The Paroxysmal Disorders

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Chapter 1

Paroxysmal attacks: diagnostic gold standards and history-taking

Peter Wolf

Gold standard case history

Can the case history ever be considered a gold standard? Generally, a gold standard is a firmly established reference that serves as a reliable comparison for other measures or procedures. We think of gold standards as hard, quantitative and cross-checked data. By comparison, the case history is soft, subjective and carries numerous risks of misunderstandings, prejudices and even the possibility of leading questioning. However, there is no doubt that the results of even the most exact and advanced investigations are of clinical significance only when they make sense in the context of the clinical background as provided by the history. In other words, the history is used in the same way as a gold standard, that is, as a counter-check for the plausibility of the findings.

Specific aspects of the case histories for paroxysmal disorders

The case history is of particularly high importance in paroxysmal disorders because many seizures may comprise a variety of subjective symptoms or may even consist exclusively of these symptoms. Because of their transient character, these symptoms are often difficult to document or can only be discovered through a detailed clinical history of the event. This, too, is not always straightforward because patients relate their experiences in their own language, which may be naïve, imprecise, or metaphorical and may need further interpretation to be correctly understood (Surmann 2005). This level of interpreter involvement can vary from structuring, to clarification by cross-questioning, to veritable "translation." In such a situation, it is very important that the doctor avoids asking questions that lead to answers which conform to his or her preconceived hypotheses and prejudices, rather than realistically reflecting patients' experiences. On the other hand, to get any result it is often necessary to structure the interview, which will rely on the background of the doctor's knowledge of typical seizure symptoms. It is important to find the correct balance of critically reflecting the physician's own experience with epilepsy but always be prepared to reconsider a working hypothesis if the patient's report does not reflect it. Ideally, the doctor taking the history should be absolutely neutral and block out his or her subjectivity so as not to influence the patient's free flow of remembering the events in question. This sounds difficult to achieve, but it is the method used in the project that established the linguistic criteria in the differential diagnosis of epilepsy from psychogenic non-epileptic seizures (PNES) and is currently used in clinical trials to define the differences experientially between various types of epilepsy. In these studies, the patient is invited by two or three standardized initial questions to speak freely about his or her experiences. The reports are recorded and transcribed, and undergo a detailed analysis using formal linguistic criteria. The method was first developed in Germany (Wolf et al. 2000) and the results have since been duplicated in England (Schwabe et al. 2007). The method used in the research is time-consuming but may end with the development of software programs to provide rapid, even online, diagnostic conclusions.

With this method, it is possible to generate objective and quantitative data based on the patient's subjective reports. It examines exclusively one of several aspects of these reports: their linguistic form, not their contents. However, this aspect is one that has hitherto been neglected. It seems highly questionable whether a similar degree of objectivity can be attained with respect to contents.

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Taking a seizure history is primarily for diagnostic purposes, largely to differentiate epilepsy from other seizure-like disorders but also to distinguish between different types of epileptic seizures and to understand the anatomy and etiology of these events. Hypotheses are formed, followed-up or refuted, and they determine the subsequent course of the interview. The questions asked in a differential diagnosis of epileptic seizures versus migraine accompagnée are entirely different from those to distinguish focal from generalized epileptic seizures. Even if the latter differentiation is only preliminary, it needs to become much more detailed; it can require rather sophisticated history-taking in and of itself. This may be the case when the patient reports some subjective experiences at or near the onset of the seizure. Are these auras indicating a focal seizure onset, absences immediately preceding a generalized tonic-clonic seizure, or non-specific sensations preceding a seizure? In addition to such difficulties inherent in the matter in question, the linguistic abilities of the patients may additionally require adjustments of the interview technique.

Taking a seizure history can and should lead far into the diagnostic process. For example, at the end of taking a history of focal epileptic seizures the doctor should develop a relatively precise hypothesis about where in the brain the seizures originate and how they spread. For this, it is essential to know the exact sequence of seizure symptoms. But this is not necessarily the way they are spontaneously described by the patients, whose reports may instead reflect hierarchies of symptoms according to their subjective importance. They are not aware that symptoms which appear minor to them may provide pivotal anatomical information. Taking a seizure history is therefore typically a structuring process, the dynamics of which are the result of an interaction between doctor and patient, and the course of which is fundamentally unpredictable.

