STRABISMUS

Dr. Saif Ibrahim
MBChB, CABophth

CONGENITAL CRANIAL DYSINNERVATION DISORDERS

Congenital cranial dysinnervation disorders (CCDD) or congenital innervation dysgenesis syndromes constitute a group of disorders originally believed to be the result of congenital muscular fibrosis, but now known to be the result of brainstem or cranial nerve developmental disturbance, in most cases having an identifiable genetic basis. Systemic associations are recognized for several

Duane retraction syndrome

- In Duane retraction syndrome (DRS) there is failure of innervation f the lateral rectus by a hypoplastic sixth nerve nucleus, with anomalous innervation of the lateral rectus by fibres from the third nerve.
- Up to half of patients have associated systemic defects such as deafness, external ear abnormalities, speech disorder and skeletal abnormalities.

Clinical features

1) A face turn is typical and confers BSV with the face in the turned position, thus avoiding amblyopia.
2) Complete or partial restriction of abduction.
3) Restricted adduction, usually partial.
4) Retraction of the globe on adduction as a result of cocontraction of the medial and lateral recti with resultant narrowing of the palpebral fissure.
5) An up-shoot or down-shoot in adduction may be present.
6) In some cases this is produced by a tight lateral rectus muscle slipping over or under the globe to produce an anomalous vertical movement.
7) Deficiency of convergence

Classification(huber)

1) Type I, the most common, is characterized by
- Limited or absent abduction.
- Normal or mildly limited adduction.
- In the primary position, straight or slight esotropia.

2) Type II, the least common, is characterized by
- Limited adduction.
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3)Type III is characterized by:
- Limited adduction and abduction.
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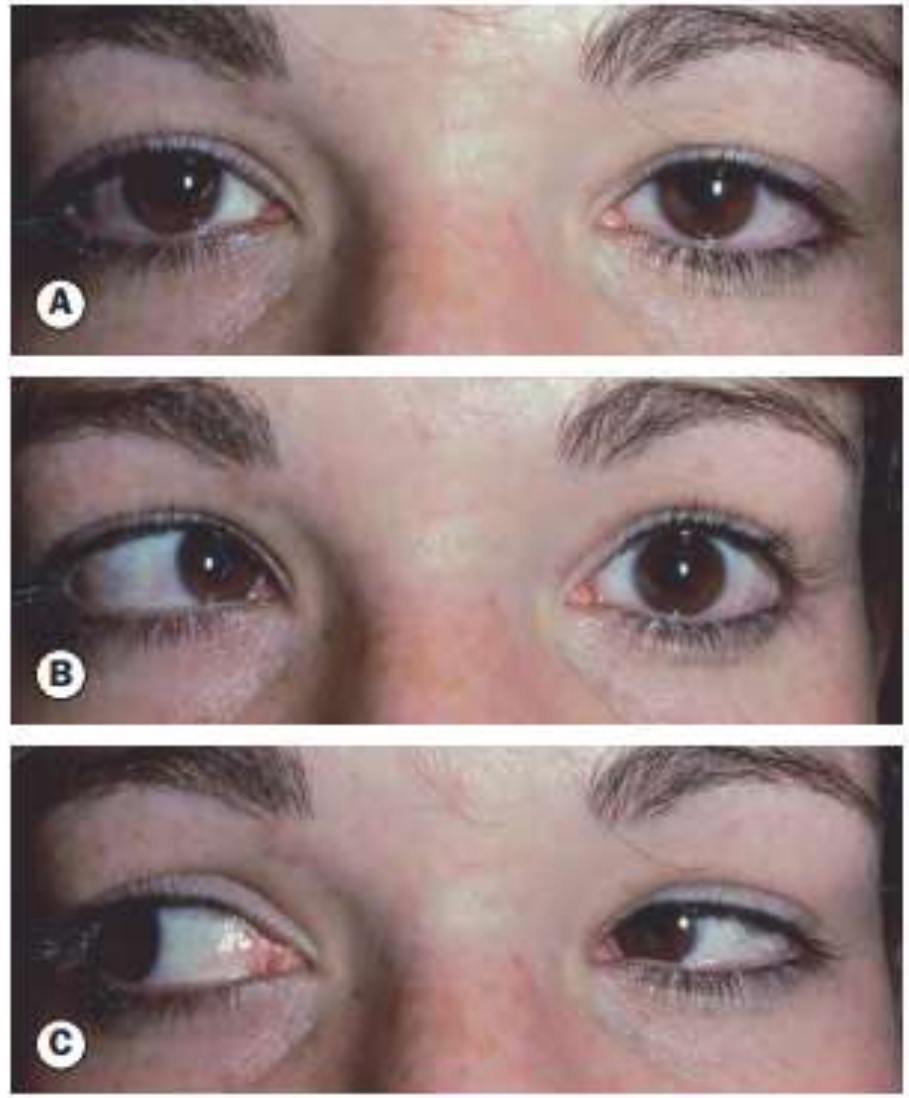

