

Strabismus: - Symptoms, Pathophysiology, Management & Precautions

Dr. Jayantilal Shah¹, Dr. Shrikant Patel²

¹Associate Professor, Department of Ophthalmology, Gujarat Adani Institute of Medical Science, Bhuj, Gujarat

²Senior Lecturer, Department of Oral Pathology, Pacific Dental College & Hospital, Debari, Udaipur

Abstract: *Strabismus, more commonly known as cross-eyed or wall-eyed, is a vision condition in which a person cannot align both eyes simultaneously under normal conditions. One or both of the eyes may turn in, out, up or down. An eye turn may be constant (when the eye turns all of the time) or intermittent (turning only some of the time). Strabismus typically involves a lack of coordination between the extraocular muscles, which prevents directing the gaze of both eyes at once to the same point in space; it thus hampers proper binocular vision, and may affect depth perception adversely. Strabismus is primarily managed by ophthalmologists, optometrists, and orthoptists. Strabismus is present in about 4% of children. Treatment should be started as early as possible to ensure the development of the best possible visual acuity and stereopsis. Strabismus in children does not go away on its own and strabismus in adults is treatable, so strabismus treatment is necessary.*

Keywords: Strabismus, Etiology, Management.

1. Introduction

Whereas many animals have eyes located on either side of their head (such as horses, for example), the eyes of humans look forwards – in the same direction. When normal, the eyes move in a coordinated manner, so that the object being looked at is centered in each eye. Because the eyes are set a small distance apart, the image in each eye is slightly different. The brain fuses the images coming from both eyes to produce a three-dimensional image that has depth.

This three-dimensional vision, also known as stereoscopic or binocular vision, gives us depth perception. This allows us to judge distances more accurately, especially with objects close to us. Try to thread a needle with only one eye open and you will see the advantage of binocular vision! In order to achieve normal binocular vision, the eyes must see well, be aligned (looking in the same direction), and be focused properly on the same object. To maintain alignment, the eyes must also move in a coordinated manner, a process involving twelve different muscles (six in each eye). The four rectus muscles move the eyes up, down, to the right, and to the left, and the two oblique muscles have more complex actions, helping the eyes to look down and in (towards the tip of the nose) or up and in (towards the bridge of the nose).[1]

Strabismus is a visual disorder where the eyes are misaligned and point in different directions. This misalignment may be constantly present, or it may come and go. Sometimes, only one eye is affected — turning inward (esotropia), outward (exotropia) or downward — while the other eye is directed straight ahead. Strabismus can also be described by its cause. The 3 cranial nerves (III, IV, VI) responsible for eye movement can be weak or paralyzed and cause strabismus. Some examples of paralytic strabismus include third nerve palsy and superior oblique palsy.[2]

Strabismus prevents proper binocular vision and prevents both eyes from gazing the same point. Either peripheral

vision or side vision may be affected. A patient's perception of depth is distorted. Perception of depth is the ability to recognize the order of objects in three dimensions. Patients will also experience a limited field of view. Some common terms for strabismus are "cross eyed," which means that one or both eyes turn toward your child's nose or "wall eyed," which means one or both eyes turn out toward your child's ears.[3]

Etiology

There are 6 muscles that work together to move your child's eye. Strabismus can happen when those muscles do not work together. This may be caused by a problem with the muscles, nerves, or a problem in your child's brain. Most people with strabismus are born with it and it tends to run in families. Strabismus may also be caused by:[4]

- Eye or head injuries
- Diseases that affect the nerves or muscles such as cerebral palsy or Down syndrome
- Brain tumors

When your child's eyes do not work together to look at an object, your child's brain pays attention to the image from one eye and ignores the image from the other eye. This is called amblyopia or "lazy eye." If treatment does not take place early, the lazy eye may never see as well as the stronger eye. Sometimes the cause of strabismus is unknown.

Other disorders associated with strabismus in children include: Apert syndrome, Cerebral palsy, Congenital rubella, Hemangioma near the eye during infancy, Incontinentia pigmenti syndrome, Noonan syndrome, Prader-Willi syndrome, Retinopathy of prematurity, Retinoblastoma, Traumatic brain injury, Trisomy 18.

