



Assessing abnormal head posture: a new paradigm

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Purpose of review

We aim to give a systematic approach on how to assess the cause of a patient's abnormal head posture (AHP).

Recent findings

Over the decades, many important clinical observers and teachers have described diagnostic techniques for patients with AHP. Recently, Wong has added a new set of office-based diagnostic criteria that expand and solidify the office assessment of AHP.

Summary

We describe old and new, well known and poorly known office techniques for the assessment of AHP.

Keywords

abnormal head posture, congenital nystagmus, oblique palsy, skew deviation, strabismus

INTRODUCTION

Assessing abnormal head posture (AHP) can be biased by the physician's training, experience and the spectrum of patients seen. The pediatric ophthalmologist will have a different perspective on AHP based on whether his or her practice has a predominance of genetic cases, one with developmental issues or one (like ours) whose practice is limited to strabismus [1–3].

Many important authors have contributed to our understanding of diagnostic and therapeutic techniques for patients with AHP. Literature of particular assistance to us has been authored by Gobin [4], Hertle [5^{*}] and Wong [6,7].

In assessing a new case with AHP, several questions have to be asked and answered.

Is the abnormal head posture driven by visual activity?

Instruct the patient to 'close your eyes, now hold your head straight', or with the patient's eyes closed, you tilt the head into an unusual position then instruct 'keep your eyes closed and now hold your head straight' (Fig. 1 right side).

If the AHP persists with the eyes closed then it is not related to visual activity, and is due to orthopedic issues or vestibular problems. The usual vestibular problem seen with AHP is the ocular tilt reaction (OTR), comprising a vertical misalignment (skew deviation) and a head tilt [8]. The hypertropia of the skew deviation is not improved by the head

tilt, that is the head tilt is not therapeutic (unlike the head tilt seen with superior oblique palsy) and the hypertropia is improved by changing from erect to supine [6].

Otolith and orthopedic abnormalities can be both causes and sequelae of AHP.

If the AHP is consistently fixed by 'close your eyes...' then the AHP is visually driven, and this article will help explain these findings.

What precise visual input drives the abnormal head posture?

You need to find out if the AHP is driven by:

- (1) Left eye fixation. Cover the right eye and ask the patient to read the chart with the left eye. If this reproduces the AHP that is seen with both eyes open (and right eye fixation does not drive the AHP) then the AHP is driven just by the left eye (Fig. 2 left side).
- (2) Right eye fixation. Cover the left eye and observe for similar findings as above.

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KEY POINTS

- AHP can be driven by monocular fixation, either eye fixing, only by both eyes fixing, or be unrelated to visual fixation. Tilt, turns and tips of the head can be driven by similar mechanisms, and there are some mechanisms unique to each type of AHP. Types of congenital nystagmus are common causes of AHP.

- (3) Either eye fixing.
- (4) Only when both eyes are fixing. The AHP is driven by strabismus.

This examination must be done while the patient is viewing an age appropriate suprathreshold visual target, ideally one that can be changed to approach threshold as the patient is being tested. For example, if a patient has 20/20 acuity in each eye, the initial evaluation might be performed while quickly reading a 20/80 line that changes every 4 s [LK uses the M&S acuity tester (M&S Technologies, Skokie, Illinois, USA)], and then increase the target difficulty to 20/60, 20/40 and so on. This process will usually accentuate an acuity-dependent AHP.

For near, the visual target can be a changing Snellen – style suprathreshold chart (LK uses 'Eye Chart HD' for iPhone from Apple Apps Store).

Nystagmus-associated AHP is usually acuity-dependent. Nystagmus degrades the acuity, and the patient will adopt the AHP that minimizes the

nystagmus (expands foveation time) and maximally improves acuity (at the null point or null zone) [5th].

Refraction-associated AHP is also usually acuity dependent. This situation can be because of an incorrect refraction or an incorrectly made spectacle lens. As the visual demand increases an AHP develops, typically driven by one eye. The AHP is abolished by using a pinhole to read the chart.

Describing the components of the abnormal head posture: the three T's – tilt, turn and tip

Most cases of AHP manifest as: head tilt (to left or right shoulder), face turn (to left or right) or head tip (up/down), and there will be some multiplanar cases (tilt and turn, turn and tip, etc) (Fig. 3 bottom).

