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Original Articles

Paroxysmal Non-epileptic Movements in Childhood

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Abstract Paroxysmal movements in childhood can be epileptic and non-epileptic. Non-epileptic events comprise a wide range of disease entities. It can be challenging for a clinician to make an accurate diagnosis. This review article will focus on the clinical approach to various non-epileptic paroxysmal movements in childhood with particular emphasis on detailed history taking and examination of the abnormal events. Prompt diagnosis is extremely important for commencement of appropriate treatment if indicated and to avoid unnecessary use of medications including anticonvulsants in certain benign non-epileptic self-limiting conditions. Timely parental counselling is also important to reduce unnecessary anxiety.

Key words Childhood; Epileptic; Movements; Non-epileptic; Paroxysmal

Introduction

Paroxysmal phenomena during childhood cause intermittent motor, behavioural or other somatic symptoms or signs. Paroxysmal occurrence means there is always a return to baseline, with resulting sign-free intervals.[1] There is a wide variety of presenting symptoms such as abnormal movement, collapse and / or loss of consciousness, respiratory irregularities, headache, abdominal pain, vomiting, dizziness, vertigo, sleep-related phenomena and emotional or psychiatric problems. Paroxysmal movements refer to paroxysmal occurrence of abnormal movements which are caused by different disorders listed in Table 1. An accurate and early diagnosis of a specific disorder can be challenging as many of the conditions listed in Table 1 can mimic an epileptic seizure. This review will focus on the approach to paroxysmal non-epileptic movements in childhood which may have caused diagnostic challenges to clinicians.

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Seizures or Seizure-like Conditions

Seizures

In 2005, the International League against Epilepsy (ILAE) defined an epileptic seizure as a transient occurrence of signs and / or symptoms due to abnormal excessive or synchronous neuronal activity in the brain.[2] One year later, a report on the ILAE Classification Core Group discussed on various epileptic seizure types. They can present as abnormal focal or generalised movements which could be tonic (continued muscle spasm), clonic (rhythmic 1-2 Hz events), myoclonic (single or irregular recurrent events), hypermotor (bilateral forceful limb movements),[3] automatisms (lip smacking, chewing, tooth grinding, swallowing, semi-appropriate / repetitive motor activities) or dystonic posturing.[4] Sleep is a possible aggravating factor for seizures.[5]

Convulsive Syncope

However, syncope can have a convulsive element which mimics an epileptic seizure. Syncope is an abrupt, transient,

Table 1 Causes of paroxysmal movements in childhood

Paroxysmal movements in childhood
Seizures / seizure-like conditions
1. Epileptic seizures
2. Convulsive syncope
3. Psychogenic non-epileptic seizures
Hyperkinetic, paroxysmal movement disorders
1. Tremor
2. Ataxia
3. Myoclonus including exaggerated startle response e.g. hyperekplexia
4. Tics
5. Stereotypies
6. Paroxysmal dyskinesias
7. Psychogenic paroxysmal movement disorders
Transient and developmental movement disorders in children
1. Jitteriness
2. Shuddering
3. Benign paroxysmal torticollis of infancy
4. Infantile masturbation
5. Benign paroxysmal tonic upward gaze
6. Sandifer syndrome
7. Breath-holding attacks
8. Spasmus nutans
Sleep-related disorders
1. Parasomnia
a. Confusional arousals
b. Sleep terror
c. Sleep walking
d. Rapid-eye-movement sleep behavioural disorders
2. Restless leg syndrome

and self-limiting loss of consciousness associated with loss of postural tone, caused by a sudden fall in cerebral perfusion.[6] Myoclonic jerks are common in syncope.[7] However, these jerks are not cortical in origin but are thought to be a brainstem phenomenon.[7,8]

