Clinical features of pediatric ocular torticollis in Dakahlia Governorate, Egypt

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Short title: Pediatric ocular torticollis

Abstract

Purpose: To determine the relative frequency of the most common ocular causes and types of torticollis in pediatric patients presented with abnormal head posture (AHP) attending pediatric ophthalmology and strabismus clinic of Mansoura Oophthalmic Center.

Method: A descriptive crosssectional study has been done. A total of 300 pediatric patients presented with abnormal head posture attending pediatric ophthalmology and strabismus clinic of Mansoura Ophthalmic Center underwent complete ophthalmologic examination. Demographic and orthoptic findings were recorded.

Results: Among the 300 patients, there were 159 females (53%) and 141 males (47%). The median age was 7 years (range 6 months to 18 years). Head tilt was the most common AHP pattern observed (68.7%) followed by combined variety (14.7%) then face turn position (12.3%) and finally abnormal chin position (4.3%). The most common ocular causes of torticollis were superior oblique palsy (SOP) in (60%), dissociated vertical deviation (DVD) in (10%), Duane syndrome in (7%) and nystagmus in (5%). Amblyopia was detected in 115 patients (38.3%). Lang test was negative even while adopting the AHP in (26%) of examined patients.

Conclusion: Our study has revealed a wide variety of AHP patterns, causes and clinical features among pediatric patients presented with torticollis. Head tilt was the most common AHP pattern in our study. Different patterns of AHP can occur in patients with the same diagnosis. Superior oblique palsy, DVD, Duane retraction syndrome and nystagmus were the most common causes of AHP in our study cohort. Amblyopia and defective binocular vision represented a great concern in our cases. A population-based study is recommended for better evaluation of ocular torticollis among Egyptian population.

Key words: Torticollis, AHP, strabismus, Nystagmus, Diplopia.

INTRODUCTION:

Torticollis is a deformity of head and neck position[1]. The head forms an angle with the body on the horizontal, vertical or anteroposterior axis[2].

The prevalence of AHP is about 3% of pediatric ophthalmology practice[3]. However, Dumitrescu has reported a higher prevalence (24.7%) in patients with Down syndrome[4]. It is presented in a form of head tilt, face turn, abnormal chin positions or a combination of previous patterns[5, 6].

Torticollis may be congenital or acquired. Congenital torticollis is usually presented since birth due to contracture or fibrosis of the cervical muscles[7]. Acquired Torticollis may occur due to trauma, inflammation, tumors or psychological disorder[8].Torticollis is posttraumatic in (10-20%) of cases[9, 10].

Ocular problems account for up to 23% of all cases of torticollis[11]. Strabismus such as (superior oblique palsy, lateral rectus muscle palsy, pattern strabismus, Duane's retraction syndrome and Brown's syndrome), nystagmus, and refractive error are the most common ocular causes of torticollis. Most cases of pediatric ocular torticollis are usually presented within the first few months after birth[5].

Appropriate management and treatment of patients presented with abnormal head posture is very important for prevention of amblyopia, loss of binocularity and permanent neck deformities as a result of muscular and soft tissue changes[12].

Careful etiological diagnosis is very important in management as each ocular cause has a different strategy of

treatment[13].

Several studies have been conducted in developed countries about torticollis, but there was no studies conducted in developing countries (including Egypt) about the most common causes and types of ocular torticollis.

Our study has been conducted to investigate the most common ocular causes, types and clinical features of torticollis in pediatric patients presented with abnormal head posture attending pediatric ophthalmology and strabismus clinic of Mansoura ophthalmic center.

PATIENTS AND METHODS:

This is a cross sectional, observational non-interventional study that was conducted in the period between March 2021 and March 2022 in Mansoura ophthalmic center, Mansoura University, Egypt.

This study included total number of 300 children with age up to 18 years of age who were presented with abnormal head posture attending pediatric ophthalmology and strabismus clinic.

Age more than 18 years old, neurological disorder and non-ocular causes were excluded from the study.

Full history was obtained from the patients including birth history and history of growth and development.

Full detailed ophthalmic examination was done for all cases of the study. Visual acuity (VA) assessment in co-operative patients was done using Landolt's VA chart in older children and Matching test by lea charts (symbols charts) in younger children.

Objective refraction was done using autorefractometer followed by subjective refraction with trial frame and computerized visual acuity testing system. Cycloplegic refraction was obtained using cyclopentolate 1%.

Uniocular patch test was used to differentiate between ocular and non-ocular causes of pediatric torticollis.

The types of abnormal head posture are (head tilt, face turn, chin up or down or a mixed type of previous positions). The angle of abnormal head posture was measured using orthopedic goniometer.

