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Amblyopia and Strabismus in Congenital Ptosis

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Summary

Seventeen per cent of 216 cases of simple congenital ptosis developed amblyopia and 19% had a squint. Of those patients with amblyopia, 14% had amblyopia attributable to stimulus deprivation, 21% had anisometropic amblyopia and 51% had strabismic amblyopia. Early refraction, orthoptic assessment and treatment and, where the pupillary axis is occluded , surgery to prevent stimulus deprivation amblyopia are recommended.

It has been stated that severe ptosis probably does not interfere with the development of visual function.[1] This view has been supported by Beneich[2] and Yasuma[3] but challenged by Anderson[4] and Merriam.[5] If the assertion is true there is no indication to correct a ptosis in a young child in order to prevent amblyopia and all ptosis surgery may be delayed until the optimum time for cosmetic correction. Our practice has been to perform this at about 4 years of age when the child is sufficiently co-operative for a reasonably accurate assessment to be obtained, but has not yet started proper schooling. The child has by then grown sufficiently for autogenous fascia lata to be obtainable where necessary. However if the ptotic eyelid of a young child was thought to endanger the pupillary axis the ptosis was corrected with a brow suspension procedure using non-autogenous material. If a severe ptosis never interferes with the development of visual function this practice cannot be justified. We have therefore examined the records of 424 consecutive patients presenting with ptosis to assess the incidence and causes of squint and amblyopia.

Patients and Methods

The case notes of 424 consecutive patients with ptosis who presented to the oculoplastic service at Moorfields Eye Hospital over the past 5 years were reviewed. The ptosis had been present since birth in 300 patients and was acquired in the remaining 124. The causes and associations of the congenital and acquired groups are shown in Tables I and II. Ptosis following surgery for conditions such as dysthyroid eye disease or orbital disease was included under the heading of surgical trauma.

There were 216 cases of congenital ptosis which were not associated with any specific syndrome and in whom the cause was thought to be either a dystrophy of the levator muscle, an aponeurotic weakness or a combination of both. In patients with a dystrophy of the levator muscle there was lid-lag on downgaze whereas the ptosis was constant if there was an aponeurotic weakness.

Results

Further attention was directed at the 216 cases of simple congenital ptosis. (Table III). The ptosis was unilateral in 172 (80%) and bilateral in 44 (20%). In 29 cases (13%), a first degree relative was affected. Thirty seven patients (17%) had amblyopia at some stage. This we defined as best corrected visual acuity of 6/12 or less and greater than two Snellen lines of difference between the

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Table I *Causes of congenital ptosis.*

Simple congenital	216
Marcus Gunn	31
Blepharophimosis	35
Congenital 3rd or aberrant innervation	6
Cyclic 3rd	2
Neurofibromatosis	3
Others	7*
Total	300

Others: Ocular fibrosis, Duane's, craniofacial dysostosis, coloboma plus dermoid, Horner's syndrome, Beckwith's Syndrome and dystichiasis with epiblepharon.

Table II *Causes of acquired ptosis.*

Trauma	25
Surgical trauma	11
Disinsertion	36
Myopathy	26
Dysthyroid eye disease	7
Neurological disease	7
Senile	4
Blepharochalasis	3
Miscellaneous	5*
Total	124

Miscellaneous: haemangioma (1), post-inflammatory (1), unknown aetiology (2), microphthalmos (1).

Table III *Characteristics of simple congenital ptosis*

Unilateral	172	80%
Bilateral	44	20%
Positive family history	29	13%
Amblyopia	37	17%
Squint	42	19%
Superior rectus weakness (Unilateral)	17	7.9%
Superior rectus weakness (Bilateral)	1	0.04%

Table IV *Aetiology of amblyopia in simple congenital ptosis*

Squint	20	54%
Astigmatism	6	16%
Stimulus deprivation	5	14%
Anisometropia	2	5%
Multifactorial	4	10%
Total	37	

two eyes in the absence of eye disease, or in the very young, two successive orthoptic reports detailing inability to detect 100s and 1000s, objection to occlusion of the better eye and in those with strabismus definite fixation preference. There were no cases of bilateral amblyopia. The aetiology of the amblyopia was sought in each case and was found to be attributable to squint in 20 (54%) of cases (Table IV). In 6 patients (16%) amblyopia was associated with astigmatism of more than 3.50 dioptre cylinders (Table V) and in 4 of these cases there was associated anisometropia. In two further cases (5%), there was anisometropia of greater than 5 dioptres. In 4 patients the exact cause was difficult to ascertain and a combination of factors was thought to be responsible.

