

## Congenital Double Elevator Palsy with Sensory Exotropia: A Unique Surgical Management

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### Abstract

**Purpose:** To report a unique surgical approach for congenital double elevator palsy with sensory exotropia.  
**Case Report:** A 7-year-old boy with congenital double elevator palsy and sensory exotropia was managed surgically by Callahan's procedure with recession and resection of the horizontal recti for exotropia without inferior rectus recession, followed by frontalis sling surgery for congenital ptosis.  
**Conclusions:** Favourable surgical outcome was achieved without any complication.

**Keywords:** Callahan's Procedure; Congenital Double Elevator Palsy; Ptosis

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### INTRODUCTION

The term double elevator palsy (DEP) was first used by Dunlap.<sup>[1]</sup> DEP is a condition characterized by a monocular elevation deficit in all superior gaze positions due to the unilateral restriction of elevation as well as weakness of one or both elevator muscles, the superior rectus (SR) and inferior oblique. Metz believed that both elevator muscles of one eye are weak and leads to restricted elevation and thus hypotropia.<sup>[2]</sup> Scott and Jackson<sup>[3]</sup> stressed the importance of concomitant inferior rectus (IR) restriction as the cause or sequel of DEP. Ziffer et al<sup>[4]</sup> classified DEP into three subsets, namely, primary SR palsy, primary IR restriction, and congenital supranuclear palsy. Few available studies have investigated the surgical management of these cases and have reported variable

outcomes with appropriate surgical planning. However, the surgical management of congenital DEP as performed in this case is not available in the literature.

### CASE REPORT

A 7-year-old boy presented to the Outpatient Department of Ophthalmology, Himalayan Institute of Medical Sciences, with a complaint of drooping of the left upper eyelid with downward and outward position of the eye ball since birth, and it was associated with poor vision without diplopia. He had no past history of birth trauma or ocular surgery. His best corrected visual acuity (BCVA) for the right and left eyes were 20/20 and counting fingers at 3m, respectively. The pupils were equal, round and reactive without evidence of a relative afferent papillary defect. The results of right eye examination was unremarkable, but the left eye showed severe ptosis with 3 mm of vertical palpebral aperture without a lid crease and poor (2 mm) levator action. Bell's phenomenon was good and corneal sensations were normal. Jaw-winking phenomenon, head tilt and chin elevation were not seen.

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Elevation was absent in left eye in all upward gazes [Figure 1a-c]. The rest of the extraocular movements for both eyes were normal. His left eye had 50 prism diopters (PD) of exotropia (XT) and 30 PD of hypotropia [Figure 2a]. The forced duction test (FDT) was performed for restriction of the IR, but the result was negative. Fundus findings were unremarkable for the both eyes.

Full neurological and pediatric work up was done to rule out acquired DEP. A diagnosis of left eye congenital DEP with sensory XT and sensory amblyopia was established.

In view of strabismic and sensory amblyopia, surgical management was mandatory. Other indications for surgery were the vertical deviation in primary gaze, ptosis, and poor cosmesis. Surgery for strabismus of left eye was planned under general anaesthesia. Left eye lateral rectus recession of 8 mm and medial rectus resection of 6 mm was performed along with the vertical transposition of the upper split half of the lateral and medial recti to the temporal and nasal half of the split SR [Figure 3].

Postoperatively, the Hirschberg test was central without any evidence of anterior segment ischemia or overcorrected hypotropia [Figure 2b]. After 1 month, left eye ptosis surgery was performed via a frontalis sling by using silicone rod. Postoperatively, no residual strabismus or ptosis was seen. Proper closure of left eye was observed without any evidence of the lagophthalmos [Figure 4a-c]. Thereafter, amblyopia therapy was instituted for the left eye, and the BCVA improved to 20/120 after 3 months.

