

Long-Term Developmental Outcomes in Patients With Deformational Plagiocephaly

Robert I. Miller, MD*‡, and Sterling K. Clarren, MD*§||

ABSTRACT. *Objectives.* To determine whether there was an increased rate of later developmental delay in school-aged children who presented as infants with deformational plagiocephaly without obvious signs of delay at the time of initial evaluation.

Methods. A retrospective medical record review of 254 patients evaluated at the Craniofacial Center of the Children's Hospital and Regional Medical Center in Seattle, Washington, from 1980 through 1991 was completed. Consenting patient families were interviewed via telephone to determine what, if any, special medical or educational problems had occurred for the children who had had plagiocephaly in infancy or their siblings with normal head shapes.

Results. A total of 181 families from the medical record review could be notified about the study and 63 families agreed to participate in a telephone interview. The sample of participants for the telephone interview was random to and representative of the group as a whole. The families reported that 25 of the 63 children (39.7%) with persistent deformational plagiocephaly had received special help in primary school including: special education assistance, physical therapy, occupational therapy, speech therapy generally through an Individual Education Plan. Only 7 of 91 siblings (7.7%), serving as controls, required similar services ($\chi^2 = 21.24$). Delays could not be specifically anticipated at the time of the diagnosis of deformational plagiocephaly from any simple set of factors including treatment with helmet therapy, although affected males with reported uterine constraint were at the highest risk for subsequent school problems.

Conclusions. Infants with deformational plagiocephaly comprise a high-risk group for developmental difficulties presenting as subtle problems of cerebral dysfunction during the school-age years. There is a need for additional research on the long-term developmental problems in infants with deformational plagiocephaly. *Pediatrics* 2000;105(2). URI: <http://www.pediatrics.org/cgi/content/full/105/2/e26>; plagiocephaly, facial asymmetry, torticollis, developmental delay.

ABBREVIATIONS. CHRMC, Children's Hospital and Regional Medical Center; IIEP, Individual Education Plan.

From the *Center for Human Development and Disability, University of Washington School of Medicine, Seattle, Washington; ‡Madigan Army Medical Center, Tacoma, Washington; §Craniofacial Center, Children's Hospital and Regional Medical Center, Seattle, Washington; and the ||Department of Pediatrics, University of Washington School of Medicine, Seattle, Washington

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Reprint requests to (SKC) CHRMC, CH-47, Box C-5371, Seattle, WA 98105-0371. E-mail: sclarr@chrmc.org

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Plagiocephaly or crooked head shape can result from 3 very different etiologic processes including abnormalities in brain shape and subsequent aberrant directions in brain growth, premature fusion of a single coronal or lambdoidal suture, or prenatal or postnatal external constraint. Postnatal external constraint is by far the most common cause, although the incidence is not fully established because of the lack of consensus on the criteria to be used in making the diagnosis. Clarren et al¹ reported that the literature suggested an incidence of 1 in 300 live-born infants. Bruneteau and Mulliken² reported the incidence to be as high as 48% in otherwise healthy newborns. In this article, plagiocephaly associated with external deforming forces will be referred to as "deformational plagiocephaly." Deformational plagiocephaly may be initiated prenatally when the fetal head rests for a prolonged period on a hard surface, such as a portion of the maternal pelvis or against the limb of a sibling in a multiple birth. Such consistent fetal positioning could be attributable to maternal constraining forces or to factors within the fetus that reduce the usual rate of spontaneous movement. More commonly, deformational plagiocephaly occurs postnatally and is associated with congenital torticollis, vertebral anomalies, neurologic impairment, or forced sleeping position.³⁻⁵ Few reports document any late effects of deformational plagiocephaly other than potential cosmetic concerns and the potential for strabismus, especially involving vertical eye movements.^{6,7} Plagiocephaly may be morphometrically evident in as many as 14% of adults, but it is rarely recognized.⁸

Deformational plagiocephaly generally becomes more severe in the first weeks of life, as the infant holds his head in a fixed position (regardless of the cause for the fixed position); then the headshape begins to improve with normal developmental progression involving head control and a full range of neck motion. In ~10% of affected infants with plagiocephaly, there will be a permanent deformity with a mild to severe cosmetic effect.⁵ Patients with moderate to severe asymmetry are often referred to a craniofacial center for consideration of treatment. Cranial surgery is almost never indicated for plagiocephaly without synostosis unless the presentation is very severe. Helmet therapy is an effective treatment option based on the mechanism that pressure from a rapidly growing brain against a concave surface should round flattened areas caused by earlier pressure against a flat surface.^{5,9} Helmet therapy is gen-

crally recommended between 6 and 18 months of age in an attempt to use remaining brain growth to redirect head shape.

