other targets. Thestandard response is equivalent to finger counting of 3–6 feet.This is a good test to evaluate fixation as well as vision in infantsand younger children.

Other ways of assessing visual function are the PreferentialLooking tests and the PVEP. These have been already described in chapter on visual acuity.

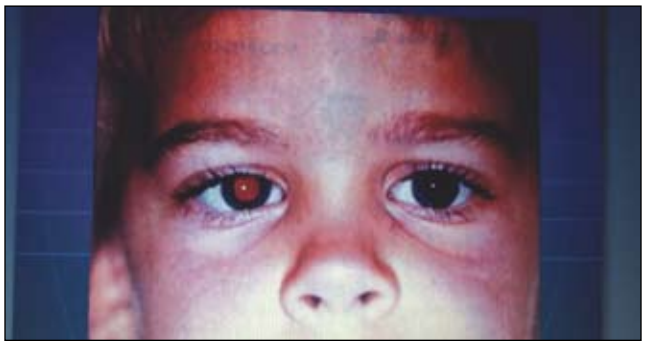
In preverbal children, Allen's figures, Lea symbols, and HOTV charts are the standard means of evaluating the visual acuity.

Corneal reflection test: Evaluation of misallignment of eyes (strabismus) is extremely important in infants and young children. Congenital esotropias, or exotropias, both have their importance with regard to vision, amblyopia or any ominous sign. Faint Leucocoria may not be evident to the parents but an obvious tropia may bring them for examination. As of strabismus per se, cover, cover-uncover testing can reveal qualitatively the type of strabismus. Though sixth nerve palsy is rare, but the author has seen cases being referred as sixth nerve palsy in infants, but actually having infantile esotropia with contracture of medial rectus. Some of these may be accommodative in nature, therefore the author emphasizes on cycloplegic refraction under atropine and a full fundus examination. Some infants or a child may resist the cover tests; in such circumstances, the Hirschberg's corneal reflection test may be helpful. Since this is done from a distance, the child may not be alarmed and a gross idea of strabismus can be achieved. Remember, this is a corneal 'reflection'of a point light from the front surface of cornea (first purkinjee image) and its position on both cornea, will give assessment of the tropia and to some extent, quantitatively also.

Pupillary responses: Newborns have small, miotic pupilswhich increase in size to about 6–7 mm by teenage and thengradually decrease in size throughout life. It is difficult to elicitdirect pupillary response due to extreme miosis and uncontrollednear reflex. Bright light should be avoided as the infantmay close the lids; also effort should be made to have the baby fix at a distance toy target to avoid the near reflex. Olderchild can control their near reflex but still it is wise to letthem look at a distance. It is important to identify any 'afferentpupillary defect', especially in unilateral amblyopias and visionloss due to macular or optic nerve disease. The 'swinging lighttest' is a good way of knowing the afferent pupillary defects asthe 'paradoxical' dilatation to light is an ominous sign ofmacular or optic nerve disease.

The Red Reflex: With the induction of high power bimicroscopy and othertechnologically advanced evaluation methods, the simpleevaluation modules have taken a back seat. Nevertheless, invery young children who would be uncooperative, the 'redreflex' from the fundus, has its own place to begin with. It would instantly show any media opacities and gross refractiveerrors, and subtle misalignment.

Bruckner described a very useful test to determine theseanomalies. He used a directophthalmoscope in a darkenedroom and examined the 'red reflex' from the pupil simultaneously in both eyes.In case ofstrabismus the affected eye would show a brighter reflex witha slightly larger pupil. It has been demonstrated that as smallas 5 pd of deviations can be ascertained by this method. Aneye with refractive error will show a darker reflex. Amblyopiatoo can be detected, as when the slit beam is focused on theaffected eye, non-fixation meansthat the eye is amblyopic.Other information like media opacities showing a dark spot or fundus anomaly showing a 'pale' reflex, can be obtained from Bruckner's test.



Bruckner's red relex: The brighter reflex in right eye shows esotropia.

PHOTO SCREENING:

This instrument based testing for visual anomalies has gained lot of importance in recent times. This is used for large scale evaluation in general population to screen children for various visual problems. This has become a standard practice pattern for testing of ocular anomalies in developed countries, where a normal protocol is that the first testing should be done at 6 months of age, then at 3 years and finally at 5-6 years age, before the child starts formal schooling.

Purpose of early vision screening and ocular examination is to identify children who may have eye disorders which may contribute to development of severe visual impairment, amblyopia, and lack of cognitive development of a child, at an early age, so that effective treatment may be initiated. Although there is limited direct evidence demonstrating the effectiveness of 'preschool vision screening' in reducing the prevalence of amblyopia or improving other milestones, a convincing indirect evidence supports this practice. The earlier vision threatening refractive errors and particularly amblyopia are detected, the higher the likelyhood of visual recovery.

Photo screening uses off-axis photography and photorefraction of the eye's red-reflex to evaluate refractive errors and small angle strabismus and thus identify risk factors in both eye simultaneously. A multicentric study revealed that photo screening was superior to optotype-based screening for children between ages 3 and 6 years and children who underwent their first photo screening at 2 years of age had superior eventual outcomes of treatment. Instrument–based vision screening techniques are more useful alternatives to visual acuity testing using optotype charts for very young children and children with developmental delays. But they are not superior to quantitative vision testing with charts in children who can participate in those tests. Instrument-based vision screening detects the presence of risk factors for amblyopia, strabismus, media opacities, retinoblastoma, and retinal diseases.

REFRACTION :

Determination of refractive errors is mostimportant in all examinations. It is mandatory for not only knowing refractiveerror in cases of strabismus and vision impairmen, but for a host of other complaints.

The clinician may be surprised to detect refractive errors in somany vague complaintsbychildren. It should be rememberedthat the adequacy of cycloplegia, not dilatation, isimportant.

Also the type of cycloplegic agent used according to age, the presence of any co-morbidity, and color of iris, should be kept in mind. The details these drugsare listed in Table 1.

In infants: Objective Refraction (retinoscopy) is indicated in all infants with defective fixation, preferential fixation,tropias, nystagmus, premature births, or any abnormality noted. The choice of cycloplegic agent is 1.0% Atropine sulphate ointment, instilled twice or thrice daily for 3 days. Some infants may not show full dilatation even after this period, particularly premature or dark iris infants. In these cases a diluted 2.5% phenylephrine can be instilled half an hour prior to examination, whence sufficient pupillary dilatation would occur.Normally sedation with phenergan syrup, chloral hydrate, or hydrozyne hydrochloride (Atarax) is sufficient in an infant for retinoscopy and fundus examination.

TABLE 1:

Characteristics & Dosages of Cycloplegic agents :

Cycloplegic drug	Concentration	Age Range	Dose	Onset of Effect	Mydriasis	Cycloplegia
1) Atropine sulphate oint.	1%	Below 1 year	TDS-3days	2-3 days.	10-12 days	15 days
2) Atropine sul.sol.	0.5-1.0%	1-5 years	1 drop-3 days	2-3 days	10-12 days	15 days
3) Homatropine hydro-bromide sol	2%	3-18 years	1 drop-10 minapart- 3 times.	60 mins.	3 days	7 days
4) Cyclopentolate drop	0.5%--1%	5-21 years	1drop-10min apart	45 min	24 hours	24 hour
5) Tropicamide drop	0.5%, 1.0%, 2%	12 years above	2 drop-10min apart	30 min	6-12 hours	6-12 hos.
6) Scopolamine sol. & oint.	1-2%	3-12 years	2 drop -10 min apart	60 min.	3 days	3- 5 days
7) In infants , alternatively, a 'mixture of 0.5% Tropicamide and 2.5% phenylephrine' can be used.						

Toddlers do not sedate well with the above medications, and where proper evaluation is necessary, general anesthesia should be used. We have now very safe agents for short-time anaesthesia. Loose lenses or a lens-rack is recommended for retinoscopy. In infants, where atropine is contraindicated or previous use showed allergic reaction, a mixture of 0.5%tropicamide and 2.5% phenylepherine serves good purpose for fundoscopy and reasonably accurate retinoscopy. In preschool children, homatropine 2% is a good alternative; and in children 5 years and above, cyclopentolate 1% can be used. In author's clinical view, cyclopentolate is normally used above 5 years of age except in Down's syndrome, cerebral palsy, or any neurological disorder,or if child showed 'abnormal behavior' on previous use of this drug. In such situations, Homatropine 1% or even Tropicamide 1% is recommended. Children of school going age, who come with complaints of ocular asthenopia or headaches or tropias,usually esotropia, tropicamide may not be effective to unleash the full hyperopia, and a stronger cycloplegic drug is required.

For information purpose, following types of refraction techniques are available:

1- Static retinoscopy, non-cycloplegic, using a distant fixation target, followed by subjective correction.

2- Non-cycloplegic Refraction using an Auto-refraction equipment.

3- Cycloplegic refraction, using retinoscope or an autoref.

4- Mohindra near non-cycloplegic retinoscopy.

Mohindra Near-Retinoscopy, without use of any cycloplegia,, is another objective method of estimating refractive error in infants and small children. The technique involves performing retinoscopy at near, about 50 cms., in an otherwise dark room ,as the patient fixates at the retinoscope light with one eye, while the other eye is occluded. However it is not very reliable for quantification of refractive error and the gross refractive error is usually 1.0 to 1.5 D on myopic side. Near-retinoscopy may be useful in the following situations:

- When frequent follow-up is required.
- When the child extremely anxious for instillation of any drops.
- When the child showed any adverse reaction to any of the above agents.

Cycloplegic retinoscopy: As a rule, 'cycloplegic' retinoscopy is mandatory in all infants, preschool and school going children, as the full and proper error can only then be established. The author uses cycloplegia till age 21 years. A common mistake is using Auto-refractor in older children without cycloplegia. It should be mentally noted that children, particularly hyperopic, automatically accommodate more on autorefractor; which gives a false reading of myopia. Thus, a myopic prescription now, would exaggerate their asthenopic symptoms and a vicious circle ensues. Even in 'static non-cycloplegic retinoscopy', a child may not be able to relax accommodation at distance, particularly a hyperopic child. Therefore, there is no substitute for a 'cycloplegic refraction' using a proper cycloplegic agent.

Dynamic retinoscopy: Dynamic retinoscopy is a type of near retinoscopy where the child first focuses on a distant target and then at a handheld near target. The change in retinoscopic findings gives an idea of the accommodative amplitude. It is useful in determining the 'accommodative lag' (insufficiency of accommodation), where the required amount of accommodation is not available for near work. The technique helps to gauge the accommodation in preverbal and schoolgoing children who cannot comply with he RAF Ruler,and who have a plethora of vague complaints. Associated with hyperopia, accommodative lag, can have serious effect on a child's reading and writing capabilities.

Visual fields: As soon as the child begins to fix steadily, say around 2 years, visual fields should be routinely tested. The easiest and quickest way is by

'confrontation method' using an interesting target. Both uniocular and binocular fields shouldbe assessed. If the child resists patching, binocular testing willalso yield homonymous defects. Even in an infant, a fixation target may be used to fixatecentrally and then a different attractive target may be broughtin the peripheral field. Owing to good saccadic reflex, the infantmay suddenly look at the peripheral target once it is broughtin the child's field of vision.

Color vision: Although color vision is not routinely done inchildren but may be helpful in decreased vision of uncertainetiology and constant monitoring in progressive macular disease and optic neuropathies. More often than not, a parentmay bring the child to the clinician claiming that he/sheconfuses between red and green pencil while during drawingfor home-work. Congenital red-green color defects areprevalent in about 8–10% of male population and the early it isdiagnosed, the better.

The easiest way to determine color defects are the colorplates. There are two popular types of plates which are helpfulin specific situations. The 'Ishihara pseudoisochromatic colorplates' work on the principle of 'color confusion' and are usefulfor detection of red-green defects. Most acquired color defectsshow in the blue-yellow range, and will be missed on Ishiharaplates, unless the defect has extended to red-green range. Theadvantage of this test is it can be done on illiterate patients aswell as children of preverbal age, as only fingers have to bemoved on the color lines.