Such an in-depth dialogue also opens up perspectives beyond diagnostics. Most patients are not used to being interrogated by somebody who has a detailed understanding of their subjective experiences and is interested in them. They often feel they are taken seriously in a way they are not accustomed to, so they are often very appreciative. This attitude goes a long way to establish the necessary interpersonal confidence for a subsequent therapeutic relationship.

Beyond providing diagnostic clues and anatomical understanding, another frequent consequence of these interviews is indications for the best further therapeutic strategy. Examples are the development of self-control strategies to avoid specific or non-specific seizure triggers (which the patient needs to be asked about) or to prevent traumas or clusters of seizures.

Verbal communication is only successful when the partners in dialogue find a common language. This is highly important in the diagnostic interview. A doctor who takes a case history or seizure history should be aware that the dialogue is asymmetrical, with the doctor holding the more dominant role. Therefore it is the doctor's responsibility that the communication works, and they need to be aware that misunderstandings may occur in either direction.

To be understood by the patient

To get useful answers from the patient, the doctor needs to ask questions which the patient can understand. Very few patients have studied medicine and know its professional language. To express a complex medical matter to lay people in plain language is not always easy but it is not only a matter of politeness – it is a basic requirement for communication.

In this asymmetrical dialogue situation, the patient cannot be expected to question something he or she has not understood. He or she may be too shy or embarrassed to do that. It is the doctor who needs to make sure that he or she has been understood by the patient (in a polite way, of course). "Did I express myself understandably?" is a much better way to ask the question as opposed to, "Do you follow me?"

To understand the patient

It is by no means trivial to point out that the patient's reports also need to be understood correctly. There may be dialects, accents and colloquialisms which do not have the same meaning everywhere. However, much more important are vague terms and undeclared similes and metaphors. Here are some examples.

- A patient who reports during a seizure to "stand beside himself/herself" may be using a relatively common metaphor for reduced presence of mind, and the interlocutor may understand it like this and pass on. But it may actually need to be understood literally and signify the experience of a double. This is a rare but characteristic possible symptom of parietal lobe seizures and is often described in this way. In a seizure history, it needs to be followed up by the question, "On which side

of yourself do you stand?" If it is a double experience, the patient will usually be able to promptly tell the side (typically, contralateral to the focus). If he or she was using a metaphor, the question will not make sense to them and it may be necessary to explain that you were not joking.

- The subjective experiences in epileptic auras are often so unnatural and different from common experiences that they are extremely difficult to describe. Quite a few patients are in the habit of using surrogate terms to get around this difficulty. These are often not descriptive, even if they may seem so to the patient. Dizziness and giddiness are such terms. They should never be taken at face value. The patient should always be asked to be as descriptive as possible. If it turns out that the symptoms are indescribable and cannot be compared to any natural experiences, this is helpful because it is a quality well-known from epileptic seizures and otherwise extremely rare.
- One of the best-known traditional terms in epilepsy – aura – is probably a misunderstanding which arose in such a situation. According to Temkin (1971), "this word, taken from the Greek, originally meant a 'breeze.' It was introduced into medical terminology not by a physician but by a patient. When still a young man, Galen, together with other physicians, visited a 13-year-old boy. The patient told them that the condition originated in the lower leg, and that 'from there it climbed upwards in a straight line through the thigh and further through the flank and side to the neck and as far as the head: but as soon as it had touched the latter he was no longer able to follow.' When the physicians asked him what exactly rose up to the head, he could not tell, but another youth, who was a better observer, said 'that it was like a cold breeze.'" The story in this version does not really make sense because it was a subjective experience, and there was nothing for the other youth to "observe." The scene can easily be imagined. Being interrogated by several physicians and having described the march of a sensory Jacksonian seizure, the boy was embarrassed because he was unable to set words an indescribable quality of the sensation and gratefully grasped the first way out that was presented to him. I have never met a patient who compared his or her aura experience to a cold breeze.

Patients with ictal hallucinations and illusions typically do not report them spontaneously because they are afraid of being considered either mentally disturbed or of becoming so. This is not only true for epileptic hallucinations but also for hypnagogic hallucinations, which the patient needs to be asked about if the differential diagnosis is in the field of narcolepsy or cataplexy. It is important in the interview to create an atmosphere of confidence where the patient can talk about such matters freely. Nonetheless, it is usually necessary to ask directly about them, and this should not be forgotten whenever there is a possibility that these conditions are present – for example, if there are other indications of an epileptic focus in the parieto-occipital area.

