EVALUATION OF PEDIATRIC SPELLS IN THE EMU

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Seizures and epilepsies present with multiple etiologies, multiple clinical features, and change across the lifespan. Epilepsy is not a single disease but a diverse group of disorders which have in common an abnormally increased predisposition to epileptic seizures. A systematic approach to epileptic seizures and epilepsies is a first step towards the diagnosis and treatment of these disorders. Determination of clinical seizure type, epilepsy localization, underlying etiology, related medical conditions and if possible epilepsy syndrome follows the general neurological approach in clinical practice, including description of symptomatic presentation, localization, and etiological investigation. Description of findings is crucial for selection of the most helpful diagnostic and therapeutic approach, for defining relationships to sleep and sleep related interactions and comorbidities.

DIAGNOSTIC APPROACH

The diagnostic approach to seizures and epilepsy involves four independent steps as follows[3]. These steps have been implemented in the most recent suggestion for a revised epilepsy classification[4]

1. Clinical Presentation (What are the symptoms?)
2. Epilepsy Localization (Where is the lesion?)
3. Etiology (What is causing the epilepsy?)
4. Syndrome (Other related features that may assist with diagnosis making or fit with known syndromes?)

Independent investigational techniques include the collection of a detailed history and clinical course, seizure semiology analysis either by history or video-review, physical examination, electrophysiological studies, structural neuroimaging and functional and metabolic neuroimaging, laboratory testing including genetic and histopathological studies, and these studies aid in answering the questions outlined above.

Recognizing non-epileptic events

When gathering information on events, always consider the option that events may be non-epileptic. In general, when witnesses report the ability to modify or stop patient's ictal movements during an epileptic seizure, a non-epileptic event should be considered.

The distinction between epileptic and non-epileptic events may represent a significant challenge for the clinician. The most commonly encountered non-epileptic events are the so-called "psychogenic" non-epileptic seizures (PNES). It is estimated that about 70% of PNES cases develop between the second and fourth decades of life, but PNES can also present in children and senior individuals. Patients with PNES may have concurrent epileptic seizures or have had epileptic seizures before presenting with PNES.

Some clinical features can help differentiating PNES from epileptic seizures, but most features are highly non-specific and no single feature is pathognomonic for PNES. When events occur during the night, and out of sleep, this distinction may be even more difficult. Disorders that may present with nocturnal non-epileptic paroxysmal events may include (1) specific sleep-related disorders, such as NREM 3 parasomnias and REM sleep behavior disorder (RBD), and (2) psychiatric and behavioral conditions, such as panic attacks[8-10] or PNES among others. These phenomena usually involve complex motor activity as seen in parasomnias and may have a wide spectrum of alterations of consciousness/awareness which makes their differentiation from seizures difficult.

An abrupt onset is typically more suggestive of epileptic seizures, although other events such as cardiac or vasovagal syncope can also present with rapid onset. Epileptic seizures tend to have a stereotypical pattern, which means that witnesses and patients tend to report events with similar dynamics and features. At times, more than one type of event can be recognized by the witnesses/patients and the clinician. Ictal activity is also involuntary and uncontrollable, which means that during the seizure the behavior is not goal-directed. Some patients during complex partial seizures (especially right temporal in origin) can follow commands often then accompanied by automatisms and varying degrees of amnesia. At times, behavior in response to an ictal aura can be seen, which is primarily not ictal in nature, but related to the epileptic seizure, such as hand shaking due to a painful aura in the hand. Post-ictal alterations including amnesia for the event, lethargy/sleep, and transient neurological deficits are also more likely in epileptic seizures and are typically reported with consistency by family members and caretakers.

When typical seizures can be recorded, video-EEG is the gold standard diagnostic tool for non-epileptic events, and a diagnosis of these disorders can be made with high accuracy.[6,11] When video-EEG reveals no ictal epileptic activity before, during or after the ictus, thorough neurological, sleep and psychiatric histories are crucial to confirm the diagnosis of these disorders.

Seizure history and semiology

Identification and classification of seizures and epilepsies is based on clinical presentation. In epilepsy, '*semiology'* refers to the study of the signs and symptoms of seizures. The seizure semiology provides important clues to seizure and epilepsy localization. The collection of information on seizure history and semiology, together with the analysis of seizure semiology with video-EEG data, are essential for understanding the clinical seizure type and provides important information on the type of epilepsy and localization, guiding treatment and addressing prognosis. Furthermore, semiology provides important lateralizing and/or localizing information.

We find it most useful to analyze the main phases of a seizure along the timeline of occurrence, therefore envision the timeline of a seizure, and inquire of features in this timeline separately, specifically:

1) Seizure setting;
2) Seizure onset;
3) Seizure presentation and evolution; and
4) Post-ictal symptoms

1. Seizure setting

Information on seizure setting requires a reliable witness capable of accurately describing the details of the entire event. If the patient is amnestic for all or part of the seizure, an independent witness is crucial. Circumstances immediately prior to the event should be noted, such as the activity in which the patient was engaged, changes in the environment, changes in the patient's behavior before seizure onset, and other prodrome, i.e. the symptoms reported within minutes, hours or even days before the seizure onset. The time of day and relationship of the events to sleep may also be elicited. For example, knowing that the events have a specific circadian pattern, arise from a particular sleep/wake state, or occur in the context of sleep deprivation helps to distinguish different seizure types and epilepsy syndromes.[16-18] The presence of fever, concomitant illness, occurrence in high altitude settings, preceding head injury, and peri-menstrual timing may be helpful. Information of seizure setting includes also prior medical history, current medical health, cognitive deficits, developmental achievements, and family history.

2. Seizure onset

Seizure onset in many people with focal epilepsies often is a so-called "aura", i.e. a sensory, gustatory, visual, olfactory, auditory or abdominal, or more complex experiential sensation(s) experienced by the patient. Auras in some patients occur in combinations. However, an aura is not necessarily present and other clinical features described below may also present as the first seizure symptom depending on the relationship of seizure onset and symptomatogenic cortical areas.

3. Seizure presentation & evolution

Seizure presentation and evolution includes symptoms and behaviors that the patient displays during the events. For practical purposes of gathering and analyzing reported information, we present information in the following major categories: 1) motor features; 2) automatisms; 3) language features; 4) autonomic features; and 5) impairment of consciousness.

Symptoms which manifest later in the seizure usually represent spread of the ictal activity to neighboring cortical regions.

4. Postictal symptoms

Immediately after the end of a seizure, patients may present with a variety of symptoms that may at times only become obvious during an examination of the patient. These may provide additional important information, because they may indicate the localization of the seizure focus. Todd's paresis (or postictal paresis/paralysis) is focal weakness in a part of the body after a seizure. Postictal weakness secondary to a seizure focus is typically contralateral to the localization of the weakness, may affect the arm, leg, and/or face. It usually subsides completely within 24 to 48 hours after the seizure. Similarly, hemianopia (partial vision loss) can also present after a seizure, self-resolves, and is indicative of a contralateral seizure focus. Postictal aphasia and dysphasia are usually suggestive of a seizure arising from the dominant hemisphere. Nose wiping can be observed after a seizure, and it has been seen more frequently ipsilateral to the side of seizure onset.

References:

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