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Correspondence

Benign External Hydrocephalus in Children: A Condition in Search of an Etiology

Dear Editor,

We read with great interest the recently published article by Zahl et al.[1] We applaud the authors for studying the clinicoradiological and demographic profile of children with benign external hydrocephalus (BES). However, there are certain points we would like to highlight.

In the present study, 86.4% children were boys. Selectively affected male gender indicates a genetic etiology, probably an X-linked inheritance pattern. Assessment of head circumference in three generations would guide the ascertain of a genetic etiology. Certain genetic syndromes such as glutaric aciduria type 1 and Sotos syndrome also present with macrocephaly and BES, so information about other affected family members and a history of parental consanguinity would have been helpful.

BES is a benign condition and most of the patients do not require surgical management. Surprisingly in the author's cohort, 80% children had ventriculomegaly and 25% required ventriculoperitoneal shunt.

Norway is situated in the northern part of the globe and has long winter nights, limiting the anount of sunlight and increasing the vulnerability to vitamin D deficiency. The prevalence of vitamin D deficiency in Norway[2] and rest of the world is very high.[3] Clinicoradiological features of vitamin D deficiency have many similarities with BES. Children with vitamin D deficiency or rickets may present with gross motor delay, hypotonia, wrist widening, delayed closure of fontanels, tense fontanels or benign intracranial hypertension, frontal bossing, and large head. Macrocephaly in rickets is usually because of parietal and frontal bossing and a neuroimaging classically shows benign enlargement of subarachnoid space and mild ventriculomegaly.[4] Assessment of vitamin D status and its correlation with head circumference and neuroimaging characteristics might have given a clue to the association or causation.

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