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A unique case of isolated lambdoid synostosis presenting with intracranial hypertension: an examination of aetiology and management by posterior skull vault expansion

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Introduction: Lambdoid synostosis as an isolated condition is uncommon, with an estimated incidence of approximately 2% (Shillito and Matson 1968). Unlike other single suture synostoses, in which intracranial hypetension (ICP) has a reported prevalence of 15-25% of cases (Renier et al 1982) there are no published case reports or series to identify the frequency that patients with isolated lambdoid synostosis are affected. We present a unique case of a child presenting with lambdoid synostosis who on investigation was found to have intracranial hypertension. We propose that the anomalous pattern of venous drainage identified in this child provides a likely explanation for presentation and that successful treatment can be effected by techniques to expand the skull using either static expansion or dynamic distraction techniques.

Methods: A single case report with a review of the literature.

Results: 3D reconstruction of planar CT imaging demonstrates the unusual phenotype associated with this uncommon single suture synostosis. Examination confirms eye field changes consistent with elevated intracranial pressures and this is supported by intracranial pressure monitoring showing both an abnormal median ICP and pathological wave form changes. Both CT venography and MR venography identified anomalous venous drainage and excluded other causes of presentation with symptoms consistent with and findings of raised pressure. Treatment resulted in successful treatment evidenced by restoration of normal eye examination and behaviour.

Conclusions: Intracranial hypertension can be associated with this uncommon form of single suture synostosis and that anomalous venous drainage should be considered a possible underlying aetiology. Successful treatment can be with skull vault expansion techniques.