Corneal light reflex tests and cover tests were used to assess ocular alignment. Alternate prism cover test is used to quantify the angle of misalignment. Ocular motility assessment was done for both ductions and versions in both eyes. Lang stereotest was used for assessment of binocular vision and stereopsis while the child was adopting the AHP position.

Nystagmus was described according to plane, manifest, latent or manifest-latent nystagmus, bilateral or unilateral, Conjugate or Disconjugate and jerk nystagmus or pendular nystagmus

The study was conducted after approval from the Institutional Review Board of Mansoura faculty of medicine (IRB) (approval number: MS.21.08.1602.R1) and obtaining an informed written consent from the legal guardians of all included participants.

Statistical analysis:

Statistical analysis was done using (SPSS 24.0, IBM/SPSS Inc., Chicago, IL) software for analysis. Baseline characteristics of the study population were presented as frequencies and percentages (%) or mean values ± standard deviations (SD) and median (Range).

Chi-Square test was used to compare two or more independent groups of qualitative data. Kruskal -Wallis test was used to compare between more than two independent groups with quantitative non-parametric data. Probability (p value) ≤ 0.05 was considered to be statistically significant.

RESULTS:

The current study included 300 pediatric patients presented with abnormal head posture to strabismus clinic of Mansoura ophthalmic center. The median age of the children was 7 years (6m-18y). There were 141 males (47%) and 159 females (53%). Positive family history was found in 30 children (10%).

The presenting complaints in the included children were strabismus in 287 children (95.7%), blurred vision in 128 children (42.7%), AHP in 114 children (38%), nystagmus in 17 children (5.7%), diplopia in 7 children (2.3%) and ptosis in 2 children (0.67%).

The best corrected visual acuity (BCVA) could be assessed in 232 children (77.3%). The median BCVA (logmar) was 0.2 (0 – 1.1). Amblyopia was detected in 115 patients (38.3%) and there was no significant difference in the prevalence of amblyopia in different patterns of AHP (p=0.425).

The most common types of cycloplegic refraction in the included children were hyperopia + astigmatism in 177 children (59%) followed by myopia + astigmatism in 56 children (18.7%), astigmatism in 29 children (9.7%), myopia

in one eye and hyperopia in the other eye in 17 children (5.7%) with no anisometropia, myopia in 11 children (3.7%) and finally hyperope in 10 children (3.3%). The median for myopic spherical equivalent was -1 with range**[**0 - (-15.75)**]** and the median for hyperopic spherical equivalent was +1 with range [0 - (+9.25)].

Limited ocular motility was shown in 112 eyes (18.67 %) of 85 children (28.3%). limited abduction in 83 eyes (74.1%), limited adduction in 36 eyes (32.1%),limited upward motility was shown in 22 eyes (19.6%) and limited downward motility in 14 eyes (12.5%).

The most common type of AHP was head tilt position in 206 children (68.7%) followed by mixed position in 44 children (14.7%) then face turn position in 37 children (12.3%) and finally abnormal chin position in 13 children (4.3%). The median angle of face turn was 9° with range (1° - 28°), In head tilt, it was 8° with range (1° - 17°) and in abnormal chin positions, it was 6° with range (-9° - 15 °. Types of AHP in the cases of the study are illustrated in figure (1).

