

Double elevator palsy – Complexities and outcomes

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Abstract

Purpose: To evaluate the surgical outcome and stability of surgical correction in patients with double elevator palsy (DEP). **Methods:** This retrospective study was conducted on hospital records of patients with Double elevator palsy over 3 year period from 2013 to 2016. The cases with positive forced duction test (FDT) had inferior rectus (IR) recession with full tendon width muscle transposition Knapp surgery, while those without positive forced duction test underwent transposition procedure alone. Transposition surgery was combined with recession and resection of horizontal rectus muscles in patients with horizontal strabismus according to the amount of deviation. Eyelid surgery was done in patients with ptosis following strabismus surgery. **Results:** The average age of 9 patients was 10.55 ± 5.36 years (range, 4–19 years). Six patients (66.66%) were female and 3 patients (33.33%) were male. The mean preoperative hypotropia was decreased from 31.22 ± 12.54 prism diopters (PD) (range, 16–45 PD) to 1.11 ± 2.20 PD (range, 0–5 PD) postoperatively. Mean follow-up period was 12.33 ± 14.11 months (range, 3–48 months). Four patients (44.4%) underwent ptosis surgery, and all achieved cosmetically satisfactory results. **Conclusion:** Transposition surgery alone or combined with IR recession is an effective procedure in treatment of double elevator palsy but Surgery for double elevator palsy must be individualized.

Key Words: Double elevator palsy, knapps procedure, force duction test.

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Received Date: 02/12/2017 Revised Date: 19/01/2018 Accepted Date: 03/02/2018

DOI: <https://doi.org/10.26611/1009523>

Access this article online

Quick Response Code:



Website:

www.medpulse.in

Accessed Date:
05 February 2018

Primary inferior rectus (IR) restriction, primary superior rectus palsy, and congenital supranuclear elevator deficiency are among the principal causes that have been described.³⁻⁶ The presence of hypotropia and ptosis at primary gaze and/or abnormal head position are the indicators for surgical procedures. The ideal way to manage. DEP is not settled and several surgical approaches have been described. In this study the surgical outcome and stability of surgical correction in patients with double elevator palsy (DEP) is evaluated.

INTRODUCTION

Double elevator palsy is classically defined as a congenital inability to elevate one eye, with the limitation occurring in adduction and abduction. The term double elevator palsy is a misnomer because, in most cases, the cause for the limited elevation is not a palsy of both elevators but is a tight inferior rectus muscle.^{1,2} Its pathophysiology is not well understood, and various hypotheses have been proposed to explain the condition.

MATERIAL AND METHODS

We reviewed the hospital record of strabismic patient operated at Aravind eye hospital Pondicherry from 2013 to 2016. Cases with limited elevation due to Brown syndrome, thyroid ophthalmopathy, orbital fractures and malignancies as well as cases with previous extraocular surgeries were excluded. 9 patients with congenital DEP were identified and included in the study. Preoperative evaluation included assessment of visual acuity, pupillary

function, cycloplegic refraction, eye movements, slit lamp examination and ophthalmoscopy. Visual acuity was measured using standard Snellen chart. Angle of deviation was measured by the alternate prism cover test in patients with good visual acuity and cooperation and by the modified Krimsky test in young children. Binocular status was assessed with worth four-dot test and randot tests. The degree of motility restriction was recorded using a conventional method from -1 to -4, each grade representing 25% upward limitation from the horizontal midline. All surgical procedures were done under general anesthesia. Forced duction test was performed in all the patient pre operatively. Patients with negative FDT underwent knapp's procedure and those with positive FDT underwent knapp's procedure with inferior rectus recession. Transposition surgery was combined with recession and resection of horizontal rectus muscles in patients having horizontal deviation according to the amount of deviation. An acceptable outcome was defined as absence of no more than 6 PD hypotropia and 10 PD horizontal deviation at the last follow up. The surgical results were obtained on postoperative day 1, 1month thereafter until the last follow-up. Frontalis sling surgery was performed in patients with ptosis which did not disappear after strabismus surgery.

