

Congenital DOUBLE ELEVATOR PALSY: Diagnosis & Management

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Abstract

This paper presents a case of congenital double elevator palsy in a six-year-old boy. It discusses diagnosis, etiology, differential diagnoses, and management, including surgical options for this condition. This case demonstrates the important role optometrists play as primary eye care providers.

Key Words

congenital double elevator palsy, monocular elevation deficit, head tilt, vertical deviation, ptosis, Knapp procedure

Congenital double elevator palsy is a condition characterized by a monocular elevation deficit in all superior gaze positions. Clinically it presents as a unilateral restriction of the superior rectus (SR) and inferior oblique (IO) muscles. A head tilt, vertical deviation in primary gaze, and a ptosis may also be present. A case report will present congenital double elevator palsy in a six-year-old boy. Diagnosis, differential diagnosis, and management are discussed.

Case Report

A 6-year-old African American boy presented to the clinic for an annual eye exam. His visual, ocular, and medical histories were negative. However, there was a history of poor reading performance in school. Uncorrected distance visual acuities were 20/20 OD/OS. Pupils were equal, round and reactive without evidence of a relative afferent pupillary defect, and with normal convergence. Visual fields by confrontation were full OD/OS. Extraocular muscle testing revealed a restriction of the left IO, and left SR. Note in Figures 1A, 1B, and 1C the inability of the left eye to elevate in all directions of superior gaze. However, in lateral and inferior gaze positions (see Figures 2-3) there are no restrictions. Cover testing, in primary gaze, revealed orthophoria at distance, and 6 diopters exophoria at near. Subjective refraction revealed +050sph OU, OD and OS, with visual acuities of 20/20 OD/OS. Phorias

using the Von Graefe technique indicated vertical orthophoria at distance and near, and 4 diopters exophoria at near with compensating BO ranges of x/28/19. Amplitude of accommodation, using the minus lens technique, was 10 diopters OD, and 6 diopters OS. Slit lamp examination was negative. A dilated fundus exam revealed distinct, pink optic nerves, with cup to disc ratios of 0.35 OD and 0.3 OS; retinal vessels of normal caliber OU; maculas were clear OU. Peripheral retinal examination revealed no holes, breaks, or tears in either eye. Neurological examination revealed an intact Bell's reflex and OKN. A diagnosis of congenital double elevator palsy and accommodative insufficiency was made. The patient was asymptomatic in primary gaze, and without a head tilt or ptosis. The patient was referred for a vision therapy evaluation to further determine the accommodative and binocular sensory status. Additionally a visual perceptual evaluation was requested in view of the child's academic performance.

Discussion

The patient presented with a congenital double elevator palsy. A diagnosis (Table 1) is made based upon the clinical presentation of a unilateral elevation deficit in all superior directions of gaze. The patient may also present with a head tilt, a vertical deviation in primary gaze, and a ptosis, which may either be a pseudo or true ptosis. If there is a vertical tropia in primary gaze, a pseudo ptosis may be

