Congenital ptosis - A clinical and demographic study in a tertiary eye care hospital

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Abstract

Aim: To study the clinical and demographic characteristics of Congenital Ptosis at a tertiary eye care centre.

Material and Methods: This is a retrospective study of patients admitted for surgical correction of ptosis under the department of Oculoplastics and Orbital Diseases in Sarojini Devi Eye Hospital, Hyderabad, Telangana State, over a period of 3 years from Feb 2013 to Jan 2016. Demographic data regarding age at presentation, gender, family history of ptosis and any other associated abnormalities were recorded. A detailed clinical examination was done for all the cases to determine the cause of ptosis, degree of ptosis, amount of LPS action, presence of strabismus and/or amblyopia and fundus examination. All cases of Acquired Ptosis were excluded from the study.

Observations: A total of 123 cases were reviewed in our study 56.90% cases presented after 16yrs of age. Children with congenital ptosis presenting at less than 5yrs of age constituted 8.1%. In our study 71.54% cases were unilateral. Involvement of Left eye was predominant at a rate of 60.22%. Severe ptosis (degree of ptosis 4mm and more) was present in 90.24%. Function of Levator palpebrae superioris was poor in 73.17%.

No sex predilection was noted in our study.

A family history of congenital ptosis was present in 14% of cases. 3 cases of congenital ptosis were because of 3rd nerve paralysis.

Conclusions: Simple congenital ptosis was the most common form of childhood ptosis. Uniocular involvement was found in 71.54% and a predominant involvement of the left eye in 60.22% of cases.

Keywords: Congenital ptosis, LPS function, Frontalis sling surgery.

Introduction

Blepharoptosis or commonly known as ptosis is drooping of upper eyelid below its normal position, is a relatively common form of eyelid malposition in children. Congenital ptosis comprises a group of diseases in which there is drooping of the upper eyelid due to developmental dystrophy of the Levator Palpebrae superioris[1] congenital 3rd nerve palsy, synkinetic ptosis, ptosis with elevation abnormality and Blepharrophimosis syndrome. Ptosis present either at birth or developing within the first year of life is called congenital ptosis. Most cases of congenital ptosis are idiopathic in nature. Familial occurrence suggests genetic or chromosomal defect. They can be unilateral or bilateral. Most of the congenital cases are unilateral and vision is not usually affected. There is no sex predilection. This may be associated with anisometropia, astigmatism, strabismus and amblyopia[2]. Simple Congenital ptosis can be corrected surgically.

Materials and Methods

This is a retrospective study of patients admitted for surgical correction of ptosis under the department of Oculoplastics and Orbital Diseases in Sarojini Devi Eye Hospital over duration of 3 years from Feb 2013 to Jan 2016.

A detailed clinical examination was done for all the cases to determine the age of onset, degree of ptosis, amount of LPS action, and presence of strabismus and/or amblyopia and fundus examination.

An analysis of age at presentation, sex distribution, severity of ptosis, amount of LPS action, presence of Bell's phenomenon and other eye protective mechanisms like corneal sensations, Schirmer's test were done. Examination of visual acuity, ocular movements and presence of Marcus-Gunn jaw winking phenomenon were noted.

All cases of Acquired Ptosis were excluded from the study.

All the children were subjected to cycloplegic refraction and fundus examination and pre-operative clinical photographs taken. A basic haematological workup was done. After pre-anaesthetic assessment patients were posted for surgery under general or local anaesthesia.

A follow up was done for a period of 1 year.

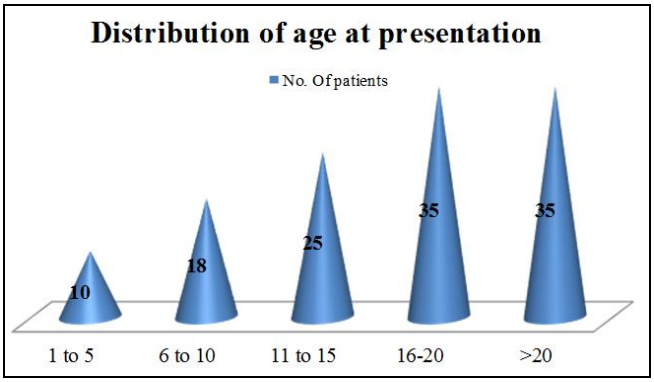
Commonly observed post-operative complications were under-correction, sutural granuloma and exposed silicon sling.

