Funny Turns (Non-Convulsive Paroxysmal Disorders)

Definition / Supporting Information

Non-convulsive paroxysmal disorders ('funny turns') occur with a wide range of clinical features that may mimic epileptic seizures in children. Breath-holding episodes, syncope, benign paroxysmal vertigo, shuddering attacks, benign neonatal sleep myoclonus, night terrors, and narcolepsy may all manifest as 'funny turns', and distinguishing these from epileptic seizures is important so that the child is not treated inappropriately with anticonvulsant medication.

Essential History

A thorough history with detailed open and closed questioning of an eye witness to the events is required.

Ask about:

- Situation of events
  - Identify an eye-witness if at all possible.
  - What was the patient doing at the time?
  - Associated triggers
    - For example, association with feeding?
  - Did the witness see the beginning?
  - Did the patient retain awareness?
  - Was the patient able to communicate throughout?
  - Were there abnormal eye or limb movements?
  - Was posture retained?
    - If lost, at what stage?
  - Did the patient become stiff or floppy?
  - Was there a change of colour?
  - Duration (often overestimated)?
  - Did the patient recover immediately or need to sleep?
  - If more than one event – were they all identical?

'Red Flag' Symptoms and Signs

Ask about:

- The presence of seizure-like movements
  - Note that generalised tonic–clonic, clonic, or myoclonic movements may be seen in any form of syncope.
- Focal neurological signs such as weakness or sensation

- Drowsiness / disorientation / confusion after event

Look for:

- Evidence of underlying cardiac disease
  - Cardiac murmur may indicate aortic or pulmonary stenosis, or hypertrophic cardiomyopathy if murmur becomes audible only when the patient is upright
  - Heart rate and blood pressure
- Altered mental status (see Altered Conscious Level)
- Any abnormality on detailed neurological examination

Differential Diagnosis / Conditions

See also Differential diagnosis of epilepsy in children, young people and adults [NICE clinical guideline 137, Appendix D].

- Epileptic seizures
- Cardiac arrhythmias
- Breath-holding spells (age 1–5 years) can be divided into two types:
  - Cyanotic (also known as blue breath-holding attacks)
    - More common than the pallid form
    - Usually precipitated by frustration or anger
    - During spells, children cry vigorously and then hold their breath in expiration
    - Apnoea is followed by cyanosis, with opisthotonic posturing and loss of consciousness
    - Recovery is usually quick, with return of respiration and consciousness within 1 minute.
  - Pallid (also known as reflex anoxic seizures)
    - Usually provoked by sudden fright or minor injuries
    - The child gasps or cries briefly and then abruptly becomes quiet, loses consciousness, and becomes pale and limp
      - He or she may develop clonic jerks
- Syncope
  - These children are usually older than 8 years
    - If a child has evidence of syncope on exercise, consider cardiac assessment.
  - Often history of unpleasant precipitating event; from standing
  - Patients have pre-syncopal symptoms that may include:
    - Light-headedness
    - Anxiety
    - Sweating
    - Nausea

        - Generalised numbness
        - Visual changes described as constriction or darkening of vision
        - Observers may notice marked pallor and clammy skin
    - Pre-syncopal symptoms are followed by loss of consciousness and slumping to the floor
        - Usually from standing
        - Once the patient is recumbent and cerebral perfusion is restored, consciousness returns within a few seconds.
        - If the patient is held with the head above the body and cerebral perfusion is not restored, clonic movements may occur.
        - May be associated incontinence
        - Patients may be tired but are not disoriented or confused after an episode of syncope
- Benign paroxysmal vertigo
    - Short episodes of intense vertigo on moving the head in certain directions (age range usually 1–5 years)
    - Characterised by abrupt onset
        - The child appears fearful and cannot maintain normal posture and gait
        - The child may seek support and clutch the parent, then abruptly sit down or fall.
        - In severe cases, the child may be limp and incapable of using the extremities.
        - Pallor is usually apparent
        - Vomiting and nystagmus sometimes occur
    - An episode typically lasts < 30 seconds
        - In rare cases, may last minutes
        - A brief period of postural instability may follow that usually resolves within a few minutes
        - Consciousness is not altered during the episode
        - The child rarely feels sleepy afterwards
    - Careful history and physical examination are required to differentiate from:
        - Brain stem lesions
        - Posterior fossa tumours
        - Epilepsy
- Benign neonatal sleep myoclonus
    - Myoclonic jerks begin in the first month of life, sometimes as early as the first day.
    - Myoclonus presents only during quiet sleep and disappears when the infant awakens.

