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**INCOMITANT STRABISMUS UPDATE**

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**Orbital causes of incomitant strabismus**

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USA**Abstract**

Strabismus may result from abnormal innervation, structure, or function of the extraocular muscles. Abnormalities of the orbital bones or masses within the orbit may also cause strabismus due to indirect effects on the extraocular muscles. This paper reviews some disorders of the orbit that are associated with strabismus, including craniofacial malformations, orbital masses, trauma, and anomalous orbital structures.

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Available from: <http://www.meajo.org/text.asp?2015/22/3/286/159714>**Full Text****Introduction**

Most strabismus results from abnormal innervation of the extraocular muscles. The muscles themselves are usually structurally normal. The abnormal eye alignment is caused by aberrant signals from the brain that cause excessive movement of the muscles in a certain direction, such as the disproportionate contraction of the medial rectus muscles in patients with infantile esotropia. In addition, the orbital structures that surround the globe are normal in the majority of patients with strabismus. The anatomy of the orbit continues to be elucidated [1] (see article on the pulley system elsewhere in this volume), but for most patients the orbital structures are not important considerations when making treatment decisions.

Less commonly, strabismus results from anatomic abnormalities of the orbit. In these disorders, the muscles are usually normal, but act abnormally due to contiguous masses or other structural anomalies. This article will review some of these orbital causes of incomitant strabismus.

**Craniofacial Anomalies**

Craniofacial anomalies in children arise from mal development of the bones that form the skull. Many of these disorders involve the orbit. In these patients, the abnormal orbital anatomy affects the function of the extraocular muscles, usually by either changing the position of the muscle insertion or by an alteration of the muscle pathway [Figure 1] and [Figure 2]. In some cases, the muscles themselves may be absent. (Figure 1) (Figure 2)

During skull development, bone is laid down along suture lines. If these sutures close prematurely, the bony skull cannot form properly, typically causing overgrowth parallel to the suture lines and undergrowth perpendicular to the lines. The most severe malformations are seen in a group of disorders called the craniosynostosis syndromes. Most of these are inherited in an autosomal dominant fashion, involve fusion of multiple sutures, and frequently have associated limb anomalies. The most common craniosynostosis syndromes are Crouzon, Apert, Pfeiffer, and Chaethre-Chotzen syndromes. Ocular problems in addition to strabismus in these disorders include globe protrusion, corneal exposure, and optic nerve compression.

Strabismus in patients with craniosynostosis syndromes is common, with reported incidences ranging from 40% to 90%. [2],[3] Strabismus can result from a variety of causes. Because the orbit is often hypoplastic, the muscle pathways may be abnormal, either by shortening of the muscles or torsion of the globe creating eccentric force vectors. Posterior displacement of the trochlea due to shallow orbits has been postulated as an etiology for superior oblique underaction. Absence of the extraocular muscles has been reported in many affected patients. [4],[5]

One of the most common types of ocular misalignment in patients with craniofacial disorders is V-pattern strabismus. This is characterized by apparent inferior oblique overaction (overelevation in abduction). A feature that is more prominent in affected patients with craniofacial disorders compared to other patients with V-pattern strabismus is a frequently striking overdepression in abduction. Understanding the etiology of this form of strabismus is important in determining a treatment plan. Although the inferior oblique muscles appear to be overacting, weakening procedures of these muscles is often not effective in resolving the pattern. [6] This is likely because the apparent overaction results from the abnormal anatomy of the globe. Tan et al. reported a series of patients with craniosynostosis in whom orbital imaging was performed. [7] They found a significant association between V-pattern strabismus and excyclorotation of the muscle cone due to orbital malformations. It is postulated that the abnormal vertical movements result from the effects of Hering's law on the rotated globe. This theory has been supported by the results of mechanical modeling of such patients. [8] Apparent inferior oblique overaction may also be caused by absence of the superior oblique tendon. [6]

Surgical treatment of strabismus in patients with craniofacial disorders depends on the etiology of the ocular misalignment. Tan et al. recommend routine imaging to assist in surgical planning. [7] If the muscles are absent or malformed, balancing of the deficits by weakening the yoke muscle in the contralateral eye may be considered. The effects of transposition surgery are difficult to predict in these patients because of the rotated muscle cones. For patients with V-pattern strabismus and excyclorotated orbits, superior transposition of the inferiorly displaced lateral rectus muscles is usually more effective than inferior oblique muscle weakening procedures, although both procedures may be necessary to effect improvement in some patients.

