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BILATERAL CONGENITAL ADDUCTION PALSY WITH SYNERGISTIC DIVERGENCE: A CASE REPORT

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Abstract:

Synergistic divergence (SD) is a rare congenital ocular motility pattern, characterized by paradoxical abduction during attempted horizontal gaze to the contralateral side. SD is generally unilateral and always associated with limited adduction of the affected eye. The present study introduces a case report on a bilateral congenital adduction palsy with synergistic divergence. The case was a ten year old boy with a unique case of synergistic divergence and normal monocular movement, which has not been documented before. The case was a Middle Eastern ethnicity, referred to the ophthalmology clinic in Ahvaz Imam Khomeini Hospital, Ahvaz, Iran with complaints of eyelid drooping, squinting and abnormal eye movements of both eyes. The study presents the findings of complete orthoptic and ophthalmologic examinations including dilated fundoscopy.

Keywords: *Synergistic divergence, Case Report, Bilateral Congenital Adduction*

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INTRODUCTION:

Synergistic divergence (SD) is a congenital ocular motility pattern characterized by paradoxical abduction during attempted horizontal gaze to the contralateral side. This rare congenital is generally unilateral and is always associated with limited adduction of the affected eye. The pathophysiology of anomalous abduction remains unclear but has been variously attributed to mechanical factors, anomalous innervation of the ipsilateral medial and lateral recti muscles (1, 2) and even anomalous cross innervation between the two lateral recti (3).

SD is usually an isolated ocular motility abnormality, but it has been described several times in conjunction with ocular motility phenotypes consistent with congenital fibrosis of the extraocular muscles types (CFEOM1) and (CFEOM3) (1, 4, 5).

Recently, certain congenital ocular motility disorders formerly thought due to congenital fibrosis of the extraocular muscles were reclassified as congenital cranial dysinnervation disorders (CCDDs) (6).

These disorders result from the congenital absence or misdirection of specific brain stem lower motor neurons, leading to the loss of correct innervation of certain extraocular and/or cranial muscles, often with subsequent anomalous innervation (dysinnervation) by other nerves (7-9). We report bilateral congenital adduction palsy with synergistic divergence.

CASE REPORT:

We report a unique case of synergistic divergence and normal monocular movement, which has not been documented before.

A 10 year old boy, of Middle Eastern ethnicity, referred to the ophthalmology clinic in Ahvaz Imam Khomeini Hospital, Ahvaz, Iran with complaints of eyelid drooping, squinting and abnormal eye movements of both eyes which was first noted by his parents when he was 4 months of age. The case was born at term with normal vaginal delivery with no complications, With an APGAR score of 9. The patient had complete orthoptic and ophthalmologic examinations including dilated fundoscopy.

During the examination, the patient had a pronounced chin-up in primary position and severe ptosis (Fig. 1). The eyes were aligned in primary position and monocular eye movements were normal in all directions of gaze (Fig. 2). However, in attempted dextroversion or levoversion, SD was observed (Figs 3 and 4).The eye movements were not associated with lid or globe retraction. The interesting observation was that the patient did not meet the clinical criteria for CFEOM (1-3).

Fixation was binocular and fusion measured by worth 4-Dot Test responses at near and distance was in normal range. The slit lamp examination and Brain and extraocular CT-scan were normal.

