ALPHABET PATTERN STRABISMUS

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Duane first described "V" pattern in 1897 in a patient with bilateral superior oblique palsy[1]. But emphasis on importance of performing measurements in straight, upgaze and downgaze in strabismic patients was first given byUrrets-Zavalia in 1948[2,3].He also called attention to the fact that oblique overactions and underactions are associated with increased or decreased convergence or divergence in these positions. Urist[4]introduced this concept to American literature in 1951 and Albert suggested the excellent descriptive terms A and V patterns, which have now found worldwide acceptance.[5]

Definition and Classification

The terms "A" pattern and "V" pattern describe horizontal strabismus that is vertically incomitant. It is characterized by a substantial increase or decrease in the horizontal deviation in the midline position in upgaze as compared to downgaze.

1) **"V" pattern:-**The eyes are more converged in downgaze (more esotropic or less exotropic) than in upgaze.

2) **"A" pattern:-**The eyes are more converged (more esotropic or less exotropic) inupgaze as compared to downgaze.

By convention, the difference between upgaze(25°) and downgaze(35°) must be 15 PD (prism diopters) or greater to diagnose a clinically significant "V" pattern, and 10 PD diopters or more to diagnose an "A" pattern.

3) **"Y" pattern:-**The deviation changes minimally from downgaze to the primaryposition and diverge in upgaze.

4) **"λ"(lambda)pattern:-**The deviation changesminimally from the upgaze position to the primary position, but diverge in downgaze.

5) **"X" pattern:-**The eyes diverge in both upgaze and downgaze as compared to the primary position.

6) **Neutralizing pattern:-**There is orthophoria in the primary position and either divergence in upgaze and convergence in downgaze (neutralizing "V' pattern), or convergence in upgaze and divergence in downgaze (neutralizing "A" pattern).

Due to these variants in the European literature it has therefore become customaryto speak of strabismus with an *alphabetical pattern*.

Prevalence

A or V pattern co exist in about 12.5% to 50% cases of horizontal strabismus[6].According to the 1964 American Academy of Ophthalmology and Otolaryngology panel V esotropia is by far the most common anomaly, followed in order of frequency by Aesotropia, V exotropia, and A exotropia[7].A" and "V" patterns are relatively frequent in patients who have had congenital strabismus.

Etiology

Different mechanisms may be responsible in different patients. The proposed theories are:

Oblique muscle Dysfunction

The most popular theory, suggested by Knapp in 1959[8] attributes most cases of "A" and "V" pattern to the role of oblique muscle dysfunction and the contributing effect of the accompanying torsion. Abduction is a tertiary action of the superior and inferior oblique muscles. Thus, if

A)The superior oblique muscle is overacting, and the antagonist inferior oblique is underacting, there is relative divergence in downgaze and convergence in upgaze; resulting in an "A" pattern

B)The inferior oblique muscle is overacting and the superior oblique muscle is under- acting, there is convergence in downgaze and divergence in upgaze, resulting in a "V" pattern.

The torsion that accompanies oblique muscle dysfunction also contributes to the associated "A"-or "V" pattern[9], V-pattern is associated with excyclotropia due to inferior oblique muscle overaction and A-pattern is associated with incyclotropia due to superior oblique muscle overaction.

Horizontal rectus muscle dysfunction

Urist(1958) believed that horizontal rectus muscles were responsible for this incomitance[6,10], in V esotropiaoveraction of the medial rectus muscles caused the increased convergence in downward gaze and overaction of the lateral rectus muscles was responsible for the increased divergence in upward gaze. Conversely, increased divergence in downward gaze in Aexotropia was thought to be caused by underacting medial rectus muscles and in A esotropia by underacting lateral rectus muscles.

Vertical rectus muscle dysfunction

Brown(1953) had the opinion that A or V patterns may be caused by primary anomalies in the function of the vertical rectus muscles in which adduction is the tertiary action. For example, if the superior rectus muscles are primarily underacting, their adductive effect in upward gaze will decrease; in fact, the eyes will diverge in upward gaze because of secondary overaction of the inferior oblique muscles. In downward gaze, secondary underaction of the superior obliques will cause decreased abduction and secondary overaction of the inferior rectus muscles, resulting in increased adduction of the eyes,which, according to Brown, would produce a V pattern.