Regarding the timing of surgery, there have contradictory reports as to whether strabismus surgery should be postponed until craniofacial reconstruction is completed. Most reconstructive procedures involve the displacement of the orbit, which potentially could affect ocular motility. However, Diamond and Whitaker reported ocular motility findings before and after craniofacial surgery in 140 patients. Only four patients had an increase in the magnitude of preexisting strabismus by >10 prism diopters, and only two had new strabismus in primary position (caused by cranial nerve palsies). [9] On the other hand, Morax reported that eight of nine patients with Crouzon syndrome had improvement in horizontal strabismus following orbital reconstruction. [10] An important factor to consider in these patients is the potentially deleterious effect of delaying surgery on the development of binocular vision, which lends support to the early surgical repair.

### Orbital Masses

Any mass in the orbit that displaces the extraocular muscles may cause strabismus. A wide variety of disorders cause orbital masses, including neoplasms, congenital or acquired vascular or cystic lesions, infections, and inflammatory lesions. The initial presenting signs and symptoms of these masses depend on the location, size, and growth of the underlying lesion. [11] If the lesions are large, proptosis (or another globe displacement) and secondary eyelid changes are usually the most prominent features. Inflammatory and infectious lesions usually present with pain and periocular inflammation. Many patients, particularly those with inflammatory lesions, also have systemic symptoms. Smaller lesions, particularly if they are adjacent to an extraocular muscle, may initially present with strabismus. [12]

Depending on the etiology of the mass, orbital lesions may potentially be life threatening (e.g., neoplasms) or vision threatening. Management of the strabismus is usually a secondary consideration. Motility disorders often improve with successful treatment of the underlying orbital mass, as the following case illustrates.

#### Case Report

A previously healthy 13-year-old male presented with progressive pain and swelling of the right eye. Examination revealed marked periocular edema and inflammation on the right [Figure 3]. Visual acuity was 20/20 in both eyes. Motility examination revealed orthophoria in primary gaze, but motility was restricted vertically, with a right hypotropia in upgaze and a right hypertropia in downgaze [Figure 4], [Figure 5], and [Figure 6]. Computed tomography revealed orbital cellulitis and a subperiosteal abscess on the right [Figure 7]. Blood cultures grew *Peptostreptococcus*. The patient was treated with intravenous antibiotics, and his abscess was surgically drained. On follow-up examination 2 weeks later his vision remained 20/20 in both eyes, and his motility limitation had almost entirely resolved. [Figure 3]{Figure 4}{Figure 5}{Figure 6}{Figure 7}

### Orbital Trauma

Strabismus may result from trauma for a wide variety of reasons. Intracranial injury may cause cranial nerve palsies affecting the third, fourth, or sixth cranial nerves. Direct or indirect orbital trauma may cause intraorbital hemorrhage with a displacement of the muscles. Injury to the extraocular muscles themselves may occur, usually due to a penetrating injury or hemorrhage within the muscle.

The most common form of strabismus related to orbital injury results from fractures of the bony orbit secondary to blunt trauma. The history and associated findings make the diagnosis obvious in most cases, although the history may sometimes be withheld due to patients' fears of repercussions. Rarely, occult fractures are found on imaging studies of patients with complex strabismus. [13] The type of strabismus is related to the location of the fracture and the extent of extraocular muscle involvement.

