Correction of Exotropia with A-or V-Patterns

Lamiaa Ahmed Attia El-aidy, Yasser M. Ibrahim, Mohamed A. Elmarakby, Manar A. Ghali

Ophthalmology Department, Faculty of Medicine - Zagazig University, Egypt

Corresponding author: Lamiaa Ahmed Attia El-aidy

E-mail: Lelaidy14@gmail.com, Laelaidy@medicine.zu.edu.eg

Conflict of interest: None declared.

Funding: No funding sources

Abstract

Pattern strabismus is usually accompanied by cyclotorsion. The patient also may attain an abnormal head posture to maintain binocular fusion. Head position will be toward the direction of maximum deviation (role of thumb). For example, V-pattern esotropia and A-pattern exotropia are associated by chin depression. However, chin elevation may occur in V-pattern exotropia and A-pattern esotropia to bring the eyes downward where the deviation is least. Vertically incomitant pattern strabismus is used to describe the type of strabismus wherein the amount of horizontal deviation changes during the excursion of the eye from upgaze to downgaze. It is a common association with many types of strabismus, especially infantile esotropia and intermittent exotropia. Several patterns have been described for the type of vertical incomitance observed (eg, “A” or “V” patterns), depending upon the relative increase or decrease in the horizontal deviation during the vertical eye movement. The pathophysiology of this phenomenon is multifactorial and has been attributed to factors including oblique muscle dysfunction, horizontal or vertical recti anomaly, displacement of muscle pulleys, and orbital anomalies. The identification of the pattern and its underlying mechanism is essential to plan a proper surgical management in strabismus.

Keywords: exotropia, A-or V-patterns

Tob Regul Sci. ™ 2023; 9 (1): 8393 - 8402

DOI: doi.org/10.18001 /TRS.9.1.592

Introduction

Twenty to 50% of patients with horizontal deviations have also incomitant ocular deviations in the form of A- or V-pattern. A-pattern is defined when the difference in horizontal deviation between upgaze and downgaze is equal or more than 10 prism diopters, with more convergence or less divergence upwards. V-pattern has a difference of 15 prism diopters or more, with more divergence or less convergence upwards. Normally the eyes are more convergent downwards which is the reading position, so a larger V-pattern position is required to be considered pathological. [1] V-pattern is the most common associated pattern. [2] Pattern deviation can occur in some cases without strabismus in primary position. [3]

Presentation:

Pattern strabismus is usually accompanied by cyclotorsion. The patient also may attain an abnormal head posture to maintain binocular fusion. Head position will be toward the direction of maximum deviation (role of thumb). For example, V-pattern esotropia and A-pattern exotropia are associated by chin depression. However, chin elevation may occur in V-pattern exotropia and A-pattern esotropia to bring the eyes downward where the deviation is least. [4, 5, 2]

If deviation is greater in downgaze the patient usually complains of diplopia and difficulty in reading. [4] Some patients with A-pattern exotropia become symptomatic only after becoming presbyopes and need bifocal glasses. They experience diplopia in downgaze as they cannot keep fixation through the near add segment. If the patient is orthophoric in primary position, management can be done by using monofocal glasses for reading or bifocal, but with a higher position of the near add segment. [6] In general, presentation of pattern strabismus usually depends on the angle of horizontal deviation in primary position. As if the angle is large, fusion will be poor, no head position is acquired. If the angle is small, the patient will be presented by abnormal head posture in order to maintain fusion (e.g., chin elevation or depression in V- or A-pattern respectively). [6]

Assessment:

A- or V-pattern could be diagnosed by measuring horizontal deviation in primary position, upgaze and downgaze by prism cover and alternate cover test. [1]

B- Measurements of upgaze and downgaze should be taken 25-30° from midline, wearing full optical correction, looking at a far target to avoid near reflex. [6]

Detection of abnormal head position which may accompany pattern deviation. [2]

Detection of overelevation or overdepression in adduction to detect the degree of oblique muscle overaction. [6]

Objective assessment of ocular torsion by indirect ophthalmoscope should be done. [6] Normally the fovea is slightly below the horizontal raphe by about 0.3-disc diameter. In excyclotorsion it takes a lower position and the reverse in incyclotorsion where the fovea is higher in position. Ocular torsion is a sign of oblique muscles overaction. [2]

