ORIGINAL RESEARCH

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Cosmetic and Cognitive Outcomes of Positional Plagiocephaly Treatment

Abstract

Purpose: Positional plagiocephaly is an acquired deformation of an intrinsically normal infant skull by sustained or excessive extrinsic forces. Non-surgical techniques include counter-positioning, supervised prone time and orthotic molding for more refractory cases. Long-term effects of positional plagiocephaly on development remain undefined, and this study evaluated cosmetic and cognitive outcomes of plagiocephaly management.

Method: Surveys were administered to parents of patients treated for positional plagiocephaly through the Children's Hospital of Eastern Ontario. Categorical responses interrogated cosmetic outcome, school performance, language skills, cognitive development and societal function. Pearson coefficient analysis tested outcomes dependency on gender, age, and plagiocephaly side at the 0.05 level of significance.

Results: Eighty respondents (51 male, 29 female) were divided as 58 right- and 22 left-sided pathology. Positional therapy was uniformly applied, and a helmet orthosis was utilized in 36% of cases. Median follow-up age was nine years with normal head appearance in 75% of cases. Only 4% of parents and 9% of patients observed significant residual asymmetry. These results did not vary by gender, age or deformity side. Left-sided disease predicted poorer language development and academic performance. Expressive speech abnormality occurred in twice as many patients with left-sided disease (36% versus 16%, $p=0.04$) along with three-fold greater special education requirements (27% versus 10%, $p=0.04$).

Conclusions: Non-surgical plagiocephaly management achieved good cosmetic outcome among patients in this study. Children with left-sided disease frequently encountered difficulties with cognitive and scholastic endeavors, although the roles of the underlying disease and the treatment measures in this delay cannot be differentiated.

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Positional plagiocephaly (PP) is an acquired cranial deformational malformation observed in children of normal development and in whom presentation of craniosynostosis has been excluded. The most consistent presentation is one of occipital flattening, with advancement of the ipsilateral ear and ipsilateral frontal bossing; features that provide differentiation from the isolated and rare lambdoid suture synostosis. The incidence of this abnormality has been climbing in the last two decades owing to the initiatives against sudden infant death syndrome (SIDS) with consequent supine positioning during infant sleeping. While such maneuvers are effective to decrease the likelihood of SIDS by approximately 40%, a remarkable increase in the incidence of positional posterior plagiocephaly (PP) has been consistently observed [1]. The deformity is more common among males and more frequently right-sided. While PP occurred in 1 in 300 live-births in 1974, the reported incidence in 1996 was 1 in 60 [2]. Other common causes of PP include congenital torticollis, fetal positioning in utero, abnormal birth position, and a low level of activity have also been implicated in the development of PP [3].

Positional plagiocephaly is usually diagnosed early in life, with most cases being mild and self-limited, resolving with conservative management. Active re-positioning during sleep and play, physiotherapy to address torticollis and stretch otherwise tight musculature, and orthotic molding helmets can improve the cosmetic outcome, with surgery almost never required. In more severe cases, orthotic molding can provide for more complete and rapid resolution of PP than with re-positioning alone, with more favorable cosmetic outcomes observed when such treatment is applied earlier in the disease course between 6 and 12 months [4]. Mulliken and coworkers [5] describe the natural history of plagiocephaly without intervention to yield residual deformity among 45% of patients at 24 months of age. The literature surrounding the cognitive outcomes among patients treated for positional plagiocephaly remains scarce and ill-defined.

Objective

The objective of this work was to define the cosmetic and cognitive outcomes observed following management of positional plagiocephaly at the Children's Hospital of Eastern Ontario. An extension of this analysis was to evaluate if the side of plagiocephaly could impact on the child's neurocognitive development and outcome. Such information remains uncertain in the literature surrounding positional plagiocephaly, far more defined among patients with underlying structural craniosynostosis.

