Common Medical Concerns in the First Few Months / Newborn and Infantile non epileptic events

01 WHAT DO I NEED TO KNOW?

Unusual movements in newborns and infants occur frequently and are often a source of concern for parents who may be concerned that these may represent a seizure disorder. This is an excellent excerpt from an excellent reference on the topic.

02 WHAT OTHERS SAY

Excerpt from Imitators of Epilepsy, 2nd edition. Kaplan PW, Fisher RS editors

New York: Demos medical publishing 2005

NEONATAL NON EPILEPTIC EVENTS

As discussed, some clinical events such as "subtle seizures," generalized tonic, and focal/multifocal myoclonic events, do not necessarily have an EEG correlate. These events may reflect brainstem-release phenomena (therefore, nonepileptic events) or may represent subcortical epileptic seizure activity. Newborns also exhibit a variety of periodic normal movements that can be readily identified by experienced observers as being nonepileptic. First-time parents may not be familiar with the character of these movements. When attacks recur frequently, parents can be asked to videotape the events. Subsequent review of the tapes can help to establish the correct diagnosis, and may also be used to reassure apprehensive parents. Some examples of common nonepileptic paroxysms include nonconjugate eye movement, sucking movements without associated eye abnormalities, and nocturnal myoclonus. Other specific nonepileptic events are described below. Age ranges are somewhat arbitrary, and some of the nonepileptic events that have been classified under infancy may begin in the newborn period.

Apnea

Apnea, or cessation of breathing for greater than 15 seconds, is usually not due to an epileptic seizure, particularly if apnea is the sole manifestation, and the patient has not been already treated with anticonvulsant medication. Apnea may be secondary to centrally mediated hypventilation or an obstructive etiology. Apnea occurs commonly in the premature child, especially during active sleep. This “apnea of prematurity” is likely to be secondary to brainstem immaturity and is typically associated with bradycardia (A’s and B’s). Apnea may occur in the older infant and present as an acute life-threatening event (previously called sudden infant death syndrome (SIDS)). Since apnea may also be an important sign of neurologic diseases such as hypoxic-ischemic encephalopathy, intraventricular hemorrhage, infections, hypoglycemia, and medication side effect, an extensive search for an underlying etiology should be undertaken. If there are other concurrent manifestations, such as eye opening, eye deviation, mouth movements, tachycardia, or hypertension, an ictal etiology should be suspected.

Jitteriness

Jitteriness is a common movement phenomenon in the newborn period. Jitteriness is associated with drug withdrawal, hypocalcemia, hypoglycemia, and hypoxic-ischemic encephalopathy. Movements typically have an oscillating quality, with tonic and foci oscillations of equal frequency and amplitude. Jitteriness can occur spontaneously, but is also very stimulus-sensitive and can often be precipitated by touch or loud noise. In addition, the movements can be dampened by consoling the child, removing the stimulus, and relaxing the affected limb. The child is typically awake during these events and no associated autonomic activity is noted. These movements may need to be distinguished from either clonic or repetitive myoclonic seizures. The quality of the jittery movement is different either from the two-phase movement of clonus or the quick jerks of myoclonus. The extreme stimulus-sensitivity, suppression of the movements, and lack of associated autonomic disturbances also suggest nonepileptic episodes.

Benign Neonatal Sleep Myoclonus

Repetitive myoclonic jerks occurring during non-REM sleep constitutes a well-described clinical phenomenon. The myoclonic jerks usually begin in the first few weeks of life and resolve by 2 to 3 months. The jerks are typically bilateral and symmetric, involving the arms and legs. However, focality and migration of the myoclonus among different muscle groups has been noted. The EEG and neurologic outcome is normal. The jerking may be significantly repetitive so as to mimic clonic seizures. Helpful features to distinguish these myoclonic jerks from epileptic phenomena include the absence of autonomic activity, no other associated seizure types, myoclonic jerks occurring only at night, and a normal neurologic and developmental examination. In addition, if the child is awakened during an episode of benign neonatal sleep myoclonus, the movements cease. It should be noted that isolated hypnogogic myoclonic jerks can occur in all age groups and is a normal physiologic phenomena.

