

Do Patients with Craniosynostosis Have an Increased Incidence of Auditory Neuropathy as Newborns?

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OBJECTIVE: To investigate the incidence of auditory neuropathy, abnormal auditory brainstem response (ABR) with normal otoacoustic emissions, in newborn patients with craniosynostosis as compared to published standards.

DESIGN: A retrospective review of consecutive patients with single or multiple-suture craniosynostosis who were seen between 2002 and 2009. Patients identified by the diagnostic code of craniosynostosis were divided into groups based on suture involvement. The newborn ABR screening and, if patients were referred, diagnosis from audiologic diagnostic testing were obtained from the Missouri Department of Health. Institutional review board approval was obtained.

PATIENTS: One hundred and thirty-five patients were identified. Seventy-two were excluded; 3 were listed as “missed” and 69 were not born in-state. The 63 patients included in the study were grouped by involved sutures: 2 left coronal, 7 right coronal, 2 nonsyndromic bicoronal, 3 syndromic bicoronal, 13 sagittal, 17 operative metopic, 15 nonoperative metopic, 1 pansynostosis, and 3 multiple-suture.

MAIN OUTCOME MEASURES: The newborn screening results for each patient were recorded as well as the diagnosis from audiologic diagnostics if the patient was referred.

RESULTS: Of the 63 patients, 94% (59/63) passed their ABR screening. Four were referred for diagnostic exam in both ears. Of those, one had a normal exam (right coronal) and three did not have diagnostic exams on file (right coronal, bicoronal syndromic and bicoronal non-syndromic).

CONCLUSIONS: According to the Centers for Disease Control, 1.8 percent of newborns failed their ABR screening in 2007. Of those, 37% were found to have

normal hearing on diagnostic exam. Although our study was inconclusive due to inadequate state records, it does demonstrate an increased incidence in abnormal ABR's in patients with coronal craniosynostosis. This is consistent with a recent publication that demonstrated higher incidence of abnormal ABR's in syndromic coronal craniosynostosis. If auditory abnormalities are present at birth, as our study suggests, the etiology would be unrelated to increased intracranial pressures.

KEYWORDS: Craniosynostosis, ABR, Auditory Neuropathy