Methods

This study was a retrospective analysis of patients managed for plagiocephaly at CHEO from 1996 to 2000 after 2000, please add "...in a dedicated clinic for patients with positional plagiocephaly, developed and coordinated by the senior author (M.V.) and managed by a nurse practitioner. Research ethics board approval was obtained to initiate data accumulation and patient contact. Patient consent was not required for the retrospective component of this work with de-identified subjects. Consent for the questionnaire data was implicit on parents completing and returning the survey, although, again, patients have been de-identified. For baseline analysis, the charts of all patients treated for positional plagiocephaly were reviewed and, from them, information regarding gender, age at presentation, side of pathology, course of treatment, and baseline cognitive function (subjective parental concern and physician-defined developmental delay) was obtained. The severity of each patient's plagiocephaly was prospectively recorded in the chart according to Argenta's five-point plagiocephaly severity score, [6] previously validated to have moderate interobserver reliability. Following identification of the patients, a survey questionnaire was mailed to each patient's guardian to assess various cosmetic and cognitive outcome variables. Examples of these questions included cosmetic enquiry into residual asymmetry (Q3: "Do you think that there is any residual asymmetry at the back of your child's head?"), parental or child concerns about head shape, and teasing behavior at school (Q4: "In the last year, has your child commented to you about being teased at school because of asymmetric appearance of his head?") Cognitive enquiry included whether the child was at age-appropriate grade at school (Q7: "What grade is your child in at school?" Q8: "Is this the normal grade level for their age?"), utilization of special education resources, as well as gross and fine motor skill development and ease of communication for both expressive and receptive language (Q13e "Do you have concerns about your child's development in the area of language comprehension?").

Exclusion criteria included those patients with craniosynostosis and those for whom chart information was incomplete.

Patients were classified by both gender and side of pathology. Demographic differences between groups were analyzed by χ^2 -test for categorical variables and Mann-Whitney test for age at presentation. All cosmetic and cognitive outcome variables were categorical and tested by χ^2 -test. All analyses were performed at the $\alpha = 0.05$ level of significance.

Table 1. Demographic Information for Plagiocephaly Patients

Feature	Overall	Male	Female	Left	Right
N	80	51	29	22	58
Age in years (median, range)	6 (4 – 9)	6 (5 – 8)	7 (4 – 9)	7 (5 – 9)	6 (4 – 9)
Gender (male)	64%	N/A	N/A	59%	66%
Side (right)	73%	75%	69%	N/A	N/A
Torticollis	30%	33%	24%	27%	31%

Table 2. Comparison Between Respondent and Non-Respondent Populations

Feature	Overall	Respondents	Non-Respondents	p-value
N	346	80	266	
Gender (male)	70%	64%	72%	0.11
Side (right)	74%	73%	74%	0.97
Head Circumference (%ile)	61%	65%	60%	0.17
Positional Therapy (%)				
Recommended	100%	100%	100%	
Compliance	92%	95%	91%	0.42
Helmet Therapy (%)	30%	37%	27%	0.11
Developmental Delay	16%	18%	15%	0.25

Results

Review of the CHEO charts revealed 346 treated patients for inclusion in the study. Among these patients, there were 80 respondents (response rate 23%, at a median age of 9 years) with distribution of their baseline demographic characteristics detailed in Table 1. There were no differences in the distribution of the overall patient population and the survey respondents for gender, side of pathology, implemented type of therapy or baseline cognitive assessment (Table 2, $\alpha = 0.05$). The median age at presentation for this group was six months, with torticollis afflicting 30% of children at presentation. This group was predominantly male (64%) with a marked preponderance of right-sided plagiocephaly (73%). Developmental delay was noted in infancy among 18% of our respondent population; higher than reported baseline population levels.

Fifty-four percent of patients presented with only mild deformity and 8% were classified as severe. The overall distribution of case severity and decision for helmet orthosis molding is demonstrated in Figure 1.

Application of helmet orthosis as a therapeutic modality was performed in 36% of cases and expectedly bore a high as-

sociation with disease severity (χ^2 -test, $p < 0.001$). There was no statistically significant difference between the severity of plagiocephaly and gender or side of pathology.

Cosmetic outcomes in this patient group were acceptable, with fewer than 20% of parents expressing concern about the patient's head shape and only 4% classifying the shape as severe deformity, with no difference by gender or side of pathology. Fewer than 10% of patients were themselves aware of any asymmetry or reported being teased about their head shape.

Among the 80 patients surveyed, parent-reported developmental delay occurred frequently, distributed as 21% having language difficulties, 28% having motor difficulties, and 15% requiring special education. This exceeds the population averages for developmental delay ($p < 0.01$), which occurs in 5-6% of children. The side of pathology was related to these cognitive outcomes as shown in Figure 2, with left-sided disease strongly related to the need for special education classes (27% versus 10%, $p < 0.05$) and the observations of fine motor delay (41% versus 22%, $p < 0.05$) and speech delay (36% versus 16%, $p < 0.05$). There was no difference in language comprehension among patients by side of pathology.

