

Multiple Suture Synostosis and Increased
Intracranial Pressure Following Repair of Single
Suture, Non-syndromal Craniosynostosis

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Abstract

Objective: Increased intracranial pressure, frequently associated with closure of multiple cranial sutures, has been reported to occur in 36% of cases following correction of syndromal craniosynostosis. Although much more uncommon, multiple suture closure may occur following repair of single suture, non-syndromal craniosynostosis and we present two such children.

Results: Two children with non-syndromal craniosynostosis, one metopic and one left coronal, underwent fronto-orbital advancement at age 3 months. At ages 19 months and 5 years, respectively, they both re-presented with headaches, decrease in head circumference percentile, and acceptable cosmetic outcome. Both had CT evidence of multiple closed cranial sutures and increased intracranial pressure (ICP) determined by monitoring. Both improved following a cranial expansion procedure. Etiologic possibilities are discussed.

Conclusion: Delayed closure of multiple sutures and resultant increased ICP may occur following correction of non-syndromal, single suture craniosynostosis. This may be more likely when the initial suture involved is contiguous with the facial sutures. Children should be followed for many years following craniosynostosis repair with cranial, neurologic and possibly fundoscopic examinations as well as head circumference measurements to detect delayed closure of cranial sutures.

Introduction

Untreated increased intracranial pressure in children with craniosynostosis can lead to blindness or cognitive impairment (Newman, 1991; Thompson et al., 1995). Release of the involved sutures and utilization of craniofacial techniques are thought to be effective in preventing these complications (Renier, 1989). Recently, however, Pollack, et al. (1996), reported that 8 of 22 children with syndromal craniosynostosis demonstrated increased intracranial pressure following an initial procedure to open the closed suture. As discussed by Humphreys (1996) in an accompanying editorial comment, this is an alarmingly high incidence and demonstrates the need for long term surveillance in this patient population. From our experience, however, increased intracranial pressure following craniosynostosis surgery is not limited to children with syndromal craniosynostosis.

Between February 1988 and December 1996, 230 children have undergone surgery for craniosynostosis at our center. The children were evaluated by a multidisciplinary craniofacial team that included a pediatric neurosurgeon, plastic surgeon, and a geneticist. Computed tomography (CT) with three-dimensional reconstruction was performed to confirm the diagnosis and to exclude associated abnormalities.

Of these 230 patients, 20 had syndromal craniosynostosis and were excluded. Of the remaining 210 children, two infants, one with metopic and one with coronal synostosis developed symptomatic, increased ICP following initial fronto-orbital advancements for single suture, non-syndromal craniosynostosis. Both infants were found to have synostosis of multiple cranial sutures. In both cases, the initial result was cosmetically acceptable and symptoms resolved following a second procedure to expand the cranium. This report discusses these cases and suggests a management protocol following fronto-orbital advancement.

Case Reports

Case 1

A term infant, product of a pregnancy complicated by gestational diabetes was noted to have trigonencephaly at birth. His examination was otherwise normal. CT confirmed metopic synostosis and patency of all other sutures (Figure 1). He was evaluated by a geneticist and his craniosynostosis was not felt to be syndromal. His development was normal. At 3 months of age he underwent a metopic synostectomy and fronto-orbital advancement of 20 mm. The surgical procedure performed in both cases was essentially the same: a variation of the "floating" fronto-orbital advancement described by Marchac and as such no attachment was left between the advanced frontal construct and the intact cranium. Postoperatively, the head was wrapped with a pressure dressing for 72 hours.

