

Paroxysmal non-epileptic events

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Abstract

Objective: this publication aims at reviewing one of the most important problems faced by the pediatrician in the field of child neurology. The paroxistic non-epileptic events are also a frequent reason for pediatric neurology consultations and admission for diagnostic video-electroencephalogram monitoring.

Methods: literature review on the subject was perform on Medline, data was also collected from the main Pediatric Neurology Textbooks, which were found to be important and unique source of information on the subject.

Results: many of entities discussed in this paper are very common in the pediatric population like syncope, breath-holding spells and the movement disorders associated with gastroesophageal reflux. Other syndromes are less frequent such as the pararoxymal dystonias and the Segawa Syndrome (dystonia with diurnal variation).

Conclusions: the basic knowledge of these syndromes is very important since it may avoid unnecessary procedures and the wrongful diagnosis of epilepsy. Patients who are mistakenly diagnosed as epileptics are exposed to anti-convulsant medications, which are probably not going to be effective and may expose them to the risk of side effects.


Introduction

About 10-30% of the children referred to neurology-epilepsy clinics have an incorrect diagnosis.1-3 According to Bye and Nunan up to 1/3 of the patients with suspected seizures evaluated with video-EEG have in actuality non-epileptic events. A large variety of events may mimic at least in part epileptic seizures.4 These events show resemblance with one or many of the aspects of epileptic seizures such as loss of consciousness, behavioral arrest, autonomic changes and repetitive motor or psychological behavior. This review aims at discussing only some of the most common paroxysmal non-epileptic seen in pediatric neurology practice. The tables are organized alphabetically but not all the diseases mentioned included in the tables are discussed. Table 1 listsalphabetically the most common types of paroxysmal non-epileptic events, which are seen in pediatric neurology practice.
Table 1 - Paroxysmal non-epileptic events

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**Transient cerebral hypoperfusion-hypoxemia causing paroxysmal signs and symptoms**

Among the most common seizure imitators are breath-holding spells and syncope. We start this review with these two entities because they are very common in the general population but also due to their similar physiopathology. The basis of the symptoms of both syncope and breath-holding spells is a decrease in perfusion and delivery of oxygen delivery to the brain leading to energy metabolism failure. Authoritative sources list a few critical values which in an adult individuals would lead to CNS dysfunction: bradycardia of less than 40 beats per minute, tachycardia of more than 150 beats per minute, asystole of more than four seconds, systolic pressure of less than 50 mmHg or venous 02 pressure of less than 20 mmHg.5,6 This cortical hypoxia-ischemia leads to a sequence of event including: loss of consciousness and of postural tone, increase in tone (decorticato/opisthotonus) followed by a few beats of clonus. This sequence is probably due to liberation of corticoreticular inhibition. Cortical hypoxia produce slowing of frequency of the brain electrical activity (loss of consciousness-hypotonia), followed by complete flattening of the EEG tracing during the tonic phase. As the hypoxia gets corrected the slow waves re-appear and at the same time that a few jerks of the limbs are seen before resumption of normal EEG activity.7,8

**Syncope**

Syncope is a commonly confused with epileptic seizures. Syncope accounts for about 1% of pediatric emergency room visits.9,10 One study the prevalence of syncope among adults was 3% for men and 3.5% for women.11 One third to half of the cases of syncope show when followed up for 5 years.12

During a syncope there is a transient cerebral hypoperfusion, followed by spontaneous recovery which is associated loss of consciousness and postural tone.7,13-15 An interruption of cerebral blood flow lasting 8 to 10 seconds causes loss of consciousness.13 There are many causes and triggers of syncope (see Table 2).
The diagnosis of syncope rests mainly on clinical grounds. The syncope is often precipitated by assuming the upright posture (orthostasis), prolonged standing, heat, fatigue, hunger, Valsalva maneuver, physical or emotional stress including venipuncture, pain, and fear, are also commonly identified. Several activities which produce a valsalva maneuver and/or parasympathetic activation can cause syncope including cough, defecation, deglutition, diving, hair grooming, micturition, sneezing, trumpet-playing, weight-lifting. Other stimuli include getting into or out of a bath, or stretching. Less commonly syncope may result from primary cardiac events (see primary cardiac syncope below).

Syncope is often preceded by a sensation of lightheadedness, blurred vision, “things going far away” (sometimes described as “tunnel vision”), pallor, nausea, epigastric discomfort or diaphoresis. In most cases the loss of tone is characterized by a progressive and slow slumping to the ground. Less frequently the loss of posture may be abrupt with a sudden fall which may be associated with tongue lacerations. Less commonly syncope may result from primary cardiac events (see primary cardiac syncope below).

The loss of consciousness during a syncope usually lasts from a few seconds to a few minutes. During the period of impairment of consciousness the patient has decreased postural tone, which in the prolonged events is followed by tonic posturing and a brief clonic activity. Less frequently incontinence may be seen. After the syncope a spontaneous recovery without persistent neurologic deficits is the rule but nausea, pallor, diaphoresis and fatigue may be seen. The recovery takes place in less than an hour. Even though most cases syncope in children are benign and self-limited falls may lead to physical injuries.

Even though syncope is usually can be distinguished from epilepsy both by the clinically and EEG picture in some cases both diseases may co-exist.

### Syncopes of cardiac origin

Syncopes of cardiac origin should be considered apart since they require a different focus in their work-up. Syncopes of cardiac are less frequent than reflex vasovagal or cardioinhibitory attacks. The conditions that cause these syncopes are associated with sudden death, which is the reason why it is important to recognize them. Both congenital and acquired (rheumatic) aortic stenosis that can produce syncopes on effort and the same is true for cardiomyopathies and disturbances of cardiac conduction. Among the conduction abnormalities associated with syncope are the long Q-T syndromes, Wolff-Parkinson-White syndrome, and congenital atrioventricular block.

### Table 2 - Causes-precipitators of syncope

| Cardio-inhibitory syncope of infants (a.k.a. breath holding spells) |
| Cardiovascular |
| Atrio-ventricular conduction abnormalities (AV heart-block) |
| Cardiac arrhythmias (long QT syndrome) |
| Congenital Heart Disease |
| Rheumatic Heart Disease |
| Congestive Heart failure |
| Drug-induced syncope |
| Hyperventilation syndrome-related syncope |
| Neurocardiogenic |
| Situational-valsalva-parasympathetic activation syncope: cough, defecation, deglutition, diving, hair grooming, micturition, sneezing, trumpet-playing, weight-lifting |
| Suffocation syncope |
| Psychogenic syncope |
Jervell-Lange-Nielsen syndrome features an associated neurosensory deafness and is recessively inherited and the Ward-Romano syndrome, which is dominantly transmitted. Both syndromes can mimic epilepsy and in a few cases secondary epileptogenesis may be seen can result in sudden death in some children.25,26 The episodes may occur during sleep, exercise and may be also precipitated by emotion or stress but since similar precipitants may be seen in epilepsy EKG and EEG confirmation is mandatory in all cases. A family history of sudden death or ‘epilepsy’ or a personal history of chest pains, palpitations or of a surgically repaired heart defect should trigger a full cardiac evaluation. The EKG examination should include Q-Tc interval calculation. The Q-T interval is measured from the onset of the first QRS waveform to the end of the T wave on the EKG. The corrected (QTc) is calculated by dividing the QT interval value by the square root of the RR interval, QTc values above 0.44 seconds are abnormally elevated.27 A prolonged EKG recording and an effort test may be indicated when there is a high index of suspicion, especially in association with history of familial syncope or sudden death is present. The treatment with pacemaker placement may prevent sudden death. There are at least three different genetic substrates for long Q-T syndromes: one linked to the ras-1 protein gene on chromosome 11p15; another associated to a potassium channel mutation on chromosome 1q35-q36 (LQT2), and a third linked to a sodium channel protein mutation on chromosome 3 (LQT3).28

Another type of syncope of cardiac origin are the attacks of hyperventilation and cyanosis seen in congenital cyanotic cardiopathies such as the Tetralogy of Fallot. These attacks if severe will result in loss of consciousness. One should be careful since congenital cyanotic cardiopathies are associated with a high risk of thromboembolic phenomena, which may lead to strokes and true epileptic foci.

Patients with poor cardiac reserve who are in chronic heart failure may also faint when they try to exert themselves or when their fragile cardiovascular system is overtaxed such is the case during severe infections.

**Treatment of syncope**

The idiopathic syncope is best managed acutely by increasing the blood flow (venous return) to the heart by placing the child flat with legs at the level of the heart or above it. Patients with situation related syncope should avoid the precipitating environment such as extreme heat. In all cases of repetitive syncope before the diagnosis of “idiopathic syncope” is made it is wise to have a proper cardiac evaluation with good history, careful cardiac auscultation and EKG interpreted by someone familiar with pediatric patterns. In selected cases consultation with a pediatric cardiologist is strongly advised especially.

“Breath-holding” spells (Cardio-inhibitory or Pallid syncope and Cyanotic breath holding spells)

“Breath-holding” spells are frequently confused epileptic seizures. One of the main reason for that are the clonic movements which can be seen at times towards the end of these spells.

**Cardio-inhibitory or Pallid syncope**

A better denomination for some these episodes would be cardio-inhibitory syncope (CIS or pallid “Breath-holding” spells) since the main problem is transient cerebral hypoperfusion (syncope) induced by bradycardia leading to facial pallor. Other cases a Valsava maneuver with or without bradycardia causes a similar syncope during which the patients are more or less cyanotic (Cyanotic “Breath-holding” spells).

The age onset range is between 6 and 24 months but cases with onset in the neonatal period and as late as 42 months have been described.29 About 4.9% of the pediatric population is thought to have these episodes.29 The sequence of events tends to be helpful when the parents are observative. A mild head injury or any type of pain or even extreme emotional upsetting will start the episode. The trigger fever may be also a trigger for similar events.30 After that the patient will begin to cry and than suddenly stops crying (giving the impression of “holding the breath”) and turns pale (thus the name pallid syncope), quickly they become hypotonic and most of the times the episodes will end on this phase with gradual recovery of the patient’s color and normal activities. Some times the parents will touch the patient’s chest and will notice the heart rate to be slow or they will not be to feel the heart beating or the pulse (which is usually weak) in the limb arteries so the worried parents will start doing CPR in the child if they know how to do so. In the more prolonged cases after the hypotonic phase the patients will become rigid (hypertonic) and may even show a few clonic beats mostly in the upper extremities. These patients will be often labeled as having seizures or epileptics and are treated with anticonvulsants. Phenobarbital, which one of the anticonvulsants most used in this age group may selectively depress the transmission in sympathetic ganglia and has a mild effect in the heart rate and blood pressure that may be potentially harmful for patients with a tendency to bradycardia. Around 18% of these patients will have episodes of syncope as adolescents.31

The diagnosis is made mostly by the typical history. In the past ocular compression has been done to confirm the diagnosis.29 Nonetheless this test has fallen out of favor due to some potential ocular side-effects.32

Treatment is rarely needed and reassurance as well as proper teaching on how to monitor vital signs (such as it is done in good CPR training) can help convince the parents
that the patient is not dying during the events. Parent education should also mention that putting the patient on a vertical position may worsen the symptoms by decreasing cerebral perfusion. Even though this seems to be very obvious during the crises the parents may instinctively pick up the child with idea of consoling her/him. Patients with anemia should be properly treated since a low hemoglobine is associated with an increase in the number of crises. Atropine 0.01mg/kg/day (40-240 microgram/kg/dose) can be sometimes helpful in children with frequent episodes.

The physiopathology of the cyanotic variant of breath holding spells is probably multifactorial in nature still imperfectly known. Among the several factors that have been implicated in the genesis of these crises are hyperventilation (causing alkalosis and pCO2 reduction), apnea, Valsava maneuver (causing an increase in the intrathoracic pressure e decreased venous return/cardiac output). The latter factors decreased cerebral perfusion and oxygenation. The latter factors are much to slow to explain the quick onset of cyanosis. An imperfect matching of ventilation to perfusion resulting in intrapulmonaty right-to-left shunt is one possible explanation.

