

Complex Strabismus & Syndromes

Some patients exhibit complex combinations of vertical, horizontal, and torsional strabismus. Dr. Shin treats patients with complex strabismus arising from, but not limited to, thyroid-related eye disease, stroke, or brain tumors as well as strabismic disorders following severe orbital and head trauma. The following paragraphs describe specific ocular conditions marked by complex strabismus.

Duane Syndrome

Duane syndrome represents a constellation of eye findings present at birth that results from an absent 6th cranial nerve nucleus and an aberrant branch of the 3rd cranial nerve that innervates the lateral rectus muscle.

Duane syndrome most commonly affects the left eye of otherwise healthy females. Duane syndrome includes several variants of eye movement abnormalities. In the most common variant, Type I, the eye is unable to turn outward to varying degrees from the normal straight ahead position. In addition, when the patient tries to look straight ahead, the eyes may cross. This may lead a person with Duane syndrome to turn his/her head toward one side while viewing objects in front of them in order to better align the eyes. When the involved eye moves toward the nose, the eye retracts slightly back into the eye socket causing a narrowing of the opening between the eyelids. In Type II, the affected eye possesses limited ability to turn inward and is generally outwardly turning. In Type III, the eye has limited inward and outward movement. All three types are characterized by anomalous co-contraction of the medial and lateral rectus muscles, so when the involved eye moves towards the nose, the globe pulls back into the orbit and the vertical space between the eyelids narrows. The eye may also drift up (upshoot) or down (downshoot) as it moves toward the nose.

Although restoration of full eye movements in patients with Duane syndrome may not be possible, eye muscle surgery can effectively correct eye misalignment in primary (straight ahead) gaze, eliminate an abnormal head position, and expand one's field of vision, especially if performed early in the patient's life. If the abnormal up and down movements of the eye or the narrowness of the eyelid opening significantly affects eye appearance, surgery may also be of benefit.



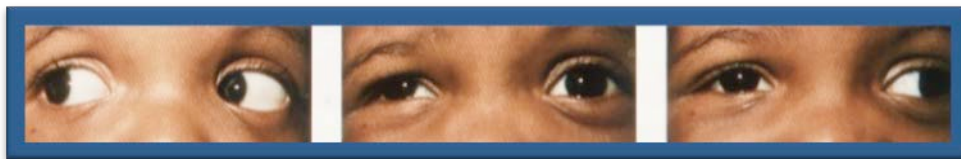
Duane syndrome, Type I, Left Eye

Left eye has difficulty turning outward and pulls back when the patient looks to the right.



Duane Syndrome, Type I, Left Eye-Limitation to Turn Left Eye Outward

When the patient tries to look to the right, his left medial rectus (inside) muscle and his left lateral rectus (outside) muscle co-contract and cause retraction of the left globe into the eye socket, narrowing of the opening between the eyelids, and upshoot (top left photo) or downshoot (top right photo) of his left eye.

