Duane Syndrome

What is Duane Syndrome?

Duane syndrome is a congenital eye movement disorder characterized by the following clinical features: horizontal eye deviation on primary gaze, contralateral face turn, limited abduction, lid fissure narrowing on adduction and lid fissure widening on abduction.

Vision is almost always normal unless there is an associated anisometropia and amblyopia. Gaze towards the side of the unaffected eye and contralateral face turn are compensatory head postures to allow binocular single vision.

The condition is unilateral in most cases but bilateral cases occur in about 15-20%. It occurs more commonly in females and the left eye is more commonly affected than the right.

About 30-50% of patients with Duane syndrome have associated congenital neurologic deficits that localize to the brainstem such as crocodile tears, sensorineural hearing loss, or structural defects involving ocular, skeletal, and neural structures.

What is the cause of this condition?

It is caused by a congenital absence of the sixth cranial nerve nucleus with misdirection of the medial rectus nerve, innervating both the medial rectus and the lateral rectus muscle. Since both the lateral rectus and medial rectus are innervated by the nerve to the medial rectus, both muscles fire and contract simultaneously on attempted adduction. This co-contraction of the muscles causes globe retraction and lid fissure narrowing on attempted adduction.

Are there variations in clinical presentation?

There are three clinical types based on the direction of the eye turn on primary gaze. Duane I shows esotropia on primary gaze and has good adduction and poor abduction. Duane II shows exotropia and has variable adduction and decreased to normal abduction. Duane III shows variable horizontal deviation on primary gaze and has poor adduction and abduction. All types show globe retraction on adduction and lid fissure narrowing on adduction.

Does the condition run in families? Any associated systemic conditions?

Duane syndrome may be associated with Goldenhar syndrome and prenatal thalidomide exposure, a known teratogen. However, the condition is most often isolated, sporadic, and of unknown cause.

Familial unilateral and bilateral cases occur, and is usually is transmitted in an autosomal dominant pattern. The clinical presentation in familial cases varies.

How is this condition managed?

The basic problem in Duane syndrome cannot be fixed by any means; however, any associated strabismus, amblyopia, or refractive error must be treated and reviewed appropriately.
The majority of patients with Duane syndrome have few problems other than a head turn and do not require surgery if mild. Parents should be reassured about this compensatory position. Surgery is done to correct unacceptable head posture associated with strabismus, relieve marked globe retraction, improve palpebral fissure changes on eye movement, and improve severe upshoots or downshoots of the affected eye. Although strabismus surgery is very effective for correcting face turn and improving abduction, full abduction is rarely achieved.

References: