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Preface: Craniosynostosis: Current Perspectives xi
Srinivas M. Susarla

Historical Perspectives on the Management of Craniosynostosis 333
Tyler J. Holley, Nathan J. Ranalli, and Barry Steinberg

The history of craniofacial surgery is one of many fundamental advances by monumental figures. Although craniosynostosis has been known to exist for multiple centuries, modern management has evolved over roughly the last century. An overview of early history, early scientific exploration, the advancement of surgical treatment of craniofacial deformities and the current state of craniosynostosis management is discussed. To fully appreciate the evolution of craniosynostosis surgery, one must understand the gradual advancements that have brought the specialty to this modern era.

Epidemiology, Genetics, and Pathophysiology of Craniosynostosis 341
Matthew Blessing and Emily R. Gallagher

Craniosynostosis, the premature fusion of the infant cranial skulls, can be recognized by characteristic head shape differences that worsen with head growth. Craniosynostosis can be syndromic or nonsyndromic and can involve one suture or multiple sutures. Timely cranial vault surgery is recommended to expand and reshape the skull, with a goal of preventing increased intracranial pressure and providing sufficient space for brain growth. Several gene variants and environmental exposures are known to increase the risk of single suture craniosynostosis (SSC), including in utero constraint, exposure to specific toxins and medications, and medical conditions such as thyroid dysregulation and metabolic bone disorders.

Multidisciplinary Care Considerations for Patients with Craniosynostosis 353
Emily R. Gallagher, G. Kyle Fulton, Srinivas M. Susarla, and Craig B. Birgfeld

Infants and children with craniosynostosis require multidisciplinary care, and this is best accomplished when care is provided on a craniofacial team. Most patients with craniosynostosis will have non-syndromic presentations; however, longitudinal care remains critical to ensure appropriate growth and development throughout childhood. In patients with syndromic craniosynostoses, coordinated longitudinal care becomes even more paramount because of the high level of complexity across many different specialties or disciplines. Care delivery that includes perspective and expertise from multiple disciplines is important to help patients reach their full potential and optimal outcomes.

Fronto-Orbital Advancement for Metopic and Unilateral Coronal Craniosynostoses 367
Benjamin B. Massenburg, Philip D. Tolley, Amy Lee, and Srinivas M. Susarla

Fronto-orbital advancement remains a powerful technique for the correction of anterior cranial vault differences related to metopic (trigonocephaly) or unilateral coronal (anterior plagiocephaly) craniosynostoses. Traditional fronto-orbital advancement requires access to the forehead and superior 2/3 of the orbit via a coronal incision. The frontal bone and orbital segment (bandeau) are then separated from the skull and reshaped.
In patients with metopic craniosynostosis, the bandeau and frontal bone will need to be advanced and widened. In patients with unilateral coronal craniosynostosis, the bandeau will need to be “untwisted” to address the supraorbital retrusion on the affected side, the affected orbit will need to be shortened and widened, and the frontal bone flap will need to be proportionately advanced on the affected side. Overcorrection of the affected dimension should be undertaken to account for growth and relapse.

Management of Unicoronal and Metopic Synostoses: Minimally Invasive Approaches

Gabriel M. Hayek, David F. Jimenez, and David M. Yates

Early endoscopic-assisted correction of unicoronal and metopic synostosis is an excellent, safe, cost-effective, and highly effective option for affected patients. Although open calvarial remodeling has a place in the armamentarium of the craniofacial team, the skull base changes seen in endoscopic-assisted techniques are unparalleled. The procedures are associated with low morbidity and no mortality. There is minimal blood loss, decreased operating time, significantly reduced blood transfusion rates, decreased hospitalization length, decreased cost, and less pain and swelling. Early diagnosis and referral for surgical evaluation are critical to obtaining these results.

Management of Sagittal and Lambdoid Craniosynostosis: Open Cranial Vault Expansion and Remodeling

Michael R. Markiewicz, Matthew J. Recker, and Renée M. Reynolds

The prevalence of sagittal and lambdoid suture craniosynostosis differs considerably, as they are notably the most and least prevalent sutures involved in isolated suture craniosynostosis, respectively. The goals of reconstructing the cranial vault in both entities is the same: to release the fused suture, expand cranial volume, restore normal head shape and morphology, and allow for normal growth of the cranial vault. With regards to sagittal suture synostosis, opinions vary on whether reconstruction should focus on either the anterior or poster cranial vault. In contrast, the poster cranial vault is always targeted in lambdoid suture craniosynostosis.

