Original Article

Screening of Clinical Data of Patients with Abnormal Head Posture and Investigation of Abnormal Head Posture Change After Treatment

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Abstract

Objectives: To analyze the clinical characteristics of patients with abnormal head posture (AHP) due to ocular causes and investigate the effect of treatment on the change in AHP.

Materials and Methods: Patients with AHP admitted to the strabismus unit of our clinic between 2011 and 2022 were retrospectively analyzed. The patients' clinical and demographic data and change in AHP after treatment were recorded.

Results: A total of 172 patients, 86 females (50%) and 86 males (50%), with a mean age of 14.1±13.9 years were included in the study. The most common ocular causes of AHP were fourth cranial nerve palsy (50%), Duane retraction syndrome (16.9%), and A-V pattern strabismus (15.1%). Sixth cranial nerve palsy, third cranial nerve palsy, nystagmus blockade syndrome, extraocular muscle fibrosis, Brown syndrome, oculocutaneous albinism, and heavy eye syndrome were diagnosed less frequently. The most common AHP type was head tilted position (52.3%), followed by head turned (40.1%), chin down/up (3.5%), and combined form (4.1%). There was a significant relationship between AHP type and diagnosis ($p<0.001$). Amblyopia was present in 55 (35.7%) and absent in 99 (64.3%) patients. There was a significant relationship between amblyopia and both diagnosis ($p<0.001$) and AHP type ($p=0.003$). Of 172 patients, 100 (58.1%) underwent strabismus surgery, 10 (5.8%) had botulinum toxin injection, and 2 (1.2%) were prescribed prism glasses. Sixty patients (34.9%) were only followed up. Among 94 patients who continued follow-up, AHP was reduced in 77.3% and completely resolved in 16.7% of patients treated surgically, and was reduced in 50% and completely resolved in 25% of those treated with botulinum toxin.

Conclusion: The causes of AHP are varied. Ophthalmological and orthoptic examinations should be performed in patients presenting with AHP, and strabismus surgery or botulinum toxin administration may reduce or completely correct AHP in eligible patients.

Keywords: Abnormal head posture, strabismus, strabismus surgery, botulinum toxin, amblyopia

Introduction

Abnormal head posture (AHP) is an adaptation mechanism in which the head may be turned or tilted and the chin up or down in order to increase visual acuity, prevent diplopia, or provide comfortable binocular vision.[1] AHP is not a diagnosis but a symptom of an underlying disease, although there may be no apparent cause in some patients.[2] The main etiologies of AHP are excessive contraction of the sternocleidomastoid muscle due to congenital muscular torticollis, ocular diseases, and central nervous system anomalies.[3] The most common ocular causes are fourth cranial nerve palsy, Duane retraction syndrome, sixth cranial nerve palsy, Brown syndrome, and nystagmus blockage syndrome.[3]

While there are studies examining the types and causes of AHP in the literature, fewer studies have also examined the changes in AHP after treatment. This study aimed to examine the AHP types, etiologies, and clinical data of patients with AHP together with the changes in AHP observed after different treatment options.

Materials and Methods

We retrospectively analyzed the data of 172 patients with AHP who presented to the strabismus unit of the Ondokuz Mayıs University Faculty of Medicine Hospital, Department of Ophthalmology between 2011 and 2022. Our study was

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conducted in accordance with the 2013 Helsinki Declaration and received approval from the Ethics Committee of Ondokuz Mayıs University (decision no: OMÜ KAEK 2022/563, date: 28.12.2022).

Patients who had previously undergone ocular surgery and those whose AHP was due to non-ocular causes were not included in the study. The patients' age, sex, comorbidity, best-corrected visual acuity on Snellen chart, AHP type, eye movements, and amount of deviation measured by prism or Krimsky test were recorded. We also analyzed the results of Worth 4-dot, fusion, and Titmus three-dimensional stereo tests performed before AHP correction.