The other test called 'Richmond pseudoisochromatic plates',previously known as 'Fardy-Rand-Rittler' plates works on theprinciple of 'color saturation' and can detect both red-greenand blue-yellow defects. Unfortunately these do not come inilliterate plates and is difficult for young children.In general, optic nerve disease will more likely show red-greendefects ,while retinal disease will show blue-yellowdefects.

Slit lamp examination: Slit lamp in young children is difficultdue to obvious reasons. Infants would not open eyes and brightlight is not appreciated by infants and young children. Hand–held slit lamps are available which are useful in a cooperative child. In infants and smallchildren, examination under general anaesthesia is the bestway for microscopic, indirect ophthalmoscopicexamination, and retinoscopy.

Fundus examination: Last but not the least, an adequate fundusexamination is imperative for children. For most patients, visualization of posterior pole (optic disc and macula) usuallysuffices. For detailed peripheral examination, general anesthesia isusually required. Infants below 1 year, can be sedated andexamination can be performed with slightly dim light. Young

children around 2 years may not get sedated, and general anesthesiamay be required. Children who are older than 3 years are morecooperative and periphery can be examined in sittingpositionby explaining them the procedure, which is more acceptable tothe child.

IN A NUTSHELL:

1- In all pediatric examinations, a subdued, non-scary light should be used. A small 'pen-light'or a direct opthalmoscope may suffice.

2- The child should be alert and cooperative, well fed and comfortable.

3- Examination of child on parents or any known relatives lap, is an ideal position.

4- 'Focussed' history is the key prior to any physical examination.No time should be wasted as children loose interest very quickly. If necessary, direct questions should be asked relevant to the complaints.

5- Observation of the child while the brief history is taken, gives enormous amount of information.

6- Try to establish some sort of rapport with the child by doing a hand-shake or calling by nicknames are helpful and drives out the fear from the child. Toddlers and preschool children will be more cooperative wih this kind of gesture.

7- Always use the 'age appropriate' toy target for examinations of fixation, motility testing, cover-testing , etc.

8- Visual acuity, even qualitative is important to assess.

9- Examine under anesthesia , whenever necessary.

10- Call the child some other time, if examination is improper due to any reason. Enquire the time of day when the child is most cheerful, and try examination at that time.

11- Insist on photographs- current and previous- as they are valuable in knowing the time onset of the disorder.

12- Fundus examination is mandatory in cases of suspicious red-reflex, strabismus, leucocoria,etc.

13- Do not alarm the parents, if some sight-threatning or life-threatning disorder is detected.

Never spell out disaster at first meeting. And refer to a higher centre if further evaluation facilities do not exist at your centre.

14-There is no set protocol in the routine of examination of child.The examiner should understand the gravity of situation and mould the physical examination accordingly. The examiner should also be innovative and adaptive to attract the childs attention during examination.

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Compartmentalization of Innervation of Extraocular Muscles

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Donder's and Listing's laws avidly illustrate how ocular torsion, a prominent part of ocular movements in lower lateral eyed animals has been compressed to mere 7-8 degrees in side / up/ down gazes for frontal eyed binocular vision. Additional vergence movements of a binocular visual plant entail an added burden for stemming torsional movements with some trace extorsion in down and intorsion in convergence.To give effect to such complexoculomotor kinematics,the horizontal recti (anterior and posterior rectus in lower animals) additionally acquired torsional and vertical vectors with the obliques taking backseat. The pulleys and compartmentalization of innervation are some of the mechanisms that have evolved to meet daunting challenges of stereoscopic binocular single vision, not only in primary gaze but also in all gaze positions as well as during vergences.

The active pulley hypothesis (APH) proposed by Demer et al and subjected to polemics by Jampel et al proposes that orbital layer shifts connective tissue structures known as pulleys to influence pulling directions, the global layer directly exerting the oculomotor torque. Aside from APH, lot of evidence has been accumulating regarding compartmentalization of innervation of human extraocular muscles(EOMs). Human EOMs are believed to differentiate from a superior mesodermal complex, the superior rectus, superior oblique (SO) and upper halves of the horizontal recti whereas from inferior complex originate inferior halves of horizontal recti and inferior oblique and inferior rectus muscles. The lateral rectus (LR) and SO have been studied most extensively but compartmentalization has been documented in others as well. Avian LR is known to arise from somatomeres, 4 and 5 and abducens from both rhombomeres 5 and 6.In humans the origin of LR is often from two roots. Attesting to this anatomical fact islongitudinal splitting of LR muscle that has been observed on MRI in congenital cranial dysinnervation disorders including CFEOM, Duane syndrome and congenital 3rd and 4rth cranial nerve palsies[1]. In other animals including monkeys the CN6 innervates LR after giving branches to retractor bulbi, a muscle that is extinct in humans showing that phylogenetically LR is not only having more than one progenitor but is programmed to innervate multiple functional units. Recent insights into Duane's retraction syndrome and 6th nerve palsybuttress this precept[1].

Histologic reconstruction also attests to above construct. Both the 6th nerve and nerve to medial rectus (MR) divide into distinct superior and inferior divisions that innervate minimally overlapping neuro muscular compartments, vertically segregated over the entire antero- posterior length of the muscle[2,3]. Segregated innervation also predicts differential response to pathologic injury to nerve branches innervating different functional compartments. A compressive, demyelinating, neuro vascular insult or trauma may denervate only one compartment. A weak superior compartment with preservation of structure and function of inferior compartment betrayed greater abduction force than those with palsy of both compartments[2, 3]. Such patients also display ipsilateral extorsion and hypotropia, both could result from intact inferior compartment force[2, 3]. Upto 1/3 cases of 6NP may show greater atrophy of superior compartment. Since unexplained cyclo vertical strabismus may entail costly neuroimaging, understanding of cyclo-vertical effects of horizontal rectus palsy would avert costly testing, wrong diagnosis of a cyclovertical muscle palsy / skew deviation and preempt unnecessary surgeries[4]. The MR motor nerve also arborises into non overlapping equal superior and inferior compartments. The inferior oblique is innervated by distinct medial and lateral roots that arborise in the muscle and generate differing contractile responses. Compartmentalization is less clear for inferior rectus that has a distinct lateral trunk with arborization with another trunk, thus selective neural control of lateral inferior rectus may be possible. The evidence for arborization of the superior rectus is tenuous[4].

The 2 divisions of trochlear nerve innervate non overlapping medial and lateral compartments of the SO. The medial SO compartment has predominantly torsional vector and inserts on the equatorial sclera. The lateral SO compartment is inserting on posterior sclera and has mechanical advantage in generating depression and abduction[5].

The diversity of presentation in SO palsies may relate to differential involvement of both the compartments. Based on MRI imaging it has been documented that Parks – Bielschowsky 3 step test, the cornerstone for diagnosis of cyclo-vertical muscle palsies may fail to detect 30% cases of SO atrophy on MRI. Compartmentalization of innervation of EOMs is thus an evolving concept and future concrete insights may help resolve many abstruse dilemmas confronting ocular motility.

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Facial Features of Alagille Syndrome

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Abstract : *Alagille syndrome is a multisystem disorder involving the liver, heart, vertebrae, facial features, and the eyes. In this case report, we document a case of Alagille syndrome with typical facial features presentation. The diagnosis of Alagille syndrome is necessary in cases of ongoing bile duct damage in the setting of early-onset jaundice, cholestasis, hepatosplenomegaly, posterior embryotoxon in the eyes, and butterfly vertebrae.Posterior embryotoxon also occurs in the general population with a frequency of 8-15%.*[4]

Keywords: *Alagille syndrome, cholestasis, jaundice, AGS (alagille syndrome).*

INTRODUCTION:

The incidence of Alagille syndrome (arteriohepatic dysplasia) occurs in 1 in 30,000 to 50,000 live births.[1] The most common symptoms associated with this syndrome are cholestasis (the obstruction or slowing of biliary flow), congenital heart disease (pulmonary artery stenosis), butterfly shaped vertebrae, anterior chamber eye defects, and dysmorphic facies.

In most cases, the diagnosis is clinical and almost 90% of cases are due to mutations in JAG1 (20p12) that are inherited in an autosomal dominance pattern.[1]

Facial features incorporates broad forehead, deep-set eyes, upslanting palpebral fissures, prominent ears,bulbous tipped nose and pointed chin. Children with Alagille syndrome have both increased prevalence of intellectual disability and motor delay when compared to the general population.[2,3]

A spectrum of retinal pigmentary changes have been reported in Alagille syndrome patients.[5,6,7,8,9]

There is inter-observer disparity in identification of these features and that they are a common result of early and chronic cholestasis ("cholestasis facies") rather than typical of Alagille syndrome.[2] In the majority of patients, visual prognosis is good, although mild decreases in acuity have been reported.

CASE REPORT :

A 8 year old boy presented to us for routine screening. The visual acuity were 6/6 for distance for both eyes and N6 for near Dysmorphic facial features as seen in Alagille syndrome noted. Anterior segment findings showed a upslanting palpebral fissure with conjunctival xerosis,mild icterus with posterior embryotoxon. Fundus findings were normal.

A liver biopsy-cofirmed the diagnosis. A finding of intrahepatic bile duct paucity on liver biopsy was found. The other family members had no specific medical history. These family members didn't look similar to the patient. A diagnosis of AGS (ALAGILLES SYNDROME) was made on the combination of, characteristic facial features, cholestasis and the presence of the result of liver .

Forehead	Broad
Eyes	Deep set eyes, up slanting palpebral fissure, conjunctival xerosis, mild icterus, posterior embryotoxon
Ears	Prominent ears
Nose	Bullous tipped
Chin	Pointed with triangular appearance of face.

DISCUSSION :

AGS is a genetic condition that is characterized by chronic cholestasis due to a paucity of bile ducts and multi-organ involvement of varying severity.[5,12,13] The manifestations generally become evident at a pediatric age. AGS is caused by defects in the Notch signaling pathway due to mutations in JAG1 or NOTCH2.[14,15,16] It is inherited in an autosomal dominant pattern with a high degree of penetrance, but variable expressivity results in a wide range of clinical features.

A classic diagnosis of AGS has been based on the finding of intrahepatic bile duct paucity on liver biopsy associated with three to five major features: chronic cholestasis, cardiac disease, skeletal abnormalities, ocular abnormalities, and characteristic facial features.3Though AGS is inherited in an autosomal dominant manner, a substantial portion of AGS is sporadic. In this case, the patient's family members have no definite features associated with AGS. According to previous reports, approximately 30%-50% of affected individuals have an inherited pathogenic variant and about 50%-70% have a de novo pathogenic variant.[14,15,16]

IMPORTANCE OF IDENTIFICATION

To label the condition predictive value of systemic associations

To guage if steps can be taken improve the general status of the patient

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Bilateral Acute Onset Accommodation Paresis as Presenting Symptom of Myasthenia Gravis in A Young Male - A Case Report

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Abstract : *Bilateral accommodation paresis with bilateral Adies tonic pupils in a young Male is a very rare. This indicates underlying systemic neurological condition. We describe a case of a young male presenting with sudden bilateral blurring of vision for near. Clinical and diagnostic evaluation revealed myasthenia to be the causative factor. Though presence of associated factors like trauma and alcoholic hepatitis seemed to be misleading, high index of suspicion lead to unveil correct diagnosis and management .This indicates that myasthenia should be highly suspected based on clinical findings in accommodation paresis or in early onset presbyopic patients.*

Keywords: *bilateral accommodation paresis , myasthenia gravis ,early onset presbyopia, bilateral Adies tonic pupil.*

Key message : *Pupil evaluation is important in early onset presbyopes. High index of suspicion and diagnosis on clinical findings are important in treatment of serologically negative myasthenia cases.*

INTRODUCTION:

Young patients in third decade of life coming with sudden onset difficulty in accommodation are usually presumed as early onset presbyopes. But pupillary abnormalities with accommodation weakness need to be investigated for systemic, neurological and local causes.

Adies tonic pupil usually presents as unilateral glare with accommodation weakness and is more commonly reported in middle aged females.

Association of myasthenia and bilateral Adies tonic pupils in young male has not been yet reported. Our patient had atypical presentations of myasthenia in terms of age, sex, and mode of presentation.

CASE HISTORY:

A 34 year old male patient, chronic alcoholic by nature presented to us with alleged history of blunt trauma over left eye one week back. He was diagnosed elsewhere as subconjunctival hemorrhage in left eye. After three days he reported sudden onset blurring of vision for near in both eyes. Patient was known alcoholic with hepatitis. He also mentioned difficulty in feeding himself and found sometimes difficult to lift his right hand to mouth while eating .

