Profile and Management of Blepharoptosis Patients in North India - A Two-Year Study

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ABSTRACT

BACKGROUND

Blepharoptosis is a relatively common form of eyelid malposition in both children and adults. Along with being a cosmetic blemish, ptosis is associated with high incidence of amblyopia, especially in young children with severe ptosis. This can have a negative impact on psychosocial development of a child. Hence, it becomes necessary for a general physician as well as for a general ophthalmologist sitting at a primary health care centre, to know the clinical profile of such patients. We wanted to study the demography, clinical presentation and management of blepharoptosis patients seen in our institute over a period of 2 years.

METHODS

45 patients with ptosis who reported at our tertiary care hospital in North India from January 2017 to December 2018, were included in the study. Demographic data, clinico-etiological profile, management of ptosis including outcome of surgeries performed was analysed retrospectively. Data was presented as frequencies and mean ± standard deviation (SD).

RESULTS

The study comprised of a total of 45 patients. The mean age of presentation was 17.4 years. Maximum number of patients i.e. 17 (37.7 %) were from the age group 11 - 20 years. Male to female ratio was 2.75 : 1. Congenital ptosis was found to be the most common type seen in 36 (80 %) patients. Simple congenital ptosis (34 patients) being the commonest among congenital type. 4 (17.7 %) patients had aponeurotic ptosis. 3 (6.6 %) patients had neurogenic ptosis. Myogenic ptosis was seen in 2 (4.4 %) patients. Majority of our cases had severe degree of ptosis (57.7 %) and poor LPS action (46.6 %). LPS resection was done in 21 (46.6 %) patients, LPS reinsertion in 1 (2.2 %), sling surgery in 18 (40 %) and Fasanella Servat operation in 1 (2.2 %) patient. 2 (4.4 %) patients were medically managed and 2 (4.4 %) were kept under observation. The outcome of surgery was good in 28 (62.2 %) patients, fair in 7 (15.5 %) and poor in 6 (13.3 %) patients. Presence of amblyopia was noted in 21 patients i.e. 46.7 % of the total patients. Marcus Gunn Jaw winking phenomenon was elicited in 2 (4.4 %) patients. Bell's phenomenon was found in 4 (8.8 %) patients. 5 (11.1 %) patients had exotropia, 3 (6.6 %) had hypotropia and 1 (2.2 %) patient had complete ophthalmoplegia.

CONCLUSIONS

The study highlights the demographics, clinical profile and management of congenital blepharoptosis patients seen at a centre of excellence in North India. The study is helpful for ophthalmologists in guiding diagnosis, evaluation and management of such patients and hence, long-term successful outcome of this disease.

KEYWORDS

Blepharoptosis, Levator Resection, Frontalis Sling Surgery

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BACKGROUND

Blepharoptosis is defined as drooping of the upper eyelid below its normal position. It is a relatively common form of eyelid malposition in both children and adults. Along with being a cosmetic blemish, ptosis is associated with high incidence of amblyopia, especially in young children with severe ptosis. Based on age of onset, ptosis can be classified as congenital and acquired. Based on etiology, it can be classified as aponeurotic, myogenic, neurogenic, and mechanical.[1,2] Various studies have reported and described the clinical characteristics and management of ptosis. Most patients with ptosis require surgery. Ptosis in young children is more challenging than adult ptosis because of extra considerations like difficulty of examination, associated amblyopia, counselling of worried parents, deciding the age at which surgery should be performed and the unpredictable post-op results. Selection of the most effective procedure to correct ptosis depends upon the degree of ptosis and the amount of levator function action. Therefore, accurate preoperative work - up becomes essential. But even then it remains difficult to achieve satisfactory results in patients with severe blepharoptosis. Studying the clinico - etiological profile and classifying the type of ptosis helps in management of each case individually for a good outcome. This study reports the prevalence, etiological profile, clinical features, management and outcome of ptosis surgery in patients of ptosis diagnosed over a period of two years. This study highlights the clinical spectrum of this disorder in a tertiary eye hospital in northern India.

