

Long-Term Outcomes of Nonsyndromic Isolated Unilateral Lambdoid Craniosynostosis

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BACKGROUND

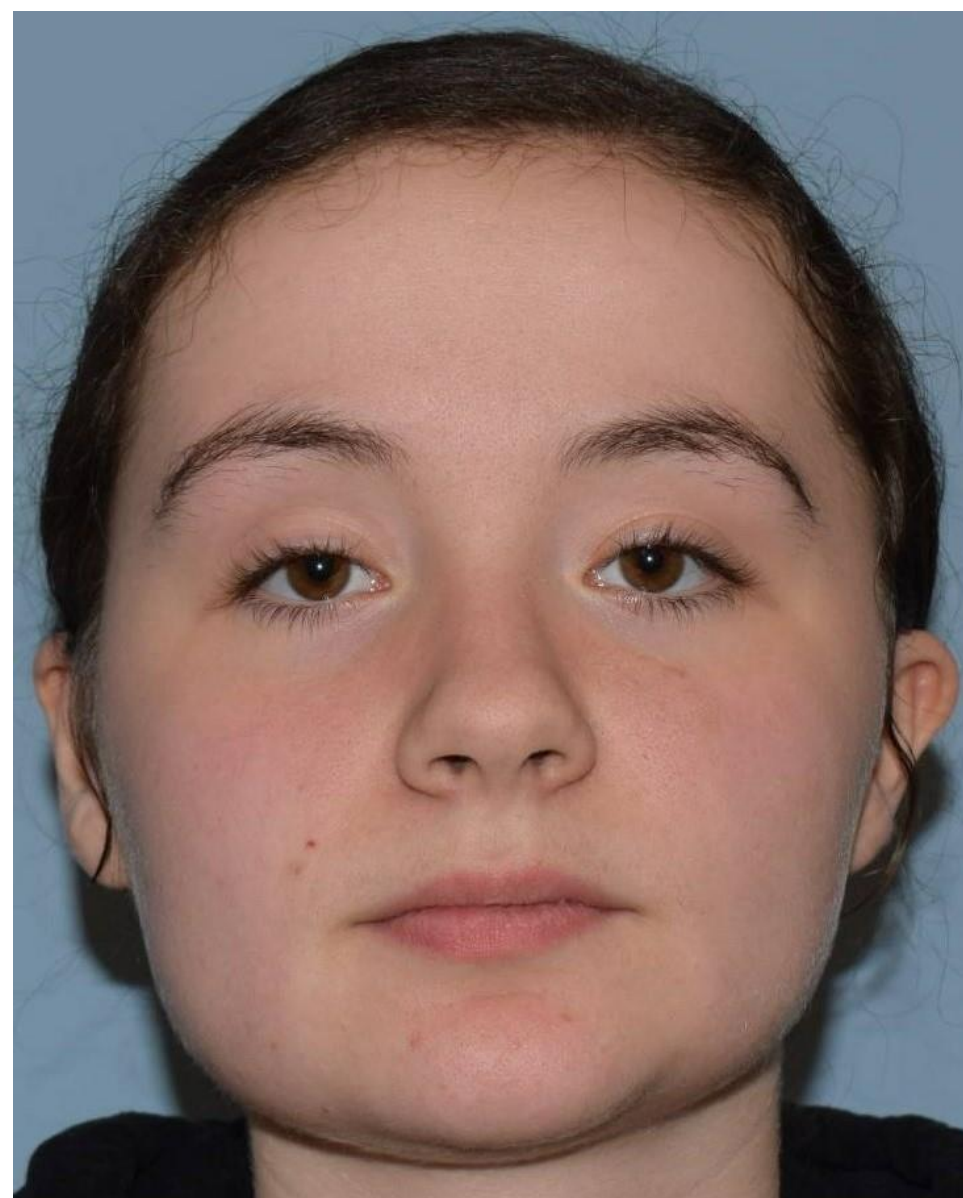
- Isolated unilateral lambdoid synostosis is the rarest form of craniosynostosis comprising 3% of operated cases.
- It is characterized by ipsilateral occipital flattening and a compensatory occipitomastoid bulge, contralateral parietal and frontal bossing, posterior and inferior displacement of the ipsilateral ear, and significant hemifacial deficiency.

