Management of ipsilateral ptosis with hypotropia

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SUMMARY Thirty-one patients presented for surgical correction of unilateral hypotropia of the globe and blepharoptosis. The hypotropia and pseudoptosis were corrected by Knapp procedures. The Bell's phenomenon was thereby improved, allowing safe correction of the true ptosis, generally by an anterior levator resection whose magnitude depended on measured levator function.

The association of unilateral ptosis and hypotropia of the globe arises in a number of clinical situations, most commonly in congenital double elevator palsy. Their surgical correction was approached by Beard, who found double elevator palsy in 6% of his cases of unilateral congenital ptoses. He noted levator resection was less effective where superior rectus weakness coexisted, and recommended an additional 4 mm approximately of levator resection. He approached the correction of hypotropia by superior rectus resection and inferior rectus recession with release of the lower lid retractors. Knapp subsequently described transposition of the horizontal rectus tendons to that of the superior rectus. This improved the position of the globe and its elevation. Scott and Jackson recognised, by forced duction tests, that inferior rectus restriction plays a part in some cases and advocated recession of the muscle with release of the lower lid retractors. This was not combined in their work with the Knapp procedure as they feared anterior segment ischaemia. Callahan modified the approach (to reduce the risk of ischaemia) by routinely recessing the inferior rectus combining this with a Jensen style procedure, in which the active horizontal rectus muscles were split and half of each muscle was sutured to the weak superior rectus.

Lee, Collin, and Timms recommend that a forced duction test is done initially. If the eye moves freely, a Knapp procedure alone is done. If there is limitation, the inferior rectus muscle is recessed, and this is combined with a Knapp procedure except in those cases considered to be at risk of ischaemia, that is, those patients over 25 years of age or with a large globe, in whom they do a Callahan procedure.

Contralateral superior rectus recession with a Faden procedure is used in selected cases to increase the drive to the paretic superior rectus where these measures prove inadequate. Elevation of the hypotropic globe is considered to be the primary step, followed by correction of the true blepharoptosis, which is the subject of this paper.

Patients and methods

Thirty-one patients presented for surgical correction of unilateral hypotropia of the globe and associated blepharoptosis. The aetiology of the combined abnormalities included congenital double elevator palsy (15 patients), Marcus-Gunn jaw-winking syndrome (eight patients), third nerve palsy (three patients), neurofibromatosis with lid involvement (three patients), and superior rectus sling-induced (two patients).

The surgical approach was to improve active elevation of the globe, thereby enhancing the Bell's phenomenon and correcting the element of pseudoptosis. A Knapp procedure was adopted, elevating the insertions of the horizontal rectus muscles to the superior rectus insertion. In addition any horizontal deviation of the globe was corrected at the same time by appropriate recession or resection of the horizontal rectus muscles, as described by Alvaro. A forced duction test was performed in each case. Inferior rectus restriction, where found, was relieved by inferior rectus recession. Careful recession of the lower lid retractors was performed to preserve normal lower lid position.

The true blepharoptosis was corrected by a levator resection, which was denoted large (greater than 20 mm), medium (15–20 mm), or small. The extent of the resection required depended on the levator...
function (good greater than 10 mm, moderate 4–10 mm, or poor). The minimum delay between these procedures was four months but ranged from four months to two years. An anterior approach was preferred for the large resections, affording good exposure.

**CONGENITAL DOUBLE ELEVATOR PALSY GROUP (15 patients)**

Five patients had previously undergone both vertical muscle surgery (inferior rectus recession, superior rectus resection, or both) and ptosis surgery. Three patients had undergone vertical muscle surgery alone and three ptosis surgery alone. Hence, four of the 15 were cases not previously operated upon. Of the patients previously operated upon one required a mucous membrane graft to his scarred upper fornix and two required scleral grafting to raise adequately the lower lid following inferior rectus surgery. A Knapp procedure was indicated in all patients in this group with additional inferior rectus recession in only two. In one case inferior rectus recession was supplemented by a contralateral superior rectus recession with a Faden procedure. Horizontal rectus recession or resection was performed simultaneously with a Knapp procedure in seven cases. Two patients were mainly concerned by their ptoses and elected not to undergo strabismus surgery.