Fig. 18.62 Duane syndrome – Huber type I ('eso'). **(A)** Straight eyes in the primary position; **(B)** extremely limited left abduction; **(C)** narrowing of the left palpebral fissure on adduction *(Courtesy of ADN Murray)*

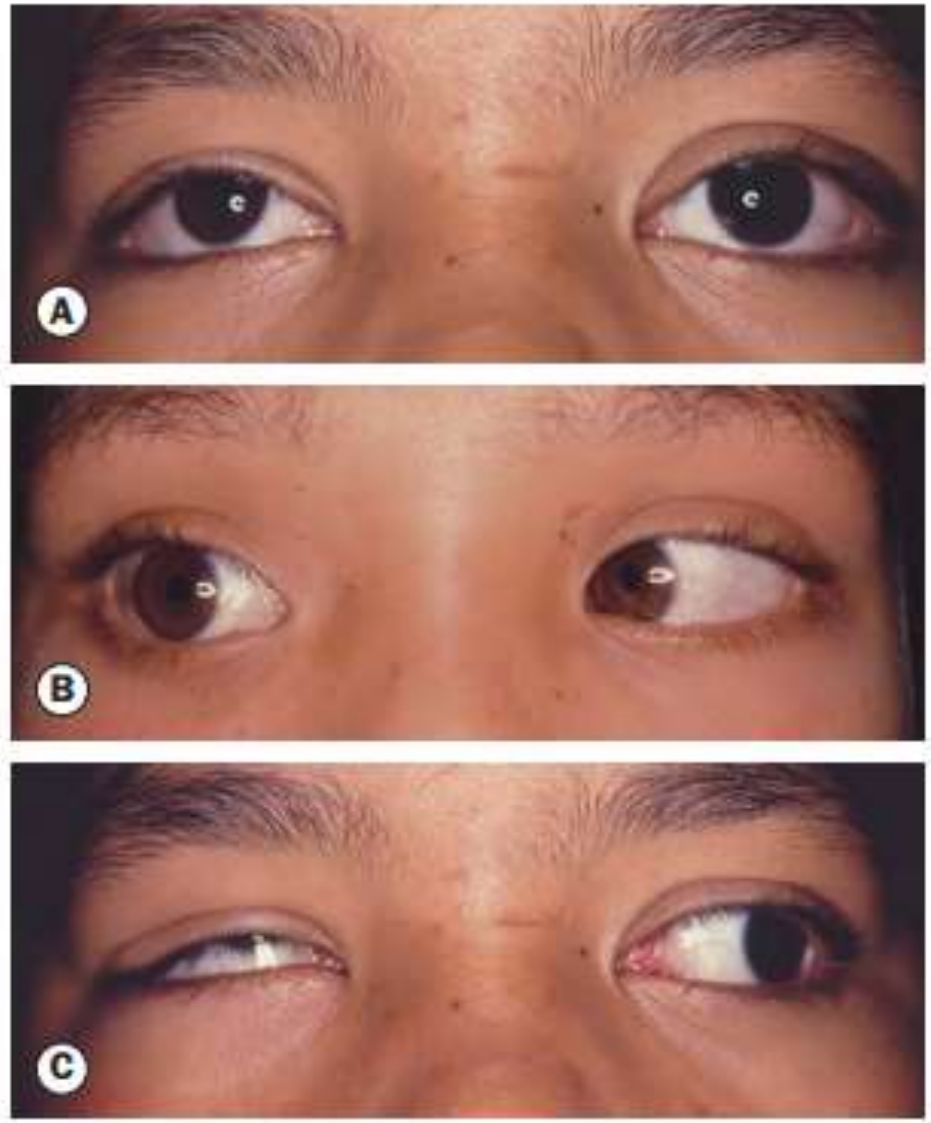

Fig. 18.63 Duane syndrome type II ('exo'). **(A)** Slight exotropia in the primary position; **(B)** limitation of right abduction with widening of the right palpebral fissure; **(C)** grossly limited right adduction with narrowing of the palpebral fissure
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Indications for surgery