Management of Sagittal and Lambdoid Craniosynostosis: Minimally Invasive Approaches

Sameer Shakir, Melissa Roy, Amy Lee, and Craig B. Birgfeld

The resurgence of strip craniectomies began in the mid-1990s with advances in surgical technique and anesthesia coupled with the critical observation that earlier interventions benefitted from an easily molded skull. Jimenez and Barone’s pioneering introduction of endoscopic approaches to strip craniectomies coupled with postoperative helmeting in newborns and young infants and Claes Lauritzen’s introduction of spring-mediated cranioplasty began the era of minimally invasive approaches in the surgical correction of craniosynostosis. This article provides technical descriptions of these treatment modalities, a comparative literature review, and our institutional algorithms for the correction of sagittal craniosynostosis and unilambdoid craniosynostosis.

Management of Minor Suture Craniosynostosis

Alisa O. Girard and Robin Yang

Although most reported cases of minor suture involvement include multiple sutures, isolated suture involvement has been reported. Morphologic differences such as scaphocephaly and anterior plagiocephaly have been reported. Management should involve proper identification and multidisciplinary treatment. Surgical treatment should involve the expansion of the cranial vault as well as the correction of the deformity.
Syndromic Craniosynostosis: Cranial Vault Expansion in Infancy 443
Sameer Shakir and Craig B. Birgfeld

Syndromic craniosynostosis (CS) represents a relatively uncommon disease process that poses significant reconstructive challenges for the craniofacial surgeon. Although there is considerable overlap in clinical features associated with various forms of syndromic CS, key extracranial features and close examination of the extremities help to distinguish the subtypes. While Virchow’s law can easily guide the diagnosis of single suture, nonsyndromic CS, syndromic CS traditionally results in atypical presentations inherent to multiple suture fusion. Coronal ring involvement in isolation or associated with additional suture fusion is the most common pattern in syndromic CS often resulting in turribrachycephaly.

Syndromic Synostosis: Frontofacial Surgery 459
Kevin Chen, Katelyn Kondra, Eric Nagengast, Jeffrey A. Hammoudeh, and Mark M. Urata

Frontofacial surgery, encompassing the monobloc with or without facial bipartition and the box osteotomy, can treat the frontal bone and midface simultaneously, providing comprehensive improvement in facial balance. Complex pediatric patients with genetic syndromes and craniosynostosis are most optimized by an interdisciplinary team of surgeons, pediatricians, geneticists, speech pathologists, audiologists, dietitians, pediatric dentists, orthodontists, and psychosocial support staff to manage the myriad of challenges and complications throughout early childhood and beyond. Despite early treatment of the anterior and posterior cranial vault, these patients frequently have resultant frontal and/or midface hypoplasia and orbital abnormalities that are best managed with simultaneous surgical treatment.

Subcranial Midface Advancement in Patients with Syndromic Craniosynostosis 467
Benjamin B. Massenburg, Srinivas M. Susarla, Hitesh P. Kapadia, and Richard A. Hopper

Patients with syndromic craniosynostosis can present with midface hypoplasia, abnormal facial ratios, and obstructive sleep apnea. These symptoms can all be improved with midface advancement, but it is essential to evaluate the specific morphologic characteristics of each patient’s bony deficiencies before offering subcranial advancement. Midface hypoplasia in Crouzon syndrome is evenly distributed between the central and lateral midface and reliably corrected with Le Fort III distraction. In contrast, the midface hypoplasia in Apert/Pfeiffer syndromes occurs in both an axial and a sagittal plane, with significantly more nasomaxillary hypoplasia compared with the orbitozygomatic deficiency.

Orthognathic Surgery in Patients with Syndromic Craniosynostosis 477
Jesse T. Han, Mark A. Egbert, Russell E. Ettinger, Hitesh P. Kapadia, and Srinivas M. Susarla

Patients with syndromic and nonsyndromic synostosis may have end-stage skeletal discrepancies involving the lower midface and mandible, with associated malocclusion. While orthognathic surgical procedures in this population can be reliably executed, the surgeon must be aware of the unique morphologic characteristics that accompany the primary diagnoses as well as the technical challenges associated with performing Le Fort I osteotomies in patients who have undergone prior subcranial midface distraction.