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Late spontaneous resolution of congenital Brown syndrome

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Abstract

Brown syndrome is characterized by restricted elevation in adduction. Congenital Brown syndrome is usually diagnosed during early childhood. It is believed to be due to an abnormality of the superior oblique tendon as it passes through the trochlea. The natural history of Brown syndrome is poorly characterized. Many patients with congenital Brown syndrome undergo strabismus surgery during childhood in an attempt to correct the problem surgically. This report describes spontaneous regression of congenital Brown syndrome in an adult.

Case Report

A 2 1/2-year-old white girl presented to the author at the Emory Eye Center with a history of exotropia and left hypertropia noted in upgaze when she looked to the left. The findings were first noted when she was 6 months of age. The parents reported the findings were more pronounced when she was tired or ill. The patient's medical history was otherwise unremarkable.

Visual acuity was 20/30 in both eyes. Cycloplegic refraction was +1.50 D in both eyes. Ocular motility testing was notable for –3 restriction of elevation of the right eye in adduction. In primary position she was orthotropic, but she had a 10^A exotropia in upgaze and a large right hypotropia in upgaze to the left. There was no torticollis. She was diagnosed as having congenital Brown syndrome and the decision was made to follow her longitudinally without surgical intervention.

When 3 years old, she developed sinusitis. Computed tomography of the orbits revealed mucosal thickening of the ethmoidal air cells and normal superior oblique muscles and tendons in both eyes. When 4 years old, she had –4 restriction of elevation of the right eye in adduction, but she was still orthotropic in primary position. She had 120 arcsec of stereopsis using the TNO test (Lameris, Utrecht, Netherlands). When 5 years of age, elevation of the right eye in direct upgaze was also limited (Figure 1). Elevation in left upgaze continued to be severely restricted, while elevation in right upgaze was normal. She reported diplopia in upgaze, but she used a chin-up head posture to compensate. She was examined annually until 12 years of age with no change in her ocular motility. She played multiple sports and excelled academically in school. She next returned for follow-up in December 2009, at 22 years of age. At that time, she reported that she had noted gradual improvement in her diplopia in upgaze. She denied hearing an audible click or having pain in upgaze. Visual

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acuity was 20/20 in both eyes. Ocular motility was normal (Figure 2). However, on direct upgaze, she had a 1^Δ right hypotropia and on left upgaze she had a 2^Δ right hypotropia. In all other positions of gaze, she was orthotropic. She had 30 arcsec of stereopsis using the TNO test. She was diagnosed with migraine headaches when 20 years of age. A neurological evaluation, including magnetic resonance imaging of the brain at that time was unremarkable.

Discussion

Spontaneous resolution of congenital Brown syndrome has been reported; however, the incidence of this phenomenon is not known. In 1958 Costenbader¹ reported on the spontaneous resolution of Brown syndrome in a 6 1/2-year-old boy after a 3 year follow-up. In 1959 Adler² also reported on the spontaneous resolution of Brown syndrome in a 7-year-old boy after a 3-year follow-up. In 1986 Leone and Leone³ reported on the spontaneous resolution of Brown syndrome in a 21-year-old man that he attributed to daily eye exercise from age 16 to 18 years and that was also associated with an audible click as it resolved. The authors postulated that these exercises may have loosened a fibrous obstruction interfering with the function of the superior oblique tendon sheath or the trochlea. Capasso and colleagues⁴ also noted the resolution of Brown syndrome in one eye of a 4-year-old girl with bilateral Brown syndrome after a follow-up of 7 months. Kaban⁵ reported that ocular motility normalized without surgical intervention in 5 of 45 (11%) patients with congenital Brown syndrome who were followed for a mean of 4.6 years. Only two patients in their series experienced a worsening of their motility during the follow-up period. Gregersen and Rindziunski⁶ reported spontaneous resolution in 3 of 10 (30%) patients and improvement in 9 of 10 (90%) patients with congenital Brown syndrome after a mean follow-up of 13 years. Finally, Dawson and colleagues⁷ noted improvement in 24 of 27 (89%) patients with congenital Brown syndrome after follow-up ranging from 1 to 10 years.

The indications for surgical correction of Brown syndrome have been reported to be strabismus in primary position, an anomalous head position or a cosmetically unacceptable downshoot of the eye in adduction.⁸ It has been suggested that observation alone may adversely affect binocular function.⁹ A variety of surgical corrections of Brown syndrome have been described including a superior oblique tenotomy, a combined superior oblique tenotomy and inferior oblique recession, and lengthening of the superior oblique tendon using a silicone band or a nonabsorbable suture.^{10,11} These operations can result in a number of complications including iatrogenic superior oblique palsy and restricted motility secondary to scarring of a silicone expander.^{11,12}

The natural history of congenital Brown syndrome is poorly understood. It seems likely that a significant percentage of patients with Brown syndrome will have spontaneous improvement or resolution of their motility disturbance without surgical intervention. Most longitudinal series of patients with congenital Brown syndrome have relatively short-term follow-ups and as a result likely underestimate the true incidence of spontaneous resolution. While spontaneous resolution has been reported to occur in as little as 7 months, in most cases it occurs years later. Brown syndrome almost completely resolved in our patient at some point 11 to 20 years after she was initially examined. The patient described it as being a gradual process, and it is possible that further improvement may occur with a longer follow-up. Unlike the patient described by Leone and Leone,³ our patient did not hear an audible click or experience pain as her motility improved. Interestingly, there was no improvement in her motility at age 13 years, whereas her ocular motility was almost completely normal at age 22 years. In addition to nearly complete resolution of her motility disturbance, our patient maintained high-grade stereopsis. While it is not known what her outcome would have been with surgical intervention during childhood, it seems unlikely that

it would have been better than that which occurred with observation alone. The possibility of spontaneous improvement or resolution of congenital Brown syndrome after long-term follow-up should be seriously considered prior to recommending surgery.

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FIG 1.

Motility at age 5 years showing restricted elevation of the right eye in adduction. In addition, there is restricted elevation of the right eye in direct upgaze. Elevation of the right eye in right gaze is normal.

FIG 2.

Motility at age 22 years showing normal versions in all positions of gaze. The patient was orthotropic except for a 1^{Δ} right hypotropia in direct upgaze and a 2^{Δ} right hypotropia in left upgaze.