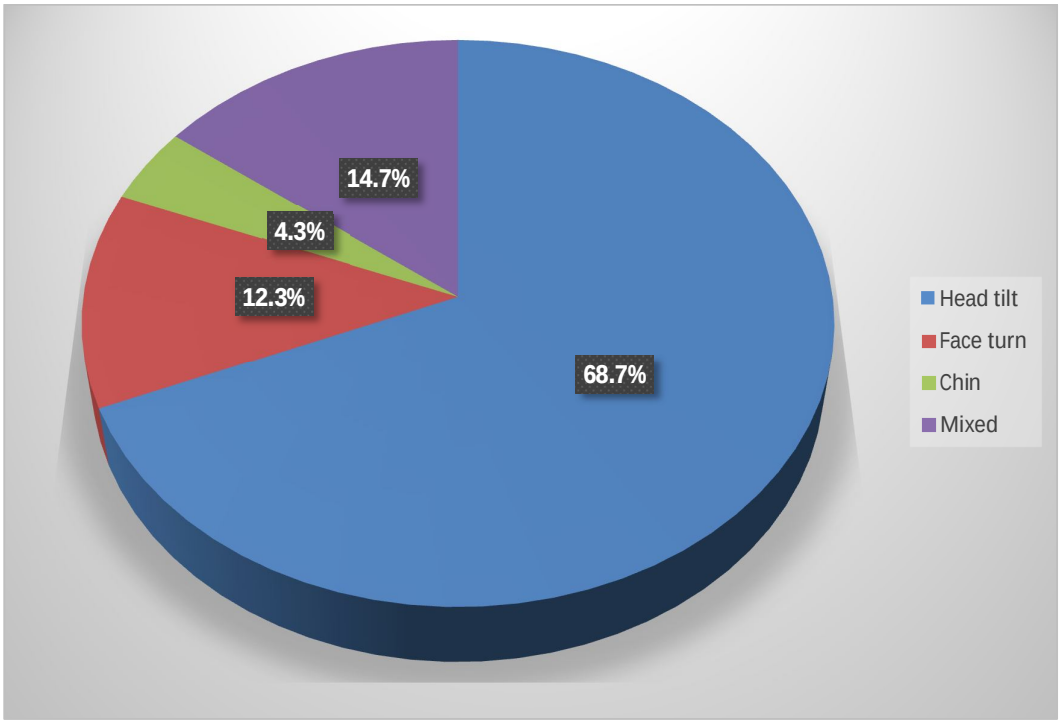

Figure (1): Types of AHP in the cases of the study.

The most common diagnoses in patients with AHP were SOP in (60%) followed by DVD in (10%), Duane syndrome in (7%) and nystagmus in (5%). As regard the different patterns of AHP in our study, Inferior oblique overaction (IOOA) was the most common cause of the head tilt position (68.9%), Duane syndrome was the most common cause of the face turn position (45.9%), Pattern of deviation was the most common cause of the abnormal chin position (30.8%), DVD was the most common cause of the mixed position (38.6%) (Table 1).

Table (1): Main causes of AHP in the cases of the study.

	Head tilt (N=206)	Face turn (N=37)	Chin (N=13)	Mixed (N=44)	Total (N=300)
Esotropia	3	2	0	1	6 (2%)
Exotropia	0	7	0	0	7 (2.33%)
Pattern of deviation	2	1	4	1	8 (2.67%)
DVD	11	2	0	17	30 (10%)
SOOA	1	0	0	0	1 (0.33%)
Hypertropia	1	0	0	0	1 (0.33%)
Hypotropia	3	1	2	1	7 (2.33%)
Duane syndrome	1	17	0	3	21 (7%)
Brown Syndrome	0	0	0	3	3 (1%)
MED	1	0	3	2	6 (2%)
CFEOM	1	0	2	0	3 (1%)
IOOA	142	2	1	5	150 (50%)
SOP	28	0	0	2	30 (10%)
Sixth nerve palsy	2	2	0	0	4 (1.33%)
Third nerve palsy	3	1	0	1	5 (1.67%)
Ptosis	0	0	1	1	2 (0.67%)
Astigmatism	1	0	0	0	1 (0.33%)
Nystagmus	6	2	0	7	15 (5%)

SOOA: Superior Oblique Overaction.

MED: Monocular Elevation Deficiency.

CFEOM: Chronic Fibrosis of the Extra-Oocular Muscles.

In the cases of the study, strabismus was the main cause of AHP in 282 (94%). Paralytic strabismus was found in 189 (67%) of cases and restrictive strabismus was found in 40 (14.2%) of cases.

14 children (4.7%) have shown patterned deviation. A pattern was documented in 5 children and V pattern was documented in 9 children.

Nystagmus was found in 17 children (5.7%) and this included vertical nystagmus in 1 child and horizontal nystagmus in 16 children. Nystagmus was manifest in12 children, manifest/latent in 4 children and latent in 1 child. Nystagmus was unilateral in 3 cases and bilateral in 14 cases. Nystagmus was jerk in 11 children and pendular in 6 children. Head nodding was detected in 1 case.

Ptosis was detected in 13 children (4.3%), it was the main cause of the abnormal head posture in 2 cases only (severe ptosis). Ptosis in the other cases was mild or moderate and was accompanied with the main cause AHP such as (CFEOM, MED and third nerve palsy).

The Lang test was done in 235 children (78.3%) while they were adopting their AHP position and it showed positive results in 74% of them.

DISCUSSION:

Abnormal head posture is an alert, commonly missed sign in the pediatric age group. It can be easily overlooked by parents, teachers and pediatricians. The prevalence of AHP in ophthalmic practice is 3.19% in pediatric age group[3]. Patients with an underlying ocular pathology usually adopt an AHP to avoid diplopia and obtain better fusion and binocular vision. Long standing AHP may cause secondary stiffness of the neck muscles, facial asymmetry or even scoliosis[11].

