

Further Insights into Developmental Errors Causing Duane's Syndrome

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EMG studies and a few autopsy cases have indicated that many cases of congenital Duane syndrome are due to a miswiring of the horizontal recti with abnormal innervation of the lateral rectus by a branch from the 3rd nerve. This produces co-contraction of the lateral and medial rectus in attempted adduction resulting in retraction of the globe. This unique type of strabismus raises many questions on the timing, location, and specificity of the error in development of innervation.

Although most cases of Duane syndrome are isolated events, the association of the motility anomalies with inner ear malformations (Wildervank syndrome) and outer ear and cervical anomalies (Goldenhar syndrome) is well documented.

We have studied 40 cases of individuals manifesting congenital malformations due to ingestion of thalidomide by their mothers during early pregnancy. Ocular motility disturbances resembling Duane syndrome and/or a gaze paresis have been observed in a large number (30-35%). These patients will be described in addition to other cases of Duane syndrome with associated systemic anomalies. The developmental implications of timing and location are discussed.