Nasal Root Deviation in Unicoronal Craniosynostosis: A Craniometric Analysis of Early and Late Postoperative Outcomes

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Purpose: Current operative techniques for correcting unicoronal craniosynostosis (UCS) leave the nasal bones untouched, resulting in an unclear long-term impact on nasal root deviation. The purpose of this study is to quantify nasal root deviation in the preoperative and late postoperative setting in patients who have undergone conventional single-staged UCS correction.

Methods: The authors performed a retrospective, craniometric analysis of nasal root deviation comparing preoperative computed tomography scans, with those of the early, and late postoperative period. Three vectors were analyzed to measure nasal root deviation, one extending from the nasion to the rhinion (nasal bone vector), the second from the rhinion to the anterior nasal spine (nasal aperture vector), and the third from the nasion to the anterior nasal spine (nasal longitudinal vector).

Results: Twenty-five subjects were included in the study. Average ages at the time of preoperative, early, and late postoperative imaging were 0.6 ± 0.3, 0.9 ± 0.6, and 9.3 ± 2.7 years, respectively. Improvement of angular deviation of both the nasal aperture vector and nasal longitudinal vector was observed. Mean angular deviation of the nasal aperture vector was 6.0 ± 1.9 degrees preoperatively, 6.0 ± 2.1 degrees early postoperatively ($P = 0.952$), and 2.4 ± 2.1 in the late postoperative period ($P = 0.013$). Mean angular deviation of the nasal longitudinal vector was $5.7{+}2.0$ degrees preoperatively, 5.8 ± 2.3 degrees early postoperatively ($P = 0.948$), and 3.7 ± 1.6 degrees in the late postoperative period ($P = 0.019$).

Conclusion: Nasal root deviation decreased significantly only in the late postoperative period, lending credence to the notion that though UCS correction does not directly address nasal root deviation, this pathology improves significantly over time.

Key Words: Craniosynostosis, nasal root, synostosis, unicoronal

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There exists a pathognomonic set of features associated with unicoronal craniosynostosis (UCS), namely, ipsilateral supraorbital retrusion, elevation of the supraorbital rim, flattening of the frontal bone, and contralateral compensatory frontal bossing.[1–6] Studies have also shown deviation of the nasal root to be present in this population to a variable degree.[7,8] Traditional, 1-stage operations for correcting UCS usually incorporate fronto-orbital advancement and remodeling of the anterior cranial vault to the level of the nasofrontal suture medially and do not directly manipulate the nasal bones. The long-held belief and justification for the lack of a direct intervention on the deviated nasal bones is that by releasing the synostosis one allows for natural correction of the aforementioned deviation.[8–11]

Multiple studies have examined nasal root deviation in UCS patients postoperatively, but these studies were subjective in nature, relying on patient photographs and a subjective grading of nasal root deviation.[7,12–16] The most likely explanation for this paucity of quantitative evidence is the complex requirements inherent to a study that would rigorously test this hypothesis–preoperative and postoperative computed tomography (CT) scans, both early and late, and software that allows for thorough craniometric analyses. The purpose of this study was to objectively quantify nasal root deviation in patients with UCS in the preoperative, early postoperative, and late postoperative periods to measure whether this abnormality corrects over time following primary reconstruction.

METHODS

A retrospective review of all patients who underwent surgical correction for UCS at the Children's Hospital of Philadelphia between 2000 and 2016 was conducted. Data collected included demographic information, operative notes, full operative course, and CT imaging data. Complete preoperative and postoperative head CTs were required for study inclusion. Any surgical intervention on the nasal bones was cause for exclusion from the study.

Operative Technique

This study includes the patients of 2 plastic surgeons (JAT and SPB). While the authors' operative technique has changed slightly over the study period, there has been consistency regarding frontonasal bone osteotomy and repositioning.[1] Additional bone grafts are placed at the lateral aspect of the tenon, in the lateral aspect of the orbit for stability, and in the contralateral orbital portion of the bandeau to achieve orbital symmetry. No surgical alteration of the nasal bones is made. Release of the bandeau is made with bilateral cuts at the level of the zygomatic-frontal suture or slightly below, and at the level of the nasal-frontal suture. The bandeau is split in the midline and bone graft is utilized to increase its width; this construct is then positioned and attached in the midline (Fig. 1).

