Double Elevator Palsy, Subtypes and Outcomes of Surgery

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Purpose: To describe the clinical manifestations of subtypes of double elevator palsy and to report the outcomes of surgery in these patients.

Methods: This retrospective study was conducted on hospital records of patients with double elevator palsy at Labbafinejad Medical Center over a ten-year period from 1994 to 2004. Patients were classified into three subgroups of primary elevator muscle palsy (9 subjects), primary supranuclear palsy with secondary inferior rectus restriction (4 subjects) and pure inferior rectus restriction (7 subjects) according to forced duction test (FDT), force generation test (FGT) and Bell's reflex. Patients in the first group underwent Knapp procedure, the second group received Knapp procedure and inferior rectus recession simultaneously and in the third group vertical recess-resect or mere inferior rectus recess operation was performed. Success was defined as final residual deviation ≤5 PD and ≥25% improvement in restriction after all operations.

Results: Overall 20 subjects including 10 male and 10 female patients with mean age of 12.6±9.3 (range 1.5-32) years were operated during the mentioned period which included 9 cases of primary elevator muscle palsy, 4 patients with primary supranuclear palsy and secondary inferior rectus restriction, and 7 subjects with pure inferior rectus restriction. Mean follow-up was 22.0±20.0 (range 3-63.5) months. Mean pre and postoperative deviation was 32.0±8.0 PD and 3.8±8.0 PD (P<0.001) respectively, and mean restriction before and after the operation(s) was -3.5±0.7 and -2.3±1.2 (P<0.001), respectively. Success rate was 77% for correction of deviation and 80% for improvement in muscle restriction.

Conclusion: Surgery for double elevator palsy must be individualized according to FDT, FGT and Bell's reflex. The outcomes are favorable with appropriate surgical planning.

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INTRODUCTION

Unilateral inability to elevate the eye may occur due to a variety of local, and peripheral or central nervous system etiologies.[1] A specific form of the condition was initially described by White[2] in 1942 but the term double elevator palsy (DEP) was first used by Dunlap.[3] DEP implies that both elevator muscles of one eye are weak leading to restricted elevation and hypotropia.[4] Since DEP is usually unilateral, it is sometimes referred to as monocular elevation deficiency as well.[5] Metz[4] believed that true paralysis of the elevator muscles is present in

only one-fourth of cases and that only one of the elevator muscles may be involved in certain cases. In the same manner, Von Noorden[6] attributed DEP to long-term paralysis of the superior rectus (SR) in the presence of a normally functioning inferior oblique (IO) muscle, indicating that the motor signs may be attributed to SR paralysis alone. There are different opinions regarding the etiology of DEP. Jampel and Fells[1] suggested a unilateral supranuclear lesion in the pretectal area near or inside the third cranial nerve nucleus in some cases. Scott and Jackson[7] stressed the importance of concomitant restriction caused by a tight inferior rectus (IR) muscle as the cause or sequel of DEP. Using a scleral search coil technique for evaluating saccadic velocities, Ziffer et al[8] classified DEP into three subgroups of IR restriction, complete or incomplete SR paralysis and supranuclear palsy.

There are different surgical methods for treatment of DEP which reflect the variety of causative mechanisms. Dunlap[3] believed that muscle transposition is justified only when no restriction is detected on forced duction test (FDT). Knapp[9] recommended transposition of the horizontal rectus muscles to the SR insertion. The initial version of this operation included complete transposition of both horizontal rectus muscles such that their new insertions straddled the SR insertion on either side. Scott and Jackson[7] found that IR restriction plays a role in some cases based on FDT findings. In such cases they recommended IR weakening and release of the retractor ligament without transposition of the horizontal recti to avoid the risk of anterior segment ischemia. They also believed that transposition alone would suffice in cases without IR restriction. Callahan[10] established a method to reduce the risk of ischemia by simultaneous IR weakening in conjunction with a modification of Jensen's procedure. This method includes splitting the horizontal rectus muscle bellies along their length and uniting each half with the adjacent half of the weak SR. Lee et al[11] suggested planning the operation according to the result of FDT; in eyes with free movement, Knapp procedure alone would suffice. However, when the IR is restricted, simultaneous IR weakening in addition to muscle transposion should be performed. They believed that the Callahan procedure would be preferred in patients at high risk of anterior segment ischemia such as subjects older than 25 years of age and those with large globes.

The stability of ocular alignment following transposition surgery has been studied in a limited number of reports. Burk et al[12] evaluated long-term stability and efficacy of muscle transposition surgery and concluded that the Knapp procedure has increasing effect over time. However the amount of deviation reduction was not a function of preoperative angle of deviation, furthermore they noted the correlation to be even less marked with simultaneous weakening and transposition surgeries.