A checklist for seizure histories

Taking a seizure history does not necessarily need to follow a rigid scheme; however, it is useful to have an internal standard checklist of topics and questions, both when the history is taken from the patient themselves and/or from witnesses.

The patient

Patients often do not spontaneously report all seizure symptoms but only the most prominent ones. Also, the first symptom mentioned is not always the first in a sequence of symptoms but the one which impresses or bothers the patient the most. A dialogue is frequently necessary to get the full picture of all subjective seizure symptoms and their sequence.

Questions for the patients:

- What is the very first indication of a commencing seizure? Patients sometimes misunderstand the question and report a suspected seizure trigger.
- Can this first symptom be preceded by something else? Many patients understand auras not as part of the seizure but as a warning that precedes it.
- What is the sequence of symptoms? Triggers of seizures (e.g., reflex epileptic seizures); syncope triggered by cough, micturition, pain; emotional triggers of cataplectic fits; movements in kinesigenic paroxysmal choreoathetosis.
- Is this dependent on certain postures? (e.g., orthostatic syncope)
- Can the patient arrest his or her seizures? Sometimes? Always? How?
- What is the duration of the seizure? Relation to time-of-day or sleep-wake cycle (e.g., generalized

tonic-clonic seizures or myoclonic seizures after awaking); frontal lobe epileptic seizures in sleep; REM and non-REM sleep parasomnias; sleep paralysis; PNES never in sleep but perhaps at night.
- Are there patterns of recurrence (e.g., clusters at intervals in temporal lobe epilepsy)?

The witnesses

Some witnesses are in the habit of stubbornly not reporting what they have observed but how they reacted to it. Patience is needed to get a seizure description. However, the witness may have some knowledge which may save a lot of unnecessary investigations.

Questions for the witnesses:

- What drew the observer's attention first to the seizure?
- What was the first observed seizure sign?
- What was the sequence of signs and symptoms?
- What movements were observed, including extent, speed and direction?
- Were there jerks? Stiffness? Where was it? Was it unilateral or bilateral? How long did it last?
- Did it spread?
- Were there automatisms? These are often not spontaneously described even if they are quite prominent.
- Was there a fall? If so, in what direction? Did the patient's tone change, and did he or she become flaccid or rigid?
- Were their eyes open or closed? Was there deviation of the eyes?
- What was their facial expression? Did they stare?
- Was there a change of color?
- Did they salivate?
- Did they bite their tongue? If so was it lateral or apical? Does that happen always, often or occasionally?
- Was the patient incontinent or enuretic?
- What was the level of responsiveness?
- Was there speech during and/or after the seizure, or was there speech arrest – speech similar to a foreign language – or speech that was grammatically correct but nonsensical or paraphasic?
- What was the duration of the seizure? Was that an estimate or was it accurately timed? Most observers often tend to overestimate duration, especially with severe seizures or with the first observed seizure. Most often, they include the postictal phase.
- Were the onset and offset sudden or phased?
- Were there postictal symptoms such as a speech disturbance, a paresis or weakness, disorientation or aggressive-defensive behavior?
- If they have observed many seizures in the same person, are the events stereotyped or variable?

It is often a good idea to take the seizure history from patients and witnesses together in one setting and point out that many seizures have both subjective symptoms which are only known to the patient and objective signs which can be visible to the observer, even as the patient may be unaware of them. This helps all parties to understand the weighting of their observations. For many patients, this is novel because their experience is often that their own knowledge is not appreciated. For every sign and symptom, it needs to be clarified if it is only subjective or only objective, or has both aspects.

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Chapter 2

Paroxysmal disorders in childhood

Gerhard Kurlemann

Introduction

Non-epileptic infantile paroxysmal dyskinesia is a common disorder that is not easily distinguished from epileptic seizures at first glance. Whenever parents can provide a good description of symptoms or it is possible to observe the incident firsthand or with video, one can usually diagnose the disorder correctly and distinguish it from epilepsy, which is often the initial diagnosis.