Causes of syncope are reviewed by Crompton and Berkovic, which can range from relatively benign conditions such as vasovagal syncope to cardiac diseases that can be rapidly fatal. A detailed history taking can usually distinguish convulsive syncope from epileptic seizures. Syncope can occur within the setting of prolonged sitting or standing, rising to upright posture or dehydration whereas an epileptic seizure may be precipitated by stress, sleep deprivation, pain, fright, micturition, defaecation, photic triggers etc. Nausea, palpitations, dyspnoea, warm sensation, light-headedness, graying of vision or hearing becoming distant are possible prodromal symptoms of syncope. If an epileptic seizure is partial at onset, symptoms may indicate a frontal, parietal, temporal or occipital focus. During an attack, syncope can cause sudden collapse with pallor while an epileptic seizure may have tongue biting, unusual body posturing, incontinence or cyanosis. After the attack, patients with syncope can remember the event but those with an epileptic attack may develop post-ictal confusion, headache and they may not recall the event before and during the event.[6] Electroencephalographic recordings during syncope vary from high-amplitude slow activities to marked attenuation of cortical activity. No epileptiform or ictal discharges are observed.[9,10] Children diagnosed to have convulsive syncope should not receive anticonvulsants. Management is directed towards the underlying aetiology causing syncope. It may be prudent to perform a complete cardiac evaluation when cardiogenic syncope is suspected.

Occasionally, syncope may provoke a true epileptic seizure i.e. anoxic-epileptic seizure. Horrocks et al reviewed a cohort of 27 children with this diagnosis and the epileptic component could be bilateral clonic in almost all children. Sometimes there was horizontal eye deviation or rhythmically interrupted vocalisation. The duration of the syncope triggered epileptic seizures varied from 28 seconds to 40 minutes. Electroencephalographic recording was performed in 4 patients and demonstrated the epileptic nature of the events. Acute and / or prophylactic treatment with anticonvulsants was initiated in some children whose epileptic seizures were prolonged and / or recurrent respectively.[11]

Psychogenic Non-epileptic Seizures

Psychogenic non-epileptic seizures usually begin with external (place, time, witness) or internal triggers (flash backs, emotions). The eyes are commonly closed in these events but uncommon in epileptic seizures. Seizure seminology is commonly stereotypic in epileptic seizures and less common in psychogenic ones. It is not uncommon for epileptic seizures to have onset at sleep, urinary incontinence, injuries but rather uncommon for psychogenic non-epileptic seizures. Patients with epileptic seizures can rarely recall for the period of unresponsiveness while those with psychogenic ones commonly can.[12-14] Prompt recognition of psychogenic non-epileptic seizures is important to minimise the inappropriate use of

anticonvulsants and optimise the multidisciplinary team approach on psychotherapy and / or pharmacotherapy. However, it could be challenging when both epileptic and psychogenic non-epileptic seizures co-exist in the same patient. In a review, up to 40% of patients with epilepsy have concomitant psychogenic non-epileptic seizures.[15]

Hyperkinetic Paroxysmal Movement Disorders

A posture is a period of non-zero (but possibly very short) duration in which minimal movement occurs. A discrete movement is any movement that departs from and subsequently resumes a posture with no intervening postures.[16] Hyperkinetic movement disorders are defined as unwanted or excess movements that are frequently seen in children with neurologic disorders. These include dystonia, chorea, ballism, athetosis, myoclonus, tremor, tics, stereotypies[17] and ataxia.[18]

Tremor

Tremor is a rhythmic back-and-forth or oscillating involuntary movement about a joint axis with a relatively symmetric velocity in both directions about a midpoint of the movement, and the velocity or oscillation may appear sinusoidal. Tremor is labeled as a rest tremor, postural tremor, or action tremor according to the condition of greatest severity. Other than resting tremor, postural tremor or action tremor has a paroxysmal nature with sign-free intervals because the occurrence is situation-specific.

Ataxia

Ataxia is defined as an inability to generate a normal or expected voluntary movement trajectory that cannot be attributed to weakness or involuntary muscle activity about the affected joints.[19] Specific associated deficits include dysmetria (inaccurate movement to a target – undershoot or overshoot), dyssynergia (decomposition of multijoint movements), and dysdiadochokinesis (impaired rhythmicity of rapid alternating movements).[17] Children presenting with ataxia usually do not have significant diagnostic difficulties. Causes of episodic ataxia can cover a wide range of neurogenetic and neurometabolic diseases.[20,21] Table 2 gives a summary of the major types of autosomal dominant genetic episodic ataxias. A detailed history with respect to the age of onset, duration of attacks, additional manifestations during attacks and interictal manifestations can help clinicians to differentiate among these disorders. Neurometabolic diseases can also give rise to episodic ataxia such as during acute / subacute decompensations of amino acids, organic acid and urea cycle disorders. Usually these disorders have other neurological or non-neurological markers. By contrast, defects of energy metabolism may involve exclusively the nervous system and may be more difficult to diagnose e.g. pyruvate dehydrogenase deficiency, and biotinidase