Strabismus that develops in adults can be caused by: Botulism, Diabetes (causes a condition known as acquired paralytic strabismus), Graves' disease, Guillain-Barré

syndrome, Injury to the eye, Shellfish poisoning, Stroke, Traumatic brain injury, Vision loss from any eye disease or injury.[5]

Risk Factor:[6]

Low birth weight (<1250 g), particularly premature infants who have developed retinopathy of prematurity, Family history of strabismus, Neuromuscular disorders (i.e. multiple sclerosis, myasthenia gravis, botulism), Congenital ocular abnormality, Tumours of the brain or eye (i.e. retinoblastoma), Cataracts, Head injury, Infections (i.e. meningitis, encephalitis, measles), Systemic conditions with vision-threatening ocular manifestations (i.e. pauciarticular juvenile rheumatoid arthritis, which can predispose to iritis and cataracts), Drugs and toxins (ie lead and heavy metals)

Types of strabismus

One eye can be deviated inwards (sometimes referred to as being 'cross-eyed'). This is called esotropia. One eye can be deviated outward (sometimes referred to as a 'wall eye'). This is called exotropia. One eye can be deviated vertically, either upwards (hypertropia) or downwards (hypotropia).[7]

Pathophysiology[8]

The extraocular muscles control the position of the eyes. Thus, a problem with the muscles or the nerves controlling them can cause paralytic strabismus. The extraocular muscles are controlled by cranial nerves III, IV, and VI. An impairment of cranial nerve III causes the associated eye to deviate down and out and may or may not affect the size of the pupil. Impairment of cranial nerve IV, which can be congenital, causes the eye to drift up and perhaps slightly inward. Sixth nerve palsy causes the eyes to deviate inward and has many causes due to the relatively long path of the nerve. Increased cranial pressure can compress the nerve as it runs between the clivus and brain stem. Also, if the doctor is not careful, twisting of the baby's neck during forceps delivery can damage cranial nerve VI.

Evidence indicates a cause for strabismus may lie with the input provided to the visual cortex. This allows for strabismus to occur without the direct impairment of any cranial nerves or extraocular muscles.

Strabismus may cause amblyopia due to the brain ignoring one eye. Amblyopia is the failure of one or both eyes to achieve normal visual acuity despite normal structural health. During the first seven to eight years of life, the brain learns how to interpret the signals that come from an eye through a process called visual development. Development may be interrupted by strabismus if the child always fixates with one eye and rarely or never fixates with the other. To avoid double vision, the signal from the deviated eye is suppressed, and the constant suppression of one eye causes a failure of the visual development in that eye.

Also, amblyopia may cause strabismus. If a great difference in clarity occurs between the images from the right and left eyes, input may be insufficient to correctly reposition the eyes. Other causes of a visual difference between right and

left eyes, such as asymmetrical cataracts, refractive error, or other eye disease, can also cause or worsen strabismus.

Accommodative esotropia is a form of strabismus caused by refractive error in one or both eyes. Due to the near triad, when a patient engages accommodation to focus on a near object, an increase in the signal sent by cranial nerve III to the medial rectus muscles results, drawing the eyes inward; this is called the accommodation reflex. If the accommodation needed is more than the usual amount, such as with people with significant hyperopia, the extra convergence can cause the eyes to cross.

Sign & Symptoms:

When observing a patient with strabismus, the misalignment of the eyes may be quite apparent. A patient with a constant eye turn of significant magnitude is very easy to notice. However, a small magnitude or intermittent strabismus can easily be missed upon casual observation. In any case, an eye care professional can conduct various tests, such as cover testing, to determine the full extent of the strabismus.

Symptoms of strabismus include double vision and/or eye strain. To avoid double vision, the brain may adapt by ignoring one eye. In this case, often no noticeable symptoms are seen other than a minor loss of depth perception. This deficit may not be noticeable in someone who has had strabismus since birth or early childhood, as they have likely learned to judge depth and distances using monocular cues. However, a constant unilateral strabismus causing constant suppression is a risk for amblyopia in children. Small-angle and intermittent strabismus are more likely to cause disruptive visual symptoms. In addition to headaches and eye strain, symptoms may include an inability to read comfortably, fatigue when reading, and unstable or "jittery" vision.[9]

The primary sign of strabismus is a visible misalignment of the eyes, with one eye turning in, out, up, down or at an oblique angle.

When the misalignment of the eyes is large and obvious, the strabismus is called "large-angle," referring to the angle of deviation between the line of sight of the straight eye and that of the misaligned eye. Less obvious eye turns are called small-angle strabismus.