Between 0.8% and 1.0% of the population suffer from epilepsy. Epileptic seizures in infancy are widespread, occurring with an incidence between 80 and 100 per 100,000. Prevalence studies of active epilepsy in infancy show rates between 3 and 6 per 1000 children.

An epileptic seizure is diagnosed according to clinical findings, including a thorough history and neurological examination with corresponding diagnostic studies, for example, electroencephalography (EEG). An exact observation/description and the corresponding EEG result are indispensable for the diagnosis of an epileptic seizure, as for all movement disorders. If the EEG matches the clinical findings – for example, violent tonic dyskinetic arm movement in a six-month-old infant together with a documented hypsarrhythmia during sleep and wake EEG – then West's syndrome with salaam convulsions can be diagnosed reliably.

Absence epilepsy is a frequent cause for children presenting with the observation of paroxysmal "daydreaming." It can often be diagnosed in a primary care setting without the corroborative evidence of an EEG by requesting the patient to perform controlled hyperventilation. Most often the frequent symptoms of absence epilepsy become obvious during hyperventilation, which is interrupted by the discontinuous breathing pattern so elicited. Most frequently, the patient will develop open and upturned eyes, have some mild smacking of the lips and fiddling hand movement. Final corroboration can be provided by the presence of the typical EEG pattern of 3-Hz spike-and-wave discharges. Shoulder girdle localized myoclonus is associated with the Janz syndrome variant. These can be either symmetrical or asymmetrical in nature (Fig. 2.1). This myoclonus in particular can precede grand mal seizures by years. The myoclonic form of this type of epilepsy is accompanied by generalized poly-spike-and-wave discharges on the EEG. Whenever clinical symptoms are not consistent with EEG findings, the wide spectrum of non-epileptic, age-linked movement disorders should be borne in mind, especially where infants are concerned (Table 2.1).

Up to 20% of patients in large representative groups presumed to have epilepsy in fact do not have epilepsy but rather one of the numerous forms of paroxysmal dyskinesia (Scheepers et al. 1998; Uldall et al. 2006). A diagnosis of epilepsy should not rely solely on the EEG. The combination of clinical picture and EEG findings allows a more definitive diagnostic differentiation when viewed as a whole. The EEG is a valuable tool when diagnosing epilepsy; however, an interictal EEG alone can neither prove nor rule out epilepsy reliably. A single, awake EEG showing no typical epileptic pattern is seen in up to 50% of epileptic patients. As a corollary, 3%–5% of routine EEGs performed on children show patterns typical for epilepsy, even though they have not suffered an epileptic fit. If in doubt, it is advisable to wait and see how things develop and broaden the angle of diagnostic possibilities. A false-positive diagnosis of epilepsy with all its consequences is more detrimental to both child and family than waiting for the next possible epileptic fit.

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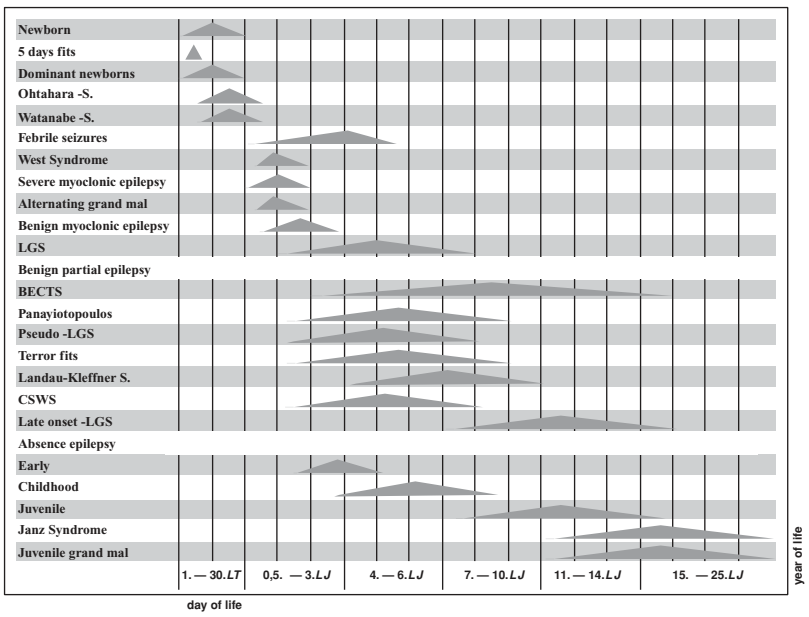

Fig. 2.1. Spectrum and age of manifestation for infant epilepsy syndromes.