The current study presents our 10-year experience with DEP in terms of the diagnosis and surgical planning, and the efficacy and complications of different surgical procedures.

METHODS

We reviewed the hospital records of 1122 strabismic patients operated at Labbafinejad Medical Center from 1994 to 2004. After excluding cases of limited supraduction due to Brown syndrome, thyroid ophthalmopathy, orbital fractures and malignancies as well as cases with previous SR weakening, 20 patients with congenital DEP were identified and included in the final analysis. Preoperative evaluation included assessment of visual acuity, pupillary function, cycloplegic refraction, eye movements, slitlamp examination and ophthalmoscopy. Visual acuity was measured using standard Snellen chart in cooperative patients and by fixation patterns in preverbal children. Ocular deviation was measured by the alternative prism and cover test in patients with good visual acuity and cooperation and by the modified Krimsky test in young children while looking at a distant accommodative target. Neutralizing prisms were held in front of the eye with DEP to measure the primary deviation. Stereopsis was measured using Titmus or

Randot tests and fusional status was assessed with the Worth 4-dot test pre- and postoperatively when feasible. 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. FDT was performed in the operating room before induction of general anesthesia in cooperative cases and after general anesthesia using non-depolarizing muscle relaxants in non-cooperative patients. The active force generation test (FGT) was also performed before general anesthesia in all cooperative patients.

The patients were classified into 3 groups according to the results of FDT, FGT and Bell's reflex as follows: (1) Patients with weak FGT, free FDT and negative Bell's reflex were classified as primary elevator paralysis and underwent Knapp procedure with or without simultaneous horizontal recess and resect (R&R) operation. (2) Patients with restricted FDT, weak FGT and visible Bell's reflex were labeled as having primary supranuclear palsy with secondary IR restriction and underwent simultaneous Knapp procedure and IR weakening. (3) Patients with normal FGT and restricted FDT were diagnosed with pure IR restriction. Bell's reflex results were not uniformly detectable in these patients which can be explained by different levels of IR restriction. Short-term measurements were obtained one week postoperatively and long-term measurements were those obtained at the final visit. Success was defined as residual vertical and horizontal deviations less than 5 PD at final follow-up. Motility success was defined as 25% or more improvement in muscle restriction.

RESULTS

Records of 20 patients with DEP including 10 male and 10 female subjects with mean age of 12.6±9.3 (range 1.5-32) years were reviewed. The left and right eyes were affected in 11 and 9 patients respectively. Mean follow-up period was 22±20 (range 3-63.5) months. Preoperatively, 9 patients had only vertical deviations while 11 patients had both vertical and horizontal deviations including esotropia in 5 and exotropia in 6 cases. Vertical deviation ranged from 20 to 45 PD (mean 32 PD) preoperatively. Upward motility restriction before the operation ranged from -1 to -4 (mean -3.4).

Fourteen patients underwent surgery once, 5 patients required surgery twice, and one patient underwent operation three times. Surgical interventions included Knapp procedure in 5 patients, Knapp procedure and horizontal R&R in 4 patients, Knapp procedure with IR recession in 4 patients, vertical R&R in 6 patients and IR recession alone in 1 patient.

Mean pre and postoperative vertical deviations were 32.0±8.0 PD and 3.8±8.0 PD respectively (P<0.001). Corresponding values for horizontal deviations were 15.6±19.1 PD and 3.0±6.7 PD respectively (P<0.001). Motility restriction was -3.5±0.7 preoperatively and -2.3±1.2 postoperatively. Success rate at final follow-up was 77% for correction of vertical deviations and 80% for improvement in restriction. The results within each subgroup are presented in greater detail below.

Primary SR Paralysis

This subgroup included 9 patients; 7 of whom underwent Knapp procedure alone. Mean vertical deviation in this group was 34.0±8.2 PD (range 20-45 PD) preoperatively. All subjects except patient #2 who developed 10 PD overcorrection, had less than 5 PD of vertical deviation after a mean follow-up period of 31.0± 17.0 months. Two other patients in this group (cases #8 and #9) underwent IR weakening 6 months after initial surgery due to undercorrection; both had less than 5 PD vertical deviation 6 months thereafter (table 1).

Primary Supranuclear Paralysis and Secondary IR Restriction

This group consisted of 4 patients who underwent simultaneous horizontal muscle transposition and IR weakening procedures. Mean vertical deviation was 24.0±4.8 PD (range 20-30 PD) preoperatively. All patients had less than 5

PD of vertical deviation after a mean follow-up period of 7.6±4.0 months (table 2).

