

# Comparative Study of Outcomes by Surgical Procedures and Monocular Elevation Deficiency Syndrome in Indian Population

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## Research Article

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# Abstract

## Introduction:

Monocular elevation deficiency syndrome (MEDS) is a monocular elevation deficiency in abduction and adduction characterised by the hypofunction of superior rectus (SR) and inferior oblique muscles. Only a limited number of studies are published on the management of this problem. Herein, the indications and types of surgery for monocular elevation deficiency syndrome are reported.

**Methods:** In this retrospective cohort study, all MEDS patients reported during 2010 to 2020 were retrieved from Electronic Medical Records after approval from the Hospital Ethical Committees. The data were exported in a pre-tested online format. The demographic and clinical information and data of surgeries, including strabismus and ptosis, were reported. The follow-up data were collected in a specific format. Strabismus was managed using Knapp procedure, Inferior Rectus recession and a combination of both approaches. Ptosis was corrected by Levator Palpebral Superioris (LPS) resection and brow suspension. The data were analysed using SPSS 22 based on descriptive analysis and cross-tabulation.

**Results:** The cohort consists of 62 cases (females 26, males 36) of patients with mean age  $17.00 \pm 12.31$  years 40/62 cases comprised the paediatric age group. Knapp was carried out in 39, IR recession was carried out in 35, and combined Knapp+IR recession was performed in 17 cases based on FDT and FGT data. The comparative study found significant difference amongst these techniques ( $P=0.04$ ). Ptosis correction was carried out by LPS resection in 21 and brow suspension in 23 cases depending on LPS function. This comparative study did not find any significant difference in the results ( $P=0.234$ ).

**Conclusion:** MEDS is a rare disease with various clinical features. The surgical management of strabismus may have more success for the combined technique than for those applied alone.

## Introduction

Monocular elevation deficiency syndrome (MEDS) is the limitation of elevation of the affected eye that is similar in both adduction and abduction. MED is one of the causes of hypotropia and can be associated with ptosis/pseudoptosis <sup>[1]</sup>.

MEDS is classified into three types. In Type 1, the aetiology is inferior rectus restriction and thus includes patients with primary inferior rectus restriction or fibrosis. The forced duction test (FDT) demonstrates tight inferior rectus. The upward saccades are usually normal until the tight inferior rectus limits the up gaze. The Bell's phenomenon is exhibited poorly <sup>[1, 2]</sup>.

In Type 2, the aetiology is deficient innervation of the elevator muscles and thus includes primary superior rectus palsy. FDT is free, and upward saccades are slow below and above the midline, while the Bell's phenomenon is absent <sup>[1, 2]</sup>.

In Type 3, the aetiology is the supranuclear type and is usually congenital. It is characterised by intact or mildly reduced vertical saccadic velocity below the midline but abnormal or absent velocity above the midline. Also, FDT is unlimited [3].

The efferent tracts for up gaze leave the rostral interstitial nucleus of the medial longitudinal fasciculus, cross the midline in the posterior commissure, cross through the pretectum, enter the superior rectus subnucleus of the oculomotor nucleus, leave the subnucleus and then cross the midline again. As a result of double decussation, the superior rectus receives innervation from the ipsilateral rostral interstitial nucleus of the medial longitudinal fasciculus, contralateral pretectum and superior rectus subnucleus. In MEDS cases, the supranuclear input from the rostral interstitial nucleus of the medial longitudinal fasciculus into the third cranial nerve nucleus is interrupted. The supranuclear deficiency affects the up gaze, therefore causing deficient elevation [1, 4].

## Methods

This study was approved by the Institutional Ethics Committee of Drashti Netralaya, Dahod, Gujarat, India (DNEC No. 23/2019). Informed consent was obtained from the patients or parents/legal guardians for clinical data, samples, and publication of photographs. All interventions were performed in accordance with relevant guidelines and regulations.

Patient cohort and data collection: In this retrospective cohort study, we recruited paediatric, adolescent and adult patients who presented outdoor ophthalmic and were ready to participate in this study; the features indicated MEDS for over a decade. All information, including demographic and clinical data, was recorded. Following documentation of anterior and posterior segment motor and sensory data, clinical information was retrieved from electronic medical records and exported to Excel. In addition, all information on blepharoptosis was documented and exported.

All information was analysed SPSS 22 (no financial interest) using the descriptive method and cross-tabulation. P-value<0.05 was considered significant.

## Results

In this study, we recruited 62 patients (26 (41.9%) females and 36 (58.1%) males) with mean age  $17.00 \pm 12.31$  (range: 74) years. The cohort consisted of 40 (64.5%) paediatric and 22 (35.5%) adult patients (Table 1).

The Bell phenomenon was observed in 41 (66.1%) patients, while it was poorly detected or absent in 21 (33.9%) patients. Also, amblyopia was observed in 42 (67.7%) eyes. Eye examination revealed that FDT was positive in 28 (41.9%), negative in 36 (58.0%) cases, while strongly positive FGT was observed in 12 (19.4%) and negative in 45 (72.6%) cases; Marcus Gun (jaw winking) was present in 14 (22.6%) cases. Furthermore, the difference between the pre ( $15.31 \pm 11.03$  Prism Diopter) and postoperative mean

vertical deviation  $3.22 \pm 1.2$  PD was statistically significant ( $P=0.05$ ). Preoperatively, 8 (12.9%) cases had horizontal deviation, and postoperatively, 46 (74.2%) were orthophoric in primary gaze.

