

A New Approach for Management of Monocular Elevation Deficiency

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Abstract

Aim: To show that the classic Knapp procedure when used for treatment of monocular elevation deficiency may not correct it completely especially when associated with pseudoptosis and to suggest a treatment option.

Methods: Thirteen patients (2 to 6 years) with congenital monocular elevation deficiency (MED) were studied. All patients had hypotropia, pseudoptosis, absent elevation above the primary position and negative forced duction test of the affected eye. Medial and lateral rectus muscles were elevated via Knapp procedure. Two months later, the inferior rectus muscle of the non-affected eye was resected 6 mm for three patients and tucked for the other two patients. On follow up, vertical deviation marginal reflex distance were recorded after each procedure.

Results: After Knapp procedure, hypotropia was moderately improved and pseudoptosis appeared worse in three of the five patients. On the other hand, a good lid height and an improvement in hypotropia were achieved after resecting the contralateral inferior rectus muscle for all patients.

Conclusion: In cases of MED with pseudoptosis and on planning for Knapp procedure, inferior rectus tuck or resection of the non-affected eye may be required for successful management of such cases.

Introduction

Monocular elevation deficiency (MED) was first described by White in 1942 as a congenital deficiency of upgaze with hypotropia and ptosis or pseudoptosis of the affected eye [1]. Watson [2] developed an effective surgical procedure for MED which was later reported by Knapp [3]. Knapp described a complete transposition of the horizontal rectus muscles to a position adjacent to the insertion of the superior rectus. Knapp's procedure is commonly practiced and is now a well-established treatment for unilateral elevation deficiency [4].

Management of pseudoptosis when associated with monocular elevation deficiency is important as if it is not managed, the lid drooping must be treated as true ptosis with special challenge in absence of Bell's phenomenon. Because the importance of pseudoptosis management, we had studied the effectiveness of Knapp procedure on the treatment of monocular elevation deficiency when associated with pseudoptosis.

Subjects and Methods

From August 2008 to March 2010, 13 patients with MED were included in our study. Patients' ages ranged between 2 and 6 years with a mean of 3.6 years (8 males and 5 females). Each patient received a complete preoperative ocular examination, which included assessment of ocular motility, head posture, pupillary function, visual acuity, cycloplegic refraction, and dilated fundus examination.

All patients had unilateral elevation deficiency, hypotropia, and pseudoptosis. Diagnosis of MED was based on the presence of normal ductions in all positions of gaze except elevation.

Pseudoptosis was determined by covering the uninvolved eye to force fixation with the ptotic eye. If the lid remained ptotic then true ptosis was diagnosed and the patient was excluded from the study. In addition, we excluded patients with horizontal deviation, inferior rectus restriction or those with a previous history of extraocular muscle surgery.

In young patients, Krimsky's Test was used for measuring the deviation. In the others, prism and alternate cover measurements were obtained at six meters. Neutralizing prisms were held in front of the eye with the MED to measure the primary deviation.

During surgery, forced duction testing was first performed after induction of general anesthesia to exclude any restrictive aetiology. Then, using Knapp's procedure, complete (full width) supraplacement of the medial and lateral rectus muscles to a position adjacent to the respective corners of the insertion of the superior rectus was performed for all patients. Two months later, we resected the inferior rectus muscle of the non-affected eye 6 mm for three patients and tucked it 6 mm for the other ten patients.

On follow up visits, we recorded the amount of hypotropia and marginal reflex distance to evaluate lid height position for all patients.

Results

Patients were observed every week until 9 weeks and every 2 months until the last follow-up, which was done after 12 months from the first surgical interference.

Vertical deviation in primary position ranged for from 17 prism dioptres (PD) to 25 PD (mean 21.4 PD, SD: 2.59931). The mean of ocular alignment in primary position after Knapp procedure was 10.8 PD (SD: 2.31495) and the mean marginal reflex distance was 1 mm (SD: 0.40825) (Table 1).