Based on the patients' diagnosis and examination findings, they were either followed up without treatment or treated with botulinum toxin A (Botox; Allergan Inc., Irvine, CA, USA), prismatic glasses, or surgical methods such as inferior oblique recession, superior oblique tenotomy, inferior oblique tenotomy, and medial/lateral rectus recession or resection.

Statistical Analysis

Descriptive statistics were expressed using mean and standard deviation for continuous variables and as number and percentage for categorical variables. Relationships between categorical variables were examined using chi-square and Cramer's V tests. The study data were analyzed using IBM SPSS Statistics for Windows, version 21.0 (IBM Corp., Armonk, NY, USA). P values less than 0.05 were considered statistically significant.

Results

The mean age of the 172 patients with ocular AHP was 14.14±13.95 years (range, 4 months-61 years); 86 (50%) were female and 86 (50%) were male. The distribution of AHPs and clinical data according to diagnosis are summarized in Table 1. The most common diagnoses were fourth cranial nerve palsy (50%), Duane retraction syndrome (16.9%), A-V pattern strabismus (15.1%), sixth cranial nerve palsy (4.1%), and nystagmus blockage syndrome (4.1%). Less common etiologies were third cranial nerve palsy, extraocular muscle fibrosis, Brown syndrome, oculocutaneous albinism, and heavy eye syndrome. In terms of AHP type, there were 90 patients (52.3%) with head tilt, 69 patients (40.1%) with head turn, 6 patients (3.5%) with chin down or up, and 7 patients (4.1%) with combined AHP. There was a significant relationship between the type of AHP patients developed and their diagnosis ($p<0.001$).

Best corrected visual acuity could be measured in 154 of the 172 patients included in the study. Fifty-five (35.7%) of the patients had amblyopia and 99 (64.3%) did not. Among the patients in whom amblyopia could be assessed, amblyopia was observed in all patients diagnosed with nystagmus blockage syndrome, oculocutaneous albinism, heavy eye syndrome, and extraocular muscle fibrosis, whereas amblyopia was not observed in any of the patients with sixth cranial nerve palsy or Brown syndrome. Of the 55 patients with amblyopia, 28 (50.9%) had head turn, 20 (36.4%) had head tilt, 4 (7.3%) had chin down/up, and 3 (5.5%) had combined AHP. The frequency of amblyopia differed significantly according to both diagnosis ($p<0.001$) and AHP type ($p=0.003$).

No deviation in primary position was observed in 53 (30.8%) of the patients, whereas 57 (33.1%) had esotropia, 20 (11.6%) had exotropia, 33 (19.2%) had vertical deviation, and 9 (5.2%) had vertical and horizontal deviation together. Of 108 patients who could undergo the Worth 4-dot test, fusion was observed in 33 (30.6%), suppression in 59 (54.6%), and diplopia in 16 (14.8%). No significant relationship was found between Worth 4-dot test result and amblyopia ($p=0.101$). Stereopsis was observed in 56.4% of 78 patients who could be assessed. Of the 167 patients whose eye movements could be evaluated, 83.2% had some degree of limitation in at least one direction.

Of the 172 patients, 100 (58.1%) underwent strabismus surgery, 10 (5.8%) received botulinum toxin injection, and 2 (1.2%) received prismatic glasses. Sixty patients (34.9%) were followed up without intervention. Of the patients who underwent surgery, 71 (71%) had fourth cranial nerve palsy, 12 (12%) had Duane retraction syndrome, 9 (9%) had A-V pattern strabismus, 4 (4%) had third cranial nerve palsy, 2 (2%) had Brown syndrome, 1 (1%) had extraocular muscle fibrosis, and 1 (1%) had heavy eye syndrome. Of the patients who received botulinum toxin, 4 (40%) had sixth cranial nerve palsy, 4 (40%) had Duane retraction syndrome, 1 (10%) had fourth cranial nerve palsy, and 1 (10%) had extraocular muscle fibrosis.