Table 2 Major types of autosomal dominant episodic ataxias (EA)

	Age of onset (years)	Duration of attack	Additional manifestations during attack	Interictal manifestations
EA 1	2-15	Seconds, minutes	None	Myokymia and jerky movements of face and limbs
EA 2	2-20	Hours, days	Downbeat nystagmus, migraine, vertigo, nausea, vomiting, weakness, dysarthria	Ataxia, nystagmus
EA 3	1-42	1 minute to 6 hours	Myokymia, migraine, tinnitus, vertigo, dysarthria	None
EA 4	23-60	Brief	Vertigo, diplopia, interictal nystagmus and abnormal smooth pursuit	Nystagmus
EA 5	3-19	Hours	Vertigo	Nystagmus, ataxia, epilepsy
EA 6	<20	Hours, days	Cognitive impairment	Epilepsy, migraine, ataxia, motor delayed milestones
EA 7	13-19	Hours, days	Vertigo, weakness, slurring, dysarthria	None

Modified from Finsterer J. Ataxias with autosomal, X-chromosomal or maternal inheritance. Can J Neurol Sci 2009;36:409-28.[20]

deficiency. Carbohydrate- and fever- sensitive ataxia is a feature of the former while the latter will have high lactate and other characteristic biochemical features present intermittently. Glucose transporter 1 deficiency usually worsens before meals. Some children improve greatly with frequent carbohydrate-rich snacks. Intermittent ataxia can also be seen in Hartnup disease; additional symptoms are pellagra-like skin changes, photic dermatitis, and psychiatric symptoms. Recently a new inborn error of metabolism caused by urocanic aciduria can cause intermittent ataxia and mental retardation.[22] A combination of detailed history taking, physical examination and appropriate neurometabolic investigations would confirm a specific inborn error of metabolism.

Myoclonus

Myoclonus is a sequence of repeated, often nonrhythmic, brief shock-like jerks due to sudden involuntary contraction or relaxation of one or more muscles. Myoclonus can be epileptic and / or non-epileptic, including various benign forms of myoclonus related to sleep e.g. hypnagogic myoclonus and benign neonatal sleep myoclonus.

Exaggerated Startle Response Including Hyperekplexia

Startle is a stereotypical response to a sudden and unexpected stimulus. In most instances, the stimulus is acoustic, but other modalities such as tactile, visual, or vestibular are also effective. Similar amongst all mammals, the response is composed of motor, autonomic, and emotional components. The motor component of startle satisfies the criteria for myoclonus.[23] It is a basic alerting reaction consisting of facial grimacing with blinking, followed by involuntary movements of head flexion, hunching of shoulders, adduction of the arms, and flexion of the trunk and the knees, causing falling without a protective reaction.[24,25]

When a pathologically exaggerated startle response interferes with normal activities, causing apnoea and frequent falls and injuries, the pathological state is termed as startle disease or hyperekplexia.[25,26] Hereditary hyperekplexia (or startle disease) may manifest shortly after birth (stiff-baby syndrome) with violent jerking to noise and touch, and massive and sustained stiffening of the trunk and limbs, clenching fists, and attacks of a high frequency trembling. Newborns are at risk for sudden infant death due to laryngospasm and cardiorespiratory failure.[23]

Exaggerated startle response can also be seen in other conditions such as startle epilepsy. The majority of patients with startle epilepsy suffer from congenital or infantile brain damage with concomitant spastic hemi-, di-, or tetraplegic cerebral palsy, mental retardation, and frequent spontaneous epileptic seizures. As in common startle, unexpected noise is the most frequently effective modality. Startle seizures typically exhibit the characteristics of tonic seizures: uni- or bilateral limb posturing, turning of the head, speech arrest. In many cases, consciousness is at least partly preserved.[23]

Tics

Tics are repeated, individually recognisable, intermittent movements or movement fragments that are almost always briefly suppressible and are usually associated with awareness of an urge to perform the movement. The movements are predictable by an observer in the sense that there is often a small and an identifiable number of different tic movements, and they are predictable by the child due to the presence of a premonitory urge to move. A characteristic feature of tics is intervening periods of normal movement. Each child has one or more recognisable tics that are repeated in a varying sequence. Individual tics have very little variability between repetitions. The sequence may appear random, but particular movements can often be identified and counted.

