

# Duane Syndrome

## Description

- Congenital abnormality of the 6th cranial nerve
  - 4th-8th week of gestation
  - Missing nucleus in most cases (4th week)
- Abnormal innervation from 3rd cranial nerve

Type	Abduction	Adduction	Deviation	Frequency
I	Poor	Normal	Ortho- or Esotropia	70%
II	Normal	Poor	Ortho- or Exotropia	15%
III	Poor	Poor	Ortho-, Eso- or Exotropia	15%

## Clinical Characteristics

- Most sporadic, 5% autosomal dominant inheritance
- More common in females than males.
- OS affected more often than OD.
- 80% of cases are unilateral, when bilateral it is usually asymmetric.
- Gender and Laterality:

Female	Male	OS	OD	OU
58%	42%	59%	23%	18%

- Gender and Laterality in 835 cases of Duane Syndrome. DeRespinis PA, Wagner RS, Guo S; Duane's Retraction Syndrome. Surv Ophthalmol 38:257, 1993

## Exam Findings

- Narrowing of the palpebral fissure and retraction of the globe of the involved eye on attempted adduction.
- The amount of limitation depends on the amount of co-contraction of the medial rectus and lateral rectus.
- Abnormal firing of the lateral rectus is found with EMG testing.
- Angle of deviation almost always less than 30D , usually less than 15D .
- Face turn toward the side of limited movement- permits fusion.
- Anisometropic Amblyopia 10-30%
- Ipsilateral congenital hearing loss 5%

## Selected Associated Syndromes

- Most cases are isolated
- **Goldenhar syndrome** (hemifacial microsomia, ocular dermoids, ear abnormalities, preauricular skin tags, eyelid colobomas)
- **Wildervanck syndrome** (sensorineural hearing loss, Klippel-Feil anomaly- shortness of neck with fused or missing vertebrae)

## Treatment

- Treat Refractive Error
- Consider treating Hyperopia > +2.50 if ET
- Treat Amblyopia
- Goals of Surgery:
  - Improve primary position alignment
  - Improve head turn
  - Decrease up-shoot or down-shoot
- Surgical Treatment Possibilities
  - Eso-Duanes
    - Medial Rectus (MR) Recession
    - Bilateral Medial Rectus Recessions
    - Superior Rectus and/or Inferior Rectus transpositions
  - Exo-Duanes
    - Lateral Rectus (LR) Recession(s)
  - Up-shoots/Down-shoots
    - Recession with Y split or posterior fixation of LR

[strabismus, syndrome](#)

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Last update: **2015/11/09 21:52**

