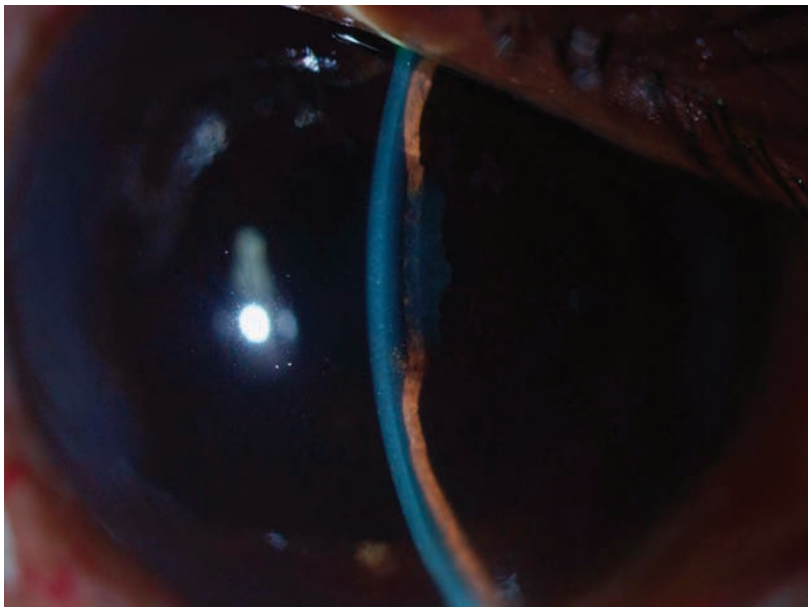

Figure 1 This slit-lamp photograph demonstrates very shallow anterior chamber, small pupil, and seclusio pupillae encountered before vitrectomy.

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Sir,

Large-angle congenital exotropia due to 'absent' medial recti: a case of congenital cranial dysinnervation disorder

Congenital cranial dysinnervation disorders (CCDD) are a heterogeneous group of disorders affecting the development and function of the extraocular muscles due to failure of normal innervation of the target muscle. The surgical management of these disorders can be complex requiring combinations of supramaximal recessions, muscle transpositions, and botulinum toxin (BT). We report an unusual case of large-angle exotropia due to absent medial recti, the subsequent surgical management, and outcome.

Case report

A 3-month-old Caucasian girl of non-consanguineous parents presented with a congenital alternating exotropia measuring 70 prism dioptres (PD) (Figure 1a). Neither eye adducted beyond the midline; other ductions were full. No ptosis, fissure changes, or synergistic divergence were observed. Apart from talipes equinovarus, there were no medical or developmental problems. A magnetic resonance imaging (MRI) scan (Figure 1c) appeared normal apart from somewhat thin medial rectus muscles. Bilateral lateral rectus BT injections reduced the exotropia to 50 PD. Adduction improved in the left eye only. At a planned bilateral maximal recession/resection procedure, only connective tissue was identified medially after recession of the left lateral rectus (Figure 1b). This eye was then fixated by traction sutures. A similar situation was encountered in the right eye; here a full tendon vertical recti transposition was combined with a BT injection to the lateral rectus. After 3 years (and two repeat BT injections), there is an alternating exotropia of 10 PD for near and 14 PD for distance. Adduction limitation is −3 in the right and −1.5 in the left eye.

Comment

Agenesis or hypoplasia of individual or combinations of extraocular muscles has been reported since the eighteenth century.[1] The extraocular muscles derive from superior and three inferior complexes of orbital