In many children, attempts at suppression of a tic lead to an increasing build-up of an urge to make the movement, often accompanied by a sensory premonition.

Stereotypies

Stereotypies are repetitive, simple movements that can be voluntarily suppressed. Stereotypies are typically a simple back-and-forth movement such as waving or flapping the hands or arms, and they do not typically involve more complex sequences or movement fragments. Movement is often but not always rhythmic and may involve fingers, wrists, or more proximal portions of the upper extremity. The lower extremity is not typically involved. Stereotypies can be unilateral or bilateral but are more commonly bilateral. There is probably no premonitory urge, and the movements tend to occur when the child is stressed, excited, distracted or engrossed. Stereotypies can be stopped by distraction or initiation of another activity. Factors distinguishing tics and stereotypies are summarised in Table 3.

Paroxysmal Dyskinesias

Paroxysmal dyskinesias represent a heterogeneous group of diseases with paroxysmal occurrence of dyskinesias including dystonia, chorea, ballism and athetosis.

Dystonia

Dystonia is an involuntary alteration in the pattern of muscle activation during voluntary movement or maintenance of posture. The involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both.

Chorea, Ballism

Chorea is an ongoing random-appearing sequence of one or more discrete involuntary movements or movement fragments. Movements appear random due to variability in timing, duration, direction, or anatomic location. Movements may therefore appear to flow randomly from one muscle group to another, and can involve trunk, neck, face, tongue, and extremities. Ballism is chorea that affects proximal joints such as shoulder or hip. This leads to large amplitude movements of the limbs, sometimes with a flinging or flailing quality.[17]

Athetosis

Athetosis is a slow, continuous, involuntary writhing movement that prevents maintenance of a stable posture. Athetosis involves continuous smooth movements that appear random and are not composed of recognisable sub-movements or movement fragments. The combination of chorea and athetosis is called choreoathetosis.[17]

Paroxysmal Dyskinesias

Paroxysmal dyskinesias represent a heterogeneous group of diseases with paroxysmal occurrence of dyskinesias including dystonia, chorea, ballism and athetosis. These include paroxysmal kinesigenic dyskinesia (PKD), paroxysmal nonkinesigenic dyskinesia (PNKD) and paroxysmal exertion-induced dyskinesia (PED). Detailed history taking can be very revealing to differentiate these 3 disorders. PKD is usually triggered by sudden voluntary movements, startles and hyperventilation whereas PNKD has spontaneous onset which is provoked by alcoholic beverages, coffee, excitement, stress, exhaustion, tea, beverages with caffeine. PED is induced by exercise. PKD has the shortest duration of attack which lasts for seconds to a few minutes with a frequency of up to 100 per day. A typical attack in PNKD usually lasts for minutes to several hours with a frequency of up to 3 per day. PED does not have more than one attack per day which lasts for 15 to 60 minutes. The clinical approach and management of these conditions are reviewed by Strzelczyk et al.[27] In addition, a group of secondary symptomatic paroxysmal dyskinesias has to be excluded. Interesting, a benign condition known as transient paroxysmal dystonia of infancy exists. The age of onset is before 1 year with no other neurodevelopmental defects. The attacks, which occur only in the awake state, are characterised by opisthotonus, symmetric or asymmetric increased tone in the arms with extreme pronation of the wrists, and preserved consciousness. Each attack can lasts from minutes to days. The condition is non-progressive with spontaneous recovery in the second year of life.[28,29]

Conceptually, different hyperkinetic movements can be distinguished by rhythmicity, repeatability and suppressibility (Table 4). Rhythmic movements imply a periodic or cyclic behaviour with similar movements on successive cycles. Repetitive movements include movements or movement elements that may revisit spatial positions or joint configurations but without a cyclic or rhythmic timing.[16]

Table 3 Factors distinguishing tics and stereotypies

	Tics	**Stereotypies**
Age of onset	6-7 years	<3 years
Pattern	Variable, wax and wane	Fixed, identical, patterned
Movements	Blink, grimace, twist, shrug	Arms / hands (flap, wave), body rock / head nod
Rhythm	Rapid, sudden, random	Rhythmic
Duration	Intermittent, brief, abrupt	Intermittent, continuous prolonged
Premonitory urge	Yes	No
Precipitant	Excitement, stress	Excitement, stress, also when engrossed
Suppression	Brief, voluntary (but have increased "inner tension")	With distraction, rare conscious effort
Distraction	Reduction of tics	Stops

Modified from Mahone EM, Bridges D, Prahme C, Singer HS. Repetitive arm and hand movements (complex motor stereotypies) in children. J Pediatr 2004:145;391-5.

