Elevating the hypotropic globe

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SUMMARY Twelve consecutive cases are reviewed of unilateral hypotropia due to various causes. In most of them the presenting complaint was of ipsilateral blepharoptosis. Some patients had undergone previous ineffective surgery. The surgical techniques employed and the results obtained are discussed, with emphasis on the necessity to test for inferior rectus restriction. Some interesting sensory results are also noted.

The combination of unilateral congenital ptosis with hypotropia of the ipsilateral eye is common.¹ Hypotropia may also accompany the Marcus Gunn jaw-winking syndrome and buphthalmos secondary to neurofibromatosis. Third nerve palsy may lead to hypotropia even when there has been some recovery. In all these disorders it is necessary to defer surgery to improve the ptosis until the best vertical alignment of the visual axes has been obtained. The results of surgery on 12 hypotropic eyes is the subject of this study.

Patients and methods

All patients attended Moorfields Eye Hospital, London, between October 1979 and November 1982. The majority (9/12) had been referred to one of us (JROC) for consideration of surgery for ptosis. The age range was from 5 to 23 years, mean 12.5 years. There were six females and six males (Table 1). Seven patients had congenital ptosis with hypotropia of the ipsilateral globe. Two patients had buphthalmos with ptosis due to neurofibromatosis. Two patients had third nerve palsy with misdirection, one congenital, the other due to a motor cycle accident. One patient had Marcus Gunn jaw-winking syndrome.

VISUAL ACUITY

Eight patients had deficient visual acuity in the affected eye. In five cases this was thought to be due to amblyopia, in two cases complicated by high myopia. The two eyes affected by neurofibromatosis had the very poor acuities of 6/60 and light percep-

tion. The patient with the traumatic third nerve palsy also suffered extensive damage to the anterior visual pathways with extensive field loss, worse in the paretic eye, and evidence of chiasmal damage.

PREOPERATIVE DEVIATION

All cases showed hypotropia in the primary position, increasing on upgaze. The size of the deviation ranged from 9 prism dioptres to 70 prism dioptres, with a mean of 30.5 prism dioptres. All cases showed deficient elevation of the affected eye. Eight cases had an associated horizontal deviation of greater than 10 prism dioptres. Four had an esodeviation, four had an exodeviation. The two cases of third nerve palsy showed signs of misdirection with elevation of the lid on adduction and especially on depression in adduction. Seven cases showed pseudoptosis with elevation of the lid to near-normal height when the hypotropic eye was forced to take up fixation.

BINOCULAR VISION

Two cases could fuse targets in the lower field of gaze, with a symmetrical cross on Bagolini striated glass testing and stereoacuities of 60 and 140 seconds of arc respectively on the Titmus stereotest. Both suppressed the hypotropic eye on upgaze.

PREVIOUS SURGERY

Four cases (nos. 1, 3, 5, and 8) had undergone previous extraocular muscle surgery. Of these one had had superior rectus resection only, two had had combined superior rectus resection and inferior rectus recession, and one (case 8) with congenital third-nerve palsy had a history of three previous procedures on the horizontal recti of both eyes.

Six cases (nos. 1, 3, 4, 5, 9, and 10) had undergone

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Fig. 1 A: *Knapp procedure.* Horizontal rectus muscles are elevated and sutured to the globe in order to straddle the superior rectus insertion. B: *Modified Knapp procedure.* The technique is similar to that in the standard Knapp procedure, but the suture for the recessed muscle is brought through long scleral bites and tied adjacent to the superior rectus insertion.

previous surgery for ptosis, usually some form of levator resection. Two cases had also had frontalis suspension procedures. In three cases the amount of levator resection was known, the figures being 10, 12, and 13 mm.