Orbital imaging may be done in patients with craniofacial abnormalities and in cases with no obvious cause. [2]

V-pattern subtypes:

1- Y-pattern: in which divergence is more in upgaze, with little difference in horizontal deviation between primary position and downgaze. It occurs mainly in IOOA associated with infantile esotropia or less commonly intermittent exotropia. Other conditions in which Y-pattern may occur as in Brown's syndrome, Duane syndrome with upshooting. [7] In this type, binocular vision is usually preserved more than other alphabetical types as the deviation is only occurring in upgaze. [8]

2- Pseudo IOOA with Y-pattern or less commonly V-pattern: This may occur secondary to bilateral aberrant innervation to lateral recti muscles or heterotopia of SO muscle pulley. Pseudo IOOA can be differentiated from true cases by: a) Absence of overelevation on adduction.

b) No torsion by fundus examination.

c) No accompanying SO underaction

d) By asking the patient to make quick saccades from downgaze to primary position, there will be rapid exotropia followed by inward corrective movement. This occurs in cases of aberrant innervation to lateral recti. Treatment is done only if there is deviation in primary position which requires treatment, in this case pattern can be collapsed by doing supraplacement of lateral recti in addition to conventional horizontal surgery. [6] Pseudo V-pattern esotropia may occur in patients with uncorrected accommodative esotropia with more deviation in primary position and downgaze (reading position), can be detected by collapsing of the pattern after good correction of the refractive error. [2]

3- Arrow pattern: with more convergence in downgaze than in primary position. It occurs in bilateral superior oblique muscle palsy and characterized by adduction and extorsion in downgaze by the unopposed action of inferior rectus muscles. [7]

A-pattern subtypes:

1- Lambda (ƛ) pattern: in which divergence is more in downgaze, with little difference in horizontal deviation between primary position and upgaze. It occurs mainly in bilateral superior oblique overaction. It may also occur in cases of overrecessed or lost inferior rectus muscle with secondary superior oblique overaction. [7]

A-pattern exotropia with incyclotorsion occurs usually after bilateral inferior rectus recession in patients with Graves ophthalmopathy. [1]

Inferior oblique muscle palsy causes more convergence in upgaze. [7]

2- X-pattern: Divergence occurs both in up- and downgazes. It occurs more frequently in patients with long-standing exotropia due to tight lateral recti muscles. Also occurs in Duane's syndrome, mostly type 3 "limitation of both adduction and abduction", with upshoots and downshoots occurring on attempted adduction. [7]

Superior and inferior obliques overaction also has been recorded in some cases with X-pattern. [2]

3- Diamond pattern: with convergence in both up and downgazes, may be secondary to tight medial recti. [2]

Etiology of A- or V-pattern:

Exact mechanisms are not well defined. However, possible etiologies may be mechanical (peripheral) or neural (central). [4]

- **Oblique muscle dysfunction:**

  Primary oblique muscles overaction is commonly associated with A- or V-pattern strabismus. [7] As the tertiary action of oblique muscles is abduction, superior oblique muscle overaction is associated with more abduction in downgaze and therefore causes A-pattern. On the other hand, inferior oblique overaction causes V-pattern as it causes more abduction in upgaze. [1]

  Another explanation is intorsion of the eye in SOOA is accompanied by lateral displacement of inferior rectus insertion, thus decreasing its adduction force in downgaze. [9],

  Primary position superior oblique overaction is associated with incyclotorsion. While inferior oblique overaction is associated with excyclotorsion. [9]

- **Heterotopic extraocular muscle pulleys:**

  New evidence based on MRI-imaging studies suggests that anatomical abnormalities of the extraocular muscle sleeves or pulleys may be present in cases previously diagnosed as having oblique muscle overaction. This anatomical malposition leads to overelevation or overdepression of the adducting eye simulating oblique muscle overaction. [10, 11, 9]

  Displacement of muscle pulley perpendicular to the plane of muscle action leads to vertical incomitant strabismus like those of oblique muscle overaction. (e.g., horizontal displacement of the pulley of vertical recti muscle, or vertical displacement of horizontal recti muscle). It's common in patients with connective tissue diseases. [12]