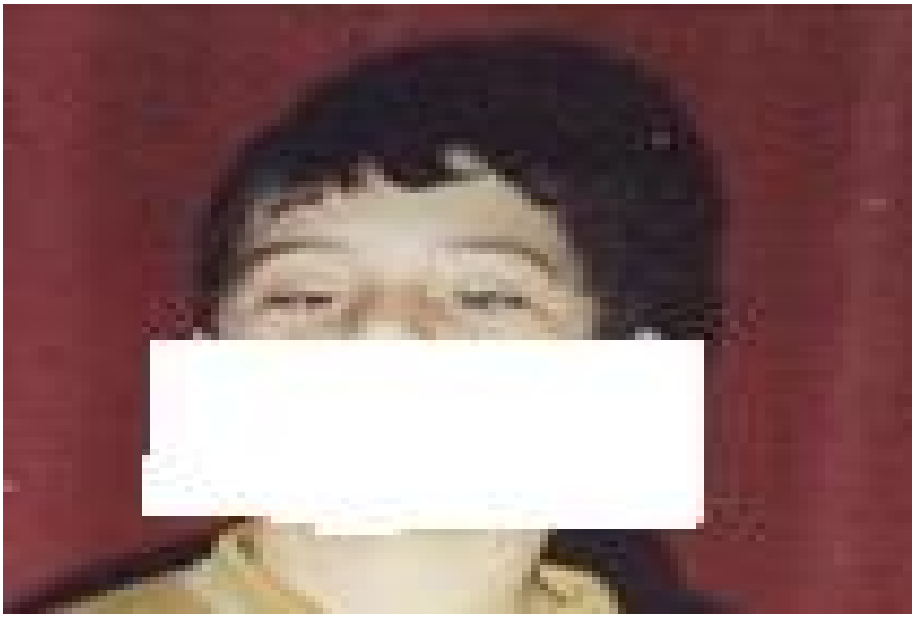

Fig 1. Chin-up in primary position and severe ptosis

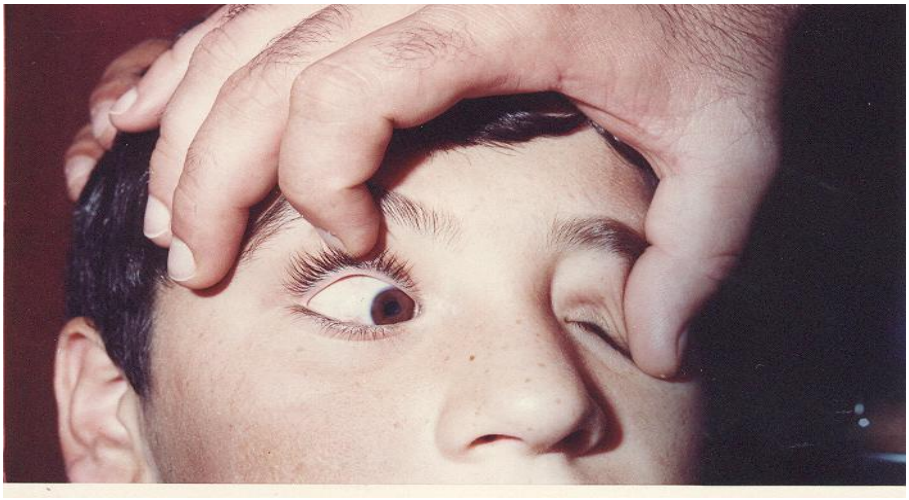

Fig 2. Monocular eye movements in the SD case.

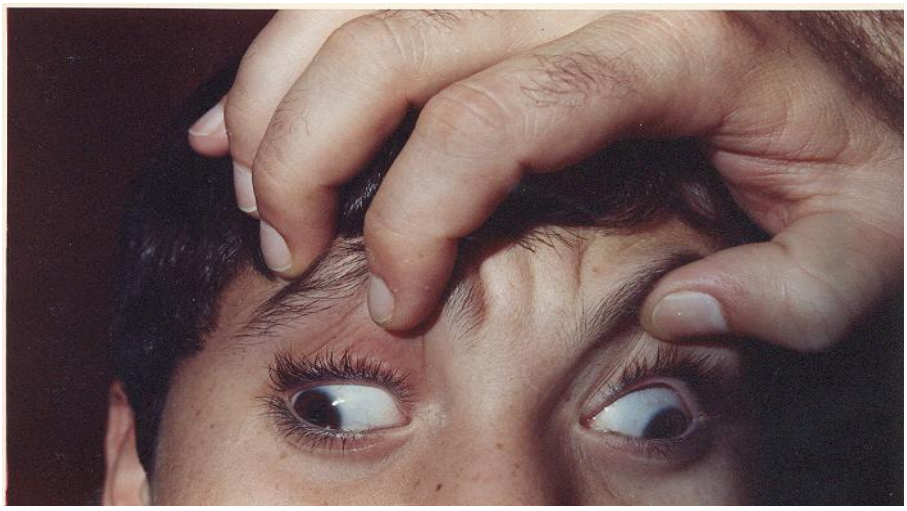

Fig 3. The attempted dextroversion, synergistic divergence.