SURGERY

In 10 of the cases the operation was performed by one of us (JPL). Eleven cases were treated with Knapp² procedures or a modification of it. The horizontal recti were transposed to the superior rectus insertion so as to straddle it (Fig. 1A). In eight cases a significant horizontal deviation was present. Six cases had a modification of the standard procedure with appropriate amounts of horizontal rectus recession and resection. Where a muscle was recessed, the points of reattachment were measured from the superior rectus insertion. Initially, two separate scleral sutures were inserted. More recently, a 'hang back' technique (Fig. 1B) has been utilised.

Eleven cases had preoperative forced duction testing of the affected eye under general anaesthesia before the start of surgery. In eight cases (66%) this was judged to show resistance to passive elevation of the globe. Seven cases were treated with inferior rectus recession in addition to the basic elevation procedure (in one case at a subsequent date). The eighth case had the area of the inferior rectus explored. The muscle was found to be surrounded by a condensation of fibrous tissue. When the muscle was dissected free, the traction test improved markedly, and the muscle was therefore not recessed. A similar case was reported by McNeer and Jampolsky.³

One case underwent the procedure described by Callahan,⁴ where the horizontal recti and superior rectus are split without disinsertion and are joined in a similar manner to the Jensen⁵ procedure for lateral rectus palsy (Fig. 2). This patient, a 12-year-old boy with von Recklinghausen's neurofibromatosis, had a

buphthalmic right eye in conjunction with a right ptosis partly due to a plexiform lid neuroma. The visual acuity of the eye was perception of light. There was extensive scleral thinning with staphyloma formation, especially superiorly (Fig. 3). He had a right Callahan procedure in April 1981. This procedure was preferred because of the technical difficulty of resuturing rectus muscles to the very thin sclera.

Results

In all cases the hypotropia was reduced as a result of surgery. The data are given in Table 1. The reduction in vertical deviation ranged from 7 to 40 prism dioptres. In those cases where additional recession-resection had been performed the horizontal devia-

Fig. 2 *Callahan procedure.* The horizontal recti are split, as is the superior rectus, and the muscle strips joined with non-absorbable sutures. In addition the inferior rectus is recessed.

Table 1 Results

Case	Age Sex	Corrected VA R/L	Aetiology	Previous squint surgery	Previous ptosis surgery	Preop. deviation (prism dioptres)	Binocular vision preop.
1	13 F	6/9 6/9	Congenital ptosis with hypotropia	RSR+	Yes—nature unknown	R hypo 11 R eso 6	Fusion on downgaze
2	13 F	6/9 6/60	Neurofibromatosis	No	No	L hypo 60 L eso 40	No
3	12 F	6/6 6/6	Congenital ptosis with hypotropia	LIR— LSR+	L levator resection 1979	L hypo 25 L eso 4	Fusion on downgaze
4	13 F	6/36 6/9	Congenital ptosis with hypotropia	No	R levator resection 1972, R frontalis sling 1975	R hypo 29 R eso 10	No
5	9 M	6/24 6/5	Congenital ptosis with hypotropia	RIR— RSR+ 1979	R levator resection 1979	R hypo 9 R eso 16	No
6	20 M	6/60 6/5	R traumatic IIIIn palsy. Misdirection. R optic atrophy and chiasmal damage	No	No	R hypo 11 R exo 50	No
7	12 M	PL 6/5	Neurofibromatosis	No	No	Gross R hypo Mod. R eso	No
8	7 M	6/9 6/6	Congenital R IIIIn palsy + misdirection	1975 { 1977 { 1979 { RLR— RMR+ RMR— RIR+ LMR— LLR+	No	R hypo 11 R exo 40 Variable	No
9	14 F	6/5 6/18	Congenital ptosis with hypotropia, L myopia	No	L ptosis surgery 1969—L levator resection	L hypo 30 L exo 6	No
10	15 F	CF 6/5	Marcus Gunn jaw-winking syndrome, R myopia	No	1971—levator resection, 1979—R frontalis sling	R hypo 20 R exo 36	No
11	23 M	6/6 6/6	Congenital L ptosis with hypotropia R myopia	No	No	L hypo 65–70 L exo 24	No
12	5 M	6/6 6/24	Congenital ptosis with hypotropia	No	No	L hypo 40 Mod L eso	No

SR=superior rectus. IR=inferior rectus. MR=medial rectus. LR=lateral rectus. ARC=anomalous retinal correspondence.
Eso=convergent squint. Exo=divergent squint. Hypo=hypotropia. —=Recession. +=Resection.