On examination, his OU UCVA was 20/20, and N 36 OU with Jaeger's chart improving to N6 with +1.50DS near add. Anterior segment examination revealed pupils 5 mm in OD and 6 mm in OS with very slow vermiform contractions in response to light. There was no evidence of RAPD, nor any evidence of traumatic sphincter tear in OS. OS showed small subconjunctival haemorrhage inferiorly. Orbital margins were intact nontender. Ocular movements were normal with normal convergence. Intraocular pressures were 16 mm of Hg in both eyes. Gonioscopy revealed normal angle structures. Fundus examination was normal in both eyes with normal disc and normal foveal reflex.

Colour vision and contrast sensitivity were normal in both eyes. Auto-mated perimetry with Humfrey's Visual Field analyser showed no de-fects. Lids were found to be drooping intermittently. Patient was referred to Neurophysician for further evaluation .

Neurological evaluation was normal except patent had subtle ptosis, transient fatiguability in right hand while having food and heaviness in tongue during talking. All these features had a demonstrable fatiguability. DTRs were normal and all other neurological examination was normal. Clinically this was highly suggestive of myasthenia and ACh Receptor antibodies titre was advised which turned out to be negative 0.12 nmol/L. Still based on clinical findings he was started on Oral Pyridostigmine (60 mg tablet) in dose of 30 mg TDS, oral Thiamine100 mg OD and oral mecobalamin. He was advised review after one week.

One week later patient reported significant improvement in symptoms in terms of near vision and improvement of power in hand.

His UCVA for near improved to N6. His pupils were 3mm with normal reaction to light. There was no drooping of eyelids . Patient was advised to continue treatment for one month and review after one month.

DISCUSSION

On a casual examination a 34 year old patient with near vision problem can be diagnosed as early presbyopia. Our patient had pupil abnormalities which warranted further evaluation. Bilaterality of complaints and intact pupillary sphincter ruled out trauma as cause.

Adies tonic pupil is usually unilateral and more common in middle aged females. Bilateral presentation in a young male suggests central involvement and should have called for MRI brain. Adies pupils shows female prepondarance (2.2:1) and bilateral involvement was seen in 20%. Autonomic dysfunction and absent tendon reflexes are associated with. Bilateral adies pupils. [1]

AchR antibodies titre is positive in about 70% cases and a negative titre does not rule out myasthenia. Clinical improvement on medical treatment of myasthenia proves the diagnosis in this case.

In past, Cases of pupillary dysfunction and fatigue have been reported in myasthenia gravis.[2] One case of bilateral idiopathic adies pupil in young pateint has been reported.[3] Two cases with accommodation paresis were reported in diagnosed cases of myasthenia.[4] One case with accommodation weakness was reported with Anti-GQ1b Antibody Syndrome.[5]

In our case, bilateral Adies pupils with accommodation paresis was initial presenting symptom which lead to diagnosis of myasthenia. In India males in sixth to seventh decades are found to be more commonly affected in MG than females, who show symptoms earlier in third decade. [6]

A great masquerader, MG is a disorder of neuromuscular junction. At neuromuscular junction when impulse travels from nerve ending to muscle cell, acetyl choline is released, taken up by striated muscle fibers through acetyl choline receptors resulting in postsynaptic depolariisation and muscle contraction. In MG, antibodies are developed against these Ach receptors which inhibit uptake of Ach and thereby inhibit muscle contraction.

Extraocular muscles have more of twitch muscle fibres which have higher frequency of synaptic firing and tonic muscle fibres responsible for maintainance of gaze. Both are more susceptible to MG, making ocular involvement very common and often presenting complaint.

Investigations for MG include Edrophonium test, neostigmine test, Ice pack test,fatigue test, AChR-Ab titres, single fibre electromyography, repetitive nerve stimulation studies and CT chest for thymoma.

Treatment of MG is mainly medical aiming to improve muscle function. Cholinesterase inhibitors improve functioning at NMJ, effective in mild cases. Oral prednisolone and azathioprine is used to maintain remission. Thymectomy may be required in case of thymoma. Plasmapheresis and immunoglobulin therapy are indicated in severe cases with life threatening symptoms. Any systemic drugs may exacerbate myasthenia and caution should be exercised while treating these patients .

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Superior Rectus Transposition for Sixth Nerve Palsy: Case Report and Review of Literature

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Abstract : *Transposition procedures have been proposed as a means of increasing vector force to the eye to compensate for the lack of a muscle function.Transposition of the superior rectus alone has become popular for patients with esotropia due to Duanes and sixth nerve palsy. We report two cases where superior rectus transposition gave a favorable outcome in cases of sixth nerve palsy and review literature in this regard.*

Keywords: *Transposition, sixth nerve palsy, abduction.*

CASE 1

A 28-year male presented to a tertiary care centre with inward deviation of right eye for 14 months. He had a history of craniotomy done for a cavernous sinus chordoma followed by gamma knife radiotherapy for the same completed eight months back. Records indicated a complete ophthalmoplegia pre-surgery which resolved partially post-surgery.

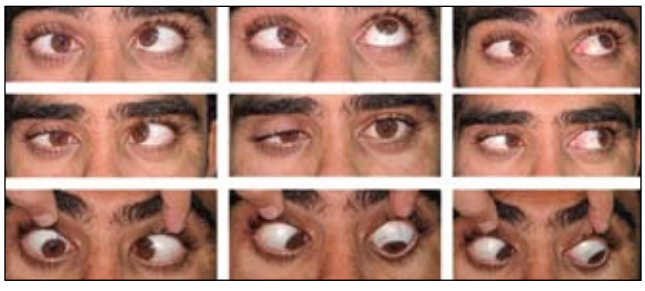

Figure 1: Resolving sixth nerve palsy in a 28-year male. There was the presence of right eye esotropia of 60 PD in primary gaze, with limitation in abduction (-4), elevation (-3) and depression (-2). SRT and 6 mm medial rectus recession was performed.

On examination he had a best corrected visual acuity of finger counting close to face in the right eye and 20/20 in the left eye. There was right eye moderate ptosis and relative afferent pupillary defect in the right eye. There was thepresence of right eye esotropia of 60 PD in primary gaze, with limitation in abduction (-4), elevation (-3) and depression (-2). After ensuring stability of deviation on repeated follow up visits, 6 mm medial rectus recession right eye superior rectus transposition to lateral rectus was performed. The superior rectus muscle was isolated taking care to clearthe muscle of attachments to the overlying levator palpebral superioris muscle andthe underlying superior oblique tendon. The muscle was secured, detached, andthen reattached to the eye along the spiral of Tillaux. Postoperatively, there was orthotropia in primary gaze at 6 weeks (Figure 2), and then six months (Figure 3). There was some improvement in abduction (-2) and slight limitation in adduction (-1). There was no vertical deviation or torsion introduced.

Figure 2: Postoperatively at six weeks, there was orthotropia in primary gaze at 6 weeks.

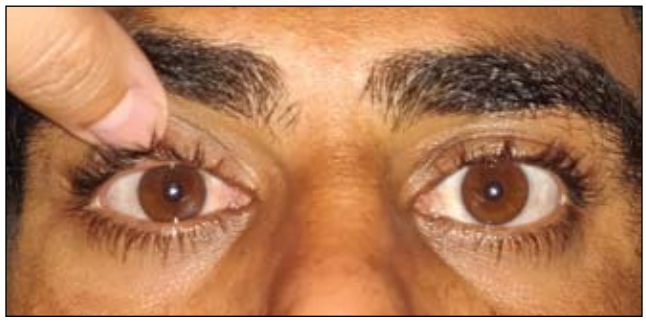

Figure 3: There was some improvement in abduction (-2) and slight limitation in adduction (-1). There was no vertical deviation or torsion introduced at the 6 months follow up.

CASE 2

A 37 year-male presented to the strabismus clinic with diplopia for six months with an inward deviation of left eye. His best corrected visual acuity was 20/20 OU. There was an esotropia of 40 PD in primary gaze with limitation in abduction(-4) (Figure 4). A diagnosis of sixth nerve palsy was made. Patient was thoroughly investigated and after six months of stable deviation, left superior rectus transposition and adjustable medial rectus recession was performed. Postoperatively the axis was parallel in the primary gaze (Figure 5) with some improvement in abduction(-3).

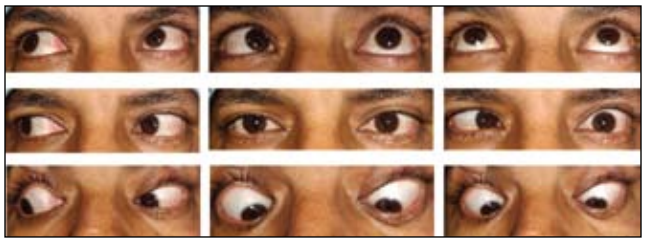

Figure 4: Sixth nerve palsy in a 37-year male patient. There was esotropia of 40 PD in primary gaze with limitation in abduction(-4). Patient was treated with SRT and medial rectus recession.

Figure 5: Postoperatively there was orthotropia in primary gaze at six weeks.

Superior Rectus Transposition in Sixth nerve palsy

If there is no lateral rectus function and there is no rotation beyond the midline in a patient with sixth nerve palsy, complete or partial transposition of the superior and inferior rectus muscles laterally with or without lateral fixation is commonly performed. However vertical rectus muscle transposition must be combined with ipsilateral medial rectus muscle recession if there is contracture of the medial rectus muscle. Full tendon transposition of only the superior rectus muscle to the lateral rectus muscle has been reported to reduce the number of vertical rectus muscles requiring surgery for treatment of sixth nerve and also reported to be effective in patients with esotropic duanes.[1-4]

Transposition procedures have been proposed as a means of increasing lateral force to the eye to compensate for the lack of lateral rectus muscle function. With the transposition of the superior rectus laterally, a portion of the muscle force vector directed vertically is displaced laterally. Abduction improves significantly in patients treated with Superior rectus Transposition (SRT) in elevation.

Previous studies have reported VRT coupled with medial rectus recession or botulinum toxin injection correcting 30 to 50 PD of esotropia1-4 and VRT with posterior fixation suture correcting 40 to 55 PD of esotropia in patients with sixth nerve palsy.[6-9] SRT/MRc with fixation suture usually corrects a mean of 36.4 PD of esotropia.

Potential complications of VRT procedures include new vertical deviations, induced torsion and anterior segment ischemia. After SRT postoperative torsional diplopia (intorsion mainly)occurs very rarely, and the incidence of vertical diplopia is approximately 7%which can be managed with secondary surgery or prism correction.[5]

Vertical alignment can change in the hypotropic as well as the hypertropicdirection, although there is a propensity toward hypotropia. It is speculated that the hypotropia does not occur when the muscle is attached along the spiral of Tillaux. A greater tendency toward hypotropic shift might result from diminished elevating force of the superior rectus muscle. Potential causes of a hypertropic shift include advancement of the superior rectus muscle along the spiral of Tillaux, dysinnervation, cicatricial changes, or residual attachment to the superior oblique tendon. Variation in lateral rectus tone can also affect outcome.Vertical deviation in affected side gaze after SRT is, however, of clinical concern, as fusion in this gaze position is prevented by persistent esotropia. Transient vertical misalignment after SRT appears to resolve within weeks to a noticeably short number of months after procedure. It is surprising that persistent vertical deviations do notpersist after this procedure. It is likely that supraduction is not significantlyaltered by SRT since the pulleys, not the insertion of the superior rectus muscles,serve as the functional origin. This might be due to the belief that the posterior portion of the transposed muscle is not as deviated toward the palsied muscle aswould be expected from transposition because of resistance from the pulleys.

Since the procedure causes a shift towards intorsion, presence of significant preoperative intorsion may prompt consideration of modified SRT that isadjustable or an alternative surgical intervention. Since anterior ciliary arteries course through the verticalrectus muscles, transposition increases the risk of anterior segment ischemiaparticularly when combined with ipsilateral medial rectus muscle recession. However, a single vertical muscle transposition i.e. SRT is associated with a minimal risk of postoperative anterior segment ischaemia.[10]

If we compare SRT with transposition of both recti, more additional procedures were needed in the VRT group in a published study.[11] Limitation of abduction is also less as compared to both recti transposition, along with better improvement in abduction. Overall, SRT procedure had a comparable efficacy to VRT.

