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Frontofacial Features of Unilateral Lambdoid Craniosynostosis: A Multicenter Assessment

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Background: Unilateral lambdoid craniosynostosis is differentiated from deformational plagiocephaly primarily by assessing the cranium from posterior and bird's-eye views. Findings include posterior displacement of the ipsilateral ear, ipsilateral occipitomastoid bossing, ipsilateral occipitoparietal flattening, contralateral parietal bossing, and contralateral frontal bossing. Diagnosis based off facial morphology may be an easier approach because the face is less obstructed by hair and head-coverings, and can easily be assessed when supine. However, frontofacial characteristics of unilateral lambdoid craniosynostosis are not well described.

Methods: A retrospective cohort review of patients with isolated, unilateral lambdoid craniosynostosis from the Children's Hospital of Pittsburgh and the Children's Hospital of Philadelphia was performed. Preoperative frontal and profile photographs were reviewed for salient characteristics.

Results: Nineteen patients met inclusion criteria. Eleven patients had left lambdoid craniosynostosis, and eight had right lambdoid craniosynostosis. All patients were nonsyndromic. Patients demonstrated contralateral parietal bossing and greater visibility of the ipsilateral ear. Contralateral frontal bossing was mild. The orbits were tall and turricephaly was present in varying severity. Facial scoliosis as a C-shaped deformity was present in varying severity. The nasal root and chin pointed to the contralateral side.

Conclusions: The combination of greater visibility of the ipsilateral ear, contralateral parietal bossing, and C-shaped convex ipsilateral facial scoliosis are hallmark frontofacial features of unilateral lambdoid craniosynostosis. Although the ipsilateral ear is more posterior, the greater visibility may be attributed to lateral displacement from the mastoid bulge. Evaluation of long-term postoperative results is needed to assess if this pathognomonic facial morphology is corrected following posterior vault reconstruction. *(Plast Reconstr Surg Glob Open 2023; 11:e5011; doi: 10.1097/GOX.0000000000005011; Published online 19 May 2023.)*

INTRODUCTION

Lambdoid craniosynostosis is the rarest of all synostotic malformations, of which isolated, unilateral lambdoid synostosis is a subset. The classic morphology consists of (1) ipsilateral occipital-mastoid bossing (mastoid bulge), (2) ipsilateral tilt of the posterior skull base, (3) contralateral frontal and parietal bossing, (4) ipsilateral occipitoparietal flattening, and (5) posterior-inferior positioning of the ipsilateral ear.[1,2] This constellation of findings is summarized as a trapezoidal head shape from the bird's-eye view.[3]

Although historically low, there has been a wide range in the proposed incidence rates in lambdoid craniosynostosis, ranging from 2.3% to 21% of all craniosynostosis.[4,5] The discrepancy may be related to confusion between deformational (positional) plagiocephaly and unilateral lambdoid craniosynostosis, especially with an increased incidence of deformational changes following the initiation of the "Back to Sleep" campaign by the American Academy of Pediatrics task force on infant positioning and sudden infant death syndrome.[6,7] Deformational plagiocephaly, in contrast to lambdoid craniosynostosis, is caused by external deforming forces from prolonged supine positioning. It is often

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described as a constellation of findings including (1) ipsilateral frontal bossing, (2) anterior displacement of the ipsilateral ear, (3) horizontal cranial base, and (4) ipsilateral occipitoparietal flattening. The head shape is a parallelogram from the bird's-eye view.[1] These changes improve over time and can be managed nonoperatively using orthotic helmets without the concern for neurocognitive or aesthetic sequela as seen in craniosynostosis.[6]

Although the comparative morphology between deformational plagiocephaly and unilateral lambdoid craniosynostosis has been described, there continues to be reports of misdiagnosis and confusion over "posterior plagiocephaly."[8–12]