The ptoses were corrected as follows. One patient with no levator function underwent bilateral brow suspension. Eight had a large anterior levator resection, one a moderate anterior levator resection, and five small resections by either an anterior or posterior approach.

**JAW-WINKING GROUP (8 patients)**

Six patients were found to require a Knapp procedure and one a Callahan operation with inferior rectus recession. A severely retarded child, in whom ptosis correction was the only cosmetic goal, had a large posterior levator resection without extraocular muscle surgery.

Where ptosis was the major problem, an anterior levator resection was performed. Two patients had a large anterior levator resection and one a cautious levator resection rather than a brow suspension, as the Bell’s phenomenon was not adequately enhanced by the Knapp procedure. Significant jaw-winking was corrected by levator excision and bilateral brow suspension. Three patients required bilateral brow suspension and another is yet to undergo ptosis surgery. The last patient declined ptosis surgery, being satisfied after the Knapp procedure.

**THIRD NERVE PALSY GROUP (3 patients)**

The first patient had a post-traumatic third nerve palsy with misdirection regeneration. The condition had been stable for five years. A Knapp procedure with lateral rectus recession and medial rectus resection was supplemented by inferior rectus recession. Later contralateral lateral rectus recession with a Faden procedure and medial rectus resection were performed to encourage medial rectus drive of the affected eye. Levator function was good, and a small posterior levator resection was finally done on the affected side with a contralateral upper lid recession.

The remaining two patients had congenital third nerve palsies. One had previously undergone both vertical muscle surgery and ptosis correction. Both had moderate levator function. They underwent Knapp procedures, achieving only reduced Bell’s phenomena. Large anterior levator resections were subsequently carried out.

**NEUROFIBROMATOSIS (3 patients)**

Debulking of the plexiform neuromata preceded correction of hypotropia and ptosis in two cases. One patient had a good Bell’s phenomenon with reasonable elevation of the globe and underwent anterior levator resection alone. Of the other two, one had a Callahan procedure with inferior rectus recession prior to a large anterior levator resection and further debulking. The third had a Knapp procedure with inferior rectus recession. This was followed by upper lid debulking and canthal raising plus a dacryocystorhinostomy. Prior to a large anterior levator resection, lower lid scleral grafting was required to lengthen the lower lid retractors.

**SUPERIOR RECTUS SLING INDUCED (2 patients)**

Both patients presented with ptosis and hypotropia following surgery elsewhere at which the superior rectus muscle had been attached to the upper eyelid to elevate it. One case in which the original ptosis was acquired following trauma required division of the superior rectus sling first, to restore proper superior rectus function. The levator disinsertion was then corrected via an anterior approach. The other case was of congenital ptosis. A Knapp procedure was performed followed by a large anterior levator resection, with freeing of all the adhesions of the previous superior rectus sling operation.

**Results**

**CONGENITAL DOUBLE ELEVATOR PALSY GROUP**

The correction of blepharoptosis may be considered in terms of both pseudoptosis and real ptosis. Following the Knapp operation as described by Lee et al., successful active elevation of the globe is accompanied by correction of the pseudoptosis and improvement of the Bell’s phenomenon. This was
achieved in three of the four cases not previously operated on; these patients underwent Knapp procedures and had good levator function. The fourth patient had poor levator function and gained improvement rather than correction of the hypotropia and pseudoptosis. Eight patients had previously undergone surgery to the vertical rectus muscles with no improvement of the hypotropia, pseudoptosis, or Bell's phenomenon. These were satisfactorily corrected by a Knapp procedure in six of the eight cases. The remaining two had considerable scarring round the common sheath, but were improved. Three patients had undergone ptosis surgery with residual ptosis, pseudoptosis, and hypotropia. Correction of the pseudoptosis and elevation of the globe were achieved by Knapp surgery.

