Does Conventional Posterior Vault Remodeling Alter Endocranial Morphology in Patients With True Lambdoid Synostosis?

Vivian M. Hsu MD, River M. Elliott MD, James M. Smartt Jr. MD, Jesse A. Taylor MD, Scott P. Bartlett MD

Department of Surgery, Division of Plastic Surgery, Hospital of the University of Pennsylvania, Philadelphia, Pennsylvania

BACKGROUND

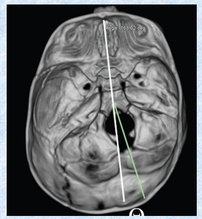
True Lambdoid Synostosis (TLS)

- Occurs in 1 in 40,000 live births
- 1-3% of all craniosynostosis cases

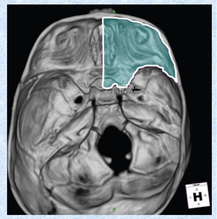
Lambdoid Phenotype

- Downward cant
- Ipsilateral mastoid bulge
- Trapezoidal head shape
- Contralateral hemifacial deficiency

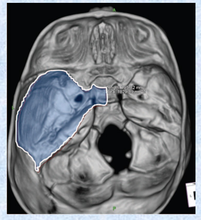
Endocranial Features



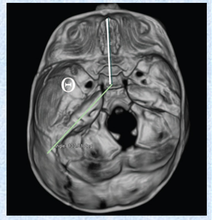
Deviation of posterior fossa toward affected suture



Normal anterior cranial fossa



Expanded contralateral middle cranial fossa



Larger contralateral petrous ridge angle

OBJECTIVES

1. We hypothesize that these endocranial features persist following surgery, causing the persistent postoperative hemifacial deficiency seen in these patients.
2. Our goal is to determine what effect conventional posterior vault remodeling has on endocranial morphology in patients with TLS.

METHODS

- Retrospective Case Series
  - All patients diagnosed with TLS at CHOP (1990-2010)
  - CT proven craniosynostosis
  - Underwent posterior vault remodeling
  - Adequate pre and post op CT scans (Slice thickness 2mm or less)
- 3D reconstructions performed on TeraRecon Aquarius workstations
- Standard measurements of endocranial base:
  - Anterior Cranial Fossa Area (AFA)
  - Middle Cranial Fossa Area (MCF)
  - Petrous Ridge Angle (PRA)
  - Posterior Fossa Deflection Angle (PFA)
  - External Auditory Meatus Angle (EAMA)
  - Position of TMJ (TMJ)

STATISTICS

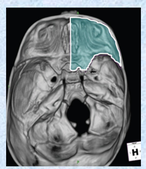
- Population too small to permit direct comparison of means
- Value of angles compared between pre and post op CT scans
- Two-tailed Student's t-test used to compare measurements
- For areas, the relative difference between sides was calculated and compared between pre and post op CT scans:

$$\text{Relative Difference}(\%RD) = \frac{100\ (\text{affected} - \text{unaffected})}{(\text{affected} + \text{unaffected}) / 2}$$

RESULTS

- Five patients met criteria for enrollment (2F, 3M)
- Mean age at pre op CT: 1.05 years
- All underwent posterior vault remodeling using a "Switch Cranioplasty" technique at a mean age of 1.33 years
- Post op CT scans were obtained at a mean age of 3.15 years
- Mean 1.82 years between surgery and post op CT

Switch Cranioplasty With Occipital Bar Advancement

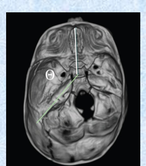


Anterior Cranial Fossa

- Symmetrical pre op (Mean %RD = -4.69)
- Symmetrical post op (Mean %RD = 1.48)
- No significant change (p = 0.21)

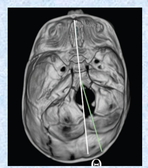
Middle Cranial Fossa

- Pre op: contralateral enlargement (Mean %RD = -34.9)
- Post op: contralateral enlargement (Mean %RD = -32.3)
- No significant change (p = 0.58)



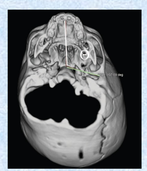
Middle Cranial Fossa

- Ipsilateral PRA changed from 120.1 to 121.1 (p = 0.95)
- Contralateral PRA increased from 130.1 to 135.1 (p = 0.02)
- Significant increase in retro-displacement in contralateral PRA



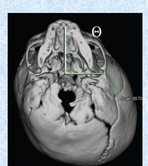
Posterior Fossa

- Pre op: posterior fossa deviated toward fused suture by mean 8.9 degrees
- Post op: posterior fossa deviation by mean 9.7 degrees
- No significant change (p = 0.76)



Ear Position

- Pre op: Unaffected side retrodisplaced relative to affected side (p = 0.01)
- Post op: Unaffected side retrodisplaced relative to affected side (p = 0.03)
- No significant change after surgery for affected (p=0.21) or unaffected (p=0.22) sides

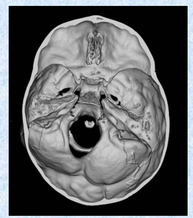


TMJ Position

- Symmetrical pre op (p = 0.24)
- Symmetrical post op (p = 0.07)
- No significant change after surgery for affected (p = 0.80) or unaffected (p = 0.57) sides

CONCLUSIONS

- Conventional vault remodeling restores calvarial shape but does not affect the abnormal growth of the endocranial base.



Age at Surgery: 0.34 years

Age at Follow-up CT: 2.64 years

- Contralateral middle cranial fossa remains enlarged relative to ipsilateral side post op and becomes more retrodisplaced.
- Ongoing retrodisplacement of contralateral MCF demonstrates that deforming forces exerted on skull base by fused lambdoid suture persist.
- The twisting and asymmetry of the contralateral MCF may explain why a persistent hemifacial deficiency is seen in these patients.

FUTURE DIRECTIONS

- Recent studies have demonstrated that expansion of the calvarial vault with distraction osteogenesis significantly alters the deformed endocranial base in patients with unicoronal synostosis. *(Choi et al, Plast Reconstr Surg. 2010 Sep;126(3):995-1004.)*
- The senior author has shown the feasibility of posterior vault distraction. *(Steinbacher et al, Plast Reconstr Surg. 2011 Feb;127(2):792-801.)*
- Our future work will be directed toward expansion of the posterior vault in patients with TLS and restoration of both the endocranial base and facial skeleton.