Superior rectus transposition alongwith medial rectus recession is not inferior to VRT forcorrecting esotropia and limitation of abduction with sixth nerve palsy. The procedure reduces the number of muscles requiring surgery at the same time, decreasing the likelihood of inducing a vertical deviation observed with VRT. Medial rectus can be placed on adjustable sutures to decrease the risk of overcorrection. The improvement in abductionobserved with SRT may be greater in elevation,which is not unexpected considering the asymmetrictransfer of force in the single muscle transposition. Long term studies in a greater number of patients would further explore the benefits and deficits of the procedure.

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Bilateral Dacryocystocele : Case Report and Review of Management Strategies

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Keywords: *congenitaldacryocystocele, respiratory distress, amniotocele, mucocele, probing and syringing.*

INTRODUCTION

Epiphora in the neonatal period is caused due to obstruction of nasolacrimal duct in most cases. Some of these cases may be associated with swelling in the lacrimal sac area, due to formation of dacryocystocele. Congenital dacryocystocele has an incidence of 0.7% in all infants[1], 0.02% in newborns[2], and 0.1% in infants with nasolacrimal duct obstruction[3]. It is seen more commonly in females[4]. 10% ofcases are bilateral.[5] 80% of bilateral cases have concurrent intranasal cysts.[6]

Clinical presentation includes unilateral or bilateral bluish, tense, cystic enlargement below medial canthus present since birth.

Pathophysiology is said to be prenatal enlargement of lacrimal sac or nasolacrimal duct due to functional block at the proximal end of lacrimal sac (at the valve of Rosenmuller) and anatomical block at the distal end (valve of Hasner)[7]. Occasionally, it can extend to nasal cavity and may cause severe respiratory distress[8]. Although amniotoceles are typically sterile, acute inflammation of lacrimal sac can develop in 20% to 60% cases shortly after birth.[4]

The treatment options for congenital dacryocystocele are varied, and depend on the clinical presentation.

Bilateral presentation , signs of infection and presence of respiratory distress are indications for early probing and syringing. Unilateral cases, with no signs of infection can be put on conservative management initially, though most of them ultimately need a timely probing and syringing. This is unlike simple congenital nasolacrimal duct obstruction, where spontaeous resolution is more common, and chances of dacryocystitis are comparatively lesser. Hence conservative management can be advocated till 1 year of age.

We describe the case of a neonate who presentedwith bilateral congenital dacryocystocele at 2 days of age, along with features of impending pre septal cellulitis. We performed bilateral probing and syringing under cover of systemic antibiotics on day 5 after birth. Bilateral dacryocystoceles have been rarely reported in literature.[5] This represents the first case of bilateral dacryocystocele from India, to be managed by prompt surgical intervention, rather than conservative approach. Patient responded very well with immediate resolution of dacryocystocele bilaterally and no recurrence seen on follows up.

CASE REPORT

We report a case of 2 days old neonate, referred to our Out Patient Department for evaluation of bilateral swelling around medial canthus present since birth. Child is of first birthorder; birth weight is 2.72 kg, delivered atterm by normal vaginal delivery in hospital. APGAR score was 5 at 9 minutes with no signs of respiratory distress. Onlocal examination, there were bilateral cystic tense swellings present below medial canthal tendon,measuring approximately 5x5mm on right side and 7x7mm on left side. Swellings were bluish in colour andassociated with erythema and bogginess, which was more pronounced on left side. (Fig 1). There was no history of breathing difficulty with feeding. Swelling was soft, nonreducible, non compressible and ROPLAS was negative. Overlying skin was red, shiny and warm to touch. Upper and lower punctum on both sides were normal in size and appearance. Rest of ocular and systemic examination was unremarkable. Otolaryngologist opinion was sought, as the swelling was bilateral. Endoscopic intranasal evaluation was done to rule out any intranasal cysts, which were not present.

Ultrasound evaluation of the masses revealed round well defined, pre septal orbital cystic swelling with hypoechoic content. There was no evidence of posterior extension or communication with eye globe.

On the basis of history, examination and investigations, the child was diagnosed with bilateral congenital dacryocystocele with left early pre septal cellulitis. Pediatric consultation was done and broad-spectrum antibiotics i.v. Piperacillin-Tazobactam 300ml TDS were started. Parents were explained to do Criggler lacrimal sac massage on both sides, 10 strokes 4 times a day. The proper technique was demonstrated to parents. Tobramycin eye drops were prescribed 4 times a day and warm compresses twice a day. The child was re examined after 3 days. Features of early pre septal cellulitis on the left side were even more pronounced than the first visit, inspite of the conservative treatment. Hence the child was planned for immediate bilateral probing and

syringing under systemic antibiotic cover under general anaesthesia.

Procedure-Nettleship's punctum dilator was used to dilate the punctum. Bowman's lacrimal probe(0000) was passed till soft stop was felt, and overcome with probe, at the level of the valve of Rosenmuller. Further the probe was passed along the direction of the nasolacrimal duct. Second soft stop was felt at the level of valve of Hasner and overcome with probe. The probe could be self retained in the nasolacrimal duct and came to rest along the supra orbital notch at the superior orbital rim. This position of the probe reflected that the probe was in the right location in the nasolacrimal duct. This was further confirmed by spring test, by lightly pushing down the free end of the probe, it was seen to spring back to position spontaneously. The correct placement of probe was further confirmed by metal-to-metal touch in the nasal cavity. The probing caused immediate decompression of swelling on both sides, along with regurgitation of contents. There was immediate (Fig 2) resolution of the dacryocystocele bilaterally. Syringing followed this, which was found to be patent on both sides.

On post-operative day 1, there was complete resolution of dacryocystocele on both sides. On compression over lacrimal sac area, there was expression of yellowish fluid from left side. The patient's father was explained about lacrimal sac massage (Criggler's maneuver) along with the use of eyedrop Tobramycin and warm compresses to be done regularly. The swelling/discharge has not recurred 4 months after the procedure.

DISCUSSION

Dacryocystocele denotes neonatal distension of lacrimal sac which occurs due to coexistence of imperforate nasolacrimal duct at the level of valve of Hasner) and proximal functional block at the level of valve of Rosenmuller, as per Jones and Wobig. As fluid goes on accumulating within lacrimal sac and duct, this causes further collapse of the valve of Rosenmuller and further expansion of the lacrimal sac by a trap door mechanism.[7]

Dacryocystoceles have also been called as amniotocele and mucocele in literature.

The termamniotocele was also described by Jones and Wobig. They proposedthat amniotic fluid comes into lacrimal drainage system by action of eyelids in utero. This term is more applicable to swellings present at birth rather than for the swellings that develop several days after birth.

The term mucocele, is used by some authors. It suggests that lacrimal sac contents are produced by secretory elements, such as serous glands and goblet cells within the lacrimal sac mucosa.[7]

Although terminology may differ, it is believed thatamniotic fluid, intrinsic mucoid secretions, tears and products of bacterial colonizationmay each be involved at some time in neonatal sac distention. Hence the term dacryocystocele may be a more comprehensive one.

Differential diagnosis of swelling in lacrimal sac area include meningoencephalocele, capillary hemangioma, dermoid cyst and sudoriferous cyst.

It is important to screen forsyndromic associations and structural abnormalities such as polycystic kidneys, polyhydramnios and Wegener's granulomatosis[4] particularly in recurrent bilateral dacryocystoceles.[5]

Bilateral swellings need a thorough ENT evaluation as they may have associated intranasal cysts which can cause respiratory distress because infants are initially obligate nasal breathers. Such cases need prompt management[4]. In our case also otolaryngological evaluation was done and no intra nasal cyst or other pathology was revealed.

The optimal approach and management of congenital dacryocystocele is largely case specific and usually depends on its clinical presentation, association with intra nasal cyst and presence of any associated anomaly.

A simple congenital nasolacrimal duct obstruction reportedly resolves spontaneously with conservativemanagement in more than 80% of newborns during their first year of life[3].

Schnall and Christian reported that 76% (16 of 21) of the dacryocystocelaes in their study resolved within 2 weeks with a nonsurgical treatment protocol consisting of warm compresses and massage.[9]

Potential complications of conservative management include the development of dacryocystitis orcellulitis. There are more chances of cellulitis and lesser chances of self-resolution as compared to a simple NLDO, because of obstruction at both ends of the sac.

Rich lymphatics of lacrimal sac along with stasis oflacrimal sac contents, vascularity of itssub mucosal tissue and proximity to sinusesadd up to the chances of flaring up of infection.

It has been reported that in eyes with congenital nasolacrimal duct obstruction, 2.9% of the cases developed acute dacryocystitis in the first year of life[15], while 37.9%–74% of eyes with dacryocystocele developed dacryocystitis[10]. Large dacryocystoceles can lead to astigmatism and permanent canthal deformities.[7]

Most dreaded complications of conservative management of bilateral cyst is imminent respiratory distress

Conservative management alone has been reported to have high failure rate due to recurrence of dacryocele or development of dacryocystitis in upto 20 % of cases.[11]

Shekunov et al have concluded from their 20-year study that conservative management worked in 33% of the cases while 67% needed surgery.[2]

Similar findings were reiterated by a collaborative study of 54 cases from 7 centres, where most of the surgeons preferred probing immediately or shortly after presentation.[4]

It has been noted that delay in probing and syringing can lead to development of frank lacrimal abscess, requiring more invasive management such as incision and drainage, which has been reported to increase the chances of recurrence and further complications.[7]

Becker et al have documented development of dacryocystitis in 37.9% (eleven of 29) of the eyes with dacryocystoceles. Hence the authorsof the study recommend that if the sac doesn't decompress into the nose with massage during the first examination, probing should be performed urgently and as early in life as possible to decrease risk of dacryocystitis and increase success rate.[12]

Polland et al have stressed on the importance of looking out for any signs of infection and to carry out immediate intervention if there are any signs of infection present at initial examination.[13]

Advantages of prompt intervention include prevention of development of lacrimal sac atony, which helps to keep the lacrimal pump mechanism intact and reduces chances of re closure. This is important because in case of permanent closure, patient has to undergo dacryocystorhinostomy, which is a more complex intervention rather than just probing.

Paediatricians also recommend timely intervention, before development of infection, as neonates are relatively immunocompromised with a significant risk of sepsis and even meningitis.[14]

Study conducted by Weinstein et al reported majority of cases were cured with single probing within 1 week of diagnosis. These patients had erythema like our patient and sac couldn't be decompressed on initial examination. Hence the authors recommended probing and syringing of sac on an emergency basis if erythema was noted on examination.[15]

On the basis of literature search and our experience, we propose early probing of dacryocystocele if

- lacrimal sac swelling is not reducible/no regurgitation on pressure over lacrimal sac.
- Presence of surrounding erythema/edema suggestive of impending dacryocystitis and pre septal cellulitis.
- Bilateral involvement with or without documented intranasal cysts.
- Large dacryocystocele (causing astigmatism and narrowing of palpebral fissure).
- Presence of respiratory distress.

Futher it is recommended that-

1. Probe under systemic antibiotics
   - To prevent infections like pre-septal cellulitis, orbital cellulitis, dacryocystitis and systemic sepsis
   - Also it is known that prophylactic treatment with antibiotics increases success of probing two folds.[6]
2. For probing to be effective, both the proximal functional obstruction and distal obstruction should be overcome with the probe.

In a case report similar to ours, having bilateral presentation along with signs of infection immediate intervention was recommended followed by good results.[16] No such case report has been reporting from India preceding our case.

Only other report of bilateral presentation from India,was different from our case as there were no signs of infection/inflammation, hence the authors opted for conservative management[17] unlike our case, where early intervention was mandatory due to impending cellulitis.

The neonate reported by us had huge distension of the left lacrimal sac. Hence even after successful probing and syringing, the child had regurgitation of contents. This was due to atonicity of the sac. Similar situation was encountered and reported by Weinstein ET al.[15] further serous and mucoid secretions from intraluminal glands and goblets lead to collection of fluid in sac. Matsuno et al, who further confirmed the wide dilatation of the lacrimal sac and nasolacrimal duct on CT-DCG 18, also noted excessive distension of lacrimal sac.

