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Management of Chiari I malformation in patients with complex craniosynostosis: concurrent posterior calvarial expansion and foramen magnum decompression

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Introduction: Patients with complex craniosynostosis could have cranio-cerebral disproportion and Chiari I malformation. Such patients are liable to develop syringomyelia, hydrocephalus and apnoea. These patients have been managed in number of ways: hindbrain decompression, posterior calvarial expansion and posterior calvarial distraction. Here we describe our experience with concurrent posterior calvarial expansion and foramen magnum decompression.

Method: The operative database of the Department of Paediatric Neurosurgery, Royal Liverpool "Alder Hey" Children's Hospital was searched for patients who have undergone posterior calvarial expansion (through posterior distraction or posterior remodeling) over the period of 2009- 2013. The demography, diagnoses, pre-op and post-op conditions were extracted from clinical notes and imaging studies.

Results: Eight patients were found to have undergone current posterior calvarial expansion and foramen magnum decompression. All these patients had complex craniosynostoses with additional Chiari I malformation. Five of the patients had posterior calvarial distraction and 3 patients had posterior calvarial remodeling. Four of the patients had Pfeiffer's syndrome, 2 patients had Crouzon's syndrome, 1 patient had Apert's syndrome and 1 patient had multiple cranial synostosis. There were 2 males and 6 females. The ages of the patients ranged from 6 months to 15.5 years (mean: 3 years and 1 month; median 1 year and 2 months). The follow-up ranged from from 1 month to 3 years (mean: 11 months; median: 3 months). The surgical technique and the outcome will be discussed in the presentation.

Conclusion: Concurrent posterior calvarial expansion and foramen magnum decompression for complex craniosynostosis patients with cephalo-cerebral disproportion and Chiari I malformation is a safe procedure. None of our patients subsequently developed syringomyelia however, patients would need longer follow-up to ascertain the long-term benefit of the combined procedure.

Competing interests: None