Double Elevator Palsy with Congenital Esotropia: A Case Study

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ABSTRACT

This case study follows Master JT, a young boy who has congenital esotropia in addition to a right double elevator palsy. Ocular assessment of the patient is outlined as well as management and surgical treatment. The characteristics of congenital esotropia and double elevator palsy are discussed in context with the child's presentation. The importance of performing a forced duction test to determine the classification of double elevator palsy and options or surgery is stressed. Rationale over surgical choices and likely prognosis are included.

Keywords: double elevator palsy, congenital esotropia, surgery, forced duction test, amblyopia

INTRODUCTION

Double elevator palsy (monocular elevation deficiency) has become a term used for any strabismus whereby there is reduced elevation in all horizontal orientations of the eye, not simply the paresis of the ipsilateral inferior oblique and superior rectus as it may imply[1,2]. The incidence is unknown but it appears more prevalent in patients with congenital ptosis or pseudoptosis and Marcus Gunn Jaw-Winking syndrome[2]. It is also not uncommon for a double elevator palsy to present in conjunction with a horizontal strabismus[3]. This case follows Master J.T. who initially presented with congenital esotropia and further testing revealed the patient also had congenital double elevator palsy.

CASE REPORT

Master J.T. presented to the New York Eye and Ear Infirmary at age 19 months for consideration of strabismus surgery following previous non-surgical management elsewhere. His mother first noticed a left esotropia when J.T. was aged 3 months and had been applying alternate patching. J.T. was carried to full term and weighed a healthy 3260 grams at birth. J.T. had no medical condition or allergies and was not on any medication. There is no family history of strabismus or any other ocular condition.

On examination, cycloplegic retinoscopy revealed a small amount of anisometropia R. +1.00 DS, L. -0.50 / -0.25 x 5°. On cover testing there was an alternating fixation with a slight preference for the left eye, indicating no or insignificant amblyopia. At near J.T. measured 25-30Δ esotropia and 10Δ R hypotropia fixing left/and a L hypertropia 10-15Δ fixing right. J.T. adopted a chin up posture when fixating with his right eye, otherwise the head posture was a 15Δ right head tilt and right head turn. The chin-up posture put J.T. into down-gaze, whilst the head tilt and turn to the side of the vertically deviated eye displaced images down and to the side of the unaffected eye, which allowed him to maximise his field of binocular single vision when he was fixing with either eye[4]. On ocular movements there was a significant limitation of all forms of elevation of the right eye by -3 (past the midline) and his left inferior oblique showed +3 over-action. Horizontal movements were full and no signs of a ptosis were present. Alternate patching for 4 hours a day was prescribed to reduce the risk of developing post operative amblyopia and maintain equal vision owing to there being a constant strabismus present.

All of the results regarding the strabismus, ocular motility and head posture remained stable at 21 months so J.T. was scheduled for strabismus surgery to correct the horizontal and vertical deviations caused by the esotropia and double elevator palsy respectively. The result of a forced duction test of the right eye was negative indicating free passive movement of the globe and no mechanical restriction. Left inferior oblique anteriorization was performed to decrease the hyper element, as well as a bilateral medial recti recession (R. 4mm, L. 3.5mm) to correct the esotropia.

Post operatively JT achieved a 2Δ esotropia with 2Δ R hypotropia with a strong fixation preference for the left eye.

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Elevation was not significantly improved as expected but the vertical deviation in primary position was decreased when fixing with his right eye. J.T. was then prescribed maintenance occlusion of one hour daily of the left eye.

DISCUSSION

Congenital esotropia presents within the first six months of life in the form of concomitant deviations of 30Δ or larger and there is generally a family history of strabismus[1,3,4]. This case showed no family history of strabismus but measurements of J.T.'s angle and age of onset are consistent with this classification. J.T.'s fixation alternated freely the majority of time allowing equal vision and alternate suppression making significant amblyopia unlikely[4]. The alternate patching aids in maintaining this balance, and by avoiding amblyopia the likelihood of maintaining stable alignment of the eyes post- operatively was improved[1]. J.T.'s left inferior oblique over-action is a common finding in congenital esotropia but may be influenced by his apparent double elevator palsy[5]. This could be due to the under-action of the left inferior oblique's contralateral synergist (right superior rectus) which requires increased innervations, leading to development of a consequent muscle sequelae according to Hering's law of equal innervations.

J.T.'s ocular movements were consistent with congenital double elevator palsy. There was very little movement past the midline during all attempts of elevation of his right eye which is the major characteristic. Additional features of double elevator palsy include a hypotropia in primary position increasing on up-gaze, ptosis or pseudoptosis and a chin up head posture with fusion in down-gaze or an extra deep lower lid fold[1,3,6]. Commonly when fixing with the unaffected eye the affected eye appears hypotropic and then conversely when fixing with the affected eye there is a large hypertropia of the unaffected eye[2]. J.T. is an excellent example of these findings and showed all signs with exception to ptosis.