From 1980 to 1991, 295 patients were recorded as evaluated at the Craniofacial Center of the Children's Hospital and Regional Medical Center (CHRMC) in Seattle, Washington for plagiocephaly and consideration of helmet therapy. Patients were followed through completion of helmet therapy when indicated; however, the developmental outcome of these patients was unknown because subsequent follow-up after completion of treatment was rarely indicated. The rate of developmental delay in this patient population at the time of initial evaluation for plagiocephaly or later in life was unknown. The primary question to be addressed by this study was whether deformational plagiocephaly could be a marker for early central nervous system disorganization that might later manifest as school problems that required additional services. Subtle problems of cerebral dysfunction would include learning disabilities, language disorders, visual-perceptual problems, motor delays, or problems with attention span. If delays were found to be present, an additional question would be what early factors related to the deformational plagiocephaly were associated with this adverse outcome. The rate of strabismus, as a documented late effect of deformational plagiocephaly, was also determined.

METHODS

According to medical records at CHRMC, 295 patients had been evaluated in the Craniofacial Center for deformational plagiocephaly between 1980 and 1991. Of these, 41 charts were excluded from additional review because the actual diagnosis found in the chart was not consistent with deformational plagiocephaly, no record of a Craniofacial Center assessment was found, or, in 3 instances, the records could not be located. The remaining 254 charts were reviewed and the following information was collected: position of plagiocephaly, head shape at birth with associated deformations or malformations, evidence of other deformations or malformations, age when asymmetry was first noted, involvement with helmet treatment, and history of developmental delay at the time of presentation. Information was also collected from the medical record to allow contact with families to address the question of developmental outcome.

A letter requesting permission for a telephone interview to discuss the developmental outcomes of their children was mailed to 244 families from the previous chart review (10 families were excluded because no address was available for mailing). In 73 cases, all attempts to find an accurate address failed. The remaining potential population consisted of 181 families whose letters of invitation were not returned to us by the postal service as undeliverable. These families each presumably received a 1-page letter briefly discussing the research project and a self-addressed, stamped reply postcard to convey their decision about participating in the project. The letter identified their child as having previously been seen at CHRMC because of an asymmetric head shape. The telephone interview was described. Specifically the letter noted that "questions being addressed in this study involved learning more about how all your children have done in school and the effect of the asymmetric head shape on eye positioning," and an example question was given. The letter then dealt with clarification of the telephone call process and parent/patient rights. Finally, the letter stressed that the project involved nothing other than the single phone call.

Information obtained from parents during the telephone interview included the patient previously diagnosed with plagiocephaly and information about all siblings serving as controls. Siblings were chosen as the control group to minimize bias, although they were obviously not age- or sex-matched.

Statistics from the State of Washington were obtained to determine the percentage of students in each grade that required special services in 1997 as per the Individuals with Disabilities Education Act, Part B. This figure was shown to be fairly consistent over the last several years and was applicable to both patients and their siblings. Interview questions included: involvement in any class or program, during or after school, which was designed for children who needed additional help such as a special education class, physical therapy, occupational therapy, or speech therapy; completion of an Individual Education Plan (IEP) by the school to document these services; and/or evaluation by an "eye doctor" because of a problem with eye alignment (strabismus). Asking parents about the special services their children might have received or were receiving as a measure of developmental outcome was considered the most likely way of obtaining pertinent history of developmental delay. Only students meeting standardized criteria for a delay would qualify for special services through the schools. These special services are documented through the IEP and are reviewed with the family on an annual basis.

Data were arranged in 2×2 tables using χ^2 analysis with Yates corrections and Fisher's exact test when cell numbers were <5 for analysis.