- **Horizontal Rectus muscle dysfunction:** This theory was considered in the early beginning of the description of alphabetical patterns. It stated that some sort of over or under activity of horizontal recti muscles is responsible for horizontal deviation in up- or down gaze. This theory lacks strong evidence to prove abnormal innervation to horizontal muscles in different gaze directions. [8]

  For example, underactivity of both medial recti leading to weak convergence downwards with development of A-pattern exotropia, and overaction of both medial recti results in V-pattern esotropia. Also, bilateral underactivity of lateral recti muscle results in weak divergence upwards with the development of A-pattern esotropia, and overactivity of lateral recti results in V-pattern exotropia. [5]

- **Vertical rectus muscle dysfunction:**

  Brown adopted this theory. It stated that A and V patterns are secondary to primary anomalies in the vertical recti muscle which are responsible for adduction in their tertiary action. The explanation that is in primary superior rectus underaction there is secondary overaction of IO muscle (its yoke muscle) which leads to more abduction in upgaze. Also, there is secondary underacting SO and overacting IR which leads to more adduction in downgaze and a net result of V-pattern strabismus. However, this theory doesn't explain the occurrence of pattern in normal functioning vertical recti muscles. Therefore, this theory doesn't gain so much acceptance. [8]

  Inferior rectus underaction results in poor adduction on downgaze and A-pattern. [5]

- **Bony orbital (craniofacial) abnormalities:**

  As in hydrocephalus, bony orbital abnormalities may cause anterior malposition of superior oblique pulley (trochlea) with stretching of SO tendon and mechanical SOOA. [9]

  Others such as plagiocephaly, craniofacial abnormalities (e.g., upslanting and downslanting of palpebral fissure) [12]

  In plagiocephaly, there is posterior malposition of the trochlea causing desagittalization of the oblique muscles (become more parallel to the coronal plane), resulting in pseudo SO muscle palsy and V-pattern strabismus. [2]

  Upslanting is frequently associated with A-pattern esotropia and inferior oblique underaction. While downslanting is associated with V-pattern exotropia and inferior oblique overaction. [6]

  Cases with shallow orbits usually result in V-pattern strabismus, may be due to an increased angle between the inferior oblique muscle axis and visual axis with more abduction on upgaze. [5] Craniosynostosis syndromes are associated with orbital rotation. For example, in Crouzon syndrome, there is excyclorotation of the orbit with an abnormal higher position of the medial rectus, resulting in more elevation on adduction and V-pattern strabismus (Pseudo or apparent IOOA). [5]

- **Heterotopic extraocular muscle insertion and cyclotorsion:**

  V-pattern strabismus is found in patients with a higher position of MR insertion and a lower position of LR insertion. This causes more abduction of LR on upgaze and more adduction of MR on downgaze. The opposite is seen on some cases of A-pattern.

  This malpositioning may be true or apparent which is caused by cyclotorsion of the whole globe caused by abnormal overaction of oblique muscles.

  The first type is usually corrected by surgical correction of recti muscle position, while the second type is corrected after appropriate surgical operation on oblique muscles. [8]

  Abnormal neurological connectivity which leads to abnormal directions of the pulling force of the extraocular muscles. It's often the cause in infants with pattern strabismus. [12]

- **Abnormal fusion and ocular torsion:**

  Early loss of binocular fusion can lead to ocular torsion in the non-fixating eye, which in turn results in a change in the direction of pull of recti muscle simulating oblique muscle overaction. For example, in V-pattern strabismus there is excyclotorsion in both eyes, so medial rectus acts as a partial elevator and lateral rectus acts as a partial depressor, thus simulating inferior oblique overaction in sidegazes. [12]

  Also, excyclotorsion makes superior rectus act as a partial abductor and the inferior rectus as a partial adductor. The same occurs in incyclotorsion with A-pattern strabismus. [2, 13] However, Kushner believes that the role of torsion is minimal in pattern strabismus and is mainly secondary to oblique muscle overaction as: [13]

a) The rise or fall of the eye in sidegazes is curvilinear rather than linear. If the cause is secondary to changing in the pulling forces of recti muscles, it's expected to be linear which is not true.

b) Objective excyclotorsion is present in infants with esotropia before V-pattern strabismus or overelevation in adduction are manifest. If torsion is the main cause, pattern strabismus will occur simultaneously not sequentially.

c) Vertical transposition of recti corrects pattern strabismus successfully but worsens the underlying torsion on the other hand.

d) Surgery for correction of torsion such as Harada-Ito operation corrects only excyclotorsion without affecting pattern deviation.