Final head position could be evaluated in 94 patients who continued regular follow-up. Of these, AHP was reduced in 55 patients (58.5%), completely resolved in 12 patients (12.8%), and remained the same in 27 patients (28.7%). Table 2 shows changes in AHP according to treatment method for the 94 patients who had regular follow-up. AHP improved or decreased in 44 of 71 patients with fourth cranial nerve palsy, 8 of 12 patients with Duane retraction syndrome, 6 of 9 patients with A-V pattern strabismus, 2 of 4 patients with third cranial nerve palsy, and the 1 patient with heavy eye syndrome.

Discussion

AHP refers to a head position that deviates from the normal angle to the body in the vertical, horizontal, and/or anterior-posterior axis.[4] The prevalence of ocular causes of AHP has been reported to be 18-25%.[3] In a study investigating ocular causes of AHP, Erkan Turan et al.[5] determined that paralytic strabismus, nystagmus, and Duane retraction syndrome were the most common. Dikici and Kızılkaya[6] showed in their study that the most common mechanism of AHP was conditions associated with incomitance, with paralytic strabismus being the most common cause (48.6%). Among these conditions, superior oblique palsy and third cranial nerve palsy were the most common causes of vertical incomitance, while sixth cranial nerve palsy and Duane retraction syndrome were the leading causes of horizontal incomitance.[6] Mitchell[7] demonstrated incomitant causes in the etiology of 52.4% of AHP cases. Similar to these studies, we observed that incomitant causes were frequently involved in the etiology of AHP, and fourth cranial nerve palsy and Duane retraction syndrome were the two most common causes in this study.

Table 1. Distribution of AHP types, treatment methods, and clinical findings by diagnosis (n=172)

Diagnosis	Frequency, % (n)	AHP type, % (n)	Amblyopia, % (n)	Treatment method, % (n)	Deviation in primary position (%)	Binocularity, % (n)
Fourth nerve palsy	50 (86)	Head turned: 7 (6) Head tilted: 90.7 (78) Chin down/up: 1.2 (1) Combined: 1.2 (1)	26.2 (21/80)	Surgery: 82.6 (71) BT: 1.2 (1) Follow-up: 16.3 (14)	Orthotropia (43) Vertical deviation (32.6) Esotropia (16.3) Esotropia + vertical deviation (3.5) Exotropia + vertical deviation (2.3) Exotropia (2.3)	Fusion: 31 (18/58) Stereopsis: 70.8 (34/48)
Duane retraction syndrome	16.9 (29)	Head turned: 93.1 (27) Combined: 6.9 (2)	26.9 (7/26)	Follow-up: 44.8 (13) Surgery: 41.4 (12) BT: 13.8 (4)	Esotropia (55.2) Orthotropia (31) Exotropia (6.9) Vertical deviation (3.4) Esotropia + vertical deviation (3.4)	Fusion: 41.1 (7/17) Stereopsis: 100 (12/12)
A-V pattern strabismus	15.1 (26)	Head turned: 65.4 (17) Head tilted: 34.6 (9)	50 (11/22)	Follow-up: 65.4 (17) Surgery: 34.6 (9)	Esotropia (61.5) Exotropia (38.5)	Fusion: 35.7 (5/14) Stereopsis: 87.5 (7/8)
Nystagmus blockage syndrome	4.1 (7)	Head turned: 85.7 (6) Combined: 14.3 (1)	100 (6/6)	Follow-up: 85.7 (6) Prism: 14.3 (1)	Orthotropia (57.1) Esotropia (28.6) Exotropia (14.3)	Fusion: 25 (1/4) Stereopsis: 100 (1/1)
Sixth nerve palsy	4.1 (7)	Head turned: 100 (7)	0 (0/6)	BT: 57.1 (4) Follow-up: 28.6 (2) Prism: 14.3 (1)	Esotropia (100)	Fusion: 0 (0/5) Stereopsis: 66.6 (2/3)
Extraocular muscle fibrosis	3.5 (6)	Chin down/up: 66.7 (4) Head turned: 16.7 (1) Combined: 16.7 (1)	100 (5/5)	Follow-up: 66.7 (4) Surgery: 16.7 (1) BT: 16.7 (1)	Exotropia (33.3) Vertical deviation (33.3) Esotropia (16.7) Esotropia + vertical deviation (16.7)	Fusion: 33.3 (1/3) Stereopsis: 100 (1/1)
Third nerve palsy	2.9 (5)	Head turned: 60 (3) Head tilted: 20 (1) Combined: 20 (1)	50 (2/4)	Surgery: 80 (4) Follow-up: 20 (1)	Exotropia (60) Vertical deviation (20) Exotropia + vertical deviation (20)	Fusion: 0 (0/3) Stereopsis: 66.6 (2/3)
Brown syndrome	1.7 (3)	Head tilted: 66.7 (2) Head turned: 33.3 (1)	0 (0/2)	Surgery: 66.7 (2) Follow-up: 33.3 (1)	Orthotropia (66.7) Vertical deviation (33.3)	Fusion: 0 (0/2) Stereopsis: 100 (1/1)
Oculocutaneous albinism	1.2 (2)	Head turned: 50 (1) Combined: 50 (1)	100 (2/2)	Follow-up: 100 (2)	Orthotropia (50) Esotropia (50)	Fusion: 50 (1/2) Stereopsis: 0 (0/1)
Heavy eye syndrome	0.6 (1)	Chin down/up: 100 (1)	100 (1/1)	Surgery: 100 (1)	Esotropia + vertical deviation (100)	Fusion: 0 (0/1) Stereopsis: 100 (1/1)