Table 2.1 Differential diagnosis of paroxysmal dyskinesia

Altered state of consciousness
Syncope
Affective respiratory spasms – breath-holding spells – cyanotic vs. pale
Narcolepsy / cataplexy
Cardiac dysrhythmia – supraventricular
Atypical migraine / "confusion" migraine
Reflex pattern in sleep
Nightmare
Jactatio capitis / corporis nocturna
Benign infantile sleep myoclonus
Unaltered state of consciousness
Tics
Shuddering attacks
Benign paroxysmal dizziness
Self-stimulation / auto-stimulation
Benign paroxysmal torticollis
Nodding spasm
Sandifer syndrome
Benign paroxysmal upward gaze
Kinesigenic choreoathetosis
Hyperekplexia
Transient infantile dystonia
Kinsbourne syndrome
Alternating infantile hemiplegia
Hyperventilation attacks
Panic attacks
Psychogenic non-epileptic fits
Pulse synchronous bulbus movement in spheno-orbital dysplasia
Munchausen syndrome by proxy

The clinical symptoms of the most frequently occurring movement disorders that should be considered in differential diagnosis are described herein.

Jactatio capitis/corporis nocturna

Jactatio capitis/corporis nocturna is characterized by rhythmic stereotype head-rolling or body-rocking movements while falling asleep during sleep stage I, or during short arousals during sleep stage II. The most frequent movement is head-rolling or banging. Head-banging can occasionally even cause callus formation on the forehead if the bed head and sides are not padded. Jactatio duration varies from between 30 seconds and 30 minutes, and in more than 60% of cases it starts around the age of nine months and generally persists until the age of five years (DiMario 2006).

Key symptoms: Rhythmic head/body movements during light sleep

Syncope

Syncope is frequently experienced by infants and adolescents, although the exact incidence is not known. It has been estimated that about 30% to 50% of children will have experienced a syncopal episode by the time they reach adolescence. A syncope is a brief, temporary loss of consciousness due to a transient reduction of cerebral perfusion. Diminished cerebral perfusion

Table 2.2 Syncope symptoms vs. epileptic seizure

	Syncope	Epileptic seizure
Onset	Sudden	Prodromes: sweating, lightheadedness, dizziness, palpitations
Position	Variable	Standing, more seldom sitting
Muscle activity	Variable	Predominantly hypotensive
Skin color	Pale	Variable – cyanotic
Tonic phase	Brief	Pronounced
Myoclonus	Brief, multifocal	Violent, symmetrical, rhythmic
Reorientation	Rapid	Variable – longer
Tongue bite	3%	Lateral, 30%
Injury	Frequent	Seldom
Bed-wetting	25%	25%

of less than 30 ml/100 g brain tissue per minute causes a syncopal episode, which in turn involves loss of posture followed by spontaneous recovery.

A distinction is made between cardiovascular and vasovagal syncope. The most frequent form in infancy is the vasovagal syncope. It is extremely rare for an epileptic fit to be triggered by a syncope episode. Depending on the clinical symptoms, in particular when violent multifocal myoclonus and bed-wetting occur, an epileptic fit is the most likely diagnosis.

A comprehensive history should always be obtained. Prodromes – such as lightheadedness or ringing in the ears in situations involving, for example, long periods of standing upright in poorly ventilated rooms – are immediately suggestive of syncope. Syncope that occurs during physical exercise could be associated with a long QT syndrome and, as such, should be ruled out (Paolicchi 2002).

Table 2.2 distinguishes syncope symptoms from those of an epileptic fit; Table 2.3 lists the possible symptoms of syncope in order of frequency.

