

# A Multi-Disciplinary Study of the Ocular, Orthopedic, and Neurologic Causes of Abnormal Head Postures in Children

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- **PURPOSE:** To determine the relative frequency that abnormal head postures in children are caused by orthopedic, ophthalmologic, or neurologic disorders, respectively.
- **DESIGN:** A prospective, consecutive case series.
- **METHODS:** Children found to have an abnormal head posture on routine pediatric examination underwent an evaluation by a pediatric ophthalmologist, pediatric orthopedist, and pediatric neurologist. The study was conducted in northwestern Italy.
- **RESULTS:** In the 63 children evaluated, the cause of the abnormal head posture was orthopedic in 35, ocular in 25, and neurologic in 5. In 8 patients, no specific cause could be found. The most common orthopedic cause was congenital muscular torticollis, which accounted for 31 patients. The most common ocular cause was superior oblique muscle palsy, which accounted for 12 patients. In 2 patients neck muscle contracture suggested an orthopedic cause, however, the tight neck muscles were secondary to a head tilt caused by superior oblique muscle palsy.
- **CONCLUSIONS:** When the cause of an abnormal head posture is not obvious, a multi-disciplinary approach including ophthalmologic, neurologic, and orthopedic specialists may be helpful. (*Am J Ophthalmol* 2005; 140:65–68. © 2005 by Elsevier Inc. All rights reserved.)

**A**N ABNORMAL HEAD POSTURE (AHP) IS A RELATIVELY common condition in children, with an estimated incidence of 1.3%.<sup>1</sup> Common causes of AHPs include congenital muscular torticollis caused by a

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tightness of the sternocleidomastoid muscle,<sup>2,3</sup> ocular problems,<sup>3–5</sup> and disorders of the central nervous system.<sup>3</sup>

Ballock and Song attempted to evaluate the prevalence of different causes of AHPs in children.<sup>3</sup> They found that 53 (18.4%) of 288 patients seen in a tertiary care pediatric orthopedic facility for an AHP had a nonmuscular etiology. Their figures, however, may be misleading as to the actual prevalence of different causes of torticollis in children for several reasons. The study was retrospective and based in a tertiary care orthopedic clinic. Most likely, children who obviously had ocular or neurologic causes for an AHP would not have been referred to the orthopedic clinic. As a result, these figures might be skewed toward reflecting a higher percentage of orthomuscular causes for AHP. To our knowledge there is no prospective study of the prevalence of different causes of AHPs in children that does not suffer from this type of bias. The purpose of this study is to investigate the prevalence of different causes of AHPs in children presenting to primary care pediatricians.

## PATIENTS AND METHODS

FIFTY-FOUR PRIMARY CARE FAMILY PEDIATRICIANS IN northwestern Italy were recruited to participate in this study. They all participated in a preliminary course instructing them in the diagnosis of AHPs in children and in the protocol for this study. This seminar, which was taught by one of us (P.N.), included a reading assignment of selected relevant articles from the literature. All consecutive patients diagnosed as having an AHP during an office examination by one of the participating pediatricians between April 2003 and October 2003 were included in this study, subject to certain exclusion criteria as outlined below. Enrolled children were referred to San Paolo Hospital, Milan, Italy, for a multi-disciplinary evaluation. They were first examined in the pediatric ophthalmology clinic by one of two of us (P.N. or M.S.). Next, a pediatric neurologist examined the child, followed by a pediatric orthopedist. The three pediatric specialists were each

masked as to the findings of the other respective specialists. The data from the three evaluations were then analyzed by two of us (P.N. and B.J.K.). In the event of any conflicts or inconsistencies, the respective specialists discussed the patient's findings in an attempt to reach consensus as to the etiology of the AHP.

Exclusion criteria included history of prior trauma to the eyes, head, neck, or shoulder before the appearance of the AHP, an acute onset of the AHP, a history of previous severe illness that required hospitalization, or a diagnosis of psychiatric problems.