  - Jerks occur every 2–3 seconds for several minutes and have been reported to last up to 12 hours.
  - Jerking movements may begin bilaterally or start in one extremity, then progress to involve the other extremities.
  - Neonatal sleep myoclonus is differentiated from seizure disorder by:
    - History of episodes during sleep only
    - Otherwise normal infant
- Night terrors
  - Affected children often have a family history of either night terrors or another sleep disorder.
  - Episodes usually occur during the first 2 hours after falling asleep, at a consistent time.
  - Child sits up in bed abruptly and screams or talks unintelligibly.
  - If the child's eyes are open, they have a glazed look.
  - During the episode, the child appears to be hallucinating and does not respond to parents.
  - Response of the sympathetic nervous system includes tachycardia and excessive sweating.
  - In some cases, the child may sleepwalk.
  - Episode usually lasts 10 minutes; the child then relaxes and abruptly falls back to sleep.
  - On awakening, the child does not remember the episode.
  - Nightmares should be differentiated from night terrors.
    - Nightmares occur during REM sleep, and therefore later in the night
    - Nightmares are associated with easy arousal and recall of the content, or at least the occurrence, of the nightmare
- Shuddering attacks
  - The pathophysiological mechanism is unclear
  - Attacks have been postulated to be an expression of an essential tremor
- Gastro-oesophageal reflux
  - Sandifer's syndrome is spasmodic torsional dystonia related to feeding with arching of the back and rigid opisthotonic posturing, mainly involving the neck, back, and upper extremities in children with gastro-oesphageal reflux, oesophagitis or a hiatus hernia.
  - See Gastro-oesophageal reflux disease [NICE guideline NG1]

Investigations

To be undertaken by non-specialist practitioners (eg, General Practitioner (GP) Team):

- Confirm normal neurological and developmental examination
- Ask parents to make a video recording of the episodes

To be undertaken by specialist practitioners (eg, Emergency Department / Paediatric / Paediatric Neurology / Paediatric Cardiology Team(s)):

- An electroencephalogram (EEG) should be performed only to support a diagnosis of epilepsy in children and young people.
- Focal neurological signs or symptoms and / or abnormalities on neurological assessment
  - Consider EEG and / or central nervous system (CNS) imaging (preferably magnetic resonance imaging) (see The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care [NICE clinical guideline 137])
- Breath-holding episodes
  - Consider full blood count for pallid breath-holding episodes, as anaemia has been described as a contributing factor
- Syncope
  - If atypical features are present (including a young age), a cardiac evaluation, including Holter monitoring, may be appropriate.
    - Absence of a precipitating factor
    - Confusion after the episode
  - Evaluation with tilt-table testing can be helpful for children who have unexplained syncope.
- Narcolepsy
  - Sleep studies are important in diagnosing narcolepsy.

Treatment Approach

To be undertaken by non-specialist practitioners (eg, GP Team) or specialist practitioners (eg, Emergency Department / Paediatric / Paediatric Neurology / Paediatric Cardiology Team(s)):

- If any diagnostic doubt, refer for specialist opinion rather than pursue investigation
- Reassure parents and carers that breath-holding episodes, syncope, benign paroxysmal vertigo, shuddering attacks, benign neonatal sleep myoclonus, and night terrors are time-limited and will not lead to a learning disability or epilepsy
  - Narcoplepsy is the exception
    - This is a lifelong disorder that must be accurately diagnosed and subsequently treated.

Specific Treatment

- Breath-holding spells
  - Anticonvulsants should not be used
    - They are not effective in treating either cyanotic or pallid breath-holding spells.