Cyanotic breath-holding spells are also provoked by pain and sudden emotional changes such as fright, anger or frustration. After the precipitant factors the infant cries, then holds her/his breath in expiration. The sequence of events is one of cyanosis, loss of consciousness and limpness. In rare cases some brief body stiffening may be seen before respiration restarts at the end of the attack. The polygraphic recordings pattern have shown a sequence slightly different of from the one seen in the pallid variant with slowing of the EEG tracing with bradycardia such as one would expect in the overshoot phase of a Valsalva maneuver. Breath-holding spells can have a frightening appearance but are rarely if ever associated with serious consequences so drug therapy is not indicated.

**Self-induced reflex syncopes**

Gastaut et at. (1987) described a type of syncope that occurs mostly in retarded and/or psychotic patients. These self-induced reflex syncopes are rare but at times can cause difficult diagnostic problems since they maybe mistaken for absences or drop-attacks. The children stop breathing, perform a Valsalva maneuver and after a few seconds, they become pale, stare and loose postural tone. Sometimes the loss of postural tone may be limited to the neck or it may involve the lower limbs with causing a fall. In some cases flenfluramine may be helpful but currently due to cardiovascular toxicity this drug is not used very often. This pattern may be rarely seen in children with normal intelligence.

**Benign neonatal myoclonus**

Commonly benign neonatal myoclonus is described as repetitive myoclonic jerks which are seen mostly during non-REM sleep involving mainly the arms. In this syndrome patients have myoclonus which is often decreased by gentle restrain even simple touch. The myoclonus is restricted to sleep periods and and stop abruptly with awakening. The myoclonus is focal, multifocal or generalized but in almost every case the focal myoclonic activity will eventually migrate to other sites. Repeated jerks lasting for a few seconds often recur every 3-15 minutes but in some cases the cluster may last up to 60 minutes. The movements may be so dramatic that even experienced pediatric neurologists may confuse it with epileptic neonatal myoclonic seizures (patient 1 reference 37). The family history may be positive in 10% to 25% of the cases. Treatment with anti-convulsant medications (lorazepam, phenobarbital, phenytoin) may be ineffective. Even though in most cases no autonomic changes are seen, in 2/10 cases described by Daoust-Roy and Seshia (1992) showed 10-30% increases in heart rate. Polygraphic studies of benign neonatal myoclonus suggest that this movement pattern is seen mostly during non-REM (quiet) sleep. Nonetheless as many 22% of the events may actually occur during in REM (active) sleep or less commonly (3% of the cases) during transitional sleep. The same study found that quiet sleep was the predominant state in their tracings and that in any particular infant with benign neonatal myoclonus the events did not occur more frequently or exclusively during quiet sleep. The electroencephalogram is normal during these events may show movement artifact. In some cases the events may be followed by a decremental reaction which is a normal pattern of arousal and/reactivity in neonatal EEG tracings after 32 weeks of post-conceptional age. Some tracings may show excessive multifocal sharp transients but one puzzling finding in one study was the presence of positive sharp waves in some patients over the temporal and central regions.

As the name implies benign neonatal myoclonus is thought to be an entity with a good prognosis with remission of the myoclonus and normal development in almost all the patients. In some instances sleep myoclonus may persist through 4 months of age. If the diagnosis is correct (infant with myoclonus with normal neurological exam and EEG) no treatment is recommended even for the cases in which the myoclonus persists for several months.

**Benign myoclonus of early infancy**

Benign myoclonus of early infancy is a syndrome in which the infants have series of brief tonic or myoclonic contractions involving the axial muscles and more prominently the neck (cephalic myoclonus). Flexion, extension or abduction of the arms can also be seen. Most of these events occur during wakefulness, the EEG’s are
normal and the long term prognosis is excellent so no treatment is recommended.41

Shuddering attacks

Shuddering attacks are an uncommon benign disorder of infancy and early childhood.42-45 This problem is characterized by shivering movements with bilateral fine tremors, at times associated with stiffening of the upper extremities.44,45 The attacks were not associated with impairment of consciousness or electroencephalographic abnormalities.44,45 These attacks last for several seconds, occur daily and may be very frequent. Neurophysiologic monitoring (electromyography/ electroencephalography) of these events shows that the activity has a similar frequency to be almost the same as that of essential tremor.45 A family history of essential tremor is not unusual.42 Shuddering attacks may represent the expression of the mechanism of essential tremor in the immature brain.42 In most patients the shuddering attacks decrease in number or disappeared overtime.45

In most cases the patients are otherwise normal but mild abnormalities on magnetic resonance imaging have been described.45

Children with shuddering spells specially those with a positive family history of tremor may develop head tremor (“yes-yes” or “no-no” movement) which can be accompanied by the development of mild dystonia.46

Shuddering attacks are usually self-limited and improve overtime becoming less frequent or remitting during the latter part of the first decade but the development of essential tremor has been seen (see above). In the cases that shuddering attacks are very disturbing propranolol therapy may be helpful.47 In most cases no treatment is necessary.

Non-epileptic seizures/Pseudoseizures/Psychogenic seizures

Pseudoseizures have been more recently called psychogenic seizures. These events are involuntary, intermittent episodes that resemble epileptic seizures, but are not associated with epileptic discharges. Psychogenic seizures can be psychologically induced especially by suggestion.

These events are often a manifestation of psychiatric disturbances such as conversion or somatization disorder. Psychogenic seizures have an age of onset 4-70, more often in teenagers and they are three times more frequent in females than in males.6 These types of events are thought to be a consequent of a “deep seated” conflict. A remote history of child abuse can be elicited in some cases. Nonetheless the psychological origin of the pseudoseizure may not found even with proper psychotherapy. Psychogenic seizures are often triggered by an unconscious mechanism may not be obvious on an initial interview but they are commonly precipitated by suggestion.

Psychogenic seizures must be distinguished from malingering seizures in which the patient will fake the event due to a secondary gain such as medico-legal compensation. In malingering seizures there is awareness of both the production of symptoms and the underlying motivation.