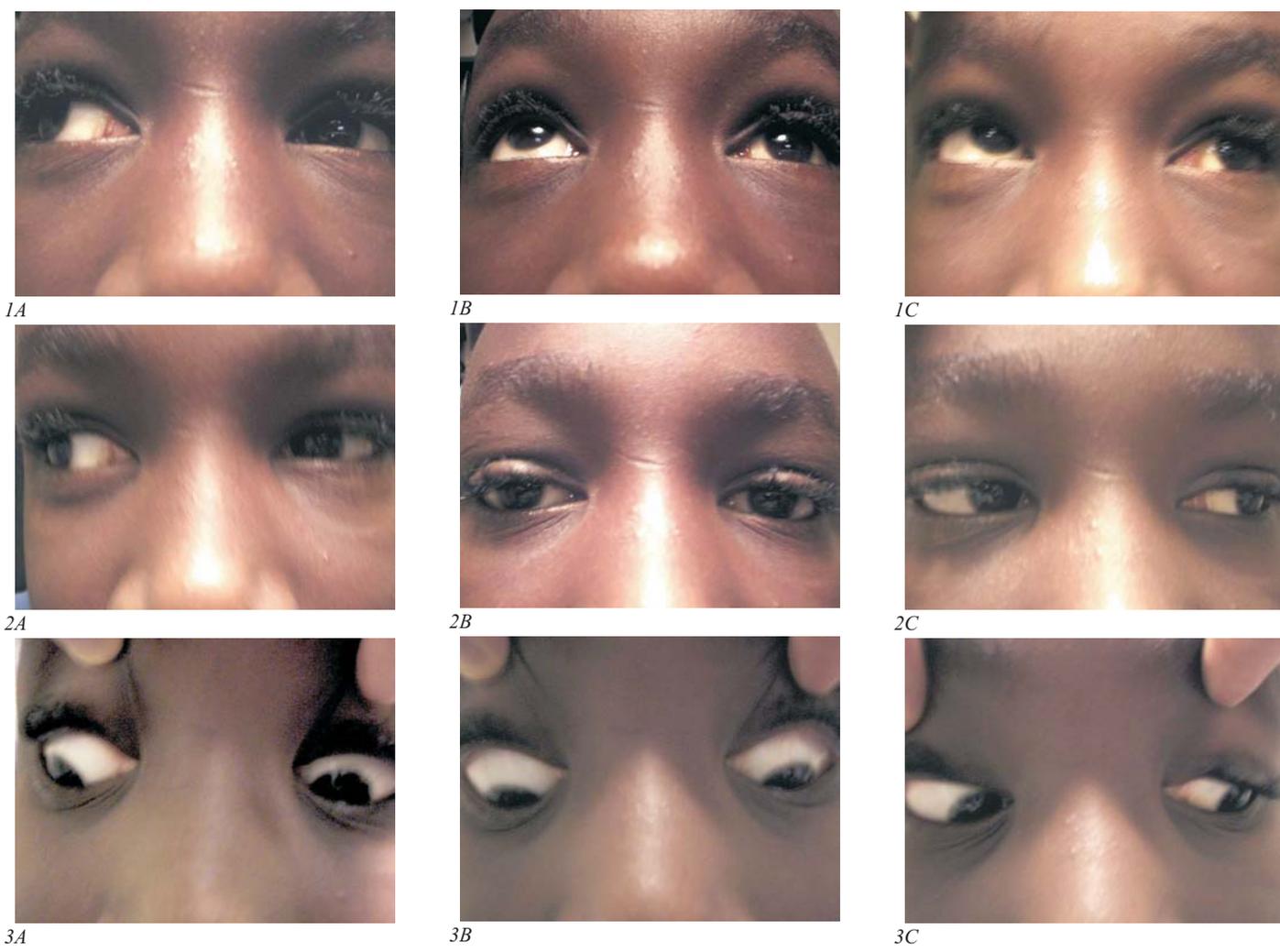


Figure 1. Extraocular motilities (note elevation deficit OS in all positions of upgaze).

present due to the lid following the ‘hypotropic’ position of the eye, as it fixates in the lower visual field.¹⁻³ In other words, when the patient is fixating with the non paretic eye the paretic eye is in a ‘hypotropic’ position, resulting in a pseudoptosis of the paretic eye. If the patient fixates with the paretic eye, and the levator is not involved, the ptosis may disappear.⁴ A true ptosis, on the other hand, is present even when fixating with the paretic eye, and is due to a weakness of the elevator muscle.^{2,3,5,6}

Although the etiology of double elevator palsy is still not well understood, it is believed to be either supranuclear^{2,7-11} or nuclear (third nerve nucleus) in origin.^{6,12-14} Because the SR has been considered the primary elevator, it is possible that what is thought to be double elevator palsy is purely a paralysis of the SR, with a functioning IO.^{4,7,8,12,15} However, the elevation deficit may actually be due to an IR restriction, rather than a palsy of both ele-

vators.^{2,7,12,15} Indeed, some studies of saccadic velocity have demonstrated normal elevator function in patients with a previous clinical diagnosis of double elevator palsy.^{12,15} Because of this, it has been proposed that congenital double elevator palsy actually be divided into three subsets: primary SR palsy, primary IR restriction, and congenital supranuclear elevation deficiency.¹⁰

It is important to differentiate double elevator palsy from other potential causes of elevation deficits (Table 2), including mechanical and restrictive etiologies. Differential diagnoses include orbital blowout fracture,^{3-5,7,8,14-18} Brown’s syndrome,^{7,8,14,16,17} thyroid myopathy,^{4,5,7,8,15,17,18} congenital fibrosis of the inferior rectus,^{4,7,12,14,16} cellulitis,^{5,15} third nerve palsy,^{5,8,15,17,18} and strabismus surgery.^{5,15} A positive forced duction test is useful in distinguishing mechanical and restrictive causes of an elevation deficiency from a true double elevator palsy.