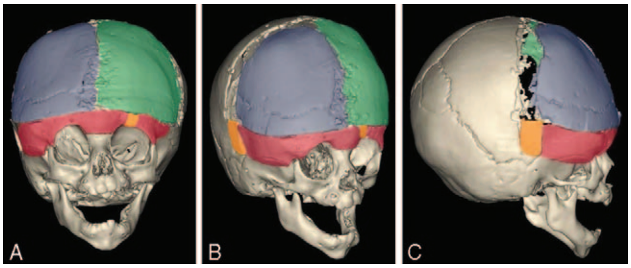

FIGURE 1. Postoperative computed tomography scan showing the authors' operative approach, specifically the placement of an interpositional bone graft in the contralateral orbit. (A) Anteroposterior view. (B) Oblique view. (C) Lateral view.

Craniometric Analysis

A craniometric evaluation of preoperative and postoperative CT scans was performed using advanced medical imaging segmentation and analysis applications (Materialise Mimics & Materialise 3-matic). Postoperative CT scans were categorized as early (less than 5 years postoperatively), or late postoperative scans (greater than or equal to 5 years postoperatively). For each scan, a mid-sagittal plane was defined using a point in the anterior cranial base (the midpoint of the tuberculum sellae), the anterior nasal spine (ANS), and the pogonion (Fig. 2). Three vectors were then created to measure nasal root deviation, one extending from the nasion to the rhinion (nasal bone vector), the second from the rhinion to the ANS (nasal aperture vector), and the third from the nasion to the ANS (nasal longitudinal vector) (Table 1). The angles at which these vectors intersect the mid-sagittal plane were the primary outcome of interest (Fig. 2).[17]

Statistical Analysis

The preoperative angular deviations of each of the 3 vectors for a given subject were compared with that same subject's early and late postoperative deviations of the corresponding vectors (Fig. 3). A paired *t* test was used to assess for a statistically significant change in angulation of each of the 3 vectors for each unique subject. All tests are defined as 2-tailed, and statistical significance was defined as $P < 0.05$ (Fig. 3).

RESULTS

One hundred ten patients underwent UCS correction during the study period, 25 of which met inclusion criteria, with pre- and postoperative CT. The majority of subjects were female (N = 20, 80%), with right-sided unicoronal synostosis (N = 15, 60%). Preoperative CT scan took place at a mean age of 0.8 ± 0.3 years. Eighteen subjects (72%) had CT scans that were categorized as "early postoperative" scans, which took place at an age of 0.9 ± 0.5 years. Eight subjects (32%) had late postoperative CT scans, at the age of 9.3 ± 2.7 years (Table 2).

Cephalometric results were analyzed in a pairwise fashion, comparing a subject's preoperative CT to the same subject's early and/or late postoperative CT scans. The nasal bone angle, defined as the angle at which the vector from nasion to rhinion intersects the mid-sagittal plane, was 4.7 ± 2.8 degrees preoperatively. This angle did not differ significantly from that of the early postoperative period (5.1 ± 3.2 degrees, $P = 0.787$) or the late postoperative period (5.1 ± 1.9 degrees, $P = 0.670$). The nasal aperture angle, defined as the angulation of the vector extending from rhinion to anterior nasal spine, was 6.0 ± 1.9 degrees preoperatively. The change in nasal aperture angle in the early postoperative period was not statistically significant (6.0 ± 2.1 degrees, $P = 0.952$). In the late postoperative period, however, a statistically significant decrease in the nasal aperture angle was seen (2.4 ± 2.1 degrees, $P = 0.013$). The nasal longitudinal angle, defined as the angulation of the vector extending from nasion to anterior nasal spine, was 5.7 ± 2.0 degrees preoperatively. This value did not differ significantly from that of the early postoperative period (5.8 ± 2.3 degrees, $P = 0.948$), but a statistically significant decrease in the nasal longitudinal angle was seen in the late postoperative period (3.7 ± 1.6, $P = 0.019$) (Table 3).