Results

A total of 123 patients with congenital ptosis admitted under the department of Oculoplastics and Orbital diseases in Sarojini Devi Eye Hospital, Hyderabad were included in the study.

56.90% of cases presented after 16yrs of age. Children with congenital ptosis presenting at less than 5yrsof age constituted 8.1%. Children under the age of 10yrs constituted 22.73%. [Graph 1]

Graph 1



The late age of presentation in our study could be because of the fact that the child is able to carry out his activities, lack of awareness among parents regarding amblyopia and improved anaesthetic techniques for children, poor socio-economic conditions and non-availability of oculoplasty services widely.

Out of 123 cases males were 69 (56.09%) and females 54 (43.90%)

Severe ptosis (degree of ptosis 4mm and more) was present in 90.24%. Function of Levator palpebrae superioris was poor in 73.17%. No sex predilection was noted in our study. A family history of congenital ptosis was present in 14% of cases. 3 cases of congenital ptosis were because of 3rd nerve paralysis.

Various types of ptosis noted in our study were- Congenital simple ptosis in 89 patients, Blepharophimosis syndrome in 14, Synkinetic ptosis in 6, ptosis with elevation defect in 11, Congenital 3rd nerve palsy in 3. [Table 1]

Congenital simple ptosis was the commonest type noticed in our study accounting for 72.35% of the total. Blepharophimosis syndrome accounting for 11.38% and Congenital 3rd nerve palsy for 2.43% [Table 1](Fig. 1-5)

Table 1

Type of ptosis	No. of patients	Percentage
Simple congenital	89	72.35%
Blepharophimosis syndrome	14	11.38%
Ptosis with elevation defect	11	8.94%
Synkinetic ptosis	6	4.87%
Congenital 3rd nerve palsy	3	2.43%

In our study 71.54% of cases were unilateral.

Involvement of Left eye was predominant at a rate of 60.22%. 35 patients had bilateral ptosis.

A family history of congenital ptosis was present in 14% of cases (Fig. 6)

Amount of ptosis was determined by comparing Margin Reflex Distance 1 (MRD) of both eyes or in cases of bilateral ptosis MRD 1 of <2.5 mm is taken as ptosis. In children who are uncooperative, absence of lid crease and presence of compensatory chin elevation were taken as indicators of severe ptosis.

In our study, an amount of 2mm ptosis is considered mild, 3mm as moderate and 4mm or more as severe. Out of 123, 90.24% [111 patients] were having severe ptosis. [Table 2]

Table 2

Grade of ptosis	No. of patients	Percentage
Mild	10	8.13%
Moderate	2	1.62%
Severe	111	90.24%

LPS function is measured by Berke's method and in children who are uncooperative absence of lid crease and lid covering the pupil and child adopting compensatory chin elevation were taken as indicators of poor LPS function. LPS function is graded as good, fair and poor. 73.17% of patients [90 patients] were having poor LPS function of 4mm or less. [Table 3]

Table 3

Amount of LPS Action	No. of patients	Percentage
Good >12mm	2	1.62%
Fair 5-1mm	31	25.20%
Poor <=4mm	90	73.17%

