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Restrictive fibrous bands originating from the oculomotor nerve (CN3) in familial Duane retraction syndrome (DRS)

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Introduction: Fibrous bands cause restrictive strabismus, yet their pathogenesis is mysterious. We studied a familial case suggest neural origin of bands.

Methods: We employed high resolution, T2 weighted, surface coil orbital MRI to investigate the anatomy of incomitant strabismus in a father and son with DRS, correlating with clinical motility.

Results: The two year old son had right enophthalmos with markedly limited abduction, supraduction, and infraduction, and mildly limited adduction accompanied by palpebral fissure narrowing. Forced duction testing under anesthesia revealed diffuse restriction. The 30 year old father was orthotropic in central gaze but had limited right eye abduction and palpebral fissure narrowing in adduction. In unilaterally affected right orbits of both patients, no abducens nerve was visible. The inferior division of CN3 both entered the inferior compartment of the lateral rectus muscle (LR), and was contiguous with dense bands running anteriorly to the inferolateral scleral entry of the short posterior ciliary nerves. In the son, another short band was inserted on the posterior sclera inferior to the optic nerve.

Discussion: In these cases, the inferior division of CN3 was both the source of mis-innervation and of fibrous bands targeting sites of normal ciliary nerve perforation of the sclera. Recognizing that familial Duane retraction syndrome is caused by mutation in nerve pathfinding molecules such as $\alpha 2$ -chimaerin, we speculate that fibrous bands may represent abortive nerves mistargeted to scleral emissary canals.

Conclusion: Occurrence of fibrous bands in familial DRS suggests that bands are caused due to aberrant axons pathfinding by CN3.

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