Treatment

- Conventional treatment for nonsyndromic unilateral lambdoid synostosis is posterior vault remodeling (PVR), which is reported to subjectively improve craniofacial dysmorphism in comparison to nonoperative management.
- Recent quantitative evidence suggests that PVR normalizes cranial vault asymmetry with sustained correction seen at 2 years postoperatively.
- The long-term outcomes are unclear due to the rarity of the condition, with most studies consisting of case reports or small series with limited follow-up.

OBJECTIVES

This study combined qualitative and quantitative approaches to evaluate outcomes of isolated nonsyndromic unilateral lambdoid synostosis.

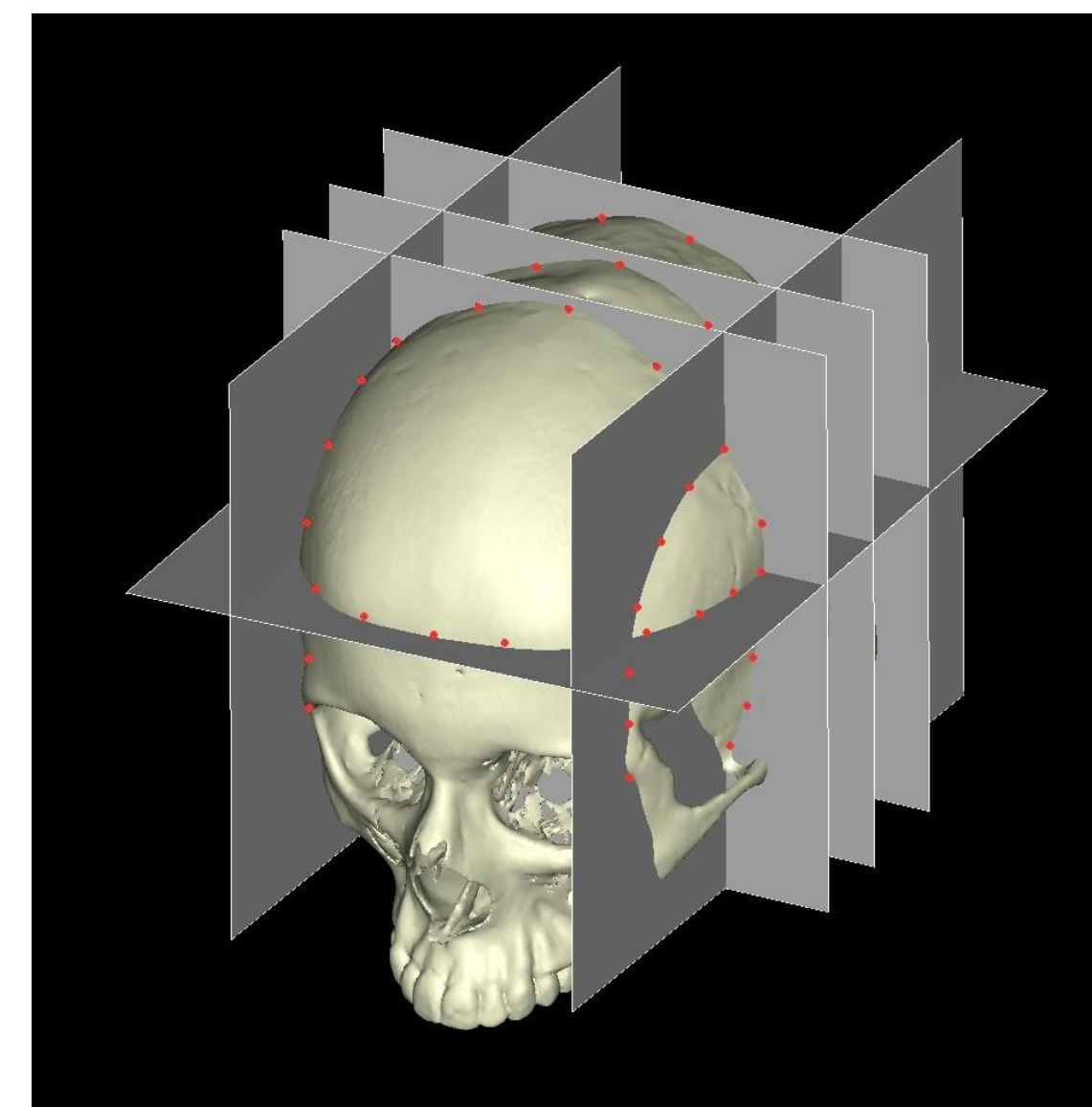
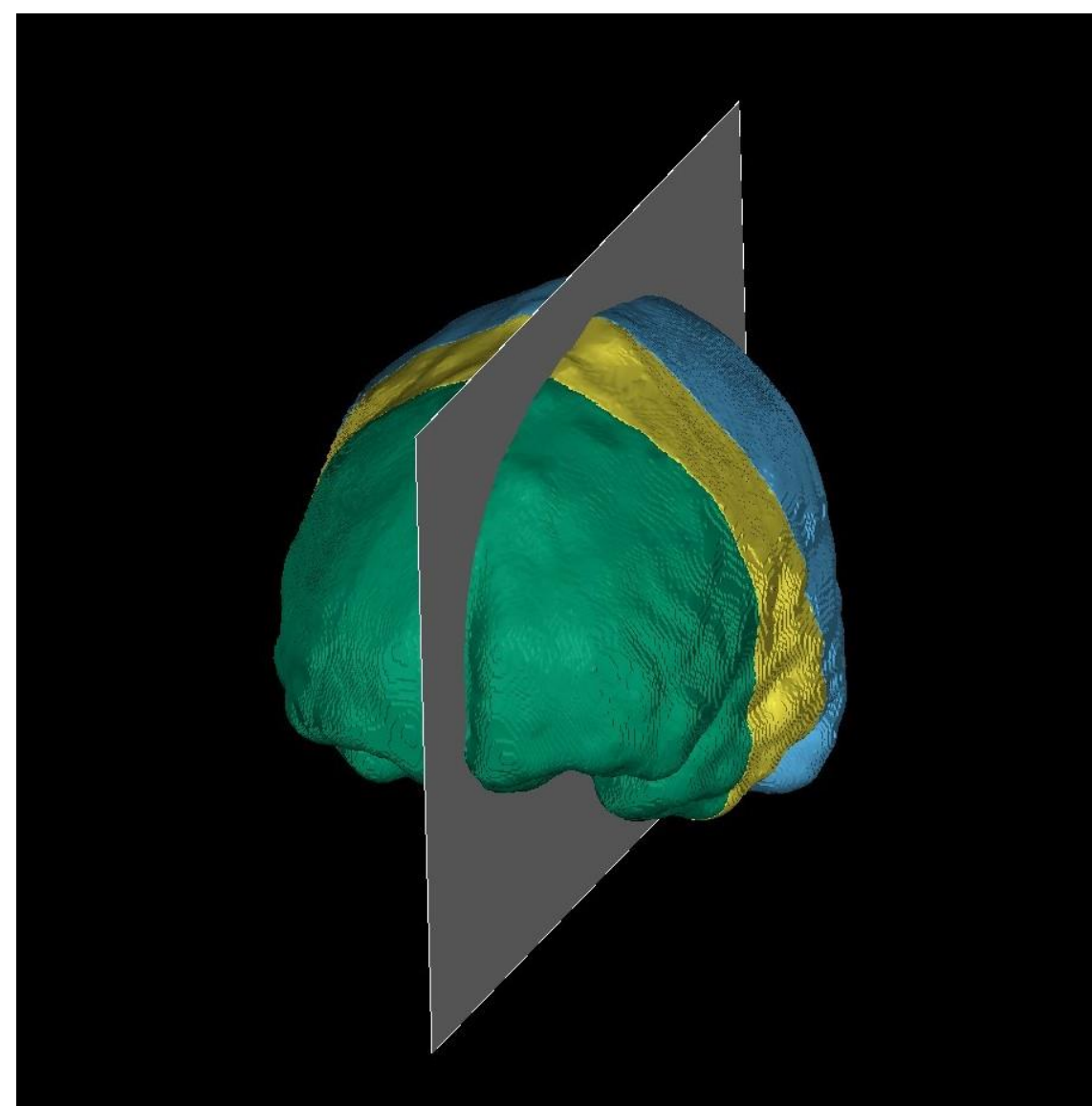


Preoperative photographs (left and middle) of a patient with right lambdoid synostosis demonstrating left-sided parietal bossing, ear asymmetry, and right-sided c-shaped deviation of the face. Following posterior vault remodeling (PVR) at age 7 months, the patient had resolution of facial scoliosis as demonstrated in a postoperative photograph (right) taken at age 18.