METHODS

This study is an institute-based retrospective observational study. Patients with blepharoptosis who reported at our tertiary care hospital in North India from January 2017 to December 2018, were included in the study. Demographic and clinical data of all the patients was collected from the oculoplasty clinic files and was analysed retrospectively. Photographs of some patients, that were available in our records, were also taken into account. The type of the ptosis was classified according to Beard's classification[1] and Freuh's mechanistic classification[2]. The patients were classified as having congenital or acquired ptosis based on onset of the ptosis. Laterality of eye involved, degree of ptosis and levator palpebrae superioris (LPS) muscle action was recorded. In unilateral cases, the amount of ptosis was calculated as the difference in mm between the heights of the palpebral apertures of the two eyes: mild ptosis was defined as 2 mm or less difference, moderate ptosis was defined as 2 – 4 mm difference, and severe ptosis was defined as 4 mm or more difference between the two eyes. In bilateral cases, the ptosis was assessed with reference to upper lid margin to reflex distance (MRD1). Ptosis was classified as severe if MRD1 was less than 0.5 mm, moderate if MRD 1 was between 0.5 mm to 2.5 mm and mild if MRD1 was more than 2.5 mm. Levator function was measured as the maximum lid excursion from maximal downgaze to upgaze, with frontalis function abolished. This was recorded as poor if less than 4 mm, as moderate if between 4 – 7 mm, and good if more than 8 mm. The presence of amblyopia, strabismus and Marcus Gunn jaw winking were noted. A record of Bell's phenomenon was also made. The types of surgeries performed and their outcomes were analysed as well. The outcome was classified as good, fair and poor as per Brincat et al, UK study.[3] A good outcome was defined as single operation, good cosmesis, no complications with both surgeon and patient / parents satisfied with the results. A fair outcome was defined as single operation but fair cosmesis, +/- complications, either surgeon or patient / parents dissatisfied with the result. A poor outcome was defined as more than one operation with poor cosmesis, complications occurred, both patient / parents and surgeon dissatisfied with the results. The data was then subjected to descriptive statistical tabulation and analysis. Data was presented as frequencies and mean ± Standard Deviation (SD).

RESULTS

45 patients of blepharoptosis were seen at our Regional Institute of Ophthalmology in North India from January 2017 to December 2018. The mean age of the study sample was 17.4 years ranging from 2 years to 70 years. Maximum number of patients i.e. 17 (37.7 %) were from the age group 11 - 20 years, followed by 16 (35.5 %) patients in age group 0 - 10 years, 6 (13.3 %) patients in 21 - 30 years, 4 (8.8 %) patients in 31 - 40 years, 2 (4.4 %) in 61 - 70 years and none in 41 - 60 years (Figure 1). Out of 45 patients, 33 (73.3 %) patients were males and 12 (26.6 %) were females. The male to female ratio was 2.75 : 1. Unilateral eye was involved in 40 (88.8 %) patients (right eye in 17, left eye in 23 patients) while bilateral eyes were involved in 5 (11.1 %) patients. While analysing best corrected visual acuity, amblyopia was seen in 21 (46.6 %) patients.

Congenital ptosis was seen in 36 (82.2 %) patients, which were further classified as simple congenital ptosis (32 patients), Marcus Gunn synkinetic ptosis (2 patients), Blepharophimosis syndrome (1 patient) and congenital third nerve palsy(1 patient). 2 (4.4 %) patients had involutional aponeurotic ptosis and 2 (4.4 %) patients presented with aponeurotic dehiscence after trauma. 3 (6.6 %) patients had neurogenic ptosis. Out of these 3, 1 patient had traumatic third nerve palsy, 1 had blow-out fracture of orbit and 1 had mucormycosis in cavernous sinus leading to multiple cranial nerve palsies. Myogenic ptosis was seen in 2 (4.4 %) patients. 1 patient had cysticercosis in superior rectus and the other had Chronic Progressive External Ophthalmoplegia (CPEO).

