Anesthesia for Craniofacial Surgery in Infancy

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KEYWORDS

• Craniofacial surgery • Craniosynostosis • Pediatrics • Transfusion • Endoscopic

KEY POINTS

- Complex cranial vault reconstruction remains a significant challenge for anesthetic management.
- Primary concerns include blood loss and its management.
- Evolution of procedures to treat craniosynostosis has resulted in improvements in perioperative morbidity with less blood loss and shorter operations and length of hospital stays.
- An understanding of the procedures performed to treat craniosynostosis is necessary to provide optimal anesthetic management.

INTRODUCTION

Craniosynostosis is a disorder of skull development that occurs as a result of the premature fusion of one or more cranial sutures, occurring with an incidence of approximately 1 in 2000 live births. The observed deformity relates to which sutures are affected, with characteristic deformities associated with specific suture involvement (Fig. 1). Although the relationship between craniofacial dysmorphism and fusion of the cranial sutures was observed earlier, Virchow (1851)¹ was the first to formally describe many of the more common specific abnormalities, and in particular, he was the first to describe the arrest of skull growth that occurs in a direction perpendicular to the affected suture.

Craniosynostosis most commonly presents as an isolated abnormality but can present as a component of an identified syndrome or genetic disorder (15%–40% of cases (Table 1)). Ongoing research will likely reveal genetic causes of cases currently thought to be isolated or idiopathic. In most infants, the abnormality is congenital and diagnosed within the first few months of life, whereas some infants may present later. The diagnosis is most commonly made based on the phenotype of skull deformation.

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Fig. 1. Depictions of characteristic skull deformities associated with different forms of craniosynostosis. (*A*) Plagiocephaly caused by unicoronal synostosis (approximately 20%–25% of craniosynostosis). (*B*) Trigonocephaly caused by metopic synostosis accounts (approximately 5%–15% of craniosynostosis). (*C*) Scaphocephaly caused by sagittal synostosis, the most common form of nonsyndromic craniosynostosis (40%–55% of synostosis). (*D*) Bicoronal synostosis is less common and more likely associated with syndromic craniosynostosis. (*From* Seruya M, Magge S, Keating R. Diagnosis and surgical options for craniosynostosis. In: Ellenbogen RG, Abdulrauf S, Sekhar L, editors. Principles of neurologic surgery. 3rd edition. St Louis (MO): Saunders; 2012. p. 138; with permission.)

Subsequent CT scanning and 3-D reconstruction allow for accurate diagnosis of specific suture involvement and can be used for surgical planning (Fig. 2). Untreated craniosynostosis can lead to elevated intracranial pressure (ICP) and disturbances in intellectual and neurologic development. Syndromic craniosynostosis is more commonly associated with multiple suture involvement and is also more often associated with increased ICP. Children with syndromic craniosynostosis also require multiple operations throughout infancy and childhood.

From both a cosmetic and neurodevelopmental perspective, optimal outcomes are achieved when these procedures are performed before a year of age, and earlier surgical intervention may translate to a less extensive operation. Surgical treatment of craniofacial dysmorphism associated with craniosynostosis was principally pioneered by Dr Paul Tessier. Dr Tessier presented his initial work in 1967 and went on to train the first generation of craniomaxillofacial surgeons. As a result, Dr Tessier is widely regarded as the father of modern craniofacial surgery. The development of these surgical techniques has led to dramatic improvements in cosmetic, neurodevelopmental, and psychosocial outcomes in children afflicted by these conditions.^{2,3}

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