Typically, constant large-angle strabismus does not cause symptoms such as eye strain and headaches because there is virtually no attempt by the brain to straighten the eyes. Because of this, large-angle strabismus usually causes severe amblyopia in the turned eye if left untreated.

Less noticeable cases of small-angle strabismus are more likely to cause disruptive visual symptoms, especially if the strabismus is intermittent or alternating. In addition to headaches and eye strain, symptoms may include an inability to read comfortably, fatigue when reading and unstable or "jittery" vision. If small-angle strabismus is constant and unilateral, it can lead to significant amblyopia in the misaligned eye.

Both large-angle and small-angle strabismus can be psychologically damaging and affect the self-esteem of children and adults with the condition, as it interferes with normal eye contact with others, often causing embarrassment and awkwardness.

Newborns often have intermittent crossed eyes due to incomplete vision development, but this frequently disappears as the infant grows and the visual system continues to mature. Most types of strabismus, however, do not disappear as a child grows.[10]

Routine children's eye exams are the best way to detect strabismus. Generally, the earlier strabismus is detected and treated following a child's eye exam, the more successful the outcome. Without treatment, your child may develop double vision, amblyopia or visual symptoms that could interfere with reading and classroom learning

Diagnosis[11]

During an eye examination, a test such as cover testing or the Hirschberg test is used in the diagnosis and measurement of strabismus and its effect on vision. Several classifications are made when diagnosing strabismus.

Latency

Strabismus can be manifest (-tropia) or latent (-phoria). A manifest deviation, or heterotropia, is present while the patient views a target binocularly, with no occlusion of either eye. The patient is unable to align the gaze of each eye to achieve fusion. A latent deviation, or heterophoria, is only present after binocular vision has been interrupted, typically by covering one eye. This type of patient can typically maintain fusion despite the misalignment that occurs when the positioning system is relaxed. Intermittent strabismus is a combination of both of these types, where the patient can achieve fusion, but occasionally or frequently falters to the point of a manifest deviation.

Onset

Strabismus may also be classified based on time of onset, either congenital, acquired, or secondary to another pathological process. Many infants are born with their eyes slightly misaligned, and this is typically outgrown by six to 12 months of age.^[23] Acquired and secondary strabismus develop later. The onset of accommodative esotropia, an overconvergence of the eyes due to the effort of accommodation, is mostly in early childhood. Acquired non-accommodative strabismus and secondary strabismus are developed after normal binocular vision has developed. In adults with previously normal alignment, the onset of strabismus usually results in double vision.

Any disease that causes vision loss may also cause strabismus. Sensory strabismus is strabismus due to vision loss or impairment, leading to horizontal, vertical or torsional misalignment or to a combination thereof, with the eye with poorer vision drifting slightly over time. Most often, the outcome is horizontal misalignment. Its direction depends on the patient age at which the damage occurs: patients whose vision is lost or impaired at birth are more likely to develop

esotropia, whereas patients with acquired vision loss or impairment mostly develop exotropia. In the extreme, complete blindness in one eye generally leads to the blind eye reverting to an anatomical position of rest.

Although many possible causes of strabismus are known, in many cases no specific cause can be identified. This is typically the case when strabismus is present since early childhood.

Results of a U.S. cohort study indicate that the incidence of adult-onset strabismus increases with age, especially after the sixth decade of life, and peaks in the eighth decade of life, and that the lifetime risk of being diagnosed with adult-onset strabismus is approximately 4%.

Laterality

Strabismus may be classified as unilateral if the one eye consistently deviates, or alternating if either of the eyes can be seen to deviate. Alternation of the strabismus may occur spontaneously, with or without subjective awareness of the alternation. Alternation may also be triggered by various tests during an eye exam.

Direction

Horizontal deviations are classified into two varieties. Eso describes inward or convergent deviations towards the midline. Exo describes outward or divergent misalignment. Vertical deviations are also classified into two varieties. Hyper is the term for an eye whose gaze is directed higher than the fellow eye while hypo refers to an eye whose gaze is directed lower. Cyclo refers to torsional strabismus, which occurs when the eyes rotate around the anterior-posterior axis to become misaligned and is quite rare.

Naming

The directional prefixes are combined with -tropia and -phoria to describe various types of strabismus. For example, a constant left hypertropia exists when a patient's left eye is always aimed higher than the right. A patient with an intermittent right esotropia has a right eye that occasionally drifts toward the patient's nose, but at other times is able to align with the gaze of the left eye. A patient with a mild exophoria can maintain fusion during normal circumstances, but when the system is disrupted, the relaxed posture of the eyes is slightly divergent.