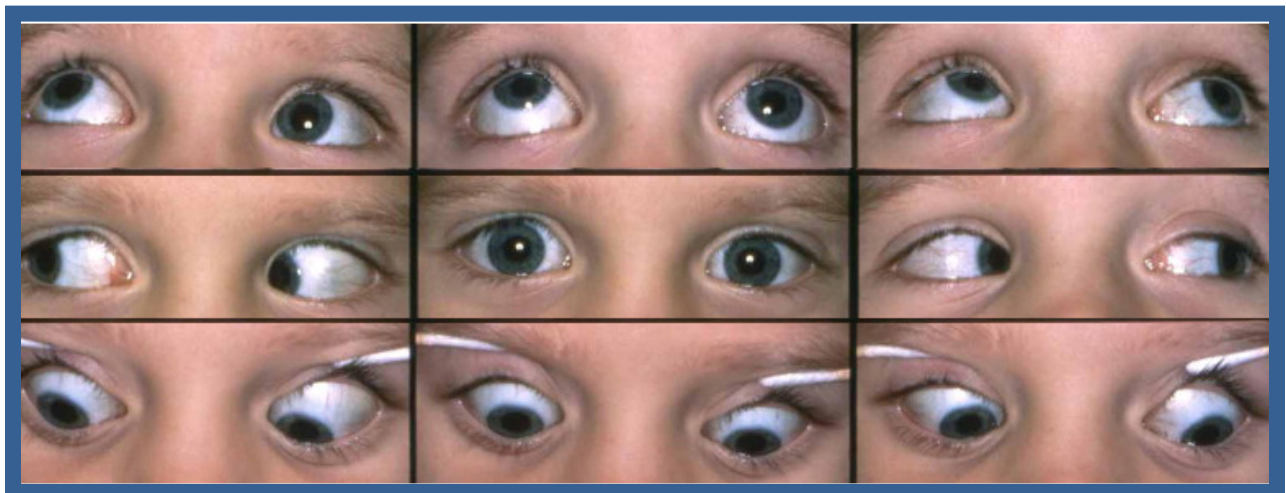


Duane Syndrome, Type II, Right Eye-Right Eye is Unable to Turn Completely Inward

Brown Syndrome

Brown syndrome is a condition typically present at birth but sometimes acquired later in life in which the affected eye is unable to move upward, especially when it is also turned toward the nose. This is caused by the inability of the superior oblique muscle, one of the muscles on the top of the eye, to slide through its natural pulley along the nasal bony wall of the eye socket. This condition is often first noted when a parent sees upward floating of the uninvolved eye as the child looks to the side opposite the affected eye. Alternately, patients may exhibit a Y pattern eye misalignment, i.e. exotropia or outward turning of the eyes in upgaze.

Brown syndrome is sometimes an incidental finding on an eye examination and does not necessitate treatment. However, if the involved eye is lower than the other eye when the individual is looking straight ahead or a chin up position is needed to keep the eyes aligned, eye muscle surgery should be considered.



Brown Syndrome of the Left Eye

Limitation of the left eye to look upward, especially when it is turned toward the nose. Mild outward turning of the eyes when the patient looks straight up. Mild depression of the left eye when the patient looks to the right.

Third Nerve Palsy

Third nerve palsy refers to a weakness of the nerve that supplies impulses to four of the six extraocular muscles, to a muscle that elevates the eyelids, and to the pupil. This may be congenital or acquired following head trauma, brain tumor, stroke, or cerebral aneurysm. The affected eye is generally turned outward (exotropia) and downward (hypotropia). At times, there is an associated droopy eyelid (ptosis) or enlarged pupil.

Occasionally a congenital or acquired third nerve palsy can “regenerate” spontaneously over the course of six months. While we wait to see if the third nerve palsy resolves without intervention, the patient may occlude one glasses lens or cover one eye to help alleviate diplopia. If the strabismus arising from a third nerve palsy remains stable but significant, eye muscle surgery can be performed to eliminate ocular misalignment and restore single vision, at least in primary gaze. A droopy upper lid can confound the patient’s ability to use the realigned eye. In that case, ptosis repair following strabismus surgery becomes warranted.

Fourth Nerve Palsy (a.k.a. Superior Oblique Palsy)

Beginning in the brainstem and travelling to different areas of the head and neck are 12 cranial nerves, each of which has a specific function. A fourth nerve palsy refers to a weakness of the nerve that supplies impulses to the superior oblique muscle, a muscle on the top of the eye whose main function is to move the eye downward and whose secondary function is to rotate the eye inward. If this muscle is weak, the eye tends to drift upward (hypertropia) and sometimes to rotate outward (a type of cyclotropia).

A fourth nerve palsy is often congenital and diagnosed in infancy. On the other hand, signs and symptoms of a congenital fourth nerve palsy may not appear until later in childhood or in adulthood. In addition, older children and adults can acquire this form of vertical strabismus, for example, with trauma. In acquired cases, it may take four to six months to improve or resolve, if at all. Most patients with an isolated fourth nerve palsy have no known underlying cause.

Signs of a Fourth Nerve Palsy

Adults with a fourth nerve palsy are frequently bothered by double vision. In acquired cases, patients may complain of apparent tilting of objects. In children and adults, a noticeable vertical deviation of the eyes is not always the initial sign. Rather, an individual with a fourth nerve palsy often manifests a head tilt (torticollis) to the side opposite the palsied eye to compensate for the strabismus and to minimize the vertical eye misalignment. The head tilt permits the affected child or adult to experience binocular vision and depth perception.



Patient with a Left Fourth Nerve (Superior Oblique) Palsy and Associated Right Head Tilt Before and After Strabismus Surgery

The two pictures on the left were taken prior to strabismus surgery and the two pictures on the right show the patient after strabismus surgery.



Right Fourth Nerve (Superior Oblique) Palsy and Right Inferior Oblique Overaction

The right eye is higher than the left which worsens when the patient gazes left. When the patient looks up and in, the right eye becomes even higher than the left.

Treatment

Prisms in the form of a plastic film that adheres to glasses (Fresnel prism) or correction that is ground into glasses may alleviate symptoms of diplopia or tired eyes (asthenopia) in adult patients with a small eye deviation. Eyeglass prisms bend light and shift the image up, down, left, or right depending on how it is positioned in or on the glasses. They allow the patient's misaligned eyes to work together. They are

usually prescribed for small degrees of ocular misalignment since the stronger they are, the more they degrade images and the thicker and heavier they make glasses if ground in. Some patients are intolerant of prisms, even if they are of small power. Moreover, prisms do not help align images if they are rotated.