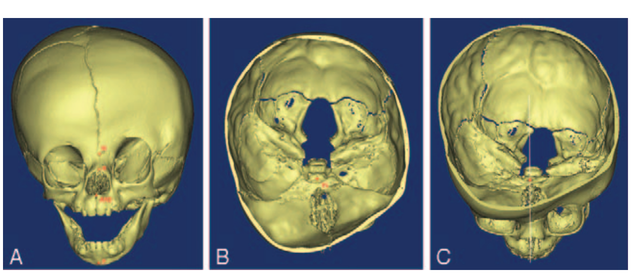

FIGURE 2. Bony landmarks for computed tomography-based cephalometric analysis. (A) Anteroposterior view. (B) Skull base.

TABLE 1. Anthropometric Landmark Definitions

Landmark (Abbreviation)	Definition
Anterior nasal spine (ANS)	Midpoint of inferior nasal aperture boarder
Pogonion (P)	The most anterior prominent point of the mandible
Tuberculum sellae (TS)	Midpoint of lateral boundaries of tuberculum sellae
Mid-sagittal plane (MSP)	The plane that intersects ANS, P, and TS
Nasion (N)	Point of intersection between nasal bones and frontal bone
Rhinion (R)	Inferior most point of internasal suture
Nasal bone vector (NBV)	Line that intersects nasion and rhinion
Nasal aperture vector (NAV)	Line that intersects rhinion and anterior nasal spine
Nasal longitudinal vector (NLV)	Line that intersects nasion and anterior nasal spine
Outcome metrics	
Nasal bone angle (NBA)	Angle of intersection between NBV and MSP
Nasal aperture angle (NAA)	Angle of intersection between NAV and MSP
Nasal longitudinal angle (NLA)	Angle of intersection between NLV and MSP

DISCUSSION

The clinical presentation of unicoronal synostosis is characterized by supraorbital retrusion, elevation of the supraorbital rim, and

TABLE 2. Summary of Patient Demographics and Imaging History

	N	%
Total subjects	25	
Gender		
Female	20	80%
Male	5	20%
Synostosed side		
Right	15	60%
Left	10	40%
Age at intervention (y; mean ± std. dev)	0.8	0.3
Age at preoperative CT scan (y; mean ± std. dev)	0.6	0.3
Number of subjects with an early postoperative CT scan	18	72%
Age at early postoperative CT scan (y; mean ± std. dev)	0.9	0.5
Number of subjects with a late postoperative CT scan	8	32%
Age at late postoperative CT scan (y; mean ± std. dev)	9.3	2.7

CT, computed tomography; Std. dev, standard deviation.

TABLE 3. Anthropometric Measurement Results

Measurement Landmark	Preoperative		Early Post-operative		P	Late Post-operative		P
Nasal bone angle (NBA)	4.7	2.8	5.1	3.2	0.787	5.1	1.9	0.670
Nasal aperture angle (NAA)	6.0	1.9	6.0	2.1	0.952	2.4	2.1	0.013
Nasal longitudinal angle (NLA)	5.7	2.0	5.8	2.3	0.948	3.7	1.6	0.019

flattening of the frontal bone on the ipsilateral side, with compensatory frontal bossing on the contralateral side.[1] Also commonly present following premature fusion of the unicoronal suture is deviation of the nasal root. An appreciation of these phenotypic features is critical when considering the degree of a patient's deformity, the optimal surgical approach, and the quality of the surgical correction.

In most types of UCS correction, both modern and historical, the surgeon directly addresses each of the aforementioned phenotypic characteristics with the exception of nasal root deviation. The logic underpinning this fact dates back to the beginnings of the modern era in craniofacial surgery as techniques published by Whitaker, McCarthy, and Marchac all adopted a nasofrontal osteotomy both for ease and to separate cranium from nose.[8–10] Their surgical approaches, as those promoted by their disciples, intentionally left the nasal root untouched, with the hypothesis that the nasal root deviation has the capacity to self-correct following release of the synostotic suture.[8–11]

The purpose of our study was to objectively test this hypothesis using pre- and postoperative CT scans of UCS patients. Both the nasal aperture and nasal longitudinal vectors improve significantly over time, while the nasal bone vector remains unchanged. When viewed in the context of the rest of the face, our results may be caused by a shift of the nasion and rhinion toward the midline over time or simply as a result of mid-face elongation. Angulation of the nasal bones, however, did not change significantly over the study period, an interesting fact given the paucity of research pertaining to rhinoplasty in UCS patients in the 10 years. One potential explanation for this negative finding is that both the nasion and rhinion shift toward the midline at similar rates, thereby preserving the relative angulation of the landmarks. Alternatively, this could be due to a relatively small amount of growth that occurs between the nasion and rhinion, when compared with the growth that occurs between each of these landmarks and the ANS (as captured by the nasal aperture and the nasal longitudinal vectors). Lastly, this negative finding could simply represent type II error, that is, a false negative due to our study's small sample size.

