An Evidence-Based Algorithm for Managing Syndromic Craniosynostosis in the Era of Posterior Vault Distraction Osteogenesis

Fares Samra MD, Jordan W Swanson MD, Brianne Mitchell MD, Andrew R Bauder BA, Ari Wes BA, Scott P Bartlett MD, Jesse A Taylor MD

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Background: Treatment approaches to syndromic craniosynostosis are variable and lack evidence. Early posterior vault distraction osteogenesis (PVDO) confers considerable cranial vault expansion and may also cause anterior vault morphologic changes that enable fronto-orbital advancement (FOA) to be delayed to a later age, with improved outcomes.

Methods: We compared treatment patterns and craniometric changes of children presenting with syndromic craniosynostosis before (2003-2008) and after (2009-2014) implementation of PVDO.

Results: 64 children with syndromic craniosynostosis presented during the study period. 40 met inclusion criteria with complete medical records and care continuity since birth: 22 prior to and 18 after implementation of PVDO (Table 1). Patients with computed tomographic studies before and after PVDO demonstrate significant reduction in frontal bossing (FNS angle decreased 125.4 to 118.8, p = 0.005.) Cranial volume increased after PVDO by a mean 21.5%, and by 28.4% in the subset of patients under 1 year of age, compared to 8.6% in patients undergoing FOA with or without posterior cranial vault remodeling (p = 0.041). Only 10 (56%) of patients who underwent initial PVDO required frontal advancement, at a mean follow-up of 4.0 years of age (range 1.5-7.5 years,) compared to 22 (100%) prior to implementation of PVDO, at a mean initial age of 1.3 years. Kaplan-Meier survival analysis indicates significant delay in need for subsequent FOA in patients who underwent early PVDO compared to early FOA or monobloc (p=0.011).

Conclusions:
Early PVDO confers improved frontal morphologic changes in children with syndromic craniosynostosis, and increased cranial volume expansion compared with those treated initially with FOA and/or posterior cranial vault remodeling. Consequently, these children are less likely to undergo FOA early in life. We have developed an evidence-based algorithm for syndromic synostosis based on these results that employs early PVDO (Figure 1).

Figure Legend:
Table 1: Demographics of study cohorts

Figure 1: Current algorithm for management of syndromic craniosynostosis

Table 1: Demographics of pre-PVDO and PVDO Cohorts				
	pre-PVDO		PVDO	
	n: 22	%	n: 18	%
Male	11	0.5	6	0.33
Female	11	0.5	12	0.66
Average age (y)	9.7		4.06	
Average follow up (y)	6.15		3.13	
Syndrome				
Apert	3	0.14	7	0.39
Crouzon	6	0.27	4	0.22
Pfeiffer	4	0.18	1	0.06
Muenke	7	0.32	2	0.11
Saethre-Chotzen	2	0.09	4	0.22