On evaluating the amount of ptosis, mild ptosis was seen in 1 (2.2 %) patients, moderate in 18 (40 %) and severe in 26 (57.7 %) patients. LPS action was recorded to be good in 13 (28.8 %) patients and fair in 11 (24.4 %) patients. 21 (46.6 %) patients had poor LPS action. Marcus Gunn Jaw winking phenomenon was elicited in 2 (4.4 %) patients. Bell's phenomenon was good in 21 (46.6 %) and was found

poor in 4 (8.8 %) patients. On assessment of association of strabismus with ptosis, it was found that 5 (11.1 %) patients had exotropia, 3 (6.6 %) had hypotropia and 1 (2.2 %) patient had complete ophthalmoplegia.

Patient No.	Age	Sex	Laterality	Type of Ptosis	Severity of Ptosis	LPS Action	Surgery done	Outcome of Surgery
1	27	M	L	Congenital	Moderate	Poor	LPS resection (maximal)	Good
2	70	M	R	Aponeurotic	Moderate	Good	LPS resection	Good
3	24	M	R	Congenital	Moderate	Good	LPS resection	Good
4	66	F	L	Aponeurotic	Moderate	Good	LPS resection	Good
5	29	F	R	Post-traumatic	Moderate	Good	LPS resection	Good
6	11	F	B/L	BPS	Severe	Poor	Sling	Fair
7	9	M	L	Congenital	Severe	Fair	LPS resection (supramaximal)	Fair
8	6	M	L	Congenital (Marcus Gunn)	Severe	Poor	Sling with LPS excision	Poor
9	2	M	B/L	Congenital	Severe	Poor	Sling	Good
10	10	M	R	Congenital	Severe	Fair	LPS resection (supramaximal)	Good
11	12	M	L	Congenital (Marcus Gunn)	Severe	Poor	Sling with LPS excision	Good
12	8	F	L	Congenital	Severe	Fair	LPS resection (supramaximal)	Good
13	8	M	L	Congenital	Moderate	Good	LPS resection	Good
14	6	M	R	Congenital	Severe	Poor	Sling	Poor
15	6	M	R	Congenital	Severe	Poor	Sling	Good
16	16	M	L	Congenital	Severe	Poor	Sling	Fair
17	12	F	R	Traumatic	Severe	Fair	LPS reinsertion n resection	Good
18	16	F	R	Congenital	Moderate	Good	LPS resection	Good
19	16	F	R	Congenital	Moderate	Good	LPS resection	Good
20	9	M	R	Congenital	Severe	Poor	Sling	Poor
21	21	M	L	Congenital	Moderate	Fair	LPS resection	Good
22	9	M	L	Congenital	Severe	Poor	Sling	Good
23	11	F	L	Congenital	Severe	Fair	LPS resection (supramaximal)	Fair
24	4	M	L	Congenital	Severe	Poor	Sling	Good
25	29	F	L	Congenital	Moderate	Good	LPS resection	Good
26	11	M	R	Congenital	Moderate	Fair	LPS resection	Good
27	20	M	R	Congenital	Severe	Fair	LPS resection (supramaximal)	Fair
28	6	M	L	Congenital	Severe	Poor	Sling	Fair
29	5	M	L	Congenital	Severe	Poor	Sling	Good
30	6	F	L	Congenital	Severe	Poor	Sling	Good
31	21	M	L	Congenital	Severe	Poor	Sling	Good
32	13	M	L	Congenital	Moderate	Good	LPS resection	Poor
33	18	M	R	Congenital	Moderate	Fair	LPS resection	Good
34	12	M	B/L	Congenital	Severe	Poor	Sling B/E	Fair
35	16	M	R	Congenital	Mild	Good	Fasanella Servat	Good
36	13	M	L	Congenital	Severe	Poor	Sling	Poor
37	6	M	B/L	Congenital	Severe	Poor	Sling	Good
38	31	F	B/L	CPEO	Moderate	Poor	Sling B/E	Poor
39	14	M	L	Congenital	Moderate	Good	LPS resection	Good
40	38	M	R	Congenital	Moderate	Good	LPS resection	Good
41	6	F	L	Cong 3rd n palsy	Moderate	Fair	LPS resection	Good