RESULTS

Records of 9 patients with DEP including 3 male (33.33%) and 6 female (66.66%) subjects with average age of 10.55 ± 5.36 years (range, 4–19years) were reviewed. The left and right eyes were affected in 3 and 6 patients respectively. Mean follow-up period was 12.33 ± 14.11 months (range, 3–48months.) Preoperatively 6 patients had only vertical deviations while 3 patients had both vertical and horizontal deviations including esotropia in 1 and exotropia in 2 cases. hypotropia ranged from 16–45 PD(mean 31.22 ± 12.54)preoperatively. Surgical interventions included Knapp procedure in 4 patients, Knapp procedure and horizontal resection and recession in 2 patients, Knapp procedure with IR recession in 3 patients. Mean pre and postoperative vertical deviations were 31.22 ± 12.54 PD and 1.11 ± 2.20 PD(range, 0–5 PD) respectively. Out of three, two patients (with exotropia) underwent horizontal muscle surgery along with knapps procedure and both of them were having no horizontal deviation on last follow up. Pseudoptosis was noted in 5 patients (56%), while clinically true ptosis was present in 4 patients (44%). Amblyopia due to ptosis and anisometropia was present in 4 patients. 4 patients underwent Frontalis sling surgery for true ptosis. Satisfactory eyelid elevation was achieved in all patients.

DISCUSSION

The surgical outcome for DEP is affected by several factors, such as visual acuity, limitation of ductions, IR restriction and associated horizontal deviations. A predilection to right side involvement has been reported in DEP; the right eye was involved in 3 of 4 patients in the series reported by Ziffer *et al*⁵ and in 10 of 18 subjects in the study by Kucak and co-worker.⁷ Likewise we found more instances of right eye involvement (6 of 9 patients). Caldiera⁸ believed that right side involvement is somewhat more common in congenital cases and left side involvement is slightly more common in acquired cases. In general, no particular diagnostic information can be obtained from the laterality of the condition. Knapp⁹ reported satisfactory results in patients with negative traction test by transposing medial and lateral rectus muscles to the level of superior rectus. It is based on the rationale that extraocular muscle transposition alters the point of tangency of the muscle with the globe, there by changing the point of mechanical action¹⁰. The efficacy of transposition surgery has also been reported to be enhanced over time.^{5,9,11-13} The majority of patients with DEP have IR restriction^{1,14}. In these patients, transposition surgery alone is not effective, and combination of IR recession is often needed^{1,7,8}. Combined surgery of IR recession and transposition was reported to provide average correction of 25.8–37.5 PD.^{6,7} Simple transposition surgery, on the other hand, has provided correction ranging from 18.6 to 38 PD.^{5-7,12} Several factors including age, duration and amount of initial deviation, and sensorial states may be responsible for the differences in outcome. In our study both the procedure met satisfactory correction of hypotropia. The optimal surgical treatment for DEP associated with a significant horizontal deviation is open to discussion. There are several possible surgical approaches described, including procedures both on the diseased and the normal fellow eye. One option is correction of horizontal deviation by a surgical intervention in the fellow eye and Knapp procedure in the affected eye. However, most patients and/or their parents do not allow surgery on the normal fellow eye. Satisfactory results may also be obtained by recessing or resecting the horizontal muscles simultaneously in the affected eye.⁷ In our study Out of 3 patient with horizontal deviation 2 patient underwent horizontal recession and resection surgery along with knapps procedure and obtained satisfactory results for horizontal deviation. One patient didnot underwent horizontal muscle surgery due to relatively small angle. Kamlesh and Dadeya¹⁵ recommended a modified Knapp procedure for DEP associated with horizontal deviation in which, transposition of the superior half of equally divided medial and lateral recti were used to correct vertical

deviation and the inferior half for horizontal deviation correction. In this study both patient underwent full tendon transposition along with recess and resect procedure.

CONCLUSION

The surgical approach for DEP must be tailored according to the degree of deviation, association of horizontal deviation, and IR restriction to obtain satisfactory results.

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Source of Support: None Declared
Conflict of Interest: None Declared