The discussion in this article will describe:

- (1) Evaluation, causes and treatment of head tilt.
- (2) Evaluation, causes and treatment of face turn.
- (3) Evaluation, causes and treatment of head tip.

HEAD TILT

The examination should elucidate whether the tilt is driven by:

- (1) Left eye fixing
- (2) Right eye fixing
- (3) Either eye fixing – tilt is driven by either eye or is independent of visual fixation

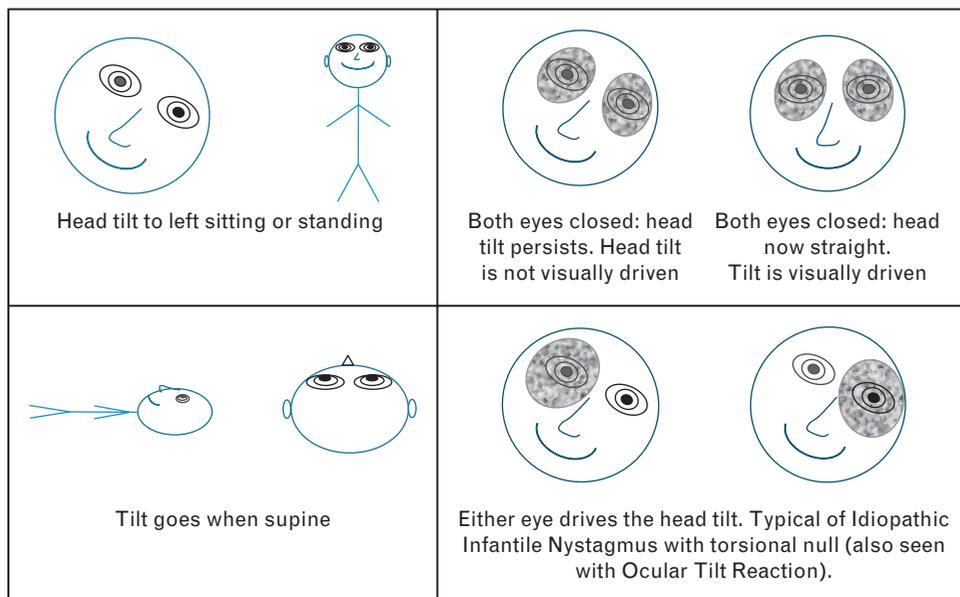


FIGURE 1. 'Both eyes closed' test to see if the AHP is visually driven. In OTR, head tilt (largely or completely) disappears when supine. In IIN, the tilt is driven by either eye. IIN, idiopathic infantile nystagmus; OTR, ocular tilt reaction [3].

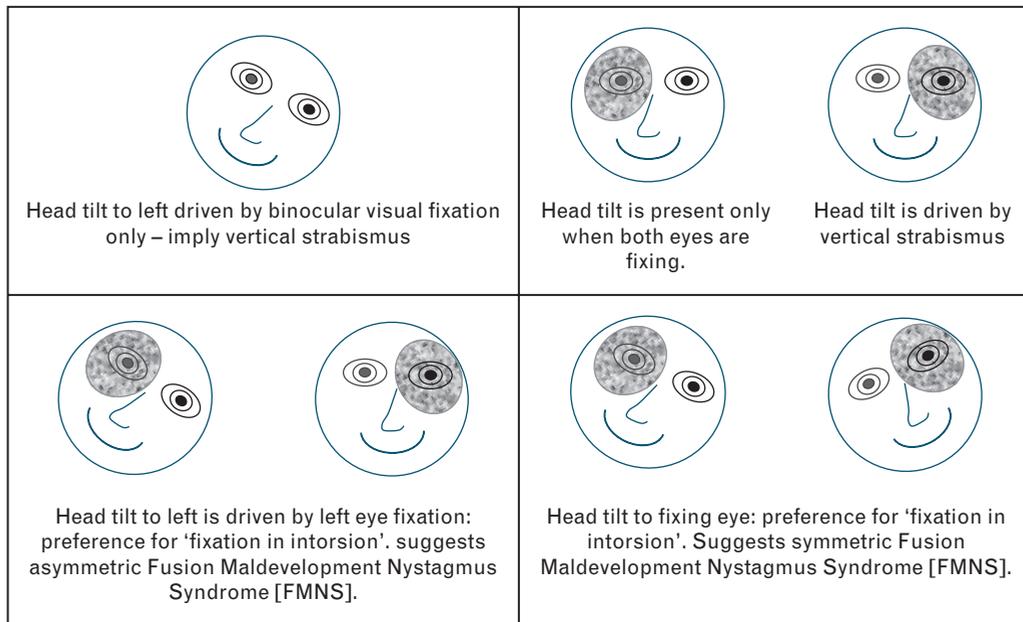


FIGURE 2. Testing whether either or both eyes only drives tilt diagnoses vertical strabismus. Monocularly driven head tilt usually due to underlying torsional FMNS. FMNS, fusion maldevelopment nystagmus syndrome.