After contralateral inferior rectus tuck or resection, both hypotropia and lid position were significantly improved for the thirteen patients (100%) where the mean vertical deviation was 0.8 PD (SD: 1.06819) and the mean of marginal reflex distance was 2.4 mm (SD: 0.29957) (Table 2).

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Case No	Preoperative Hypotropia (PD)	Postoperative Hypotropia (PD)	Preoperative Lid height (mm)	Postoperative Lid height (mm)
1	23	12	3	1
2	18	10	2	1.5
3	24	8	3	1
4	20	10	3	1.5
5	22	14	3	1
6	22	9	3	0
7	20	12	3	1
8	25	15	2	1.5
9	19	13	3	1
10	20	11	2	0.5
11	25	8	3	1
12	17	8	2	1
13	23	10	3	1

Table 1: Patients' Surgical Results after Knapp.

Case No	Surgery	Observations at final Postop Exam	
		Hypotropia (PD)	Lid height (mm)
1	IR tuck	2	3
2	IR resection	0	2
3	IR tuck	0	2.5
4	IR resection	3	2.5
5	IR tuck	2	2.5
6	IR tuck	0	2.5
7	IR tuck	0	2.5
8	IR tuck	2	2
9	IR tuck	0	2.5
10	IR tuck	0	2
11	IR tuck	1	2.5
12	IR tuck	0	2
13	IR resection	1	2.5

Table 2: Patients' Surgical Results after contralateral inferior rectus tuck or resection.

Case No	Preoperative	Post- Knapp procedure	Post contralateral inferior rectus surgery
1	2	1	5
2	2	2	6
3	2	2	8
4	1	1	6
5	2	2	5
6	2	1	4
7	3	1	5
8	3	1	4
9	3	3	3
10	3	3	6
11	4	3	4
12	3	2	3
13	2	2	3

Table 3: Upper lid Excursion (mm).

Elevation was absent at the time of presentation. This elevation did not improve after Knapp procedure but showed moderate improvement following contralateral inferior rectus surgery (Table 3). None of the patients had difficulty in up gaze of the non-affected eye.

Discussion

Our work demonstrates; for some cases of monocular elevation deficiency, resection or tucking the inferior rectus muscle of the non-affected eye can properly manage residual hypotropia and/or pseudoptosis that may persist after Knapp procedure.

The cause of MED for patients in our study was superior rectus

paresis. The classic teaching treatment is to do Knapp procedure for the affected eye to elevate it and consequently the pseudoptosis will be corrected. This is because it is assumed that in pseudoptosis, the lid is drawn down passively (lid follows the globe) by the hypotropic eye due to facial attachments between the superior rectus and levator palpebrae. These attachments are responsible for pseudoptosis improvement when we force the parietic eye to pick up fixation.

The question of what to do if after Knapp procedure the pseudoptosis did not improve is very important. This is because if it is not managed, the lid drooping must be treated as true ptosis [5]. This will be challenging, as these patients will require either frontalis suspension or levator resection procedure. However they do not have Bell's phenomenon, and consequently they are predisposed to postoperative corneal exposure.

In our work, pseudoptosis did not improve after Knapp procedure and even get worse (Table 1). We suppose that, Knapp procedure elevates the eye passively by medial and lateral rectus muscles and consequently the superior rectus activity decreases with less innervation to the levator muscle and so the lid position does not improve. Resection or tucking the inferior rectus muscle of the non-affected eye induces hypotropia in this eye with extra-innervations to the superior rectus muscle on both sides and consequently, more innervation to the levator which improves lid position.

In addition, increased innervations to the superior rectus of the involved eye had resulted in correction of the residual hypotropia and improvement in its ability to elevate.

Despite our results are encouraging, there is a need for further study involving more cases treated with this technique to ensure prediction of consistent results.

Conclusion

For some cases of MED with pseudoptosis, and on planning for Knapp procedure, inferior rectus tucking or resection of the non-affected eye may be required for successful management of such cases.

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