

PREMATURE FUSION OF CRANIAL SUTURES (CRANIOSYNOSTOSIS)

*L. MUKONGE, M.D., FRCS, DIPL TM & HGY

Summary

A personal experience in the management of craniosynostosis in 15 Zambian male children has been presented. An important finding of this study was rather late referral of the majority of these patients. The importance of early surgical intervention in the management of craniosynostosis has been stressed. It has been suggested that recent commissioning of the Paediatric Surgical Centre in Lusaka is likely to result in prompt referral of patients with surgically correctable paediatric disorders.

Introduction

Premature fusion of the cranial sutures (craniosynostosis) produces a deformed skull; causes impaired expansion of the cranial cavity and the fast growing brain eventually gives rise to an increase in the intracranial pressure. By far most common variety (80-90 per cent) is the one with premature fusion of the sagittal suture, which due to an unrestricted growth of coronal and lambdoid sutures produces a scaphocephalic type of distortion of the skull (figure 1). The two other relatively uncommon types, in descending order of frequency, are fusion of all skull sutures and of the coronal suture alone; the former produces a tall mis-shaped oxycephalic skull whilst the latter results in the formulation of a brachycephaly with short anterior fossa, flat brows and a broadened skull. This paper reports the initial experience in the management of a limited number of patients with craniosynostosis in the Paediatric Unit of University Teaching Hospital, Lusaka Zambia.

Patients and Methods

15 male Zambian children with craniosynostosis were referred to this Unit. Their ages ranged from 8 months old to 10 years; an average of 4 years. All patients attended the referring hospitals for symptoms unrelated to craniosynostosis. Two patients were the last and first borns of a mother and her daughter. The diagnosis was suspected on the appear-

ance of the skulls and later confirmed by X-rays. All 15 patients had premature fusion of the sagittal suture and all ultimately underwent ventriculograms. On the basis of an extensive neurological examination and ventriculogram findings, 8 patients (53.3 per cent) were deemed unsuitable for surgical correction. The family of 3 of the remaining 7 patients declined surgery; surgical correction was thus undertaken on 4 (26.6 per cent) patients. The type of surgical correction performed in this series consisted of linear craniectomies (figures 2 and 3) on either side of the affected sagittal suture as described by Shillito and Matson (1968); wide stripping of the adjacent periosteum with swapping of the bone edges by thin teflon were also undertaken in all cases.

Discussion

Craniosynostosis, a relatively uncommon disorder, obviously exists in Zambia. Its exact incidence, however, is very difficult to estimate. The lack of awareness among the population at large, limitation of medical manpower, difficulties in transporting a patient to a referring hospital and until recently the absence of a national centre for referral of surgical paediatric patients undoubtedly contributed to our lack of knowledge regarding the exact prevalence of various surgically correctable disorders in Zambian children. It is also not inconceivable that many physicians working in the rural areas will fail to recognise cases of craniosynostosis. No doubt recent commissioning of the Paediatric Surgical Centre in the University Teaching Hospital, Lusaka, is likely to go a long way in remedying some of these short comings.

In any discussion on craniosynostosis it is of paramount importance to emphasize the fact that the best results are obtained when a surgical correction is undertaken within the first year of life. Results of a corrective procedure leaves much to be desired when signs and symptoms of in-

*Senior Lecturer in Surgery Neonatal and Paediatric Surgical Centre, University Teaching Hospital, School of Medicine, Lusaka, Zambia.

creased intracranial pressure are present. As a matter of fact the presence of marked rise in intracranial pressure constitutes a contra-indication to surgical correction. Moreover, the major (80-85 per cent) expansion of the brain in a child normally occurs by the age of three and very little benefit is likely to be obtained by attempted surgical corrective procedures after this period. As to the surgical procedure, linear craniectomies with teflon

wrapping was found entirely satisfactory on the limited number of patients in this series. There was no mortality and no second procedure was found necessary.

Acknowledgements

My thanks are due to Miss Angela T. Zulu for typing the manuscript and Mr A. Ansary for illustrations.



Fig. 1. A patient with Sagittal Craniosynostosis demonstrating classical Scaphocephaly.

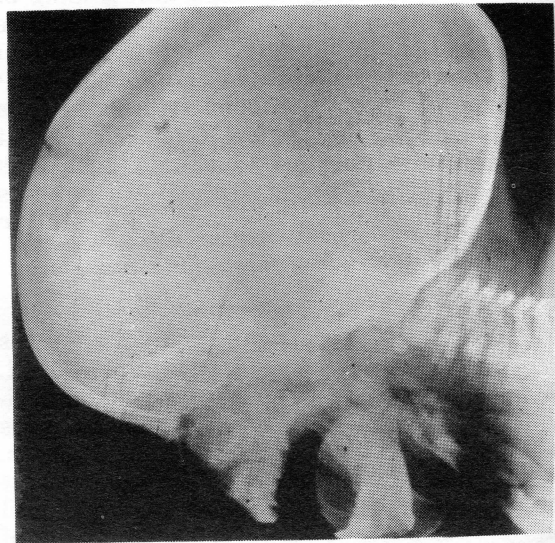


Fig. 2. Parasagittal Craniectomies (Lateral View).

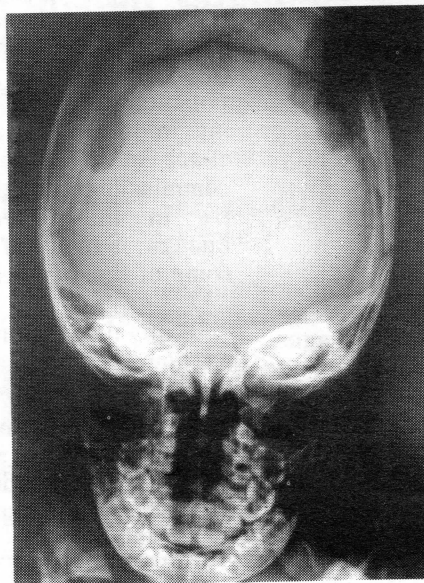


Fig. 3. Parasagittal Craniectomies (Antero-Posterior View)

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