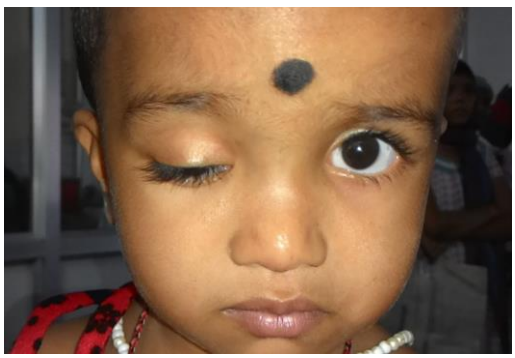


Fig. 1: Congenital ptotis

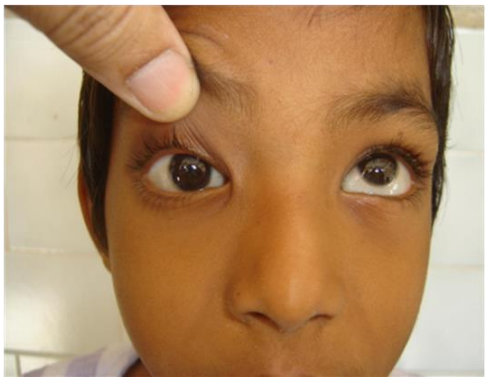


Fig. 2: Ptotis with defective elevation

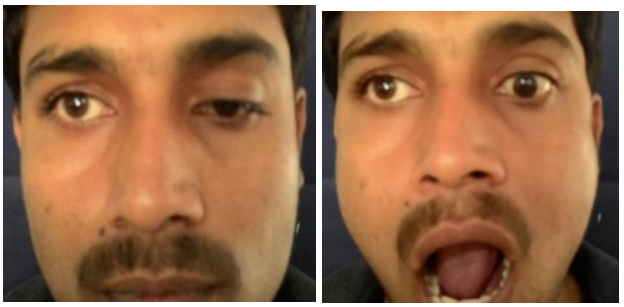


Fig. 3: Synkinetic ptosis (Marcus-Gunn Jaw winking ptosis)

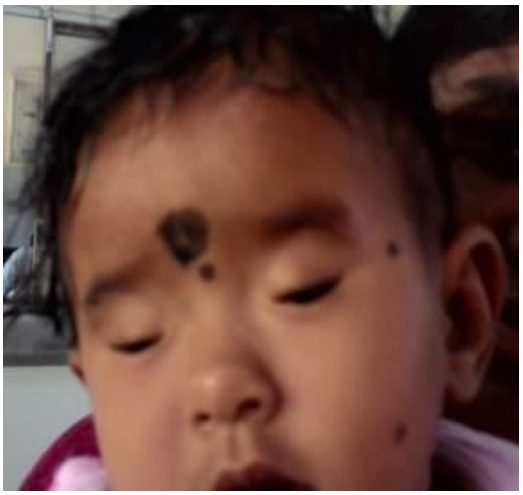


Fig. 4: Blepharophimosis syndrome

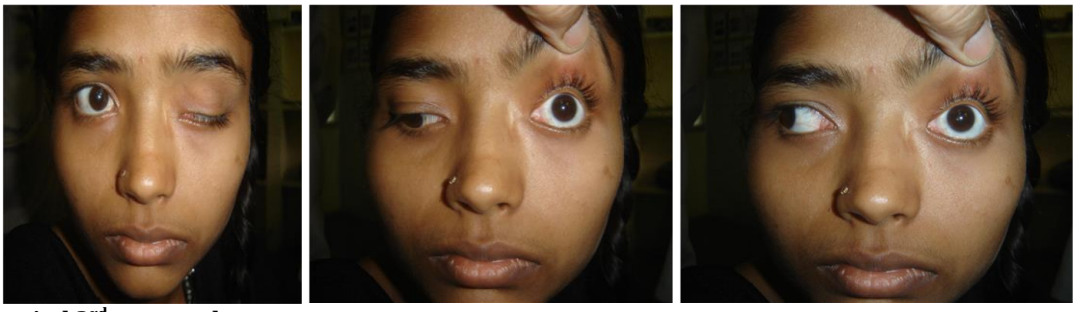


Fig. 5: Congenital 3rd nerve palsy

Discussion

A total of 123 cases admitted with ptosis were evaluated and it was found that simple congenital ptosis was the most common subtype of childhood ptosis diagnosed accounting for 72.35%. A study done by the department of Ophthalmology, Cairo University, Egypt, and a 10 year review of 155 children who presented for corrective surgery in the United Kingdom by Berry-Brincat and co-authors also reported a 68.9% and 74% of simple congenital ptosis which is similar to our study.

