Symptomatic Recurrence Following the Open Surgical Repair of Nonsyndromic Craniosynostosis: A Sixteen Year Experience

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Learning Objectives

By the conclusion of this session, participants should be able to: 1) Describe the importance of symptomatic recurrence of nonsyndromic craniosynostosis and factors associated with its occurrence. 2) Realize the importance of long-term follow-up given the long time frame of recurrence.

Introduction

Recurrence following the open repair of craniosynostosis is a known event that has been described since 1948. However, the prevalence of this complication is poorly reported in the literature. The purpose of this study is to report the prevalence of symptomatic resynostosis following the repair of nonsyndromic craniosynostosis and elucidate any factors associated with this outcome over a sixteen year time period.

Methods

All nonsyndromic patients undergoing open surgical repair of craniosynostosis between 1997 and 2012 were identified. This population was then examined for symptomatic resynostosis requiring correction in addition to associated factors (such as gender, age at initial surgery, and affected cranial suture[s]). Patients requiring reoperation for other cosmetic issues (such as persistent cranial defects) were not included.

Table 1: Criteria Used to Establish Symptomatic Post-Operative Recurrence

Clinical Symptoms
headache/vomiting
developmental/school changes
irritability
Cosmetic Signs
supraorbital retrusion
severe towering
severe frontal & occipital bossing
decreasing head circumference
Radiological Evidence
recurrent synostosis
inner table erosion (thumbprinting, copper-beaten appearance)
compressed subarachnoid spaces
obscuration of gray-white matter interface
Ophthalmologic Evidence
papilledema
cranial nerve palsies

Results

257 patients with nonsyndromic craniosynostosis underwent surgical repair at a mean age of 10.9 months (2.2-82.9 months). Fourteen (5.4%) patients required further surgery at an average age of 35.4 months (16.6 -93.1 months) for a combination of clinical, cosmetic, radiographic, and ophthalmologic evidence of resynostosis. The average length of time between the initial procedure and subsequent reoperation was 29.0 months (11.6-69.5 months). Patients with bicoronal synostosis were significantly more likely to experience recurrent synostosis ($p < 0.05$). Additionally, all patients requiring reoperation had an acceptable cosmetic outcome (defined as a Whitaker Class I or II result) with an average follow-up of 6.2 years in this cohort.

Conclusions

This study of recurrence following open surgical repair of nonsyndomic craniosynostosis represents the largest to date. The prevalence in this study of 5.4% is consistent with prior reports. Given the prolonged time interval between initial surgery and recurrence, long-term surveillance for symptomatic resynostosis up to a decade post-operatively should be highly encouraged. Additionally, parents of patients with nonsyndromic bicoronal synostosis should be informed regarding the increased risk of symptomatic recurrence in this population.

Table 2: Patient Demographics

Total Number of Patients (n)	257
Male	165 (64.2%)
Female	92 (35.8%)
Average Age at Time of Surgery (months)	10.9 +/- 12.4

Table 3: Distribution of Affected Sutures

	Total	Average Age at Time of Initial Surgery
Metopic	44 (17.1%)	7.9 +/- 4.5 months
Unicoronal	51 (19.8%)	8.4 +/- 5.6 months
Bicoronal	13 (5.1%)	11.8 +/- 11.4 months
Sagittal	125 (48.6%)	11.0 +/- 12.4 months
Lambdoid	1 (0.39%)	5.8 +/- 0.0 months
Bilambdoid	3 (1.2%)	14.8 +/- 6.8 months
Multiple	20 (7.8%)	9.6 +/- 8.6 months
Total	257	10.9 +/- 12.4 months

Table 4: Choice of Initial Surgical Procedure

Procedure	Number	Percentage
BFOA	110	42.8
Pi	92	35.8
ACR	12	4.7
PCR	2	0.8
Lambdoid Synostectomy	4	1.6
TCR	33	12.8
Other	4	1.6
Total	257	100

Legend: BFOA: Bifronto-orbital advancement. ACR: Anterior 2/3 calvarial remodeling. PCR: Posterior calvarial remodeling. TCR: Total calvarial remodeling

Table 5: Demographics of Patients Experiencing Symptomatic Resynostosis

Total Number of Patients (n)	14 (5.4%)
Male	9 (64.3%)
Female	5 (35.8)
Average Age at Time of Surgery (months)	6.5 +/- 5.6
Average Age at Reoperation (months)	35.4 +/- 19.5
Average Time Span Between Surgeries (months)	29.0 +/- 16.1

Table 6: Clinical Presentation of Patients Experiencing Symptomatic Resynostosis

Criteria	Total	Percentage
Clinical	5	35.7%
Cosmetic	14	100.0%
Radiographic	13	92.9%
Ophthalmologic	1	7.1%

Combination of Criteria	Total	Percentage
1 out of 4	1	7.1%
2 out of 4	7	50.0%
3 out of 4	6	42.9%
4 out of 4	0	0.0%

Table 7: Distribution of Initially Affected Sutures in Patients Experiencing Symptomatic Resynostosis

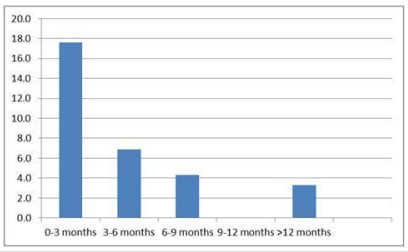
	Total	Resynostosis
Metopic	44 (17.1%)	3 (6.8%)
Unicoronal	51 (19.8%)	1 (2.0%)
Bicoronal	13 (5.1%)	4 (30.1%)*
Sagittal	125 (48.6%)	5 (4.0%)
Lambdoid	1 (0.39%)	0
Bilambdoid	3 (1.2%)	0
Multiple	20 (7.8%)	1 (5.0%)
Total	257 (100%)	14 (5.4%)

Asterisk Denotes P-Value < 0.05

Table 8: Rates of Symptomatic Resynostosis as a Function of Age at Initial Procedure

Age at Initial Procedure (mo)	Total	Resynostosis	p*
0-3 months	17	3 (17.6%)	0.07
> 3 months	240	11 (4.6%)	
0-6 months	104	9 (8.7%)	0.1
> 6 months	153	5 (3.3%)	
0-9 months	173	12 (6.9%)	0.24
> 9 months	84	2 (2.4%)	
0-12 months	196	12 (6.1%)	0.75
> 12 months	61	2 (3.3%)	

Figure 1: Rates of Resynostosis According to Age at Initial Procedure



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