Hemiconvulsion-hemiplegia-epilepsy syndrome

Contributors

Alexis Arzimanoglou MD, author. Dr. Arzimanoglou, Director of the Pediatric Epilepsy Department of the University Hospitals of Lyon (HCL) received consulting and/or guest speaker fees from Eisai, GlaxoSmithKline, UCB Pharma, and Viropharma.

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Key points

• Hemiconvulsion-hemiplegia-epilepsy syndrome is a disorder associated with a specific clinical presentation sequence: initial prolonged hemiconvulsion (at less than 2 years of age) and immediate flaccid hemiplegia (a hemiconvulsion-hemiplegia episode), and subsequent (after 1 to 3 years) development of focal epilepsy.

• The focal partial epilepsy may be temporal, extratemporal, or multifocal.

• Onset is associated with the presence of prolonged seizures often in a febrile context. Early administration of a benzodiazepine to reduce the duration of the initial event is of primary importance.

• Although the syndrome may be associated with prior infection or lesion in some cases, a possible cause is often not revealed.

• Therapy for hemiconvulsion-hemiplegia-epilepsy syndrome includes antiepileptic drugs and surgery.

Historical note and nomenclature

The term "hemiconvulsion-hemiplegia syndrome" was first used by Gastaut (Gastaut et al 1957) to describe the following sequential combination: unilateral or predominantly unilateral clonic seizures (usually of long duration), occurring during the first 2 years of life and immediately followed by a flaccid hemiplegia (usually permanent), ipsilateral to the clonic seizure.

The term "hemiconvulsion-hemiplegia-epilepsy syndrome" was used to describe the complete form of the syndrome. It included the consequent development of a focal epilepsy, usually occurring after 1 year to several years following the initial hemiconvulsion-hemiplegia episode. Focal seizures were considered to be of temporal origin.

Further studies (Roger et al 1972; 1982; Chauvel et al 1991; Chauvel and Dravet 2002) have demonstrated that the initial episode may be observed in various situations and that the subsequent partial epilepsy can be temporal, extratemporal, or multifocal.

It can be argued that, taken separately, both the initial episode (hemiconvulsion-hemiplegia) and the subsequent development of epilepsy are not proper "syndromes." In fact, the hemiconvulsion-hemiplegia episode corresponds to a unique episode of focal status epilepticus responsible for a unilateral motor deficit, and the later development of epilepsy corresponds to a focal symptomatic epilepsy. However, the stereotyped sequence of events that characterizes hemiconvulsion-
hemiplegia epilepsy allows us to consider it as a syndrome. Thus, hemiconvulsion-hemiplegia-epilepsy was reintroduced as a syndrome in the published report of the ILAE Task Force on Classification and Terminology (Engel 2001). In the published ILAE revised terminology and concepts report, hemiconvulsion-hemiplegia-epilepsy syndrome is considered a “clinically distinctive constellation” (Berg et al 2010).

Clinical manifestations

The first sign of the syndrome is a sudden, prolonged hemiconvulsion in the form of status. It occurs in a child without antecedents, between 5 months and 4 years old, with a peak incidence during the first 2 years of life. A febrile episode is almost always associated, but in many cases, no cause is obvious (Aicardi and Chevrie 1983). The onset of convulsions may pass unnoticed, the child being discovered convulsing in bed.

If untreated, hemiconvulsions may last for several hours. They present as predominantly unilateral clonic jerks. The rhythm is variable, and the jerks can be asynchronous. Impairment of consciousness is not a permanent or even constant feature. In long-lasting convulsions, jerks may diffuse to the opposite side or change sides. Adversion of head and eyes may be observed, sometimes even before the occurrence of jerks. Autonomic symptoms (hypersalivation, cyanosis, etc.) may be associated.

Massive flaccid hemiplegia immediately follows the unilateral seizure. When jerks change sides, it is usually the side involved last that remains hemiplegic. Evolution of hemiplegia is variable. It can either remain as a permanent neurologic deficit with signs of spasticity or decrease progressively, leaving behind a slight hemiparesis. To differentiate hemiplegia from Todd paralysis, a minimum duration of 7 days is arbitrarily set. In more than 80% of the cases, the hemiplegia is permanent (Gastaut et al 1960). Rarely, it may disappear completely, although some degree of spasticity and pyramidal signs usually persist. In contrast with congenital hemiplegia, the face is constantly involved, and aphasia is present in left-sided cases.

In nearly 80% of cases, focal epilepsy will develop 1 to 3 years later (hemiconvulsion-hemiplegia-epilepsy or HHE syndrome). Focal seizures with secondary generalization and episodes of status are not uncommon. In addition to focal epilepsy, most of the children with hemiconvulsion-hemiplegia-epilepsy present with some degree of mental impairment.