The element of real ptosis was corrected to within 1 mm, with acceptable cosmesis, in 13 cases with surgery based on the guidelines relating levator function to the amount of levator resection required. Where a large resection was indicated, it was facilitated by the good surgical exposure gained via an anterior approach.

Two patients had a measured overcorrection, one of whom needed an upper lid recession. Two of the 13 patients, who had cosmetically good results with lid levels within 1 mm of each other, had a functional overcorrection reflected by punctate corneal staining. These were asymptomatic and treated conservatively with lubricants.

**Jaw-Winking Group**

Of the six patients who underwent primary Knapp or Callahan procedures one was cosmetically satisfied and had good levator function. Of the three requiring levator resection, one was corrected despite the complication of orbital asymmetry due to a baso-cranial teratoma. The second had been unsuitable for brow suspension in the absence of an adequate Bell's phenomenon. The third is yet to undergo ptosis surgery. Two patients with significant jaw-winking were corrected by brow suspension and levator excision procedures. Two patients underwent levator resection alone (including the retarded child) and were found to have residual pseudoptosis requiring a Knapp operation.

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**Fig. 1** Primary gaze showing hypotropia of the right globe and blepharoptosis.

**Fig. 2** Cover test showing elevation of the right globe with correction of the pseudoptosis.

**Fig. 3** Post-Knapp procedure showing correction of the hypotropia and revealing the true blepharoptosis.

**Fig. 4** After anterior levator resection, demonstrating symmetry of globes and lids.
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THIRD NERVE PALSYS GROUP
The Bell's phenomenon was not markedly improved in this group. After the Knapp operation, supplemented by contralateral superior rectus surgery, as described above in the case with misdirection regeneration, levator resection achieved a satisfactory eyelid elevation correcting the lid levels to within 1 mm of each other. The incidence of punctate corneal staining was high in this group, as the Bell's phenomenon was characteristically poor. This was particularly marked in the case with misdirection regeneration, whose poor ocular movements had necessitated bilateral strabismus surgery. However all the patients were young, with good tear production, and have tolerated the punctate staining with the aid of lubricants over a three-year follow-up period.

NEUROFIBROMATOSIS
The ptosis was completely corrected in one case. A 50% improvement of the ptosis was achieved in the other two cases. Residual bulkiness of the lid and some hypotropia of the globe remained in all cases.

SUPERIOR RECTUS SLING-INDUCED GROUP
The ptosis was corrected to within 1 mm and the hypotropia was almost completely corrected in both cases. There was a residual minimal vertical diplopia in the post-traumatic case.

Discussion
The group with congenital double elevator palsy illustrates the principles involved in the management of combined hypotropia and blepharoptosis. In the presence of good levator function, pseudoptosis tends to account for much of the blepharoptosis and may be corrected by a Knapp procedure alone. Two such patients did not require further surgery, and two required a small posterior levator resection. Beard reported a lack of impressive improvement using a Knapp procedure alone. Our experience suggests that this depends on the levator function.

Pseudoptosis may be difficult to assess in young children, particularly where fixation by the affected eye is poor, secondary to amblyopia. Where the magnitude of the pseudoptosis is in doubt, it should be assumed to exist in the presence of a hypotropic globe. The globe should be elevated first with a Knapp procedure before ptosis surgery is performed.

Satisfactory elevation of the globe and improvement of the Bell's phenomenon was achieved in all four cases not previously operated on, including one with a large deviation (55 dioptres). Patients who had previously undergone surgery to the vertical rectus muscles were not corrected until a Knapp operation was performed. Full correction of blepharoptosis can safely be planned only in the presence of an adequate Bell's response, and in our experience the Bell's phenomenon was usually improved after a Knapp procedure. The magnitude of levator resection can be reduced where exposure problems are anticipated, as in the absence of a good Bell's phenomenon.