Key symptoms: See Table 2.2

Breath-holding spell attacks

Breath-holding spell attacks typically occur in infants. It is estimated that up to 4% of children under five years of age are so affected. A distinction is made between the frequently cyanotic form (80%) and pale breath-holding form (20%). Cyanotic attacks are brought on by different triggers, for example, the child cannot have its own way. There is a long run-up to loss of consciousness due to dead-volume ventilation. Brief shoulder myoclonus is common during the final phase of a cerebral hypoxia, when the rising CO_2 concentration is already beginning to activate the respiration reflex, marking the start of the recovery phase. However, this type of myoclonus must not be mistaken for an epileptic seizure. In contrast, the pale syncope occurs abruptly and is frequently pain-induced. The long-term prognosis is favorable, the symptoms are generally self-limiting and therapy is not necessary (Lombroso & Lerman 1967; Evans 1997; Kuhle et al. 2000).

Children who have breath-holding spell attacks should be examined to systematically rule out iron-deficiency anemia. If such an anemia is diagnosed, iron substitution is recommended (Mocan et al. 1999; Hüdaoglu et al. 2006). The mental prognosis for breath-holding spell attacks is good. A detailed history often shows familial incidence. Table 2.3

Table 2.3 Breath-holding spell attacks vs. epileptic fits

	Cyanotic breath-holding spell attacks	Pale breath-holding spell attacks	Tonic-clonic seizures
Family history	Frequent +	Frequent +	Variable
Age	Baby – infant	Baby – infant	Any age
Trigger	Anger, vexation, pain	Sudden unexpected stimulus	Sleep deprivation
Symptoms	Loud crying, dead-space ventilation, apnea, loss of consciousness, opisthotonus, short muscle spasms	All shorter	Tonic-clonic spasms, loss of consciousness
ECG	Primary tachycardia	Primary bradycardia / asystole	Ictal neuronal discharges
EEG	Normal	Normal	Typical epilepsy pattern

compares the symptoms of breath-holding spell attacks with those of epileptic fits.

Key symptoms: Emotionally triggered, dead-space ventilation, longer "run-up" time, cyanosis, loss of consciousness.

Cataplexy/narcolepsy

Narcolepsy is a disorder rarely seen in infants; however, single cases have been reported in patients under four years of age. In 30% of all cases, the disorder begins under 15 years of age with emotionally triggered cataplectic states. The full-blown syndrome includes symptoms such as increased drowsiness during the daytime, cataplexy, hypnagogic hallucinations and sleep paralysis. Sleep paralysis is characterized by persisting muscular atonia from REM sleep, which can be interrupted by body contact (Black et al. 2004). Diagnosis can be made using the multiple sleep latency test and by detection of the HLA antigen DR2 and Dqw1, found in 90% of affected patients, combined with reduced hypocretin (Overeem et al. 2002).

Key symptoms: Emotionally triggered loss of tone, sleep paralysis.

Benign sleep myoclonus

Benign sleep myoclonus in babies can only be diagnosed clinically. The main symptoms are focal, multifocal or generalized bilateral synchronous myoclonus. Myoclonus commonly affects the upper extremities and occurs in non-REM sleep, beginning often when the baby is only a few days old. It can occur in frequent clusters. An EEG reveals no pathological discharges. Myoclonus can be interrupted immediately by waking the infant. The children have no neurological impairment. The myoclonus generally ceases when the child reaches an age of about six months, and symptoms seldom persist longer (Coulter & Allen 1982).

Key symptoms: Myoclonus during non-REM sleep, interruptible by waking the infant.

Benign early infantile myoclonus

Benign early infantile myoclonus occurs in healthy children during the first year of life (age 3 to 15 months). Characteristic symptoms are neck-bending movements accompanied by shaking of the head and contraction of the axial muscles.

Symptoms occur only when the child is awake, several times a day, and frequently in clusters, which makes it easy to mistake for infantile spasms caused by West's syndrome. It may be accompanied by facial grimacing. EMG examinations have shown tonic discharges lasting longer than 200 ms. These tonic discharges are not typical for myoclonus, and the term "myoclonus" is incorrect in this case. Affected children never show focal signs of a movement disorder and remain fully conscious. There is no accompanying eye deviation, and an EEG is always normal. A 24-hour EEG is only necessary to safely rule out epilepsy in exceptionally rare cases. The syndrome is self-limiting; occasionally, it only lasts for a few weeks, but always ceases by the age of two years (Preblick-Salib & Jagoda 1997).

Key symptoms: Self-limiting myoclonus of head and axial muscles in an awake state.