Pathologic, Nonepileptic Myoclonus

Neonates with severe cerebral dysfunction from a variety of causes may present with myoclonic jerks that do not have an EEG correlate but are secondary to the underlying disease process (pathologic). These myoclonic jerks may be focal, multifocal, or generalized. The seizures may occur when the child is awake or asleep. This form of myoclonus is often stimulus-sensitive. Neurologic disease states associated with this nonepileptic pathologic myoclonus include metabolic encephalopathies (for example, hyperglycinemia), hypoxic-ischemic encephalopathy, cerebral vascular events, and infections. Medications may also produce nonepileptic myoclonic jerks (10).

Hyperekplexia

Hyperekplexia (stiff baby syndrome, startle disease) is a genetic disease involving an abnormal gene for a subunit of the glycine receptor on chromosome 5q. Inheritance is autosomal dominant or sporadic. The hallmark of the disease is a triad of symptoms including generalized stiffness of the baby, particularly while awake, nocturnal myoclonus, and an exaggerated startle reflex. Upon awakening or with an auditory or tactile stimulus, the neonates and infants may have a marked generalized episode of stiffening associated with apnea. These episodes can be severe and cause hypoxic brain injury. Manual flexion of the neck and hips may resolve an episode. These episodes lessen in severity as the child grows older. As adults, the subject may have a pathologic startle response to even a minor visual, auditory, or tactile stimulus. Both clonazepam and valproate have been cited in the literature as useful therapies.
INFANTILE SEIZURES
A number of investigators have attempted to classify infantile seizures. While no universally accepted infantile seizure classification system yet exists, the unique seizure symptomatology of infants is now recognized. Infantile seizure semiology includes the following:

Infantile Seizure Semiology.

- Tonic posturing. This extremity stiffening may be symmetric or asymmetric. Either type of tonic posturing may reflect either focal or generalized EEG changes.
- Clonic jerking of one or more limbs. Bilateral clonic jerking typically does not have the synchronous rhythmicity of adult generalized clonic seizures. Although bilateral clonic movement may reflect either a focal or generalized electrographic correlate, unilateral clonic jerking of an extremity does correlate with contralateral hemispheric discharge.
- Astatic events with loss of tone of a part or the whole body.
- Hypermotor seizures characterized by a distinct but subtle behavioral arrest. If automatisms exist, they are typically subtle and simple, involving restless extremity movements, chewing, and lip-pursing. Complex semipurposeful automatisms are not seen in infants.
- Myoclonic jerks (isolated or repetitive) can be seen in either a partial or generalized electrographic correlate.
- Versive seizures consisting most notably of forced eye deviation.
- Infantile spasms. Infantile spasms are clusters of quick extension or flexion spasms involving the neck, arms, and trunk. The spasms initially resemble a quick jerk that then sustains the posture for a few seconds. It has been debated in the literature whether these seizures should be classified as myoclonic jerks or tonic seizures. Given the distinct seizure semiology, unique interictal electrographic appearance of hypsarrythmia, and poor prognosis regarding development, a unique seizure type is proposed for these events.

Infantile Nonepileptic Events
A wide variety of events mimicking seizures in the infant. Within the first 2 years of life, while the central nervous system is maturing, children can exhibit different behaviors and events that are physiologically normal for that age, although many appear paroxysmal and unusual to care-givers. In addition, well-described paroxysmal disorders can also mimic epileptic events. The remainder of this chapter reviews spells that occur in 1-month to 2-year-old infants that may mimic myoclonic, clonic, or tonic seizures, as well as events marked by a loss of consciousness, abnormal eye movements, and unusual behaviors. The following nonepileptic events are categorized as events with excessive movements, events that mimic tonic seizures, and events of abnormal eye movements. In addition, the nonepileptic events described for neonates (apnea, jitteriness, benign neonatal sleep myoclonus, pathologic/nonepileptic myoclonus, and hyperexplexia), may also occur in infants.

