Dramatic parasomnias: recognition and treatment

As each of the many parasomnias requires its own specific treatment, confusion between them can have serious consequences. By recognizing their distinctive features, misdiagnosis can be avoided and the appropriate management decided.

Parasomnias are recurrent episodes of disturbed behaviour, experience or movements occurring predominantly or exclusively in relation to sleep. They constitute one of the three basic sleep problems, the other two being insomnia and excessive daytime sleepiness of which there are many possible causes (i.e. underlying sleep disorders).

About 30 different parasomnias (or parasomnia-like disorders) are now described in the *International Classification of Sleep Disorders* (American Academy of Sleep Medicine, 2005). Some are subtle but others are dramatic and alarming. Parasomnias can be classified as primary, i.e. primary sleep phenomena (Giglio et al, 2005), or secondary in the sense that they are manifestations of a medical or psychiatric disorder.

The issue of diagnostic error in clinical medicine as a whole has received increasing attention in recent times. It has been estimated that the rate of 'non-trivial' diagnostic error in developed countries could be as high as 15% (Graber and Berner, 2008). Studies have demonstrated the mistakes made in the initial diagnosis of obstructive sleep apnoea (Smith et al, 2002) and narcolepsy (Kryger et al, 2002). It seems probable that such errors occur with sleep disorders in general as a consequence of the relative neglect of sleep and its disorders in medical teaching (Stores and Crawford, 1998). Confusion of the parasomnias with each other may well be a common problem if it is not realized how complicated and varied behaviour can be during sleep.

The consequences of this mistake can be serious because the significance and treatment requirements of different parasomnias varies considerably. Some can be expected to remit spontaneously while others indicate a progressive disorder; some call for active intervention, whereas explanation and reassurance are all that is required for others. Patients will be referred inappropriately to various specialized clinical services if their parasomnias are not correctly diagnosed.

This article cannot be concerned with all parasomnias of a dramatic nature. Instead, prominent examples of possible diagnostic error have been chosen to emphasize that each parasomnia tends to have distinctive features, the recognition of which helps in making the correct diagnosis. This is important because advice and treatment is very different from one parasomnia to another.

Only the main clinical features and treatment requirements will be considered here in order to illustrate these issues. Detailed information is available from the references cited. Sleep disorders in general at risk of misdiagnosis are considered in Stores (2010).

Misdiagnosis of primary parasomnias

Arousal disorders

Distinctive clinical features

The common conditions of sleepwalking and the related 'arousal disorders' (Mahowald and Schenck, 2005) of sleep terrors and confusional arousals occur during non-rapid eye movement (REM) sleep, mainly early in the night.

Sleepwalking may consist of calmly walking about in a semi-purposeful manner, mumbling or talking incoherently but some sleepwalkers do much more complex things such as following complicated routes outside the house, or even driving a car. In addition, sleepwalking can take an agitated form in which (as in sleep terrors) the patient appears to be very fearful, rushing about and crying out as if escaping from danger.

Other sleepwalkers develop an eating disorder with excessive weight gain because of the amount of food they consume while still asleep at night. Yet others behave in an aggressive or destructive way, causing injury to themselves or others. Sexual or other serious offences have been committed during a sleepwalking episode (and, indeed, some other sleep disorders) (Schenck et al, 2007).

Some factors trigger arousal disorder episodes in predisposed people. These include influences which increase the amount of deep non-REM sleep, such as sleep deprivation, fever, systemic illness, and CNS-depressant medication or other substances. Other factors include sleep interruption (e.g. a full bladder, being woken forcefully, unfamiliar environment or obstructive sleep apnoea) and psychological stress. Some uncertainty has been expressed about alcohol as a precipitant. Zolpidem, antidepressants including selective serotonin-re-uptake inhibitors, lithi-

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um and neuroleptics have been associated with sleepwalking but the evidence is unclear (Pressman, 2007).

Sleep terrors mainly occur in later childhood but sometimes in adults. Classically, parents are woken by the child's piercing scream to find the child apparently terrified, with staring eyes, intense sweating, rapid pulse and cries suggesting intense distress. The same signs are seen at any age. The patient may jump out of bed and rush about frantically. Injury from colliding with furniture or jumping through windows is a serious risk. The event usually ends abruptly after a few minutes and the patient then settles back to sleep. If a person wakes up at the end of such an episode, he/she may describe feelings of primitive threat or danger, but not the extended narrative of a nightmare.

Confusional arousals occur mainly in infants and toddlers. An episode may begin with movements and moaning and then progress to agitated and confused behaviour with perhaps intense crying, calling out or thrashing about. Typically, although appearing very alert, the child does not respond when spoken to. Forceful attempts to intervene may meet with resistance and increased agitation.

