Craniosynostosis has a varied clinical spectrum, ranging from isolated single suture involvement to multi-sutural fusions. Greater understanding of the pathogenesis of craniosynostosis has led to the development of practical treatment protocols. Three stages of growth have determined the approach to managing craniosynostosis: the early period, up to 12 months; the intermediate period, from 1 to 10 years; and the late period, beginning at 10 years. This review discusses current surgical management and future perspectives in craniosynostosis.

Key Words: Craniosynostosis · Neurosurgery · Pediatric.
growing brain to expand. If urgent decompression was required, fronto-orbital advancement and multiple suture excisions were performed, resulting in lateral and posterior expansion.

The intermediate period, from 1 to 10 years

After the second year, cerebral growth slows; nevertheless, severe craniosynostosis may still lead to papilledema and potential visual failure. The cranial capacity must therefore be expanded by large bilateral decompressive craniotomies, generous fronto-orbital advancement, or a combination of these procedures.

Late period, beginning at age 10 years

Definitive facial surgery can be performed, beginning at age 10 years, although waiting until maturity may achieve better aesthetic outcomes. Patients with Crouzon syndrome who have proptosis and/or maxillary hypoplasia are especially likely to need surgical treatment.  

Early surgical correction has been limited by a late diagnosis and the risks associated with intraoperative blood loss, which was less effectively managed in the past than currently. Since the beginning of the 21st century, many centers have tended to determine whether a less invasive procedure could be performed at an earlier stage with acceptable risk. Endoscopic linear craniectomy, with postoperative application of helmets and cranial remodeling through small skin incisions, has led to cosmetic results comparable to those of more invasive procedures if performed during the first 4–6 months of life, with acceptable blood loss and operative risks. Surgical management of complex craniosynostosis has also changed significantly. For many years, the early treatment of this condition consisted of bi-frontal advancement. More recently, the combination of frontal advancement and posterior cranial enlargement during the first months of life has been found to protect both the ocular and posterior cranial structures. Currently, a free bone flap (floating technique) or springs are used to allow for cerebral growth until a rigid frontoorbito-maxillary advancement can be performed. The advent of osteodistraction has lowered the age for facio-maxillary advancement (3–5 years), and may avoid the necessity of repeating fronto-orbital procedures. This has reduced the risks of dural tears and postoperative cerebrospinal fluid fistulas in a significant proportion of patients.

As the procedures used to remodel the calvarial vault are extensive, complications can occur following surgery for craniosynostosis. Although the mortality rate has been reported to be as high as 2.3%, most international studies had mortality rates of 1.5% to 2%. Most deaths were attributed to hemorrhagic complications, but various other causes have been reported, including air emboli, cerebral edema, and respiratory infections. Attention to intraoperative hemodynamics and careful postoperative intensive care unit monitoring are critical in minimizing overall morbidity and mortality rates.

FUTURE PERSPECTIVES IN CRANIOSYNOSTOSIS

The combination of early technical success with recent advances in treatment has indicated the necessity of multidisciplinary management. The overall approach can be distilled into six principles: 1) Care should be provided by multi-disciplinary teams; 2) Care should be a protocol-driven process, with all forms of care defined and delivered optimally; 3) Care should be longitudinal, as age, healing and growth processes; 4) Secure financial support is needed to implement such longitudinal care; 5) Competent professionals should be involved in ongoing education and training in teaching and research; and 6) Research should explore causes, treatment strategies, and treatment outcomes.

CONCLUSION

Centers with the appropriate vision and infrastructure are necessary to optimize the care of patients with craniosynostosis, as well as to enhance scientific knowledge and education about this disease. Several investigations have evaluated the roles of various growth factors and cytokines in determining the fate of sutures. Fibroblast growth factors (FGFs) are particularly important, as mutations in their receptors have been implicated in many craniosynostosis syndromes. Mutations in three of the four known FGF receptors have been associated with premature pathologic suture fusions. Recent advances in developmental biology and genetics have identified some of the events governing suture fate, highlighting multiple axes of cellular signaling with the potential for clinical manipulation. Such knowledge and comprehension may facilitate therapeutic translations, ultimately enhancing or perhaps even replacing contemporary modalities for treating craniosynostosis.

Acknowledgements

This research was supported by a grant of the Korean Health Technology R&D Project, Ministry of Health & Welfare, Republic of Korea (HI13C1509, HI14C0042).

References

1. Abbott AH, Netherway DJ, David DJ, Brown T: Application and comparison of techniques for three-dimensional analysis of craniofacial anomalies. J Craniofac Surg 1:119-134, 1990
2. Cohen MM Jr: Sutural biology and the correlates of craniosynostosis. Am J Med Genet 47:581-616, 1993
3. David DJ: Advances in the management of the craniosynostoses. ANZ J Surg 73:949-957, 2003
4. Davies DW, Munro IR: The anesthetic management and intraoperative care of patients undergoing major facial osteotomies. Plast Reconstr Surg 55:50-55, 1975
5. Fearon JA: Evidence-based medicine : craniosynostosis. Plast Reconstr Surg 133:1261-1275, 2014
6. French LR, Jackson JT, Melton LJ 3rd: A population-based study of craniosynostosis. J Clin Epidemiol 43:69-73, 1990
7. Grabb WC, Smith JW, Aston SJ: Plastic surgery, ed 4. Boston: Little,
8. Graham JM Jr, de Saxe M, Smith DW: Sagittal craniosenosis: fetal head constraint as one possible cause. J Pediatr 95 (5 Pt 1): 747-750, 1979.
9. Mehrara BJ, Mackool RJ, McCarthy JG, Gittes GK, Longaker MT: Immunolocalization of basic fibroblast growth factor and fibroblast growth factor receptor-1 and receptor-2 in rat cranial sutures. Plast Reconstr Surg 102: 1805-1817; discussion 1818-1820, 1998.
10. Netherway D, Abbott AH, Abbott JR, David DJ: Techniques for characterisation of the human craniofacial skeleton using computer tomography data. Perspect Hum Biol 4: 155-165, 1999.
11. Persson KM, Roy WA, Persing JA, Rodeheaver GT, Winn HR: Craniofacial growth following experimental craniosenostosis and craniectomy in rabbits. J Neurosurg 50: 187-197, 1979.
12. Singer S, Bower C, Southall P, Goldblatt J: Craniosenostosis in Western Australia, 1980-1994: a population-based study. Am J Med Genet 83: 382-387, 1999.
13. Tamburrini G, Di Rocco F: Editorial. Childs Nerv Syst 28: 1293-1294, 2012.
14. Tessier P: [Total facial osteotomy: Crouzon’s syndrome, Apert’s syndrome: oxycephaly, scaphocephaly, turricephaly]. Ann Chir Plast 12: 273-286, 1967.
15. [Treat flap ischemia-reperfusion injury by local transplanting human umbilical cord mesenchymal stem cells]. Zhonghua Zheng Xing Wai Ke Za Zhi 28: 203-207, 2012.
16. Wilkie AO, Morris-Kay GM, Jones EY, Heath JK: Functions of fibroblast growth factors and their receptors. Curr Biol 5: 500-507, 1995.
17. Wilkie AO, Slaney SF, Oldridge M, Poole MD, Ashworth GJ, Hockley AD, et al: Apert syndrome results from localized mutations of FGFR2 and is allelic with Crouzon syndrome. Nat Genet 9: 165-172, 1995.