Triplets with Craniosynostosis: A Case Report

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Introduction

Craniosynostosis is a craniofacial abnormality characterized by the premature ossification of one or more cranial sutures. In the literature, the cause of nonsyndromic craniosynostosis is attributed to a complex interaction between genetic and environmental factors. To our knowledge, there is no report in the literature of craniosynostosis occurring in each of three triplets.

Methods

We describe a case of spontaneous male triplets, each afflicted by craniosynostosis. The patients were the product of a non-consanguineous marriage and were delivered at 35 weeks' gestation by a 38-year-old mother.

Results

The dichorionic triamniotic triplets consisted of a monochorionic-diamniotic pair (A and B) and a fraternal triplet (C). Examination of A and B revealed a palpable ridge along the sagittal suture. Examination of C revealed a palpable ridge along the metopic suture. Three-dimensional computed tomography scans confirmed sagittal synostosis in A and B and metopic synostosis in C. All patients underwent endoscopic strip craniectomy at 10 weeks of life and were discharged by postoperative day two.

Conclusions

Current understanding of craniosynostosis involves an interaction between genetic and environmental factors. Increased concordance rates in monozygotic twins supports a genetic etiology while a concordance less than 100% suggests a multifactorial process which may also involve environmental and/or epigenetic influences. Involvement of the sagittal suture in the monozygotic twin pair and the metopic suture in the fraternal triplet substantiates the current understanding of craniosynostosis. Given the 100% concordance in this set of siblings, a genetic mechanism likely underlies the observed phenotype however environmental and/or epigenetic contributions cannot be excluded. Genetic analysis is in process to identify a causative mutation if any.

Table 1

Patient		Anthropomorphic Measurement (cm)								
		HC	APD	EuD	MFZD	CI	MI	RFLO	LFRO	FPDD
A	Pre-op	38	13.5	9.2	N/A	68.15	N/A	12.8	12.5	3
	Post-op	44	14.7	11.7	N/A	79.59	N/A	14.5	14.2	3
B	Pre-op	36.6	13.8	8.2	N/A	59.42	N/A	13	12.4	6
	Post-op	42.8	14.7	10.5	N/A	71.43	N/A	14.5	14.4	1
C	Pre-op	35.9	12.7	9.5	4.5	74.80	47.37	11.2	11.2	0
	Post-op	43.5	14	12	7	85.71	58.33	14	13.8	2

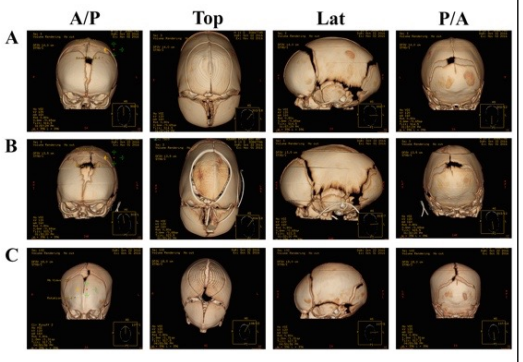
Abbreviations: HC = head circumference, APD = anterior-posterior diamteter, EuD = eurion-eurion diameter, MFZD = midfrontalzygomatic diameter, CI = cephalic index, MI = metopic index, RFLO = right frontal to left occipital frontoparietal diameter, LFRO = left frontal to right occipital frontoparietal diameter, FPDD = frontal parietal diameter difference

Anthropomorphic measurements of the patients at initial consultation and at follow-up 4 months postoperatively.

Learning Objectives

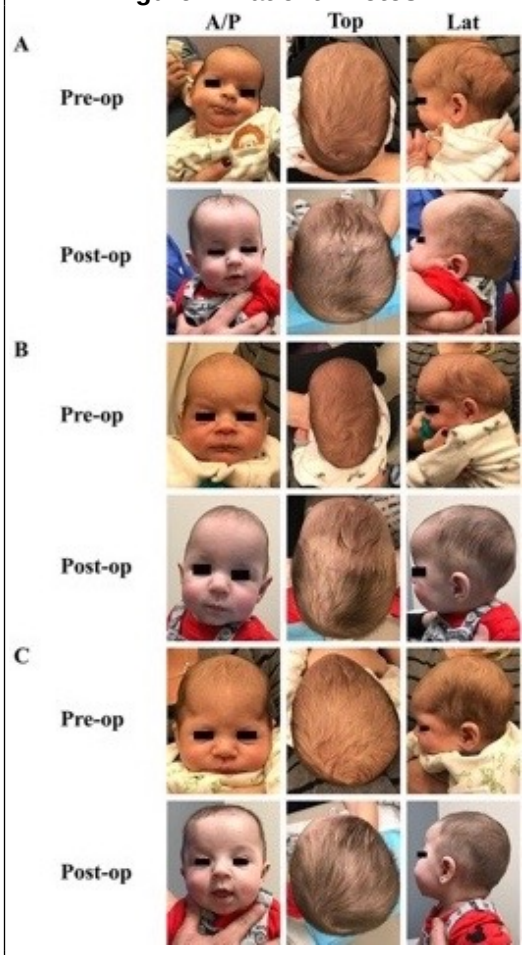
By the conclusion of this session, participants should be able to: 1) Describe the etiology of craniosynostosis, 2) Discuss the evidence for and against both genetic and environmental factors in the development of craniosynostosis

Figure 1: Preoperative 3D CT Scans



Preoperative 3D CT scans of A & B reveal a dolichocephalic calvarium with fusion of the sagittal suture (A and B). 3D CT scan of C reveals a trigonocephalic calvarium with fusion of the lower portion of the metopic suture (C).

Figure 2: Patient Photos



A and B show frontal bossing and dolichocephaly with improvement at 4 months postop. Images of C demonstrate frontal narrowing and occipital widening with significant improvement and normocephaly postop.