Distribution of Plagiocephaly Severity and Helmet Utilization

FIGURE 1. Distribution of plagiocephaly severity among this patient population, with 54% qualifying as mild and 8% as severe by the Argenta classification system. Helmet orthosis was applied in 36% of cases, most frequently among higher grades of deformity severity (χ^2 test, $p < 0.001$).

Discussion

Our study showed males to be disproportionately affected by PP over females, and a preponderance of right-sided versus left-sided disease. These findings are consistent with similar retrospective reviews of plagiocephaly with regards to both short and medium-term outcomes.[5, 7, 8] This may be related to positional preference among infants, with population studies revealing 68% preference for right-sided position, particularly among males and among supine-sleeping patients.[9]

Cosmetic outcomes after helmeting in this series of patients were acceptable, with 85% of parents noticing improvement in head shape and only 4% having residual significant deformity. Similarly, only 10% of children raised any concern about their head shape. Followup work by Hutchinson and coworkers[5] among patients aged three to four years also reveals 61% normalization of head shape and only 4% residual severe deformity.

Cognitive outcomes have been less well characterized in the literature. Associations between the need for special education and diagnosis of attention deficit and hyperactivity disorder have been made for positional plagiocephaly.[10, 11] It remains unclear if this is associated with choice of prone or supine positioning, with earlier gross motor development and tone associated with infants playing and sleeping in prone position.[12] Among patients with PP followed from infancy to three to four years of age, Ages and Stages Questionnaire (ASQ) testing revealed improvement among early develop-

Educational Level at Followup for Plagiocephaly Patients

FIGURE 2. Educational level at followup among this patient population. The majority of patients were attending age-appropriate grades at school, although a significant fraction did require special education classes. This need for special education was more prevalent among patients with left-sided skull deformity (27% versus 10%, χ^2 test, $p < 0.05$).

mental delay that was primarily gross motor in quality.[5] Those patients with most severe initial deformity were least likely to demonstrate short-term cognitive improvement.

The remarkable findings from this analysis begin with identifying possible associations between PP and cognitive development persisting into school-aged populations, with a three- to four-fold increase in language and motor difficulties than the general population. While 94% of our patients attended age-appropriate grades at school, 15% of our respondents also indicated that special education was required. These results support other findings in PP research,[13] but novel work from this project further associates this developmental delay with left-sided versus right-sided plagiocephaly. Developmental delay in plagiocephaly is felt to be related to the modified skull form; Balan and colleagues were able to demonstrate altered auditory ERPs in infants with plagiocephaly as compared with unaffected infants, suggesting that brain function is already adversely affected by the plagiocephalic skull.[14] The restrictive effect of the skull deformity on cerebral development may be responsible for the perceived cognitive delay, as has been already strongly observed in single-suture craniosynostosis and to lesser extent in non-synostotic plagiocephaly.[15, 16]

Limitations

This study has a variety of limitations, owing first to the low response rate, potentially reflecting the lack of any questionnaire incentive to elicit response or also inaccurate mailing information when performed nearly a decade after some patients were last seen in clinic. With such a low response rate, one cannot be certain whether the respondents truly represent the entire cohort, or if this group is more afflicted with the outcome sought in the questionnaire. While such a shortcoming can be addressed by a prospective study format, this can be challenging in the context of outcomes sought a decade after intervention. In future studies, subjective cosmetic outcomes could be replaced by more objective facial symmetry metrics including differences in head diagonal diameters and differences in distance from anterior superior helical fold to the ipsilateral lateral canthus. The subjective cognitive outcomes reported by the parents could be replaced by more objective metrics including school performance and class rank and perhaps more formal neuropsychological testing into adolescence. Certainly, a more appropriate comparison would be against a local peer control group for incidences of various forms of developmental delay. This study was also underpowered to define outcome differences between patients treated by helmet orthosis as a sole strategy or in combination with other more aggressive modalities.

Conclusions

This study suggests that cosmetic outcomes following treatment of positional plagiocephaly at CHEO are acceptable, with low rates of both patients and family members perceiving any residual deformity. While the vast majority of patients are currently at age-appropriate grades, parental concerns regarding cognitive development are common and more prevalent for those patients with left-sided pathology. Prominent concerns include delayed fine motor and language development, alongside a three-fold higher prevalence of special education courses. Such findings are important in identifying the need to more rigorously characterize these outcomes as well as to educate parents and primary care providers. Further, they provide motivation for invoking the aid of primary care providers to initiate early preventative management to correct positional preference among infants. While limitations include the low response rate and the crude outcome measures, the results do provide insight into what residual parental and child concerns exist following the conservative management of positional plagiocephaly.

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