Clinically pseudoseizures have being recognized since the time of Charcot. These events usually occur when other people are around (at home or at school) and are often precipitated by emotional factors. Often there is recall of the episodes. During the events there is a combination of some of the following elements: movements that tend to be uncoordinated, non-synchronous thrashing of limbs, quivering, pelvic thrusting, side to side movements of the head, opisthotonic posturing, screaming or talking, episodes are often not stereotyped and the eyes tend to be closed. When pseudoseizures resemble generalized tonic-clonic seizures the tonic phase is rarely seen, the clonic phase is associated with wild thrashing movements, there is usually no loss of consciousness and if there is loss of consciousness it may be accompanied by shouting or obscene utterances. There are some features that are often not helpful since they maybe seen in both epileptic and non-epileptic events include pupillary dilatation, depressed corneal reflexes, Babinski response and cardiorespiratory changes. Some patients may have event that are almost identical to partial motor (focal) or staring-absence seizures.6 In rare cases incontinence and even injuries may happen during the events. In some cases post-ictal confusion or abnormal clinical behavior after an attack may be seen.

During the pseudoseizure the EEG does not show any alteration of the background and no organized seizure activity or postictal slowing is seen. During an electrographic epileptic seizure the EEG shows a disruption of ongoing activity such as suppression of the posterior background rhythm, attenuation besides that there is usually the onset of a novel rhythmic electrographic activity.

Making the diagnosis by video-EEG and inducing an episode by suggestion is the preferred method of confirming the non-epileptic nature of psychogenic seizures.

One has to be careful not to confuse pseudoseizures with extratemporal (specially frontal lobe) onset seizures. Patients with frontal lobe seizures have bizzare patterns with pelvic thrusting, copulation-like or alternating leg movements.6 Besides that frontal onset seizures are at times difficult to document by EEG recordings. Video-EEG monitoring will show in frontal lobe seizures a stereotyped pattern of behavior whereas consecutive seizures tend to vary more in the psychogenic events. Patients with frontal lobe epilepsy also tend to have seizures at night, where as pseudoseizures are usually daytime events.
Postictal serum prolactin measurements have been advocated as a means of differentiating psychogenic from epileptic events. Prolactin is elevated in mostly generalized and tonic-clonic and complex partial seizure types but may be normal in simple partial seizures and in some unilateral mesial temporal seizures.

Other helpful tests include neuropsychological testing patients with psychogenic seizures are more likely to have high scores on the schizophrenia, hypochondriasis, and hysteria scales of the MMPI.

Even though as many as 50% of the patients with pseudo-seizures may also have genuine epileptic attacks in adults this occurrence is not as common in children in our experience. In these cases anti-seizure medications often reach toxic levels because psychogenic seizures continue after epileptic seizures are controlled with lower doses of these drugs.

One should be careful about making the diagnosis of psychogenic seizures since careful follow-up may show a relatively high rate of recurrence even with appropriate treatment on long-term follow-up (see reference 6 for review of the literature). The treatment of psychogenic seizures includes psychotherapy, behavioral modification, relaxation, stress management, reassurance. Psychotic patients with pseudoseizures need proper antipsychotic medications.

Hyperventilation syndrome

This syndrome is defined by symptoms, which are reliably reproduced by voluntary hyperventilation. These symptoms seen during attacks of hyperventilation include anxiety, breathlessness, light-headedness, and paresthesias. Children with hyperventilation syndrome commonly have many chronic symptoms including headaches, irritable bowel anxiety and depression. In the pediatric age group the syndrome is more frequent among adolescents, although it has been recorded as early as 6 years of age.

It is not uncommon for the children hyperventilation syndrome to have psychiatric problems before the onset of hyperventilation attacks. The prognosis of this syndrome is not very good and up to 40% of patients are still having hyperventilation episodes in adulthood.

Authoritative sources have pointed out that this diagnosis is commonly missed and requires a high index of suspicion as well as reproduction of the symptoms during hyperventilation. Voluntary hyperventilation does not reproduce these symptoms every time so empirical treatment is indicated when the diagnosis is being strongly considered. Breath-holding, slow breathing, or breathing into a paper bag may improve the symptoms and it is recommended during the crises. Treatment with reassurance, psychotherapy and psychiatric consultation is necessary in most cases.

Hyperekplexia

This disease is transmitted by an autosomal dominant trait is caused by a mutation in the alpha1 subunit of the glycine receptor. The glycine receptor is encoded in the long arm of the chromosome 5 in the 5q33-q35 chromosomal locus.

Familial cases demonstrate point mutations in the alpha-1 subunit of the inhibitory glycine receptor (GLRA1). The glycine receptors have higher density in the brainstem and spinal cord. The disease is characterized by an exaggeration of the normal startle reflex. Hyperekplexia is a term originary from the Greek meaning exaggerated jumping/jerking.

Andermann et al. (1980) described the details of the normal startle reflex which consists of alerting reaction associated with eye blinking, facial grimacing, flexion of the head, elevation of the shoulders, and flexion of the elbows, trunk, and knees. The authors call attention to the fact that tension, fatigue, will heighten and repeated stimulation will decrease the reaction. The startle reflex appears early in infancy.

This condition is characterized by episodes of excessive startle after sudden tactile, auditory (loud noises) or visual stimuli. Stimuli of similar intensity usually fail to produce a similar response in most normal individuals. The attacks are associated with increased muscle tone and spontaneous clonus. In some cases patients may have loss of muscle tone with a subjective sensation of stiffness. Patients with hyperekplexia the exaggerated startle reaction may be followed by fall, which may be due to loss of postural control.

The physiopathology of this disease is thought to be related to an impairment of reciprocal inhibition at the level of the spinal cord.

The neonatal presentation of hyperekplexia is the period has been called “major form”, stiff-baby syndrome or stiff-man syndrome in the newborn. The neonatal hyperekplexia is characterized by hypertonia starting on the first day of life (especially around the shoulder girdle) and excessive startle which can be produced by tapping the tip of the or the glabella (glabellar tap) as well as by feeding but may occur spontaneously. A clinically useful maneuver is to tap the tip of the nose or the glabella. The glabellar tap will elicit a non-habituating exaggerated startle response in almost all affected individuals. This startle reaction leads not only to an increase in muscle tone but may cause apnea in infants. Frequent choking and swallowing difficulties may be seen too. Over time the patients develop hyperekplexia, gait ataxia, rhythmic clonus and are commonly mistaken for cases of spastic cerebral palsy. Sudden death may occur in infancy starting from the neonatal period possibly due fatal apnea. In older patients the startle reaction may cause falls, which are not unlike, drop attacks except for the EEG (see below). Often the patients’ falls are
associated with a loss of postural control causing loss of protective mechanisms or fall “en statue”.6 Hypertonia tends to disappear overtime65,66 and it is absent during sleep. Due to facial muscle involvement the patients may have the appearance of being frightened or tense. Transition from awake to sleep state produces the normalization of the baseline tone patients will often have violent and repetitive myoclonic jerks of the limbs especially during quiet sleep in some cases this pattern is so dramatic that the child is lifted off the bed.55,67 This dramatic pattern starts often when the patients baseline awake tone starts to decrease, towards the end of the first year of life55 but cases with onset in the neonatal period have been described.63,68 Insecure and hesitating gait may be also present in patients with the major form of hyperekplexia.