A number of previous studies have sought to address the question of autocorrection of nasal root deviation, but the vast majority rely on subjective clinical assessment and direct anthropometry of nasal root deviation.[18] A study by Bartlett et al[12] compared outcomes resulting from 2 operative approaches for the correction of UCS. Neither approach addressed nasal root deviation directly, but subjective postoperative assessment by the authors concluded that both approaches lead to full correction of the nasal root nonetheless.[12] Similar studies that followed however had less positive conclusions relating to the nasal root. Machado and Hoffman[19] concluded that 39% of their UCS cohort had persisting deviation of the nasal root postoperatively; again, this study depended on subjective assessment by the surgeon. Similarly, a study by McCarthy et al[7] analyzed a 20-year experience treating patients with UCS and found this figure to be 51%. Prévot et al[13] categorized the severity of nasal root deviation in a similar cohort

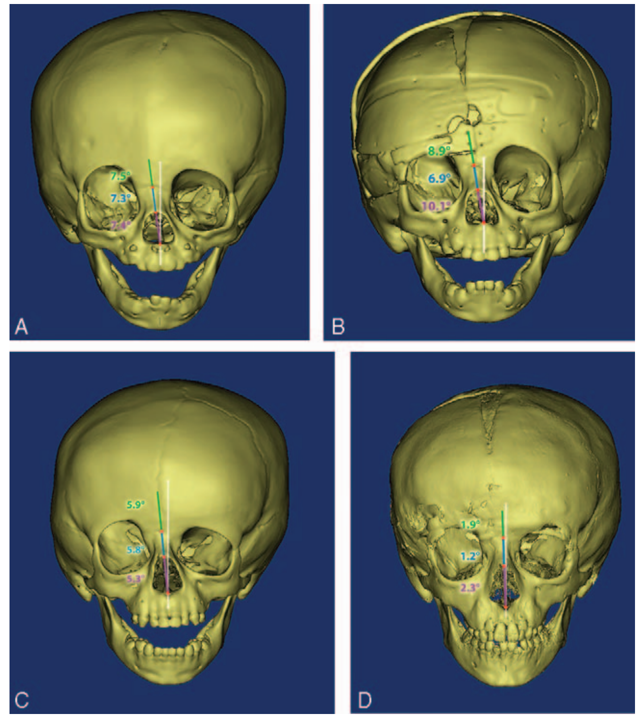

FIGURE 3. Cephalometric analyses of 2 subjects, with the nasal bone vector (blue), nasal aperture vector (purple), nasal longitudinal vector (green), and associated angles indicated. (A) Subject 1, preoperative. (B) Subject 1, early postoperative. (C) Subject 2, preoperative. (D) Subject 2, late postoperative.

both pre- and postoperatively using photographs, and found 38% to have lasting abnormality. The authors then performed a subgroup analysis, grouping patients by their age at surgery, and found that those operated on between 1 and 2 years of age had the greatest degree of nasal root correction. Conversely, the oldest subgroup, those patients operated on after the age of 4, showed the smallest degree of correction. Limitations shared by all of these studies include the subjective nature of root deviation measurement, that only soft tissue was evaluated not the underlying bony framework, and for all but the Prevot study, no correlation was made between preoperative and postoperative deviations.

Advances in CT scan resolution and image analysis software have yielded additional, more quantitative studies examining nasal root deviation in UCS. Camargos et al[17] used patient CT scans to measure nasal root deviation, as well as that of the overlying soft tissue in preoperative UCS patients. Their analysis found a 6.6 ± 2.9 degree deviation of the nasal bones toward the side of synostosis, as measured by the intersection of a vector between nasion and rhinion with a mid-sagittal plane. One potential limitation of this study is their construction of the mid-sagittal plane, which they defined with the nasion, C1, and C2. To include the nasion in such a definition implies that this point falls along the true mid-sagittal plane in patients with UCS, which is a tenuous assumption when there is already known deviation of the nasal root.

