

# ***Infantile (Congenital) Esotropia***

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# Risk Factors

## Inheritance

- *Both genetic and environmental factors appear to play a role in the causation of esotropia.*
- *Large-scale studies have documented that 20% to 30% of children born to a strabismic parent will themselves develop strabismus.*
- *The inheritance of congenital esotropia remains undefined. Maumenee et al. in an analysis of a large group of families, concluded that the inheritance is consistent with a Mendelian model in which there is an admixture of primarily autosomal recessive cases, some dominant cases, and possibly nongenetic cases.*
- *Variable patterns of inheritance for infantile esotropia speak to the heterogeneity of this syndrom.*

# **Risk Factors**

## **Environmental Factors**

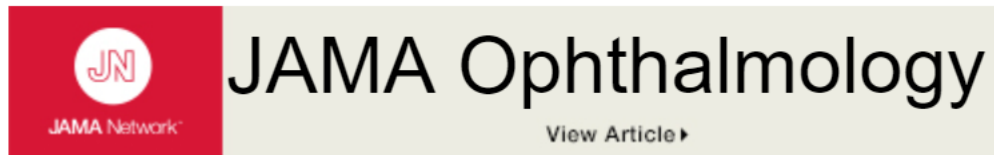
- *As for environmental factors, the prevalence of strabismus and amblyopia is substantially higher in low-birth-weight, premature infants or those who suffer perinatal hypoxia.*
- *Infants weighing less than 2500 g at birth have a prevalence of strabismus four times that of normal-weight infants.*
- *Infants born weighing less than 1500 g have a prevalence of amblyopia and strabismus seven times that of normal-weight term infant.*

# **Risk Factors**

## **Environmental Factors**

- *The increased risk of strabismus in these infants is probably due to the maldevelopment of binocular connections in the visual cortex.*
- *The striate cortex is susceptible to hypoxic injury because it has the highest neuron-to-glia ratio in the entire cerebrum and the highest regional cerebral glucose consumption.*
- *Maternal smoking as well as drug and alcohol abuse perturb cerebral development and are associated with increases in the risk of amblyopia or strabismus equivalent to those associated with prematurity or hypoxia.*

# Epidemiology



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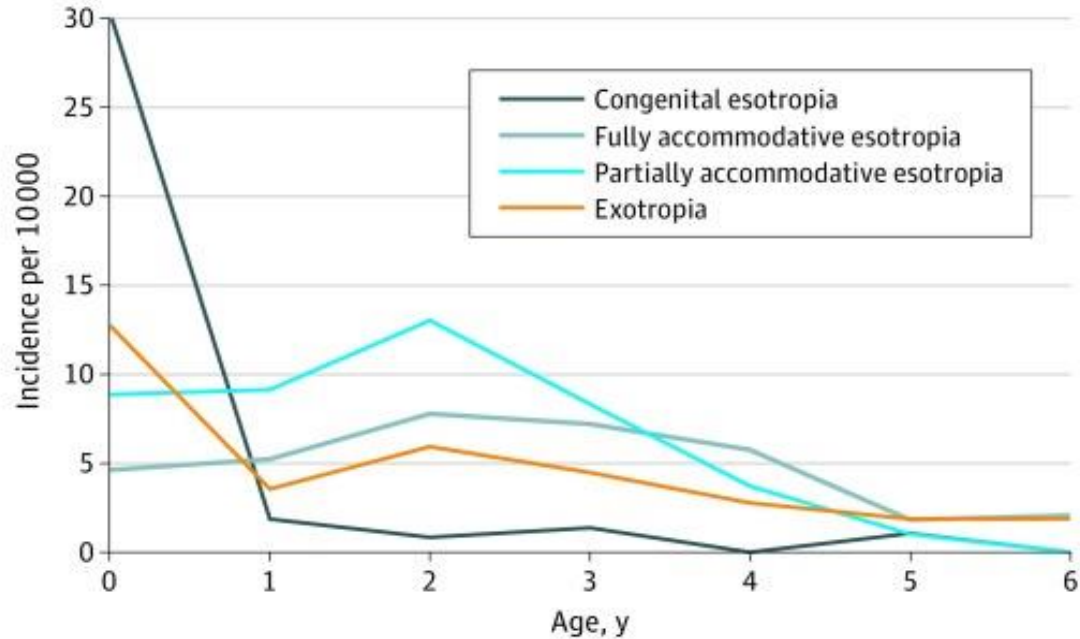
PMID: [28859196](#)

doi: [10.1001/jamaophthalmol.2017.3158](https://doi.org/10.1001/jamaophthalmol.2017.3158)