AHP: Abnormal head posture, BT: Botulinum toxin, n: Number of patients

Table 2. Changes in AHP by treatment method (n=94)

Treatment	Change in AHP, % (n)		
	AHP reduced	AHP resolved	AHP remained the same
Surgery (n=66)	77.3 (51)	16.7 (11)	6.1 (4)
Botulinum toxin (n=4)	50.0 (2)	25.0 (1)	25.0 (1)
Follow-up (n=24)	8.3 (2)	0 (0)	91.7 (22)

AHP: Abnormal head posture, n: Number of patients

Moreover, the most common AHP type in our study was head tilt, followed by head turn, combined, and chin down/up positions. This finding is similar to the frequency ranking of AHP types in the study by Erkan Turan et al.[5] In contrast, Akbari et al.[8] observed in their study that the predominant AHP type was head turn (48.3%), followed by head tilt (24.8%), combined (20.8%), and chin up (6%) positions.

Chin up or down head positions may develop due to orbital base fractures, thyroid myopathy, A-V pattern esotropia/exotropia, double elevator palsy, extraocular muscle fibrosis, congenital motor nystagmus, and congenital ptosis.[9] In our study, the chin down/up head position was the most common AHP type among patients with extraocular muscle fibrosis and heavy eye syndrome.

In nystagmus blockage syndrome, AHP occurs when the null point (the point where nystagmus is least noticeable and visual acuity is best) is not at primary gaze position. It can be variable but is often observed as the head turning away from the null point.[10] In our study, 85.7% of patients diagnosed with nystagmus blockage syndrome presented with head turn, whereas this rate was 25% in the study conducted by Erkan Turan et al.[5]

Patients with superior oblique palsy, which is one of the leading ocular causes of AHP, are expected to exhibit head tilt with chin down and ipsilateral head turn. However, some cases may present with only head tilt or only head turn.[3] Erkan Turan et al.[5] reported that 87.3% of patients with superior oblique palsy had head tilt, 3.6% had head turn, 7.3% had combined head tilt and turn, and 1.8% had a chin down position. Nucci et al.[11] reported that of 12 AHP patients with superior oblique palsy, 10 had head tilt and 2 had a combined head position. Similarly, the fourth cranial nerve palsy patients in our study mostly exhibited head tilt (90.7%), with head turn (7%), chin down/up (1.2%), and combined (1.2%) being less common AHP types.