In 5 patients (14%) there was no significant refractive error and no squint but the lid level was recorded as endangering the pupillary axis. These patients were thought to have stimulus deprivation amblyopia as a direct result of the ptosis and eyelid level. Three of these patients underwent unilateral brow suspension using stored fascia lata as an urgent procedure and one patient was aged 4 on presentation and proceeded immediately to bilateral autogenous fascia lata brow suspension. The fifth patient was noted to

Table V *Amblyopia due to high astigmatism and/or anisometropia*

Case	*VA*	*Refraction*	
		R	*L*
Case 1. J.L.	6/18	−0.50	−4.50
		+0.50 × 70	+ 3.50 × 65
Case 2. S.H.	6/18	−0.25	−5.00
			+5.50 × 85
Case 3. R.W.	6/18	−1.50	00.00
		+5.00 × 105	
Case 4. V.N.	CF	00.00	−4.25
			+ 5.25 × 90
Case 5. S.D.	6/18	−0.25	−4.00
			+ 6.00 × 70
Case 6. R.S.	6/12	+0.25	−1.25
		+0.25	+ 4.00 × 95
Case 7. N.S.	6/12	−5.00	00.00
Case 8. R.H.	6/24	−7.00	−1.25

have pupillary occlusion at the age of 3 but the parents refused to allow surgery. The child was treated orthoptically but nevertheless developed amblyopia and subsequently underwent anterior levator resection as an emergency procedure.

Forty two patients had a horizontal or vertical strabismus of which 20 (48%) developed amblyopia. The incidence of squint was identical in both unilateral and bilateral ptosis groups. There were 26 esotropes, one accomodative esotrope, 12 exotropes and 3 hypotropias; seven cases with a horizontal strabismus had an associated vertical deviation. Six cases underwent correction of their hypotropia and pseudoptosis by the Knapp procedure.

Discussion

The incidence of amblyopia in the general population is approximately 3%.[6,7] Our incidence of 17% in simple congenital ptosis is comparable to that of Merriam et al,[5], 14% and Anderson,[4] 20%. Only broad comparisons can be made between our work and that of other authors since they have included all forms of congenital ptosis in their calculations. In our study, 5 cases of amblyopia (2.3%) were directly attributable to stimulus deprivation; Anderson found 1.6%, Merriam 1.5% and Stark 4%.[8] Our findings support our policy of performing a brow suspension as an emergency procedure in those cases where the pupillary axis is threatened. A high incidence of astigmatism in congenital ptosis has been reported,[3,5,8] and in our series amblyopia was associated with astigmatism of more than 3.50 dioptres. We therefore recommend that all children with ptosis should have a refraction and that any astigmatism be corrected appropriately.

Forty two (19.4%) of our cases had a squint. The incidence in other studies ranged between 12% and 32%.[2,3,8,9,10] In ten of our cases there was a vertical deviation. It is our policy to perform a Knapp procedure[11] where there is significant hypotropia and pseudoptosis prior to the correction of any residual ptosis. At the time of surgery a forced duction test is performed and if there is limitation, recession of the inferior rectus is performed as well as the Knapp procedure.[12,13]

This study has demonstrated a high incidence of amblyopia in congenital ptosis. This should be preventable by a combination of early refraction, orthoptic assessment and treatment and surgery for those cases where stimulus-deprivation amblyopia is thought to be a risk. Eight cases developed amblyopia as a result of high astigmatism or spherical anisometropia; this emphasises the importance of early refraction in children with congenital ptosis.

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