Metz believed true paralysis of the elevator muscles was only involved in a quarter of cases[7]. Double elevator palsy is divided into three types. Type 1 is elevator paresis of both the superior rectus and inferior oblique of the deviated eye or the superior rectus alone. Type 2 is mechanical restriction of the inferior rectus and Type 3 is a combination of 1 and 2 due to a long standing paresis and consequent contracture of the inferior rectus[1,2,5]. These categories can be distinguished by saccadic velocity, a forced duction test, presence of Bell's phenomenon and forced generation testing (FGT), which then allow for appropriate intervention to follow[4,8].

The need for treatment is dependent on the result of the "forced duction test", head posture and size of the vertical deviation is in primary position[1]. In cases of mechanical restriction (positive forced duction test) inferior rectus recession is recommended, and those with paresis (negative forced duction test) commonly undergo a Knapp surgical procedure whereby the horizontal recti are transposed towards the superior rectus of the affected eye[3]. Knapp's procedure can produce successful outcomes even when performed in conjunction with horizontal squint surgery[9]. J.T.'s forced duction test showed no mechanical restriction indicating pure paretic double elevator palsy most plausibly due to lesions in the oculomotor fascicle affecting the superior rectus and inferior oblique[6]. Therefore inferior rectus recession was not required and the surgeons chose not to perform a Knapp procedure.

The procedure of bimedial rectus recession used in J.T.'s case is most commonly practiced for congenital esotropia 25-45Δ and was undertaken between age 6 months and 2 years which can be considered optimal[4]. Simultaneous surgery to weaken the overacting inferior oblique is favourable and is expected to improve the hypertropic element of J.T.'s deviation so both eyes appear more balanced (especially during up-gaze) and later possible presentation of dissociated vertical divergence would be minimal[1,4,10]. Benefits of undergoing surgery now include greater potential for binocular single vision, improved interaction with his parents and a reduced mechanical component[4,5].

J.T.'s post operative outcome was favourable, but the strong fixation preference for the left eye puts him at high risk of developing strabismic amblyopia[4]. Therefore regular follow up will be required to prevent any post operative amblyopia development, monitor the ocular position and test for binocular functions. Cycloplegic refractions should be regularly performed as a recurrence of esotropia may result from an accommodative component[4]. Further surgeries may then become necessary[1].

CONCLUSION

It is not uncommon for congenital esotropia to co-existent with double elevator palsy. Clinical investigation should include a thorough patient history, observation of head posture, cover test and ocular motility exam to confirm the expected diagnoses. A forced duction test is then essential for the further classification of the type of double elevator palsy as it detects whether mechanical restrictions are involved or if there is purely muscle weakness. This is necessary for choosing the appropriate surgical intervention to treat the vertical component of the strabismus when it is problematic. Surgery was performed in the optimal timeframe and provided good outcomes in J.T.'s case.

REFERENCES

1. American Academy of Ophthalmology. Pediatric Ophthalmology and Strabismus. USA: American Academy of Ophthalmology, 2006.
2. Gallin, P. Pediatric Ophthalmology: A Clinical Guide. New York: Thieme, 2000.
3. Dornic, D. Ophthalmic Pocket Companion. 6th Ed. USA: Butterworth-Heinemann, 2002.
4. Ansons AM, Davis H. Diagnosis and Management of Ocular Motility Disorders. 3rd Ed. UK: Blackwell Science, 2006.
5. Pratt-Johnson J, Tillson G. Management of Strabismus and Amblyopia: A Practical Guide. 2nd Ed. New York: Thieme, 2001.
6. Wong A. Eye Movement Disorders. New York: Oxford University Press, 2008.
7. Metz HS. Double Elevator Palsy. Arch Ophthalmol 1979;97:901-903
8. Bagheri A. Sahebghalam R, Abrishami M. Double Elevator Palsy, Subtypes and Outcomes of Surgery. J Ophthalmic Vis Res [serial online]. 2008; 3: 108-113.
9. Lee JP, Collin JR, Timms C. Elevating the Hypotropic Globe. Br. J. Ophthalmol. 1986; 70: 26-32.
10. Burke JP, Ruben JB, Scott WE. Vertical Transposition of the Horizontal Recti (Knapp Procedure) for the Treatment of Double Elevator Palsy: Effectiveness and Long-Term Stability. Br. J. Ophthalmol [serial online]. 1992; 76: 734-737.