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SEIZURE DISORDERS

SYNCOPE-LIKE SEIZURES IN PANAYIOTOPOULOS SYNDROME

Researchers at St Thomas' Hospital, London, and Leeds General Infirmary, UK studied prospectively the clinical features of syncope-like epileptic seizures (SLES) and their frequency in children aged 1 – 15 years with Panayiotopoulos syndrome (PS) who were referred for an EEG. PS was defined by the occurrence of at least one autonomic seizure (AS) in a neurodevelopmentally normal child and one EEG with focal spikes. PS was diagnosed in 33 of 394 consecutive children with at least one AS (8.4%). SLES occurred at least once in 17 of 33 PS cases (51.5%); 12 had SLES in all AS. Overall, 53 of 74 AS presented with SLES (71.6%); 25 occurred during the course of an AS (AS + SLES) and 28 were pure SLES without other autonomic or convulsive manifestations. Concurrent autonomic symptoms during AS + SLES included emesis, incontinence, mydriasis, miosis, and cardiorespiratory abnormalities. Pure SLES presented with sudden collapse without premonitory symptoms, while standing, sitting, lying down, or asleep; flaccidity and unresponsiveness persisted for several minutes, up to 1 hour. Recovery was spontaneous, rapid and complete. All children with follow-up >2 years are seizure-free.

A typical case report of PS describes a 5-year-old boy who at age 13 months, woke, vomited profusely, became unresponsive and floppy, and later that night, woke again, vomited, and collapsed. He remained flaccid and unresponsive for 1 hour, and had dilated nonreactive pupils. At age 20 months, he collapsed and was unresponsive and flaccid for 10 min. His last seizure occurred at age 28 months, he fell and remained unresponsive and flaccid for 20 min; cardiology exam was normal. (Koutroumanidis M, Ferrie CD, Valeta T, Sanders S, Michael M, Panayiotopoulos CP. Syncope-like epileptic seizures in Panayiotopoulos syndrome. *Neurology* 2012 Jul 31;79(5):463-7). (Response and reprints: Dr Koutroumanidis. E-mail: michael.koutroumanidis@gstt.nhs.uk).

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COMMENT. A consensus view of Panayiotopoulos syndrome (PS) determined by an international consortium concludes that PS is a common idiopathic, benign seizure disorder of childhood that should be classified as an autonomic epilepsy, rather than an occipital epilepsy (Ferrie C et al. **Dev Med Child Neurol** 2006 Mar;48(3):236-40). Recognized by the ILAE as a distinct clinical entity within the spectrum of benign focal epilepsies of childhood, PS is manifested electrographically with multiple interictal spikes (Kokkinos V et al. **Clin Neurophysiol** 2010 Jun;121(6):859-69).

Autonomic epilepsy is frequently misdiagnosed and treated as atypical migraine, syncope, or abdominal disorder, leading to unnecessary and sometimes invasive investigations. The present UK study of PS concludes that in atypical cases of suspected syncope in children having focal spikes in the EEG, a diagnosis of syncope-like epileptic seizures in PS should be considered.

DEVELOPMENTAL & AUTISM SCREENING IN EPILEPSY UNIT

Researchers at the Epilepsy Center, Ann & Robert H Lurie Children's Hospital of Chicago assessed the yield of routine screening for neurodevelopmental delay and autism in all patients, 5 years of age and younger, seen in the monitoring unit or ketogenic diet clinic. The Ages and Stages Questionnaire, a parent completed form selected for development screening, addresses communication, gross and fine motor, problem solving, and personal-social. The Modified Checklist for Autism in Toddlers (mCHAT) was used for children up to age 4 years; and the Social Communication Questionnaire (SCQ) in children 4 years and older. Of 65 who participated, 49 (75%) were established epilepsy patients, and 16 (25%) were screened in a new-onset epilepsy clinic. Mean age at screening was 2.5 years (range 2 months to 5 years), and 38 (58%) were boys. Seizure frequency at time of screening was seizure free (N=14), <1/month (N=2), monthly (N=4), weekly (N=5), and daily (N=30).

Developmental screening was positive/delayed in 47 (72%), negative in 10 (15%), and borderline in 8 (12%). Established epilepsy patients were more likely to be developmentally delayed and to test positive than new-onset patients ($p=0.0001$). Among 49 screened for autism, 24 (49%) scored positive. Of 12 with normal or borderline developmental screening, none screened positive for autism, whereas 24 of 37 (65%) of those with developmental delay also scored positive for autism ($p<0.0001$). The prevalence of positive autism screening correlated with the degree and type of developmental delay. Delays in the social-personal domain were more likely than those in gross and fine motor to discriminate between positive and negative autism screening. Positive autism screening in 20 children were better explained by the underlying developmental delays. New concerns identified through screening prompted referrals for further evaluations in 16 patients; these included physical therapy, speech therapy, early intervention mental health and school services, and psychiatry. The yield of routine screening was sufficiently high to support developmental and autism screening in all children attending an epilepsy center. (Fisher B, Dezort C, Nordli DR, Berg AT. Routine developmental and autism screening in an epilepsy care setting. **Epilepsy Behav** 2012 Aug;24(4):488-92). (Respond: Dr Anne T Berg. E-mail: atberg@luriechildrens.org).

COMMENT. The authors conclude that comorbid developmental and behavioral