RESULTS

In the chart review, plagiocephaly was found in 152 males (60%) and 102 females (40%). A flat right occiput was found in 161 (63%) patients, whereas 72 (28%) presented with a flat left occiput, 17 (7%) had an asymmetric but bilaterally flat occiput, and 4 (2%) had an unknown asymmetry. A total of 174 patients (69%) were born with a normal head shape of which 147 (84%) were believed to have associated torticollis. Other associations included 1 patient with hemivertebrae and 3 with a neuromotor deficit. Of the patients born with an abnormal head shape, the most common association was uterine constraint in 20 patients (42%). Other deformations were noted in 73 (29%) and malformations in 47 (19%) of the total patient population. Parents reported being aware of a head asymmetry eventually diagnosed as plagiocephaly at an average age of 1.5 months. Age at the initial Craniofacial Center appointment was 6.5 months on average. Helmet treatment was initiated for 91 patients (36%). Breech delivery was present in 15 (6%). There was a history of an ophthalmologic evaluation in 7 (3%). Finally, 14 patients (6%) were 1 of a set of twins.

History of any concerns about developmental delay per parent or physician was documented in the records of 33 patients (13%) at the time of initial evaluation. No formal diagnosis of developmental delay was made in the record for any of these patients at the time of the CHRMC Craniofacial Center evaluation. Four of the families of these 33 patients (12%) subsequently went on to participate in the telephone interview. Two of the 4 with initial gross motor concerns subsequently required special education assistance but no motor therapy. One with initial gross motor concerns required no special services during school-age years. One with a question of global delays at time of initial evaluation had persistent global delays during school-age years requiring support from all available services.

In all, 68 of the 181 families (37.6%) responded to the postcard solicitation. Sixty-three families agreed to participate in the telephone interview, 1 remained undecided, and 4 requested not to be included. Table 1 compares results from participants in the telephone

TABLE 1. Comparison Between 63 Participants and 191 Non-participants Based on Chart Review Data

	Participants	Non-participants	
Position of flattening			
Flat right occiput	39	122	$\chi^2 = .04$
Flat left occiput	19	53	$P = .85$
Head shape at birth			
Normal	45	129	$\chi^2 = .00$
Asymmetric	12	36	$P = .95$
Helmet used			
Yes	27	64	$\chi^2 = 1.42$
No	36	127	$P = .23$
Normal head shape at birth and			
Torticollis	39	108	Fisher
Other*	3	6	$P = .70$
Abnormal head shape at birth and			
Constraint	6	14	$\chi^2 = .11$
Other†	6	22	$P = .74$
Antecedents to torticollis			
Known	10	37	$\chi^2 = .62$
Unknown	29	71	$P = .43$
Associated anomalies			
Deformation	19	54	$\chi^2 = .24$
Malformation	15	32	$P = .62$
Developmental delay considered in infancy			
Yes	4	29	$\chi^2 = 2.06$
No	24	67	$P = .15$
Sex of subject			
Male	42	110	$\chi^2 = 1.27$
Female	21	81	$P = .26$
Breech birth			
Yes	7	8	Fisher
No	56	183	$P = .061$
Area of residence			
West Washington	54	108	$\chi^2 = .92$
Other	9	10	$P = .34$

* Forced sleep position, hemivertebrae, neuromotor dysfunction, unknown

† Asymmetric brain, unknown.

interview and nonparticipants. The sample of participants seemed to be random and representative of the whole for every category addressed with no significant difference between the 2 categories.

Table 2 compares participants in the telephone interview who reported any delays in school with those without any reported delays or need for special services. Delays could not be anticipated by any simple set of early factors. However, patients at highest risk included those born with an abnormal head-shape at birth secondary to constraint ($P = .015$) of which all were males. Breech placement was not statistically significant ($P = .061$). In addition, helmet therapy was not a risk factor for a subsequent history of delays ($P = .91$).

Table 3 compares participants in the telephone interview with their siblings in regards to history of need for special services during the school-age years. The need for any special services frequently documented on an IEP to included special education assistance, speech therapy, physical therapy, or occupational therapy was dramatically different compared with siblings ($P = .000041$). Controlling for gender, a significant difference was found for males ($P = .0003$) but not for females ($P = .13$).