1) Loss of binocular function(indicated by failure to continue to use CHP)
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Surgery:

- Depending on the clinical features, unilateral or bilateral medial rectus muscle and/or lateral rectus muscle recession are the procedures of choice.
- In order to improve abduction, transposition of the superior and inferior rectus muscles (or just the superior rectus muscle) to the lateral rectus muscle.


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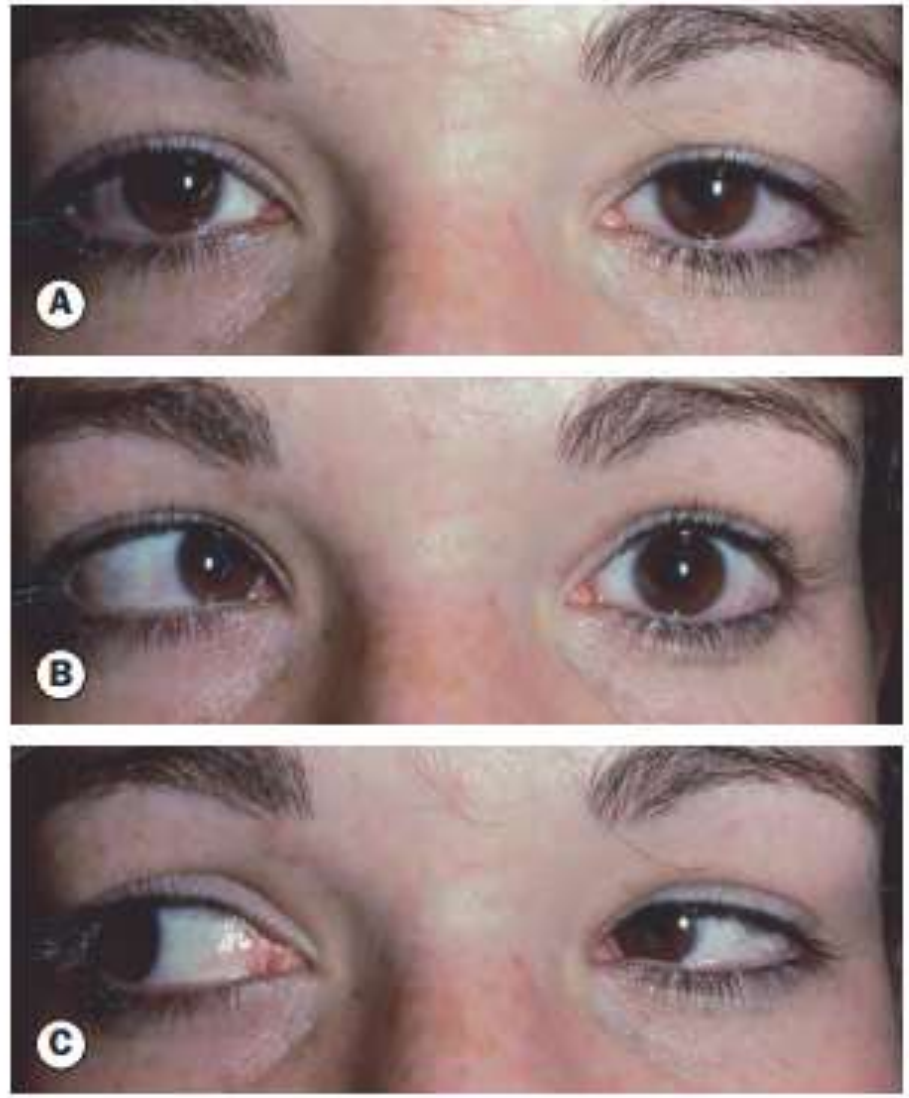