Most experienced paediatric oculoplastic surgeons recommend early surgical intervention in case of bilateral dacryocystocele or with any complication like respiratory compromise, cellulitis, dacryocystitis or large dacryocystocele causing astigmatism, permanent canthal deformities, recurrent cases or failed conservative management.[4]

CONCLUSION

The probing caused immediate decompression of swelling on both sides, along with regurgitation of contents. There was immediate (FIG 2) resolution of the dacryocystocele bilaterally. Syringing followed this, which was found to be patent on both sides.

On post-operative day 1, there was complete resolution of dacryocystocele on both sides. On compression over lacrimal sac area, there was expression of yellowish fluid from left side. The patient's father was explained about lacrimal sac massage (Crigler's maneuver) along with the use of eyedrop Tobramycin and warm compresses to be done regularly. The swelling/discharge has not recurred 4 months after the procedure.

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Contralateral Eye Surgery in Third Nerve Palsy with Aberrant Regeneration

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Abstract: *A 38 year old female presented with binocular diplopia, drooping of upper lid and outward deviation of left eye since 3 years following head injury in a road traffic accident. On examination, her unaided visual acuity in both eyes was 6/6, N6. She had mild ptosis and restricted elevation, depression and adduction in left eye. Abduction and intorsion were present. On adduction, ptosis showed improvement and there was an upper lid lag on down gaze. These are signs of aberrant regeneration in third nerve palsy. Strabismus surgery was performed in the normal eye which led to improvement in ptosis and exotropia.*

Keywords: *Aberrant regeneration, third nerve palsy, Pseudo Von Graefe's sign.*

CASE REPORT

A 38 year old female presented with complaints of binocular double vision, drooping of upper lid and outward deviation of left eye since 3 years. She had suffered a head injury in a road traffic accident following which she was unconscious for a week. A CT-scan was performed at the time and revealed a sub-arachnoid bleed for which she was treated. On regaining consciousness, she noticed that she was unable to open her left eye. The drooping gradually improved over the next few months, however, she started noticing double vision and outward deviation of the left eye.

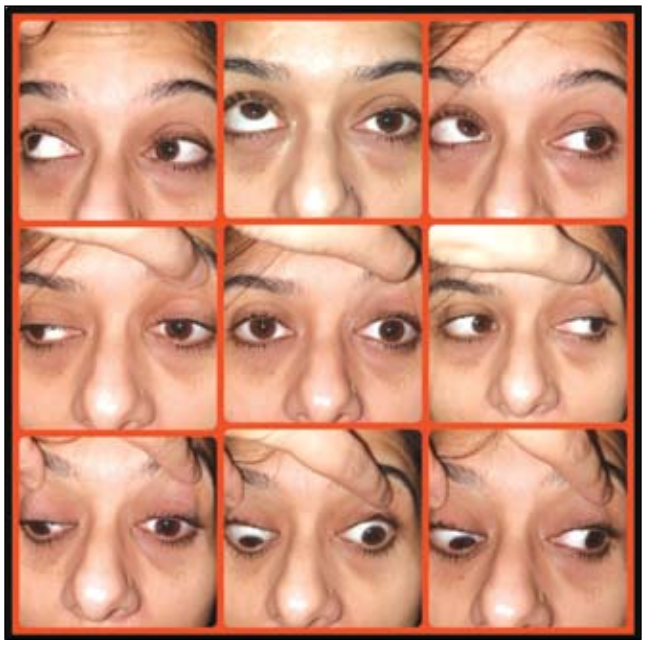

Figure 1 : (OS) ptosis and restricted extraocular motility in elevation, depression & adduction

At presentation her visual acuity was 6/6, N6 (OU) and intraocular pressure was 16mm Hg (OU). There was mild ptosis (OS) with a 3 mm difference in the palpebral fissure height between the two eyes. Bell's phenomenon was poor in the left eye. Hirschberg test showed a 15 degree exotropia with 7 degree hypotropia in the left eye. Ocular motility was restricted in elevation, depression and adduction [Figure 1]. Abduction and intorsion were intact. On krimsky test, there was an exotropia of 35PD and 10 PD hypotropia in primary position of gaze. Diplopia charting was done and fusion was obtained in primary position after prism adaptation test.

On down gaze, there was a pseudo appearance of lid lag in the left eye similar to what is seen in thyroid eye disease. This is termed pseudo Von Graefe's sign. Also, there was a decrease in the palpebral fissure height on abduction which improved significantly on adduction. This is reverse of what is seen in a classic case of Duane retraction syndrome. Hence, this is named Inverse Duane's sign [Figure 2].

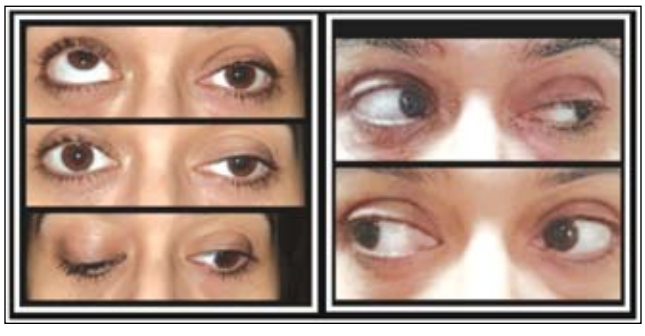

Figure 2 (a) Pseudo Von Graefe's sign and anisocoria (b) Inverse Duane's sign

Pupil diameter of right eye was 2mm and left eye was 5 mm suggestive of anisocoria. Rest of the anterior segment examination and fundus was normal in both eyes. Thus a diagnosis of traumatic, pupil involving partially recovered third nerve palsy with aberrant regeneration was made.

The patient underwent (OD) lateral rectus recession of 7mm and medial rectus resection of 5mm with half muscle upshift in the right eye under local anaesthesia. She was orthophoric in primary gaze on the next post operative day. Ptosis improved in the left eye and the palpebral fissure height was symmetrical [Figure 3]. At 3 months post-op, she maintained orthophoria with good lid symmetry.

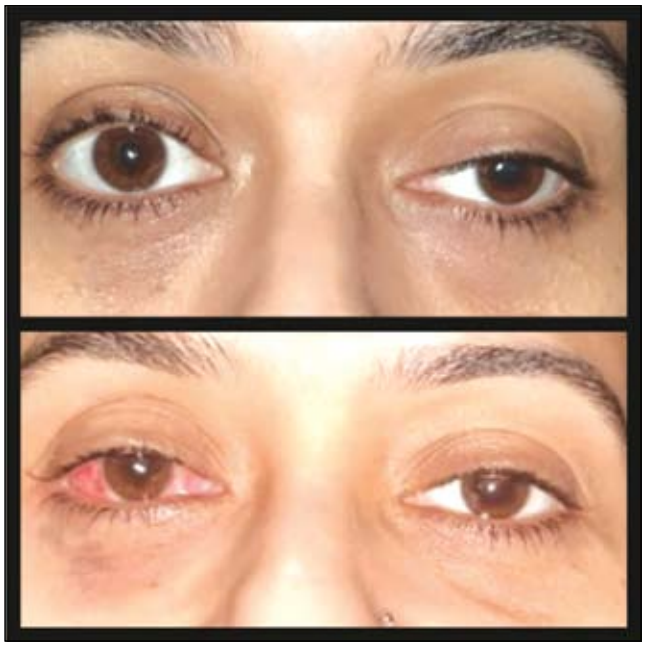

Figure 3: First post operative day- orthophoria in primary gaze with symmetrical palpebral fissure height

DISCUSSION

Non surgical modalities are not promising in cases of third nerve palsy. Surgery remains the mainstay of treatment with strategies differing for complete and partial third nerve palsy. In complete third nerve palsy, the aim of the surgery is alignment in the primary position by compromising on its motility. Supramaximal lateral rectus recession and large medial rectus resection of the paretic eye was described by Helveston[1]. Combined surgery on the horizontal and inferior rectus muscle is also described. In partial third nerve palsy, surgery aims to create a centralised enlarged field of binocular single vision, eliminating abnormal head posture and minimizing diplopia[1]. For isolated muscle involvement, the paretic muscle with residual function is resected and its antagonist muscle is recessed. If multiple muscles are involved, Knapp's procedure is recommended if forced duction test (FDT) is negative for inferior rectus. If positive, inferior rectus is recessed[1]. Since superior oblique overaction accounts for incyclotorsion and 'A' pattern, weakening of superior oblique muscle has also been described[1].

Patients with third nerve palsy are usually operated in two stages. First stage involves correction of the exotropia. Second stage usually comprises of levator disinsertion or frontalis sling suspension to correct ptosis. However, a two-step procedure is more cumbersome and is associated with higher risks of two surgical procedures and local anaesthesia. Resection of the ipsilateral medial rectus is unrewarding as the nerve supply to the muscle is already compromised. A second stage ptosis surgery will always carry the burden of corneal exposure related complications owing to poor Bell's phenomenon.

Aberrant regeneration refers to misdirection of regenerating axons, usually seen in traumatic or compressive lesions of the third nerve. They result in synergistic movements of eyelid with eye movements. This is also termed as 'oculomotor synkinesis'[2]. Common signs of aberrant regeneration include 'Pseudo Von Graefe's' sign and 'Inverse Duane's' sign. Other signs of aberrant regeneration include abnormal globe retraction on attempted vertical movements, pupillary constriction on attempted adduction (pseudo Argyll Robertson pupil) and ipsilateral adduction on attempted vertical movements[3]. Gaze-evoked segmental constriction of the pupil due to involvement of the iris sphincter is called Czarnecki's sign.[4]

Operating the contralateral eye in patients with aberrant regeneration dates back to 1980 when O'Donell et al. reported acceptable postoperative results in patients with good lid position on contralateral gaze[5]. Larger lateral rectus recession and smaller medial rectus resection with downward transposition of both horizontal recti in the contralateral eye demonstrated by Parulekar and Elston reported better correction of vertical diplopia[6].

Thanh Nguyen PT, et al. have also reported good results with the similar plan of surgery. Furthermore, the authors had planned an adjustable suture technique for optimised correction. Their patient was orthophoric and diplopia free in primary gaze 4 months post operatively[7].

The principle of surgery has been attributed to Herring's and Sherrington's law.

Herring's law confers equal and simultaneous innervation to the yoke muscles. Weakening the contralateral lateral rectus leads to increased innervation to the recessed muscle. This increased innervation is also equally and simultaneously received by its yoke muscle that is the affected medial rectus and by virtue of aberrant regeneration, the levator palpebrae superioris of the affected eye also receives increased innervation, thereby improving ptosis.

According to Sherrington's law equal, simultaneous and inhibitory impulses are supplied to the antagonist muscle in the same eye. Increased innervation to medial rectus leads to increased inhibitory impulses to its antagonist muscle, ipsilateral lateral rectus. This reduces the chance of contracture of the lateral rectus in the exotropic eye.

Thanh Nguyen PT, et al. also highlighted that by Herrings law, contralateral lateral rectus recession creates a fixation duress on its yoke muscle, that is, medial rectus of the affected eye, thus bringing the affected eye in a relative adducted position when the other eye fixates[7].

CONCLUSION

Signs of aberrant regeneration should always be looked for in cases of recovered third nerve palsy. Correct diagnosis and utilizing these synkinetic aberrant movements in planning surgery can be promising. However, counselling the patient regarding the decision of operating the better eye can be challenging. Dealing with patience, elaborating the plan and expecting realistic results is the key.

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Cover page illustration

Fraser syndrome, also known as Meyer Schwickerath's, Fraser- Francois or Ulrich-Feichtiger syndrome is an autosomal recessive disorder. It is characterized by cryptophthalmos, cutaneous syndactyly, abnormalities of the genitalia and urinary tract. Other organs can also be affectedincluding heart, larynx, ear and nose and palate. Fraser syndrome can be fatal before or shortly after birth, less affected individuals may live into childhood or rarely into early adulthood. Fraser syndrome is known to arise from mutations in FRAS1, FREM2 or GRIP1 genes. The present phenotype likely conforms to a genotype of FREM2 mutation with bilateral complete cryptophthalmos and gourd shaped eyes with systemic features of Fraser syndrome. Such a novel presentation of Fraser with gourd shaped eyes has not been reported.