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Pseudoptosis	Surgery	Traction test	Complications	Postop. binocular vision	Postop. deviation (prism dioptres)	Correction (prism dioptres)
Yes	24 Oct 79 R Knapp procedure	Not done	R lower lid retraction	Improved field of BSV	R exo 16	11 R hypo
No	29 Nov 79 L Knapp procedure LLR+6 mm LMR-5.5 mm LIR-5 mm	Pos.	No	No	Improved	
Yes	L Knapp procedure 10 Mar 80	Neg.	No	Improved field of BSV	No hypo eso 2	25 L hypo
Yes	12 May 80 R Knapp procedure + IR freeing	Pos.	No	No	R hypo 3 R eso 6	26 R hypo
Yes	14 Jul 80 R Knapp procedure	Neg.	No	No	R hypo 2	7 R hypo
No	12 Feb 81 R Knapp + RLR-10 mm RMR+7 mm 2 Apr 81 RIR-4 mm	Pos.	No	No	Straight in primary position	11 R hypo 50 R exo
Minimal	2 Apr 81 R Callahan procedure + RIR-4 mm	Pos.	No	No	Less R hypo More R eso	
Yes	16 Jun 81 R Knapp procedure RLR-4 mm RMR+5 mm RIR-3 mm	Pos.	Mild Periorbital cellulitis	No	No vertical R exo 20	11 R hypo 20 R exo
Yes	17 Sep 81 L Knapp procedure	Neg.	No	Vertical ARC (see text)	L hypo 4 + latent component	26 L hypo
No	17 Dec 81 R Knapp procedure RLR-3 mm RMR+3 mm RIR-3 mm	Pos.	No	No	R hypo 12 R exo less	8 R hypo
Minimal	15 Jul 82 L Knapp procedure LLR-5 mm LMR+3 mm LIR-5 mm	Pos.	No	Present after R surgery (see text)	L hypo 30 L exo 4	40 L hypo 20 L exo
Yes	18 Nov 82 L Knapp procedure LMR-5 mm LIR-5 mm	Pos.	No	No	No vertical Small L eso	40 L hypo

Fig. 3 Case 7 preoperatively.

Fig. 4 Case 7 after right Callahan's procedure.

tion was reduced. There was no significant difference between the size of the correction when the groups with and without inferior rectus recession were compared, the average corrections achieved being 23 and 19 respectively. In general the improvement was in reduction of the vertical deviation in the primary position, with persistently poor elevation and an increase in the deviation on upgaze. All patients or their parents reported cosmetic improvement. There were no overcorrections.

Case 7, the only case treated with a Callahan procedure, showed a reduction of hypotropia (Fig. 4) but an increase of esotropia to greater than 45 prism dioptries.

COMPLICATIONS

These were minor. Case 1 developed lower lid retraction, later treated with a scleral graft to the lower tarsus. Case 8 developed mild periorbital cellulitis which swiftly resolved on antibiotic therapy. There were no cases of postoperative uveitis or anterior segment ischaemia.

POSTOPERATIVE BINOCULAR VISION

The two patients with demonstrable fusion in down-gaze preoperatively had an extension upwards of their binocular area while still suppressing on upgaze.

Two other patients showed binocular vision post-operatively that had not been detectable pre-operatively.