Inferior rectus recession may cause lower lid retraction, which can aggravate corneal exposure. It is significant, therefore, that in this series it was not found necessary to recess the inferior rectus in most cases (two out of 15 in the congenital double elevator palsy group). This emphasises the importance of preoperative assessment by means of the forced duction test in deciding upon inferior rectus recession. The age factor, with respect to anterior segment ischaemia, must be borne in mind when considering disinsertion of more than two extraocular muscles. Most patients in this series ranged from 3 to 18 years of age, but one aged 23 underwent an uncomplicated Knapp procedure with simultaneous inferior rectus recession without ischaemic problems.

Inferior rectus recession was routinely performed by Callahan and inferior rectus retraction requiring recession was reported in 73% of cases by Scott and Jackson. The need for this high rate of inferior rectus recession was not confirmed in this series.

The management of the ptosis in patients with jaw-winking has received much attention. This series points to the relatively high incidence of jaw-winking in patients presenting with a ptosis and hypotropia (25%) and should alert the ophthalmologist to look for the Marcus-Gunn phenomenon in all patients presenting in this way. The importance of extraocular muscle surgery in achieving symmetry of the eyes and an improved Bell's phenomenon cannot be overemphasised, as this governs the type of ptosis surgery which is subsequently indicated.

The cases with third nerve palsy illustrate the importance of normal horizontal rectus muscles in the achievement of good results following Knapp procedures. The patient with misdirection regeneration required supplementary contralateral surgery to the horizontal rectus muscles with a Faden procedure on the lateral rectus to increase adduction of the affected eye. This in turn aided active elevation of the hypotropic globe. Poor elevation of the globe limited the amount of levator resection it was safe to perform. Symmetry was therefore achieved by a planned, small, ipsilateral levator resection combined with contralateral upper lid recession. Both patients with congenital third nerve palsy were found to have persistently poor elevation following their Knapp procedures. They did not suffer from exposure problems, despite full correction of their
true ptosis, but it should be remembered that tear production is generally adequate to compensate in young patients.

The patients with neurofibromatosis have a continuing problem with recurrence of abnormal lid tissue during the growing period. Thus debulking must often be repeated. The levator is often grossly infiltrated, such that measurement of a resection is inaccurate. The globe may be buphthalmic as well as hypotropic. A Callahan procedure was preferred in one case because of the large abnormal globe, which was at risk of ischaemia. Levator resection following either a Callahan procedure or a Knapp operation achieved good results in two patients. The third patient was educationally subnormal, and debulking with levator resection provided a satisfactory result.

The last two cases show that the superior rectus may be weakened mechanically by a superior rectus sling which incompletely corrects the original ptosis. The management follows the same general principles of active elevation of the globe and levator surgery. Thus removal of the sling with division of adhesions to the superior rectus was the first step, and it could be supplemented by a Knapp operation. This was followed by appropriate levator surgery—reinsertion in the traumatic case and resection in the congenital ptosis case.

In summary, hypotropia, ptosis, and levator function must be assessed carefully. Surgical correction is best achieved by the logical sequence of a Knapp procedure followed by levator resection. This eliminates the confusion introduced by pseudoptosis and provides a bonus safety factor of an improved Bell’s phenomenon at the time of lid raising. Horizontal deviations are conveniently corrected by modifications of the Knapp procedure. Inferior rectus recession is seldom required, and it is known to increase complications related to the lower lid position and normal lid closure. Superior rectus resection when the muscle is known to be weak is illogical and unhelpful. It can be associated with disturbance of the upper fornix, which may require mucous membrane grafting. Superior rectus surgery is said to impair levator function, since the attachment of the common sheath is interfered with. This may make the ptosis worse, often without any significant effect on the position of the globe. After a Knapp procedure a levator resection may be quantified and reliable ptosis correction thereby achieved. A brow suspension may be substituted where levator function is very poor or in jaw-winkers in whom the levator muscle is excised.

References

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