The existing literature focuses on malformations of the skull, cranial base, and facial skeletal differences present in patients with both lambdoid synostosis and deformational plagiocephaly.[13–15] In a study by Ploplys et al, significant posterior cranial base and facial skeletal asymmetries were identified in cases of lambdoid synostosis that were not present in deformational plagiocephaly, namely deviation of the ipsilateral posterior fossa, enlargement of the contralateral middle cranial fossa, and ipsilateral acute petrous angle.[13] In terms of facial morphology, this study suggested the external auditory meatus (EAM) and articular fossa are displaced anteriorly on the affected side in cases of lambdoid synostosis and deformational plagiocephaly, although severity of this displacement was significantly greater in those with craniosynostosis. According to these authors, lambdoid synostosis patients demonstrated more shallow midfacial depth of the affected side than patients with deformational plagiocephaly. These findings are similar to those described in our previous studies.[14] Recent investigations using computed tomography (CT) imaging on a larger series of patients describe anterior, inferior displacement of the ipsilateral ear in lambdoid synostosis that was not present in deformational plagiocephaly.[15] Allam et al also confirmed previous findings of posterior fossa deviation and mastoid bulging present in synostosis.

It is suggested that deformity of the middle and posterior fossa of patients with lambdoid synostosis drives unilateral lengthening of the contralateral midface and displacement of the EAM and temporomandibular joint.[14] Thus, distinct frontofacial features can be expected for patients with lambdoid synostosis. Although the classic comparative morphology has been primarily based on the bird's-eye and posterior views, these may be difficult to accurately assess with certain hairstyles. Certainly, determining additional characteristic morphology from the frontal view in particular, as is frequently used to diagnose unilateral coronal synostosis, would be helpful in further clarifying the diagnostic differences between deformational plagiocephaly and unilateral lambdoid craniosynostosis. This article focuses on characterizing the frontofacial morphology of unilateral lambdoid craniosynostosis through a multi-institutional collaboration.

PATIENTS AND METHODS

A retrospective chart review was performed at the Children's Hospital of Pittsburgh and the Children's Hospital of Philadelphia of patients treated in the craniofacial center from 2005 to 2017 and 1990 to 2017, respectively. Patients with a unilateral lambdoid craniosynostosis diagnosis, confirmed by CT, were included in the study. Patients with multisuture synostosis or deficient frontal-view photography were excluded.

Preoperative, frontal, left lateral, and right lateral view photographs were primarily utilized. If available, posterior, left lateral, and right lateral view photographs were also utilized. Six craniofacial surgeons collectively reviewed the photographs (J.Y.L., S.N., J.A.T., J.A.G., S.P.B., J.E.L.), and were blinded to the laterality of synostosis. Four anatomic regions were descriptively evaluated: (1) cranial shape, (2) comparative ear position, (3) facial meridian, and (4) orbital shape and symmetry.

Takeaways

Question: Can frontofacial features assist in diagnosing unilateral lambdoid craniosynostosis (CS)?

Findings: A retrospective analysis of photographs of 19 patients with unilateral lambdoid CS revealed these patients have notable contralateral parietal bossing and greater visibility of the ipsilateral ear while contralateral frontal bossing is mild. The orbits are tall and turricephaly may be present. Facial scoliosis as a C-shaped deformity is often present.

Meaning: The combination of greater visibility of the ipsilateral ear, contralateral parietal bossing, and C-shaped convex ipsilateral facial scoliosis are hallmark frontofacial features of unilateral lambdoid CS.

RESULTS

A total of 19 patients across both institutions were included in the study. Eleven patients were diagnosed with left-sided lambdoid craniosynostosis and eight patients with right-sided lambdoid craniosynostosis. All patients were nonsyndromic. All patients had frontal view photographs taken at the time of presentation. The age at which preoperative images were obtained ranged from 2.1 months to 12 years of age.

All patients demonstrated obvious cranial shape anomalies on frontal-view. Contralateral parietal bossing was present in all patients. Minimal to no frontal bossing was observed (n = 2/19). Turricephaly was observed in varying severity for half of the patients (n = 8/19). Ear position was also noticeably asymmetric for nearly all patients (n = 17/19). More specifically, the ipsilateral ear was more easily visible (prominent) than the contralateral ear; however, vertical positioning was not observed to be consistently asymmetric. Nearly all patients (n = 17/19) demonstrated a C-shaped deviation of the face, with the root of the nose and chin shifted to the contralateral side. The facial midline was curvilinear, with convexity towards the ipsilateral affected side. Orbital shape also demonstrated anomalies in half of patients (n = 10/19). The superior orbital rim appeared peaked