Psychogenic Paroxysmal Movement Disorders

Psychogenic movement disorders are a group of disorders that an organic basis will not be found for the symptoms or that a psychiatric disorder is the primary substrate from which the atypical symptoms blossom. The average age of onset is 12 to 14 years, with a range of 7 to 18 years. Common psychogenic movement disorders involve hyperkinetic movements such as dystonia, tremor, myoclonus, tics, hemiballismus and chorea.[18] Useful clues for diagnosing psychogenic movement disorders in children are summarised in Table 5. Revised diagnostic criteria have recently been proposed.[30] One should bear in mind that organic and psychogenic movement disorders can co-exist in a patient which poses more diagnostic challenges.

Transient and Developmental Movement Disorders in Children

Many movement disorders in childhood are benign and related to normal stages of development. These are typically associated with complete resolution of the abnormal movements and ultimately normal development and neurologic function.[18]

Jitteriness

Jitteriness is a term to describe a series of recurrent tremors in infants.[31] It is the most common abnormal movement encountered in neonates.[32] Jitteriness can be differentiated from seizure if the following characteristics are observed – the jitteriness can be brought on with stimuli and can be stopped with gentle passive flexion and restraint of the affected limb; it is not associated with ocular phenomena, such as forced eye deviation; and is not associated with significant autonomic changes such as hypertension or apnoea.[33] Jitteriness can be benign or pathological.

Shuddering

Shuddering attacks are benign nonepileptic events that typically begin in infancy.

The clinical events consist of rapid shivering of the head, shoulder, and occasionally the trunk. Events are brief, usually lasting not more than a few seconds. Frequency can be up to more than 100 events per day. Attacks seem to be precipitated by feeding, eating, head movements and certain tasks (pressing toys together or sticking a fork into a piece of bread).[34] Reassurance of parents is crucial since relatives are often frightened by the unexpected appearance and often high frequency of the attacks. Spontaneous remission can be expected.[35]

Benign Paroxysmal Torticollis of Infancy

Benign paroxysmal torticollis is a rare paroxysmal dyskinesia characterised by recurrent stereotypic attacks of torticollis. Attacks first manifest during infancy, between ages 2-8 months. They resolve by age 3-5 years.[36] Typically, the frequency and duration of attacks decline as the child grows older.[1] The diagnostic criteria are listed in Table 6. Patients with benign paroxysmal torticollis may develop benign paroxysmal vertigo, cyclic vomiting syndrome, abdominal migraine, motion sickness, or migraine. If the episodes recur, their management is unclear. No clinical trials have been reported. Some may suggest a trial of cryproheptidine if an individual's episodes are painful.[37]

Infantile Masturbation

Infantile masturbation (self stimulatory or gratification behaviours) can affect children of both sexes. Age of presentation typically ranges between 3 months to 5 years.

Table 4 Distinguishing features among different hyperkinetic movements

	Rhythmic	Repeated stereotyped movement	Suppressible
Dystonia	Rarely	Sometimes	Partial or only briefly
Chorea	No	Rarely	No
Athetosis	No	No	No
Myoclonus	Sometimes	Usually	No
Tremor	Yes	Yes	Sometimes briefly
Tics	No	Yes	Usually
Stereotypies	Yes	Yes	Yes

Modified from Sanger TD, Chen D, Fehlings DL, et al. Definition and classification of hyperkinetic movements in childhood. Mov Disord 2010;25: 1538-49.[17]

Table 5 Useful clues for diagnosing psychogenic movement disorders in children

Historical clues

1. Abrupt onset
2. Static course
3. Spontaneous remission or inconsistency over time
4. Remission when the child is not aware of being observed
5. Presence of secondary gain

Clinical clues

1. Inconsistent character of the movement (amplitude, frequency, distribution, selective disability)
2. Paroxysmal movement disorder
3. Movements increase with attention to the movement, or decrease with distraction
4. Ability to trigger or relieve the abnormal movements with unusual or nonphysiologic interventions (e.g. body trigger points)
5. False weakness or sensory findings
6. Deliberate slowness of movements
7. Entrainment of tremor with voluntary rapid alternative movements of different frequencies
8. Functional disability out of proportion to examination findings

Therapeutic responses

1. Unresponsiveness to appropriate medications
2. Response to placebos
3. Remission with psychotherapy

Modified from Singer HS, Mink JW, Gilbert DL, Jankovic J. Psychogenic movement disorders. Movement disorders in childhood. Saunders, Elsevier, 2010.