## Strabismus Incidence in a Danish Population-Based Cohort of Children

*This study cohort included 96 842 children born between 1996 and 2008. Overall, 1309 cases of strabismus were identified in the cohort. We found an overall cumulative strabismus incidence of 2.56% at 7 years. The overall incidence was similar among boys and girls. 216 participants (16.5%) had congenital esotropia, 177 (13.5%) had fully accommodative esotropia, 252 (19.3%) had partially accommodative esotropia, and 181 (13.8%) had exotropia. The esotropia:exotropia ratio was 5.4 : 1*

2017 Oct; 135(10): 1047–1053.



Person-years:	95 845	95 144	93 942	81 424	62 424	42 076	21 954
Cases:							
Congenital esotropia	291	18	8	11	0	5	0
Fully accommodative esotropia	45	50	73	59	36	8	5
Partially accommodative esotropia	85	87	123	68	23	4	0
Exotropia	122	34	56	37	17	8	4

# Clinical Characteristics

## Onset

- *The classic presentation is a infant who develops strabismus begins to manifest a chronic esodeviation of the visual axes at 2 to 4 months of age, Transient episodes of misalignment may precede this by several weeks.*
- *Variable, transient, intermittent strabismus is commonly observed in the first 2-3 months of life. Also, it is common to see both intermittent esotropia and exotropia in the same infant (termed ocular instability of infancy).*

*This condition should resolve by 3 months of age but can sometimes persist, especially in premature infants.*

*If an esotropia is present after age 2 months , and is constant, it is unlikely to resolve.*

# Clinical Characteristics

- *Classic infantile esotropia is constant and cosmetically obvious, typically exceeding 20 PD on corneal light reflex measurement.*
- *The magnitude of the strabismus may increase in the first few weeks or months of observation.*
- *the angle can vary depending on the level of attention. In general, the greater the degree of attentiveness, the larger the esotropia.*
- *The esotropia is comitant in right gaze, left gaze, upgaze, and downgaze.*



# Clinical Characteristics

## Refractive Error

- *Refraction using cycloplegia classically reveals a small amount (+ 2.00 D or less) of hypermetropia.*
- *The Congenital Esotropia Observational Study (CEOS) showed that mild to moderate hypermetropia was present in most patients, with about 20% being above 3.00 D, 12% above 4.00 D, and less than 10% being myopic. Similar results have been reported in other infantile esotropia series .*
- *It seems, from a review of the literature, that infants with esotropia have, on average, refractive errors similar to the normal age-matched population.*

# ***Clinical Characteristics***

- *Other variants include either a combination of refractive (hypermetropic) and "baseline" infantile esotropia or high accommodative convergence/accommodation ("high AC/A") esotropia with infantile esotropia.*
- *The combined refractive-infantile esotropes show a substantial reduction in the total angle of strabismus when wearing hypermetropic spectacles. Spectacles alone, however, seldom restore full orthotropia.*

# **Clinical Characteristics**

## **Cross-fixation**

- *The infant cross-fixates : so that the targets of interest presented to the left of the head elicit fixation with the right eye and targets presented to the right of the head elicit fixation with the left eye.*
- *The cross-fixation pattern may be so habitual as to obviate normal abduction of either eye under casual viewing conditions.*

# Clinical Characteristics

## *Ciancia's syndrome*

- *Ciancia's syndrome is a large-angle congenital esotropia with cross-fixation, and both eyes appear to be "stuck" in toward the nose.*