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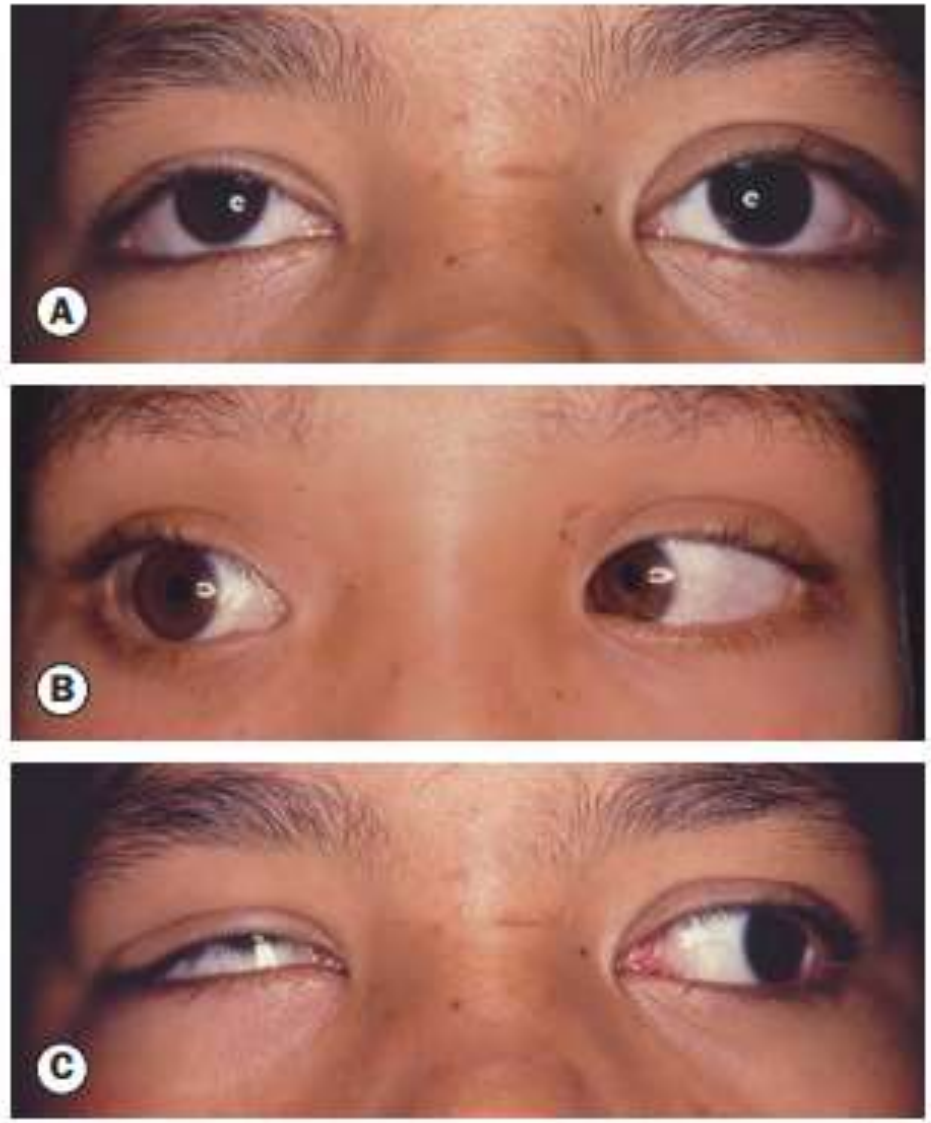