Patient history, and ocular signs, such as enophthalmous, exophthalmous, eyelid swelling, lid retraction, and ptosis are also important in ruling out differential diagnoses, as are x-rays when indicated.

Acquired double elevator palsy (Table 3) may present with similar characteristics as congenital cases, but must be differentiated due to potentially serious outcomes if left untreated (Table 4). The neurological structure most reportedly affected is the preteectum.^{7-9,11,19,20} However, a lesion of the pineal gland has also been reported.²¹ A patient with an acquired double elevator palsy will present with an elevation deficit of sudden onset that was not previously present, and diplopia.^{8,9,19} The elevation deficit may be an absolute or relative deficit as compared to the other eye, and other extra ocular motility deficits such as an abduction paresis may be present.^{8,20} In addition, ocular signs such as pupillary abnormalities, convergence weakness, vertical deviation in primary gaze, ptosis,

Table 1 Diagnosis of congenital double elevator palsy [(+/-): may/or may not be present]
Diagnosis of CDEP
Monocular elevation deficit
Congenital
Unilateral
Vertical deviation in primary gaze (+/-)
Anomalous head position (+/-)
Ptosis (pseudo or true) (+/-)

Table 2 Differential diagnosis of congenital double elevator palsy
Differential diagnosis
Orbital blow out fracture
Brown's syndrome
Thyroid myopathy
Inferior rectus fibrosis
Cellulitis
Strabismus surgery
Third nerve palsy

Table 3 Diagnosis of acquired double elevator palsy
Diagnosis of ADEP
Sudden onset
Diplopia
Pupillary abnormalities (+/-)
Convergence weakness (+/-)
Vertical deviation in primary gaze (+/-)
Ptosis (+/-)
Impaired OKN movements (+/-)

Table 4 Reported causes of acquired double elevator palsy
Causes of ADEP
Pineocytoma
Occlusive vascular disease
Polycythemia
Metastatic tumors
Sarcoidosis

impaired OKN movements, and nystagmus may also be present.^{8,9,11,17,19-21} Reported causes for acquired cases include occlusive vascular disease,^{8,20} polycythemia,¹⁹ metastatic tumors,¹¹ sarcoidosis,⁹ and pineocytomas.²¹ If an acquired double elevator palsy is suspected a full neurological work-up and

physical examination should be recommended.

Treatment of congenital double elevator palsy consists of surgical intervention, when warranted. Indications include vertical deviation in primary gaze, head tilt, ptosis, and poor cosmesis. A surgical procedure that has been reported to be effective in treating double elevator palsy, is the Knapp procedure.^{4,7,14,16} It involves a complete vertical transposition of the horizontal recti. Specifically, it consists of disinserting the medial and lateral rectus tendons and moving them superiorly, near the insertion of the superior rectus.¹⁶ For vertical deviations of less than 25 diopters a graded transposition, rather than a full, or complete transposition, has been recommended.⁷ In some instances a long-standing double elevator palsy, with a hypotropia in primary gaze, may lead to a contracture of the inferior rectus.¹⁸ This is confirmed by a positive forced duction test. In order for the Knapp Procedure to be effective in these cases, an initial inferior rectus recession is needed to release any restrictions.^{2,7,12,15,18} By performing the inferior rectus recession prior to the Knapp Procedure, a greater vertical correction will be obtained.¹⁸ Once surgery has been performed to attempt to vertically align the eyes, any remaining ptosis can be evaluated. Because correction of the vertical misalignment will correct pseudoptosis, the remaining ptosis is a true ptosis and can be treated by resection of the elevator.^{1,7,17,22}

Conclusion

Congenital double elevator palsy has been described. As optometrists, it is important to first differentiate from other conditions that may present with similar findings of an elevation deficiency. Then, if a double elevator palsy is suspected, one needs to determine if it is congenital or acquired. Vascular and neurological disease must be ruled out in suspected instances of acquired double elevator palsy. Treatment of congenital cases consists of surgical intervention for symptomatic presentations, i.e. hypotropia in primary gaze, head tilt, ptosis, and poor cosmesis. For patients that are asymptomatic, surgery is contraindicated, and these patients are monitored on a routine basis. However, as in the case presented, other accompanying conditions such as the accommodative insufficiency and the child's poor academic

performance should be addressed and appropriately managed.

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