Knapp's procedure was carried out in 39 (62.9%) cases, inferior rectus recession was conducted in 35 (56.5%), and combined Knapp+IR recession was carried out in 17 (27.4%) cases (Table 2). The comparison between the three techniques revealed significant differences (Table 3).

The preoperative superior rectus movement was weakly positive in 24 (38.7%) and negative in 38 (61.3%) cases. Interestingly, no significant difference was detected while investigating the superior rectus movement on presentation ( $P=0.576$ ).

While considering ptosis, the mean LPS function was  $6.08 \pm 5.90$ , and the mean amount of droop was  $5.24 \pm 2.8$ . Herein, we operated using LPS in 21 (33.9%) and brow suspension in 23 (37.1%) cases. The surgical results did not show any significant differences between the two techniques ( $P=0.234$ ).

## Discussion

MEDS or "double elevator" palsy is a descriptive term denoting a congenital deficiency of monocular elevation that is equal in abduction and adduction [5, 6]. Luo et al reported the mean pre- and postoperative deviation is  $32.0 \pm 8.0$  PD and  $3.8 \pm 8.0$  PD ( $P < 0.001$ ), respectively. In the current study, these values were  $15.31 \pm 11.03$  PD and  $3.22 \pm 1.2$  PD ( $P=0.00$ ), respectively [2]. Luo et al. also reported similar findings [6].

The genetic basis of MED is yet unclear. Previously, it was speculated to arise due to a supranuclear abnormality [7, 8]. In the current study, paediatric (64.5%) revealed phenomena similar to those in the study by Luo et al. [6]. We also reported a jaw winking phenomena in 14 (22%) cases, which has not been shown previously.

The Bell phenomenon reported in identical twins [5] was not observed in the current study. Khan et al. reported a case with double elevator weakening for unilateral congenital superior oblique palsy with ipsilateral superior rectus contracture and lax superior oblique tendon [9].

Nagpal et al. reported congenital double elevator palsy with sensory exotropia. [10]. Verma et al. reported a rare case of golden harr syndrome associated with MEDS [11]. Khawam et al. demonstrated coexistence of MEDS with Duane's retraction syndrome [12]. Munoz et al. reported acquired MEDS in association with pineocytoma [13].

Typically, the choice of surgery in the management of MED depends on the FDT results. In patients with a positive FDT, inferior rectus recession is the first choice of management. If there is no restriction, the transposition of the medial and lateral rectus muscles to the superior rectus muscle could be performed (Knapp procedure). Other options include resection of the ipsilateral superior rectus muscle or recession of the contralateral superior rectus muscle [1, 14, 15].

The surgery for double elevator palsy must be customised according to FDT, FGT, and Bell's reflex. The outcomes are favourable with appropriate surgical planning, as reported by Bagheri et al. [2].

According to Metz et al., superior rectus muscle and inferior oblique muscle insufficiency are the primary factors in the aetiology of congenital double ocular elevator palsy. Vertical rectus recession and superior oblique tenectomy on paretic eyes are effective surgical procedures for the treatment of the disease [16]. Burke et al. reported that vertical transposition of the horizontal recti (Knapp procedure) could be applied to treat double elevator palsy due to its effectiveness and long-term stability [17].

The Knapp procedure had an increasing effect in a time-dependent manner, but the amount of vertical correction was not correlated with the size of the preoperative vertical deviation and was less predictable when a prior IR had been performed. The current study had similar findings ( $P=0.06$ ).

Caldeira et al. studied a similar technique (Knapp) in early-age acquired cases. This transposition procedure could reduce or eliminate the distal and proximal vertical deviation; similar findings with increased effects were observed in the current study in a time-dependent manner [18].

Kocak-Altintas et al. and Yurdakul et al. also studied Knapp procedure or inferior rectus recession without any comparison. We proposed that the surgical intervention should be selected according to DEP clinical features. The surgical effect of transposition surgery may be enhanced by IR recession; a similar finding was reported previously [19, 20].

Li et al. tried Knapp and modified Knapp procedure with Foster sutures for this condition [21]. Snir combined rectus muscle transposition and posterior fixation suture for MEDS [22]. Luo et al. compared Knapp and Foster procedures and revealed that the surgery selection is based on the angle of vertical deviation.

## Conclusion

Surgery for MEDs may be selected based on FDT, FGT, and Bell's phenomenon in order to improve the deviation in primary gaze. In addition, ptosis or pseudo ptosis may be treated according to LPS activity.

## Declarations

**Conflicting interests:** None to declare.

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## Tables

Table-1 Age sex distribution

AGE GROUP	SEX		TOTAL
	FEMALE	MALE	
0-10	7	4	11
11-20	15	19	34
21-30	3	7	10
31-40	1	3	4
41-50	0	1	1
51-60	0	1	1
>60	0	1	1
TOTAL	26	36	62

Table-2 Type of surgical techniques

SURGERIES	FREQUENCY	PERCENT
IR RECESSION	15	24.2
KNAPP'S PROCEDURE	20	32.3
KNAPP+IR RECESSION	21	33.9
NOT OPERATED	6	9.7
TOTAL	62	100.0

Table-3 Comparative study of post operative deviation in primary gaze with different surgical management

POST OPERATIVE DEVIATION IN PRIMARY GAZE	TYPE OF SURGERY			TOTAL	
	IR RECESSION	KNAPP	KNAPP+IR RECESSION	N	
DEVIATION	2	3	7	4	16
ORTHOPHORIA	13	17	14	2	46
TOTAL	15	20	21	6	62

P=0.040