In a related study, Marianetti et al[15] used CT scans to quantify nasal root deviation in skeletally mature UCS patients. This analysis found significantly larger degrees of root deviation than the previous study, with a mean nasal bone deviation of 17.9 ± 6.5 degrees. The large disparity between these 2 studies may be due to their vastly different definitions of the mid-sagittal plane. Unlike the Camargos study, Marianetti created a mid-sagittal plane using landmarks in the posterior skull base. Though this definition is strengthened by the fact that it does not use any nasal landmarks in defining midline, it is nonetheless problematic. In using posterior cranial base landmarks to assess facial structure, one assumes that the vectors of the posterior and anterior cranial bases are parallel to, and coincident with, one another. This assumption, however, has been disproven, with potentially persistent deviations between anterior and posterior skull base trajectory reaching upward of 13 degrees in UCS patients.[20,21] To address this complexity in our study, we used landmarks from the anterior skull base and facial skeleton that were unaffected by the pathology to define the mid-sagittal plane. Namely, the midpoint of the tuberculum sellae, the ANS, and the pogonion were used.

In a UCS cohort however, while consideration of the aforementioned principles is beneficial, defining a mid-sagittal plane remains problematic. Our analysis used the pogonion in defining the mid-sagittal plane, as the authors felt it was the least-bad option. Though deviation of the pogonion is often described as a hallmark of UCS, a review of the literature reveals remarkably few studies addressing this issue directly. Bruneteau and Mulliken[22] published one of the earliest papers to examine chin-point deviation, wherein the surgeon simply noted the presence or absence of multiple phenotypic features in UCS patients based on clinical examination or photographs. No attempt was made to quantitatively assess given characteristics or to have multiple raters. According to their results, the authors found that 96% of the UCS cohort had chin-point deviation, but the examination of both the paper's figures showing UCS patients from this cohort fails to mention or show this same phenotype.[22] The other study to assess chin-point deviation in UCS patients was performed by Kane et al,[23] and took a much more quantitative approach to assessing the mandibular phenotype in this cohort. This CT-based analysis showed that their cohort of 20 UCS patients did, in fact, have slight lateral shift of the chin point, as evidenced by the difference in distance from the pogonion to both the condylion and the tip of the coronoid when comparing the affected and unaffected sides of the mandible. Importantly though, these authors' analysis showed a chin-point shift of only 1.75% of the length of the hemi-mandible.[23] In our study, the mid-saggital plane was noted to bisect the maxillary central incisors in all patients, lending further credence to our methodology.

An additional limitation of our study is the inherent selection bias in our late postoperative cohort, as the presence of a CT scan at this late date implies that there may be some abnormality. This bias however would lessen the measured improvement in nasal root deviation if anything, not amplify it. Additionally, we do not control for variations in operative technique over the 16-year study period, although no fundamental changes in operative approach occurred during this time.

Our results showed a decrease in nasal bone angulation following surgical correction of UCS, this finding may be the result of mid-face lengthening or the migration of the nasion and rhinion toward the midline takes place. Of note, this correction was only found in the late postoperative period. Despite significant improvement at this late time point however, our analysis indicates that there remains some persistent angulation of the nasal bones. It is the authors' subjective experience, however, that this patient cohort does not go on to require rhinoplasty, as the degree of angular correction is adequate. However, this may in part represent a practice pattern within the authors' institution, wherein UCS patients are rarely considered for a nasal-angulation correcting procedure as their nasal phenotype continues to improve subjectively. Correlating these results to soft tissue deviations as well as quantitative clinical impressions of deviation remain important.

CONCLUSIONS

We report preoperative, early postoperative, and late postoperative nasal root deviation in a cohort of patients with UCS. Nasal root deviation as measured by nasion and rhinion shift decreased significantly only in the late postoperative period, lending credence to the notion that though UCS correction does not directly address nasal root deviation, this pathology improves significantly over time.

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