METHODS

Cohort

- Those with an associated genetic syndrome, inadequate medical records, and/or who underwent initial treatment at an outside institution were excluded.
- Whitaker grades were assigned to patients by the senior author (S.P.B.)
- All patients underwent a posterior switch cranioplasty.



Morphometric and Vector Analyses

- 3D craniometric analyses were performed on high-resolution CT scans taken at 3 years postoperatively or later.
- Preoperative scans, when available, were also analyzed.
- The control group was comprised of CT scans from age- and sex-matched patients who underwent imaging for non-craniofacial indications
- Morphometric and vector analyses were based on previously described methodology (Nguyen, Purnell et al. 2023).

Frontofacial Feature Analysis

- Subjects with clinical photographs taken preoperatively and greater than 3 years postoperatively were included in the frontofacial feature analysis.
- Frontal, right lateral, and left lateral photographs were used for all time points.
- Three attending craniofacial surgeons (S.P.B., J.A.T., J.W.S.) independently reviewed the photographs and were blinded to laterality of synostosis.
- Reviewers commented on the presence and laterality of the listed features
- Disagreements were resolved by majority opinion.

RESULTS

Cohort Characteristics

- Among 26 patients, median age of surgery was 10 months (IQR 8-15), and follow-up was 5.9 ± 5.7 years.
- Whitaker grades at age 7.3 ± 5.5 (n=22): I: 91%, II: 5%, III: 5%
- 2 (8%) patients underwent secondary vault procedures
- 2 (8%) reported intermittent headaches at 5.9 ± 5.7 years postoperatively

3D Morphometric Analysis

- Mean age at postoperative scan was 11.5 ± 5.3 years (n=9)
- In comparison to the control group, the lambdoid cohort had:
 - Increased anterior vault volume (fused side), posterior fossa deflection angle, cranial base angle
 - Decreased posterior vault volume (fused side), posterior vault volume ratio
 - Inferior and posterior ear position (fused side)

Frontofacial Feature Analysis

- Among 10 patients who were included in the frontofacial feature analysis, mean age was 0.8 ± 0.3 years at the preoperative images and 8.8 ± 4.6 years at the postoperative images.
- Preoperatively, all subjects had one or more frontofacial abnormalities, 9 (90%) had ear position asymmetry, among whom 8 (80%) demonstrated parietal bossing on the unfused side, and 5 (50%) had c-shaped deviation of the face toward the stenosed side.
- Postoperatively, 5 (50%) had one or more residual dysmorphic frontofacial features, among whom 4 (40%) had ear position asymmetry, 2 (20%) had parietal bossing, and 1 (10%) had frontal bossing of the patent side.
- C-shaped deviation resolved in all patients

Predictors of Long-Term Outcomes:

- Total abnormal features preoperatively correlated with number of residual deformities postoperatively on frontofacial feature analysis ($r(8)=.676$, $p=.032$).
- Among patients with consecutive craniometric data, posterior vault volume ratio normalized over time (0.76 ± 0.07 versus 0.89 ± 0.12 , $p=.002$), with the most dramatic correction following initial PVR
- Additionally, cranial base angle decreased from 5.1 ± 2.9 degrees preoperatively to 2.8 ± 3.0 degrees long-term postoperatively ($p=.05$).
- Age of surgery correlated with postoperative middle cranial fossa deviation ($r(7)=.823$, $p=.044$).

CONCLUSION

Most patients did not require secondary cranial vault procedures with long term follow-up. Relative to normal controls, most patients had mild persistent volumetric asymmetry, ear displacement, and skull base abnormalities. Most did not have clinically significant frontofacial differences at long-term follow-up, despite these craniometric abnormalities.

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