Pure IR Restriction

This group included 7 patients who underwent either vertical R&R or IR weakening procedures. Mean vertical deviation was 36.0±6.0 PD (range 30-45 PD) preoperatively. Mean follow-up period was 7.0±5.5 months (table 3). Case #14 had 25 PD under-correction 5 months after vertical R&R and underwent a Hummelsheim operation but was still 7 PD under-corrected 9 months after the second operation; Botulinum injection was performed at that time but the patient was lost to follow-up. Case #19 had 30 PD hypotropia in the right eye and underwent IR recession alone due to severe restriction noted on FDT. The patient was 10 PD under-corrected 6 months after surgery.

Table 1 Characteristics of patients with primary superior rectus paralysis

No	Sex	Age (yr)	Eye	FDT	FGT	Bell's reflex	Upward Limit		Downward Limit		Eye deviation		Surgery	F/U (mo)	Post-op drift (Δ)	Ptosis	Ptosis surgery (Result)
							Pre-op	Post-op	Pre-op	Post-op	Pre-op	Post-op					
1	F	16	OS	-	-	Weak	4-	3-	0	1-	30Δ Lh 14Δ ET	0	Knapp	68	-	Bilateral true ptosis+ JW	Bilateral levator disinsertion (Recovery)
2	M	6	OD	-	NP	Weak	3-	0	0	0	20Δ Rh 20Δ XT	0	Knapp	63.5	10Δ at 1st mo	Pseudoptosis	-
3*	M	3.2	OD	-	NP	Weak	4-	0	0	0	40Δ Rh 60Δ XT	0	Knapp+ horizontal R&R+ 2 other surgeries to correct MR sliding and residual XT	37	30Δ	No	-
4	M	3	OD	-	NP	Weak	4-	3-	0	0	25Δ Rh 40Δ ET	20Δ ET	Knapp+ horizontal R&R	33.5	-	True ptosis	Sling (No change)
5**	M	21	OS	-	-	Weak	4-	1-	0	1-	30Δ Lh 20Δ ET	10Δ Lh	Knapp+ horizontal R&R	32	20Δ	Pseudoptosis	-
6	F	3	OD	-	NP	Weak	4-	3-	0	0	35Δ Lh	0	Knapp	36.5	5Δ	True ptosis	Levator resection and then sling (Recovery)
7	F	9	OS	-	-	Weak	4-	4-	0	0	40Δ Lh 50Δ XT	0	Simultaneous Knapp and horizontal R&R + another surgery to correct MR sliding	15.5	-	True and false ptosis	Levator resection (Recovery)
8	F	14	OS	-	-	Weak	4-	3-	0	1-	40Δ Lh	0	Knapp and IR recess 6 mo later	14	-	True ptosis	Levator resection and then sling (Recovery)
9	F	12	OS	-	-	Weak	4-	3-	0	1-	45Δ Lh 45Δ ET	0	Knapp and IP recess 6 mo later	12	-	True ptosis	Sling (NA)

FDT, forced duction test; FGT, force generation test; Limit, limitation; op, operation; F/U, follow-up; F, female; M, male; OS, left eye; OD, right eye; NP, not possible; Lh, left eye hypotropia; Rh, right eye hypotropia; ET, esotropia; XT, exotropia; R&R, recess and resect; MR, medical rectus, IR, inferior rectus; JW, jaw winking; NA, not available.

*Patient #3 developed right eye exotropia of 40Δ with limitation of abduction and underwent advancement of the left medial rectus and marginal myectomy of left lateral rectus (LLR) due to right medial rectus (RMR) sliding one year after the first operation. He developed relapse of right eye esotropia about one year later and underwent RMR recess of 8 mm together with LLR recess of 7 mm which resulted in orthotropia thereafter.

**Patient #5 was a case of superior rectus aplasia.

Table 2 Characteristics of patients with primary supranuclear paralysis and secondary inferior rectus restriction

No	Sex	Age (yr)	Eye	FDT	FGT	Bell's reflex	Upward limit		Downward limit		Eye deviation		Surgery	F/U	Post-op drift(Δ)	Ptosis	Ptosis surgery (Result)
							Pre-op	Post-op	Pre-op	Post-op	Pre-op	Post-op					
10	F	17	OS	+	-	Good	4-	2-	1-	2-	30Δ Lh	0	Knapp+ IR recess	14	-	True ptosis	She had history of sling with supramid 15 yr ago and underwent sling with silicon rod following strabismus surgery (Recovery)
11	M	5	OD	+	NP	Good	4-	4-	0	3-	20Δ Rh 20Δ XT	0	Knapp+ horizontal recess and R&R	6.5	5Δ at 3.5 mo	Pseudo-ptosis	-
12	F	11	OD	+	-	Good	3-	2-	0	1-	20Δ Rh	0	Knapp+ IR recess	6	-	True ptosis	Levator resection (lost to F/U)
13	M	22	OD	+	-	Good	3-	2-	0	0	25Δ Rh	0	Knapp+ IR recess	4	-	Pseudo-ptosis	-

FDT, forced duction test; FGT, force generation test; Limit, limitation; op, operation; F/U, follow-up; F, female; M, male; OS, left eye; OD, right eye; NP, not possible; Lh, left eye hypotropia; Rh, right eye hypotropia; IR, inferior rectus; R&R, recess and resect .