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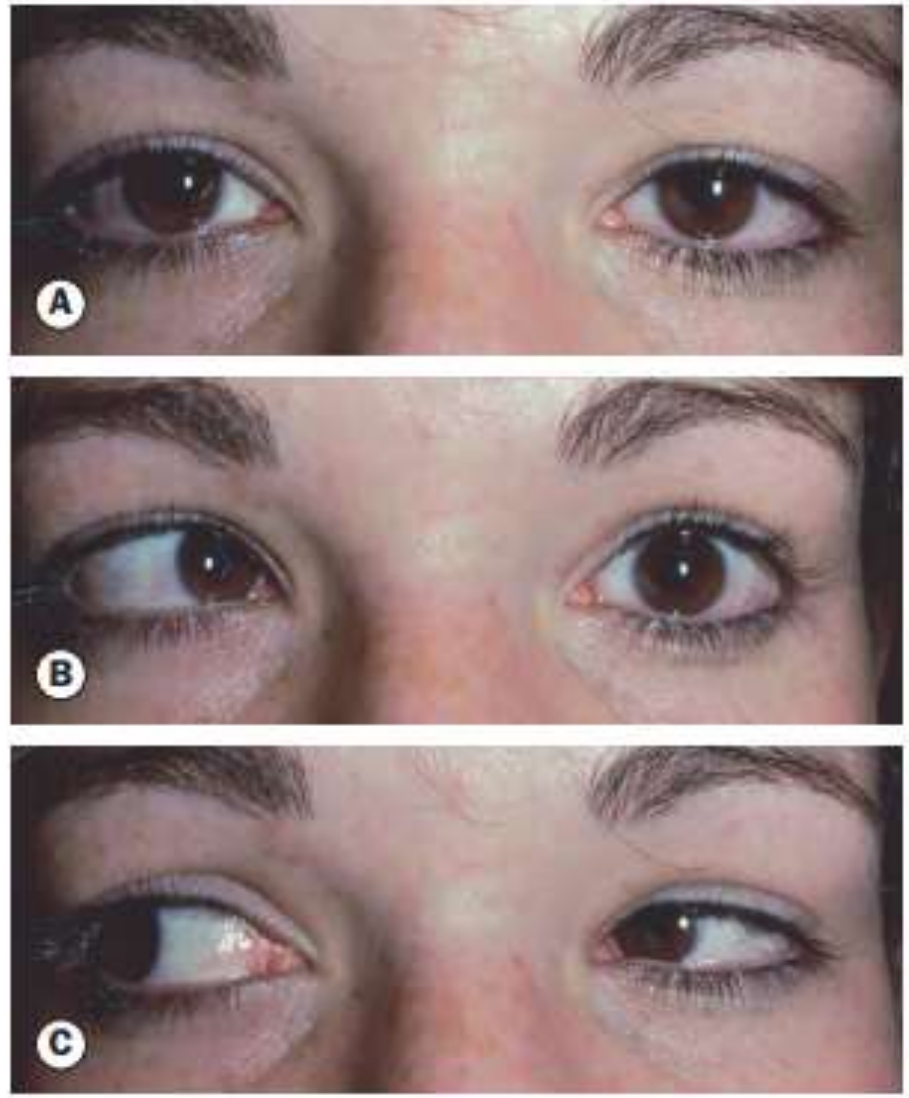

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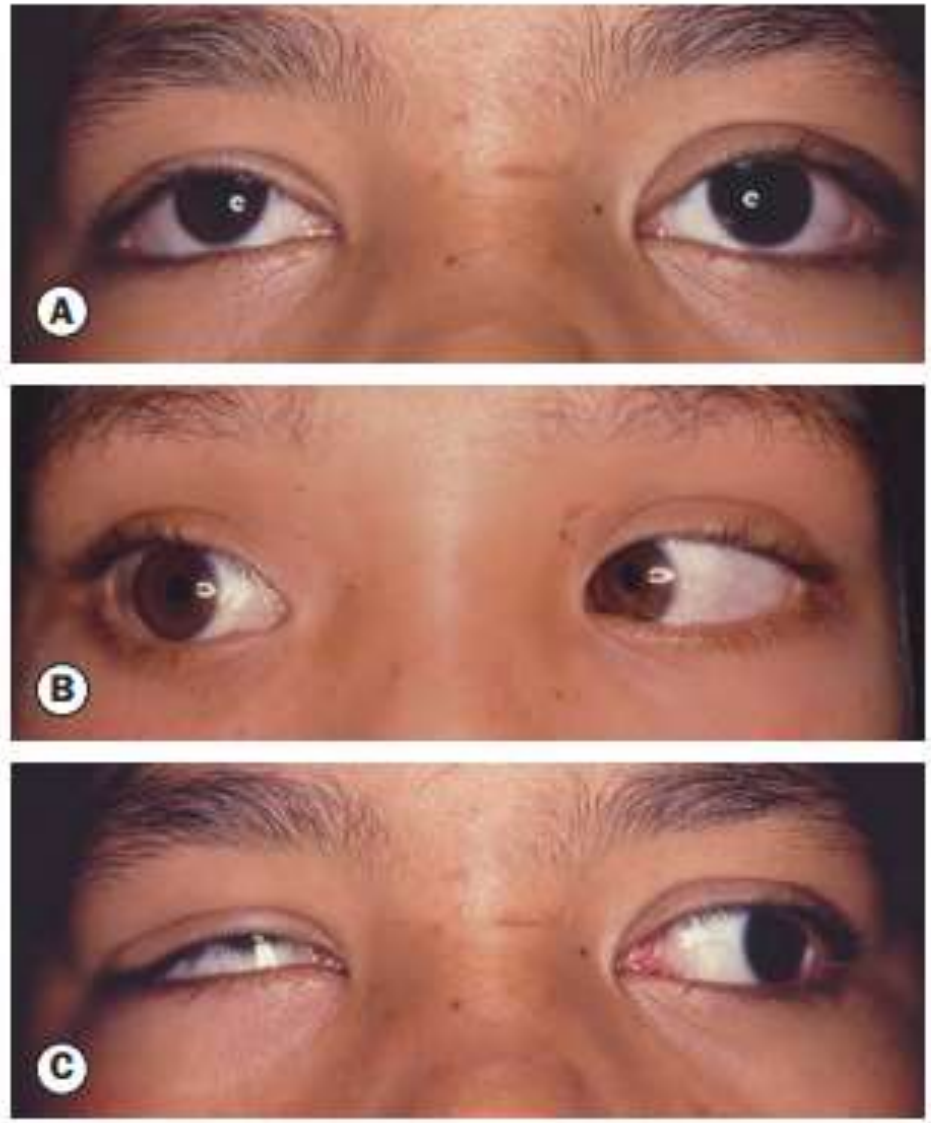