Case 9. This 14-year-old white schoolgirl had congenital left ptosis (Fig. 5). The acuity of the right eye was 6/5, with a refraction of -0.25 dioptre. The left eye's corrected acuity was 6/18, with a refraction of $+0.50$ DS/ -5.00 DC $\times 175^\circ$. There was a left ptosis with 6 mm of levator function. There were 30 dioptres of left hypotropia and 3° of left exotropia. Orthoptic testing preoperatively showed 'left suppression.' A Knapp procedure performed on 17 September 1981 improved the hypotropia (Fig. 6). She was then found to have a vertical form of anomalous correspondence, with anomalous fusion at left hypo 4 prism dioptries, with an angle on dissociation of 22–28 prism dioptries.

Case 11. This 23-year-old Asian male student had a congenital left ptosis. The refraction of the right eye was -5.00 DS, giving an acuity of 6/6. The left refraction was -0.50 DS, giving an acuity of 6/6. With spectacles he fixed with the right eye and had a left hypotropia of 65–75 prism dioptries, and a left exotropia of 12° . Without spectacles he fixed with the left eye, adopting a chin-up head posture, and showed a marked right hypertropia. There was a left ptosis of 5 mm with about 5 mm of levator function.

Fig. 5 Case 9 preoperatively.

Fig. 6 Case 9 after left Knapp procedure.

On preoperative testing no evidence of binocular function could be found. A left Knapp procedure with inferior rectus recession and horizontal rectus recession/resection was performed on 15 July 1982, with reduction of the deviation to 30 prism dioptres of left hypotropia. On 30 September 1982 he underwent a right superior rectus recession of 5 mm, with posterior fixation sutures at 14 mm from the insertion. Remarkably, after this he showed fusion, with a 15° range of motor fusion on the synoptophore, a symmetrical cross on the Bagolini striated glass test, and a stereoacuity of 80 seconds of arc on the Titmus stereotest.

Discussion

The technique of elevation of the horizontal rectus insertions to relieve hypotropia is usually termed the Knapp procedure, although Dunlap⁶ has pointed out that Alvaro,⁷ Watson,⁸ and Dunlap himself⁹ described the procedure before Knapp's own description² in 1969. He reported 15 patients in whom the procedure gave a correction from 21 to 55 prism dioptres of vertical deviation. Two cases also had inferior rectus recession. Six of his patients had had unsuccessful Berke-Motais procedures, with an attempt to elevate the ptotic lid with a slip from the superior rectus. He found little effect on horizontal rotations, and noted that elevation remained poor, even when a good result was obtained in the primary position. In 1981 Callahan⁴ reported three cases of failed ptosis surgery in the presence of hypotropia of the affected eye. He described a new procedure in which the superior and horizontal recti were split and united similarly to the Jensen procedure. Inferior rectus recession was also performed. The rationale was that the anterior ciliary circulation would be less affected by such surgery, as the recti were not disinserted. Our experience is confined to one patient, in whom the hypotropia was improved, but a pre-existing esotropia was worsened. We cannot therefore recommend the procedure, but it may be of value in older patients where anterior segment ischaemia is a risk with detachment of rectus muscles.

In general we find the Knapp procedure reliable, especially where there is a pre-existing horizontal deviation, as appropriate adjustments of horizontal rectus muscle forces may be incorporated. The literature on 'double elevator palsy' is somewhat confusing in that some authorities, such as Dunlap,⁶ apply the term only to cases without mechanical restriction of elevation. He reported 22 cases of double elevator underaction, 16 of whom had supraplacement of the horizontal recti, 10 having horizontal recession/resection in addition. Results were generally good, and he provided a table of average

correction in prism dioptres per millimetre of supraplacement.

Other authors have used the term to describe a group of patients with unilateral ptosis and hypotropia, and have emphasised the necessity of detecting inferior rectus restriction by a preoperative traction test. Scott and Jackson¹⁰ reported a high incidence (11/15) of inferior restriction and considered that this represented a form of secondary contracture. They described two signs which they thought were commonly associated with inferior restriction. These were deficient Bell's phenomenon and an accentuation of the lower lid fold on attempted upgaze of the affected eye. McNeer and Jampolsky³ reported a case in which an additional anomalous insertion of the inferior rectus was found at surgery. Division of this improved rotations and led to a good cosmetic result. Case 4 in our series is reminiscent of this.

