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.