prognosis. However, the early use of Miglustat, prior to presentation of the characteristic neurologic signs of the disease, may lead to improvement in life quality.

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HYPERVITAMINOSIS A - IS IT STILL A CAUSE OF IDIOPATHIC INTRACRANIAL HYPERTENSION IN CHILDHOOD?

T. Jacinto¹, C. Sousa¹, I. Sobreira¹, A.T. Raposo¹, L. Lima², C. Pereira Duarte¹

¹*Pediatrics, ²Ophthalmology, Hospital Divino Espírito Santo HDES - Ponta Delgada, Ponta Delgada, Portugal*

Idiopathic intracranial hypertension (IIH) in prepubertal children is rare and different in presentation from the adult, occurring with no apparent preference for sex or weight. Clinically, strabismus with VI nerve palsy and stiff neck are more frequent, and secondary causes more common. Vitamin A toxicity has been described mainly in adolescents treated with topic retinoids for acne, rather than oral intake as dietary supplement.

We present the case of a previously healthy 9-year-old male, with strabismus, ptosis of the left eye and diplopia about a month after a cold with occipital headache and vomiting (medications included supplements containing vitamin A). Fundoscopy revealed bilateral papilledema with enlargement of blind spot and endotropium of the left eye. Brain CT was normal. Lumbar puncture (LP) at admission revealed opening pressure of 500 mmH₂O, with normal CSF results. Admitting IIH, he was started on acetazolamide, with good clinical results. Regular eye exams revealed visual acuity 8/10 bilaterally and stabilization of papilledema. Elevated serum levels of vitamin A were the only positive findings. He was discharged after two weeks, asymptomatic; completed two months of acetazolamide, then repeated LP with opening pressure of 200mm H₂O.

Hypervitaminosis A has been associated with IIH in children, although its causative role is yet to be determined. A limiting factor is that IIH does not correlate directly with increased vitamin A serum levels, but with CSF levels, which are not tested routinely. Nevertheless, vitamin intake in children should be started only in cases with formal indication and monitored carefully.

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RECURRENT MENINGITIS AS A COMPLICATION OF SKULL BASE MENINGOCELE

I. Sedighi¹, B. Borzouei², S.T. Sadrosadat², M. Ghodsi³

¹*Department of Pediatrics, Hamedan University of Medical Sciences/ Besat Hospital, ²Hamedan University of Medical Sciences, Hamadan,*

³*Department of Neurosurgery, Tehran University of Medical Sciences/ Shariati Hospital, Tehran, Iran*

Background: Malformations of the skull base are rare anomalies, but can cause severe complications such as meningitis. They are often detected not until after a history of recurrent meningitis.

Methods: A 6-year-old boy was admitted in our hospital due to change in mental status, fever, and vomiting. His past medical history revealed two episodes of bacterial meningitis in 12 and 22 months of age. His parents were cousins and had lost their 3 sons before this boy was born. One of them had died due to respiratory distress 15 days after birth, another one had been born dead, and the last one had died due to meningitis in 3 years of age.

Results: He received Vancomycin and Ceftriaxone upon diagnosis of recurrent meningitis. Due to seizure, intravenous Phenytoin was started. Complementary diagnostic evaluations included computed tomography (CT) and magnetic resonance imaging without any abnormal finding. Streptococcus pneumoniae was isolated from CSF culture. Serum immunoglobulins, complement C3, C4, and CH50 levels, NBT slide test, and flow cytometry (CD3, CD4, CD8, CD19, and CD16+56) were normal. After ruling out primary immunodeficiency, we repeated CT and this time a skull base bony defect behind sphenoidal sinus with a meningocele was noted. Endonasal surgery confirmed this bony defect and closure was accomplished. No further meningitis has been observed.

Conclusions: Congenital skull base defects may be difficult to detect. All children with recurrent meningitis should be investigated for these defects. Early recognition of skull base defects is important to avoid delay of definitive surgical management.