

Management of a case of divergent strabismus fixus secondary to a congenital fibrosis of extraocular muscles type 2

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A 17-year-old boy presented with a large exotropia with both eyes fixed in an abduction and upgaze, pupillary involvement since childhood. He had mild optic nerve hypoplasia in the right eye and situs inversus of the retinal vessels in the left optic disc. His ocular motility showed restriction of eye movements in all gazes. He was diagnosed with congenital fibrosis of extraocular muscles, type 2 (CFEOM2) and operated upon in a staged procedure with a satisfactory eye alignment using hang-back sutures in one eye and periosteal fixation in the other. This report highlights the surgical course and final outcome in this case of CFEOM2.

Key words: Congenital fibrosis of extraocular muscles, congenital fibrosis of extraocular muscles type 2, divergent strabismus fixus, exotropic strabismus fixus

The extraocular fibrosis of muscles syndromes are rare, congenital ocular motility disorders that arise from dysfunction

of the oculomotor, trochlear, and abducens nerves and/or the muscles that they innervate.^[1] Four clinical phenotypic forms of familial congenital fibrosis of extraocular muscle (CFEOM) have been identified.^[2] CFEOM type 2 (CFEOM2) has been described in literature with bilateral ptosis, exotropia, severe restriction of horizontal and vertical eye movements with variable abduction being present, pupillary abnormalities with miotic, poorly reactive pupils, and a positive forced duction test.^[3] Its surgical management is unsatisfactory with few cases reported in literature. We report a case of CFEOM2 that was satisfactorily managed in a staged manner.

Case Report

A 17-year-old male presented with a history of outward deviation of both eyes since childhood. He was the second child born of a nonconsanguineous marriage, with no family history of similar eye disease. He came with the globes “frozen” in extreme abduction with the inability to adduct and depress both eyes [Fig. 1a]. On examination, he maintained chin depression with a slight right face turn while fixing with his left eye [Fig. 2a] and a chin depression of more than 50° with a large left face turn while fixing with his right eye [Fig. 2b]. Cycloplegic refraction was $-2.0/-1.50 \times 90^\circ$ in the right eye and $-2.25/-2.0 \times 100^\circ$ in the left with a best-corrected visual acuity for distance of 20/80 and 20/40, respectively. Extraocular movements were severely limited in all gazes in both eyes except for the abduction of -2 in the left eye. Anterior segment was normal in both eyes. Pupillary examination showed a 5 mm fixed pupil, not reacting to light, and near reflex in the right eye and 2 mm size pupil, with normal reaction to light and near reflex in the left eye. Fundus examination revealed myelinated nerve fibers with a mildly hypoplastic disc in the right eye and situs inversus

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Figure 1: Clinical photograph of the eyes; (a) before surgery showing a large exotropia with hypertropia of both eyes in primary position, (b) 5 months postoperative follow-up showing recurrence of the large exotropia and hypertropia in the right eye, (c) final postoperative alignment in the primary position with a small residual exotropia in the right eye measuring 15° with corrected head posture

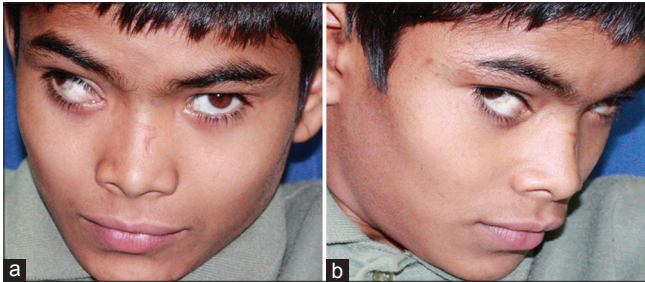


Figure 2: Preoperative clinical photograph showing the head posture; (a) a chin depression adopted when fixing with the left eye. (b) a chin depression with a large left face turn adopted when fixing with the right eye



Figure 3: Magnetic resonance imaging orbit; (a) both the globes in exotropia, (b) the displacement of the extraocular muscles. SR: superior rectus muscle, MR: medial rectus muscle, IR: inferior rectus muscle

