

Pseudostrabismus



In this picture you the eyes appear crossing due to the skin folds that cover the medial corners of the eyes

Pseudostrabismus is a condition where the eyes are well aligned (orthotropic) but appear to be misaligned. This is essentially an optical illusion which is created by certain morphological features of the face although it can be associated with ocular/orbital disorders. The most common form of pseudostrabismus is pseudoesotropia where the eyes appear to be crossed. The other forms are pseudoexotropia, where eyes appear to be deviated outwards and pseudohypertropia where eyes appear to be vertically misaligned.

Prevalence

Pseudostrabismus, typically pseudoesotropia is a very common diagnosis in a busy pediatric ophthalmology practice although the true prevalence of this entity is unknown. Of note some patients with a diagnosis of pseudostrabismus can later present with eye misalignment. The incidence of manifest strabismus following pseudostrabismus diagnosis has been reported as high as 12% in a retrospective review of 83 patients diagnosed with pseudostrabismus.

Etiology

Pseudoesotropia

Pseudoesotropia is the most common type of pseudostrabismus and can be seen due to certain facial morphological features such as orientation, shape and size of the orbits, size and shape of globes, volume and viscosity of retrobulbar tissue all of which can create an illusion of misaligned eyes. Most commonly this occurs in infants who have a wide nasal bridge with prominent epicanthal folds (semilunar fold of skin at the medial canthus). Patients with small interpupillary distance may also appear to be esotropic. A negative angle kappa (angle formed by pupillary axis and visual axis at the pupil) where corneal light reflex appears to be on the temporal side of the pupillary center can simulate an esodeviation.

Pseudoexotropia

Like pseudoesotropia, certain morphological features of the face can result in a false appearance of eyes to be drifted outwards. Most commonly hypertelorism, which is widely set eyes, can result in pseudoexotropia. Traction of the retina resulting in pathologic ectopia of the macula temporally can cause a positive angle kappa resulting in nasal displacement of the light reflex on the cornea simulating a true exotropia. Pseudoexotropia from positive angle kappa is mostly seen in retinopathy of prematurity which results in temporal dragging of the macula, it can also be seen in ectopic macula resulting from toxocara retinal scars, high myopia or congenital retinal folds.

Pseudohypertropia

Facial asymmetry may create an appearance of vertically misaligned eyes where one eye appears to be higher than the other. Certain orbital tumors or trauma to the orbital floor can also in rare occasions create hypoglobus where the entire globe is higher/lower than the other side simulating a vertical misalignment.

Risk Factors

- Prematurity: Retinopathy of prematurity with temporal dragging of macula can result in positive angle kappa and pseudoexotropia
- Facial Morphology: Asian children have prominent epicanthal folds resulting in pseudoesotropia
- Orbital tumors: can result in pseudohypertropia
- Orbital trauma: can cause hypoglobus resulting in pseudohypertropia in some cases
- Chorioretinal Infections: can cause chorioretinal scarring with temporal dragging of the macula resulting in pseudoexotropia
- Vertical eyelid asymmetry due to various conditions- like Horner's syndrome, thyroid eye disease, trauma may cause pseudo vertical strabismus.

Diagnosis

History

Detailed history regarding birth weight, gestational age, the health of the child, history of prior procedures to treat retinopathy of prematurity may give diagnostic clues. History of first presentation aided by photographs of the child in the first few months of life can assist in documenting the onset, detecting the stability of the condition and confirming the diagnosis.

Physical examination

A good physical examination comprising of inspection of facial, orbital, ocular morphology with special attention to eyelid structure and its characteristics is imperative to diagnosing pseudostrabismus. In infants presence of an epicanthal fold should be noted. An epicanthus is a semilunar fold of skin running at the side of nose with its concavity directed toward the inner canthus. Hirschberg's light reflex test is the best means of estimating the relative position of the eyes

whereby the patient fixates at a penlight held at 2/3rd of a meter and the light reflex at the cornea is noted. A light reflex centered in the pupils of both eyes in general indicates no misalignment of the eyes. This test should be demonstrated to the parents so that they can check for the alignment of the eyes at home. This test is quick and easy however it will not diagnose a small angle strabismus. A detailed ocular motility exam comprising of cover-uncover and alternate cover test should be performed as the gold standard test for true strabismus. A cycloplegic refraction should be performed in every case of pseudostrabismus to rule out high hyperopia which may be a sign of intermittent esotropia in patients with accommodative strabismus which can sometimes be misdiagnosed as pseudostrabismus.

Symptoms

Pseudostrabismus in infants is typically noted by astute parents or by pediatricians during the well child exams, mostly crossing of eyes in cases of pseudoesotropia. Pseudoesotropia can be exaggerated when the child looks in side gazes due to the nasal sclera being buried in the epicanthal fold.

Clinical diagnosis

Clinical Diagnosis of Pseudostrabismus is reached after ruling out the presence of true manifest or intermittent strabismus.

Diagnostic procedures

Visual evaluation

Any patient that presents with pseudostrabismus should have a full age appropriate visual acuity assessment.

Motor evaluation

An appropriate motor evaluation should be performed to rule out any true strabismus. Hirschberg's light reflex test is the best mean of estimating the relative position of the eyes in an uncooperative infant. Cover/uncover testing is used to rule out the presence of true strabismus. It is necessary to test the function of each extraocular muscle (ductions and versions) and the patient's control over the deviation. Additional tests may include prism adaptation and diagnostic occlusion.

Sensory evaluation

Complete sensory evaluation in a preverbal child is difficult. Some signs may give a clue about the sensory development. Detection of fixation preference for one eye can be performed by the use of vertical prism test (10-prism diopter test).

Differential diagnosis

True Strabismus should always be ruled out with a thorough exam before diagnosing a patient with pseudostrabismus

Management

Once the diagnosis of pseudostrabismus is confirmed it is important to reassure the family and educate them about the signs of true strabismus in any patient with pseudostrabismus. Family should report back if any evidence of true strabismus is noted as it is important to diagnose a new onset manifest strabismus as early as possible in these patients which can be easily missed in view of a prior diagnosis of pseudostrabismus.

Medical follow up

Most pediatric ophthalmologists follow these patients in 6-12 months to rule out any evidence of true strabismus in future, especially if risk factors are identified like family history of hyperopia more than 2D.

Prognosis

The natural history of pseudostrabismus is variable, most cases of pseudoesotropia resolve by 2 or 3 years of age because the epicanthal folds diminish as the bridge of the nose enlarges. Pseudostrabismus secondary to positive or negative angle kappa typically persists in adulthood.

Additional Resources

- AAPOS Frequently asked questions about Pseudostrabismus
- Scott Larson, MD. Pseudostrabismus. American Association for Pediatric Ophthalmology & Strabismus.
<https://aapos.org/browse/glossary/entry?GlossaryKey=eafce745-dfbf-48e4-aac4-66b9b05868e8> Accessed July 01, 2019.

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