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Seizures

Neonatal Seizures and Infantile Spasms

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Neonatal Seizures

Most seizures in a newborn are symptomatic—they are caused by acute or remote insults to the developing brain. Some of these may be eliminated or reversed; others may require concurrent treatment. Therefore, a diagnostic evaluation should precede or coincide with therapeutic intervention (see later). A second unique feature of many neonatal seizures is their unusual or subtle nature compared with seizures later in life. In addition to tonic, clonic, focal, or apparent generalized motor seizures, neonatal seizures may consist of fragmented, nonrhythmic movements, eye blinking, single-extremity posture, or electroencephalographic seizures without overt motor manifestations. Thus, the clinician should have a high level of suspicion for seizures in an ill neonate. Tachycardia or bradycardia, unexplained oxygen desaturation, or abnormal mental status may signify ongoing seizures. Finally, the outcome of neonatal seizures is best predicted by their cause. For example, hypocalcemia-induced seizures in an otherwise normal fullterm infant typically remit, with good outcome. In contrast, seizures following severe hypoxia/ischemia, severe neonatal infection, or congenital brain malformation may respond to treatment, but the infant's prognosis would be guarded.

TREATMENT (Figure 1)

There is no “absolute best” drug. Phenobarbital is recommended here because it is both rapid and long acting and has no “ceiling”; repeated boluses can be used in severe cases as monotherapy with high likelihood of success. Ignore serum levels; in any event, they are not steady-state. Use remission as your guideline. With phenobarbital monotherapy, respiratory depression

is uncommon. If you add benzodiazepines, be prepared to intubate. If the seizures or the cause are focal, phenytoin may be considered. Here, if a dose of phenytoin, 20 mg/kg, with serum levels greater than 20 µg/mL is ineffective, then further doses are unlikely to be helpful.

Infantile Spasms

Infantile spasms (West's syndrome) are a severe, relatively common (~1:2400 births), and often missed diagnosis. It is important to recognize infantile spasms because they respond poorly to conventional anticonvulsants, but remit in most infants when treated with *high-dose* adrenocorticotrophic hormone (ACTH) (see later) (Figure 2). A broad consensus suggests that cognitive outcome is better in infants with infantile spasms who are treated successfully.

Infantile spasms are considered a form of myoclonic seizures that occur in clusters. They are prevalent in 3- to 12-month-old infants and are associated with a highly abnormal (interictal), chaotic electro-encephalogram (EEG) (hypsarrhythmia). This pathognomonic EEG pattern is most commonly observed during sleep. The seizures may be flexor, extensor, or mixed, subtle, or massive, and “flattening” of the EEG during a spasm is typical. Most infantile spasms are symptomatic, resulting from a large variety of insults or genetic causes. A variant associated with tuberous sclerosis may be particularly responsive to vigabatrin (100 to 150 mg/kg/day). This medication is available outside the United States, and the side effects of visual-restrictive retinal changes should be considered.

In some cases the causes of infantile spasms may be treatable, with seizure remittance. Mostly, treatment of the spasms with a goal of eliminating them and normalizing the EEG is required. Early, successful therapy seems to improve cognitive outcome. The latter is grim in symptomatic cases but excellent in remitting idiopathic cases.

The most efficacious treatment is high-dose ACTH, with greater than 85% success. A 2-week treatment with this potent hormone results in unpleasant but rarely dangerous side effects such as acne, hypertension, voracious appetite, and irritability. Long-lasting treatment may lead to immunosuppression or gastric bleeding. Because efficacy is a function of the dose, and

side effects are a function of the duration of treatment, it is best to initiate ACTH treatment at the high dosage and limit duration to 2 weeks, with a 2-week taper. Complete dramatic remission of the spasms typically occurs during the first week. Note that twice-daily dosing at the high (ActharGel, 150 U/m² of body surface area) is required. Occasional “bad” batches of the hormone have been described.