The compensatory head posture is a motor adaptation developed to achieve binocular single vision.[12] The development of AHP may not be expected in the presence of amblyopia or suppression because AHP is a compensatory mechanism that occurs in patients whose fusion capacity and visual acuity are over a certain level.[13] In our study, 30.6% of the patients had fusion, 14.8% had diplopia, and 54.6% had suppression. However, similar to the study conducted by Erkan Turan et al.[5], no significant relationship was found between Worth 4-dot test results and amblyopia. In addition, the significant relationships between amblyopia and both diagnosis and AHP type demonstrated in our study emphasizes that amblyopia should be considered in diseases such as nystagmus blockage syndrome, oculocutaneous albinism, and extraocular muscle fibrosis. Although the relationship between AHP type and amblyopia was statistically significant, drawing a definitive conclusion may not be advisable due to the varying number of patients in the AHP groups.

Different treatments can be used to correct AHP, depending on the etiology and orthoptic examination findings. Teodorescu[1] explained the mechanism of AHP as "the head moves where the eye cannot, to avoid diplopia" and emphasized that an adequate surgical treatment can correct deviation, diplopia, and AHP. Zheng et al.[14] reported resolution of infantile nystagmus syndrome in 95% of patients at postoperative 1-3 weeks and resolution of AHP in 82% of patients at 2-year follow-up. Gündüz et al.[15] reported that AHP resolved postoperatively in all patients with Duane retraction syndrome, primary inferior oblique hyperfunction, and superior oblique palsy. In our study, we observed AHP reduction in 77.3% and resolution in 16.7% of patients treated surgically.

AHP may also develop to provide binocular vision and prevent diplopia in patients with congenital non-progressive Duane retraction syndrome, which is characterized by limitation of horizontal eye movements and globe retraction. Prism glasses or extraocular muscle surgery can be performed to correct the compensatory head position.[16] Surgery may also be useful in correcting cosmetically intolerable globe retraction and upward or downward displacement of the affected eye upon adduction.[17] In our study, AHP resolved or decreased in 8 of 12 patients with Duane retraction syndrome who underwent corrective surgery.

Study Limitations

Our study has the limitations inherent to retrospective research. In addition, some of the patients included in our study were too young for binocularity, stereopsis, and amblyopia assessment. The fact that some patients did not continue follow-up also resulted in the inability to observe their post-treatment change in AHP. A study with larger diagnostic groups can be conducted to obtain more detailed results.

Conclusion

AHP can be caused by many different conditions. The results of our study revealed a significant relationship between patients' AHP types and diagnoses, which suggests that a patient's AHP may provide clues to their diagnosis at first sight. However, conducting a detailed examination for amblyopia, binocular vision, and eye movement limitations is still essential. The association between amblyopia and both AHP type and diagnosis highlights the need to be vigilant for amblyopia depending on the patient's diagnosis and AHP type. Reduction or resolution of AHP was noted in most patients treated by surgery or botulinum toxin administration. In light of this information, it should be kept in mind that AHP can be modified with a correct diagnosis and the right treatment.

Ethics

Ethics Committee Approval: Ethics Committee of Ondokuz Mayıs University (decision no: OMÜ KAEK 2022/563, date: 28.12.2022).

Informed Consent: Retrospective study.

Declarations

Authorship Contributions

Surgical and Medical Practices: L.N.Ş., Concept: L.N.Ş., Design: L.N.Ş., B.E., Data Collection or Processing: B.E.,

Analysis or Interpretation: B.E., Literature Search: B.E., Writing: B.E.

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