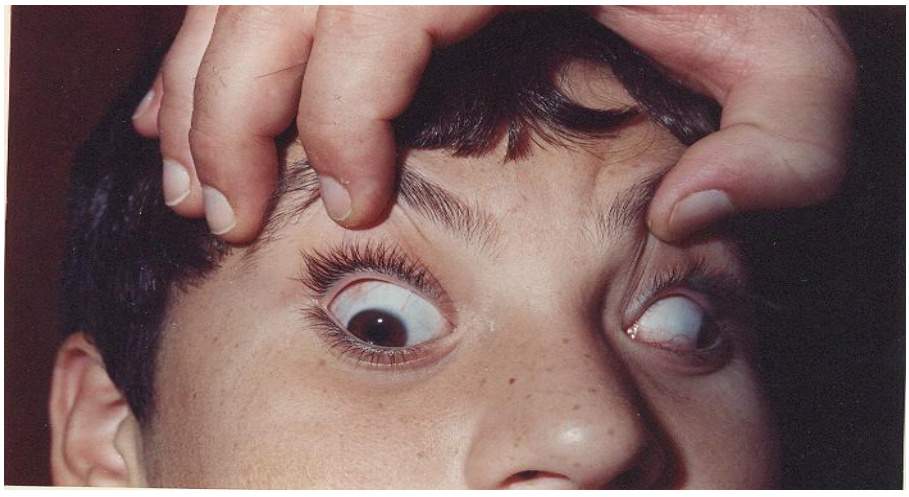

Fig 4. Attempted levoversion, synergistic divergence

DISCUSSION:

The SD has specific characteristics similar to the Duan's Retraction Syndrome (DRS), the most common of them is similar characteristics in CCDD ocular motility pattern. Both SD and DRS affect predominantly horizontal ocular muscles, they both occur unilaterally or bilaterally (10) and they sometimes coexist (11). However, the SD clinical

phenotype differs from DRS in several ways. It involves a different motility pattern from all three DRS types; the muscle most involved is the medial rectus rather than the lateral rectus; and it does not include the DRS clinical hallmarks of globe retraction and lid fissure narrowing. DRS is more common in females for unclear reasons (10). In contrary, reviewing the recent case report studies on the SD showed that this dysfunction is more common among males (12-15).

A possible scenario to explain the different prevalence of SD between the male and female genders is that a developmental anomaly in SD prevents the inferior branch of the oculomotor nerve from correctly innervating the medial rectus. The presence of synergistic divergence of the affected globe suggests that oculomotor fibers that should innervate the medial rectus actually innervate the lateral rectus, causing anomalous abduction on attempted contralateral gaze.

SD is sometimes associated with other evidence of miswiring such as Marcus Gunn jaw winking (13, 16). Moreover, anomalous innervation of the medial rectus by the motor branch of the trigeminal nerve (12). In all previously documented cases, there was a unilateral SD with adduction deficit reported in ocular examination (17, 18). While, in our case, version was normal in all directions of gaze, and the SD was bilateral.

SD deserves recognition as a distinct ocular motility pattern of CCDD, comparable to DRS but much less common. It is possibly caused by congenital denervation of the medial rectus with dysinnervation of the ipsilateral lateral rectus, resulting in the characteristic anomalous abduction bilaterally on attempted contralateral gaze. Future studies may elucidate the genetic and/or teratogenic factors during development that cause this CN misdirection pattern.

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