Anomalies of orbit

Urrets- Zavalia and coworkers,[11] in a study of Bolivian Indian children, found out that in *mongoloid* type of facial development (hyperplasia of the malar bones, upward slanting of the palpebral fissures, and a straight lower lid margin)esotropia was frequently associated with underacting inferior oblique muscles (A esotropia) and exotropia with overacting inferior obliques (V exotropia). In white children with *antimongoloid*features (hypoplasia of the malar bones, downward slanting of the palpebral fissures, and S-shaped contour of the lower lid margin) the opposite was found, esotropia associated with overacting inferior oblique muscles (V esotropia) and exotropia with underacting inferior oblique muscles (A exotropia).

Sagittalization of oblique muscle insertions (figure 4)

In Sagittalization the oblique muscle becoming more parallel to the sagittal (anteroposterior) axis and indesagittalizationoblique muscles become more parallel to the coronal plane concept of which was given

by Gobin[12]. If the superior oblique is desagittalised due to the retroplacement of trochlea (as in plagiocephaly), it becomes a poorer depressor. And relatively the inferior oblique becomes a stronger elevator. Similarly with a more frontally placed trochlea (as in hydrocephalus with frontal bossing), superior oblique becomes more sagittalised in relation to the inferior oblique making it a stronger depressor. This relative action can cause A and V patterns .

Heterotropia of muscle pulley

Demer et al[13] proposed the presence of fibromuscular pulleys of the recti and inferior oblique muscles. Just like the trochlear pulley for superior oblique tendon. If these pulleys are displaced, incomitant deviations can be caused. For example: upward displacement of medial rectus pulleys and downward displacement of lateral rectus pulleys results in A pattern.

Sensory Deprivation

Guyton and coworkers had the view that loss of fusion predisposes the oculomotor system to cyclodeviations of the eyes which, in turn, cause A and V patterns according to the mechanism proposed by Weiss[14]. Guyton and Weingarten showed that formerly fusing patients with intermittent exotropia who lost fusion after surgical overcorrection may develop A or V patterns.

Presentation:

1) **Asthenopia and diplopia:** common complaints in patients with A and V patterns, the increase in a deviation in downward gaze (with A exotropia or V esotropia) may cause acute visual discomfort during reading ,On the other hand, an increase in the deviation in upward gaze (with V exotropia) is best tolerated by most patients since little or no interference with binocular vision.

2) **Anomalous head posture:** The patient with Aesotropia and V exotropia and fusion in downward gaze may hold his or her chin in an elevated position. Conversely, V esotropia and A exotropia may cause chin depression .

   Some adults with an "A" or "V" pattern may not become symptomatic until they become presbyopic; until they need to get their eyes into downgaze to read through their bifocal segment.

Examination

Vision assessment, refraction, detailed orthoptic evaluation with full correction which includes: Abnormal head posture, cover test, examination of ocular movements, Prism bar cover test, sensory evaluation and fundus examination should be done.

Motor Examination:

Alternate prism cover testing is performed with head held in primary gaze using an accommodative target at 20ft (6m). The measurements are made in primary position,upward (25°) and downward gaze (35°) to establish whether an A or V pattern is present and if so whether it is clinically significant. Stuart and Burian established that divergence of the visual lines in upwardgaze and convergence in downward gaze are physiologic variants. Thus V pattern in which the difference in deviation between upward and downward gaze is 15 Δ or more should be considered a significant vertical incomitance. Since an A pattern is never found as a normal variant, a limit of 10 Δ has been set beyond which an A pattern is thought to be significant.

Pseudo A and V Patterns: A pseudo V pattern may be seen in patients with accommodative esotropia. This occurs if the patient having a small amount of hyperopia is tested without using hyperopic correction.

Uncorrected hyperopia gives rise to accommodation in primary and downgaze, as opposed to upgaze, simulating a V pattern. Similarly V- Pattern strabismus may also be seen in cases of intermittent exotropia. Hence full optical correction must be given prior to motor examination.

It is also important to recognize that a "V" pattern may simulate a high AC/A ratio if care is not taken to keep the fixation target in the primary position at near.

Sensory Examination:

Patients with "Y" patterns, or "λ" patterns may be well aligned in the primary position and may have surprisingly good fusion.

