

Craniosynostosis and the paediatrician

Recent years have witnessed increasing interest in the treatment of craniosynostosis and there are now three centres in England that are funded on a supraregional basis by the Department of Health specifically to carry out the complex craniofacial surgery required.

Clearly, such work is highly specialised. What is the role, if any, of the general paediatrician?

The craniosynostoses cover a wide variety of congenital abnormalities which all have in common the premature fusion of one or more of the skull sutures. The resulting abnormality of growth of the cranial vault leads in all cases to an abnormal head shape that may be sufficiently distressing to demand treatment on cosmetic grounds alone. Also, as more sutures become involved, there is an increasing risk of the subsequent restriction to normal brain growth affecting the child's neurodevelopmental progress, in some cases leading to clinically obvious raised intracranial pressure.

Again, as the severity of the condition increases, so does the incidence of associated problems. Prominent among these are airway obstruction, hydrocephalus, craniovertebral anomalies, exposure of the eyes and complex abnormalities of eye movement, disorders of the facial skeleton (particularly maxillary hypoplasia), difficulties with feeding, and a general failure to thrive.

Surgical treatment of craniosynostosis used to involve no more than a simple excision of the fused sutures which, it was hoped would, if performed early enough, allow future head growth to evolve along more normal lines. Unfortunately, except for some cases of sagittal suture synostosis, such a procedure is rarely successful and most patients need a more complex operation involving not only the removal of the affected sutures but also some form of reconstruction of the skull itself. For example, children with bicoronal synostosis (which includes those with Crouzon's and Apert's syndromes) require the removal of the frontal bones, the supraorbital ridges, and part of the lateral orbital walls. These are all reshaped to provide a more pleasing contour and then replaced in a sufficiently advanced position to make up for the previous restriction of growth imposed by the premature suture fusions.

Many subsequent operations may be required, not only for further remodelling of the skull vault (some children 'grow out' of their previous reconstruction) but also for deformities of the facial skeleton. Squints and other associated abnormalities may also require surgical correction.

Clearly, the management of these complex cases requires a multidisciplinary approach, with a team made up not only of plastic surgeons, neurosurgeons, and faciomaxillary surgeons, but also ophthalmologists, otologists, psychologists, anaesthetists and radiologists, geneticists, etc. The list is obviously a long one and I have not, so far, included the specialist nurses, both on the ward and in the operating room, whose contributions are so vital.

Children with craniosynostosis should, therefore, be managed only in centres capable of fielding the appropriate expertise in a wide variety of specialties. There is absolutely no justification for treatment to be carried out on an occasional basis in units where only a few cases are likely to be seen each year. Not only is such a policy dangerous but the overall results will always be less satisfactory (and complications dealt with less expeditiously) compared with management in a specialist centre.

But to return to the general paediatricians. There can be no doubt that their role in the management of these cases is of the greatest importance. There is first the question of the early recognition and appropriate referral of children with craniosynostosis. The efficacy of some of the surgical procedures depends vitally upon their timing. For example, the treatment of sagittal synostosis is best carried out around the age of 6 weeks and the results of a simple suture craniectomy become much less impressive after the age of 3 months. Therefore, all children suspected of suffering from craniosynostosis, whether or not the skull radiographs performed at the local hospital appear to support this diagnosis, should be referred to a supraregional craniofacial centre as early as possible. These centres exist in Birmingham, in London (at The Hospital for Sick Children, Great Ormond Street), and in Oxford. Cases in Scotland and the border areas should be referred to The Southern General Hospital, Glasgow.

As I have already implied, many affected children have developmental problems and may fail to thrive. None of the specialists so far listed as members of the craniofacial team are really qualified to supervise this aspect of a child's welfare and, therefore, it is most important that general paediatric care and supervision are available at all times. Most children will spend only a comparatively small amount of time in the supraregional centre itself which, for obvious reasons, is likely to be far from home. The general day to day management of the child will, therefore, devolve upon the local paediatric (and general practitioner) services.

There is no doubt that success in the management of patients with craniosynostosis has increased in proportion to our experience and that there have been remarkable achievements in treatment since the policy of referring cases to selected specialist units has been adopted. Despite the sophistication in terms of personnel and equipment to be found in supraregional centres, however, there is no doubt that without early recognition and referral, and the subsequent management of the child and his or her family, by local paediatricians, all our specialist attention may well be in vain.

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