Table 3 Characteristics of patients with pure inferior rectus restriction

No	Sex	Age (yr)	Eye	FDT	FGT	Bell's reflex	Upward limit		Downward limit		Eye deviation		Surgery	F/U	Ptosis	Ptosis surgery (Result)
							Pre-op	Post-op	Pre-op	Post-op	Pre-op	Post-op				
14*	M	16	OS	+	+	Good	4-	3-	0	3-	30Δ Lh 20Δ XT	7Δ Lh 15Δ XT*	Vertical R+R, Hummelsheim 5 mo later and Dysport injection in the IR 9 mo later	16.5	Pseudo-ptosis	-
15	M	31	OS	2+	+	Weak	4-	3-	0	2-	40Δ Lh	14Δ XT, ortho vertically	Vertical R&R	9.5	Ture ptosis	Sling with silicon rod (Recovery)
16	M	6	OS	+	+	Good	3-	2-	0	1-	30Δ Lh	0	Vertical R&R	7.5	True ptosis + JW	Levator disinsertion and sling with 240 band early in childhood (Recovery)
17**	M	1.5	OS	+	NP	NP	3-	2-	0	1-	45Δ Lh 20Δ XT	0	Vertical R&R	7.5	Pseudo-ptosis	-
18	F	1	OD	3+	NP	NP	4-	4-	0	2-	30Δ Rh	10Δ Rh <20Δ ET	RIR recess	6	True ptosis	Previous sling with supramid (Recovery)
19	F	32	OD	+	+	Good	2-	1-	0	1-	40Δ Rh 10Δ ET	0	Vertical R&R	3	True ptosis	Levator retraction (Recovery)
20	F	18	OD	+	+	Good	2-	2-	0	0	30Δ Rh	0	Vertical R&R	3	True ptosis	Sling with one week support (recovery)

FDT, forced duction test; FGT, force generation test; Limit, limitation; op, operation; F/U, follow-up; F, female; M, male; OS, left eye; OD, right eye; NP, not possible; Lh, left eye hypotropia; Rh, right eye hypotropia; ET, esotropia; XT, exotropia; R&R, recess and resect; RIR, right inferior rectus; JW, jaw winking.

* Patient #14 was lost to follow-up after Dysport injection.

** Patient #17 was a case of inferior rectus restriction and elevator paralysis.

DISCUSSION

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[8] and in 10 of 18 subjects in the study by Kucak and coworker.[13] On the other hand, Khawam and Younis[14] reported that the condition affected the left eye in 3 of 4 patients. Likewise we found more instances of left eye involvement (11 of 20 patients). Caldiera[15] 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.

According to the underlying mechanisms of the disorder, Ziffer et al[8] classified DEP into 3 subgroups of primary IR restriction, primary paresis or paralysis of the SR and those with supranuclear defects. Both Metz[4] and Scott[7] noted IR restriction in the majority of their patients. Scott attributed IR restriction to long-term paralysis of the elevator muscles while Metz considered it as a primary defect. Maruo et al[16] referred to these patients as a subgroup of congenital fibrosis syndrome. Some degree of IR restriction was reported by Metz[4] in 73% of their patients. This figure was 60% in our study (12 of 20 patients). In cases with restricted FDT, we found only 2 patients with simultaneous SR paralysis. Our results therefore confirm Metz' theory on the primary role of IR restriction.

Metz[4] noted two interesting points in the evaluation of 15 patients; all patients with hypotropia in straight gaze had simultaneous IR restriction but no elevator paralysis. On the other hand, in patients with orthotropia in primary position, one half had IR restriction and the other half had elevator paralysis. We evaluated only DEP cases who required surgical correction of hypotropia, therefore we cannot prove the first point but the presence of elevator paralysis in 9 out of 20 patients can be considered to be consistent with the second observation by Metz.

In summary, unilateral DEP includes at least three subgroups with different underlying mechanisms. The surgical procedure should be individualized for each case based on the etiology and mechanisms of involvement which can be determined by a comprehensive clinical

examination and careful evaluation of FDT, FGT and Bell's reflex. Such an approach should lead to appropriate surgical planning and an acceptable outcome. The pure paretic type seems to have the best prognosis due to less induction of downward limitation and less need for reoperation. In contrast, the pure restrictive type had the worst prognosis because of more severe downward limitation postoperatively and more need for reoperation.

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