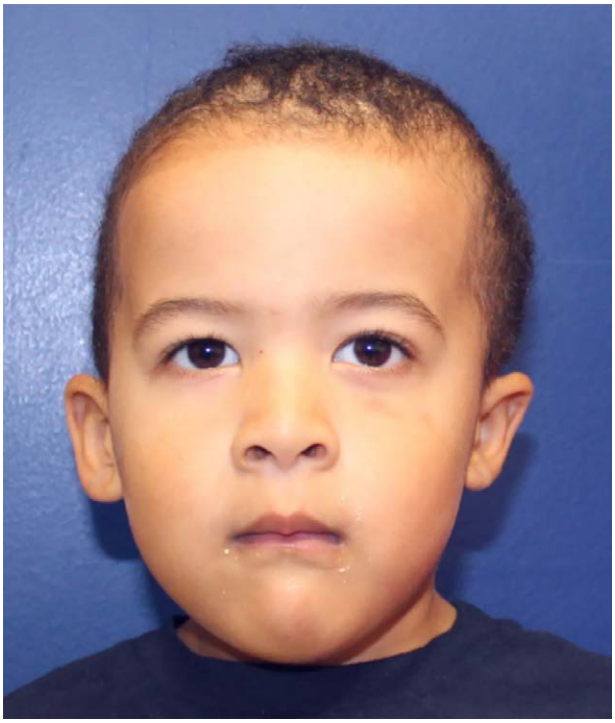

Fig. 1. Preoperative frontal view photograph of a young patient with right unilateral lambdoid craniosynostosis demonstrating most salient features: greater visibility of the ipsilateral ear, contralateral parietal bossing, C-shaped convex ipsilateral facial scoliosis.

in the central-medial portion, giving the appearance of a tall orbit. Orbital asymmetry, however, was not readily observed (n = 4/19). Thus, the most consistent and noticeable anomalies on frontal-view were (1) contralateral parietal bossing, (2) greater visibility of the ipsilateral ear, (3) and C-shaped facial scoliosis with convexity of the facial midline to the ipsilateral side (Fig. 1).

DISCUSSION

The classic morphology of unilateral lambdoid craniosynostosis is characterized by an ipsilateral occipitoparietal flatness, ipsilateral mastoid bulge, posterior cranial base tilt, ipsilateral posterior/inferior position of the ear, and overall trapezoid shape of the head. These features are in contrast to deformational plagiocephaly, allowing the evaluator to achieve a proper diagnosis, oftentimes without the need for radiographic studies. Nonetheless, confusion and misdiagnosis are still reported. The posterior and bird's-eye view, which predominates current assessment, can be limited, and additional defining morphology in the frontal view may help aid the proper diagnosis of unilateral lambdoid craniosynostosis.

The results of this study demonstrate that the constellation of contralateral parietal bossing, ipsilateral ear prominence, and C-shaped facial scoliosis with ipsilateral convexity are highly salient for unilateral lambdoid craniosynostosis. Although largely a posterior cranial anomaly, the sequelae of premature unilateral lambdoid suture obliteration clearly have a noticeable impact on frontofacial features (Fig. 2).

The contralateral parietal bossing is a consequence of Virchow's law, defined by compensatory growth parallel to the synostotic lambdoid suture.[16] Since the contralateral parietal cranium is in direct alignment with the lambdoid suture, the compensatory cranial growth as bossing is highly noticeable and salient (Fig. 3A). Although contralateral frontal bossing is also classically described, that morphology was not readily observed in our patient cohort in the frontal view. This may be secondary to the frontal bone being further away from the line of compensatory growth, reducing the severity of deformity. Additionally, the bird's-eye view would allow better detection of forehead contour as opposed to the frontal view, which would depend upon shadowing and light source position on the face below to detect asymmetries.

The ipsilateral mastoid bulge is also a consequence of compensatory growth, and likely contributes to the ipsilateral ear prominence on frontal view. The ipsilateral ear is typically in an inferior position compared to the contralateral ear; however, anterior/posterior positioning is debated.[14,15,17] This study observed a more posterior positioning of the ear. This positioning would then place the ear closer to the mastoid bulge, resulting in a lateral displacement of the ear (Fig. 3B). Therefore, even though the ipsilateral ear is posterior, it projects further from the cranium laterally and becomes more visible on the frontal view.