His postoperative course was uneventful and his development remained normal. At 19 months of age his mother stated that he would frequently wake up at night, hold his head, and cry. His neurologic examination was normal and he was alert and playful. His fundi were not visualized due to lack of cooperation. His cosmetic result was good with a normal shaped forehead and only mildly deficient supraorbital ridges. His head circumference, which was in the 98% at birth, decreased steadily to the 35% at 19 months of age. A CT scan demonstrated the absence of subarachnoid space, normal ventricles, digital markings, and closure of both coronal, the metopic and sagittal sutures (Figure 1) The lambdoid sutures remained patent. An intracranial pressure monitor demonstrated resting intracranial pressures of 20-30 mm Hg. A total calvarial reconstruction with fronto-orbital advancement was performed. At surgery the lambdoid sutures were open but both coronals and the sagittal suture were closed. Postoperatively his symptoms resolved and his head circumference is now in the 50%.

Case 2

A term product of an uncomplicated pregnancy was noted to have left coronal synostosis. Her examination was otherwise normal. Left coronal synostosis was

confirmed by CT and the other sutures were noted to be patent (Figure 2). She was evaluated by a geneticist and her craniosynostosis was not felt to be syndromal. At three months of age, she underwent a fronto-orbital advancement of approximately 20 mm. She did well initially, but after the first year she did not return for evaluation for 4 years. She presented again at age 5 complaining of nightly headaches. Neurologic examination was normal and there was no papilledema. She was very slightly brachycephalic, but an otherwise acceptable cosmetic result. Her head circumference, which was in the 50% at birth, had decreased to the 10%. A CT scan demonstrated no subarachnoid space, normal ventricles, and closure of all sutures except for the posterior sagittal suture (Figure 2). Intracranial pressure monitoring demonstrated resting pressures of 25-30 mm Hg with waves of pressures as high as 50 mm Hg when she was sleeping. At surgery, all sutures were closed except of the posterior one-half of the sagittal suture. A total calvarial reconstruction with fronto-orbital advancement was performed. Following surgery, she is symptom free and her head circumference has increased to the 50%.

Discussion

Increased ICP following initial suture release has been reported in children with syndromal craniosynostosis. Siddiqi, et al.(1995), found 6 cases of postoperative intracranial hypertension in a retrospective study of 107 cases of syndromic craniosynostosis. They surmised that the cranial vault in these children lacked adequate growth potential to accommodate the growing brain. All 6 children were improved by procedures that expanded the cranial vault. Pollack, et al. (1996), discovered increased ICP in 8 of 22 children with syndromal craniosynostosis. Three of these children were asymptomatic, but were found to have papilledema on routine postoperative eye examinations. Two children had ventriculomegaly and were treated by placement of a ventriculo-peritoneal shunt. The other 6 were treated with cranial expansion procedures; one of these children subsequently required a shunt. Both papers propose long term interval assessment by a craniofacial team, including periodic fundoscopic examination, in children with syndromal synostosis following

Cohen, et al. (1993), reported cranial expansion techniques for treatment of increased ICP. Of the 7 cases reported, 3 children had previously undergone craniosynostosis surgery: 1 parasagittal strip craniectomy for sagittal synostosis, and the other 2 had fronto-orbital advancements for metopic and left coronal synostosis. Time from initial surgery to diagnosis of increased ICP was 1, 3, and 3.5 years, respectively. All three children were not felt to have syndromal synostosis. In an accompanying commentary, Marchac and Renier (1993) stated that in their series of 542 patients under the age of one year with synostosis, they had re-operated on 25 (4.5%) for "recurrence" of increased ICP. They do not state how many of the cases were syndromal. They attributed most cases to technical pitfalls such as inadequate advancement. They also discuss avoidance of frontal pressure dressings to maintain the expansion.

Reddy, et al. (1990) reported 7 children with progression from single suture to multiple suture craniosynostosis. Three had not had prior craniosynostosis surgery. While they did exclude children with Crouzon's syndrome, it is not clear whether the reported children were syndromal or not.