Table 6 Diagnostic criteria for benign paroxysmal torticollis of infancy

1. Episodic attacks with normal neurological examination in between attacks, in an infant, with all of the following including criterion 2:
   - Tilt of the head to one side (not always the same side), with or without slight rotation
   - Lasting minutes to days
   - Remitting spontaneously and tending to recur monthly
2. During attacks, signs of one or more of the symptoms and signs including pallor, irritability, malaise, vomiting, ataxia
3. Other disorders excluded

Modified from Headache Classification Committee. The international classification of headache disorders, cranial neuralgia and facial pain. 2nd edition. Cephalalgia 2004;24(Suppl 1):1-160.

Events occur numerous times per week and last for several minutes. Most episodes in children lack the obvious manual stimulation of genitalia but have behaviours such as dystonic posturing, rocking, grunting, facial flushing with diaphoresis, posturing of the lower extremities allowing pressure on the perineum, cessation of the behaviour with distraction, and no alteration of consciousness. Episodes can occur in a variety of situations including sleep, while in a car seat, a walker or high chair, when tired, or watching television. A potentially helpful diagnostic clue is that the gratification event can be stopped with distraction, which in turn leads to the child becoming angered and annoyed.[38] As these behaviours are a normal occurrence in development, reassurance is the key in counseling families.[39]

Benign Paroxysmal Tonic Upward Gaze

This condition is characterised by periods of constant or variably sustained tonic conjugate upward eye deviation, downbeating saccades in attempted downward gaze with apparently normal horizontal eye movement, relief after sleep or rest, with or without chronic or intermittent ataxia. The duration of each attack can lasts from a few seconds to days. Onset appears to occur at the first month to the end of the second year of age. The course of the disease appears to be characterised by spontaneous improvement after 1-2 years, but there are rapidly resolving cases.[40] Benign paroxysmal tonic upward gaze is usually associated with a normal neurodevelopmental profile. However, this condition is recently reported to be associated with other neurological conditions including syndromal disorders and white matter diseases while spontaneous resolution is not seen.[41,42]

Sandifer Syndrome

Sandifer syndrome is a paroxysmal movement disorder characterised by abnormal movements of the head, neck, and trunk in association with gastroesophageal reflux disease.[43] Most cases are neurodevelopmentally normal children with symptom onset in early childhood. Infants often have retrocollis and opisthotonic posturing, whereas older children have the side-to-side head movements.[44] Nodding and rotation of the head, neck extension, gurgling sounds, writhing movements of the limbs and severe hypotonia have been reported. Although the intermittent stiff tonic posture and periods of crying and apparent discomfort may suggest seizures, absence of rhythmic clonic component is unlikely to be seizures.[45] The key to this diagnosis is the association between the movements

and gastrointestinal symptoms such as abdominal pain and regurgitation. The movements usually respond to anti-reflux medications and / or fundoplication.[43,44]

Breath-holding Attacks

Breath-holding spells affect children aged 6 months to 5 years. Typically, a clear trigger is present, with the child being upset and crying. At the end of expiration, the child is unable to relax and inhale and becomes apneic and cyanotic. The child may appear angry and upset about this uncomfortable feeling, loses consciousness, may have urinary incontinence, and becomes stiff or even opisthotonic. The EEG during the event typically shows high-amplitude slowing followed by suppression, as is seen in syncope of any cause. When the child relaxes and breathes again, consciousness is gradually regained. These cyanotic breath-holding spells could be easily confused with epileptic events, but they are not primarily epileptic phenomena.[46] They can evolve into epileptic seizures and even status epilepticus, but the initial event is not epileptic. Cyanotic breath-holding spells are to be distinguished from pallid infantile syncope, which is associated with brief cardiac asystole and overlap with seizures.[12] Pallid infantile syncope is usually provoked by sudden fright or pain.[47]