Twists and Turns in Strabismus - A Case Series of Patients with Abnormal Head Postures

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Abstract:

Aim: *To study the cases of head postures presenting at a Tertiary Eye care centre.*

Materials & Methods: *A total of 15 patients with head posture who had presented to our out-patient sub-specialty clinic were included in the study. Five patients presented with diplopia in one or more gazes, three patients presented with head tilt to either side, four patients with horizontal deviation, one patient each with nystagmus, face turn and chin elevation. All patients underwent detailed ocular examination and squint work-up. Depending on the cause of incomitance, they were taken up for surgery or managed conservatively. After surgery, they were followed up at one week, one month and three months. The post-operative deviations and other related findings (head posture, diplopia) were recorded. The data was analyzed at the end of the study.*

Result: *The mean patient age was 20.36 years (range 4.5 – 71 years). Out of the total 15 patients, 12 were male and three were female. 12 (80%) patients had best corrected visual acuity of > 20/40 whereas three (20%) patients had BCVA of 19/40-20/100 (both eyes). Out of 15 patients, seven patients were diagnosed as a case of Superior oblique palsy, two patients each were diagnosed as esotropia and sixth nerve palsy and one patient each as Exo-DRS, manifest latent nystagmus with left face turn and exotropia, isolated inferior rectus palsy and acquired Brown's syndrome. Seven patients with superior oblique palsy underwent, total anterior positioning of inferior oblique in five cases and antero-nasal transposition of inferior oblique in two cases. In two cases of sixth nerve palsy and two cases of Esotropia, medial rectus recession was done. Patient of Exo-DRS underwent 08 mm lateral rectus recession with Y split whereas patient of nystagmus underwent Augmented Anderson procedure. One patient of acquired Brown's syndrome was managed with oral albendazole whereas one case of isolated IR palsy (myasthenia gravis) was treated with oral pyridostigmine, prednisolone and Mycophenolate mofetil. Head postures was fully corrected in 12 (80%) patients and partially corrected in remaining three (20%) patients.*

Conclusion: *Head posture can be present in various strabismus namely nerve palsies, restriction (congenital, traumatic or acquired) or neuro-muscular junction problems (ocular myasthenia). The treatment may range from observation only during initial phase, followed by surgical intervention in cases of extra-ocular nerve palsies and in some cases, it can improve by medical management alone. Surgical correction can result in resolution of head posture, diplopia and improvement in stereo-acuity.*

Keywords: *Incomitant squint, diplopia, head tilt.*

INTRODUCTION:

Incomitant strabismus is a deviation which is different in various fields of gazes and can be caused by oblique muscle dysfunction, extra-ocular muscle paresis, ocular restriction or can be associated with a primary A or V pattern deviation[1]. Limited ductions on extra-ocular muscle movements associated with squint is secondary to extra-ocular muscle paresis, ocular restriction or both. The three major causes of muscle paresis are cranial nerve paresis, primary muscle disease and mechanical disadvantage of muscle pull[1].

Mechanical restriction of eye movement is caused by adhesions to an extra-ocular muscle or sclera, a tight or inelastic extra-ocular muscle or an orbital mass. The principle diagnostic tests that differentiate paresis from restriction include saccadic velocity measurements, forced ductions and forced generation test[1]. The deviation of the squinting eye with the normal eye fixing is called the primary deviation whereas the deviation of the normal eye with the paretic eye fixing is called secondary deviation. In incomitant squint, secondary deviation is more than primary deviation. There are three main distinctive characteristics to distinguish incomitant from comitant squints – history of diplopia, past pointing, head posture, associated neurologic findings and lack of sensory adaptations[2].

Management of incomitant squints involves releasing the restriction, if it is the cause of limited motility of extra-ocular muscle. If it is secondary to poor rectus muscle function, then we have to address the muscle weakness. In cases of normal eye movements or little restriction, we can operate on the good eye to

match ocular rotations of the deviated eye[1]. The followed principle is recession procedures have their greatest effect in the field of action of the recessed muscle, and the resections produce a leash with the greatest effect occurring when the eye rotates away from the resected muscle. Sometimes, Faden operation has also been suggested to reduce incomitance[1].

MATERIALS & METHODS:

This was a retrospective hospital based study performed to ascertain various presentations and management of cases of incomitant squint who presented in our squint clinic. The study sample was comprised of 15 patients (12 male and three female) over a period of one year. Patient follow-up data was reported three months after surgery.

The inclusion criteria were patients with incomitant strabismus, diplopia, abnormal head posture or acute onset strabismus. Patients having comitant strabismus or history of any previous squint surgeries were excluded from the study. A well informed consent was taken from the patients.

A standardized examination protocol was followed which included measurement of distant visual acuity, near vision, detailed ocular examination followed by detailed squint evaluation including recording head posture and extra-ocular motility. All patients underwent PBCT, diplopia charting and sensory evaluation with Bagolini, TNO and lang's two pencil test if no gross stereo-acuity was found on TNO. All patients with nerve palsy or acute onset strabismus underwent imaging of brain. Forced duction test was done in all the cases as part of pre-operative assessment. All patients were followed-up at one week, one month and three months. At the end of three months, pre-operative and post-op data was analysed.

RESULTS:

The mean patient age was 20.36 years (range 4.5–71 years). 10 patients were less than 25 years of age whereas four were in 25- 50 years age group and one patient was above 50 years. Out of the total 15 patients, 12 (80%) were male and three (20%) were female.

Clinical Presentation:

Of the total 15 patients, 05 (33%) patients presented with diplopia in one or more gazes, three (20%) patients presented with head tilt to either side, four (27%) patients with esotropia or exotropia, one (7%) patient with nystagmus, one (7%) with face turn and one (7%) patient with chin elevation.

Best corrected visual acuity (BCVA) on presentation (Snellen's chart): 12 (80%) patients had best corrected visual acuity of > 20/40 whereas 3 (20%) patients had BCVA of 19/40- 20/100 (both eyes).

Diagnosis:

Based on the clinical evaluation, seven (46.7%) patients had Superior oblique palsy, two (13%) had esotropia and sixth nerve palsy each and one patient (6.5%) each was diagnosed as Exo-DRS, manifest latent nystagmus with left face turn and exotropia, isolated inferior rectus palsy and acquired Brown's syndrome.

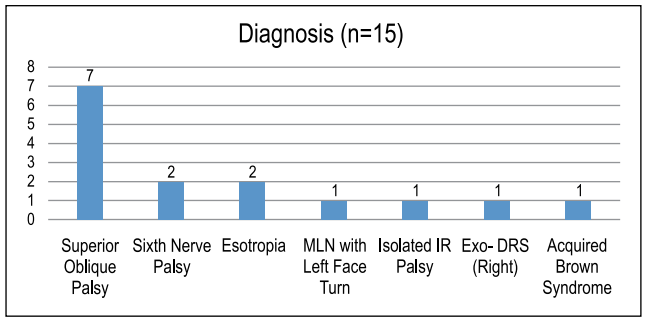

Fig1- Diagnosis of head posture at presentation

Surgery done:

Out of total seven (46.7%) patients with superior oblique palsy, five (33.3%) cases underwent total anterior positioning of inferior oblique and two (13.3%) cases underwent antero-nasal transposition of inferior oblique. In two (13.3%) cases each of sixth nerve palsy and Esotropia, medial rectus recession was done. Patient of Exo-DRS underwent 08 mm lateral rectus recession with Y split whereas patient of Exotropia with MLN underwent part of Augmented Anderson procedure with only LR recession of 12 mm.

Non-Surgical treatment:

One (6.5%) patient each of acquired Brown's syndrome and isolated IR palsy (myasthenia gravis) were treated with oral albendazole and oral pyridostigmine respectively.

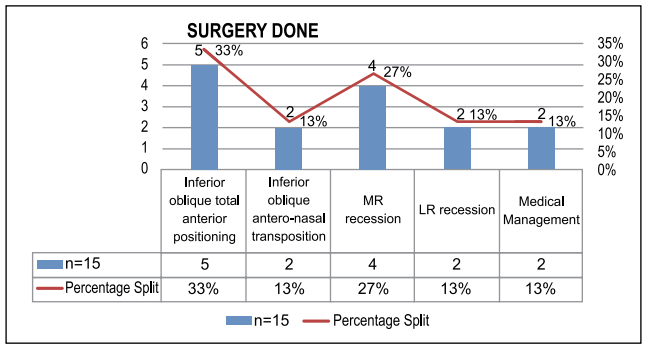

Fig 2- Surgery Performed for correction of head posture

Stereo-acuity:

On presentation, 10 (66.6%) patients had gross stereo-acuity (stereo-plates were identified in plates I-III of TNO (The Netherlands Organisation for Applied Scientific Research)) whereas remaining five (33.3%) had no gross stereo-acuity. Stereo-acuity was measured initially with the help of TNO test and in patients having no stereo-acuity on TNO, Lang's two pencil test was done to check for any gross stereopsis.

Post-operative/treatment showed improvement in stereo-acuity in 6 patients whereas 9 patients had no change in their stereo-acuity after surgery.

Deviation (in prism diopters):

Out of the total 15 patients, two (13.5%) had horizontal deviation of < 30 PD whereas three (20%) had horizontal deviation of > 30 PD. Six (40%) patients had mixed deviation (both horizontal and vertical deviation) whereas three (20%) patients had vertical deviations only.

Diagnosis	Count (n=15)	Deviation
Superior Oblique Palsy	7	Mean: 9.83 PD (Max: 14 PD, Min: 3 PD), vertical deviation
Sixth Nerve Palsy	2	25 PD BO/4L/R and 85 PD BO
Esotropia	2	30 PD BO
MLN with Left Face Turn	1	35 PD BI
Isolated IR Palsy	1	3 L/R
Exo- DRS (Right)	1	35 PD BI
Acquired Brown Syndrome	1	8 PD BI/ 12R/L

Post-operatively, mean deviation (vertical) in cases of superior oblique palsy was 3.5 PD (Max: 5 PD, Min: 2 PD). In cases of sixth nerve palsy and esotropia post MR recession, mean deviation was 9 PD (Max: 13 PD, min: 4 PD). Post treatment deviation in case of MLN and Exo-DRS (right) was 10 PD BI. Post treatment deviation in Acquired Brown Syndrome was 6 PD BI in primary gaze and orthotropia in patient of isolated inferior rectus palsy.

Fig.3- Preop primary deviation

Head Posture and diplopia was corrected in nine (60%) of patients and small residual head posture with diplopia in few gazes was seen in six (40%) of patients.

DISCUSSION:

Incomitant squint can be caused by ocular restriction, muscle paresis or an oblique muscle dysfunction[1]. It can present with varied clinical presentation such as diplopia, abnormal head posture, deviations of eye or nystagmus. Evaluation involves saccadic velocity measurements, forced duction test, forced generation tests of suspected muscle, intra-ocular pressure change on eye movement and lid fissure change on eye movement besides complete ocular examination including refraction and squint evaluation.

Surgery is mostly indicated in cases of long standing and stable nerve palsies or cases of restrictive squints, as it carries a risk of errors and/or complications, which can occur during assessment or per-operatively. Irsch has provided an overview of clinically-relevant optical issues in squint evaluation with emphasis of sources of error in angle measurements and ways to avoid pitfalls when using prisms to measure ocular deviations and when observing the corneal light reflex[3]. Olitsky and Coats reviewed the most common and serious surgical complications of strabismus surgery and various means to reduce them[4].

Complexity of extra-ocular muscle orbital anatomy should be recognized and is the most significant scientific advancement as reviewed by Clark who studied concepts of extra-ocular muscle pulleys, pulley disorders as a cause of incomitant squint and surgical techniques specifically focusing on pulley disorders[5]. Leuder reviewed common orbital factors that can underlie incomitant squint other than EOM pulleys[6]. Stager and colleagues reviewed the inferior oblique muscle anatomy, different weakening procedures, and recent surgical techniques that take advantage of the muscle's unique anatomy for the treatment of additional indications such as excyclotorsion and hypertropia in primary gaze[7].

