Congenital Left Superior Oblique Palsy

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ABSTRACT

This was a resort of a rare case of congenital fourth nerve palsy, its clinical signs and management. A 6-year-old boy came with the chief complaint of deviation of his eyes and abnormal head position since he was born. Visual acuity are 6/6 both eyes. At primary position, vertical deviation as 15 prism diopter hypertropia was present in the left eye. Bielschowsky Three Step Test revealed left superior oblique palsy with minimal inferior oblique overaction. This patient was diagnosed with congenital fourth nerve palsy. Ipsilateral inferior oblique weakening with recession and anteriorization procedure was performed in this patient and shows satisfying results.

In conclusion: Our case represents a rare case of congenital fourth nerve palsy managed with surgical procedures.

Key words: congenital, fourth nerve palsy, ipsilateral inferior oblique weakening

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INTRODUCTION

Congenital fourth nerve palsy is a condition present at birth characterized by a vertical misalignment of the eyes due to a weakness of paralysis of the superior oblique muscle. The superior oblique muscle primarily intorts the eye, with secondary actions of depression (downgaze) and abduction (looking away from the nose). When this muscle’s function is diminished due to a fourth cranial nerve palsy, the affected eye will extort, deviate upward (hypertropia), and, to a smaller extent, drift inward.1,2,3

Congenital fourth cranial nerve palsy may not become symptomatic until later childhood or adulthood. Young children adopt a compensatory head position in order to compensate for the underacting superior oblique muscle. The usual head posture in superior oblique palsy is the head tilted to the opposite side with the chin depressed.1,3,4

Treatment is based on prism cover measurement findings, torsion, and the results of superior oblique traction testing indicating the state of the tendon. The primary goal of surgery is to eliminate the head posture and promote binocularity in the most important fields of gaze, that is, straight ahead and in down gaze or the reading position. In congenital superior oblique palsy with tendon anomaly, surgery is concentrated on the antagonist, the yoke, and the lax superior oblique tendon. The safest surgical procedure in any superior oblique palsy is weakening of the antagonist inferior oblique.5,6

The most commonly used techniques in weakening inferior oblique include myectomy, recession, and anteriorization. Myectomy weakens the inferior oblique, as removing a portion of muscle reduces the chance of local reattachment. An inferior oblique recession acts by inducing muscle slack, thus reducing muscle tension and anteriorization changes the vector of forces making the inferior oblique muscle more of a depressor rather than an elevator. Selection of the appropriate surgical procedure is based on the amount of inferior oblique overaction which is clinically estimated on a scale of +1 through +4. The advantage of the recession and anteriorization procedures is that they are very effective and produced a controlled weakening that can be tailored to the amount of inferior oblique overaction.6

CASE REPORT

A 6-year-old boy presented in outpatient eye clinic with deviation of his eyes since he was born and abnormal head position right head tilt with chin depressed. Visual acuity
revealed 6/6 in both eyes. At primary position, vertical deviation as 15 prism diopter hypertropia was present in the left eye (figure 1).

The Bielschowsky *Three Step Test* was performed to the patient. The first step revealed left hypertropia which was more prominent in cover test (figure 2). Left hypertropia was greater to the right as the second step, with left inferior oblique overaction (+1) (figure 3) and the third test revealed left hypertropia greater on tilting head to the left (figure 4). The patient was diagnosed as left superior oblique palsy.

We decided to choose surgical procedure weakening ipsilateral inferior oblique muscle. We performed inferior oblique recession and anteriorization 4 mm posterior and 2 mm lateral to inferior rectus muscle with no complication during the surgery (figure 5).

The patient was followed up day 1, 7 and a month after surgery, and in the first follow up, the patient was already in orthophoria. Visual acuity were 6/6 in both eyes. Next follow up in day 7 and a month after surgery revealed orthophoria (figure 6).

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**Figure 1.** Primary position with right head tilt and vertical deviation.

**Figure 2.** Left hypertropia more prominent with cover test. First step of Bielschowsky *Three Step Test*.

**Figure 3.** Left hypertropia was greater to the right. Second step of Bielschowsky *Three Step Test*.

**Figure 4.** Left hypertropia greater on tilting head to the left. Third step of Bielschowsky *Three Step Test*.

**Figure 6.** Day 1 after surgery with orthophoria.
The two causes of fourth nerve palsy are congenital and acquired. The most common type of the disorder is congenital. Acquired cases most commonly result from closed head trauma. Patients with a congenital trochlear nerve palsy do not have associated pathology and these patients are usually not “worked up” with neuro-imaging.

In this patient, we found abnormal head posture to the right shoulder with a little bit chin depressed. Bielschowsky Three Step Test used to determine which muscle was involved. From the first step, cover test has determined that a left hypertropia was present. This reduced the diagnostic possibilities to a paralysis of the right superior rectus or left superior oblique muscles. The third step revealed that left hypertropia greater on tilting head to the left which in superior oblique palsy, hypertropia increased on tilting head toward paralyzed side.

The superior oblique muscle is innervated by the trochlear nerve and it has three functions. These functions are dependent upon the gaze position that the eyes are in. The primary function of the superior oblique muscle is to depress the globe when the eye is adducted (this is important for reading). The second function is excycloversion of the globe (this helps to compensate for head tilting). The third function is abduction of the eye in down gaze.

The antagonist to the superior oblique muscle is the inferior oblique muscle. This muscle has three functions which are also field of gaze dependant. The primary function of the inferior oblique is to elevate the globe when it is in an adducted position. The second function is excycloversion and the third function is to abduct the globe when the eyes are in an upgaze position. In this patient we found left inferior oblique overaction (+1).

Based on the Knapp type, this patient was Knapp I, which overaction of antagonist inferior oblique was found with deviation about 20 prism diopters or less in the field of action of the antagonist. The purpose of the surgery was to weaken antagonist inferior oblique. In this patient, based on prism test and the amount of inferior oblique overaction, we performed inferior oblique recession and anteriorization 4 mm posterior and 2 mm lateral to inferior rectus muscle and the result was orthophoria.

CONCLUSION

This case represents a rare case of congenital fourth nerve palsy with minimal antagonist inferior oblique overaction managed with ipsilateral inferior oblique recession and anteriorization procedures, shows satisfying results.

REFERENCES