Spasmus Nutans

Spasmus nutans is a rare, idiopathic disorder of childhood comprising the clinical triad of nystagmus, head nodding, and torticollis. This triad classically presents in the first year of life and symptoms typically resolve by 3-6 years of age.[48] Spasmus nutans may have paroxysmal symptoms. One must rule out associated intracranial pathologies such as neoplasms, arachnoid cysts, optic nerve hypoplasia, opsoclonus-myoclonus, diencephalic syndrome and subacute necrotising encephalomyelopathy (Leigh disease) etc. Retinal diseases have also been associated with spasmus nutans including achromatopsia, cone or rod dystrophy, congenital stationary, night blindness and Bardet-Biedel syndrome.[49]

Sleep-related Disorders

Parasomnias

Parasomnias are unpleasant or undesirable behavioural or experiential phenomena that occur predominantly or exclusively during sleep. It could be a challenge to distinguish parasomnias from seizures such as non-rapid-eye-movement (non-REM) arousal disorder parasomnias (confusional arousals, sleep terrors and sleep walking) or REM sleep behaviour disorder (RBD).[50] Individual disorder will be described below. Major clinical features differentiating parasomnias from nocturnal seizures are summarised in Table 7.

Confusional Arousals

Confusional arousals are characterised by sudden arousals, disorientation, and prolonged confusion, sometimes associated with complex behaviours, but never with conscious awareness. They are very common, especially in young children, but under-recognised by nonsleep pediatricians, and frequently misinterpreted as nocturnal epileptic events.[51]

Sleep Terrors

Sleep terrors are typically seen in toddlers and are characterised by sudden arousals with a loud and

Table 7 Major clinical features differentiating parasomnias and nocturnal seizures

Parasomnias	Nocturnal seizures
1-2 episodes per night	>3 episodes per night
1-4 episodes per month	>10 episodes per month
Episodes occur during rapid-eye-movement or slow-wave sleep	Episodes occur during stage 1 and 2 sleep
Episodes occur more likely after 90 minutes of sleep onset	Episodes may occur at any time of the night
Episodes lasts up to 30 minutes	Episodes lasts up to 1-2 minutes
Variable movements and actions	Stereotyped movements
Physical and verbal interaction	Rare physical and verbal interaction
Failure to fully arouse after the event	Postictal confusion may be present but the patient generally is fully arousable after the event

Modified from Vendrame M, Kothare SV. Epileptic and nonepileptic paroxysmal events out of sleep in children. J Clin Neurophysiol 2011;28:111-9.[55]

inconsolable scream. The toddler seems pale and terrified. There is no awakening and the child usually falls asleep and does not recall the event at a later time.[52]

Sleep Walking

Sleep walking is common among the adolescent population and consists of episodes of complex and elaborate activities including walking, but without recollection of these events.[53]

REM Sleep Behaviour Disorder

REM behaviour disorder is characterised by motor and behavioural manifestations associated with dreaming during REM sleep. The presence of bizarre movements and complex behaviour, including laughing, talking, moaning, punching, kicking, and running can mimic epileptic events.[54] Video-polysomnography may help in differentiating seizures from REM sleep behaviour disorders as events of this parasomnia occur during REM sleep, representing a major differentiating finding from nocturnal seizures.[55]

Restless Leg Syndrome

Restless leg syndrome is not only confined to the adult population. This can occur in children. Diagnostic criteria included the four essential factors used in adults: a history of an urge to move the legs accompanied or caused by uncomfortable sensations, the urge to move or unpleasant sensations worsened during periods of rest or inactivity, the urge to move relieved by movement (quisescegenic), and a circadian pattern with symptoms being worse in the evening or night. In addition, for a definite diagnosis in children aged 2-12 years, the children must express leg discomfort in their own words (e.g. tickle, bugs, shaky, etc.) or have two out of three of the following: a sleep disturbance, parent with definite restless leg syndrome, or elevated periodic limb movement index on polysomnography. Another finding was the coexistence of restless leg syndrome with other comorbid conditions such as parasomnias, attention-deficit hyperactivity disorder, anxiety, and depression. Dopaminergic agonists have been used with success in children.[39]

Clinical Approach to Paroxysmal Movements in Childhood

The clinical approach to paroxysmal movements in childhood can be very challenging.