## DISCUSSION

A head tilt, vertical deviation in primary gaze, and ptosis may also be present in DEP. This lowered eyelid position may result from the globe's hypotropic position (pseudoptosis) or intrinsic levator weakness (true ptosis) or both.<sup>[2]</sup>

Depending on the etiology, different surgical methods are available for treatment of DEP. The technique of elevation of the horizontal rectus insertions to relieve hypotropia is usually termed as the Knapp procedure. In 1969, Knapp reported successful correction of DEP by disinserting the entire medial and lateral rectus



**Figure 1.** (a-c) Extraocular movements showing the elevation deficit in the left eye at all gaze positions.

muscle tendons and transposing them superiorly near the insertion of the paretic SR muscle. This enhanced the elevation of the globe, which was not possible by resection of the paretic SR muscle.<sup>[5]</sup> Lee et al recommended FDT to rule out IR restriction; if it were present, the Knapp procedure could be combined with IR recession, and otherwise, the Knapp procedure alone should be done.<sup>[6]</sup> In 1981, Callahan reported three cases of failed ptosis surgery in the presence of hypotropia of the same eye. He described a new procedure in which the superior and horizontal recti were split and united similarly to the Jensen procedure along with IR recession.<sup>[7]</sup> The rationale was to avoid the effect of surgery on the anterior ciliary circulation, as the horizontal recti were not disinserted.

Scott and Jackson<sup>[3]</sup> reported a high incidence of inferior restriction due to secondary contracture, which led to deficient Bell's phenomenon and an accentuation in the lower eyelid fold on attempted upgaze of the affected eye. They also reported overcorrection of the upper eyelid after proper vertical ocular alignment in two patients with previous history of ptosis surgery.

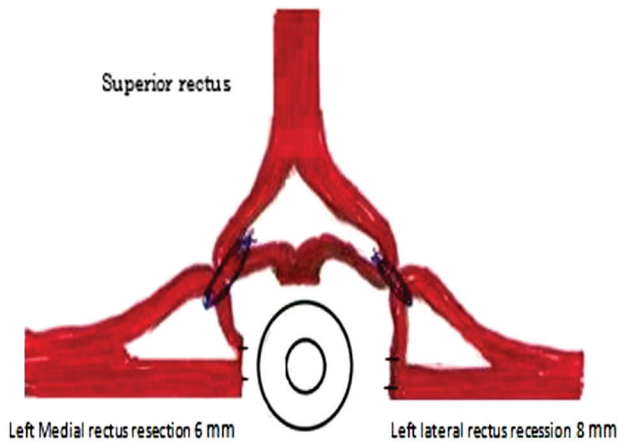
Ficker et al<sup>[8]</sup> reported simultaneous horizontal rectus recession or resection with a Knapp procedure in cases of congenital DEP.

Beard<sup>[9]</sup> stated that vertical muscle surgery should be attempted before final evaluation and correction of ptosis because the correction of the vertical tropia will correct the pseudoptosis along with the residual true ptosis. SR resection yielded poor results because paretic or congenitally hypoplastic SR is associated with levator muscle dystrophy. Bagheri et al<sup>[10]</sup> stated that pure paretic DEP has the best prognosis because of less chances of downward limitation and resurgery in contrast to the pure restrictive type, which had the worst prognosis.

In the present case, XT was corrected by recession and resection of the lateral and medial recti, respectively, and hypotropia in the same eye was corrected by the vertical transposition of the upper split half of the recessed lateral



**Figure 2.** (a) Preoperative image showing hypotropia and exotropia of the left eye. (b) Image after Callahan's procedure.



**Figure 3.** Schematic diagram showing Callahan's procedure for hypotropia with additional simultaneous recession and resection of the horizontal recti for exotropia.

and resected medial recti to the temporal and nasal half of the split SR without IR recession in the same sitting. Severe left eye ptosis with poor levator action was corrected using a frontalis sling surgery after 1 month. To the best of our knowledge, the surgical management of hypotropia and XT performed in the present case has not been reported in the literature. Anterior segment ischemia was absent in the present case in which both the horizontal recti were disinserted for recession and resection of XT without IR recession, as compared to Callahan's procedure in which the horizontal recti remained attached to their insertions.

We conclude that Callahan's procedure without IR recession is a safe and effective procedure along with recession and resection of the horizontal recti in cases of XT with DEP. Such a surgical procedure can be considered prior to surgical intervention for the associated congenital ptosis in view of obtaining correct and precise quantitative assessment of the eyelid position after elevating the globe.

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**Figure 4.** (a) Preoperative image showing ptosis of the left eye. (b) Image after the frontalis sling procedure (1 week). (c) Image after the frontalis sling procedure (6 weeks).

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### Conflicts of Interest

There are no conflicts of interest.

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