Pavor nocturnus

Paroxysmal panic attacks at night occur in 3.5% of children aged between 4 and 12 years. These attacks are frequently of a dramatic nature and are accompanied by frightened crying containing quite articulate speech. The children "wake up" 1 to 2 hours after falling asleep, sit up scared in bed, wander about the room and often refuse to be comforted by their parents. Their eyes are wide open and the pupils dilated. Amnesia exists for this time span. Pavor nocturnus is linked to the transition phase from non-REM to REM sleep (phase 3 and 4) and thus does not influence dreaming. The typical behavior shown by these children allows a clinical diagnosis to be made.

The syndrome is self-limiting. Stress has repeatedly been discussed as a possible trigger; however, this has not been verified. Frontal lobe epileptic seizures at night, which are often stereotypic, associated with hypermotoric features can resemble pavor nocturnus. If symptoms also occur during the day, then they can be interpreted correctly. Attacks generally cease spontaneously and seldom persist into adulthood.

Key symptoms: Occurring in first two hours after onset of sleep, agitation and confusion, refusal of parental comfort and retrograde amnesia.

Tic disorders

The clinical spectrum of infantile tic disorders is varied and ranges from simple "nervous" tics to complex

disorders that frequently show clinical manifestations that are not easily recognized (Kotagal et al. 2002).

Often bizarre movement patterns exist, with the frequency increasing with attention and decreasing when ignored. They are often of an inconsistent pattern. The most frequent movement pattern for girls is rhythmic knee buckling. There also exists the ability to suppress the tic at will for short periods of time.

Additional diagnostic measures such as EEG and further electrophysiological examinations produce no tangible pathological results. Many tic disorders are self-limiting. Depending on the psychological strain, which is often more of a problem for the parents than for the child, psychotherapy may be helpful, and in particularly persistent cases medication may be indicated (Alsaadi & Marquez 2005).

Key symptoms: Bizarre uncoordinated movement patterns, more pronounced in certain situations, can be suppressed at will.

Shuddering attacks

These episodes present as a sudden change in posture, as if cold water was being poured over the body, with brief muscle contraction and shaking movements (Vanasse et al. 1976). The head is bowed or inclined to the side, the arms and legs are close to the body and the arms may be bent. The condition begins in babies and infants with varying frequency of occurrence. Attacks are self-limiting, and an EEG is always negative (Holmes & Russman 1986).

Key symptoms: Paroxysmal tension, entire body shudders.

Benign paroxysmal dizziness

This condition is a sudden emergence of dizziness, occurring between the ages of 1 and 3, in short spells of generally up to one minute duration without aura. Affected children are pale, anxious, cling to their parent or lie down so that they do not fall over, frequently accompanied by nystagmus. Children are fully conscious. Functional disorders of the labyrinth have been reported. The disorder is self-limiting by the time children reach school age. There is no need for therapy.

As with paroxysmal torticollis, a connection between benign paroxysmal dizziness and migraine is also suspected. In the history, it should always be asked if migraines run in the family. Due to the inability of a young child to give an exact description of their condition, the symptoms can easily be mistaken for a complex partial seizure. However, normal postictal behavior and the fact that there is no loss of consciousness should enable the correct classification of symptoms.

Key symptoms: Brief dizziness with vegetative symptoms, patient fully conscious.

Benign paroxysmal torticollis

Benign paroxysmal torticollis becomes manifest during the first year of life and is probably closely related to benign paroxysmal dizziness. Torticollis attacks can last between several minutes or hours and days accompanied by relapsing vomiting. Rolling eye movements may occur when the labyrinth is affected. The syndrome is self-limiting. Cerebral diagnostic imaging is required to rule out any pathological intracerebral processes when the first attack occurs.

The syndrome may run in families. A relationship to migraine is possible as many children suffer from migraines in later life, and it is therefore advisable to routinely ask whether there is a history of migraines in the family. In the differential diagnosis of an initial attack, this syndrome may be confused with the Sandifer syndrome or paroxysmal vertigo, as the symptoms are similar (Deonna & Martin 1981).

Key symptoms: Paroxysmal wryneck of varying duration, vomiting, seldom eye movement disorders.