History taking is the first and extremely important step to make an accurate diagnosis. A detailed history should be obtained from the patient and eye witnesses, which could be parents, family members, not infrequently teachers or even ambulance officers. Attention could be made particularly to the age of onset (Table 8) and different situations during which the movements occur (Table 9), examination of the nature of abnormal movements by the clinicians either during the attack or using videotape recordings of previous attacks may dramatically improve the diagnostic accuracy. Other details including frequency and duration of each attack, natural disease course, past medical history of the patient including perinatal, developmental aspects, family history and physical examination would all contribute to a correct diagnosis. When history and analysis of movements cannot reach a diagnosis, long term video-electroencephalographic study to capture paroxysmal events may be required to rule out epileptic phenomenon, and classify the underlying disorder or even help parents understand the potential psychogenic nature of some disorders. This is especially useful for benign paroxysmal torticollis of infancy and benign paroxysmal tonic upward gaze when each attack is short and cannot be easily distinguishable from an epileptic seizure; and psychogenic non-epileptic seizures or paroxysmal movement disorders. Long term follow up is necessary to ensure the benign and self-limiting nature of some conditions like transient and developmental movement disorders in children, distinguish them from more serious disorders, and be able to provide reassurance when appropriate.[18]

An accurate diagnosis is extremely important to differentiate between various epileptic and non-epileptic conditions. Clinicians should have knowledge and be aware of various paroxysmal movements mentioned in this article to ensure timely and appropriate treatment for certain conditions and to avoid unnecessary use of anticonvulsants in benign non-epileptic conditions.

Conclusion

Paroxysmal abnormal movements in childhood comprise a wide range of differential diagnoses which can be epileptic and non-epileptic. History taking and examination of the movements may dramatically increase the diagnostic accuracy. A long-term video-electroencephalographic recording may be necessary to provide ultimate diagnosis in some situations. Prompt and accurate diagnosis is

Table 8 Classification of paroxysmal movements in childhood by age of onset

Paroxysmal movements in childhood		
Across different ages	***Infantile onset***	***Childhood onset***
• Epileptic seizures	• Jitteriness	• Tics
• Convulsive syncope	• Exaggerated startle responses	• Restless leg syndrome
• Ataxia	• Shuddering	• Psychogenic non-epileptic seizures
• Tremor	• Benign paroxysmal torticollis of infancy	• Psychogenic paroxysmal movement disorders
• Myoclonus	• Infantile masturbation	• Confusional arousals
• Paroxysmal dyskinesias	• Benign paroxysmal tonic upward gaze	• Sleep walking
• Sandifer syndrome	• Stereotypies	• Sleep terror
	• Breath-holding attacks	• Rapid-eye-movement sleep behavioural disorders
	• Spasmus nutans	

Table 9 Approach to paroxysmal movements in childhood by specific situations

Situation	**Possible cause(s)**
Precipitation / Aggravation by	
• Sleep	• Seizures • Confusional arousals • Sleep walking • Sleep terror • Rapid-eye-movement sleep behaviour disorders • Restless leg syndrome
• Prolonged sitting or standing, rising to upright posture, dehydration, pain, fright, micturition, defaecation etc.	• Convulsive syncope • Pallid breath-holding attacks
• Feeding	• Sandifer syndrome • Shuddering attacks
• Sensory stimuli (visual / auditory / tactile etc.)	• Seizures • Jitteriness • Exaggerated startle responses
• Crying	• Cyanotic breath-holding attacks
• Movements	• Paroxysmal dyskinesias • Shuddering attacks
Pre-existing conditions	
• Cardiac diseases	• Convulsive syncope
• Psychological / psychiatric / psychosomatic symptoms	• Psychogenic non-epileptic seizures • Psychogenic paroxysmal movement disorders
Preceding events	
• Aura	• Seizures
• Urge to move part of the body	• Tics • Restless leg syndrome
• Nausea, palpitations, dyspnoea, warm sensation, light-headedness, graying of vision or hearing becoming distant	• Convulsive syncope
Distractibility / Suppressibility	• Tics • Stereotypies • Infantile masturbation • Jitteriness

extremely important for commencement of the most appropriate treatment if indicated and to avoid unnecessary use of anticonvulsants in certain benign non-epileptic self-limiting conditions. Last but not the least, timely reassurance to parents of benign conditions is also very important to reduce unnecessary parental anxiety.

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