Harvey Cushing’s experience with cranial deformity

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Surgery for cranial deformity was associated with significant surgical morbidity during the early part of the 20th century. For this reason, Harvey Cushing was initially not in favor of surgical treatment of craniosynostosis. Later in his career, Cushing began to operate on these children, although it never became a major focus of his practice. Several examples of his patients with cranial deformity are presented, and his limited role in the development of this field is discussed. (DOI: 10.3171/2010.9.FOCUS10191)

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Although there are numerous premodern descriptions of craniosynostosis, the scientific basis for the surgical correction of craniosynostosis dates to the early 19th century. Early pioneers in this work included Samuel von Sömmerring (1755–1830) and Rudolf Virchow (1821–1902). In 1800, von Sömmerring’s description of cranial sutures and their role in cranial vault development and growth suggested that premature suture closure would lead to subsequent cranial deformity.18 In 1851, Virchow published his paper, giving rise to Virchow’s law: “When premature fusion of the cranial vault occurs there is an inhibition of the normal growth of the skull in a direction perpendicular to the suture which is fused, which gives a compensatory growth in a direction parallel to the fused suture.”1,15 It was in this same publication that Virchow coined the term craniostenosis.3

Progress with the surgical correction of this disorder lagged several decades behind the early pathophysiological descriptions. In August 1888, strip craniectomies were performed in a child with craniosynostosis in San Francisco by L. C. Lane (1830–1902). Unfortunately, the child died of anesthetic complications.12 In 1890, the novel use of a double parasagittal linear craniectomy was reported by Lannelongue with an excellent outcome.13 For every surgical success, however, there were numerous failures that led to open verbal attacks on those surgeons who would perform such operations. Abraham Jacobi (1830–1919) was a well-known New York pediatrician who openly criticized craniectomies:11

The last subject I dare to discuss before you is that of linear craniotomy… such rash feats of indiscriminate surgery, if continued, moreover in the presence of fourteen deaths in thirty-three cases, are stains on your hands and sins on your souls. No ocean of soap and water will clean those hands, no power of corrosive sublimate will disinfect the souls.11

The high mortality rate associated with craniectomy for craniosynostosis during this period made this operation especially controversial during the early years of Harvey Cushing’s practice.

Cushing’s Early Opinions on Cranial Deformity Surgery

Secondary microcephaly, resulting from primary brain abnormalities rather than craniosynostosis, was often incorrectly diagnosed as craniosynostosis during the early years of Cushing’s practice. Cushing described the introduction of linear craniotomy as “a lamentable instance of the juror operandi running away with surgical judgment.”15 Cushing believed that “educational rather
than surgical or medicinal measures are indicated…” and furthermore, “only when epilepsy or hydrocephalus is superadded to the defect can we have any expectation—and that a remote one—of benefiting the condition…” By 1908, the improper surgical treatment of microcephalic patients led to this warning from Cushing on the indiscriminate performance of craniectomies:

It is unaccountable that an idea should have arisen attributing microcephalus and other conditions of congenital imperfection, as well as those of development arrested by disease, to premature closure of the cranial bones. All of our knowledge goes to show that an early closure of fontanel and suture is due to a primary failure of growth of the encephalon; not the reverse, a failure of growth due to a primary closure. Experiments by d’Abundo and others show that animals whose skulls in early life have been firmly enclosed, so that there is no possibility of cerebral expansion, cannot survive. Even firmly closed sutures may give way before the pressure of growth. We see this even in young adult life, as a separation of sutures in acquired hydrocephalus, and in cases of tumor growth.

Perhaps Cushing’s opinion at that time is best summarized by his remark, “There has been a high mortality in these operations, and though death cannot be lamented, the surgeon is not a barbarian to execute the helpless.” Since surgical and anesthetic techniques were in their infancy at this time and the methods of blood transfusion were rudimentary, children with limited blood volume were especially at risk in such operations, which often involved significant blood loss due to manipulations of the adjacent dural sinuses.

Controversy surrounding the use of craniotomies delayed further development in the surgical management of craniosynostosis until the early 1920s when Mehner in 1921 advocated the use of simple extirpation of the synostosed suture and successfully used strip craniectomy to reduce further development in the surgical management of craniosynostosis until the early 1920s when Mehner in 1921 advocated the use of simple extirpation of the synostosed suture and successfully used strip craniectomy to remove a fused suture.2,6 At that time, further recognition of the risk to normal brain function and vision associated with craniosynostosis was used to justify these early surgeries.1,8

**Cranial Deformity in Cushing’s Practice During the 1920s**

We present photographs of 4 patients treated by Dr. Harvey Cushing at the Peter Bent Brigham Hospital between 1921 and 1924 (Figs. 1–4). Photographs of 2 of these patients (Figs. 3 and 4) have been presented previously without discussion,4 and the other patients (Figs. 1 and 2) are presented now for the first time. The first patient in this series (Fig. 1) was an infant admitted to the Brigham with an appearance consistent with secondary microcephaly. The primary cause of the microcephaly is not known. Images of the child seem to demonstrate an overriding of the coronal sutures, which would be consistent with the type of primary failure of growth of the encephalon described by Cushing. This child was not treated surgically, which is not surprising given Cushing’s recorded opinions on unnecessary surgery for this condition.