Management

Parents of a child with an arousal disorder can be reassured that arousal disorders usually remit spontaneously by later childhood or adolescence. Regular sleep habits and adequate sleep should be encouraged, and other identified precipitating factors avoided where possible. The risk of injury needs to be minimized by making the environment safe.

Parents (and others) are often very alarmed and, mistakenly thinking the patient is awake and distressed, try to waken and console him/her. This should be discouraged. Waking someone from sound sleep is difficult and, if achieved, the patient will be genuinely confused and frightened. Instead, it is best if the patient is gently guided back to bed.

Behavioural and pharmacological treatments can be used for arousal disorders where the episodes are frequent, particularly dangerous, embarrassing or alarming. However, the efficacy of both types of approach has not yet been assessed adequately (Harris and Grunstein, 2009).

In some cases, associated factors such as medical or psychiatric conditions, or other sleep disorders (such as obstructive sleep apnoea) will require attention.

Nightmares

Distinctive clinical features

Nightmares are a REM ('dreaming') sleep parasomnia (Neilsen and Zadra, 2005). They consist of frightening dreams that awaken the sleeper in a highly distressed state, usually in the later part of overnight sleep when REM sleep is most abundant. 'Bad dreams' are similar but do not waken the patient.

Occasional nightmares are common. Most occur in childhood but some are lifelong. Usually there is no apparent cause and no serious significance. However, they are more common in people with anxiety states such as post-traumatic stress disorder, chronic alcoholics and when undergoing substance withdrawal. In these circumstances, the nightmares qualify to be called secondary parasomnias.

Typically, a frightening or otherwise very upsetting sequence of dream events (i.e. a narrative) is experienced, the distressing effect of which increases until the patient wakes up very frightened, fully alert and able to describe the nightmare. Fear persists making it difficult to return to sleep for some time despite being comforted.

The content of children's nightmares varies from frightening creatures at an early age to later dreams based on frightening TV or films, or events at home or school. Common adult themes include being pursued, trapped or threatened in other ways. The responsible traumatic experience is likely to be re-lived in the dream where nightmares follow trauma.

Nightmares may be spontaneous or (like arousal disorders) precipitated by illness or psychological stress. They can be associated with other sleep disorders such as obstructive sleep apnoea, sleep terrors or narcolepsy. Various medications (e.g. beta-blockers, anti-parkinsonian agents, selective serotonin-re-uptake inhibitors and amphetamines) are reported to be capable of triggering nightmares and bad dreams.

The term 'nightmare' is often mistakenly used for any sort of dramatic nocturnal episode in which the patient appears to be very frightened. For example, 'nightmare' and 'sleep terror' are sometimes used synonymously. In fact, nightmares and arousal disorders are clinically very different. The cardinal features of nightmares are that the patient describes a sequence of increasingly distressing dream events, wakes up afraid and is temporarily difficult to console. The episodes usually occur in the latter half of the night. In contrast, arousal disorders tend to occur earlier in the night, with no memory for the events or, at most, recall of fragmented impressions rather than clear narratives. In some cases, sleep terror episodes end with awakening but no detailed dream narrative is recalled – usually only an alarming experience of some kind of primitive threat such as drowning.

Management

Occasional nightmares require no special measures apart from providing reassurance and comfort, avoiding disturbing experiences before bedtime, and attention to good sleep habits which promote sound sleep – 'sleep hygiene' (Hauri, 1998).

Patients with more persistent, frequent or otherwise distressing nightmares (including those which are part of post-traumatic stress disorder) require more active intervention such as cognitive-behavioural treatment and/or, in severe cases, medication (Maher et al, 2006).

Sleep paralysis with hallucinations

Distinctive clinical features

'Isolated' sleep paralysis (Cheyne, 2005) is distinct from the sleep paralysis associated with narcolepsy. It is not uncommon but often unreported unless it is frequent. Brief episodes of inability to move or speak occur when going to sleep or on waking up. Consciousness is preserved and eye movements are possible. Respiration is usually unaffected but often there is a sensation of not being able to breathe. Episodes end spontaneously after several seconds or longer, or with external stimulation such as being touched or moved. Sleep loss or disruption, as well as alcohol ingestion are said to be associated factors.

