

HEADACHE DISORDERS

HEADACHE WITH FOCAL NEUROLOGICAL SIGNS

Investigators at Robert Debre Hospital and Paris Diderot University, Paris, France, and Basildon University Hospital, Essex, UK, conducted a prospective study of 101 children aged 6-18 years presenting to the ED of a tertiary hospital with moderate to severe headache and focal neurologic deficit with focal brain disturbance. Children with a history of trauma, fever, or neurosurgical intervention were excluded. After one-year follow-up, the final diagnosis was primary headache in 66% cases (94% migraine with aura), and 34% received a final diagnosis of secondary headache (76.5% with focal epilepsy). Children with bilateral localization of pain had a higher likelihood of having secondary headache ($p < 0.001$).

In the secondary headache group, 26 children had a seizure: 14 had benign childhood occipital epilepsy (Panayiotopoulos syndrome), 5 had temporal lobe epilepsy (2 with dysembryoplastic neuroepithelial tumor), 4 had epilepsy with centrotemporal spikes, and 2 had occipital and 1, parietal lobe epilepsy. Other secondary headaches were associated with arterial ischemic stroke (2 cases), cerebral venous sinus thrombosis (2), arteriovenous malformation (2), and 2 with intracranial neoplasm (1 astrocytoma, 1 medulloblastoma). The headache lasted longer than 24 hours in 15 of the primary group and 8 of the secondary headache group. Neurologic deficit was no longer present by 1 hour after initial onset in 72 patients (71.3%).

MRI disclosed brain disorders related to the acute headache in 10 of 78 children (12.8%). In patients suspected of having epilepsy, an awake and asleep EEG was performed during the ED admission in 15 patients (14.9%) of whom 9 had epilepsy, and within 1 week in 47 patients (46.5%), of whom 26 were considered to have experienced a seizure. (Massano D, Jullian S, Kanagarajah L, et al. Headache with focal neurologic signs in children at the emergency department. *J Pediatr* 2014 Aug;165(2):376-82).

COMMENTARY. The 2nd edition of the International Classification of Headache Disorders (2004) (ICHD-2) separates headache into 4 categories of primary headache (migraine, tension-type headache, cluster headache, other trigeminal autonomic cephalgias), and other unclassified primary headaches, and 8 categories of secondary headache [1][2]. Secondary causes of headache are considered in the following situations [3]:

- escalating frequency and/or severity of headache
- change of frequency and severity of headache
- headache associated with fever
- headache accompanied by seizures

Comorbidity of headache and epilepsy. The ICHD-2 defines 3 kinds of association of headache and epileptic seizure: (1) migraine-triggered seizure or ictal epileptic headache, (2) hemispheric epileptic headache (very rare variant of epileptic headache), and (3) pre-ictal headache [4]. Headache and epilepsy are a common comorbidity in childhood and occur mostly in children older than 10 years with idiopathic epilepsy [5].

Among 100 children with chronic recurrent headaches treated in neurology practice, 15% had a history of epileptic seizures. EEG showed epileptiform discharges in 18%. Headaches were diagnosed as migraine in 42% and tension headaches in 18%. A trial of antiepileptic medication controlled headaches in 77% of 30 children with migraine, but a positive response was unrelated to an abnormal EEG. Beneficial response rates were 61% and 88% in 13 with abnormal and 17 with normal EEGs, respectively. Migraine patients with normal or abnormal EEGs were benefited. An abnormal EEG and response to AED are insufficient criteria for a diagnosis of epileptic headache [6].

References.

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2. Lipton RB, et al. Neurology. 2004 Aug 10;63(3):427-35.
3. Winner P, et al. Headache in Children and Adolescents. 2nd ed. Hamilton, Ontario: BC Decker; 2008:344.
4. Clanchetti C, et al. Seizure. 2013 Nov;22(9):679-85.
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MUSCLE DISORDERS

CEREBRAL ABNORMALITIES IN DUCHENNE MD

Investigators from Leiden University, the Netherlands, used quantitative MR imaging to study brain microstructure in 30 patients with Duchenne muscular dystrophy (DMD) and 22 age-matched controls (age 8-18 years). DMD patients had smaller total brain volume, lower white matter fractional anisotropy, and higher white matter mean and radial diffusivity than healthy controls. DMD patients also performed worse on neuropsychological examination. Subgroup analyses showed that isoform expression DMD_Dp140 subjects contributed most to the gray matter volume differences and performed worse on information processing. Dp 140 dystrophin isoform has an important role in cerebral development. (Doorenweerd N, Straathof CS, Dumas EM, et al. Reduced cerebral gray matter and altered white matter in boys with Duchenne muscular dystrophy. *Ann Neurol* 2014 Jul 10).

COMMENTARY. In addition to cardiomyopathy, DMD patients should be tested for central nervous system disorders. These include cognitive impairments (1 SD below normal) involving verbal short-term memory, visuospatial long-term memory, and verbal fluency, and higher incidence of autism, ADHD, ODD, and learning disorders [1].

References.

1. Hendriksen JG, et al. J Child Neurol. 2008 May;23(5):477-81.