In our study out of seven (46%) patients of superior oblique palsy, five (33.3%) underwent total AP of IO, which is the most commonly operated muscle in cases of SOP as observed by Pilar Merino Sanz et al[8]. and also seen in our study. Two (13.3%) patients underwent IO ANT with good results. Inferior oblique antero-nasal transposition has resulted in good long-term outcomes in patients of SOP presenting with hypertropia, inferior oblique overaction and extorsion in primary gaze as also concluded by Stager D et al[7].

In our study, we performed recession of 8 mm with Y-split of LR which resulted in correction of exotropia, upshoot, downshoot and globe retraction, same has been recommended by Venkateshwar B Rao et al[9].

Management of sixth nerve palsy can be divided into those directed at first six months after onset which involves occlusion to treat amblyopia, temporary Fresnel prisms and medial rectus chemodenervation as recommended by Robert A Clarke, MD[10]. If the palsy persists beyond 6 months, surgical options like medial rectus recession or lateral rectus resection can be considered. In two of our cases of sixth nerve palsy which persisted after more than a year, MR recession was done to improve head posture.

One of our case of isolated IR palsy was diagnosed as myasthenia gravis without ptosis and was started on oral pyridostigmine. The etiology of isolated IR palsy can be congenital, traumatic, myasthenic or vascular[11]. The presenting symptoms can be abnormal head posture, diplopia (in our case) or disfiguring diplopia as reported by Colavito J et al[11]. Our case improved with medical management alone.

Acquired Brown syndrome due to myocysticercosis of superior oblique is not so common condition, as superior rectus muscle is most commonly involved muscle in extraocular muscle cysticercosis[12]. The most common symptoms are proptosis, pain and restriction of extraocular movements. It should be differentiated from orbital metastasis, myositis other parasitic infections like hydatid cyst[13]. Diagnosis is often based on the findings of imaging namely high-resolution ultrasonography, CECT and MRI. It is important to rule out any CNS or intraocular involvement. Treatment is in the form of oral

steroids (1 mg/kg body weight) and Tab Albendazole. (15mg/kg body weight)[14]. Our patient also showed good recovery with the same medical management.

In cases of Manifest latent nystagmus, Augmented Anderson procedure can be done where recession of the yolk muscle is performed[15]. For a face turn to the left, the LR of the right eye is recessed and the MR of the left eye is recessed[15]. In our case, we did part of Augmented Anderson procedure which involved only LR recession in right eye which showed improvement in head posture.

In our study we found that each case of head posture when evaluated and treatedindividually, can result in improvement of head posture, stereoacuity and most importantly diplopia free primary and down gaze atleast.

CONCLUSION:

Head posture is a motor adaptation in strabismus and can be a presenting feature in various types of incomitant strabismus namely nerve palsies, restriction (congenital, traumatic or acquired) or neuro-muscular junction problems (ocular myasthenia). Meticulous history, clinical examination supported by investigations can help in diagnosis in most of the cases. The treatment may range from observation only during initial phase, followed by surgical intervention in cases of stabilized or long standing extra-ocular nerve palsies, although sometimes it may be caused due to myasthenia gravis or myocysticercosis in which case most of the times, medical management alone is required.

The main aim of the surgery is resolution of diplopia (in primary and downgazes), increase in binocular single field of vision and correction of head posture.

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Case Report of Syndromic Bilateral Complete Cryptophthalmos with Gourd Shaped Cystic Eyeballs; A Novel Fraser Syndrome Phenotype

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Abstract: *Syndromic Cryptophthalmos, more common than isolated, is eponymously seen in Fraser syndrome. However, it is not a sine-qua-non for diagnosis and ocular involvement in Fraser syndrome is protean with cryptophththalmos(CO), microphthalmos, anophthalmos, microcornea, iris colobomas, lid colobomas and myriad other presentations. Fraser syndrome, has mutations in FRAS1, FREM 2 and GRIP2 genes. FREM 2 mutations can give rise to both isolated CO with gourd shaped eyes and syndromic CO associated with Fraser syndrome. Gourd shaped cysts have been previously reported with isolated complete cryptophththalmos, however, not with Fraser syndrome and we present this hitherto unreported novel presentation of bilateral cryptophthalmos with gourd shaped cysts.*

Keywords: *Cryptophthalmos, Fraser syndrome, Renal aplasia, syndactyly, umbilical hernia*

Key Messages: *Cryptophthalmos is a psychologically traumatising disorder presenting to an ophthalmologist which may have serious underlying systemic associations and requires thorough workup. Here we present a case of large gourd shaped cysts in a patient of Fraser syndrome with syndactyly, umbilical hernia, ambiguous genitalia, renal agenesis and tongue tie.*

INTRODUCTION

Fraser syndrome is a constellation of features including bilateral cryptophthalmos, syndactyly, abnormal genitalia and may be associated with systemic abnormalities like renal agenesis and cardiac disorders. Its incidence is estimated to be around 0.043 per 10000 live birthsand around 200 cases have been reported worldwide with only few anecdotal reports from India.[1] We present a case of large gourd shaped cysts in a case of Fraser syndrome not described before.

CASE HISTORY:

A 1 year old child presented with inability to open eyes, ambiguous genitalia and fused digits since birth. A product of third-degree consanguineous marriage, child was a full-term normal vaginal delivery with birth weight of 2.5kg with bilateral complete cryptophththalmos (CO), hypertelorism, flat broad nasal root, bifid nasal bridge and everted stenotic choanae. (Fig 1). Microtia with low set ears was noted. Eye brows were absent and a tongue of hair descended towards brow area from the temple. A cystic structure was palpated under the skin on either side of the nose and child responded by crying. No lid structures or lacrimal appendages were discernible.

Fig 1: Bilateral cryptophthalmos, flat nasal root and hypertelorism

Incomplete cutaneous syndactyly between the 1st 2nd and 3rd fingers of the right hand and the 2nd and 3rd finger of the left hand along with thenar hypoplasia was present. Incomplete cutaneous syndactyly in the feet between the 2nd 3rd and 4th toe on the right and the 2nd and 3rd toe on the left was also noted. There was an umbilical hernia which increased on crying (fig 2A). The child had ambiguous genitalia with clitoromegaly and absent vagina. The frenulum of the tongue was short with microstomia (Fig 2 B).

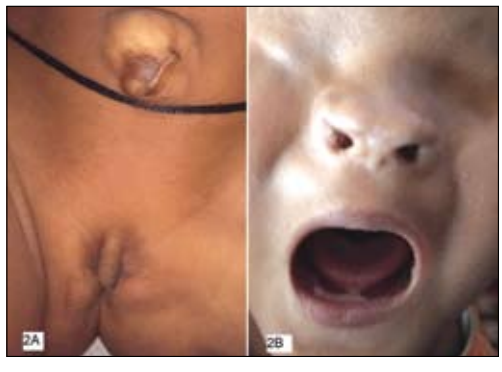

Fig 2A: Umbilical hernia with ambiguous genitalia
2B: Tongue tie seen on crying

Contrast enhanced MRI of the brain and orbits revealed bilaterally elongated gourd shaped cystic eyeballs, extraocular muscles appeared normal in dimensions and insertions, optic nerve appeared normal in diameter but was kinked, pushed posteriorly and medially. The intraocular structures like crystalline lens, cornea, anterior chamber and iris were not discerned. Ultrasound (USG) orbits confirmed gourd shaped cysts with no evidence of ciliary body, choroid or retina. The axial length was 28 mm and 29 mm right and left side respectively (fig 3A).Contrast enhanced MRI pelvis revealed anterior abdominal wall defect with mild herniation of small bowel loops consistent with umbilical hernia. (3B)Uterus, vagina and ovaries were not visualised. USG Kidney, ureter, bladder revealed right renal agenesis (fig 3C).

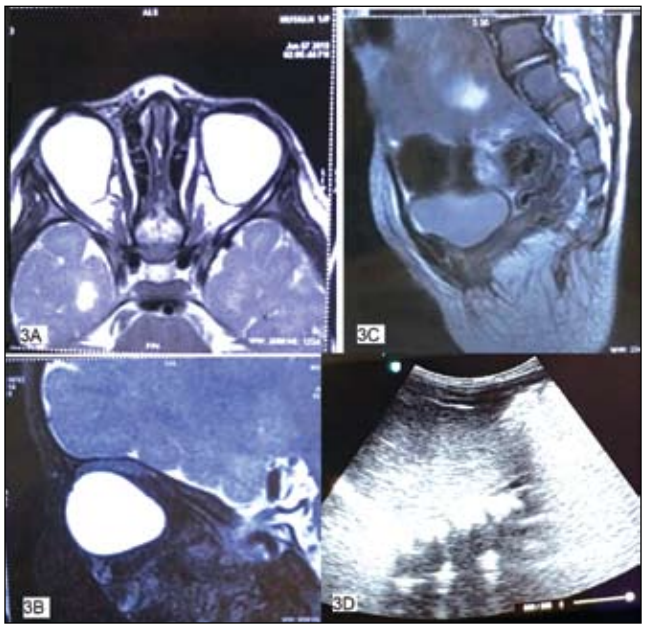

Fig 3A&B: Elongated cystic eyeballs with no discernable cornea, lens, anterior chamber seen on contrast enhanced MRI(CEMRI)
Fig 3C Anterior abdominal wall defect and mild herniation of small bowel loops seen on CEMRI pelvis
Fig 3D: Ultrasound KUB: Right renal agenesis

Roentgenograms of limbs revealed simple soft tissue syndactyly with no bony fusion of right 1st, 2nd and 3rd finger and left 2nd and 3rd finger with absent distal end of distal phalanx of thumb bilaterally and distal phalanx of 4th finger bilaterally. lower limb showed syndactyly of the 2nd and 3rd toe (fig 4). Echocardiography showed no cardiac abnormality. Linkage analysis could not be done. No surgery was contemplated and parents were counselled regarding systemic implications and nil visual prognosis.

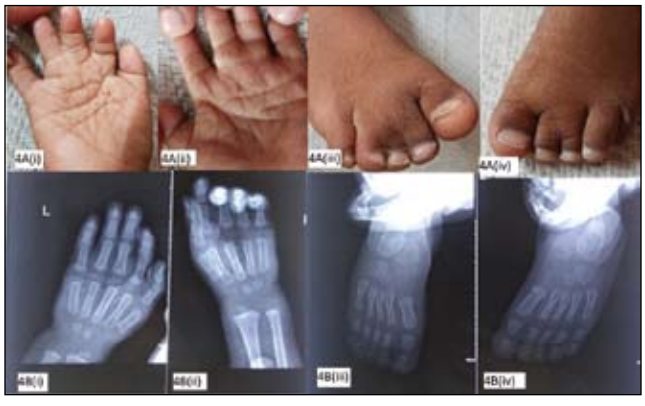

Fig 4a(i-iv) Clinical photographs showing syndactyly of both upper and lower limbs
4B(i-iv) Roentgenograms showing no bony fusion; absent distal end of distal phalanx of thumbs bilaterally and distal phalanx of 4th finger bilaterally

DISCUSSION:

Cryptophthalmos is the congenital absence of eyelid with skin passing continuously from the head to the cheeks over a malformed eye seen in syndromic or isolated form. Fraser syndrome showcases syndromic CO, ocular findings are protean including unilateral or bilateral CO, microphthalmos, anophthalmos, lid colobomas, absence/malformation of ocular adnexa.[2-4] Syndromic CO is also seen in Manitoba oculotrichoanal (MOTA) syndrome.[5] Isolated CO is seen far less frequently[6-8] and could be inherited as autosomal dominant/autosomal recessive or be sporadic.

The constellation of findings of cryptophthalmos, syndactyly and abnormal genitalia in our case conforms to the major criteria for diagnosis of Fraser syndrome which enjoys tremendous phenotypic and genotypic heterogeneity prompting many authors to suggest major and minor criteria for diagnosis, nevertheless there is no unanimity on the subject and no single anomaly including CO is universally present.[3,4] This child met the diagnostic criteria as proposed by multiple authors as noted above.

Fraser syndrome is known to arise from mutations in FRAS 1, FREEM 2 and GRIP1 genes. The genes encode proteins of extracellular matrix that are essential for adhesions between basement membrane of the epidermis and adjoining connective tissues of the dermis during embryological development. FRAS1 related extracellular matrix protein 2 is a member of FRAS1, FREM 2 family that directly interacts with connective tissue constituents through chondroitin sulphate proteoglycan motifs contributing to epithelial mesenchymal adhesions. FREM 2 is produced by epithelial cells and deposited in the basement membranes of epidermis, peri ophthalmic region, lungs, neural tube and kidney and may thus may share great genotypic and phenotypic heterogeneity.

