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HEADACHE DISORDERS

SLIT VENTRICLE SYNDROME AND HEADACHE

The management of slit ventricle syndrome and shunt-related headache is reviewed by researchers at the Barrow Neurological Institute, St Joseph's Hospital, Phoenix, AZ. Five syndromes of shunt-related headache are described: 1) *Intracranial hypotension* with headache that develops later in the day and with the erect position and is relieved by lying down. Treatment involves replacement of the valve mechanism and incorporating a device that retards CSF siphoning (DRS). 2) *Intermittent proximal obstruction* with sudden increase in intracranial pressure (ICP) with activity. As ICP increases, the headache worsens until the ventricular catheter reopens and the pressure is normalized. In chronically shunted patients, proximal shunt failure is most commonly caused by over-drainage of CSF and collapse of the ventricular walls around the catheter. The valve and DRS system are replaced. 3) *Normal volume hydrocephalus* with symptoms of ICP and headaches developing in the morning and progressing, a common problem with congenital hydrocephalus. All have elevated venous sinus pressure. Older patients have symptoms of pseudotumor cerebri. Treatment involves lumboperitoneal or cisternal shunt. 4) *ICP with working shunt* associated with Chiari I malformation and hindbrain herniation. Some cases are associated with craniofacial abnormalities and cephalocranial disproportion. 5) *Migraine* headaches complicating shunted hydrocephalus may require ICP monitoring to exclude slit ventricle syndrome as the cause of headache. The author estimates that one third of his patients with shunted hydrocephalus followed more than 5 years will have chronic headache disorder requiring intervention. In 20% of cases of shunt-related headache the ventricles do not enlarge with shunt failure and the headaches are associated with normal volume hydrocephalus. (Rekate HL. Shunt-related headaches: the slit ventricle syndromes. **Childs Nerv Syst** April 2008;24:423-430). (Dr HL Rekate, Neuroscience Publications, Barrow Neurological Institute, 350 West Thomas Road,

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COMMENT. The cause of headache in shunted hydrocephalus is often not identified with a CT scan. Headache relieved by lying down may point to over-drainage of CSF, intracranial hypotension and slit ventricle syndrome. Headache exacerbated by exercise points to an intermittent obstruction of CSF flow. If the ventricles do not expand with shunt failure, a normal volume hydrocephalus with increased ICP is suspected. All require immediate neurosurgical intervention. Seizures are an additional complication of slit ventricle syndrome. The development of spike and sharp wave EEG abnormality following a shunting operation for hydrocephalus may indicate shunt malfunction and over-drainage of CSF. (Saukkonen A et al. **Child's Nerv Syst** 1988;4:344-347; **Ped Neur Briefs** May 1989).

SEIZURE DISORDERS

GENETICS OF FEBRILE SEIZURES AND EPILEPSY (GEFS+)

Mutations in 3 genes *SCN1A, SCN1B* and *GABRG2* have been shown to cause GEFS+ in families of various ethnic origins. The occurrence of mutations in these genes in 19 families of Scandinavian origin with a history of GEFS+ was studied at Ulleval University Hospital, Oslo, Norway, and centers in Denmark. Families were identified from population-based twin registries in Denmark and Norway. One mutation in *SCN1A* was identified in a Danish family with phenotypes consistent with GEFS+. The mutation was not found in healthy and unrelated controls. No mutations were found in any of the other families. (Selmer KK, Egeland T, Solaas MH et al. Genetic screening of Scandinavian families with febrile seizures and epilepsy or GEFS+. **Acta Neurol Scand** April 2008;117:289-292). (Respond: Dr Keja K Selmer, Ulleval University Hospital, Kirkeveien 166, 0407 Oslo, Norway).

COMMENT. GEFS+ is an autosomal dominant disorder characterized by multiple febrile seizures persisting beyond age 5 years and complicated by afebrile seizures of absence, myoclonic or atonic types. Seizures cease in mid-childhood. (Scheffer IE et al. **Brain** 1997;120:479-490; Idem. **Epilepsia** 2005;46:41-47). Genes on chromosomes 2q24 and 19q13 encode subunits of the voltage-gated sodium ion channels, while the gene on 5q31 codes for the g-subunit of the g-aminobutyric acid (GABA) receptor. The genes responsible for GEFS+ show considerable heterogeneity and variable expressivity. GEFS+ is an evolving composite of many syndromes, with shared genetic susceptibility. (Nordli DR Jr. **Epilepsia** 2005;46(Suppl 9):48-56). While the definition of GEFS+ is continually changing and probably involves many genes, the common denominator is the association with febrile seizures.

Failure of replication of epilepsy gene associations is discussed by researchers from Columbia University Medical Center, and New York State Psychiatric Institute, New York, NY. (Pal DK, Strug LJ, Greenberg DA. **Epilepsia** 2008;49:386-392). Over 50 genetic associations with various idiopathic epilepsy syndromes are reported but most have not been replicated. Genetic heterogeneity is a confounder in population-based studies, in both