This study had approval of the University of Milan ad hoc committee and conformed to the principles outlined in the Declaration of Helsinki guiding physicians in biomedical research involving human subjects. Informed consent was obtained in writing from all parents or legal guardians.

## RESULTS

THIS SERIES CONSISTS OF 73 PATIENTS. INITIALLY 81 PATIENTS were referred to San Paolo Hospital for evaluation. Five were excluded from this study because they did not complete all of the three required evaluations, and 3 were eliminated because of one or more of the aforementioned exclusion criteria. Of the 54 pediatricians recruited to participate in this study, 4 did not refer any patients, 30 referred 1 patient, 13 referred 2 patients, 3 referred 3 patients, and 4 referred 4 patients, respectively. The mean age at the time of referral of the children in this study was 39.0 months  $\pm$  27.0; there were 38 males and 35 females. Patients with superior oblique muscle palsy tended to present at an older age (mean 78.0 months  $\pm$  13.3) than patients with other causes for an AHP (mean 32.7 months  $\pm$  25.4). This difference was significant ( $P < 0.001$ ; Student's 2-tailed  $t$  test). Table 1 presents the different etiologies found for the torticollis subdivided according to the patients' age. In all patients except 2, the three subspecialty evaluations clearly identified a single category for the etiology of the torticollis. In these 2 patients, neck muscle contracture was found, which suggested an orthopedic etiology. These 2 patients were also found to have superior oblique muscle palsy. After a discussion between the pediatric ophthalmologist and pediatric orthopedist, it was concluded the neck muscle contracture was secondary to the superior oblique muscle palsy. Consequently, these patients were included in the ocular group. In 8 patients, no specific cause for the torticollis could be found. These are listed in Table 1 as having an unknown cause. Table 2 presents the specific entities responsible for the neurologic, ocular, and orthopedic entities encountered in this study and the types of AHP they caused respectively. For the purpose of data analysis, we reserved inclusion in the "com-

**TABLE 1.** Etiology of Abnormal Head Posture as a Function of Age

Etiology	1/2 to 2 years of age, N	2-5 years of age, N	5-8 years of age, N	Total - all ages, N
<b>Ocular</b>				
Duane's syndrome	2	4	0	6
Nystagmus	2	1	1	4
SOP	0	2	10	12
Brown's syndrome	0	1	2	3
Total ocular	4	8	13	25
<b>Orthopedic</b>				
CMT	17	8	6	31
Klippel-Feil	2	1	1	4
Total orthopedic	19	9	7	35
<b>Neurologic</b>				
Psychomotor delay	0	3	1	4
Brain tumor	0	1	0	1
Total neurologic	0	4	1	5
Unknown	6	1	1	8

SOP = superior oblique muscle palsy; CMT = congenital muscular torticollis.

binéd" category in Table 2 for those patients in whom there was a substantial element of abnormal posture from more than one category. The respective specialist made that judgment subjectively. For example, many patients with superior oblique muscle palsy had a substantial head tilt but also a minimal face turn, chin elevation, or chin depression. These patients were included in the "head tilt" category instead of the "combined" category. Although no patients in Table 2 are listed as solely having a chin up head posture, this type of AHP was seen in several of the patients listed in the combined category. In general, patients with superior oblique muscle palsy predominantly manifested a head tilt, and patients with nystagmus or Duane's syndrome manifested a face turn. An AHP in the form of a face turn was exclusively caused by ocular motility problems in this series. Most of the patients with congenital muscular torticollis had a head tilt. However, there were many exceptions to these general trends so that the nature of the head posture could not reliably predict the underlying cause.

Six (24%) of the 25 patients in the ocular category and 22 (63%) of the 35 patients in the orthopedic category had a positive family history for ocular motility problems or orthopedic problems, respectively. None of the 5 patients in our neurologic group had a positive family history for neurologic problems.

Orthopedic evaluation revealed a normal range of neck motion in all cases that were in the ocular or neurologic category; however, in some there was palpable tightness in the absence of true contracture of the neck muscles.