Table 1. Various Types of Surgeries Done with Regard to Degree and Type of Ptosis and Success Outcome

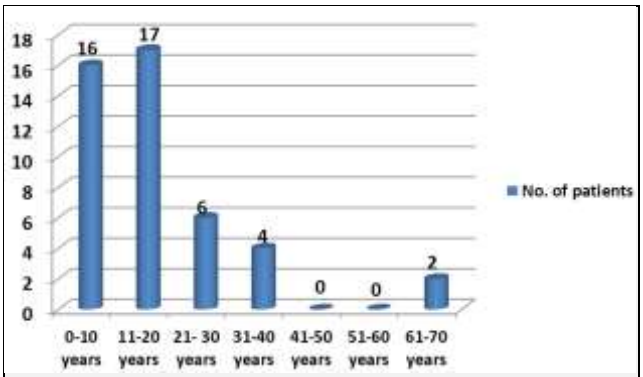

Figure 1. Age-Wise Distribution of Ptosis Patients

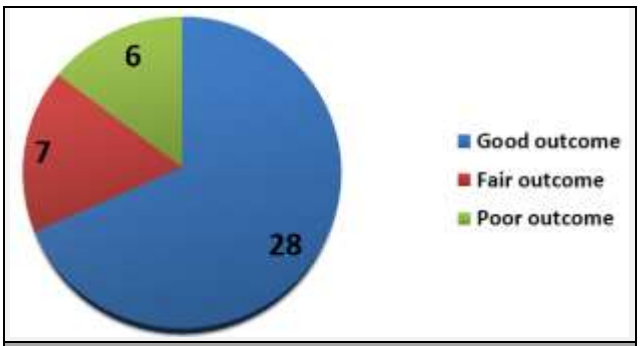

Figure 2. Outcome of Surgical Procedures

Figure 3. Photograph of a Patient Who Underwent LPS Resection (Pre-Op and Post-Op)

Figure 4. Photograph of a Patient before and after Sling Surgery

% of Different Types of Ptosis	Current Study	Berry et al[3]	Griepentrog et al[4]	Lee et al[5]	Prabha et al[6]	Agrawal et al[7]
Location	India	United Kingdom	United States of America	South Korea	India	India
No. of patients	45	155	107	2328	123	35
Congenital dystrophic	71.1	74	75.7	73.7	72.35	65.71
Blepharophimosis syndrome	2.2	4.5	2.8	2.5	11.38	2.86
Marcus Gunn synkinetic ptosis	4.4	5	3.8	1.3	4.87	17.14
Congenital third nerve palsy	2.2	7.7	2.8	0.04	2.43	5.71
Double elevator palsy	-	-	-	0.4	8.94	-
Childhood myasthenia gravis	-	0.6	1.9	-	-	8.57
Congenital Horner's syndrome	-	0.6	1.9	-	-	-
Mechanical ptosis	-	5	-	1.4	-	-
Traumatic ptosis	8.8	0.6	1.9	1.6	-	-
Involutional aponeurotic ptosis	4.4	-	-	11.5	-	-

Table 2. Comparison between Types of Ptosis in Different Studies

After the clinical data analysis, the management profile of patients was compiled. It was found that, out of 45, 41