In our study Marcus Gunn Jaw winking ptosis is accounting for 5% and Blepharophimosis syndrome for 11.38 of all cases of congenital ptosis. In our study Blepharophimosis syndrome accounted for 11.38% of 123 cases which is higher than the UK study, which reported 4.5% incidence but similar to Cairo study with a reported incidence of 16.7%.[3-5]

In our study males were relatively more affected but there was no statistically significant difference in sex distribution of patients with congenital ptosis.

In our study left eye was involved in 60.22% of unilateral cases which is similar to other studies.[3]

Severe ptosis with 4mm or more was present in 90.24% with poor levator palpebre superioars action in 73.17% which is similar to the study done by Rania El Essawy et al.

Amblyopia was present in 38.21% of cases in our study which is higher than the Cairo study which reported an incidence of 10% of occlusion amblyopia. The higher incidence in our study could be because of inclusion of all cases of amblyopia like anisometropic, strabismic and stimulus deprivation which is similar to other studies reported.[6,7]

Frontalis sling surgery was done in 71.55% cases. The higher percentage of this procedure in our study could be because of fact that majority of patients had severe ptosis with poor levator palpebre superioars action.

We preferred autogenous fasia lata as sling material whenever possible and silicone band in young children of less than 4 year.[8-10] (Fig. 7, 8)

Similar to other studies this study also demonstrates the prevalence of congenital dystrophic ptosis as the leading cause for blepharoptosis in children. Though congenital ptosis presents no difficulty in diagnosis patients presented as late as 60yrs for management for cosmetic purpose.

Uncorrected congenital ptosis may lead to amblyopia or abnormal head posture in cases with severe ptosis. Uncorrected Ptosis can have psychosocial impact on the development of the child. As surgical correction of ptosis gives excellent functional and cosmetic results, parents need to be informed and educated regarding early correction of congenital ptosis.

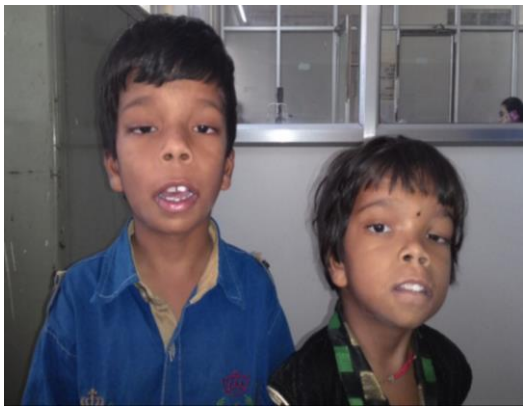


Fig. 6: Ptosis in siblings

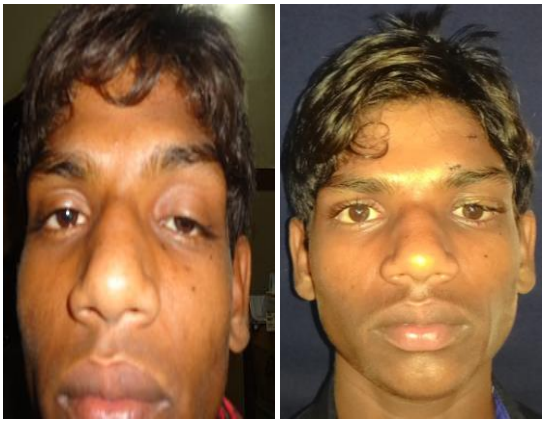


Fig. 7: Pre and post-operative photographs

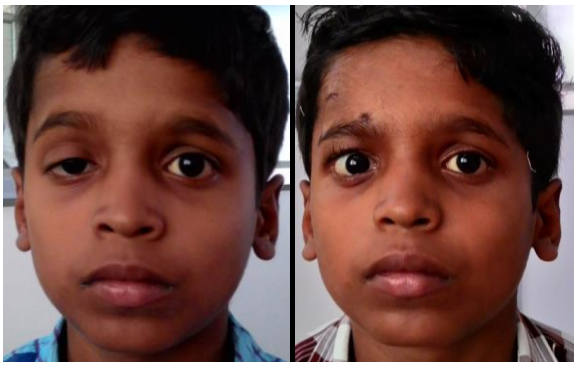


Fig. 8: Pre and post-operative photographs

Conflict of Interest: Nil.

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