- **Abnormal supranuclear control:** It is believed to be secondary to abnormal circuits at the level of saccadic eye movement center in brainstem (parapontine reticular formation, PPRF) and vertical gaze center. Theories are under research and based on animal studies in which either early loss of binocular vision is made by prism goggle or early vision deprivation is made by lid suturing. [12]
- **Abnormal vestibular function:**

  Primary SOOA is usually associated with neurological diseases. It is believed to be secondary to abnormal balance between anterior and posterior semicircular canals. They described this type of A-pattern as a special type of bilateral skew deviation. [9, 2]

  Primary IOOA occurring in infants with congenital esotropia is supposed to be due to early loss of single binocular vision. This causes imbalance of central vestibular inhibitory impulses to anterior canals which leads to increased impulses toward muscles of upgaze (inferior oblique and superior rectus) and V-pattern strabismus. [9]

- **Iatrogenic pattern strabismus:** It can occur after surgery for strabismus as in:

  a) Anterior transposition of inferior obliques in antielevation syndrome can cause a picture like IOOA with overelevation in adduction.

  b) Bilateral inferior rectus recession in thyroid eye disease can cause A-pattern strabismus due to weakening of the adduction force of the recessed inferior recti when looking inferiorly, also by overacting superior obliques.

  c) Overcorrection after surgery for A- or V-strabismus.[13]

Pattern exotropia

Surgical treatment of pattern strabismus

Indications for surgery: [6]

1- Significant head posturing to maintain fusion (chin up or down) secondary to pattern strabismus.
2- Children undergoing surgery for horizontal deviation with significant A- or V-pattern to maintain binocular vision if both eyes have good vision.,

N.B: In dense amblyopia, pattern is usually ignored.

If deviation is mainly affecting primary position and downgaze.

N.B: Asymptomatic Y-pattern can be ignored, especially if its correction will be at the expense of either primary position or downgaze.

3- Cosmetically unaccepted strabismus with pattern deviation.

We can divide the surgical plan in two main items:

Cases with oblique muscle overaction: Operation should be symmetrical in both eyes; otherwise, vertical deviation can occur. [6]

A) V-pattern: in cases of inferior oblique overaction (+2 or more), inferior oblique weakening procedures (recession, myectomy or disinsertion) are done (usually bilateral), oblique muscle surgery doesn't affect the amount of horizontal muscle correction. [14, 5]
B) Bilateral IO weakening procedures usually corrects 15-25 diopters of exotropia in upgaze with no effect on horizontal deviation in primary position or downgaze. However, some increase in divergence may occur in downgaze (10-15 prisms) secondary to increased action of SO secondary to IO weakening. [6]
C) A-pattern: with significant superior oblique overaction (+3 or +4), superior oblique weakening can be done. Tenotomy is usually unpredictable surgery and can lead to postoperative superior oblique palsy and diplopia (i.e., due to loss of bifoveal fixation secondary to muscle torsion). Predictable procedures such as superior oblique tendon expander and split tendon elongation are recommended, or otherwise horizontal surgery with transposition is preferred. [14]
D) Nasal tenotomy of SO tendon is more powerful than temporal tenotomy as it corrects up to 40 prisms of exotropia in downgaze. Posterior tenectomy can correct up to 15-20 prisms. A greater effect than posterior tenectomy can be obtained by complete muscle disinsertion, tendon expansion or graded recession. [6]
E) Posterior tenectomy weakens up to the posterior 90% of SO muscle fibers which are responsible for depression, sparing the anterior ones which serve for intorsion. [5]
F) A-pattern secondary to pulley heterotopia is diagnosed by good orbital imaging and can be treated surgically by stabilization or repositioning of muscle pulley. [6]

Cases without significant oblique muscle overaction:

Is treated with vertical transposition of horizontal recti muscles' insertion. The transposition is in the direction of the apex of the pattern in case of medial rectus transposition and the reverse for lateral rectus muscle. For example: infraplacement of medial rectus muscle in V-pattern strabismus and supraplacement of the lateral rectus muscle. [1]