(91.1 %) patients were surgically managed (Table 1). In 22 (48.8 %) patients, levator muscle surgery was done. Out of these 22, LPS resection was performed in 15 (33.3 %) patients, maximal LPS resection in 1 (2.2 %) and supramaximal LPS in 5 (11.1 %) patients. One patient who had traumatic aponeurotic dehiscence underwent LPS reinsertion. 18 (40 %) patients underwent frontalis suspension (sling) surgery. Fasanella Servat operation was done in 1 (2.2 %) patient. 2 (4.4 %) patients were kept under observation and 2 (4.4 %) were medically managed. The outcome of surgery was good in 28 (62.2 %) patients, fair in 7 (15.5 %) and poor in 6 (13.3 %) patients (Figure 2,3,4). Out of 41 operated patients, 12 (26.6 %) patients had complications after surgery. Out of these 12 patients, 8 patients had been operated by brow suspension using

Silicone sling. The complications included lagophthalmos, under-correction, over-correction, knot / suture granuloma, sling exposure / extrusion, asymmetry in both the lids after bilateral sling surgery, notching of upper lid margin, exaggerated lid fold, post op entropion and corneal exposure. 6 (13.3 %) patients had to undergo reoperations.

DISCUSSION

In our study, the clinico-aetiological profile, the demographic profile, other associated features, management of ptosis along with the outcome of surgeries performed, were analysed. The results of our study were also compared with the previous large-scale studies. In our study, congenital ptosis was found to be the most common type seen in 36 (80 %) patients. Simple congenital ptosis (34 patients) being the commonest among congenital type. A study done by Brincat et al (2009) reported the relative proportion of various forms of ptosis in a large 10 - year review of 155 children in the United Kingdom (UK) who presented for corrective surgery.[3] The most common form of ptosis noted was myogenic developmental abnormality (74 % patients), which in the current study is termed simple congenital ptosis. Another study done in USA by Griepentrog et al (2011) is the first population - based report on the incidence of childhood ptosis. A total of 107 children (< 19 years) were diagnosed with ptosis during the 40 - year period, yielding an incidence of 7.9 / 100,000. 96 (89.7 %) were congenital in onset, 81 (75 %) of which had simple congenital ptosis.[4] A similar retrospective study was done by Lee et al (2018) in a tertiary referral hospital in South Korea. Of the 2,328 patients, 1,815 (78 %) had congenital ptosis and 513 (22 %) had acquired ptosis. Simple congenital ptosis was the most common type overall (73.7 %), Simple congenital ptosis without any concurrent anomalies comprised 94.5 % of the cases of congenital ptosis. Among the cases of acquired ptosis, aponeurotic (52.1 %) and myogenic (25.5 %) were the most common.[5] Some Indian studies were also reviewed to compare our data with similar Indian population. Prabha et al (2018) reviewed 123 cases of congenital ptosis presenting to their institute in South India. Simple congenital ptosis was the commonest type noticed in their study accounting for 72.35 % of the total (89 patients). Blepharophimosis syndrome was found in 11.38 % patients and congenital 3rd nerve palsy in 2.43 % patients.[6] In a prospective study done by Agarwal (2019) in Western India, 35 blepharoptosis patients were enrolled. Myogenic ptosis due to a dystrophic levator muscle included 23 cases (65.71 %). Marcus Gunn jaw winking synkinesis was seen in six (17.14 %) cases. Congenital myasthenia was documented in three (8.51 %) cases. Third nerve palsy was seen in two (5.71 %) cases and blepharophimosis syndrome in one case (2.86 %).[7]All these studies are in consistence with our study. Table 1 shows the comparison between types of ptosis in these different studies.

We have enrolled pediatric as well as adult ptosis patients in our study. So, the mean age of presentation of the study group was 17.4 years. Lee et al also reported the mean age of presentation of their study population as 17.26 years.[5] Our study reports a male predominance. The male to female ratio in our study is 2.75:1. Lee et al also reported a male predominance, male to female ratio as 1.34:1.[5]

In the present study, majority of the patients (88.8%) had unilateral ptosis. Previous studies have also reported high percentage of unilateral ptosis. UK based study reported bilateral involvement in 20% of patients and left lid involvement in 45.81% of patients.[3] In a study by Griepentrog et al, three (4%) of the simple congenital ptosis cases were bilateral, and 55 (68%) of the unilateral cases involved the left upper eye lid.[4] Lee et al reported 82 % cases as unilateral ptosis.[5] In the study by Prabha et al, 71.54 % cases were unilateral and involvement of left eye was predominant at a rate of 60.22 %.[6] With respect to laterality of eye, our finding is in consistence with all these studies as in our study also left eye was involved in 57.5 % of unilateral cases. Predominance of left eye in unilateral cases as seen in most of the studies is not a very well understood phenomenon. Further studies would be required to know the exact cause of this predominance.