A chronic increase in the tone may lead to hip dislocation as well as increased intra-abdominal pressure and subsequent umbilical, inguinal and diaphragmatic hernias formation.6,66,69

A “minor form” which begins in infancy has been described. In these cases as the patients get older emotional stress plays a major role. Exaggerated startle is often isolated and may be inconstant. One patient was reported with onset in adolescence that responded well to valproic acid.70 In some pedigrees the “major and minor forms” may coexist in the same family and sporadic cases have also been described.71

Neurophysiologic studies demonstrate that hyperekplexia is not only an exaggerated normal startle response since the EMG latencies are shorter than normal.72 The EEG may show centro-parietal spikes followed by slowing and desynchronization during the events.55 The EEG’s changes have been questioned to be due to artifact and/or not related to seizure activity. Somatosensory and auditory evoked potentials have been described to be increased or normal in this disease.55,72,73

The author agrees with the other authoritative sources that the relationship of hyperekplexia with other nonepileptic startle disorders, such as jumping (jumping Frenchmen of Maine), latah (ticklishness associated with echopraxia and coprolalia in Malaysia), and myriachit (to act foolishly as in Siberia and other parts of Asia), remains speculative at this point.20,55 An exaggerated startle response may be a component of reflex epilepsy (startle epilepsy), which is outside of the scope of this review.55

Valproate and clonazepam are the treatments of choice for hyperekplexia. Clonazepam is used in relatively high doses 0.1 to 0.2 mg/kg/day but symptoms may only partially controlled.9 Clonazepam may help prevent neonatal problems.60 At times the episodes of increased tone may not respond to benzodiazepines but forced flexion of the legs, head and trunk may abort them.63,67 It is possible that familial cases are more likely to respond to clonazepam.50 In some cases in which clonazepam proved ineffective valproic acid, 5-hydroxytryptophan, or piracetam may reduce abnormal startle.74 Valproic acid has been helpful in cases of late onset.70 Valproic acid is usually started on a dose of 15-20 mg/kg/day, hematologic and liver toxicity can be seen in any patient taking this drug and it is especially common with polytherapy and below age 2 years. Even though one is tempted to say that clonazepam and valproic acid exert their effect through their actions in the GABA receptor52 another GABAergic drug Vigabatrin was not effective in controlling the startle reactions.75

The prognosis is variable and the main concern in neonatal hyperekplexia is apnea. Early identification and treatment may help improving the outcome in these cases. There is a tendency for the neonatal form to improve spontaneously during the two years of life, although later on but delayed motor development is often seen.20,61 In some cases hyperekplexia may persist into adult life. Complete heart block has also been described in one case of non-familial hyperekplexia.76

Sandifer syndrome

Children with hiatus hernia, gastroesophageal reflux or esophageal dysmotility may have contortions of the neck associated abnormal postures.77-79 In spite of the abnormal neck position and movements the tone of the neck muscles is not increased.6 Some of the head and neck movements resemble in part those seen in adverse seizures. In some cases head nodding and gurgling sounds may be seen.6 In one study of 126 infants and children with gastroesophageal reflux 7.6% had Sandifer’s syndrome.80 This pattern of movements has been named Sandifer syndrome consist of sudden extension or torsion spasms of the neck and it is often associated with continuous twisting of the head from side to side.78 This pattern of movements be mistaken by dystonia may be made. Less dramatic patterns often go unrecognized and undiagnosed and it is not uncommon for the patients to carry another diagnosis such as dystonia or epilepsy are entertained before the Sandifer Syndrome is diagnosed.81,82

The presence of hiatus hernia or reflux in association with such a dyskinesia allows the correct diagnosis to be made and appropriate therapy to be given. Medical treatment may not be sufficient and fundoplication is often necessary.

The recognition of Sandifer’s syndrome may lead to effective medical or surgical treatment (fandoplication) of gastroesophageal reflux. Sandifer syndrome should be included the differential diagnosis of pediatric patients with presuptive paroxysmal torticollis, paroxysmal dystonia of the neck without palpable increased tone of the corresponding muscle groups, or adverse seizures without alteration of the level of consciousness.
Dystonia and hypermotor movement disorders

The typical dystonic reactions are not very hard to differentiate from epileptic seizures nonetheless there is a border-zone of symptom overlap between epilepsy and dystonia. In dystonia there is co-contraction of agonist and antagonist muscle groups often leading to twisting of body parts. Dystonias are not associated with impairment of consciousness but may be preceded by a subjective sensation over the body part affected. Some cases of frontal lobe epilepsy there is a dystonic-type motor phenomenon and loss of consciousness is not an obligatory the partial seizure in that epileptic syndrome. Patients with hemiplegia may develop dystonia too. One of my personal patients had a large congenital infarct in the left internal carotid territory and the cavitation which formed after the stroke made remaining functional cortex three centimeters or more removed from the internal skull surface. The boy had a history of partial seizures with dystonic posturing leading to secondary generalization but he had also some plain dystonic posturing without any EEG changes. The lack of EEG changes could have been due to the large distance between cortical generators and the recording electrodes. It took a while and some therapeutic trials of antiepileptic medications to get to the conclusion that he had two problems partial seizures and dystonia.