*It consists of the following characteristics:*

- (1) large-angle deviation (>60 PD)*
  - (2) bilateral limited abduction with intact abduction saccades*
  - (3) fixing eye in adduction*
  - (4) nystagmus on attempted abduction with no nystagmus in adduction*
  - (5) face turn to the side of the fixing eye*
- *the abduction deficit is most likely secondary to tight medial rectus muscles.*



**A**

***A : the patient is fixing the right eye, with face turn to the right***



**B**

***B : patient is fixing the left eye and has a face turn to the left.***

# Clinical Characteristics

## Amblyopia

- *The ability to alternate fixation, or hold fixation well with either eye, indicates equal vision. Strong fixation preference, on the other hand, indicates amblyopia of the nonpreferred eye, and should be treated by patching the preferred eye before strabismus surgery.*
- *In the CEOS, amblyopia was diagnosed in 19% of patients at the first visit (2 months of age) and doubled to 42% at subsequent visits (after 6 months of age).*
- *It is more likely, however, that this higher rate of amblyopia reflects the higher incidence of amblyopia having a longer duration of esotropia.*

# **Clinical Characteristics**

*Associated **motor phenomena** (usually present after 2 years of age)*

- 1. Inferior Oblique Overaction (IOOA) (60%)*
- 2. Dissociated Vertical Deviation (DVD) (40%)*
- 3. Latent Nystagmus (40%)*
- 4. Pursuit Asymmetry*

*Incomitance may be observed; the most common type is a V pattern, in which esotropia is greater in downgaze and less in upgaze. V-pattern infantile esotropia is commonly associated with overaction of the inferior oblique muscles.*

# Clinical Characteristics

## DVD

- *Dissociated vertical deviation (DVD) is characterized by an upward-directed slow movement of the nonfixing eye.*
- *There are 2 explanations for the origin of DVD. One theory is that it is the result of mechanisms to compensate for latent nystagmus, with the oblique muscles playing the principal role.*
- *An alternative theory suggests that deficient fusion allows the primitive dorsal light reflex, which is prominent in other species, to emerge.*
- *It is nearly always bilateral, but of differing magnitude in the two eyes.*
- *It is rarely detected in infants. Typically DVD appears in preschool-age and school-age children who have had horizontal muscle surgery to correct esotropia earlier in life.*