(4) Only with both eyes fixing – tilt is driven by strabismus

Left fixation: tilt is driven by the left eye; with right fixation there is no head tilt (Fig. 2 left bottom)

This can be caused by nystagmus or by restrictive strabismus. If nystagmus, this is usually caused by the torsional component of fusion maldevelopment nystagmus syndrome (FMNS), previously called latent manifest latent nystagmus. This is a type of congenital nystagmus usually seen with infantile onset strabismus [9,10*].

The left eye seems to prefer to fix in intorsion. The left superior oblique is recruited (this recruitment is achieved by tilting to the left) to act as a brake on the torsional component of the FMNS and this improves acuity. If the nystagmus is not obvious to the examiner’s naked eye, it is easily seen with the slit lamp and will often have both horizontal and torsional components. A similar mechanism has been described by Brodsky [11], and a similar mechanism also causes DVD [12].

Left fixation – tilt is driven by the left; right fixation there is no head tilt; both eyes fixing also drives the same tilt as left fixation alone: usually caused by a restrictive left extraocular muscle

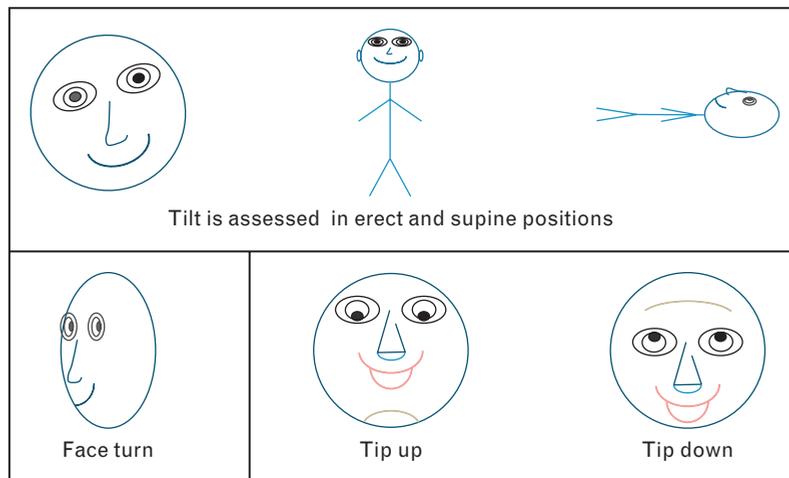


FIGURE 3. Each AHP has three components, tilt, turn and tip. Tilt has to be assessed in erect and supine positions. AHP, abnormal head posture [3].

anomaly, revealed by examining versions, ductions and cover test.

Right fixing: tilt is driven by the right; with left fixation there is no head tilt

This is the opposite appearance to the previous example above. Usually, this is due to torsional FMNS asymmetrically affecting the right more than the left eye or to restrictive strabismus.

Special case: head tilt to the fixing eye (Fig. 2 bottom right)

Here, left eye fixation results in tilt to the left, right eye fixation results in tilt to the right. The mechanism seems to be preference for fixation in intorsion, and the patient has fairly symmetric torsional FMNS. This is the torsional variant of the more frequently seen face turn to the fixing eye driven by horizontal FMNS, popularized by Ciancia and when combined with idiopathic infantile esotropia is known as Ciancia's syndrome [13]. Sometimes, the FMNS-associated face turn and head tilt can be seen together [10⁷].

Either eye fixing: tilt is driven by either eye (Fig. 1 bottom right)

This is usually caused by the type of congenital nystagmus known as idiopathic infantile nystagmus, (IIN), the nystagmus typically associated with eccentric nulls and a convergence null. There will be a horizontal nystagmus with a torsional or oblique null that is the same with either eye fixing or with both eyes fixing.