Management

As with other binocular vision disorders, the primary therapeutic goal for those with strabismus is comfortable, single, clear, normal binocular vision at all distances and directions of gaze.

Whereas amblyopia (lazy eye), if minor and detected early, can often be corrected with use of an eye patch on the dominant eye and/or vision therapy, the use of eye patches is unlikely to change the angle of strabismus. Strabismus is usually treated with a combination of eyeglasses, vision therapy, and surgery, depending on the underlying reason for the misalignment. For parents it is important to know that strabismus surgery does not remove the need for a child to wear glasses.[12]

In cases of accommodative esotropia, the eyes turn inward due to the effort of focussing far-sighted eyes, and the treatment of this type of strabismus necessarily involves refractive correction, which is usually done via corrective glasses or contact lenses, and in these cases surgical alignment is considered only if such correction does not resolve the eye turn.

In case of strong anisometropia, contact lenses may be preferable to spectacles because they avoid the problem of visual disparities due to size differences (aniseikonia) which is otherwise caused by spectacles in which the refractive power is very different for the two eyes. In a few cases of strabismic children with anisometropic amblyopia, a balancing of the refractive error eyes via refractive surgery has been performed before strabismus surgery was undertaken.[13]

Strabismus surgery attempts to align the eyes by shortening, lengthening, or changing the position of one or more of the extraocular eye muscles. The procedure can typically be performed in about an hour, and requires about one or two weeks for recovery. Adjustable sutures may be used to permit refinement of the eye alignment in the early postoperative period.[14]

Orthoptics is the medical term for eye muscle training procedures, provided by orthoptists and/or optometrists, which address eye teaming and visual clarity (acuity) only. Technically, there are broad distinctions between Orthoptics and Vision Therapy (which includes Orthoptics). Orthoptics regards strabismus as an eye muscle problem and treatment is directed toward muscle strength. Optometrists who provide Vision Therapy look at the neurological control system of the eyes and thus treat the whole visual system (and whole person). Vision Therapy alters the entire nervous system and reflexive behavior, thus resulting in a lasting cure. In general, orthoptics is home-based therapy. In general, Vision Therapy is performed under supervision in an optometrist's office and home therapy is an adjunct. Recent scientific research has shown that office-based Vision Therapy with homework is more successful than home-based therapy alone.

Optometric Vision Therapy[15]

Optometric Vision Therapy is an individualized, supervised, non-surgical treatment program designed to correct eye movements and visual-motor deficiencies. Vision Therapy sessions include procedures designed to enhance the brain's ability to control:

- eye alignment,
- eye teaming,
- eye focusing abilities,
- eye movements, and/or
- visual processing.

Visual-motor skills and endurance are developed through the use of specialized computer and optical devices, including therapeutic lenses, prisms, and filters. During the final stages of therapy, the patient's newly acquired visual skills are reinforced and made automatic through repetition and by integration with motor and cognitive skills.

While Vision Therapy includes the [eye muscle training methods of orthoptics](#), it has advanced far beyond it to include training and rehabilitation of the eye-brain connections (neuroplasticity) involved in vision. Clinical and research developments in Vision Therapy are closely allied with developments in neuroscience and research continues.

In Vision Therapy programs, developmental optometrists look at the neurological control system and thus are treating the whole visual-motor system and altering reflexive behavior, which results in a lasting cure. Also, most optometrists rely on office based therapy, which they believe is more accurately performed and monitored.

Additional research by the National Eye Institute showed that older children with lazy eye (7 years - 17 years) improve significantly with therapy. If one considers the benefits of in-office therapy combined with home therapy and the likelihood that the older child will be more cooperative...it makes a case for more treatment of older children

Double vision can rarely result, especially immediately after the surgery, and vision loss is very rare. Glasses affect the position by changing the person's reaction to focusing. Prisms change the way light, and therefore images, strike the eye, simulating a change in the eye position.^[39]

Early treatment of strabismus in infancy may reduce the chance of developing amblyopia and depth perception problems. Most children eventually recover from amblyopia if they have had the benefit of patches and corrective glasses. Amblyopia has long been considered to remain permanent if not treated within a critical period, namely before the age of about seven years;^[23] however, recent discoveries give reason to challenge this view and to adapt the earlier notion of a critical period to account for stereopsis recovery in adults.