Fraser syndrome can be caused by 3 different loci. FRASRS1 is caused by homozygous or compound heterozygous mutations in gene 607830 on chromosome 4q21.21. FRASRS2 is caused by homozygous mutations on chromosome 13q13 (OMIM617666) on FREM2 protein. FRASRS3 is caused by homozygous mutations on GRIP1 gene on chromosome 12q14.3, only 3 consanguineous families have been reported. FREM 2 mutations on chromosome 13q13 (OMIM 123570) also give rise to isolated bilateral complete CO without systemic associations. Bilateral gourd shaped cystic eyes seen in this child have been reported with isolated CO with FREM 2 mutations (6089450004) but not with Fraser syndrome with FREM 2 mutation (6089450001).

Egier et al reported a case of isolated bilateral CO with retinal detachment, optic nerve coloboma and Peter's anomaly.[7] Gourd shaped eyes were not reported and an autosomal dominant inheritance was proposed. Kanhere et al reported a single case of isolated bilateral complete CO with gourd shaped eyes, no genetic studies were carried out.[8]

Yu et al reported a three year old girl with isolated right unilateral complete CO with gourd shaped eyes due to missence mutation in FREM 2.[9] The other eye was normal.

Zhang et al reported three unrelated Chinese children from non-consanguineous families with isolated bilateral complete CO with bilateral gourd shaped cystic eye balls and documented FREM 2 nonsense mutations.[10] FREM 2 was also localized to the outer plexiform layer of the retina implicating profound influence of FREM 2 on retinal differentiation. No lid structures like corneal, lens, iris were identified, vascular and pigment tissues could not be identified as retinal and choroidal tissues. No other systemic anomalies were detected and all 3 attained normal developmental milestones.

After neural tube formation, the lateral part of non-neural ectoderm makes contact with developing optic vesicle and begins to differentiate into lens. This non neural ectoderm known as the pre placoid ectoderm includes the presumptive fields of lens, nasal, ear, -adeno-hypophyseal, trigeminal and epibranchial placodes, explaining diverse involvement. Signalling from the neural retina is responsible for formation of secondary lens fibres. FREM2 expression has been shown in outer plexiform layer of the neural retina, if that does not happen retina may not differentiate and lens may also fail to develop as a downstream effect. This may explain gourd shaped cysts with aborted embryogenesis. The extraocular muscle develop normally as globe is present and normal optic nerve portends that optic stalk has developed normally.

The present phenotype likely conforms to a genotype of FREM2 mutation with isolated bilateral complete CO and gourd shaped eyes, however systemic features of Fraser have not been documented with gourd shaped cystic eyes and bilateral complete CO.

CONCLUSION:

Present case may represent a new mutation in FREM2 with more extensive affectation and underscores extreme genotypic and phenotypic heterogeneity between isolated Cryptophthalmos and Fraser syndrome and both may be running conterminous. Future studies in genetics and neuro imaging will offer new insights into these rare disorders and locate missing pieces of the jigsaw.

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Congenital Cryptophthalmos Treated Successfully by Surgical Treatment

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Abstract : *Cryptophthalmos is a congenital condition usually recognized by skin fold from forehead to cheek. The ocular content is usually rudimentary. It is classified into three types: complete, incomplete and abortive. Failure of eyelid separation can be associated with maldevelopment of the underlying cornea and microphthalmia. We found a case with congenital Cryptophthalmos, which was associated with cystic globe and maxillary hypoplasia. We decided to treat this patient surgically in three sessions.*

METHOD:

A 2 months old girl was brought to us with abnormality in her left eye since birth. The patient was born by spontaneous vaginal delivery after full term gestation. Antenatal history was unremarkable. There was no history of consanguineous marriages in the family. Ocular examination did not reveal any abnormality in her right eye. The lower eye lid, except in the lateral 4mm portion which had lid margin and eyelashes, was replaced by a fold of skin Patient was having rudimentary eyeball and rudimentary tarsal plate which was confirmed on ultrasound B scan, puncta without canalized lacrimal system. Extending from the lateral part of the nose was a large cystic mass. (Figure-1 A) Patient found to have absent bone during surgery, Histopathological sample showed dermoid cyst during histopathological examination.

Fig-1: Presentation (A) Image following cyst removal (B)

The patient underwent surgery, first for removal of the cyst (Figure-1 B) fornix reconstruction (Figure-2 A) followed by lid reconstruction with skin graft (Figure-2 B) and fitted artificial (Figure-2 C) There are only 2 cases reported in till now.[1,2]

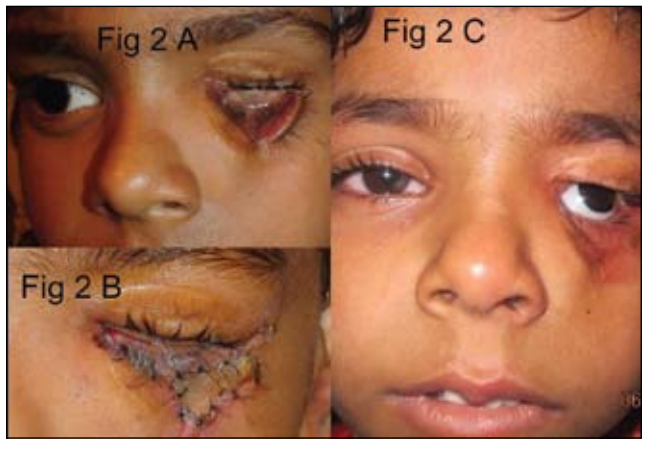

Fig-2: Image following fornix reconstruction(A) Image following lid reconstruction and skin grafting(B) Final Image with prosthesis(C)

Author is not aware about many reports of rudimentary cystic eye ball treated surgically and achieved cosmetic correction with follow up of 6 years.

CONCLUSION:

Cryptophthalmos can be treated successfully by surgical treatment.

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The 'Evil Eye'; from Antiquity to Posterity

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Since antiquity, the evil eye has persistently and incessantly chased the mankind, taking diverse forms and templates, transcending almost all cultures. From petrifying gaze of Greek Gorgons to Irish folk tales of men able to bewitch horses with a single stare, virtually every culture has fables relating to evil eye. The genesis of evil eye is lost in antiquitybut devices to ward it off are in evidence since 3300 BC. As a source of vision, awareness and knowledge, its natural that such beliefs were woven around eyes. The universal belief at the core is that glances full of frustration, envy, jealousy, revenge can have devastating consequences.

The underpinnings could lay in the ancient Greek and Roman beliefs that vision was possible by directing the light outward onto objects rather in the reverse by receiving it and processing it. Plutarch, the Greek philosopher and a priest at the Delphic oracle of Apollo suggested that the human eye released invisible rays of energy that in some cases were potent enough to kill. Plutarch claimed that some people possessed even stronger ability to fascinate and uncannily proficient in bestowing the curse. Heliodorus of Emesa, came as a Greek ambassador to India and built the Heliodorus pillar at Vidisha (MP), in Aethiopica, writes "When anyone looks at what is excellent with an envious eye, he fills the surrounding atmosphere with a pernicious quality and transmits his own envenomed exhalations into whatever is nearest to him" The evil eye is so deeply embedded in human civilization that in-spite of it's totally pagan connotations, it even finds place in religious texts like the Bible and the Quran.

'Eat thou not the bread of him that hath an evil eye, neither desire thou his dainty meats;'-Proverb 23.6, Bible (KJV version 1900).

The eye thus was believed to be charged with tremendous power that could be translated into maliciousness. Brides and children (boys) were considered natural targets of envy. The veil, the black teeka on forehead of newborn, the kajal, the black Kardhani around the waist of the young boys and Nazar Battu hanging at the door entrances are all devicesto ward off evil spirit. Turkey and many other countries have a nazaramulet to ward off evil eye. The blue eye shaped amulet is hung everywhere from window frames to handbags. The blue color distracts evil energy, the amulet averts the bad energy and protects you. Evil eye has also entered world of fashion with launching of countless eye bracelets, necklaces and key chains to avert it. In ancient Egypt, the Eye of Horus also known as Wadjet pendant was buried with Pharaohs to protect them in afterlife. Even the Rx symbolused today can be traced to Egyptian 'Eye of Horus' symbol, emanating from a myth about protection from evil.

Cross eyed people have had their own share of superstitions attributed to them, to prevent ill luck from meeting a squint eyed person, you must spit 3 times over your left shoulder. Ancient high status Mayan mothers attempted to induce esotropia by hanging a piece of thread between infant's eyes with a stone or ball of resin attached that caused infant's eyes to focus on it , eventually causing esotropia. This was done in honor of Kinich Ahau, the cross eyed Sun God in their culture, crossed eyes were considered a thing of beauty for the Mayan women.

It was held in many societies that If a cross eyed person looks at you, you will have ill luck all day, for such people can see right through you and know your thoughts. Persons with one eye set lower than other (hypertropia) have been commonly regarded as witches. This is the evil eye, the dreadful eye that seems to regard us not but 'sees' all there is to see. Another superstition that children with strabismus will grow out of it is a dangerous old wives' tale with tremendous public health connotations. In many regions and cultures in India, it is held that a child born with crossed eyes, brings good luck to the family hence no treatment is warranted.

In Slavic mythology, Likho / Liho/ Licho, a creature with one eye often depicted as an old skinny woman in black or as a male goblin of forests, entails evil fate and misfortune. In many parts of India if you encounter a one eyed person early in the morning, your day is ruined.The disfigured eye was also considered a visionary in worst sense. It knows already what will befall us, we tend appear reflected upside down. Even blindness could be forestalled by wearing gold earrings. Sailors wore them to staunch ocean blindness. Children wore them to stall sore eyes (trachoma). Depending upon the gender and eye of the person, eye twitching has also been a source of good or bad luck across many civilizations.

Iris color has traditionally been associated with evil / not so evil eye. Blue eyes are sign of good luck, People with blue eyes are thought to be intelligent, pure and insightful. Blue eyed have best memories, possess honest nature, gifted to see the truth, and have lasting relationships. Those with pale blue are thought to be deviant and evil and not to be trusted. Brown is the commonest color, people with brown are thought to be trustworthy, humble. Brown eyes are a sign of independence and such people have a great sense of humor. Green is one of the rarest colors. Green eyes have been center of superstitions and beliefs worldwide related to witches, magic, vampires and evil spirits. Heterochromia has been a source of many a superstition including that such people can see earth and heaven at the same time.

The evil eye is thus a vestigial remnant from the very dawn of human civilization, transcending geographical, cultural and religious barriers, a repository to our most esoteric and everlasting beliefs. Over many millennia, the superstitions live on and refuse to die out.

A deep man believes that the evil eye can wither, the heart's blessings can heal, and that love can overcome all odds... Ralph Waldo Emerson.

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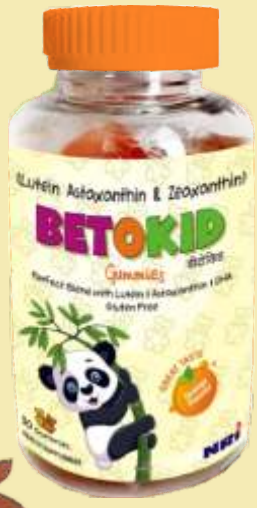

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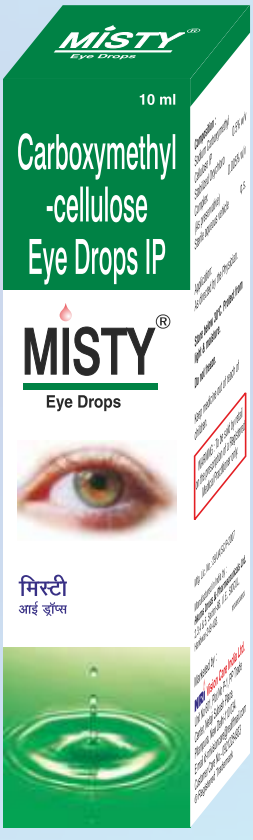

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