If there is no nystagmus, and tilt seems to be driven by either eye but on further examination is independent of visual fixation and goes away with closing both eyes can be caused by the OTR [8]. There is usually other evidence of OTR, for example,

- (1) A hypertropia that largely disappears when changing from erect to supine (Fig. 1 left).
- (2) A hypertropia that is not fixed by the head tilt that is the head tilt is not therapeutic.
- (3) Recent or past vestibular disease.
- (4) Conjugate torsion on fundus examination – one eye (the higher one if there is a hypertropia) intorts, the other extorts.

The hypertropia seen in a–d above is a skew deviation. OTR is sometimes seen in the cochlear implant population [14].

Only with both eyes fixing: tilt is driven by strabismus (Fig. 2)

When the tilt is driven by both eyes open but disappears when either eye fixes, then vertical strabismus is the cause, and the head tilt is compensatory for the hypertropia revealed by the abnormal cover test. Hypertropia is the commonest cause of only with both eyes fixing. Torsion is usually not the cause [15].

Spasmus nutans and other neurological disease

Spasmus nutans present as an asymmetric nystagmus (may seem to be monocular nystagmus) sometimes associated with head tilts (can be variable) and head bobbing. Variable head tilts can be seen in other neurological diseases, for example, opsoclonus–myoclonus syndrome and some developmental/chromosomal disorders in which the examination does not allow precise subtyping according to the guidelines above. Head bobbing or shaking are not a sine qua non of spasmus nutans and can be seen also in congenital nystagmus [16]. There should be a low threshold for quick referral to pediatric neurology and imaging if spasmus nutans is suspected; some will be associated with tumor.

FMNS is more common in abnormalities of the visual cortex such as periventricular leukomalacia (PVL) and can drive face turns, head tilts, head bobbing and shaking. (Ref [17], and personal communication from R. Hertle). It is not necessary to routinely image children with possible PVL; a pediatric or pediatric neurology opinion should usually be sought as there may be nonocular issues present also.

FACE TURN

The diagnostic approach is similar to that adopted for head tilt, with some differences.

You still have to ask:

Is the face turn driven by visual activity?

Instruct the patient to 'close your eyes, now hold your head straight', and then observe for changes to the face turn.

What vision input drives the face turn?

Is the AHP driven by right eye fixation, left eye fixation, either eye fixing or only when both eyes are fixing.

Face turn driven by monocular fixation

The nystagmus that causes face turn driven by monocular fixation is usually the horizontal component

of FMNS. This will cause apparent preference for fixation in adduction. So, left fixation will cause face turn to the left; right fixation, face turn to the right.

Face turn driven by monocular fixation can also be caused by imperfect correction of abnormal refraction (will be 'cured' with a pinhole) or a restrictive orbital anomaly with associated incomitant strabismus.

Alternating face turns can be caused by:

- (1) FMNS with spontaneous alternation of fixation. This might be driven in part by subtle refraction asymmetry, for example, the slightly hyperopic right eye is used for distance fixation and the face is turned to the right for reading a vision chart. The fixating right eye is drawn into adduction to suppress the FMNS and the face will turn right to view the distance target. If fixation is switched to a myopic left eye for reading a near card, then the left eye is adducted and the face turn will be to the left for near fixation.
- (2) Periodic alternating nystagmus (PAN). This is an uncommon variant of IIN and is under-recognized. 'Congenital' PAN often has asymmetric periodicity, for example, 5 min to left, 2 min to right (unlike acquired PAN, 2 min each way; commonly due to diencephalic disease). Eye movement recordings can be more helpful than the office findings in some cases of PAN, especially in cases in which one null is in or near the primary position. Recordings can also be misleading if the periodicity of the PAN is very asymmetric (e.g. 10 min with face turn to left, 1 min to the right) and the recordings are not continued for long enough to detect this [18].

A rare variant of congenital PAN is periodic alternating gaze deviation, (PAGD). The face slowly rotates from one position to another and back again with similar asymmetry of periodicity to 'congenital' PAN. Acquired PAGD secondary to neurological disease is called ping-pong gaze, and is qualitatively different.

(Cogan's) oculomotor apraxia can sometimes seem to cause an alternating face turn.

Conditions that cause face turns apparently driven by either eye are:

- (1) Large homonymous field defects. Huge congenital or infant onset field defects may not be reported by the patient, and recognized only during a targeted examination. The field defects causing a face turn are never small subtle ones requiring automated perimetry, and are easily detected by confrontation techniques.