TABLE 2. Comparison Between 25 Delayed Participants and 36 Not Delayed Participants Based on Interview

	Delayed	Not Delayed	
Position of flattening			
Flat right occiput	17	22	$\chi^2 = .97$
Flat left occiput	5	14	$P = .33$
Head shape at birth			
Normal	16	29	Fisher
Asymmetric	5	7	$P = .74$
Helmet used			
Yes	11	16	$\chi^2 = .01$
No	14	22	$P = .91$
Normal head shape at birth and			
Torticollis	14	25	Fisher
Other*	1	2	$P = 1.00$
Abnormal head shape at birth and			
Constraint	5	1	Fisher
Other†	0	6	$P = .015$
Antecedents to torticollis			
Known	5	5	$\chi^2 = .48$
Unknown	9	20	$P = .45$
Associated anomalies			
Deformation	7	12	$\chi^2 = .02$
Malformation	5	10	$P = .88$
Developmental delay considered in infancy			
Yes	3	1	Fisher
No	10	14	$P = .31$
Sex of subject			
Male	20	22	$\chi^2 = .12$
Female	5	16	$P = .12$
Breech birth			
Yes	20	22	Fisher
No	5	16	$P = .061$

* Forced sleep position, hemivertebrae, neuromotor dysfunction, unknown

† Asymmetric brain, unknown

Questions about attention deficit hyperactivity disorder were not specifically asked during the telephone interview; however, parents provided this information when discussing services on the IEP for 4 of the patients previously diagnosed with plagiocephaly and in none of the siblings ($P = .03$). In regards to the type of therapy required for the patients with plagiocephaly, 10 received speech therapy, 1 occupational therapy, and 1 physical therapy. The 2 siblings who required therapy services both received speech therapy. The percentage of patients on an IEP with plagiocephaly was 34.9% and their siblings 6.6%. The percentage of children qualifying for special services on an IEP in the state of Washington for 1997, as per the Individuals with Disabilities Education Act, Part B, was 11.9%.

Strabismus was not found to be statistically significant in this study, with only 1 patient with plagiocephaly and 1 sibling with a history of strabismus resulting in evaluation and treatment by an ophthalmologist.

DISCUSSION

The population of children with deformational plagiocephaly evaluated at CHRMC from 1980 through 1991 was consistent with previous studies in regards to position of plagiocephaly. Right-sided occipital flattening was more common as previously reported with percentages ranging from 61% to 73%,

TABLE 3. Comparisons Between 63 Participants and Their 91 Siblings Based on Interviews

	Participants	Siblings	
Was there an IEP?			
Yes	22	6	$\chi^2 = 18.4$
No	40	84	$P = .00002$
Were there special education classes?			
Yes	16	6	$\chi^2 = 9.27$
No	47	85	$P = .002$
Was there ST/OI/PT?			
Yes	12	2	$\chi^2 = 10.8$
No	51	89	$P = .00099$
Was there a diagnosis of ADHD?			
Yes	4	0	Fisher
No	59	91	$P = .03$
Was there any form of special help?			
Yes	25	7	$\chi^2 = 21.24$
No	38	84	$P = .0000041$
Sex difference--males			
Delays	20	2	$\chi^2 = 17.3$
No delays	22	39	$P = .0003$
Sex difference--females			
Delays	5	4	Fisher
No delays	16	41	$P = .13$

ST/OI/PT indicates speech therapy/occupational therapy/physical therapy; ADHD, attention deficit hyperactivity disorder (any type)

compared with 63% in this study.^{7,10,11} Bruneteau and Mulliken² reported torticollis as an associated finding in 64% of infants with deformational plagiocephaly; however, our population had a higher association with torticollis in 84%. Bruneteau and Mulliken also noted other congenital anomalies in 43% of patients, which was similar to 47% of patients in this sample with other deformations or malformations. Demographics were similar to the results reported by Hellbusch et al⁹ with the age of deformity first being noted at an average age of 1.6 months and the age that the patient was first seen in a doctor's office reported to be an average of 7.3 months. Results from the CHRM population was an average age of 1.6 months and 6.5 months, respectively.

Concern about developmental delay in children who have a history of plagiocephaly secondary to deformation has not previously been evaluated in the literature. There is reportedly no increased risk of functional neurologic disability in patients with plagiocephaly without synostosis.¹² Rekate⁸ stated that there were few reports documenting any late effects of occipital plagiocephaly in his comprehensive critical review of the literature. Late effects discussed by Rekate included visual disturbances, orthodontic/oral surgical issues, cosmetic concerns, and psychosocial issues. A statement was made in that review that the possibility that cranial distortion without increased intracranial pressure or definable damage being done to the distorted underlying areas of the brain could lead to overt or subtle neuropsychological problems, had not been reported to that point. Although this study does not attempt to explain an underlying mechanism, there clearly seems to be a difference in children with plagiocephaly, compared with their siblings in regards to need for special

services during the school-age years. Problems noted related to subtle problems of cerebral dysfunction, which were manifest during the school-age years, involving language disorders, learning disability, and attention deficits. These are clearly issues that primary care providers need to be aware of, because they may care for children in the high-risk group.