  - Cyanotic episodes
    - Often precipitated by temper tantrums, anger, and frustration
    - Advice about behaviour management may be helpful
  - Pallid breath-holding spells
    - Treating anaemia may reduce the incidence of the episodes
- Syncope
  - Teach the patient and family about managing an episode
  - For pre-syncopal symptoms:
    - The patient should sit or lie down as soon as symptoms begin to prevent progression to loss of consciousness.
    - If the patient loses consciousness, place the child in a recumbent position, with the head lower than the trunk.
    - Parents should be cautioned against picking up a child who has fainted, as this may prolong the period of unconsciousness.
- Benign paroxysmal vertigo
  - In most cases, no treatment is necessary as it is a self-limiting condition.
  - Anticonvulsants are not effective.
  - Antihistamines (eg, cinnarizine) have been used in some patients who have frequent episodes.
    - Some have noted an apparent reduction in the number of episodes.
    - Assessing the effect of therapy accurately is difficult because the frequency of attacks varies.
- Benign neonatal sleep myoclonus
  - No treatment is necessary
  - Reassure parents that these infants do not subsequently develop epilepsy or cognitive delay
- Night terrors
  - The nature of the episodes should be explained to the parents.
  - Parents tend to try to wake and reassure the child.
    - Attempts to awaken the child are not helpful and may increase agitation.
    - If the episodes continue to occur at the same time of night, it may be more helpful to wake the child just before this time.
  - Parents should be told that the child is not aware of their presence.
  - If the child is sleep deprived as a result of night terrors, parents should take steps to increase the amount of sleep the child is getting.
  - Sleep hygiene advice is also useful
    - For example, a pre-bed routine and no distractions in the bedroom area

  - If night terrors persist despite adequate sleep:
    - A sleep study may be needed to evaluate for obstructive sleep apnoea, which can trigger night terrors.
  - In most cases, no medication is indicated
- Shuddering attacks
  - In most cases, no treatment is necessary
  - Anticonvulsants are ineffective and should not be used

To be undertaken by specialist practitioners (eg, Paediatric (with advice from Paediatric Neurology) / Paediatric Neurology Team(s)):

- Night terrors
  - If episodes are frequent or severe, after full evaluation and in consultation with Paediatric Neurologist / Sleep Physician, medications may be helpful
    - Benzodiazepines
    - Tricyclic antidepressants
- Pallid breath-holding spells
  - Atropine is effective for pallid breath-holding episodes, but its use is rarely warranted.
- Narcolepsy
  - Requires full sleep evaluation for diagnosis
  - Central nervous system stimulants help to reduce the frequency of naps
    - Methylphenidate hydrochloride (unlicensed indication)
    - Modafinil (unlicensed in <18 years)
  - Tricyclic medications are used to treat cataplexy and the other associated symptoms
    - Imipramine hydrochloride (unlicensed indication)

When to Refer

Refer to paediatric specialist if:

- A diagnosis cannot be clearly established by history and physical examination
- Epileptic seizures are suspected
- Narcolepsy is suspected
- A need exists for subspecialty expertise

When to Admit

- Admission may be necessary if the child needs video EEG monitoring to evaluate an episode

‘Safety Netting’ Advice

- Advise parents and carers to record (eg, video) further episodes and seek medical advice.

Patient / Carer Information

Please note: whilst these resources have been developed to a high standard they may not be specific to children.

- Narcolepsy (Web page), the NHS website

Resources

National Clinical Guidance

The epilepsies: the diagnosis and management of the epilepsies in adults and children in primary and secondary care (Web page), NICE clinical guideline CG137, National Institute for Health and Care Excellence.

Gastro-oesophageal reflux disease in children and young people (Web page) NICE clinical guideline NG1, National Institute for Health and Care Excellence.

Suggested Resources

Please note: these resources include links to external websites. These resources may not have national accreditation and therefore PCO UK cannot guarantee the accuracy of the content.

Patient information (Web page), Syncopy Trust and Reflex Anoxic Seizures (STARS).

Living with reflux (Website), Living with reflux.

Fits (Web page – requires log-in), Spotting the Sick Child.

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Clinical Expert Reviewer: Dr Helen Cross

GP Reviewer: Dr Zoe Cameron

AAP Reviewer: Jane Meschan Foy, MD, FAAP

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