Clinically dystonia and hypermotor movement disorders may affect only one of a few limbs but trunkal stiffness, hyperextension of the head, opisthotonus, oculogyric crises may be seen as well. Oculogyric crises are frequently related to drugs use. Among these drugs are psychotropic drugs such as phenothiazines and butyrophenones and metoclopramide. Many other agents have been reported to induce movement disorders this is especially true of the drugs with DOPAminergic effects. Young males are more prone to dystonic reaction after exposure to psychotropic medications such as haloperidol. Weaning patients after treatment with haloperidol or other dopamine agonist drugs may cause emergent withdrawal reactions which include oro-facial dyskinesias, dystonia and akathisia. Akathisia is a sensation of restlessness which accompanied by the need to move incessantly. The diagnosis can be confirmed by urine examination for the commonly responsible drugs. In most acute drug reactions are self-limited and the treatment with intravenous anti-histaminics such as diphenhydramine (1 mg/kg/dose intravenous slowly) can control dystonic or dyskinetic symptoms and oculogyric crises, but in some episodes may not respond to high doses.

Other causes of dystonia include intracerebral lesions (tumors, vascular malformations, parasitic cysts) and more rarely the attacks may be caused by a mastocyctoma. Dystonia which is not related to drug use may respond to anticholinergic drugs. These medications are rather toxic and should only be used a trained Pediatric Neurologist since they are associated with a multitude of side-effects such as dry mouth, constipation, postural hypotension, drowsiness etc. In the recent years the recommended initial treatment for dystonia to be a trial L-DOPA. The rationale for using L-DOPA as the first-line therapy is to rule-out an infrequent syndrome named DOPA-sensitive dystonia (a.k.a. Dystonia with diurnal variation or Segawa Syndrome).

Patients with the latter syndrome typically have dystonia which gets worse or appears in the afternoon by but many atypical cases can be diagnosed with the empirical treatment with L-DOPA. The onset of dystonia is typically in the preschool years but atypical cases have been described. Untreated cases develop parkinsonism in adolescence. The L-DOPA dose is dependent on the patient’s weight and it is usually given in association with a peripheral DOPA carboxylase inhibitor such as carbidopa to avoid nausea. The initial dose in pre-school children in 50 mg of L-DOPA but the dose can be increased to 250 mg if some response is seen. In most cases the response is obtained with low dose but atypical cases may required up to 750 mg/day.

The second line of treatment of dystonia (for patients who fail L-DOPA trial) is usually one of the anti-cholinergic agents such as trihexyphenidyl on an initial dose of 1-2 mg/ day increasing by 1-2 mg/week up to doses of 40-80 mg/day if tolerated. Anti-cholinergic type of side-effects are quite common specially with the higher doses including dry mouth, difficulty initiating micturition, tachycardia, precipitation of glaucoma etc. Other alternatives in the treatment of dystonia include baclofen (initial dose 2.5 mg tid increase up to 30 mg tid by 2.5 mg/day/week), clonazepan (initial dose 0.01 mg/ kg/day increase to up to 0.2 mg/ kg/ day), valproic acid (see Hyperekplexia above) and carbamazepine on an initial dose of 5 mg/kg/day increased to up to 30 mg/kg/day.

Chorea and tics are often easily differentiated from seizures. The exception are cases of simple motor tics which can be confused with myoclonus, in these cases an EEG easily differentiate cortical/epileptic myoclonus from tics. Movement disorders do not need to be treated in all patients. In many cases the tics a mild and transient and do not need treatment. When the tics are very socially disruptive they can be treated initially with alpha-receptor agonist such as guanfacine (0.5 mg bid) or clonidine on an initial dose of 0.05 mg/day increased to up to 0.1 mg tid. We routinely obtain an ECG before initiating treatment with alpha-agonists. Patients who do not respond to guanfacine or clonidine need treatment with dopamine antagonist which are drugs with more complex toxicity including potentially irreversible side-effects such as tardive dyskinesia and akathisia that are better prescribed in consultation with a experienced Pediatric Neurologist or Psychiatrist.

Spasmus nutans

Spasmus nutans is characterized by the asymmetric ocular oscillations, head nodding, and anomalous head positions (tilting) with onset in infancy between ages 1-15.
months.\textsuperscript{20,91-93} Asymmetric nystagmus and head nodding may be missed during the exam and many cases of spasmus nutans often do not demonstrate all of the typical diagnostic features.\textsuperscript{20,91-93} The nystagmus in spasmus nutans can be conjugate (binocular), monocular or dissociated.\textsuperscript{94} The nystagmus in these children is enhanced by the presentation of a visual target (baby toy or colored books).\textsuperscript{91} Much of the pathophysiology of spasmus nutans is unknown but the cause of the head nodding is a normal compensatory oculovestibular reflex.\textsuperscript{91} This conclusion is secondary to the fact that infants with spasmus nutans. Speculations that spasmus nutans results from deprivation of sunlight have not been confirmed.\textsuperscript{20} Even though the typical syndrome of spasmus nutans is thought to be often benign and self-limiting condition but other more serious conditions may have some of the symptoms and need to be differentiated before one makes the correct diagnosis.\textsuperscript{20} Among the "secondary" causes of spasmus nutans are cases of congenital nystagmus, chiasmatic lesions,\textsuperscript{94} diencephalic syndrome, porencephalic cysts, opsoclonus-myoclonus,\textsuperscript{95} empty sella, ependymoma, and retinal disorders.\textsuperscript{20,91-93}

One compared the clinical findings in spasmus nutans, with spasmus nutans-like disease (CNS lesions and congenital nystagmus).\textsuperscript{96} These authors found that in the spasmus nutans group the mean onset of nystagmus and head nodding was 8 months, and the mean onset of head tilt was 15 months. Nystagmus was asymmetric, intermittent and had a later onset in spasmus nutans compared with the other causes of infantile nystagmus. The optokinetic nystagmus was frequently present in spasmus nutans and absent in the majority of cases infantile nystagmus. Head nodding was also more frequent, of larger amplitude, and clinically easier to detect in spasmus nutans. Head tilt was not found to be a useful differentiating criterion.

