Duane Syndrome

WHAT IS DUANE SYNDROME?

Duane syndrome, also called Duane retraction syndrome (DRS), is a congenital (present since birth) and non-progressive (does not get better or worse over time) type of strabismus (eye movement problem). It is one of a group of eye diseases called the Congenital Cranial Dysinnervation Disorders (CCDDs). The eye problems in this group all have abnormal eye movement. In Duane syndrome, it can be hard to move one or both eye(s) outward toward the ear (abduction) or inward toward the nose (adduction), or both. There may also be eyelid movements when the eyes are trying to look around.

WHAT IS THE CAUSE OF DUANE SYNDROME?

Duane syndrome comes from a miswiring of nerves to the eye muscles.

In Duane syndrome, the 6th cranial nerve that controls the lateral rectus muscle (the muscle that pulls the eye out towards the ear) does not develop normally. Why the nerve does not develop well is not fully understood. Because the nerve does not develop normally, sometimes the muscle also does not develop normally, it can be weak or tight. In Duane syndrome there can also be miswiring of the 3rd cranial nerve, which normally controls the medial rectus muscle (the muscle that pulls the eye toward the nose). This is why there may be problems looking both left and right in people with Duane syndrome.

WHO GETS DUANE SYNDROME?

Most people with Duane syndrome get it spontaneously, no one else in the family has the problem. And most often only one eye is affected. Around 20% of people with Duane syndrome have problems in both eyes. For reasons unknown, the left eye is more often affected than the right eye. Duane syndrome also affects girls more often than boys. No particular race or ethnic group is more likely to get Duane syndrome. Duane syndrome is also seen

with other problems that affect growth and development. 30% of people with Duane syndrome have other medical problems from birth.

WHAT IS SEEN IN PEOPLE WITH DUANE SYNDROME?

- Strabismus: the eyes may be misaligned (point in different directions) some of the time or all of the time
- Head position: people often turn their head to one side to try to keep the eyes straight
- Amblyopia (blurry vision, weak vision, lazy eye): occurs in 10% of those with Duane syndrome
- Eyelid narrowing: the eye with Duane syndrome may look smaller than the other eye. This eyelid narrowing or squinting is easier to see when the eye moves left and right.
- Upshoot or downshoot: when looking in certain directions, the eye may sometimes roll upward or downward

IS DUANE SYNDROME CONGENITAL (PRESENT FROM BIRTH)?

YES. Duane syndrome is present from birth, even if it is not always seen during infancy. Often times, a head tilt or turn and eye movement problems can be seen in old photographs taken in early childhood for people later found to have Duane syndrome.

IS DUANE SYNDROME HEREDITARY?

Most of the time, it is not hereditary (does not run in families). In 90% of cases, there is no family history of Duane syndrome. Only 10% of people will have an affected family member and these tend to be cases where both eyes have Duane syndrome.

ARE THERE DIFFERENT TYPES OF DUANE SYNDROME?

Yes! Duane syndrome generally has three main forms. In type I Duane syndrome it is hard for the eye to move outward toward the ear. In type II Duane syndrome it is hard for the eye to move inward toward the nose. In type III Duane syndrome it is hard for the eye to move inward AND outward. Type I is the most common form of Duane syndrome. People with type I have a head turn towards the eye with problems moving and will look like they

have esotropia (crossed-eye, eye turned toward nose) when trying to look straight ahead.

DO DUANE SYNDROME PATIENTS HAVE OTHER EYE PROBLEMS?

Sometimes, Duane syndrome may be seen with other eye problems including other forms of strabismus (eye movement problems), nystagmus (back-and-forth movement of the eyeball), cataract, amblyopia (blurry vision, weak vision, lazy eye), optic nerve problems, microphthalmos (abnormally small eye), and crocodile tears.

BUT, most of the time, a person with Duane syndrome is otherwise completely normal. With careful follow-up eye exams, long-term vision is usually excellent.

DO PEOPLE WITH DUANE SYNDROME HAVE OTHER MEDICAL PROBLEMS?

Not usually, however, some patients with Duane syndrome can have hearing problems, Goldenhar syndrome (a medical problem affecting eyes, ears, the spine and the face), and spine problems. Children with Duane syndrome in both eyes may have weakness of the muscles of the face and muscles involved in sucking (a problem called Moebius syndrome).

WHEN IS DUANE SYNDROME TREATED WITH SURGERY?

For most patients, Duane syndrome does not require surgical treatment. Surgery for Duane syndrome may be needed for one of four reasons:

1. To make the eye turn smaller when looking straight ahead
2. To help improve a head turn
3. To help improve the eye rolling up or down when looking off to the side
4. To help improve how the eyelid looks

Surgery can also help to prevent amblyopia (weak vision or lazy eye).

HOW SUCCESSFUL IS SURGERY FOR DUANE SYNDROME?

Surgery cannot actually fix the problem of nerves that are miswired. It cannot make the nerve or muscle work “normally.” However, eye muscle surgery

(strabismus surgery) can (and usually does) improve the eye movement. Some people with Duane syndrome may need multiple eye surgeries and the full effect of the surgery may take some weeks to months to show itself.

WHERE CAN I FIND MORE INFORMATION REGARDING DUANE SYNDROME?

- National Human Genome Research Institute
- EyeWiki