If a patient is tropic in all fields of gaze, suppression and varying depths of anomalous retinal correspondence (ARC) may be found.

Ciancia and Helveston et al. found that in patients with "A" or "V" pattern and ARC, the angle of anomaly varies with the angle of deviation, thus resulting in the ARC being harmonious in all fields of gaze.[15]

Treatment:

Goals:

1) To maintain,improve or regain binocular single vision.
2) To restore patients normal facial configuration i.e to eliminate chin elevation or depression.
3) To establish binocular fusion in functional position of gaze.(primary and reading position)

General Principles

- Both sensory and motor components should be addressed.
- Full refractive correction should be prescribed.
- Remove suppression and treat amblyopia before surgery.
- Ocular movements should be carefully assessed.
- Surgery done if the deviation causes symptoms, cosmetic defect and/or produces sensory anomalies like suppression/amblyopia.

Surgical methods:

Vertical transposition of horizontal recti: (figure 3)

Basis: The action of a muscle is weakened in the direction in which its insertion is shifted. This procedure should be considered only if the obliques are not overacting.

1/2 width transposition: corrects upto 15 PD of pattern.

Medial rectus is transposed towards the apex of V.

Lateral rectus is transposed towards the base of V.

Slanting muscle recessions:

Basis: Horizontal muscle tensions are different between the upper and lower margins of the muscle with variant amounts of gaze[16], i.e, horizontal muscle tension at the upper margin is stronger than that at the lower margin in upward gaze.

In the slanting surgery, the upper margins of the LR are recessed more than the lower margins to gain a greater effect on horizontal deviation in upward gaze in patients with "V" exotropia. The usual amount of the selective shift of the tendon to slant its insertion is 2-mm backward.

Surgery on oblique muscles

Inferior oblique surgery

It is indicated in presence of inferior oblique over action and V pattern strabismus. The usual resulting correction after any of the procedures is about 20 PD in elevation[17]. Various weakening procedures include myectomy, Inferior oblique recession or anterior transposition. Bilateral IO myectomies has no effect on the horizontal alignment in primary position[18,19]. Inferior oblique anterior transposition gives more effect than IO recession, but it results in limitation of elevation, so is preferred for patients with dissociated vertical deviation (DVD).

Superior oblique surgery

- Superior oblique weakening should be performed very cautiously as it affects the reading gaze of the patient. Therefore, it is indicated only in patients with significant superior oblique overaction.
- Superior oblique tenotomy is the most commonly performed procedure for superior oblique weakening.

Specific Management

A-Pattern Esotropia

(a) *Patients without superior oblique (SO) muscle overaction*- Recession and symmetric supraplacement of the tendons of the MR muscles by one-half tendon width.

(b) *Patients with SO muscle overaction*- Bilateral SO tenotomy/posterior tenectomy and horizontal rectus recessions to correct esotropia in primary gaze.

A-Pattern Exotropia

(a) *Patients without SO muscle overaction* - Recession and symmetric infraplacement of the tendons of the LR muscles by one-half tendon width.

(b) *Patients with SO muscle overaction* - Bilateral SO tenotomy/posterior tenectomy combined with symmetric surgery on the horizontal rectus muscles to correct exotropia.

V- Pattern Esotropia

(a) Patients without inferior oblique (IO) muscle dysfunction-Recession and symmetric infraplacement of the tendons of the MR muscles by one-half tendon width.

(b) Patients with IO muscle dysfunction - Weakening of IO muscles is combined with appropriate MR recession.

V-Pattern Exotropia

(a) Patients without IO muscle dysfunction - Recession of the LR muscles with supraplacement by one-half tendon width.

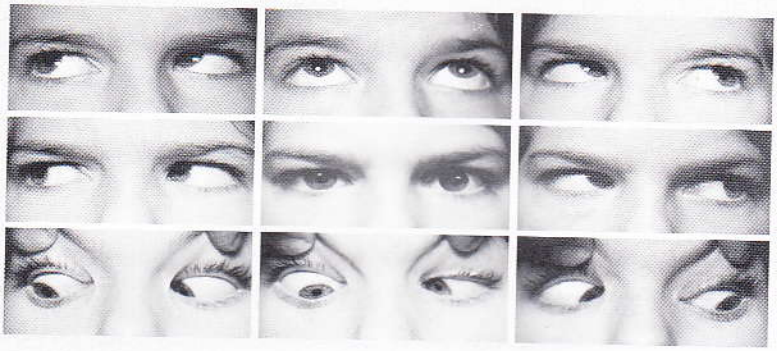
(b) Patients with IO muscle dysfunction- IO muscle should be weakened symmetrically and appropriate recession of the LR muscle performed to correct exodeviation in primary gaze.