Long-term follow-up is important to confirm the diagnosis of spasmus nutans since most patients attain good visual acuity but subtle nystagmus may persist until age 12 years.\textsuperscript{93}

Tetany

Tetany is in most cases due to hypocalcaemia or hypomagnesemia or both. The term is used to designate increased muscle tone due to increased excitability of the muscle membrane due to low calcium and magnesium concentrations. Sometimes the total serum calcium may be normal but ionized calcium level is low a condition which may be due to alkalosis secondary to hyperventilation or repeated vomiting such as in pyloric stenosis.\textsuperscript{6} In third world countries babies who are fed cow’s milk improperly diluted may develop hypocalcaemia secondary to the excess phosphorus in the feedings. Vitamin D deficiency is now a rare cause of tetany. In developed countries congenital (inborn errors of calcium and vitamin D metabolism) and postoperative hypoparathyroidism and pseudohypoparathyroidism are among the most frequent the causes of hypocalcaemia.\textsuperscript{6}

The classical tetany symptoms often do not appear before the age 3 months. Carpopedal spasm appears abruptly and affecting mostly the fingers, which become flexed at the proximal joint and extended at the distal joints, with the thumbs adducted and opposed. The feet become tonically extended. No alterations in consciousness are noted which is a key differentiating factor in older children. Laryngospasm may occur with or without carpopedal spasm and it is diagnosed when inspiratory stridor is heard due to vocal cord adduction. Latent tetany may be tested with the Chvostek sign, which is the presence of facial muscle contraction after tapping the zygomatic arch region. Chvostek sign may occur in some normal children.\textsuperscript{6} Hypoparathyroidism and pseudohypoparathyroidism may be also associated with headaches, extrapyramidal signs and calciﬁcation of the basal ganglia.\textsuperscript{97} Pseudohypoparathyroidism is associated with short metacarpals, moon-shaped facies, enamel defect, decreased olfaction and auditory acuity, besides other nonspeciﬁc signs such as mental retardation, cataracts, obesity and decreased height.\textsuperscript{6,98} Epileptic seizures can occur at any age in patients with hypocalcaemia and are more commonly observed than the classical muscle spasms of tetany.\textsuperscript{6} In infants especially in neonates, bona ﬁde convulsions that are persistently focal may be seen due to hypocalcaemia. The treatment of tetany consists of parenteral administration of calcium which requires careful monitoring.\textsuperscript{99,100} Calcium given by an umbilical vein catheter may cause liver necrosis if the tip of the catheter is not pushed into the inferior vena cava, the infusion site should be checked frequently since extravasation of calcium into the subcutaneous tissues may cause severe necrosis.\textsuperscript{99,100} Calcium chloride causes more severe reactions when extravasation occurs thus calcium gluconate is the preparation of choice.\textsuperscript{99,100} Calcium should not be mixed with sodium bicarbonate in the same vial or infusion line since precipitation of calcium carbonate in the solution may occur. Animal experiments suggest that calcium infused quickly through the aorta decreases the intestinal perfusion so slow infusion should be also used with umbilical artery catheters.\textsuperscript{99,100} Acute therapy of symptomatic hypocalcaemia (tetany, seizures and apnea) should be 1-2 ml/kg of 10% calcium gluconate (1 ml of the Calcium gluconate 10% solution = 100mg = 9mg elemental Ca) by intravenous infusion over 5 minutes under cardiorespiratory monitoring.\textsuperscript{99,100} The dose may be repeated if there is no clinical response and the serum calcium remains low. Ionised Ca correlates more reliable of neuron membrane irritability and should be used whenever possible to guide the beginning and termination of acute therapy especially in neonates since in this age group total serum Ca correlates poorly with ionized Ca\textsuperscript{99,100}. Vitamin D and magnesium are indicated when low levels are documented.
Apnea-bradycardia episodes, Sudden Infant Death Syndrome/Apparently Life-Threatening Events (ALTE)

In many instances patients presenting with apnea may be thought to have seizures. Apnea as the sole manifestation of seizures is rare but may occur infants especially full term neonates.\(^{101-102}\) Commonly neonates with seizure related apnea eventually show other signs such as tonic eye deviation or clonic activity. Apnea of preterm infants is usually due to respiratory centers immaturity an it is only rarely seizure related. A full discussion of all causes of apnea is beyond the scope of this review (see reference 6 for review). A partial list of the common causes of apnea is neonates and infants includes: acute presentation of asphyxia, neonatal seizures (with or without asphyxia), spinal cord and brain birth injury, closed-head injury (accidental), non-accidental trauma (shaken-baby syndrome), acute sepsis, shock-dehydration, respiratory infection (for example respiratory syncicial virus-RSV), mechanical cord compression (ostegogenesis imperfecta), increased intracranial pressure, hypokalemia, arterial hypotension, feeding intolerance, hypoglycemia, metabolic complications, congenital heart disease, congenital malformations, and other causes such as cerebral palsy, central nervous system disorders, and neuromuscular disorders. Apnea of the newborn has been estimated to occur in about 2% of live births, and is not uncommon. Apnea of preterm infants is usually due to respiratory centers immaturity, and it is only rarely seizure related. A full discussion of all causes of apnea is beyond the scope of this review (see reference 6 for review).

Sudden infant death syndrome (SIDS) and other ALTEs are a heterogeneous group of events due to the association of apnea with hypotonia or near-miss SIDS cases may be easily mistaken for epileptic events due to the association of apnea with hypotonia or stiffness and cyanosis or pallor. Severe cases may produce hypotensive and hypoxic-ischemic injuries and may be followed by epileptic seizures.\(^{108}\) The majority of the recent publications have found no clear-cut correlation between polygraphic recordings and the later occurrence of sudden death (see reference 6 for review). The indications and usefulness of home electronic monitoring for apnea remains unclear.\(^{110}\)

Paroxysmal vertigo

A syndrome of recurrent brief (one to several minutes) attacks of vertigo and nystagmus, which come on suddenly without known precipitant has been described in children. The onset of symptoms is between the ages 1-5 years and boys and girls equally affected.\(^{111}\) The usual frequency of attacks is low usually one to three per month but in some case several per day have been reported.\(^{20,112}\) Most children cease having attacks before age 7 years and in many cases less than five attacks are recorded in their life. During the attacks the child appears in pale and frightened but conscious, and may stagger and even fall. The patients will often hold onto something (usually furniture or someone) at the onset of the event or she/he will lie down on the floor, refusing to move for the duration of the symptoms.\(^{112}\) In most cases the patients have normal examinations are negative, but abnormalities in inner ear/labyrinthine function have been reported.\(^{111}\)

The relationship between childhood migraine and benign paroxysmal vertigo remains speculative but a positive family history of migraine among immediate relatives is not uncommon. That possibility that benign paroxysmal vertigo represents a “migraine equivalent” is supported by cases of children with the typical syndrome with or without associated headache that subsequently develop classic or vertebrobasilar migraine (reference 20; and several of my personal cases).