The facial scoliosis is likely secondary to asymmetries of the cranial base.[14] Although the posterior cranial base is tilted inferiorly on the ipsilateral side, the cranial base is also shifted in two other axes. The foramen magnum is shifted to the ipsilateral side, and the contralateral TMJ is displaced posteriorly (Fig. 3C). These changes result in deviation of the root of the nose and the chin to the contralateral side, resulting in the C-shaped scoliosis of the face with ipsilateral convexity.

Smartt et al described the facial scoliosis in a smaller cohort of unilateral lambdoid craniosynostosis patients, and demonstrated that the C-shaped facial meridian persisted after surgical correction with an occipital switch cranioplasty.[14] Although cranial morphology was reported to steadily improve after correction, no significant differences were observed in regards to the position of the EAM. Furthermore, the mastoid bulge improved with surgery, but did not completely normalize, postulating that asymmetries in ear prominence may also persist after posterior vault reconstruction. Further analysis of frontofacial morphology post-correction and with long-term follow-up is warranted.

Orbital shape and asymmetry was not a salient characteristic in the study cohort. This finding within itself is significant in comparison to the frontofacial morphology of unicoronal craniosynostosis (UCS). UCS is also on the differential for a trapezoid head and often presents

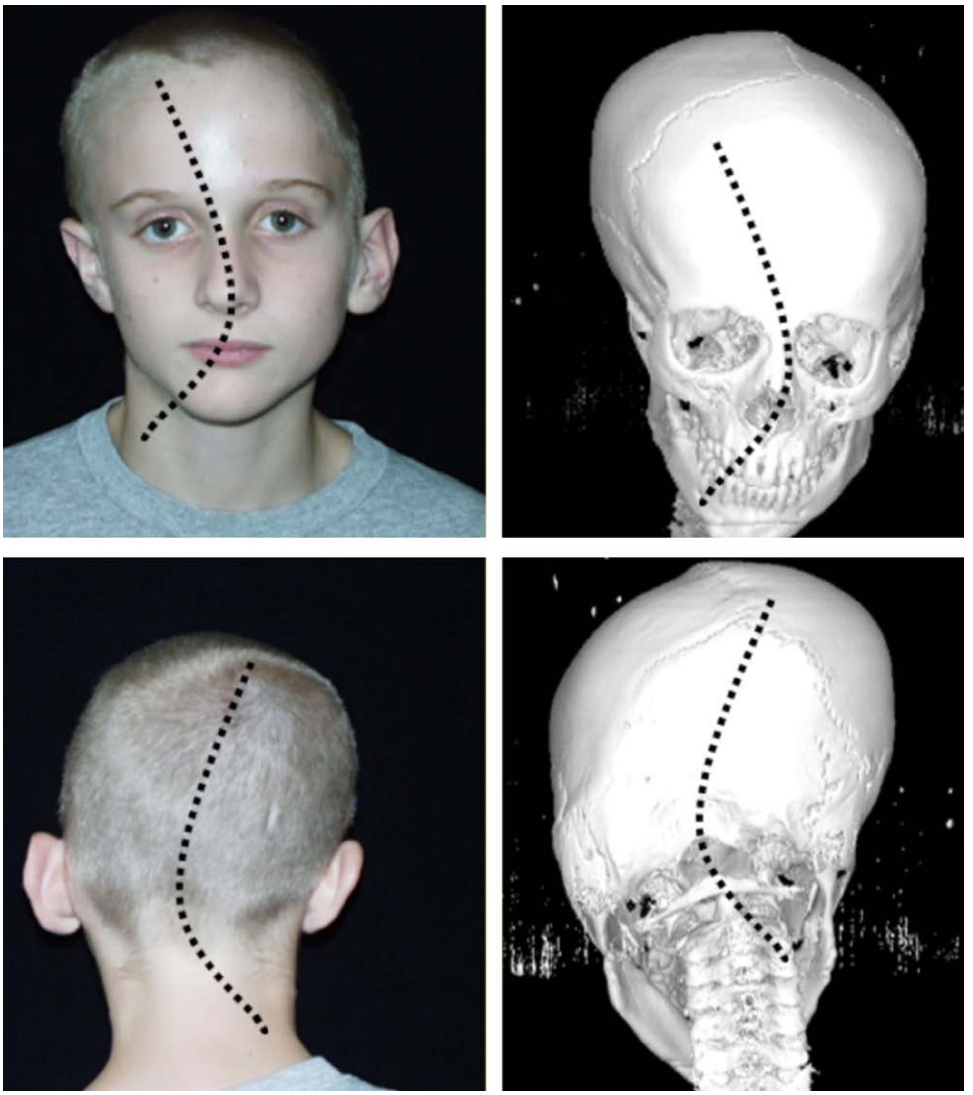

Fig. 2. Clinical photographs and the corresponding 3D head CT images demonstrating the appearance of C-shaped facial scoliosis (dotted line) seen in a 12-year-old patient. Contralateral parietal bossing, ipsilateral ear prominence, and ipsilateral occipitoparietal flattening are particularly notable. Adapted with permission from Smartt JM, Russell RR, Singh DJ, et al. True lambdoid craniosynostosis: long-term results of surgical and conservative therapy. Plast Reconstr Surg. 2007;120: 993-1003. Per the licensing agreement, the Creative Commons license does not apply. Use of this material in any format is prohibited without written permission from the publisher, Wolters Kluwer Health, Inc. Please contact permissions@lww.com for further information.