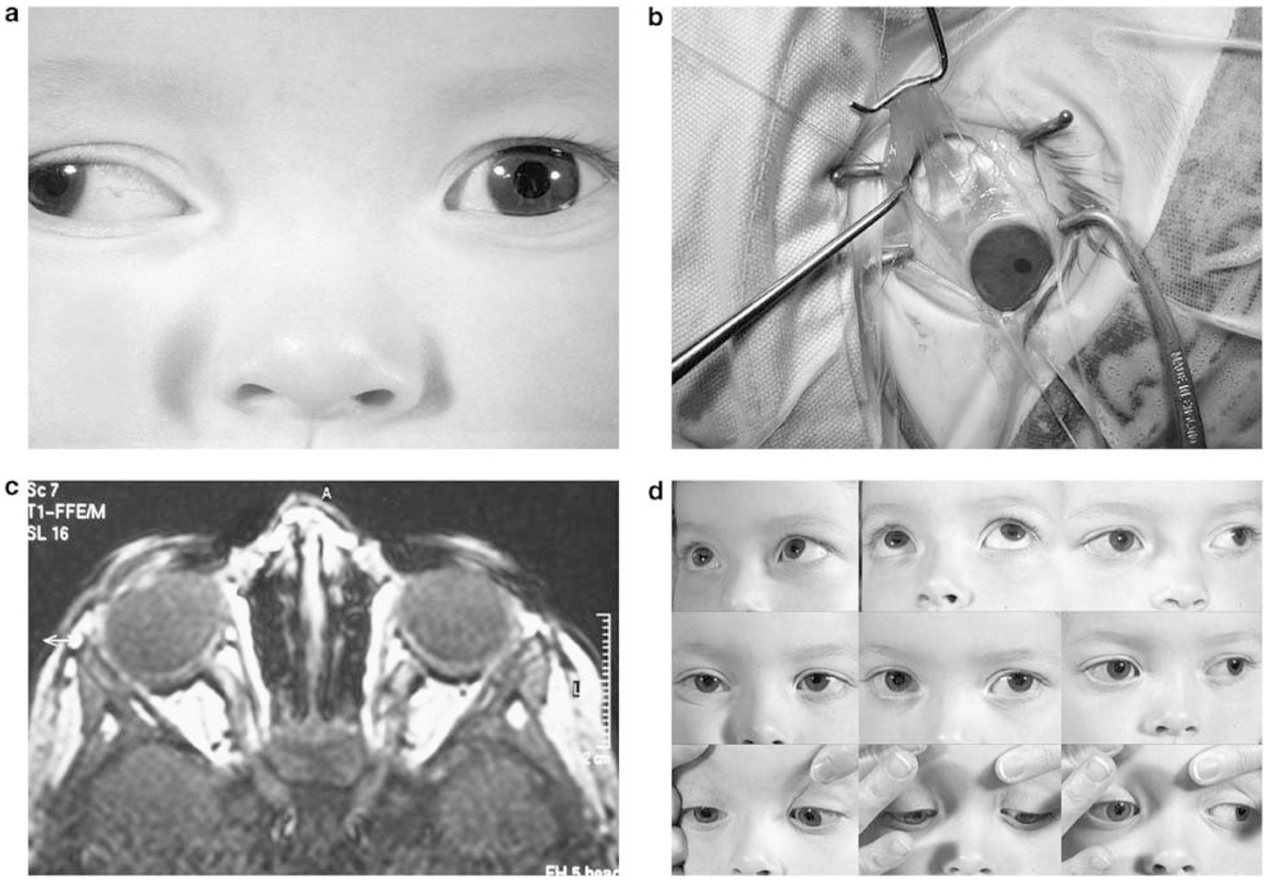

Figure 1 (a) Large-angle exotropia at presentation, (b) intraoperative view of left medial rectus muscle, (c) MRI showing thin medial rectus muscles, (d) 9-gaze template 2 months after strabismus surgery, showing improved adduction of left eye and residual effect of botulinum toxin in right lateral rectus muscle.

mesodermal tissue. The ocular motor nerves originate from mesencephalic and rhombencephalic structures and migrate to their target muscles. Cranial nerve motoneuron aplasia or failed extraocular muscle innervation may cause aberrant ocular motor patterns, ptosis, and secondary structural muscular abnormalities. The aetiology is frequently genetic.[2] These non-progressive developmental disorders are known as CCDD, and among them are Duane retraction syndrome, congenital fibrosis of extraocular muscles, and Möbius syndrome.[2,3] There are numerous associated non-ocular defects, and clinical heterogeneity is common. MRI may show affected muscles and cranial nerves to be normal, hypoplastic, or absent.[4] We believe this case to be part of the CCDD spectrum. Ideally, in this case, the MRI should have shown more detail. We would advocate high-resolution MRI before surgery for large-angle congenital exotropia with adduction deficits to facilitate diagnosis and surgical plan.

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