Of the 210 non-syndromal children who underwent surgical correction of craniosynostosis at our institution over a 9 year period, 2 have developed symptomatic, postoperative increased ICP. Until recently, we did not routinely perform fundoscopic examination so children with only papilledema may have been undetected. Both children underwent a substantial advancement of the frontal construct and had acceptable cosmetic outcomes. Frontal pressure dressings were used in both cases as we believe this diminishes periorbital swelling and reduces the child's discomfort. If technical pitfalls (inadequate advancement, frontal pressure dressing) were the primary reason for the postoperative synostosis, it seems likely that post-operative synostosis would involve only the frontal sutures involved in the initial repair and not the multiple sutures demonstrated on CT and at time of the second surgery. The closure of multiple sutures suggests an abnormality of the skull base.

Early age at time of initial fronto-orbital reconstruction has been discussed as a factor that may lead to the necessity for re-operation. Wall, et al. (1994), demonstrated

a much higher re-operation rate in those infants whose initial surgery was performed between 0 and 6 months of age rather than after 6 months of age. Criteria for re-operation consisted of unacceptable fronto-orbital appearance or clinical and radiological signs of increased ICP. Unfortunately, they did not state how many children needed repeat surgery for increased ICP. In their series, re-operation was most frequently required in children with unilateral coronal synostosis, presumably for recurrence of forehead asymmetry and not increased ICP. They recommended surgery be delayed until 12-15 months of age to lower the incidence of re-operation.

This raises the question of whether surgery at 3 months of age contributed to the postoperative increased ICP in the two children described in this report. Our decision to perform surgery between 2 and 4 months of age is based on the protocol of Marchac, et al. (1994) and is predicated upon the relatively high incidence of increased ICP found in children with untreated plagiocephaly, brachycephaly, and trigonencephaly (8-50%) (Renier, et al., 1982; Thompson, et al., 1995). Prevention of neurologic injury from increased ICP is the primary goal of craniofacial surgery, and we believe this is most reliably assured by early release of the closed sutures. While reclosure of the operated suture might be an expected complication of early surgery, closure of multiple, uninvolved sutures as seen in these two cases is not.

Debate regarding the etiology of craniosynostosis has primarily concerned whether the primary abnormality is at the cranial base or in the effected calvarial suture (Moss, 1975; Babler, 1982). With syndromal craniosynostosis, especially when there is involvement of the midface, it seems clear that the primary abnormality involves the cranial base (Hoyte, 1991). Moss (1975) has proposed that craniosynostosis results from the dura acting as a "messenger" and carrying an abnormal signal from the cranial base to the calvarium. Therefore, as the underlying pathology is not corrected, it is not surprising that surgery directed towards the calvarial sutures may result in recurrent synostosis.

In contrast, the etiology of non-syndromal, single suture synostosis is usually ascribed to compressive uterine forces that act on the individual sutures (Koskinen-Moffett and Moffett, 1989). Recurrent synostosis is less likely as the surgery is directed

at the pathologic suture. However, the development of postoperative, multi-suture synostosis in these two children following repair of single suture, non-syndromal synostosis suggests an abnormality at the cranial base in these cases. Pershing, et al. (1990), have proposed that cranial vault synostosis may be transmitted to the cranial base and that this may further deform the facial and vault skeleton. This transmission may be enhanced when, as in our two cases, the initial closed suture meets a skull base suture (metopic suture-frontonasal suture; coronal suture-frontosphenoïdal suture). Thus, a proposed etiology is that an abnormality of the cranial base induced by the initial vault synostosis remains uncorrected by standard fronto-orbital advancement. The abnormal cranial base then induces synostosis of other sutures.

While the etiology of this process is unclear, it is evident that even children with single suture, non-syndromal synostosis should be followed for several years following initial correction to insure patency of the sutures. The parents and child should be questioned about the presence of headaches or other symptoms of increased intracranial pressure. Head circumference should be measured on each visit. Investigation may be warranted if the head circumference decreases several percentiles. While an assessment of the head shape is important, an acceptable cosmetic outcome, as in our cases, does not preclude postoperative craniosynostosis. As well as a detailed neurologic examination, consideration should be given to a dilated fundoscopic examination to detect asymptomatic, increased intracranial pressure.

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