Craniosynostosis: To Study the Spectrum and Outcome of Surgical Intervention at a Tertiary Centre In India

Charandeep Singh Gandhok Mch Neurosurgery; Simran Syal; Ajay Sharma; Arvind Kumar Srivastava; Daljit Singh MS, MCh

[Maulana Azad Medical College and G. B. Pant Hospital (GIPMER), New Delhi, India]

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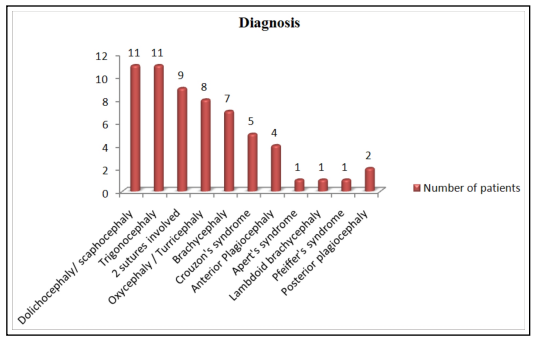
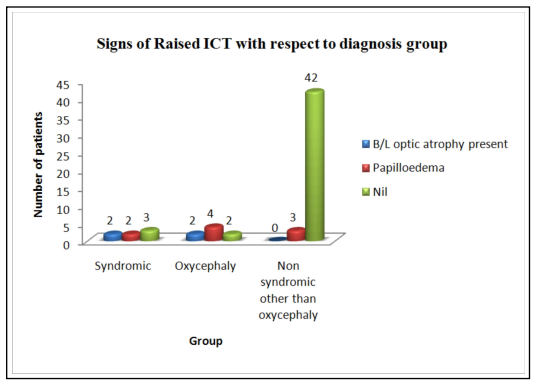
Introduction: Craniosynostosis is defined as premature fusion of the cranial sutures. This can lead to abnormal head shape, abnormal facial features, visual impairment due to intracranial hypertension, hydrocephalus due to mechanical increase of the CSF outflow resistance and venous hypertension, Chiari malformation secondary to a disproportion between the posterior fossa and growing hindbrain structures, obstructive sleep apnea due to midface hypoplasia or an impairment of neurocognitive development.

Methods: After obtaining ethical clearance, we conducted a cross sectional study in our department which included 60 cases of craniosynostosis operated in our hospital from 2010 till 2015 and who were on regular follow up. Data was collected including name, age, gender, involved sutures, other medical comorbidities, whether syndromic craniosynostosis or not, whether signs of intracranial hypertension were present or not, any findings on MRI, type of surgery performed, complications of surgery (if any) and findings at follow up. To be able to analyse the surgical results, we used the seven category classification system used by Sloan et al.

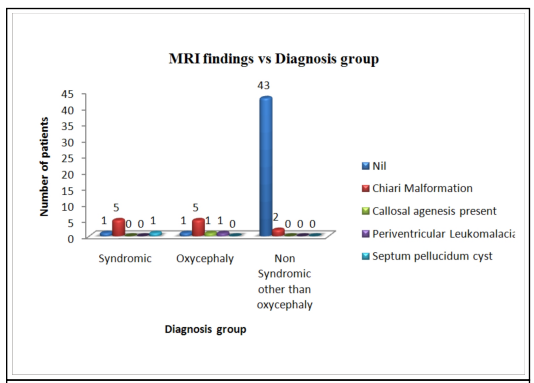
Results: 1) In non syndromic cases (53), scaphocephaly and trigonocephaly groups had the maximum number of cases (11 each). **2)** Mean age at first surgery in the 60 cases was 3.85years. Mean age at first surgery was earliest in the

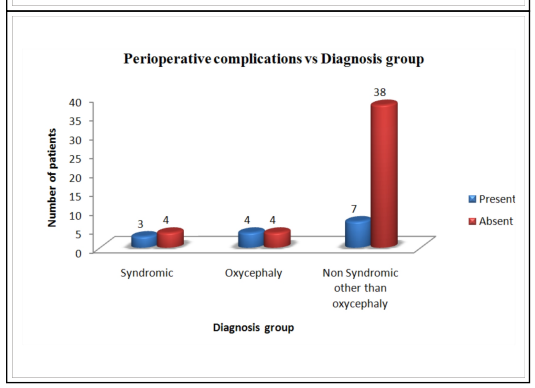


anterior plagiocephaly and trigonocephaly groups, i.e. 1.67 years and 1.83 years respectively. **3)** 4 out of 5 cases of Crouzon's syndrome (80%) and 6 out of 8 cases of oxycephaly (75%) had signs of raised intracranial pressure. **4)** There was a significant association between syndromic craniosynostosis/ oxycephaly and higher Sloan et al class (compromised correction). **5)** All 11 patients of trigonocephaly showed class I correction.



6) Chiari malformation was present in 80% of the cases of Crouzon's syndrome and 62.5% of the cases of oxycephaly.





Conclusions: Mean age at first surgery was very high due to late presentation of the cases.

The incidence of metopic synostosis is rising.

Chiari malformation, intracranial hypertension and peri-operative complications were a frequent finding in multi-sutural and syndromic craniosynostosis.

The best surgical outcome and the least peri-operative complications were seen in the trigonocephaly group.

Learning Objectives: Syndromic craniosynostosis and multi-sutural craniosynostosis surgeries are complex surgeries and require a certain level of expertise. Peri- operative complications are more common in these cases as compared to single suture craniosynostosis.

MRI Brain and cranio-vertebral junction should be done in every patient of craniosynostosis especially the syndromic cases, to look for Chiari malformation or intrinsic brain abnormalities.

References:

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Sloan GM, Wells KC, Raffel C, McComb JG. Surgical treatment of craniosynostosis: Outcome Analysis of 250 consecutive patients. Pediatrics. 1997 July; Vol 100(1):E2