In cases of fourth nerve palsies, prisms are often poorly tolerated because 1) the amount of prism necessary to address the magnitude of eye misalignment often makes images blurry, 2) prisms do not change power when patients look in different directions (vertical eye misalignment is worse when patients gaze to the opposite direction and tilt their head to the same side as the eye with the fourth nerve palsy) and 3) prisms do not eliminate tilting of objects. In addition, prisms are usually avoided as treatment for children because image degradation is undesirable for their developing visual system. Surgery is tailored to align eyes and promote stereoacuity in different gaze positions. Therefore, eye muscle surgery is generally recommended as the treatment for fourth nerve palsy in adults and children. Following a corrective eye muscle procedure, the associated abnormal head tilt usually decreases or disappears.

Since infants with a congenital fourth nerve palsy (or any other type of strabismus) may favor one eye and habitually ignore the other deviated eye, they may develop amblyopia (lazy eye) or poor vision in one eye due to inadequate visual stimulation. If present, amblyopia should be addressed before proceeding with strabismus surgery in children.

Frequently Asked Questions

Do children outgrow a fourth nerve palsy? No.

Will eye exercises help this problem? No.

At what age should the surgery be performed?

Most pediatric ophthalmologists recommend surgical correction of a congenital fourth nerve palsy sometime after age six months. Some children and adults require more than one strabismus operation to eliminate the eye misalignment.

Sixth Nerve Palsy

Sixth nerve palsy refers to a weakness of the nerve that supplies impulses to the lateral rectus muscle, the eye muscle that mainly moves the eye outward. This is usually an acquired condition which presents with gradual or sudden onset of eye crossing, double vision, and inability of the eye to move outward. An abnormal face turn to the side of the paretic sixth nerve may occur in order to relieve the double vision. In

children, the most common cause of a sixth nerve palsy is most often viral and can be recurrent. Other causes in the young age group include head trauma and brain tumors. Small blood vessel disease associated with diabetes or high blood pressure is a common cause of sixth nerve palsies in adults.

Sixth nerve palsies generally improve over the course of several months. Botox treatment to the patient's antagonist medial rectus muscle (eye muscle on the nasal side of the same eye) is sometimes recommended, particularly in acute cases, to prevent contracture while the lateral (outside) muscle regains strength. After a period of observation, if recovery is incomplete and residual eye crossing remains, eye muscle surgery can eliminate the eye crossing in straight ahead gaze and relieve symptoms of double vision. Restoring single vision in right and left gaze in addition to primary gaze depends on the severity of the sixth nerve palsy and the surgical procedure performed.

Thyroid-Related Eye Disease

Thyroid-related eye disease represents the most common cause of vertical eye deviation in adults. Extraocular muscles enlarge with fluid and lymphocytic infiltration, become inflamed, and then fibrotic. The extraocular muscles are affected in the following order: Inferior, medial, superior, and lastly, the lateral rectus muscles. These changes cause the affected muscles to become asymmetrically or unequally tight, which leads to restricted eye movements, lid retraction, and prominent-appearing eyes. Sometimes patients benefit from orbital, eye muscle, and lid surgery, in that order. These changes can occur even with normal thyroid bloodwork.

Nystagmus

Nystagmus refers to an abnormal persistent, involuntary, oscillatory motion of the eye(s), usually from side to side. The shaky, repetitive, eye movements sometimes occur in a vertical or rotary motion. Nystagmus is typically congenital (infantile) but may be acquired.

Infantile Nystagmus Syndrome

Infantile nystagmus syndrome (INS) refers to nystagmus that appears during the first six months of life. Although nystagmus may arise from an abnormality of the eyes, it can also arise as a result of a subtle imbalance of the eye movement mechanism within the brain.

Ocular albinism, optic nerve abnormalities, congenital cataracts, and retinal dystrophies are common ocular anomalies that may cause nystagmus.

Children with INS require a pediatric ophthalmologist to evaluate the child and order a relevant workup to rule out serious neurologic causes of the nystagmus.

Most patients with nystagmus have an identifiable null zone, i.e., a position of gaze where the nystagmus is least, and therefore, vision is best. This may cause the patient to adopt an abnormal head posture. For example, if the null zone occurs when the child gazes upward, then the child will assume a chin-down position to place his/her eyes in upgaze. Alternatively, if the null zone is present when the child gazes toward the right, then the child will turn his/her head to the left to shift the eyes into right gaze.

Nystagmus Surgery

Eye muscle surgery for pediatric and adult patients with INS may be effective for two reasons. For patients with an abnormal head position due to an eccentric null zone, surgery can center the null zone and thereby improve or eliminate the aberrant head posture. Additionally, even in INS patients with a normal head posture, horizontal rectus tenotomy surgery can reduce the severity of nystagmus, establish a broader null zone, and lead to improved visual acuity.