Eyes that remain misaligned can still develop visual problems. Although not a cure for strabismus, prism lenses can also be used to provide some temporary comfort for sufferers and to prevent double vision from occurring.

Botulinum toxin therapy is used for treating strabismus in certain circumstances. In 1989, the US FDA approved botulinum toxin type A (BT-A) as a treatment for strabismus in patients over 12 years old. Most commonly used in adults, the technique is also used for treating children, in particular children affected by infantile esotropia. The toxin is injected in the stronger muscle, causing temporary and partial paralysis. The treatment may need to be repeated three to four months later once the paralysis wears off. Common side effects are double vision, droopy eyelid, overcorrection, and no effect. The side effects typically resolve also within three to four months.[16]

Prognosis[17]

With early detection, the prognosis of strabismus is excellent. Thus, it is extremely important to know when to refer a child to an ophthalmologist. Under conditions of accurate diagnosis and administering proper treatment before the ages of 6 years the outlook for children diagnosed with strabismus has been positive. Unfortunately, however, once a child reaches the age of 8 to 10 years, treatment of strabismus has

been found to be unsuccessful and can result in permanent decrease in vision. When strabismus is congenital or develops in infancy, it can cause amblyopia, in which the brain ignores input from the deviated eye. Even with therapy for amblyopia, stereoblindness may occur. The appearance of strabismus may also be a cosmetic problem. One study reported 85% of adult strabismus patients "reported that they had problems with work, school, and sports because of their strabismus". The same study also reported 70% said strabismus "had a negative effect on their self-image". After surgery, the squint can return, so a second operation is sometimes required to straighten the eyes.

[17] Flom, M.C., *The prognosis in strabismus*. American journal of optometry and archives of American Academy of Optometry, 1958. **35**(10): p. 509-514.

References

- [1] Helveston, E.M., *Understanding, detecting, and managing strabismus*. Community Eye Health, 2010. **23**(72): p. 12.
- [2] Guyton, D.L., *Ocular torsion reveals the mechanisms of cyclovertical strabismus The Weisenfeld Lecture*. Investigative ophthalmology & visual science, 2008. **49**(3): p. 847-857.
- [3] Katzin, H.M. and G. Wilson, *Strabismus in childhood*. 1968: CV Mosby Co.
- [4] Leigh, R.J. and D.S. Zee, *The neurology of eye movements*. 2015: Oxford University Press.
- [5] Paul, T.O. and L.K. Hardage, *The heritability of strabismus*. Ophthalmic genetics, 1994. **15**(1): p. 1-18.
- [6] Chew, E., et al., *Risk factors for esotropia and exotropia*. Archives of ophthalmology, 1994. **112**(10): p. 1349-1355.
- [7] Matsuo, T., T. Yamane, and H. Ohtsuki, *Heredity versus abnormalities in pregnancy and delivery as risk factors for different types of comitant strabismus*. Journal of pediatric ophthalmology and strabismus, 2001. **38**(2): p. 78.
- [8] Emmett T. Cunningham, P.R.-E., *Vaughan & Asbury's general ophthalmology*. . 18th ed., Medica: McGraw-Hill.
- [9] Olson, J.H., et al., *Congenital esotropia and the risk of mental illness by early adulthood*. Ophthalmology, 2012. **119**(1): p. 145-149.
- [10] Beauchamp, G.R., et al., *The utility of strabismus in adults*. Transactions of the American Ophthalmological Society, 2005. **103**: p. 164.
- [11] Martinez-Thompson, J.M., et al., *Incidence, types, and lifetime risk of adult-onset strabismus*. Ophthalmology, 2014. **121**(4): p. 877-882.
- [12] Yorston, D.H. and D.M. McGavin, *Ophthalmology in the Tropics and Subtropics*. Manson's tropical diseases. Amsterdam: Saunders Elsevier, 2009: p. 283-332.
- [13] Finlay, A.L., *Binocular vision and refractive surgery*. Contact Lens and Anterior Eye, 2007. **30**(2): p. 76-83.
- [14] Derner, J.L., J.M. Miller, and V. Poukens, *Surgical implications of the rectus extraocular muscle pulleys*. J Pediatr Ophthalmol Strabismus, 1996. **33**: p. 208-218.
- [15] Yeung, J., *Management of Strabismus*. Medical Bulletin, Hong Kong Medical Society, 2010.
- [16] Biglan, A.W., et al., *Management of strabismus with botulinum A toxin*. Ophthalmology, 1989. **96**(7): p. 935-943.