Metz¹¹ studied a series of patients with limited vertical movement of one eye. Nine of 15 were hypotropic in the primary position. Of these cases five had normal saccadic velocities in an upward direction and a positive forced duction test. The remaining four had reduced upward saccadic velocity, only one having a positive forced test. He concluded that, where the hypotropia was of mechanical origin, horizontal rectus transposition was not indicated. He also observed that the cases without hypotropia in primary gaze but with defective elevation all had inferior restriction. Cases with hypotropia in the primary position had a 50% chance of having a positive forced duction test.

Recognising that our series is a more heterogeneous group than that reported by other authors we performed a Knapp procedure (or Callahan procedure) on all patients, with an inferior rectus recession if the forced duction test was positive. In one patient the inferior rectus surgery was performed three months later. The inferior bulbar conjunctiva was also recessed if it seemed to cause restriction. Two problems might have resulted from this surgical approach: anterior segment ischaemia, which we did not encounter, doubtless owing to the youth of our patients, and overcorrection of the hypotropia. None of our cases was overcorrected, though overcorrections have been reported in other authors' series. Our average correction of hypotropia was 21 prism dioptres.

Barsoum-Homsy¹² reported six cases of congenital double elevator palsy with results of Knapp procedures in four. Her average correction was 31.7 prism dioptres. She reviewed the literature, quoting Watson,⁸ who obtained an average correction of 30.5 prism dioptres, Knapp,² with an average correction of 38 prism dioptres (however, 40% of his cases were

post-Berke-Motais procedures), Cooper and Greenspan¹³ with an average of 26.6 prism dioptres, and Scott and Jackson,¹⁰ who obtained an average shift of 37 prism dioptres but overcorrected three of five patients.

All authors are in agreement that any ptosis surgery contemplated should be deferred until the best ocular alignment has been achieved. Six of our 12 patients had previous surgery that failed to elevate the lid, usually a levator resection. In addition three had had attempts to elevate the globe by surgery on the vertical recti. Not all of the previous records were available to us, but where they were they showed poor results from superior rectus resection. Presumably the superior rectus in these cases was either paretic or congenitally hypoplastic in association with levator muscle dystrophy. Furthermore, Beard¹⁴ states that superior rectus resection is liable to increase the apparent ptosis by an indirect effect on the levator.

Finally, there are the interesting and unexpected results in cases 9 and 11, which appeared to show a sudden acquisition of binocularity after the surgery was completed. In case 9 this appeared to be a vertical form of anomalous correspondence, whereas in case 11 there appeared to be bifoveal fixation and fusion. Each patient had a clear history of the ptosis and hypotropia being present from birth. Both were unilaterally myopic and had worn their correction constantly with relatively good vision in the myopic eye in case 9, while case 11 preferred to fixate with the myopic eye when wearing spectacles. It is well recognised that patients with double elevator palsy may have binocular function in downgaze with suppression elsewhere. Two other patients in this series showed this. Although neither case 9 nor 11 could be shown to be able to fuse in downgaze before surgery, it seems plausible to assume that they represented extreme forms of this adaptation. Certainly the other references to vertical anomalous correspondence that we have been able to locate¹⁵⁻¹⁷ do not appear to be comparable.

CONCLUSIONS

Hypotropia and ptosis, with or without pseudoptosis, has been treated surgically in 12 patients, 11 with Knapp procedures, one with a Callahan procedure. Improvement was obtained in all cases, the correction ranging from 7 to 40 prism dioptres with no over-corrections. It is concluded that these are safe and effective procedures in this situation and should be considered prior to considering direct surgical intervention for the ptosis.

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