Although actually benign, sleep paralysis can generate much anxiety and fear of having a stroke, epilepsy, cardiac disease or other forms of paralysis. Sleep paralysis can be combined with complex hallucinatory phenomena, in which case the overall experience can be so bizarre (including conversations with people or even strange creatures, as well as feelings of threat and dread, or an alien presence) that a psychotic process, especially of a schizophrenic nature, may well be mistakenly suspected (Stores, 1998). Otherwise, attention seeking or hysteria may be diagnosed.

Sleep paralysis seems to be a universal phenomenon which, especially when combined with hallucinatory experiences, in some cultures is interpreted as being caused by mystical influences such as visitation by demons, spirits or witches.

Management

Explanation and reassurance that sleep paralysis is essentially benign are important. Tricyclic antidepressants have been used for more severe or worrying cases.

Misdiagnosis of dramatic secondary parasomnias

Nocturnal epileptic seizures

Distinctive clinical features

Several types of epilepsy are closely related to the sleep-wake cycle, all of which are risk being misdiagnosed especially as sleep terrors, 'nightmares' or 'hysteria' because their clinical features include prominent behavioural changes.

Rolandic epilepsy (Gobbi et al, 2006) is an example. Most children with this common form of childhood epilepsy have seizures exclusively in relation to sleep, usually on falling asleep and around waking. Characteristic features of the seizures are abnormal hemifacial movements or sensations, strange throat sensations, excess salivation and speech impairment. These symptoms, which can be very alarming to the children and their parents, originate from discharges in the centro-temporal region of the brain.

Nocturnal frontal lobe epilepsy (Provini et al, 2000) has been described mainly in adults but also in children (Raju et al, 2007). Nocturnal frontal lobe epilepsy can be inherited as an autosomal dominant trait. Structural brain lesions are found in some cases but in most the cause is unclear.

The various types of seizures include some consisting of complex, stereotyped movements and vocalizations, with or without impairment of consciousness. Movements include kicking, hitting, rocking, thrashing, and cycling or scissor movements of the legs. There may be accompanying vocalizations such as grunting, coughing, muttering, moaning, shouting, screaming or roaring. Other presentations of nocturnal frontal lobe epilepsy include episodes that are very similar to arousal disorders.

Especially because of these dramatic manifestations, and the fact that elecroencephalograms are often normal (even during the seizures), there is a high risk of misdiagnosis. Guidelines for distinguishing nocturnal frontal lobe epilepsy from other parasomnias are available (Tinuper et al, 2007), and the frontal lobe epilepsy and parasomnias scale can help to distinguish between nocturnal frontal lobe epilepsy, arousal disorders and REM sleep behaviour disorder (Manni et al, 2008).

Management

Possibilities range from explanation and reassurance without specific treatment in many cases of Rolandic epilepsy, to epilepsy surgery if there is evidence on investigation of an operable lesion in cases of nocturnal frontal lobe epilepsy. Between these extremes, antiepileptic medication might be recommended for both of these seizure disorders if they are severe.

REM sleep behaviour disorder

Distinctive clinical features

Normally, in REM sleep (where most dreaming occurs) skeletal musculature is paralysed ('REM atonia'). In REM sleep behaviour disorder, REM atonia is lost and the patient can act out his/her dream. If the content of the dream is violent, the patient may punch, kick, jump out of bed or run about, possibly shouting, crying or swearing. As a result, the patient and family members are at risk of being hurt or injured. Such behaviour, best described by the sleeping partner, is generally out of character.

Most patients are elderly males but the condition has been described in people of both sexes and all ages (rarely) including children (Stores, 2008). The condition may be acute or chronic, often following a fluctuating course. Although some cases seem to be idiopathic (i.e. a primary parasomnia), increasingly the term 'cryptogenic' is preferred because an underlying condition is identified or eventually comes to light (Thomas et al, 2007). REM sleep behaviour disorder has a strong association with neurodegenerative disorders such as Lewy body disease, multiple system atrophy, Parkinson's disease and also with narcolepsy.

Other disorders have also been associated with REM sleep behaviour disorder as well as some forms of medication, including antidepressants and beta-blockers as well as withdrawal from hypnotics, alcohol or caffeine (Mahowald and Schenck, 2009). It can be associated with such sleep disorders as narcolepsy, obstructive sleep apnoea, periodic limb movement disorder, and also arousal disorders (called 'parasomnia overlap disorder').

REM sleep behaviour disorder may precede, perhaps by many years, the development of the underlying disorder such as Parkinson's disease. A prodromal period, over months or more, has also been described consisting of persistent sleep talking, loud vocalizations, limb twitching and limb and body jerking.