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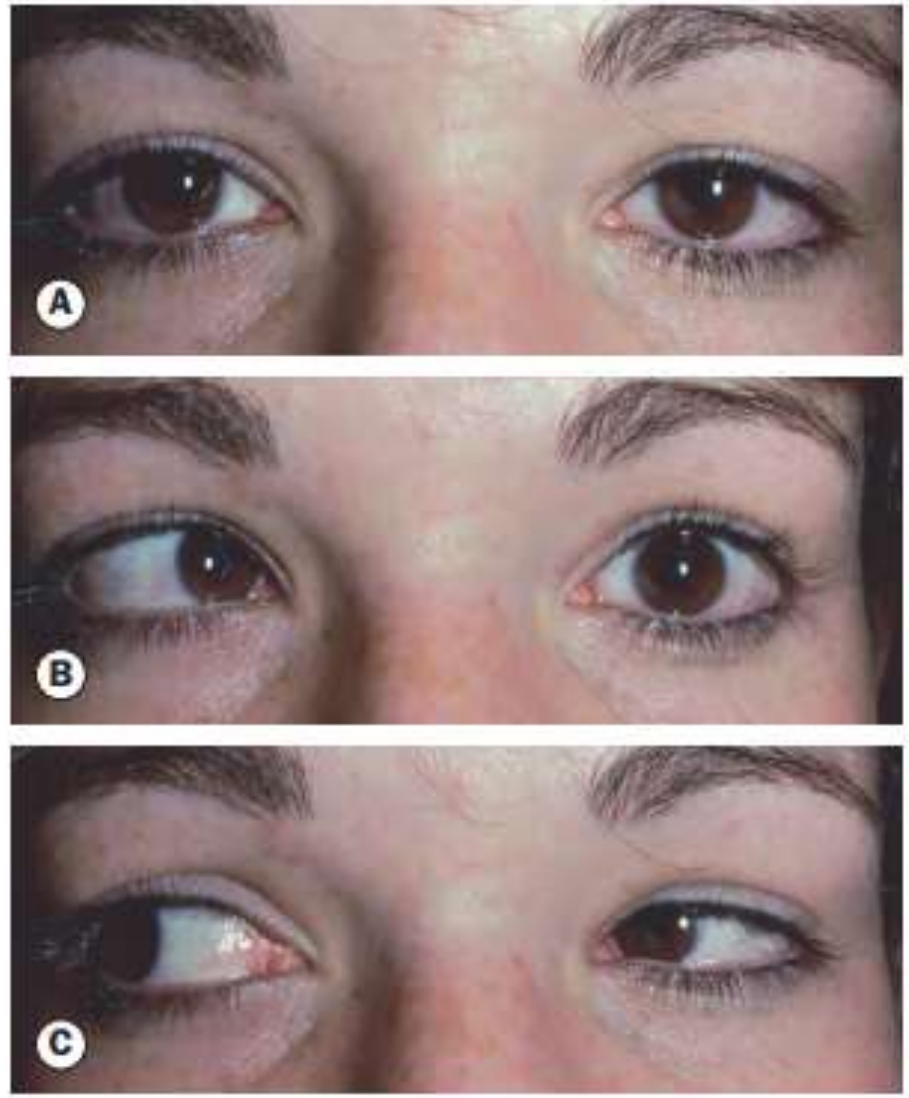

