Tarsorrhaphies are not enough in Crouzon's Syndrome

Dear Sir,

I refer to the article on luxation of the globe in Crouzon's disease, which was published in the October 96 SMJ[1]. The authors, ophthalmologists, should be congratulated for their detailed description and management of the orbital problems encountered in this case. However, there are several points in this case report which need to be addressed.

The first point is the diagnosis of Crouzon's disease. Given the information of bilateral coronal and lambdoidal synostosis and the limited illustrations (4 photographs of the upper face and orbits, and one lateral skull X-ray), it would appear to me that this child has a cloverleaf-type cranium, as evidenced by a prominent elevated tower-shaped skull and marked convexity of the squamous temporal bones, together with ocular proptosis and mid-face hypoplasia. Whilst this form of craniosynostosis can occur in Crouzon's syndrome, it is extremely rare, and children with the severe form of this condition do not survive unless operated on within the first few months of life[2]. Milder forms of course do exist and these patients may survive to adulthood. To confirm the diagnosis, it would be helpful to be able to see the CT scans, anterior-posterior projections of skull X-rays and a picture of the whole cranium, mid and lower face. If this is correct, the treatment advocated by the authors is inadequate.

Secondly, the authors mentioned confidently that in the presence of a normal CT scan of the brain and "normal intelligence", "major surgery is not warranted". However, there is enough evidence from the data presented that this child has significant raised intracranial pressure (ICP), namely bilateral papilloedema with pale discs and optic atrophy and a diffuse copper beaten cranium as seen on lateral skull X-ray (Fig 4). The presence of papilloedema has been shown to be 100% sensitive for raised ICP in children older than 8 years and is more likely to occur in multi-suture synostosis and craniofacial syndromes as in this case[3]. In addition, ICP has been shown to be significantly greater when a diffuse copper beaten pattern was seen on skull X-ray[4]. In the face of chronically raised ICP, such children will eventually develop visual problems and reduced intelligence quotients (ID)[5]. The authors have not defined normal intelligence or adequate school performance, nor have they reported the present level of the patient's visual function and mental intelligence, given that she first presented 5 years ago in 1991. These are important facts, required to substantiate their recommended management strategy of simple tarsorrhaphies for this patient.

Thirdly, the statement that the "best time for surgery is the first year of life" should not go unchallenged. While remodelling of the cranial vault is easier in the early years of life because the skull bones are softer and more pliable, most of these children also require orbital and facial reconstruction for mid-face abnormalities when they are older, ie 4 to 5 years of age, and sometimes again later, until the adult skeleton is stabilised[6]. Orbital advancements and reconstructions, together with mid-face osteotomies and advancements, are within the armamentarium of the craniofacial surgical team, which usually comprises a neurosurgeon, maxillofacial surgeon, plastic surgeon, otorhinolaryngologist, and paediatrician. Such surgery aims to relieve raised ICP primarily, remodel the cranium, prevent and correct orbital deformities so as to preserve vision, and improve mid-face cosmesis, which has been shown to have a positive psychological impact in children[5,6]. Since a coordinated craniofacial service was started in our institute this year, 8 cases of craniosynostosis, including 4 Crouzon's, have been treated, 5 of whom had orbital osteotomies and advancements in addition to reconstructive craniotomies and suturectomies.

Tarsorrhaphies alone are inadequate treatment for this patient. The correct management should be in the realm of a craniofacial surgical team.

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Authors' Reply

We refer to the letter from Dr Keith Y C Goh. We fully agree with Dr Goh that any further reconstructive surgery falls within the realm of the craniofacial team. As we have clearly stated in our article, the patient was previously offered corrective surgery but this was refused by her parents as they were opposed to major reconstructive surgery. Also the pertinent question in this child is whether there is any definite or pressing indication for further surgery. We cannot ascertain that the child has chronic raised intracranial pressure as the CT scan of the brain was normal with no signs of cerebral atrophy or ventricular dilatation. As stated in our article, there are several causes of optic atrophy in craniofacial anomalies; raised intracranial pressure being just one of them. Also, as we have stated in our article, we have referred her to a neurosurgeon who presumably would have advised surgical intervention had she been diagnosed to have chronic raised intracranial pressure.

We mentioned in our article that as at the date of writing, "she has been well since". But because our article was written and submitted a few years ago, we could not possibly have stated the fact that we did, over a 5-year period since she first presented, monitor her visual function regularly and it appeared to be in status quo, in terms of visual acuity, pupil reflexes, optic disc appearance, Goldmann perimetry, colour vision testing and orthoptic assessment.

Finally, tarsorrhaphy is indicated in this patient for the specific purpose of preventing corneal exposure and possibly further globe luxation.

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