Sandifer syndrome

Sandifer syndrome is associated with spasmodic torsional dystonia, chiefly involving the neck and back, accompanied by body-rocking movements and mood changes. There is often a time link to meals, with symptoms developing 30 minutes following ingestion. Further symptoms include regurgitation or retching. Diagnostic examinations should concentrate on gastroesophageal reflux, hiatus hernia or esophageal dysmotility, as incidence of these symptoms is common. Symptoms cease following operative correction of reflux or hernia. In many cases, vomiting persists into infancy (Somijit et al. 2004; Lehwald et al. 2007).

Sandifer syndrome is frequently mistaken for tonic epileptic fits, particularly in retarded children with epilepsy or dystonic seizures. An isolated tonic increase has only been determined as an epileptic phenomenon in 30% of cases (Kabakus & Kurt 2006; Kostakis et al. 2008).

Key symptoms: Paroxysmal tonic increase involving body-rocking, time link to ingestion, no loss of consciousness.

Benign tonic upward gazing

This rare paroxysmal upward gazing in infancy is characterized by tonic upward eye movement of varying duration or intermittent occurrence (Ouvrier & Billson 1998). If a downward glance is attempted during attack, a down-beat nystagmus occurs; horizontal eye movement is normal. Occasionally, this eye movement disorder is accompanied by ataxia. There may rarely be a family history of the disorder, and the syndrome is self-limiting. L-DOPA therapy has been reported to have positive effects (Echenne & Rivior 1992).

Key symptoms: Paroxysmal upward gazing, self-limiting.

Self-stimulation

Self-stimulation or masturbation in infancy is characterized by paroxysmal stereotypical movement patterns involving pressing the thighs together rhythmically and rocking the body to-and-fro simulating copulation, and is found more frequently in girls than in boys. In addition, vegetative symptoms such as sweating, reddened face or irregular breathing can occur; children are fully conscious – girls more frequently than boys. The children never stimulate their genitals manually (Nechay et al. 2004).

Self-stimulation frequently occurs in situations where the children receive no attention. The children do not like being interrupted during self-stimulation and often react irritably. They seem to enjoy the situation. The EEG is always normal and the syndrome is self-limiting. Parental reassurance is most important. Of course, self-stimulation also occurs in children with epilepsy, where the symptoms are frequently mistaken for an epileptic seizure (Yang et al. 2005).

Key symptoms: Rhythmic pressing together of thighs, lack of manual genital stimulation, autonomous accompanying phenomena.

Nodding spasm – spasmus nutans

As with many paroxysmal movement disorders, nodding spasms are a self-limiting disorder affecting infants that involves the triad of head-nodding, torticollis and asymmetric ocular nystagmus; monocular or dissociated nystagmus can also occur. Head-nodding is a compensatory vestibulo-ocular reflex required to avoid visual distortion. The symptoms persist in older children only in exceptional cases. As various retinal and intracerebral complications such as retinitis or diencephalic processes have been reported, an ophthalmological examination and an MRI of the CNS should be performed to rule out associated disorders.

Key symptoms: Head-nodding, torticollis and nystagmus

Paroxysmal kinesigenic choreoathetosis

Characteristic symptoms for this condition are short, bizarre, dystonic choreiform movements of single or whole muscle groups, occasionally with ballism initiated by abrupt body movements. Emotional situations or hyperventilation are seldom the trigger. Patients remain fully conscious during an attack, occasionally experiencing a brief aura, but there is no postictal impairment. The attacks are of short duration and can occur up to 100 times a day. Children between 5 and 15 years of age can be affected. An EEG shows no epileptic pattern.

Symptoms can be suppressed by low-level doses of antiepileptic medication such as phenytoin, carbamazepine/oxcarbazepine or valproic acid. The syndrome ceases spontaneously between the third and fourth decade of life. The disorder is hereditary and is autosomal dominant; if parents have no symptoms, they should be questioned about symptoms during childhood (Vidaillhet 2000).

Table 2.4 shows the differential diagnosis of paroxysmal dystonic – choreoathetoid movement disorders.

Key symptoms: Dystonia, always triggered by movement, patients remain fully conscious.

Psychogenic non-epileptic seizures

Up to 20% of epileptic children suffer from psychogenic, non-epileptic seizures. Young age does not prevent psychogenic seizures, so this diagnosis must also be borne in mind where children under six years of age are concerned (Scheepers et al. 1998; Uldall et al. 2006). Whenever the described symptoms give reason to doubt whether it is a true form of epilepsy and the EEG findings produce no pathological findings,