The orbital floor is the most frequent location of fractures that cause strabismus [Figure 8] and [Figure 9]. The inferior rectus muscle may become entrapped within the fracture, causing mechanical restriction. Depending on the nature of the injury, hemorrhage or edema of the muscle may also contribute to a motility disorder. Patients typically manifest limited ability to elevate the affected eye. In some cases, depression may also be limited by restriction or muscle dysfunction. Attempted upgaze may elicit the oculocardiac reflex, with bradycardia and syncope. [Figure 8]{Figure 9}

Two types of strabismus related to orbital trauma merit special consideration. The first is the white-eyed blowout fracture. In this type of injury, which usually occurs in children, the inferior rectus muscle becomes entrapped in a fracture of the orbital floor, usually either a trapdoor fracture or a linear nondisplaced fracture. [14] Patients often have very few external signs of trauma, and the entrapment may not be recognized on imaging studies. Motility evaluation reveals limited elevation. In many patients, depression is also limited. Early surgical repair with the release of the restricted muscle is indicated to prevent permanent ischemic muscle damage.

The second type of strabismus related to orbital trauma that merits special consideration is a flap tear of an extraocular muscle. In this condition, orbital trauma causes partial avulsion of the orbital portion of the extraocular muscle or partial disinsertion of the tendon from the sclera. [15] Scar tissue forms between the avulsed portion of the muscle and surrounding orbital tissue, creating restriction. Similar to orbital fractures, the medial, and inferior rectus muscles are most commonly involved. The most frequent presentation is a limitation of movement in the direction of the avulsed muscle, which may be mistaken for muscle palsy. Restriction in the direction away from the affected muscle may also occur, creating an appearance of entrapment. Ludwig reported generally good results with surgery in which the muscle is carefully explored, the avulsed muscle is dissected from its adhesions to orbital tissue and reattached to the globe, and the capsule is repaired. [15] The best results were found in patients in whom the repair was performed early or who had simultaneous repair of the flap tear and orbital fracture.

### Anomalous Orbital Structures

Anomalous extraocular structures are an unusual and interesting cause of incomitant strabismus. These are structures within the orbit that attach to the globe, and which may cause primarily restrictive strabismus. [16] It has been postulated that some of these structures represent an atavistic remnant of the retractor bulbi muscle, a structure found in lower animals that originates in the muscle cone and attaches to the posterior surface of the globe. [17] Contraction of the retractor bulbi muscle creates posterior movement of the eye, which may help protect the globe.

Three types of anomalous extraocular structures have been described. The first type is those that arise from the extraocular muscles themselves, but which insert in an abnormal location. These are likely more common than is usually recognized, as small slips of muscle tissue are sometimes seen when muscles are disinserted during strabismus surgery. If these structures are close to the insertion, they may be incorporated within a muscle hook when the muscle is isolated and disinserted concurrently, and therefore not recognized. Such minor variations probably are not important in the etiology of strabismus. Some of these structures, however, clearly cause strabismus. Miner and Jampolsky described such a patient with a large-angle restrictive hypotropia. [18] During surgery, forced ductions remained positive after disinsertion of the inferior rectus muscle. Upon exploration, an accessory muscle was discovered arising from the undersurface of the inferior rectus muscle, and the restriction was freed after disinsertion of the accessory muscle.

The second type of anomalous orbital structure is a fibrotic band of tissue found beneath the extraocular muscles, but which is distinct from the muscles themselves. Such structures have been reported in patients with unusual motility patterns including large-angle vertical and horizontal deviations, limited extraocular movements, globe retraction, and enophthalmos. [19],[20],[21] There is little information in the literature regarding surgical attempts to treat patients with these fibrotic bands.