Vertical transposition should be symmetrical and is accompanied by the usual recession, resection addressed for horizontal deviation. Measurements for horizontal deviation aren't affected by vertical transposition. [6]

Transposition of half tendon width (about 5 mm) for pattern from 15-20 prisms. If the pattern is more than 20 prisms, full tendon width is required after exclusion of oblique muscle overaction. Oblique muscles weakening procedures can be accompanied by recession of the vertical rectus

agonist muscle in large pattern angle (i.e., SO posterior tenectomy with inferior rectus recession). [6]

Also, in large pattern angle horizontal transposition of vertical recti can be done. [15]

Rectus muscle transposition leads to three major sequelae: [13]

A) Decrease in the primary action of the muscle in the direction of transposition and increase in the action in the opposite direction. (e.g., decrease in the adduction force of medial rectus muscle inferiorly, in case of its inferior transposition in V-pattern strabismus, therefore improving the convergence occurring in downgaze).

B) A new force of action is added to the muscle in the direction of transposition (e.g., depression force if medial rectus is transposed inferiorly).

C) Cyclotorsion in the direction towards the original insertion. (e.g., excyclotorsion occurs with inferior displacement of medial rectus muscle or supraplacement of lateral rectus muscle, excyclotorsion occurs also with nasal displacement of superior rectus or temporal displacement of inferior rectus). This point should be taken into consideration in case of recti muscle transposition to be aware of this torsional side effect. For example, excyclotorsion could be worsened in case of inferior medial rectus transposition to treat V-pattern strabismus secondary to IOOA. As IOOA is already accompanied by excyclotorsion.

It's worthy to note that this torsional effect is usually not noticeable in vertical transposition of horizontal muscles in contrast with vertical recti transposition. It can be explained partly due to poor bifoveal fusion in patients with horizontal strabismus before operation and the change in torsion is usually small (mean 6.4°). [1]

However, if there is significant oblique muscle overaction, transposition of recti muscles is not recommended. As objective torsion which already presents with obliques overaction can be exaggerated to symptomatic subjective torsion with the torsional vector created by recti muscles transposition. [6]

Slanting of the muscle insertion:

In case of pattern strabismus without obliques overaction. Slanting the insertion of recti muscles after recession or resection can be done. In V-pattern esotropia, slanting is made by more recession of the inferior part of medial rectus muscle to increase the effect of recession on downgaze. The reverse is done in A-esotropia. By the same principle in V-exotropia, recession is more in the upper part of lateral recti muscles to maximize the effect of the operation on upgaze. And the reverse is done in A-exotropia. [2, 8]

Lee YB et al found that only horizontal muscle surgery can correct a small angle of A- or V-pattern (less than 10 and 15 degrees respectively) associated with intermittent exotropia, without need for additional oblique muscle surgery or horizontal recti transposition. The study was retrospective and was carried on 58 patients (12 with A-pattern and 46 with V).

Other conditions in which this operation is useful

1. Double elevator palsy:

a) With positive forced duction (couldn't be elevated passively):

• Inferior rectus fibrosis: usually congenital or secondary to Graves ophthalmopathy in adults. May be associated with vertical retraction syndrome (retraction and esotropia on attempted elevation). Treatment: inferior rectus recession +/- lengthening in severe cases +/- inferior conjunctival recession.

• Orbital floor fracture with muscle entrapment: Treatment: early or late.

• Brown syndrome with superior rectus weakness:

b) With negative forced duction:

• Primary: neurological causes, may be associated with true or pseudoptosis. Treated by horizontal muscle transposition.

• Secondary: to certain ptosis surgery Plication with transposition versus with resection There are many approaches to manage cases of both horizontal and vertical deviation. Some surgeons can operate on both horizontal and vertical recti muscles. Also, horizontal muscle transposition can be done accompanied by usual resection or recession. [16]

Resection is done to strengthen the muscle action. However, nowadays plication gains much popularity due to preservation of vascular supply, protection against anterior segment ischemia, decreasing risk for lost muscle and being reversible in the early postoperative period. Efficacy of plication is comparable to resection postoperatively and with long-term follow-up. The amount of plication is done the same as calculated for resection. Adjustable sutures can be used with plication. [17, 16]

Transposition of plicated horizontal muscle is a new technique which offers both the advantages of plication and transposition. It is indicated in cases with horizontal strabismus associated with pattern deviation, not secondary to oblique muscle overaction. [15]

Gokyigit B et al announced this technique in 2019 and published another paper in 2021 showing the long-term follow-up of 17 cases who have done this technique which was named "sliding shape extraocular muscle transposition with plication procedure (SSMTP). They prescribed this technique to be safe, effective with good results on long-term follow-up.

