

Back to Basics - Duane Retraction Syndrome

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Introduction

Duane retraction syndrome or Stilling-Turk-Duane syndrome is a congenital cranial dysinnervation disorder characterized by abduction/adduction deficits, palpebral fissure width changes, globe retraction on attempted adduction, upshoot or downshoot. DRS is more commonly unilateral than bilateral, affecting the left eye more often. Bilateral cases of DRS have been reported to range from 10% to 24%. Type I is the most common followed by types III and II. Patients with DRS present with an abnormal head position, one of the eyes appearing smaller than the other due to globe retraction, pseudoptosis in adduction or abnormal eye movements [1-3].

Etiology

The cause of globe retraction in DRS has been attributed to an abnormally tight lateral rectus. LR receives innervation in adduction which is thought to be the cause of co-contraction. Absence of normal sixth nerve innervation to the LR (nuclear or supranuclear) or complete absence of sixth nerve has been shown on electrophysiological studies and high resolution MRI's. The pathogenesis of upshoots and downshoots is caused due to a bridle effect of the tight lateral rectus (leash effect). As the globe adducts and moves above or below the horizontal plane, there is sudden slippage of the tight LR, causing an upshoot or downshoot.

Classification

Huber's classification system groups DRS into three entities; type I (limited abduction with normal to near normal adduction), type II (limited adduction with normal to near normal abduction), type III (limited abduction and adduction). This scheme has been widely accepted for clinical and electromyographical classification.

Molecular genetic testing

This includes single-gene testing (Sequence analysis of *CHN1*), use of a multigene panel and a more comprehensive genomic testing (exome sequencing, genome sequencing and mitochondrial sequencing).

Evaluation

The following evaluation is recommended: Family history, complete ophthalmologic examination, forced duction testing and/or force generation testing, family album tomography, brain and orbital MRI and general physical examination.

Management

Nonsurgical treatment

Refractive errors should be managed with spectacles or contact lenses to prevent amblyopia. Amblyopia can be treated effectively with occlusion in the early years of life. Prisms can be used to improve the compensatory head position in mild cases.

Surgical treatment

The aim of surgery is to correct or improve compensatory head posture, to improve alignment in primary gaze position and to improve upshoot or downshoot. Esotropic DRS can be treated by unilateral MR recession if esotropia is less than 20 prism diopters. Bilateral MR

recessions may also be required in cases of esotropia with primary position deviation > 20 PD. Type 1 and 3 patients with head turn can be treated with vertical rectus transpositions combined with medial rectus recession. Type 2 patients with head turn should be treated with lateral rectus recession. Exotropic DRS is managed with unilateral or bilateral LR recession. Globe retraction can be corrected with recessions of the co-contracting muscles. MR recession, LR recession and periosteal fixation of the LR are the treatment options. Upshoots and downshoots can be corrected by Y splitting of the LR [4-6].



Figure: Photograph showing (a) Face turn, (b) Limited abduction (LE), (c) (LE) Exotropia, (d) Secondary deviation more than primary, (e) Upshoot (LE) and (f) Downshoot (LE).

Conclusion

Duane retraction syndrome if treated at the appropriate time can correct the strabismus, abnormal head posture, upshoot, downshoot and amblyopia.

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