# Clinical Characteristics

## *latent nystagmus*

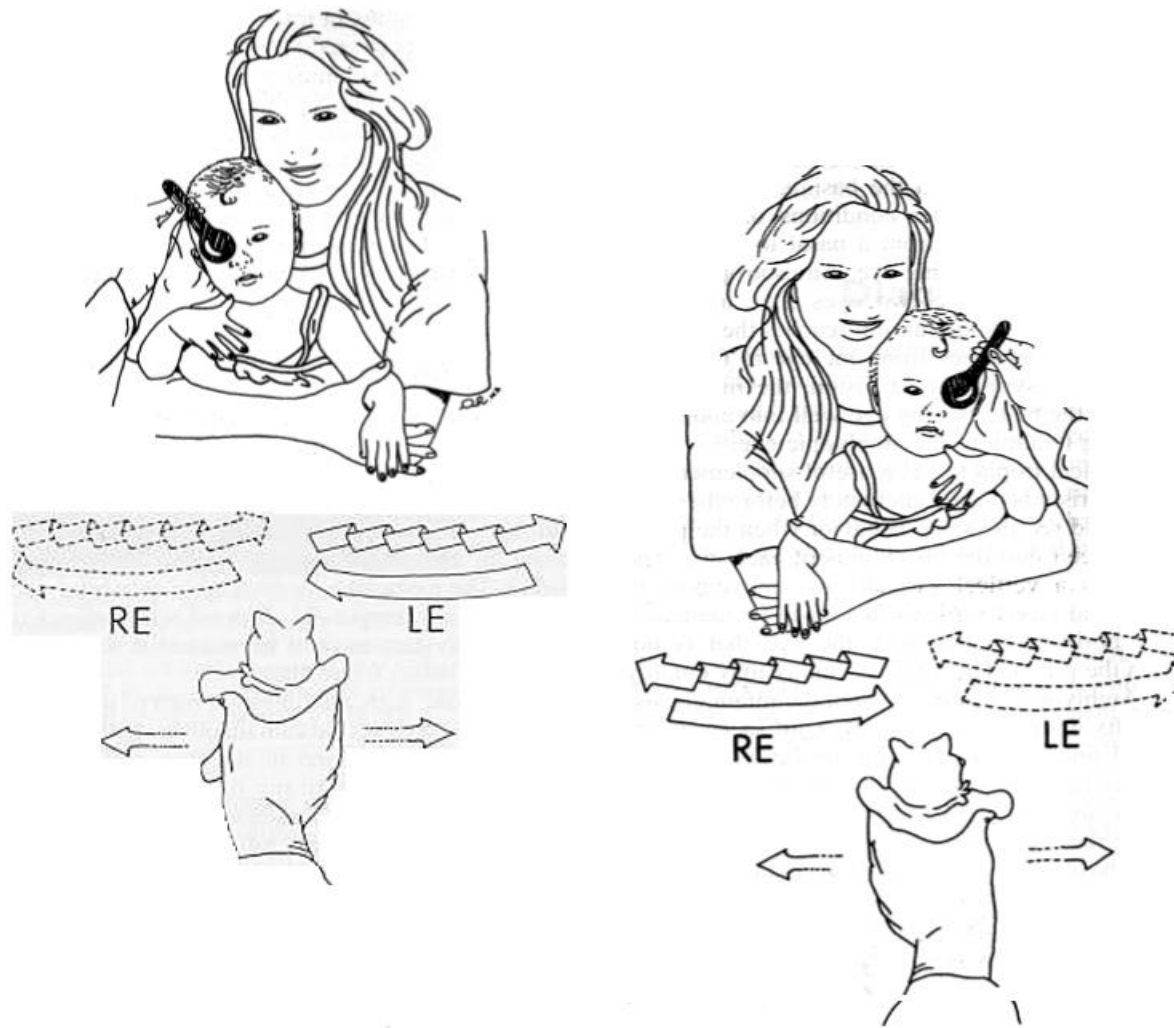
- *When one eye is covered and the infant is induced to attentively fix on a small accommodative target held at midline or in slight abduction, latent nystagmus may be observed.*
- *The nystagmus is easy to miss if the infant is inattentive or keeps the eye in the nasal orbit.*
- *In a smaller number (approximately 5%), latent nystagmus is severe enough to result in variants of the "nystagmus blockage" phenomenon, in which the angle of esotropia may increase greatly when the infant attentively fixates. In these cases, the infant uses convergence to dampen nystagmus and improve acuity.*
- *The presence of latent nystagmus and dissociated vertical deviation (DVD) in these cases allows the examiner to retrospectively date the onset of strabismus to the first year of life.*

# Clinical Characteristics

## Pursuit Asymmetry

- *Infants in whom normal binocularity fails to develop exhibit asymmetric horizontal pursuit.*
- *When one eye is occluded and a hand-held toy is moved from temporal to nasal before the fixing eye, pursuit is smooth.*
- *Pursuit is absent or jerky (cogwheel ) when the target moves nasal to temporal.*
- *The movements of the two eyes are conjugate, and the direction of the asymmetry reverses instantaneously with a change in the fixing eye, so that the direction of normal pursuit is always nasally directed with respect to the fixing eye.*

# *Clinical Characteristics*



Asymmetry of horizontal smooth pursuit evident during monocular viewing.

