

## Fourth Ventriculoceles with Extracranial Extension: One Case Report

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### SUMMARY

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Encephaloceles involving only the herniated fourth ventricle are exceedingly rare. We report a case of such a cranial malformation and review the literature regarding these malformations and their potential embryological derailment.

### INTRODUCTION

An encephalocele is a developmental abnormality in which part of the CNS herniates through a cranial defect. Occipital encephaloceles are the most common forms of encephaloceles in the Western Hemisphere (Bui et al., 2007). These entities often include neural tissue within the herniated sac. A form fruste of such encephaloceles is the rare fourth ventriculocele where the herniated sac is simply an enlarged outpocketing of the fourth ventricle that does not include neural tissue (Merlob, 1985). Fourth ventriculoceles, albeit rare, are most commonly associated with Dandy-Walker Syndrome and occasionally the Chiari II malformation. Dandy-Walker syndrome is characterized by the enlargement of the fourth ventricle with hypoplasia or complete absence of the cerebellar vermis and cyst formation within the skull. Congenital atresia of the foramina of Luschka and Magendie may be a causative factor of these anomalies (Huang et al., 2001; Oblu et al., 1976).