- (2) Gaze paresis/palsy. A left gaze paresis will result in a face turn to the left apparently driven by either eye. If subtle, (e.g. with no apparent version or duction deficit) this may only be detected in the office by looking at saccades, and especially oblique saccades. Oblique saccades allow one to qualitatively compare the speed of the horizontal saccade with the vertical saccade. If a normal patient is asked to generate a saccade from (his) down-right position to up-left, both eyes will move in a straight line from down-right to up-left (recordings might show a slight curve). If there is a left sixth, the saccade of the right eye will move in a straight line, and the saccade of the left eye will execute a curved saccade, moving up before it moves left because the vertical saccade is faster than the horizontal saccade. If there is a left gaze paresis, the up-shape then-left shape of the saccade will be seen in both eyes.
- (3) IIN with an eccentric null will cause a face turn apparently driven by either eye.

TIPS: TIP UP AND TIP DOWN

Once again, the diagnostic steps are:

- (1) Is the tip driven by visual fixation?
- (2) Is it driven by left eye fixing, right eye fixing, either eye fixing or only with both eyes fixing?

Some of the conditions we have discussed above can also cause tips:

- (1) IIN can cause a tip (vertical null) with either eye fixing [19]. Although IIN is nearly always horizontal, rarely it can be vertical. A vertical null is more common with vertical IIN. FMNS does not cause tips.
- (2) Incomitant strabismus. Many causes of incomitant strabismus will cause a tip – the alphabet patterns, the congenital fibrosis syndromes, Brown's syndrome, thyroid eye disease and so on.

Tips can be driven by supranuclear disease.

- (1) Vertical gaze paresis including the paroxysmal ones [20]. Some of these cases evolve into (and may be a forme fruste of) horizontal or vertical IIN with a vertical null (personal observation).
- (2) The spinocerebellar atrophy syndromes, usually secondary to acquired downbeat nystagmus with a null in either upgaze or in extreme down-gaze.

VARIABLE ABNORMAL HEAD POSTURE

Examination findings or photographic evidence (family album test) can indicate a variable AHP. Usually, the explanation is one that has already been discussed above or a variation:

- (1) PAN
- (2) When different AHP is dependent of which eye is fixing, when left fixation and right fixation produce different outcomes. For example, if torsional FMNS is more relevant to left eye fixation and horizontal FMNS for right eye fixation, then the AHP may vary from left head tilt (left eye fixation) to right face turn (right eye fixation). Small differences in refraction may dictate the circumstances of each component of the variable AHP.
- (3) In a patient with vertical strabismus, we may see various scenarios. There may be no AHP when the hypertropia is small and compensated for with suppression. In some positions of gaze the amount of hypertropia will exceed the size of the suppression scotoma and the patient will then use a head tilt to compensate. But, if the vertical strabismus is too great in some positions there may be acquired suppression, and the head tilt is no longer present.
- (4) Some patients with IIN have several eccentric null zones. This is different to those with PAN who will have two null zones and oscillate between them in a consistent pattern. An IIN patient may have a horizontal null (with a face turn) and a vertical null (with a tip) but 'never' uses the vertical null because it is physically less comfortable or, as a teenager, the patient becomes aware it looks worse than the face turn. The vertical null might be used for, say, looking at a wall-mounted Television, or become apparent only after surgery for a horizontal face turn!
- (5) Myasthenia and other causes of variable oculomotor conditions can cause variable AHP.

CONCLUSION

In this review, we offer a practical systematic approach to the office assessment of the patient with AHP. The most frequent ocular causes of head tilt are the types of congenital nystagmus and vertical misalignment, especially superior oblique palsy. Face turn is associated most commonly with congenital nystagmus (FMNS and/or IIN), neurogenic strabismus, refractive errors and visual field defects. Tip postures are most commonly caused by incomitant strabismus, nystagmus, neurologic

conditions and ptosis. Sometimes, two or three different null zones exist in the same patient. These may all be effective, but one is typically preferred and dictates the usual AHP.

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Conflicts of interest

The authors have no financial interest to disclosure.

REFERENCES AND RECOMMENDED READING

Papers of particular interest, published within the annual period of review, have been highlighted as:

- of special interest
- of outstanding interest

Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 516).

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