# Clinical Characteristics

## **Pursuit Asymmetry**

- *The asymmetry is identical to that seen transiently in all healthy young infants.*
- *It suggests delayed development of binocular connections in the visual cortex when it persists beyond age 6 months and is definitely pathologic when it persists beyond age 12 months.*
- *The pursuit asymmetry persists permanently into adulthood in individuals with an onset of strabismus in infancy, whether or not the strabismus is surgically corrected.*
- *Pursuit asymmetry in an adult indicates strabismus present from infancy and raises the strong possibility that the subnormal vision represents strabismic amblyopia.*

## *Differential Diagnosis*

**TABLE 7-2. Differential Diagnosis of Infantile Esotropia.**

Pseudo-esotropia  
Congenital esotropia  
Infantile accommodative esotropia  
Duane's syndrome  
Sensory esotropia  
Congenital sixth nerve palsy, usually transient  
Möbius syndrome  
Congenital fibrosis syndrome  
Infantile myasthenia gravis  
Neurological disease

# Differential Diagnosis

- *The differential diagnosis of infantile esotropia includes Duane's syndrome, congenital fibrosis syndrome, congenital sixth nerve palsy (Möbius syndrome associated with sixth nerve paresis), and infantile myasthenia gravis.*
- *These disorders all have limited abduction and, therefore, can be differentiated from infantile esotropia where the ductions should be full.*
- *This differentiation may be difficult in patients with large-angle infantile esotropia and tight medial rectus muscles. Even in these patients, however, vestibular stimulation by doll's head maneuver reveals full ductions and good abduction saccades.*

## *Differential Diagnosis*

Vestibular stimulation is best performed in infants by gently spinning the child



Infant is moved to the right, which stimulates eye movement to the left.

# Differential Diagnosis

- *Other diagnoses include infantile accommodative esotropia and pseudo-esotropia.*

## ***pseudo-esotropia :***

- *Pseudo-esotropia is a condition in which the eyes are orthotropic but appear to be crossed; this usually occurs in infants who have a wide nasal bridge with prominent epicanthal folds.*
- *Pseudo-esotropia usually resolves by 2 or 3 years of age because the epicanthal folds diminish as the bridge of the nose enlarges.*
- *Patients with a small interpupillary distance may also appear to be esotropic, especially when the eyes are in side gaze or are focusing at near.*
- *Children with pseudo-strabismus should have a full ocular examination. It is important to follow these children, as a small percentage will end up having a true esodeviation.*



## ***Differential Diagnosis***



**A**

***A : Note the large epicanthal folds giving the appearance of esotropia even though the eyes are well aligned***



**B**

***B : Pinching the epicanthal skin folds demonstrates the eyes are well aligned.***

## ***Differential Diagnosis***

- ***Infantile accommodative esotropia*** may be difficult to distinguish from infantile esotropia, it can occur in babies as young as 2 months of age.
- The key to the diagnosis of infantile accommodative esotropia is the presence of straight eyes for several months, followed by a variable small-angle esodeviation associated with hypermetropia of 3.00 D or more.
- These infants should be immediately treated with their full hypermetropic correction .

# *Systemic Association*

- *In most cases, congenital esotropia occurs as an isolated problem in an otherwise healthy child.*
- *Infantile esotropia occurs in up to 30% of children with neurologic and developmental problems, including cerebral palsy, hydrocephalus, and prematurity.*
- *Unlike the majority of infants with esotropia, more than 90% of those with constant exotropia have significant eye or brain abnormalities such as optic nerve hypoplasia, morning glory anomaly of the optic disc, retinoblastoma, microcephaly, infantile spasm, encephalomalacia.*
- *The ophthalmologist must be particularly diligent in ruling out neuro-ophthalmic abnormalities in any infant presenting with constant exotropia, as opposed to esotropia, in the first 12 months of life.*
- *This dictum does not apply to infants who display early-onset intermittent exotropia, nor does it apply to normal infants younger than 3 to 5 months of age who display a transient physiologic exodeviation in early infancy.*

## ***Systemic Association***

*Ophthalmology*. 2012 January

### **Congenital esotropia and the risk of mental illness by early adulthood**

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**Objective**—The purpose of this study is to investigate whether children with congenital esotropia are more likely than controls to develop mental illness by early adulthood.

**Conclusions**—Congenital esotropia, similar to those with intermittent exotropia or convergence insufficiency, increases the odds of developing mental illness by early adulthood 2.6 times compared to controls. The etiology of this association does not appear to be associated with premature birth.

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