A brief self-rating screening questionnaire has been developed which might be useful as an aid to recognizing REM sleep behaviour disorder (Stiasny-Kolster et al, 2007). If REM sleep behaviour disorder is suspected, referral to a neurology or sleep disorders service is required for diagnosis, including the use of sleep studies to confirm the loss of REM atonia, as well as audio-visual documentation of the episodes.

Management

Low dose clonazepam is promptly effective in the vast majority of cases, even in the setting of a neurodegenerative disease, but it is advisable to exclude obstructive sleep apnoea which may be worsened by this treatment. Melatonin or pramipexole are alternatives.

Attention is needed to any associated condition or comorbid sleep disorder, and steps should be taken to prevent accidental injury to the patient or bed partner. Reassurance should be given that the disturbed behaviour in the REM sleep behaviour disorder episodes is not a psychological disorder and that, in most instances, drug treatment will help.

Sensitively explained regular review is important to check for any signs to suggest the emergence of an underlying neurological condition. Psychometric assessment is appropriate if intellectual deterioration is suspected.

Psychiatric conditions

A number of other dramatic parasomnias are secondary to psychiatric disorders. For example, a high proportion of patients with daytime panic attacks also have nocturnal panic attacks (Craske and Tsao, 2005) and some patients' attacks are exclusively nocturnal. As with other frightening sleep disorders, insomnia and fear of sleeping may also be a problem. Clinically, the attacks are characterized by sudden awakening in a state of high autonomic arousal with dizziness, breathlessness, sweating, trembling and palpitations, as well as a fear of an impending and possibly fatal heart attack or stroke.

If panic attacks occur only at night, they might well be misdiagnosed as some other form of dramatic parasomnia such as sleep terror or nightmare, sleep apnoea or epilepsy. The symptoms of such attacks can result in frequent visits to non-psychiatric or sleep disorder services such as neurology or cardiology. Both obstructive sleep apnoea and gastro-oesophageal reflux can produce symptoms similar to nocturnal panic attacks.

Management recommendations consist of measures used for daytime panic and other anxiety disorders, i.e. cognitive–behavioural methods and/or medication where necessary, together with (as in any sleep disorder) sleep hygiene measures as a general aid to achieving sound sleep.

Pseudoparasomnias involve episodes of disturbed behaviour while apparently asleep when, in fact, the person is awake as shown by sleep recordings made at the time. This has been reported in some patients who seem, for psychological reasons, to be unaware of what they are doing ('dissociation') but others are well aware (malingering). The behaviour during such pseudoparasomnias has been similar to arousal disorders or epilepsy and sometimes bizarre or violent. Such conditions can pose particularly difficult diagnostic challenges but, clearly, the correct diagnosis is essential to determine what particular type of help is required.

Detection and assessment of parasomnias

To detect and diagnose sleep disorders in general a sleep assessment, which traditionally has been neglected (Namen et al, 2001), is essential. Three basic screening questions for any patient are:

1. Do you sleep long enough or well enough?
2. Are you very sleepy during the day?
3. Do you do unusual things or have strange experiences at night?

The patient's bed partner or other relative should also be questioned.

Positive answers call for a detailed sleep history, an account of which is provided elsewhere (Malow, 2005); a modified approach is required for children (Stores, 2001). A screening questionnaire for use with adults (e.g. Partinen and Gislason, 1995) or younger patients (such as Owens et al, 2000) can be a useful starting point in assessment.

In order to correctly identify parasomnias, a working knowledge of the distinctive features of the various types is needed, together with (ideally) the precise details of the subjective and objective features of the individual patient's episodes from the first change to return to normality, and the circumstances in which they occur, including when they occur. Home audio-video recordings can be helpful, often revealing aspects of diagnostic significance that are missed in retrospective descriptions in the clinic.

Information from these sources may well be sufficient to at least provisionally formulate the problem correctly. In a proportion of cases, special investigations such as detailed polysomnography will be required, especially in diagnostically complex cases. In such circumstances, referral to a specialized sleep disorders service is required.

Conclusions

It is important that parasomnias are correctly recognized, preferably when they first come to medical attention, to ensure that they are correctly managed and not referred to inappropriate specialist services. This can be achieved by knowing their distinctive features and making the right enquiries. More accurate recognition and treatment of the parasomnias would be increased by improving professional education about sleep disorders in general. BJHM

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KEY POINTS

- The many officially recognized parasomnias can be confused with each other, especially those of a dramatic nature.
- This is serious because each requires its own type of advice and treatment.
- Such errors can be avoided by knowing the main distinctive features of each parasomnia and assessing a patient in detail in light of these distinctive features.
- This should help to avoid referral to inappropriate specialist services and to ensure the correct clinical management.

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