To our knowledge, no previous work has comprehensively evaluated the prevalence of ocular causes, types and specific features of ocular torticollis among pediatric Egyptian patients. Therefore, our study with a descriptive cross-sectional design was conducted at the Strabismus Clinic of the

Mansoura Ophthalmic Center, where a cohort of 300 pediatric patients up to 18 years of age was comprehensively evaluated with the primary aim to investigate the most common ocular causes, types and clinical features of ocular torticollis in these patients.

The current study found that the most common AHP pattern was "head tilt" (68.7%), followed by "mixed position"(14.7%) then "face turn"(12.3%) and finally "abnormal chin position"(4.3%). These findings align with similar study conducted by Kadriye Turan and colleagues who have reported that head tilt (45.4%) and face turn (36.8%) were the most frequently occurring types of AHP in their study [(14)]. To the contrary, Dumitrescu and colleagues reported that chin up position (26.6%) was the most common AHP pattern in children with Down syndrome[4]. Boricean and colleagues have reported that face turn was the most frequent type of AHP in studied children[3]. Additionally, Turan in another study reported that face turn position also represented 40 % of their patients[15].

In the current study, the most common ocular causes of torticollis were SOP in (60%), DVD in (10%), Duane syndrome in (7%) and nystagmus in (5%). In concurrence with our research, Both Mitchell and Dumitrescu have reported incomitant strabismus as the most common cause of AHP in their patients[4, 16]. In another study conducted by Nucci and colleagues, study included 25 patients with ocular torticollis. They observed that superior oblique palsy was the main cause of AHP in 12 patients[17]. Turan and colleagues reported fourth nerve palsy (33.7%), Duane retraction syndrome (21.5%) and six cranial nerve palsy (11%) as the most common underlying causes for AHP in their prospective study[14].

On the other hand, Turan and colleagues in another study reported that comitant strabismus, nystagmus and Duane retraction syndrome were the most frequent causes of AHP in the cases of their study[15].

Each specific cause has a wide variety of AHP pattern presentations. However, IOOA was the most common cause of head tilt. Duane retraction syndrome was the commonest cause of face turn. Pattern of deviation was the commonest cause for abnormal chin position. DVD and nystagmus were the commonest causes for mixed position. This variability in presentation was detected also in previous studies conducted by Nucci, Lee,et al and Spielmann[18,19,20]. So, each patient with abnormal head posture should be examined thoroughly for different diagnoses

Our study was conducted on pediatric patients only with median age 7 years (range 6 months-18 years). In concurrence with our research, Nucci study was conducted on children with median age 39 ± 27 months [(18)]. On the other hand, Turan and colleagues have conducted 2 studies on both pediatric and adult patients[14, 15].

In the current study, strabismus was the main presenting complaint (95.7%), AHP in (40%) only and diplopia in (2.3%). AHP is an easily missed symptom and sign, it was the presentation in (40%) of our patients only. Therefore, Detection of AHP depends basically on clinical suspicion and experience particularly in pediatric age group[15].

Amblyopia was present in 38.3% of our pediatric patients with torticollis. This prevalence was higher than what was reported in Turan's study (28.2%) with no significant difference in the prevalence of amblyopia in different patterns of AHP in both studies[14]. This reinforces the notion that amblyopia can be present irrespective of the specific AHP pattern adopted.

In the current study, stereopsis was tested in 235 children (78.3%) by Lang test while they were adopting their AHP and showed positive results in 74% of them which was higher than what was reported in Kadriye Turan's study where 128 patients were examined for stereopsis and only 43.8% of them had positive results[14].

The strength of our study lies in its comprehensive evaluation of ocular torticollis in Egyptian pediatric patients. It is the first large prospective study (300 patients) to deal with the prevalence of underlying ocular causes for this important easily missed symptom and sign in pediatric ophthalmology clinic.

Despite the promising outcomes in the current study, the following limitations have to be taken into consideration: Firstly, our study cohort consisted of pediatric patients attending strabismus clinic, a large population-based study is required. Secondly, we didn’t do a quantitative assessment of facial asymmetry in our cases, although it is a potential complication of ocular torticollis.

CONCLUSION:

Abnormal head posture is an alert, commonly missed symptom and sign in the pediatric age group. It can be easily overlooked by parents, teachers and pediatricians. SOP, DVD, Duane retraction syndrome and Nystagmus are the main causes of torticollis in our study cohort. Head tilt was the most common AHP pattern. Different patterns of AHP can occur in patients with the same diagnosis. Amblyopia and decreased binocular vision and stereopsis were a great concern in our study cases despite the AHP adopted, so early proper intervention is mandatory.

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Data Availability: The authors declare that all data supporting the findings of this study are available within the article.

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Conflict of interest

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