

Duane Syndrome

What is Duane Syndrome?

Duane syndrome, also called Duane retraction syndrome (DRS), is a congenital and non-progressive type of strabismus due to abnormal development of the 6th cranial nerve. It is characterized by difficulty rotating one or both eyes outward (abduction) or inward (adduction). There may also be changes of eyelid position on attempted movement of the eyes.

What is the cause of Duane Syndrome?

Duane syndrome is due to miswiring of nerves to the eye muscles.

In Duane syndrome, the 6th cranial nerve that controls the lateral rectus muscle (the muscle that rotates the eye out towards the ear) does not develop properly. Why the nerve does not develop is not fully understood. Thus, the problem is not primarily with the eye muscle itself, but with the nerve that controls to the muscle. There can also be associated with miswiring of the 3rd cranial nerve, which normally controls the medial rectus muscle (the muscle that rotates the eye toward the nose). This is why abnormalities may be found in both left gaze and right gaze.

Who gets Duane Syndrome?

The vast majority of cases occur spontaneously and most often only affect one eye. Around 20% of Duane syndrome patients have both eyes affected. For reasons unknown, the left eye is more often involved than the right eye. Duane syndrome affects girls more often than boys. No particular race or ethnic group is more likely to be affected. 30% of cases are associated with other congenital anomalies.

What are the characteristics of Duane Syndrome?

- [Strabismus](#): the eyes may be misaligned and point in different directions some or all of the time
- [Head position](#): patients often maintain an abnormal head posture or head turn to keep the eyes straight
- [Amblyopia](#) (reduced vision in the affected eye): occurs in 10% of patients
- Eyelid narrowing: the affected eye may appear smaller than the other eye
- Upshoot or downshoot: with certain eye movements, the eye may occasionally deviate upward or downward

Is Duane syndrome congenital (present from birth)?



Duane retraction syndrome is present from birth, even if it is not recognized during infancy. An abnormal head posture and strabismus are often visible in old photographs taken in early childhood.

Is Duane syndrome hereditary?

Most of the time, it is not hereditary. In 90% of cases, the patient has no family history of Duane syndrome. Only in 10% of patients will have an affected family member and these tend to be cases where both eyes are involved.

Are there different types of Duane Syndrome?

Duane syndrome is often characterized by whether the primary abnormality is a reduced ability to turn the affected eye(s) outward (type I), inward (type II), or both (type III). Type I is the most common form of Duane syndrome, and affected patients will characteristically have a head turn towards the involved side and will appear esotropic (crossed-eye) in straight ahead gaze.

Do Duane Syndrome patients have other eye problems?

The problem with the 6th cranial nerve is usually an isolated condition and the child is usually otherwise completely normal. With careful follow-up, the long-term prognosis for good vision is usually excellent.

Occasionally, Duane syndrome may be found in association with other eye problems, including disorders of other cranial nerves, [nystagmus](#) (an involuntary back-and-forth movement of the eyeball), [cataract](#), optic nerve abnormalities, microphthalmos (abnormally small eye), and crocodile tears.

Do Duane Syndrome patients have non-ocular medical problems?

Not usually, however, some patients with Duane syndrome have other problems, such as hearing impairment, Goldenhar syndrome, spinal and vertebral abnormalities. There is also an increased frequency of Duane syndrome in patients with thalidomide exposure.

When is Duane Syndrome treated?

For the majority of patients, Duane syndrome does not require surgical treatment. Surgery for Duane syndrome is indicated for one of four reasons:

1. To reduce a significant deviation in normal straight-ahead position
2. To eliminate a significant abnormal head position
3. To eliminate a significant upshoot or downshoot.
4. To eliminate disfiguring abnormal eyelid position



The goal of treatment is to restore satisfactory eye alignment in the straight-ahead position, eliminate an abnormal head posture and to prevent amblyopia. Eye muscle surgery is not always required. Because the function of the affected nerve and muscle cannot be restored, the other eye muscles are adjusted to compensate and allow for better eye alignment.

How successful is surgery for Duane Syndrome?

Surgery cannot actually fix the problem of nerves that are miswired, but can move muscles to compensate for the miswiring. Because surgery cannot fix the problem of the abnormal nerves, surgery cannot restore normal eye movement. However, surgery can (and usually does) substantially improve the situation. The full effect of the surgery may take some weeks to become apparent.

Where can I find more information regarding Duane syndrome?

- [National Human Genome Research Institute](#)
- [EyeWiki](#)

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