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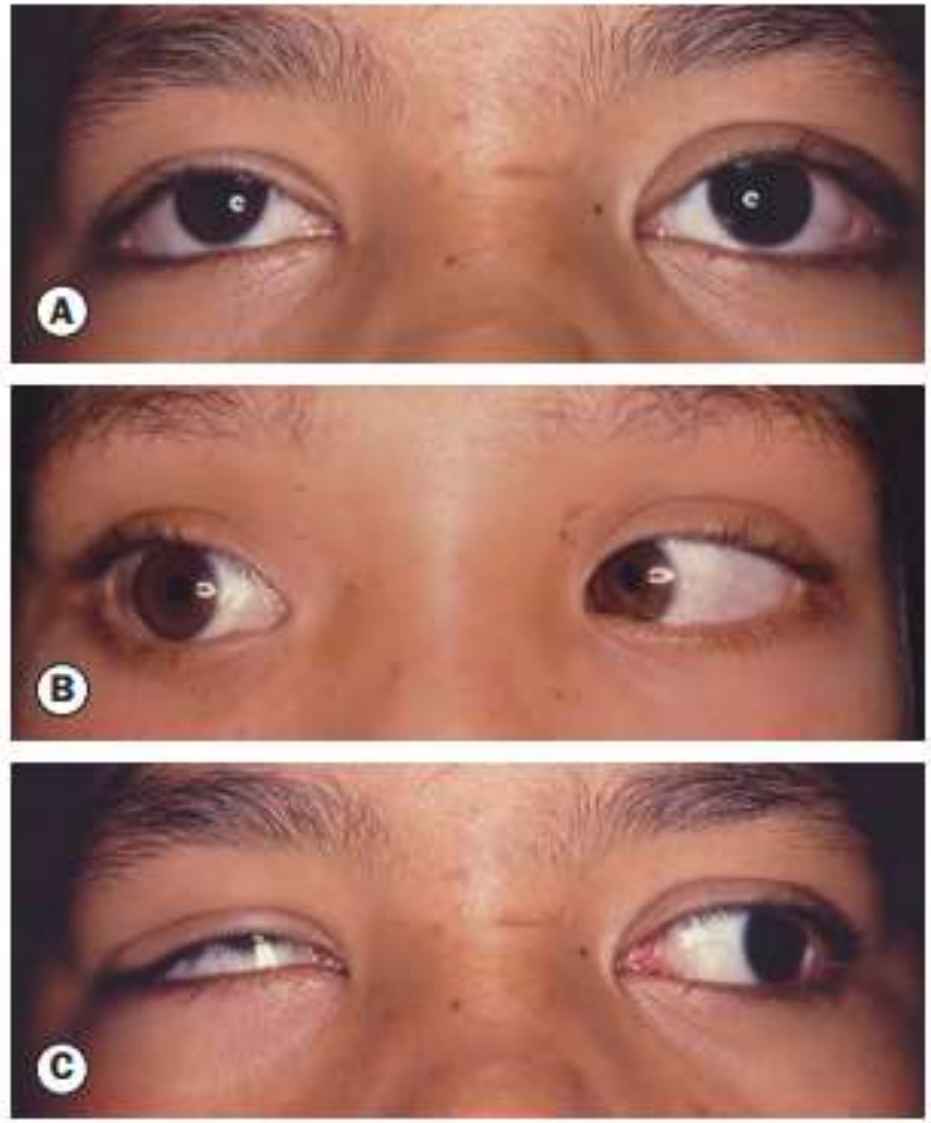

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