In infants with a strong focal element of the spasms, particularly with focal lesion (e.g., tuber) and focal EEG, surgical therapy should be considered. A trial of ACTH may still be indicated and may convert apparent infantile spasms to focal seizures.

Other therapies may be successful at a much lower rate. Consider pyridoxine (to exclude pyridoxine-dependent seizures, or as therapy), 100-150 mg/day; valproate; topiramate; and the ketogenic diet.

If seizures remit and recur, verify that these are indeed infantile spasms rather than a new seizure type with focal EEG. A second course of ACTH may be effective for infantile spasm recurrence. New seizure types, instigated by the original disorder, may respond to appropriate anticonvulsant therapy.

Febrile Seizures

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Febrile seizures occur in approximately 3% of children, making them the most common type of seizure in this age group. They are seizures in infancy or childhood (typically between 3 months and 5 years of age) associated with fever and without evidence of central nervous system infection or other defined cause, such as metabolic abnormalities due to dehydration. Ordinarily, they are generalized (tonic-clonic or tonic) in nature and brief.

The fact that they are common to pediatric practices (and often to neurologic consultation) makes it important to understand many of the concerns associated with their occurrence and to understand the information that has reshaped our approach to this problem. In fact, this has resulted in reconsideration of our overall approach to seizures.

Given the generally benign prognosis in febrile seizures, counseling the family is the primary form of “therapy.” One way to consider the issues involved in febrile seizures is to organize the information in a format that answers parents’ questions and provides a framework for providing ongoing care. This can even include anticipatory guidance, which is so vital to the practice of pediatrics.

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TABLE 1 Risk of Experiencing a Febrile Seizure

Factor	Risk of Febrile Seizure (%)
General population	2
Child in daycare	7
Slow development	10
Prolonged nursery stay (>28 days)	12
Febrile seizure in first-degree relative (mother, father, sibling)	10
Febrile seizure in two first-degree relatives	33
Any two risk factors	28

Adapted from Bethune P, Gordon K, Dooley J, et al: Which child will have a febrile seizure? *Am J Dis Child* 147:35-39, 1993.

What Is the Chance That My Child Will Have Febrile Seizures?

Families with a history of febrile seizures may ask this question, particularly when there are siblings with febrile seizures. Febrile seizures are more common in some families, and 10% to 20% of siblings of children with febrile seizures will also experience them. Certain other children may have an increased risk, as high as 25% if the child had a prolonged nursery stay, slow development, or daycare attendance. These factors suggest susceptibility to febrile seizures in a child who may have experienced some subtle neurologic changes and who is exposed on a more consistent basis to a wide range of infections in the daycare setting. When two or more of these factors exist, it may be appropriate at one of the early well-child visits to discuss management of fever and what to do if a febrile seizure occurs (Table 1).

What Should We Do If Our Child Has a Febrile Seizure?

Most febrile seizures are brief and do not need medical intervention. If, however, the seizure has persisted longer than 5 minutes, Emergency Medical Services (i.e., “911”) should be called. Less than 5% of febrile seizures occur as status epilepticus, and it is likely that the seizure will end before medications can be given. The usual intervention is either lorazepam, 0.1 mg/kg intravenously (IV), up to 4 mg, or diazepam, 0.3 mg/kg IV given slowly at less than 1 mg/kg/min. Health care providers in emergency settings should recall that diazepam could be given rectally.* In essence, care during a febrile seizure should be the same as for any other generalized convulsion.

When the seizure is over and the child has returned to baseline (often sleepiness if the seizure has occurred at night), a decision must be made concerning the

*Diastat, 0.5 mg/kg rectally for children 2 to 5 years, 0.3 mg/kg rectally for children 6 to 11 years, 0.2 mg/kg rectally for children older than 11 years, all rounded to the nearest syringe size; one dose may be repeated after 30 minutes, if necessary.

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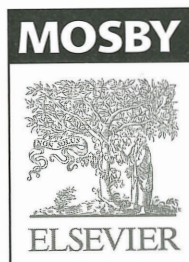
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