Benign paroxysmal torticollis of infancy

Benign paroxysmal torticollis of infancy is probably closely related to paroxysmal vertigo but it may be have different etiologies.\(^{113,114}\) The symptoms appear during the first year of life and may even be present in the neonatal period more often between ages 2-8 months.\(^{20}\) The duration of the attacks is usually several hours but they may be short (10 minutes) and may last for as long as 14 days.\(^{20}\) Clinically twisting of the neck (wryneck or torticollis) is seen without persistent or obligatory abnormal head positioning and subsequent spontaneous resolution is the rule in this syndrome.\(^{20}\) "Spread" of the dystonic posturing to the trunk may be seen in some cases.\(^{115}\) Associated symptoms include repeated vomiting, discomfort and tilting of the head to one side (may change with successive attacks) and abnormal eye movements. In some cases the attacks may be heralded by a period of irritability, distress, or vomiting.\(^{20}\) More extreme cases have been described in which one sees inclination of the trunk to the side sometimes ipsilateral stiffness, ataxia veering toward the side to which the head
is tilted. In these extreme cases a CNS tumor needs to be ruled-out unless there is a clear-cut history of previous similar episodes, which subside leaving a completely normal child free of any neurological anomaly. During the assessment of the first episode head imaging is commonly necessary. Familial clustering has been reported.

Other causes of torticollis must be ruled out including ocular, pain-related, contracture-related, dystonic, Sandifer’s syndrome. CNS tumors including those located in the posterior fossa, third ventricle (colloid cyst) and spinal cord may also produce intermittent torticollis. Syringomyelia may also have similar symptoms. One of my personal cases presented with intermittent tonic posturing of the neck as a toddler which lead to an investigation for dystonia and seizures. “Ocular torticollis” resolves with supine position or occlusion of paretic eye and it is present in the sitting position. Trauma and infection-related lymphadenitis may also produce pain and abnormal neck posturing. Paroxysmal torticollis can be differentiated from contractures since the head can be passively returned to the neutral position but the tilting recurs immediately. Like in the case of paroxysmal vertigo a relationship to migraine has been suggested for paroxysmal torticollis. Typical migraine has been seen in cases of torticollis as the child grows older. Other etiologies of vertiginous episodes may occur in children such as drug intoxication or acute labyrinthitis of infectious origin need to be taken in consideration when making the diagnosis of paroxysmal vertigo. Most cases of Benign Paroxysmal Torticollis of Infancy recover before age 2 to 3 years without medical intervention.

Paroxysmal kinesigenic choreoathetosis

In this disorder sudden movement precipitates uni or bilateral dystonia and/or chorea spells which may repeat up to 100 times. The attacks may be preceded by an aura of tightness or tingling in the affected segment. No loss of consciousness is noted during in the events, which last from one to a few minutes. The movements may be bizarre, writhing dystonia, choreic or even and ballistic in nature and sometimes cause falls. Other stimuli which have been described to precipitate the attacks include stress, hyperventilation, excitement and startle. Cases precipitated by methylphenidate have been described. Three-quarters of the cases of this disease are familial and transmitted by a dominant trait. The other ¼ of the cases probably represent denovo mutations. One needs to be careful with this diagnosis since cases which have been thought to be paroxysmal kinesigenic choreoathetosis undergoing long-term video-EEG monitoring have been shown to have an electroencephalographic seizures originating from the supplementary sensorimotor cortex and ipsilateral caudate nucleus.

Paroxysmal kinesigenic dyskinesia in many cases responds well to anticonvulsant drugs, especially those which act by inactivation of the sodium channel such as carbamazepine and phenytoin even in low doses. Patients affected by paroxysmal kinesigenic choreoathetosis have normal longevity but the attacks in public may lead social embarrassment.

This syndrome is usually treated initially with carbamazepine 5 mg/kg/day some patients can respond to even lower doses but others may required slight higher doses in the range used for children with seizures (15-25 mg/kg/day). In some resistant cases, flunarizine has been successful in controlling of the attacks but totally unresponsive cases have been reported. Flunarizine is usually started on a dose of 2.5-5mg/day in children and increased as tolerated to up to 0.5-1mg/kg/day.

Paroxysmal non-kinesigenic (dystonic) choreoathetosis

Attacks similar to the ones described under “Paroxysmal Kinesigenic Choreoathetosis” (see above) but triggered by stress, coffee or alcohol ingestion have been described. Attacks are usually longer than the ones see in the kinesigenic forms often lasting from several hours on the other hand they occur less frequently. This condition is also familial and transmitted by a dominant trait but is less common. The non-kinesigenic for of paroxysmal choreoathetosis does not usually improve with anticonvulsant agents, with the exception of clonazepam and oxazepam. One patient who also had familial ataxia responded positively to acetazolamide. The initial clonazepam dose is 0.01 mg/kg/day which is increased to 0.1 mg/kg/day over a several weeks, sedation is often one of the most limiting side-effects.

Other idiopathic paroxysmal dyskinesias

Lance described also intermediate cases that did not fit either the kinesigenic or the non-kinesigenic paroxysmal choreoathetosis groups. These odd cases are often induced by sustained exercise with attacks lasting 5-30 minutes but cases of have been reported. Wali reported also hemidystonia induced by prolonged exercise and cold. A rare form of autosomal dominant paroxysmal choreoathetosis with spasticity has been described and recently mapped to chromosome 1p near the potassium gene channels cluster.

Paroxysmal dystonia of infancy

Angelini et al. described a paroxysmal dystonia in infants aged between 1 and 5 months. Clinically the
episodes are characterized by brief, frequently and repeated episodes of dystonia of the upper limbs (symmetrical or asymmetrical) and/or opisthotonus. The symptoms abated over time and disappear altogether before the end of the second year of life.126

References