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Hydrocephalus in primary craniosynostosis

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Background

Ventricular dilatation in the presence of primary craniosynostosis is a unique condition with respect to pathogenesis, clinical significance, and management. We report on our personal experience with this condition over a period of 20 years.

Materials and methods

In a series of 613 patients treated for craniosynostosis and subjected to at least one detailed evaluation by ultrasound, CT scanning or MRI we found 134 to be affected with various degrees of ventricular dilatation. 32 of them ultimately turned out to be shunt-dependent.

Results

Hydrostatic hydrocephalus was rarely observed in non-syndromic craniosynostosis, and in these cases it was usually attributable to coincidental disorders including four cases of myelo-meningocele associated with coronal suture synostosis. Conversely, progressive hydrocephalus was a fairly common feature in syndromic craniosynostosis of the Crouzon or Pfeiffer type, whereas in the Apert syndrome the usual finding was that of non-progressive ventriculomegaly which, however, was also noted in some cases of Crouzon syndrome.

Conclusion

The pathogenesis of progressive hydrocephalus in Crouzon patients is a matter of still ongoing debate. A hypoplastic posterior fossa leading to crowding of its contents and extrusion of the cerebellar tonsils as well as venous outflow compromise are the main causative factors currently being discussed. The diagnosis of progressive hydrocephalus is hampered by the fact that intracranial hypertension may be attributed to both, CSF circulation disorder and premature sutural fusion. Moreover, in a synostotic skull accelerated head growth cannot be expected, classical clinical signs of intracranial hypertension may be absent, and ventricular dilatation may become evident only after decompressive cranial surgery. Therefore, careful surveillance of intracranial pressure and ventricular size in the pre- and postoperative period is a diagnostic mainstay in these cases. Ventriculo-peritoneal shunting remains the most reliable mode of treatment although surgical expansion of the posterior fossa has been suggested as an alternative option.