Nonepileptic Events with Excessive Movements
Benign Myoclonus of Early Infancy
Benign myoclonus of early infancy, also referred to as benign nonepileptic infantile spasms, is a syndrome in which infants have clinical events suggestive of infantile spasms but with a normal EEG and clinical development. The events, which begin between 3 and 8 months, resemble infantile spasms, with clusters of flexion or extension extremity movements. However, unlike spasms, both the interictal and ictal electrographic pattern are normal. Regardless of treatment, the events resolve spontaneously at 2 to 3 years of age. Head magnetic resonance imaging (MRI) and neurologic examination are normal, as is the developmental outcome. No subsequent seizures of any type are noted on follow-up.

Shuddering Attacks
Shuddering attacks may begin in infancy, as early as 4 months, or in childhood. The events consist of a rapid tremor of the head, shoulder, and trunk suggestive of the "shuddering" episodes from a child. The duration of the events is brief, lasting only seconds, but the events may occur multiple times a day. The spells are often associated with eating and may represent a pattern of stimulation overflow in a young child. The electrographic pattern during these spells is normal. No other neurologic abnormalities are associated with these events. The spells require no treatment and spontaneously resolve by the second decade. A family history of essential tremor has been noted for children with shuddering attacks.

Benign Paroxysmal Vertigo
Benign paroxysmal vertigo is reflected by the recurrence of events of brief dysequilibrium in young children. When the attacks occur, the child appears frightened and off balance, often reaching out to steady himself. The events may be associated with dystonia, diaphoresis, nausea, and vomiting. These attacks occur in toddlers and young children but typically resolve by age 5 years. Neurologic examination, development, and EEG are normal. A later association with migraine has been reported.

Stereotypies
Stereotypic behaviors are repetitive movements such as head banging, head rolling, body rocking, and hand flapping. These behaviors can be seen in normal children but are more common in neurologically impaired infants. The behaviors may be seen while the child is awake, falling asleep, or even in early sleep stages. These behaviors are "self-stimulating" behaviors and often comfort or relax the child.

Masturbation
Infantile masturbation can present as paroxysmal episodes of rocking or stiffening. The children may be sitting or lying prone or supine with a rhythmic rocking movement. During the events, the infants are alert but may appear to have a decreased level of responsiveness or an "unusual" look on their face. The behavior can be interrupted. A careful history or a videotape of the events can distinguish masturbation from seizure activity.

Nonepileptic Events That Mimic Tonic Epileptic Seizures
Sandifer Syndrome
Infants with gastroesophageal reflux may have intermittent paroxysmal spells of generalized stiffening and opisthotonic posturing. These spells may also be associated with apnea, staring, and minimal jerking of the extremities. A careful history will reveal that these spells are associated with feedings, often occurring within 30 minutes of a feed. Sandifer syndrome can be seen in neurologically normal children as well as children with hypotonia and tracheomalacia, which may predispose the child to the acid reflux. The extreme generalized stiffening may represent a pain response to the acidic material refluxing into the esophagus. No electrographic correlate is noted with these events. If the history suggests Sandifer syndrome, a gastroesophageal work-up should ensue.