The third type of anomalous orbital structure is a separate muscle that originates in the posterior orbit and inserts in an abnormal location. Types of these structures include single discrete structures that insert on the globe in abnormal locations, [22],[23],[24],[25],[26] or structures that branch and insert on multiple locations on the normal extraocular muscles. [17],[27] The first of these, single discrete structures, has been more commonly reported. Imaging studies have demonstrated such findings in patients with vertical strabismus. [24],[25],[26]

When associated with strabismus, anomalous orbital structures may produce unusual motility problems. Three clinical signs suggest the presence of anomalous orbital structures. Globe retraction in adduction is common in patients with Duane retraction syndrome. Globe retraction in other fields of gaze is unusual, and is the first abnormality that suggests an anomalous orbital structure may be present. Two patients reported by Mühlenyck with fibrotic bands presented with retraction in upgaze and abduction. [21] The second clinical sign is very large angle vertical strabismus, either hypertropia or hypotropia. [18],[20] The third pattern of abnormal motility is an elevation deficit that is worse in abduction (rather than that seen in Brown syndrome, which is worse in adduction, or monocular elevation deficiency, which causes limitation of elevation in both abduction and adduction). [24],[26] If any of these unusual patterns of strabismus is found, orbital imaging should be considered to evaluate for the presence of an anomalous orbital structure. Because these are so unusual, communication with the radiologist is recommended, as such structures may be overlooked or misidentified. [26]

Not all anomalous structures produce strabismus. Those that do almost always cause restriction. If surgical treatment is indicated, the presence of restriction can be verified by intraoperative forced ductions. A finding that may indicate the presence of such structures is the persistence of restriction after the normal extraocular muscle is released from the globe. [18] Disinsertion of the anomalous structure, with or without recession of the adjacent extraocular muscle usually (but not always) improves the strabismus. [18],[20],[21],[23],[26]

#### Case Report

An otherwise healthy 4-month-old female presented for evaluation of progressive left esotropia. She had a limitation of abduction on the left and lid fissure narrowing in right gaze with a presumed diagnosis of Duane retraction syndrome. By 8 months of age, she had developed a 25° left face turn and esotropia in primary gaze [Figure 9] and [Figure 10]. A left medial rectus muscle recession was planned. At the time of surgery, significant restriction in abduction was present in the left eye. The normal-appearing left medial rectus muscle was sutured and disinserted [Figure 11]. As the posterior globe was examined for placement of sutures, an abnormal muscular structure was noted inferiorly and posterior to the medial rectus muscle insertion site [Figure 12]. The structure was isolated and was found to be an accessory muscle distinct from the medial rectus muscle [Figure 13] and [Figure 14]. It was approximately half the size of the medial rectus muscle but otherwise appeared normal. The accessory muscle was dissected and clamped 10 mm posterior to its insertion. It was disinserted from the globe, and the muscle anterior to the hemostat was resected. The stump was cauterized and allowed to retract. The left medial rectus muscle was then recessed 4 mm. Pathology of the accessory structure was consistent with an extraocular muscle. Postoperatively the patient improved, but still had moderately limited abduction of the left eye. She was orthophoric in primary gaze. Her head position resolved and vision was normal in both eyes. The contribution of the accessory muscle to the original motility disorder could not be definitively determined, but it is unlikely that the patient would have improved without recognizing and removing the muscle. [Figure 10]{Figure 11}{Figure 12}{Figure 13}{Figure 14}

#### Summary

Orbital abnormalities account for a small proportion of patients with strabismus, but it is important to recognize these disorders for several reasons. First, they may have important systemic and visual consequences, particularly those disorders caused by orbital masses. Second, unlike most common forms of strabismus, imaging studies are frequently indicated in patients in whom orbital abnormalities cause strabismus. Third, recognition of the underlying disorders is important in determining a surgical plan. In some patients, such as those with orbital lesions, treatment of the underlying problem may improve the strabismus. In others, such as craniosynostosis or orbital trauma, determining the location and pathways of the muscles is necessary to effectively address the strabismus. Finally, anomalous structures are sometimes found in patients with unusual patterns of strabismus, and appropriate treatment depends on identification of these structures.

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