# PAROXYSMAL DISORDERS

## MANAGEMENT OF REFLEX ANOXIC SEIZURES

Investigators at the Roald Dahl EEG Unit, Alder Hey Children's NHS Foundation, Liverpool, UK, review the definition, pathophysiology, clinical presentation, and management of reflex anoxic seizures (RAS) in children. Reflex asystolic syncope is proposed as the most appropriate alternative term; other terms for RAS include pallid breath-holding attacks and vasovagal syncope. A sudden injury or fright precipitates an acute loss of consciousness with opisthotonus, cyanosis, and clonic movements, resembling a short generalized tonic-clonic seizure. The underlying pathophysiology is a vagal-induced cardiac asystole with resultant cerebral hypoperfusion and consequent anoxia. EEG shows diffuse, high-amplitude slow wave activity during 10-15 sec of asystole, replaced by diffuse attenuated (low-amplitude) activity. After a further 5-20 sec, the vagal discharge stops, and the EEG shows a brief diffuse high-amplitude slow wave activity before returning to normal.

The differential diagnosis is an anoxic epileptic seizure, cyanotic breath-holding spell, or cardiogenic syncope. Anoxic epileptic seizures in which an initial anoxic seizure provokes a true epileptic seizure are very rare. Examples of cardiogenic syncope include prolonged QT syndromes (autosomal dominant Romano-Ward and autosomal recessive Jervell and Lange-Nielsen syndromes).

A careful history from an eyewitness of the onset of the attack is most important in diagnosis. Pharmacological treatment (e.g. atropine, scopolamine patch) may be helpful but carries a risk of adverse effects. Cardiac pacing is the only definitive treatment, reserved for frequent, most severe episodes. (Iyer A, Appleton R. Management of reflex anoxic seizures in children. **Arch Dis Child** 2013 Sep;98(9):714-7). (Response: Dr Richard Appleton. E-mail: Richard.appleton@alderhay.nhs.uk).

COMMENT. Reflex anoxic seizure (or reflex asystolic syncope) is important in the differential diagnosis of non-epileptic paroxysmal disorders in infants and pre-school-aged children.

**Home video recordings of epileptic seizures induced by syncope** are reported in 3 patients with "anoxic epileptic seizures." (Stephenson J, et al. **Epileptic Disord** 2004 Mar;6(1):15-9). These authors also reported a cohort of 9 children with syncope in which an initial anoxic seizure provoked a true epileptic seizure (Horrocks IA, Nechay A, Stephenson JBP, et al. **Arch Dis Child** 2005 Dec;90(12):1283-7). Contrary to these reports, the above author (Appleton R), a pediatric neurologist for over 22 years, avers he has never seen a child with epilepsy as a complication of RAS.

## ELECTROCLINICAL SUBTYPES OF CINGULATE EPILEPSY

Investigators from the Cleveland Clinic, and University of Texas Southwestern Medical Center, studied consecutive cases of cingulate gyrus epilepsy identified retrospectively from their epilepsy databases from 1992 to 2009. Of 14 patients with

cingulate epilepsy confirmed by MRI and response to lesionectomy, 4 with lesions in the posterior cingulate location had electroclinical findings suggestive of a temporal origin of the epilepsy. Of 10 anterior cingulate cases, 6 in a typical (Bancaud) group had hypermotor/hyperkinetic seizures, rarely generalized, with fear, laughter, or severe interictal personality changes, and 4 were atypical, with simple motor seizures, frequently generalized, and a less favorable long-term surgical outcome. All atypical cases were associated with an underlying infiltrative astrocytoma. Posterior cingulate gyrus epilepsy is regarded as a pseudotemporal epilepsy. (Alkawadri R, So NK, Van Ness PC, Alexopoulos AV. Cingulate epilepsy: Report of 3 electroclinical subtypes with surgical outcomes. **JAMA Neurol** 2013 Aug 1;70(8):995-1002). (Response: Rafeed Alkawadri MD, Yale University School of Medicine, 15 York Street, LC17148, New Haven, CT 06510. E-mail: mhdrafeed.alkawadri@yale.edu).

COMMENT. Surface EEG is often inaccurate in localizing a deep-seated epileptogenic zone in a patient with cingulate epilepsy. Symptoms of cingulate epilepsy are heterogeneous and dependent on an anterior or posterior localization of the lesion. Anterior lesions are associated with hyperkinetic behavior, cycling and running and gelastic seizures, expressed by mirthless laughter. Posterior cingulate seizures resemble temporal lobe epilepsy. This report emphasizes the importance of an MRI-identifiable lesion in the diagnosis of cingulate epilepsy.

## INTRACRANIAL PRESSURE DISORDERS

## SPONTANEOUS INTRACRANIAL HYPOTENSION

Investigators at the Department of Neurosurgery, Cedars-Sinai Medical Center, Los Angeles, CA, evaluated 24 children (18 girls, 6 boys) with spontaneous intracranial hypotension, seen 2001-2012. Onset of symptoms was at mean age of 14.3 years (range, 2-19 years). The majority (23 patients) presented with orthostatic headaches, mainly occipital, and one with a non-positional headache. Additional symptoms included nausea, neck pain, dizziness, and hearing abnormalities. Precipitating headache factors included lifting, dance class, handstands, and football. Spinal MRI demonstrated a CSF leak in 12 (50%) patients, usually thoracic in location, and spinal meningeal diverticula without a leak in 10 (42%). Underlying connective tissue disorders in 13 patients (54%) included Marfan syndrome in 3, Marfan-like syndrome in 4, Ehlers-Danlos syndrome in 2, and hypomelanosis of Ito in 1. Dural ectasia with multiple meningeal diverticula were found in all 3 patients with Marfan syndrome.

Treatment consisted of bed rest and hydration, epidural blood patches in 23, with permanent resolution in 9 patients (39%). Injections of fibrin glue directed at the CSF leak were successful in 2 patients (25%). Surgical treatment had a good result in 10 patients (91%), with permanent resolution of symptoms. Acetazolamide for rebound high intracranial pressure headache was required in 5 patients. Overall, outcome was good in 22 patients (92%) and poor in 2 (8%). (Schievink WI, Maya MM, Louy C, Moser FG, Sloninsky L. Spontaneous intracranial hypotension in childhood and adolescence. **J Pediatr** 2013 Aug;163(2):504-10). (Response and reprints: Dr Wouter I Schievink. E-mail: SchievinkW@cshs.org).