The second patient in this series presented to Dr. Cushing with an appearance consistent with a craniofacial syndrome (Fig. 2). The almost conical appearance of the calvaria is consistent with a diagnosis of oxycephaly. In addition, given the midface hypoplasia, oxycephaly, exorbitism, broad great toes, and subtle syndactaly or webbing of the toes, a diagnosis of Pfeiffer’s syndrome may be considered. Such a precise syndromic designation was difficult or impossible in Cushing’s day. Pfeiffer’s syndrome was not described until 1964, although Crouzon’s (1912) and Apert’s (1906) syndromes had been recently described at the time that Cushing was treating this patient.7 We are fortunate in that the surgical treatment of this child was recorded in great detail. Although his diagnosis is not recorded, the careful photography of the toes suggests that Cushing appreciated the syndromic nature of this child’s condition. From an examination of Cushing’s skin incisions as well as the preserved pieces of removed cranial bone, it is evident that Cushing performed multiple small craniotomies on this child. Craniotomies were carried out over the lateral sphenoid wing bilaterally as well as over the bregma. It is difficult to say with precision the exact rationale for the specific craniotomies performed by Cushing; however, the removal of bone at the lateral sphenoid wing could have been an attempt at addressing the patient’s proptosis due to the exorbitism while simultaneously increasing intracranial volume. Although the patient’s aesthetic appearance at extended follow-up would be considered disappointing by the standards of a modern practice, the postoperative result was quite favorable in an era when mortality was still a common outcome of this surgery. It is
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interesting that this child was older than the typical child undergoing craniofacial surgery today. This likely reflects both a reluctance to perform surgery until it was obviously necessary, as well as the relative safety of operating on an older child with a larger blood volume.

In contrast with the previous example, the third child in this series presented to Dr. Cushing at a very early age (Fig. 3). The cranial shape is most consistent with plagiocephaly due to left unicoronal synostosis. Note the recessed supraorbital rim and flattened forehead on the left side. The patient also demonstrates a classic “C” deformity with deviation of the nasal root and chin point to the opposite side. Such a finding represents the facial manifestations of the compensatory changes in this condition. A small craniectomy was performed, and multiple postoperative photographs were taken. Despite the fact that this craniectomy would not be recommended today for coronal synostosis, the operative result would have been considered satisfactory under the circumstances. The follow-up photographs of this child do seem to demonstrate somewhat of a diminished progression of the deformity. Given this child’s small blood volume, hemostasis would have been a principal concern during this surgical procedure, as methods to control intraoperative bleeding were quite primitive at that time. Cushing began his initial work with intraoperative electrocautery several years after this child underwent surgery.24,10

The fourth child in this series also presented to Dr. Cushing at a very young age (Fig. 4). Although no preoperative photographs of this patient are known to exist, it is evident from photographs taken immediately postoperatively that this patient had a Kleebblattschädel-type deformity with associated exorbitism. Such children often demonstrate multiple suture synostoses and are often syndromic. This child underwent small bilateral craniectomies. Photographs obtained 10 days after surgery show significant swelling at both sites. We may speculate that this child suffered from increased intracranial pressure, whether as a direct result of a severe cranial suture deformity or due to associated hydrocephalus. No outcome information is available for this patient, a fact that is sadly telling, given Dr. Cushing’s normally fastidious recording of follow-up data.

Craniosynostosis Surgery After Cushing

Since the middle of the 20th century, surgical treatment of craniosynostosis utilizing linear craniectomies has become a well-established treatment with progressive refinements and increasingly good results. In 1948,
Fig. 3. Photographs of an infant with plagiocephaly presumably resulting from unicoronal synostosis (A) who was treated by Harvey Cushing. A craniectomy was performed centered on the bregma but including the medial portions of both coronal sutures (B). Photographs obtained 3 weeks postoperatively show a large incision was made over the convexity (C and D). The craniofacial deformity is still apparent postoperatively, especially at the left orbit. At 1 year (E) and 3 years (F and G) postoperatively, the cranium achieves a more normal shape, although some deformity persists even at the last follow-up. Photographs courtesy of the Cushing Brain Tumor Registry at Yale University.

Fig. 4. Photographs of an infant with a head deformity most consistent with Kleeblattschädel with exorbitism. Although multisutural syndromic craniosynostosis is possible, the fingers appear normal in the available images and there are no associated midface anomalies. In the immediate postoperative photographs, bilateral incisions can be seen in front of the ears (A and B). Bilateral craniectomies were carried out at both of these sites (C). Two weeks later, significant swelling is noted at the operative sites (D–F), most likely the result of persistently elevated intracranial pressure or associated hydrocephalus. Dr. Cushing’s orderly, Adolf Watzka, can be seen holding the baby for the photographer. Photographs courtesy of the Cushing Brain Tumor Registry at Yale University.
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Ingraham and coworkers\(^8\) described 50 patients, 44 of whom were treated surgically, and reported that “mortality for operation on infants with craniosynostosis is negligible when proper supportive measures are provided.” In 1954, Ingraham and Matson\(^9\) included aesthetic benefits as an indications for surgery in their textbook on pediatric neurosurgery; thus, the modern era of craniofacial surgery was born.

Cosmetic indications for craniosynostosis began to receive emphasis only after the safety of surgical correction could be assessed as minimal. This evidence came in 1968 with a review of 519 patients with craniosynostosis who were surgically treated between 1930 and 1966 with a reported mortality rate of 0.39%, a morbidity rate of 14%, and lasting undesirable sequelae of 0.58%.\(^10\) With the safety of surgical procedures established, cosmetic benefits as indications for surgery received increasing emphasis in the 1960s and 1970s.\(^2\) In 1976, Venes and Sayers\(^7\) noted that “it is generally accepted that scaphocephaly does not significantly alter brain growth and consequently the single indication for operative correction is cosmetic improvement.” It is this emphasis on cosmetic improvement that lead to the development of many new techniques to replace the simple craniectomy.\(^17\)

Conclusions

Harvey Cushing’s contributions to craniosynostosis surgery were modest in comparison with his major contributions in nearly all other areas of neurosurgery. Nevertheless, his dominance of orthodox neurosurgical opinion for many decades did influence the evolution of the surgical treatment of this condition.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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