Most of our cases had severe degree of ptosis (57.7 %) and poor LPS action (46.6 %). Similar trends were seen in other studies also.[6-8] In the study by Prabha et al, severe ptosis was present in 90.24% and function of Levator palpebrae superioris was poor in 73.17 %.[6] Agrawal reported 32 patients (91.43 %) out of 35 patients as severe blepharoptosis with poor levator function.[7] Essawyet al reported severe ptosis in 260 cases (63.7 %) and poor levator function in 293 cases (71.8 %).[8] This could be due to the fact that severe degree of ptosis is cosmetically unacceptable for the patients and their parents, so in such cases there is a tendency to report early to ophthalmologist.

Amblyopia was reported in 46.6% of our cases. In other two Indian studies,[6,7] the incidences, of amblyopia were 25.71 % and 38.21 %, while in a study by Essawy et al in Egypt,[8] amblyopia was seen in just 10% of ptosis cases. The higher incidence in our study could be because of inclusion of all cases of amblyopia like anisometropic, strabismic and stimulus deprivation in our study. Marcus Gunn jaw winking was seen in 4.4% cases. Prabha et al also reported 5 % incidence of this phenomenon in their study.[6]

The surgery performed in most of our cases was levator resection (48.8 % cases). This is in sharp contrast to other

studies mentioned earlier where the most common surgery performed was frontalis suspension using fascia lata or silicone sling.[3-8] This could be due to the fact that though most of our cases had severe ptosis, we tried managing some of those cases by maximal or supramaximal levator resection which was lacking in other studies. These patients were however found to have fair results.

In our study, 13.3 % patients had poor outcome after surgery which was defined as poor cosmesis, occurrence of complications and reoperations. Out of the total 45 patients, complications were seen in 26.6 % patients after surgery; lagophthalmos and under-correction being the commonest one. One patient with associated superior rectus palsy, with residual hypotropia even after squint surgery, was under-corrected intentionally so as to prevent exposure keratitis due to poor Bell's phenomenon. Agrawal also reported nocturnal lagophthalmos and under-corrections as most common complications in the cohort.[7] Although our study reported a greater number of levator resections, but poor outcome was seen more commonly in sling surgery cases. The results of frontalis suspension were less satisfactory as it was associated with complications like nocturnal lagophthalmos, lid notching, suture or knot granuloma and exposure/ extrusion of sling in some of the patients. Although sling surgery is contraindicated in CPEO patients, our patient with CPEO underwent sling surgery due to immense insistence by the patient.[9] In this surgery, though we intentionally under-corrected the ptosis but still the patient had post op severe exposure keratitis and the surgery had to be undone. In our study, 13.3 % patients had to undergo reoperation mostly because of exposure of silicone sling and under corrected ptosis. Similarly, Essawy et al[8] reported 16.9 % reoperation cases and Lee et al[10] reported 13 % cases and who underwent reoperations.

A thorough understanding of clinico-aetiological factors and a proper and timely management of all ptosis patients helps the ophthalmologist in a long-term successful outcome of this disease.

CONCLUSIONS

Ptosis, if not corrected may lead to amblyopia or abnormal head posture, besides having cosmetic blemish. It can also have psychosocial impact on the development of the child. Surgical treatment is the mainstay for ptosis correction. Surgical correction of ptosis gives excellent functional and cosmetic results. Patients and parents of young children should be educated and motivated for early correction of ptosis.

Data sharing statement provided by the authors is available with the full text of this article at jebmh.com.
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