Dystonic Drug Reaction
Dystonia is a sustained abnormal posture that occurs from the contraction of both the agonist and antagonist muscle groups of an extremity. Dystonic postures may be generalized or focal. Spells of paroxysmal dystonias are difficult to differentiate from tonic seizures. In infants, a common etiology of sudden dystonia is an acute drug reaction. These reactions may manifest themselves as opisthotonic posturing, torticollis, and an oculogyric crisis. Metoclopramide, a parasympathomimetic drug often used for the treatment of gastroesophageal reflux, is a common medication used in infants that can cause this drug reaction. Other medications associated with dystonic drug reactions, such as phenothiazines and haloperidol, are used less often in toddlers.
Cyanotic Syncope (Breath Holding Spells)
Breath holding spells are common events in infants and young children age 6 months to 6 years. The attacks are precipitated by minor injury, frustration, or anger. The events are heralded by crying followed by apnea that typically occurs in expiration. The child then develops significant cyanosis. If the attacks are prolonged, the child loses consciousness and becomes limp. If significant hypoxia occurs, the child may develop tonic posturing or even a few extremity jerks that may be mistaken for seizure activity. The key to the diagnosis is the preceding crying and cyanosis prior to each event. Difficulty in correct diagnosis occurs if the onset of the attacks is not witnessed. Cyanotic syncope is presumed to be a benign involuntary development response to the injury and crying that the child will ultimately outgrow.

Palid Syncope
Palid syncopal events are similar to breath holding spells. Again, events are typically precipitated by minor injury, frustration, or fright. Instead of significant crying and cyanosis, the child becomes pale and then loses consciousness. If the events are prolonged, tonic stiffening may be seen. The pathophysiology is secondary to a brief bradycardia or asystole resulting in decreased blood flow and the subsequent facial pallor. Atropine has reportedly been of benefit in decreasing the frequency of these spells.

Paroxysmal Torticollis
Torticollis is an abnormal sustained posture of the head and neck in which the head tilts to one side and the face rotates to the opposite side. In paroxysmal torticollis, the events begin and end suddenly. The attacks can be brief or prolonged. The child is alert and responsive during an attack although the patient may appear uncomfortable and irritable. The EEG is normal during the event. The etiology of the attacks is unknown, although both a focal dystonia and labyrinth dysfunction have been suggested as the cause, as has migraine. Often a family history of migraines is noted, and children with benign paroxysmal torticollis may develop typical migraines later in life. Paroxysmal torticollis usually begins in the first few months of life, and resolves by age 3 years. No treatment is required.

Decorticate/Decerebrate Posturing
Decorticate and decerebrate posturing can mimic tonic seizures. Decorticate posturing refers to flexor posturing of the upper extremities and extensor posturing of the lower extremities. In decerebrate posturing, both the upper and lower extremities show extensor stiffening. Both events can occur suddenly from a severe brain injury affecting the brainstem level. This posturing can be seen bilaterally or unilaterally in different herniation syndromes. Possible impending herniation should be considered in a comatose patient exhibiting sudden tonic stiffening (25).

Nonepileptic Events of Abnormal Eye Movements
Oculomotor Apraxia
Oculomotor apraxia is a condition in which the child has impaired saccadic eye movements. The child appears to have fixed eye positions although the visual system and eye movement ability are normal. Therefore, in order to view an object, the child will turn his head suddenly so as to move the direction of gaze. These peculiar head thrusts may be confused for epileptic seizures. The idiopathic congenital form of this apraxia is called Cogan’s oculomotor apraxia. However, oculomotor apraxia can also be seen in ataxia telangiectasia and lysosomal storage diseases.

Spasmus Nutans
Spasmus nutans consists of a triad of symptoms including nystagmus, head nodding, and head tilt. The symptoms can wax and wane during the course of the day and, therefore, be confused with epileptic seizures. The triad has been associated with mass lesions at the optic chiasm or third ventricle. A head MRI scan should be obtained in these infants. If no abnormality is revealed, no further work-up or treatment is required. The symptoms usually resolve by age 5 years.

Opsoclonus
Opsoclonus consists of random, erratic, conjugate oscillation movements of the eye (“dancing eyes”). These movements usually occur continuously including sleep, although they may wax and wane in intensity. In mild opsoclonus, a brief stable fixation on an object may be seen. These eye movements are usually seen in association with myoclonus and ataxia. Opsoclonus can be seen in children with an occult neuroblastoma, encephalitis, or without an identifiable cause.

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