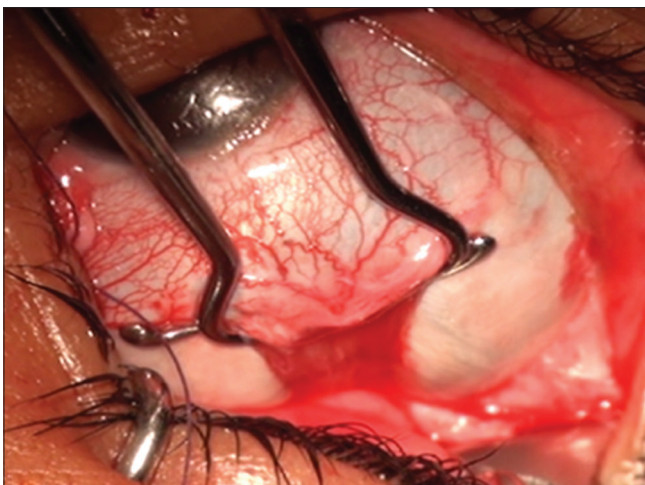


Figure 4: Intraoperative photograph of the right eye showing the tight lateral rectus muscle indenting the globe

of the retinal vessels of the left optic disc. The rest of the fundus was within normal limits. Systemic examination was normal. Magnetic resonance imaging (MRI) of the brain was normal with normal cranial nerve nuclei. MRI of both orbits showed nasal displacement of superior and inferior

recti [Fig. 3a and b] and superior displacement of the lateral rectus muscle, which was confirmed intraoperatively. Forced duction test of the extraocular muscles revealed tight lateral rectus and superior rectus muscles in both eyes with indentation of the globe, seen intra-operatively [Fig. 4]. The patient underwent sequential surgeries, a 10 mm recession of lateral rectus with transposition of the muscle inferiorly adjacent to inferior rectus using hang-back technique and 8 mm recession of superior rectus with nasal transposition adjacent to medial rectus using hang-back technique was performed in both eyes, using the limbal approach. On the 1st postoperative day, a residual exotropia of 7° and hypertropia of 7° was present in the right eye with the left eye fixing in primary position with a small right face turn. Over the next 5 months, the left eye remained aligned but the exotropia and hypertropia in the right eye increased to the preoperative position [Fig. 1b] and hence a second surgery was advised. Since the procedure of recession of the lateral and superior recti failed, a more permanent technique of periosteal fixation was planned to align the eyes in primary position in view of the CFEOM. Here, the lateral rectus and superior rectus were disinserted, tied together, and attached to the lateral orbital wall periosteum with 4-0 Prolene sutures.

A month later, there was an improvement in his right eye position [Fig. 1c] with best-corrected visual acuity of 20/70 in the right eye and 20/40 in the left eye. At the last follow-up, 6 months later, he had an exotropia of 15° in the right eye with left eye fixing with a small face turn suggesting a significant reduction in the deviation and a cosmetically acceptable alignment.

Discussion

CFEOMs is amongst the uncommon, nonprogressive, restrictive strabismic disorders^[3] included in the category of congenital cranial dysinnervation disorders.^[4] CFEOM2 is predicted to result from a defect in the development of both its superior and inferior branches of the third cranial nerve.^[1] The clinical presentation depends on the number of affected muscles and the degree of fibrosis. Ptosis with various degrees of severity is a common but not a constant feature. Patients are usually healthy, with few of them having ocular and systemic features such as optic nerve hypoplasia, globe retraction, microphthalmia, enophthalmos, oculocutaneous albinism, Marcus Gunn's jaw-winking phenomenon, ethmoidal mesenchymoma, and cerebral, cortical, and basal ganglia maldevelopment. In the involved eye, the predominantly affected muscle behaves like a fibrous band causing a loss of elasticity for the rectus muscle.^[5] Our patient had a hypoplastic right optic nerve.

A few reports describe surgical management of divergent strabismus fixus. Furthermore, one cannot plan according to the standard surgical tables, as the surgical anatomy is altered. Daniell *et al.*^[6] has described supramaximal horizontal recess-resect procedure with adducting traction sutures left

in situ for 6 weeks to prevent the eye from getting abducted and thus reducing the chance of a postoperative exotropic drift.

Hang-back sutures provide a beneficial option instead of placing sutures way back on a frozen globe but have increased chances of postoperative recurrences. Apt and Axelrod^[7] have also demonstrated that exploration and re-recession were needed for hang-back suture recessions with an eventual good outcome. Free disinsertion described by Sener *et al.* or recession of a lateral rectus, in contrast, has not been as powerful as suturing it to the lateral orbital periosteum in decreasing the exotropia in CFEOM.^[8] Merely cutting the muscle free of its attachment to the globe is often unpredictable. The muscle may reattach to the globe and continue to exert some force on the globe.^[9] In our case also, we observed that a simple hang-back recession with transposition of the superior and lateral rectus worked in the left eye, but the right eye needed lateral rectus and superior rectus muscle disinsertion with fixation on the lateral orbital wall.

Conclusion

Surgery in a case of CFEOM2 requires an individualized approach. Satisfactory outcomes are possible in significantly reducing the head posture and obtaining cosmetically acceptable alignment of the eye in the primary position with systematic planning and staged procedures as required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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