If others can confirm that deformational plagiocephaly is associated with increased rates of behavioral and learning problems during the school-age years, then the question to be asked is: "Is plagiocephaly an early sign of subtle brain dysfunction or does early brain molding lead to subtle brain dysfunction?" Referrals to craniofacial centers for evaluation of deformational plagiocephaly and brachycephaly are increasing.⁸ This increase in deformations has been temporally linked to the Back to Sleep program advanced by the American Academy of Pediatrics in 1992 that advises the avoidance of the prone sleeping position as a method of reducing the rates of sudden infant death syndrome.^{10,12,13} There is a delay in early gross motor milestones in children forced to sleep supine but these delays seem transient and have not been linked as yet to any longer term problems.¹⁴ Children who are encouraged to sleep on their backs and develop abnormal head shapes as a result are a different population than children who spontaneously restricted their movement in bed for one reason or another. It will be important to follow both groups over the next several years and document the rates of attentional and cognitive disabilities that arise.

The use of helmet therapy did not seem to affect the rate of developmental delay. A total of 44% of the delayed patients had worn helmets, whereas a total of 42% of the nondelayed patients had worn helmets. The sample seemed large enough to conclude that if the additional pressure on the head from the helmet contributed to the delay, it must be a very small part of the variance.

A limitation of this study was the number of patients included in the telephone interview compared with the chart review because of an inability to contact families previously evaluated in the Craniofacial Center. We were unable to contact 73 families because of an inaccurate address and telephone number. Two thirds of the families who presumably received our letter of introduction and invitation did not respond in any way. This is a typical response rate for a survey and obviously we could not ask the families who did not respond why they did not respond. The families who were eventually contacted were shown to be a random and representative sample of the whole, based on comparison data in the charts. The reader might speculate that families with concerns about their child's development might be more likely to reply to the letter, but most parents involved with the telephone interview did not have significant complaints about their child's developmental progress. Further, only 12% of families who had had an infant noted to have delays, and therefore, at risk for additional neurodevelopmental problems responded to our invitation, compared with 44% of families in which no early delay had

been found. Although all the interviewees were willing to discuss their children's needs for special services in the school setting, most parents reported an expectation of being asked questions about cosmetic appearance and social acceptance. Of note, most parents reported no significant psychosocial issues related to persistent facial asymmetry. It was the opinion of the interviewers that the families involved with the telephone interview were not a group in search of additional services, and they did not associate their child's need for special school services with a previous diagnosis of deformational plagiocephaly.

An additional question considered in this study as part of the telephone interview was whether patients with persistent plagiocephaly have an increased rate of strabismus related to their underlying craniofacial asymmetry. Synostotic plagiocephaly is known to be associated with vertical strabismus and contralateral head tilt.⁷ The underlying mechanism resulting in strabismus involves traction on the ocular globe induced by bone deformation caused by craniosynostosis.⁶ ReKate⁸ reports that strabismus, especially involving vertical eye movements, is common in the more severe forms of this disorder. A study by Fredrick et al⁷ evaluated 13 patients with deformational plagiocephaly, of which 9 were found to have ipsilateral torticollis and 1 of these presented with strabismus. The need for early identification of strabismus is based on the development of binocular vision by ~6 months of age, whereby late correction may be an obstacle to the development of normal visual function.⁶ History of strabismus of any type or need for ophthalmology evaluation was found in only 7 patients during the chart review. One of these 7 patients with plagiocephaly was part of the telephone interview, and only 1 sibling had a history of strabismus. The results of this study do not suggest a need for increased awareness of this issue by providers caring for children with deformational plagiocephaly.

Primary care physicians commonly diagnose deformational plagiocephaly early in its course and offer appropriate recommendations for prevention of additional cranial distortion. However, even with appropriate intervention, there will continue to be a subset of children with persistent cranial asymmetry

in need of additional evaluation and follow-up. Children with deformation plagiocephaly in the high-risk group, which includes males with abnormal head shapes at birth associated with uterine constraint, need to be closely followed for potential developmental delay presenting as subtle problems of cerebral dysfunction during the school-age years. If concerns arise, timely evaluation is indicated to ensure that children in the high-risk group are receiving appropriate services based on their underlying special needs.

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