Multidisciplinary management of craniosynostosis in school kids and adolescents.

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Introduction

Surgical decision-making in older patients who are not treated in infancy for their craniosynostosis is far from standardized. Only limited guidance for surgical intervention in these patients is available (Strahle 2011). We present a cohort of patients with non-syndromic craniosynostosis. Besides the cosmetic concerns, a significant subset of these patients have daily non-focal headaches, neurodevelopmental delay, behavioural changes and Chiari malformation with and without syringohydromyelia.

Methods

Patients with delayed diagnosis and treatment of their craniosynostosis between January 2008 and December 2013. Inclusion criteria were age greater than one year at time of evaluation, fusion of at least one suture, and adequate imaging studies including CT or MRI or both. We reviewed medical records and radiographic images. Type of craniosynostosis, age at evaluation, past medical history, surgical findings, developmental abnormalities, ophthalmologic findings and clinical course were reviewed and recorded. All relevant data on patient's demographics and findings are summarized in TABLE 1.

	DX	AGE	MRI findings (Chiari and/or abnormal CINE flow)	Headache	Developmental findings
# 1	Pan suture synostosis	6	*ABNORMAL*	+	Severe
#2	Metopic	7	*ABNORMAL*	+	None
#3	Sagittal	11	none		Mild
#4	left UCS, secondary sagittal	6	*ABNORMAL*	+	Mild to Moderate
#5	Sagittal	4	*ABNORMAL, SYRINX*		Mild
#6	Metopic and Sagittal	5	none		Moderate
#7	Metopic	2	none		None
#8	Sagittal (microcephaly)	3	none	+	Severe
#9	Pan suture synostosis	17	Post chiari decompression	+	Mild

Table 1

Results

7 out of 9 patients presented with developmental delay and behavioural changes. Five patients had daily non-focal headaches.

With the exception of the 17 yo male patient with pan suture synostosis, none of the patients were noted to have papilledema. On imaging, three patients presented with Chiari malformation w/wo syringohydromyelia and reduced CSF flow on CINE studies. Two of them required posterior fossa decompression. In the 17 yo male with pan suture synostosis, a history of Chiari decompression at age of 11, and headaches as the primary complaint, we performed diagnostic ICP monitoring. The overnight measurements revealed significantly elevated ICP with A- and B-wave activity. In total, five of the patients underwent posterior cranial vault remodeling for concern of headaches and/or developmental delay. Intraoperative epidural monitoring was done in 3 patients at the start of the cranial vault repair, which 2/3 showed increased ICP levels. All surgical procedures were done for posterior vault remodeling using the Armadillo technique (Figure 1). There were no peri- or postoperative complications. 3/4 patients who presented with headaches experienced improvement.

Conclusions

Our findings suggest that children with uncorrected craniosynostosis associated with headache, developmental delay, or Chiari malformation benefit from cranial reconstruction. The impact of our findings is to provoke the debate: Counseling these patients and their families presents a challenge given the absence of data on intervention in this age group. Older pediatric patients may require a more extensive operation and have decreased calvarial regeneration potential, increasing the risk for residual skull defects (Rottgers, Kim et al. 2011). Furthermore, since the explosive growth phase of the young brain is largely complete after 3 years of age (Chugani 1987), the functional utility of delayed cranial vault remodeling must be evaluated in greater depth.

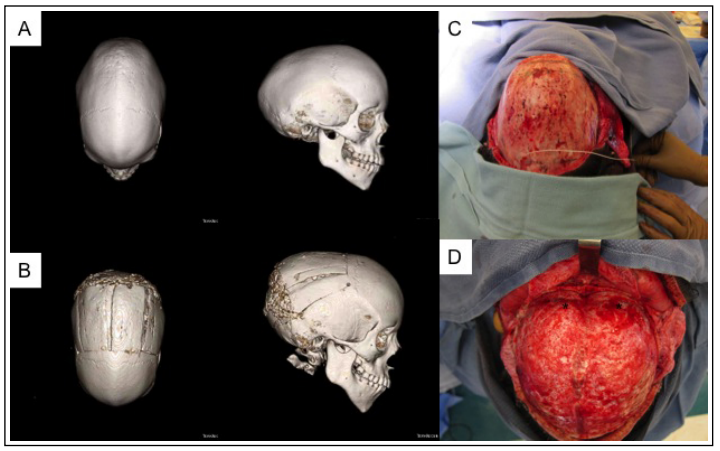

Fig.1: Split calvarial method to expand the biparietal volume. This is an 11 year old boy (#3) with mild developmental delay and psychological problems. The MRI findings were negative for posterior fossa pathology, and also, there were no headaches. (A) Preop 3 D CT scan, (B) Postop 3 D Ct scan showing the posterior cranial vault reconstruction, (C) Craniectomy sites are outlined with the pencil on the patients skull. The ICP probe is placed epidurally through a frontal burr hole and baseline ICP (D) The craniectomy extends into the posterior fossa below the inion to maximize expansion posterior vault capacity. Both transverse sinus are marked (asterisks) to show the extent of the craniectomy.

References

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