Y-Pattern

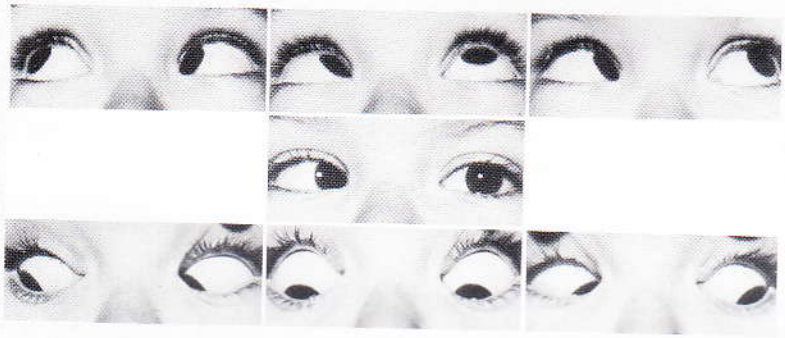
Y esotropia or exotropia usually have IO overaction with evidence of fundus excyclotorsion. Bilateral weakening of IO muscle reduces or eliminates the pattern in upgaze.

Lambda (λ) Pattern

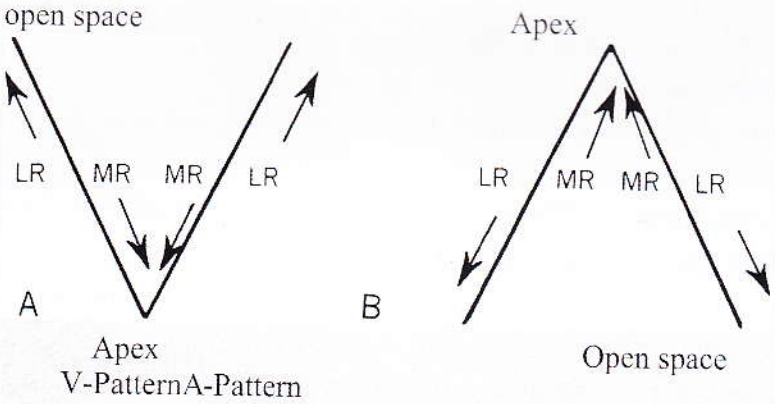
Bilateral weakening of SO will reduce the λpattern if SO overaction is present.



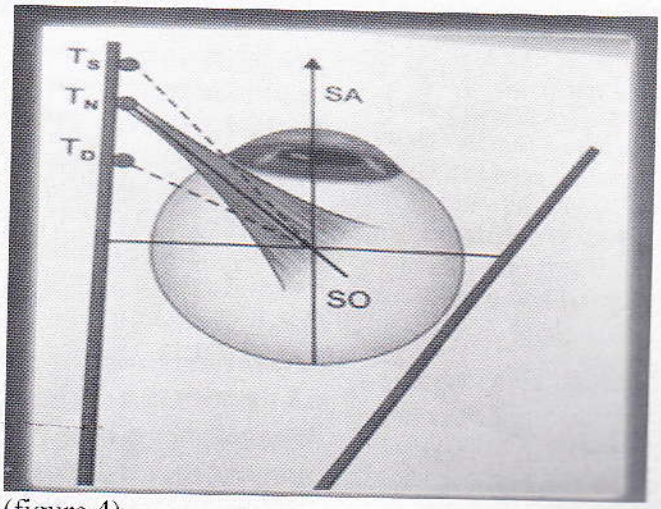
V-Esotropia



A-Esotropia



(Figure 3)



(figure 4)

Conclusion:

Various factors are responsible for pattern strabismus and hence every case is a different case. Special emphasis must be given on measurement of deviation in primary position,upgaze and downgaze, and oblique muscle dysfunction is to be looked carefully as it can significantly alter the surgical management.

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