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## Giant Occipital Meningocele as a Presenting Feature of Dandy-Walker Syndrome

A 10-month-old female infant, born of nonconsanguineous parents, full-term normal delivery presented with a large swelling over the occipital region since birth. The child could not hold the head. She was playful and recognized parents but could not sit or stand on her own. There were no other congenital abnormalities or family history of neural tube defects. On examination, there was frontal bossing, hypertelorism and mild 'sun-setting' sign. The anterior fontanelle was lax. Local examination revealed a large, pear shaped, pedunculated swelling in the occipital region. It was brilliantly transilluminant,

nontender, fluctuant and non-pulsatile. There was no bruit. The physical and mental milestones were delayed. Neurological examination showed no gross motor or sensory deficit. Magnetic resonance (MR) imaging of the brain showed features suggestive of a Dandy-Walker malformation. The posterior fossa cyst formed a giant occipital meningocele with few incomplete septae within. A primary repair of the meningocele was performed. Cranioplasty was not performed. The child developed fulminant bacterial meningitis on the 10th postoperative day and later succumbed.

In 1887, Sutton was the first to describe the association of Dandy-Walker syndrome with occipital meningoencephalocystocele on autopsy (1). The incidence varies between 11-16% in reported series. It might simply

be an expression of increased intracystic pressure compensating for the raised intracranial pressure during fetal life. The elasticity and the compliance of the meningocele sac acts like a vent for CSF and is probably life saving preventing dangerous elevation of intracranial pressure. The grotesque external appearance is due to the centrifugal expansion of the occipital meningocele. The presence of incomplete internal septae caused a 'trap door' effect and consequent further expansion of the meningocele sac. The difficulties of feeding, handling and ordinary care are great as well as the psychic trauma to parents in the presence of disfiguring and unconcealable mass, that earliest possible excision is highly desirable. The anterior fontanelle was lax and the sutures were wide open despite ventriculomegaly. It is a matter of speculation whether an initial cystoperitoneal shunt would have appreciably reduced the giant size of the meningocele. There is a increased likelihood of developing decubital ulcers due to restricted head movement caused by the occipital lesion in addition to the high probability of shunt infection and frequent

shunt malfunction(2). It is also postulated that resection of the cyst wall rarely gives permanent relief since there is a generalized dysgenesis(3). A ventriculo-peritoneal shunt may be inserted for symptomatic progressive hydrocephalus and/or associated aqueductal stenosis.

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### **Cost of Syrup versus Capsule Form of Vitamin A**

We read the article entitled "Cost of syrup versus capsule form of vitamin A supplementation"(1) with great interest and found it informative. The authors have compared the cost of supplementing vitamin A in the form of syrup versus capsules in the research manuscript but in the conclusion authors have interestingly recommended use of applicaps for vitamin A administration. However, the existing scientific evidence indicates that

supplementation of vitamin A should be continued in the form of syrup as presently being done in India under the "National Program of Prevention of Nutritional Blindness due to vitamin A Deficiency" because of the following reasons.

- (i) The present study was descriptive in nature and no mention has been made of total number of Anganwadi workers and Auxillary Nurse Midwives (ANMs) included in the study. The functionaries interviewed for distribution of vitamin A were from a research project area who may not be true representatives of