Also, Shah PR & Pihlblad MS published a successful 4 cases of complex horizontal and vertical squint, who was treated with unilateral recession/plication with vertical transposition.

In our study we choose only cases with intermittent exotropia and pattern deviation to deal with by doing medial rectus muscle plication and transposition and comparing it with a standard technique in which resection is done instead of plication followed by either superior or inferior transposition according to type of pattern (A or V).

References:

1. Straight, S.M., Bahl, R.S. (2016). A- and V-Pattern Strabismus. In: Traboulsi, E., Utz, V. (eds) Practical Management of Pediatric Ocular Disorders and Strabismus. Springer, New York, NY.

2. Saxena, R., & Dhiman, R. (2020, December 28). Pattern strabismus. Retrieved from American Academy Of Ophthalmology:

3. Gantz L, Millodot M, & Roth GL. (2020). Analysis of Alphabet Patterns of Deviations Found in Patients Without Strabismus in Primary Position. Clinical optometry, 12, 49–56.

4. Kekunnaya, R., Mendonca, T., & Sachdeva, V. (2015). Pattern strabismus and torsion needs special surgical attention. Eye, 29(2), 184-190.

5. Jain, S. (2019). Alphabet Patterns in Strabismus. In Simplifying Strabismus (pp. 125-130). Springer, Cham.

6. Kushner, BJ. (2017). A-Pattern, V-Pattern, and Other Alphabet Pattern Strabismus. In: Strabismus. P97-114. Springer, Cham.

7. Wright, K.W. (2006). Alphabet Patterns and Oblique Muscle Dysfunctions. In: Wright, K.W., Spiegel, P.H., Thompson, L.S. (eds) Handbook of Pediatric Strabismus and Amblyopia. Springer, New York, NY.

8. Von Noorden GK and Campos EC. (2002) Binocular and Ocular Motility. 6th Edition, Mosby, Maryland Heights.

9. Brodsky, M.C. (2021). Primary Oblique Muscle Overaction. In: The Evolutionary Basis of Strabismus and Nystagmus in Children. Springer, Cham.

10. Clark RA, Miller JM, Rosenbaum AL, Demer JL. Heterotopic muscle pulleys or oblique muscle dysfunction? J AAPOS. 1998;2(1):17–25.

11. Kono, R., Okanobu, H., Ohtsuki, H. et al. Displacement of the rectus muscle pulleys simulating superior oblique palsy. Jpn J Ophthalmol 52, 36–43 (2008).

12. Pyatka N, Ghasia F (2019). Pattern Strabismus: Where Does the Role of the Brain End and the Role of Muscles Begin?. In: Shaikh, A., Ghasia, F. (eds) Advances in Translational Neuroscience of Eye Movement Disorders. Contemporary Clinical Neuroscience. Springer, Cham.

13. Kushner BJ. (2017). Exotropia. In: Strabismus. Springer, Cham.

14. Wright KW and Strube YNJ. (2015). Exotropia. In: Color Atlas Of Strabismus Surgery: Strategies and Techniques. Springer, New York, NY.

15. Gokyigit B., Inal A., Ocak B., & Aygit E.D. (2019). Sliding Shape Extraocular Muscle Transposition with Plication: A Novel Method. Beyoglu Eye Journal, 4(2), 120-122.

16. Shah, P. R., & Pihlblad, M. S. (2020). Transposition of plicated horizontal muscles. Journal of American Association for Pediatric Ophthalmology and Strabismus, 24(4), 244-247.

17. Gokyigit, B., İnal, A., Ocak, O. B., & Aygıt, E. D. (2021). Sliding shape extraocular muscle transposition with plication: Long-term results. International Ophthalmology, 41(11), 3593-3598.