with facial scoliosis and parietal bossing on frontal view. However, one of the hallmark signs of UCS in frontal view is the harlequin orbital deformity. This orbital asymmetry is striking, characterized by a wider opening of the palpebral fissure secondary to peaking of the central-lateral superior orbital rim. Additional frontal-view comparative analysis among UCS, unilateral lambdoid synostosis, and other unilateral craniosynostosis is needed.

CONCLUSIONS

The combination of greater visibility of the ipsilateral ear, contralateral parietal bossing, and C-shaped convex ipsilateral facial scoliosis are hallmark frontofacial features of unilateral lambdoid craniosynostosis. When combined with the classically described features from the bird's-eye view and posterior view, better differentiation can be achieved when clinically diagnosing unilateral lambdoid craniosynostosis from deformational plagiocephaly.

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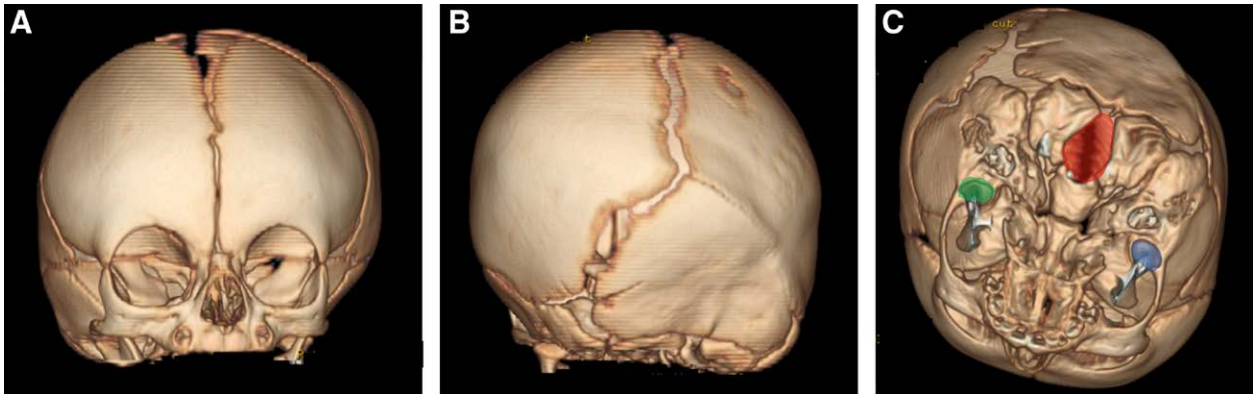

Fig. 3. A, Frontal view 3D head CT of right unilateral craniosynostosis. Contralateral parietal bone bossing is visible. B, Posterior view 3D head CT of right unilateral craniosynostosis. Greater ipsilateral ear visibility can be explained by ipsilateral mastoid bulge pushing ear laterally. C, Cranial base view 3D head CT of right unilateral craniosynostosis. Foramen magnum (highlighted in red) is shifted toward the affected side, and contralateral glenoid fossa is shifted posteriorly (highlighted in green) compared to the ipsilateral fossa (blue), resulting in C-shaped convex ipsilateral facial scoliosis.

DISCLOSURES

Joseph E. Losee receives royalties from the publication of Comprehensive Cleft Care, for which he is an editor. The other authors have no financial interest to declare in relation to the content of this article.

PATIENT CONSENT

The patient's parents provided written consent for the use of his images.

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