**TABLE 2.** Type of Abnormal Head Posture by Etiology

Cause of Abnormal Head Posture	Head Tilt	Face Turn	Chin Up	Chin Down	Combined
<b>Ocular</b>					
Duane's syndrome	0	5	0	0	1
Nystagmus	0	2	0	0	2
SOP	10	0	0	0	2
Brown's syndrome	0	0	0	0	3
<b>Orthopedic</b>					
CMT	27	0	0	1	3
Klippel-Feil	0	0	0	0	4
<b>Neurologic</b>					
Psychomotor delay	2	0	0	0	2
Brain tumor	1	0	0	0	0
Unknown	1	0	0	2	5

SOP = superior oblique muscle palsy; CMT = congenital muscular torticollis.

## DISCUSSION

AHPS IN CHILDREN MAY BE CAUSED BY ANY ONE OF A number of etiologies. In our series we found that orthopedic causes were most frequent, followed by ocular disorders and neurologic problems in decreasing order of frequency.

We found congenital muscular torticollis to be the single most common cause of an AHP, accounting for 31 of our 63 patients. When this disorder is caused by an abnormality of the neck muscles, it is usually benign and self-limiting.<sup>1,6,7</sup> If the neck muscles are extremely tight, a mass can be felt in the sternocleidomastoid region, which has been referred to as pseudotumor of infancy.<sup>1,7</sup> In some cases, muscular causes of torticollis require neck muscle surgery. Other orthopedic causes of AHP include the Klippel-Feil anomaly<sup>3</sup> and brachial plexus injury.<sup>3</sup> An AHP may also be caused by the Sandifer syndrome (hiatal hernia associated with gastro-esophageal reflux),<sup>8</sup> as well as other less common conditions.

There are numerous ocular causes of AHPs. The most frequent ones include superior oblique muscle palsy, lateral rectus muscle palsy, nystagmus, vertically incomitant horizontal strabismus (A or V-patterns), Brown's syndrome, and Duane's syndrome.<sup>3-5,9</sup> These ocular conditions either result in a head tilt, a face turn, or a chin-up or down position, depending on the specific etiology involved. They frequently require eye muscle surgery to correct the abnormal head position.

Neurologic causes of AHPs include space-occupying lesions of the central nervous system, postinflammatory conditions, and focal dystonia.<sup>3,10,11</sup> Additionally, psychomotor delay can cause an AHP, because it may result in poor control of the neck muscles.<sup>12,13</sup>

In the aforementioned study by Ballock and Song, 53 of 288 patients had a nonmuscular cause for their AHP.<sup>3</sup> In these 53 patients, the AHP was attributed to the

Klippel-Feil anomaly in 16, ocular disorders in 12, brachial plexus palsy in 9, and central nervous system disorders in 6. The remaining 8 patients had a miscellaneous or an unknown cause.

In our series, superior oblique muscle palsy was the most common diagnosis in older children, possibly because this is a condition that becomes more symptomatic with time. The other causes for AHPs found in our series tend to become more evident at younger ages. The one child in our neurologic group with a brain tumor had a pontine glioma. Of the four children with psychomotor delay, two had Down syndrome; however, we could not attribute the head tilt to an associated ocular motility problem.

The majority of our patients with an AHP had congenital muscular torticollis. Nevertheless, in a substantial number of patients an underlying ocular motility problem caused the AHP. Most commonly this was superior oblique muscle palsy. The presence of a tight neck muscle did not preclude the possibility of superior oblique muscle palsy as the cause of the AHP. It is therefore important that when a child is being evaluated for an AHP, the possibility of neurologic and ocular causes must be considered in addition to congenital muscular torticollis. It would be inappropriate for a child to undergo physical therapy directed at loosening the neck muscles when there is a nonmuscular cause for the AHP. When the diagnosis is not clear, a multidisciplinary approach may be appropriate to help plan the appropriate treatment protocol.

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### **Biosketch**

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