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Parasomnias

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1. Introduction

Sleep is a vital physiological process with important restorative functions. Parasomnias are characterized by undesirable physical or verbal behaviors, such as walking or talking during sleep and occur in association with sleep, specific stages of sleep or sleep-wake transitions. The category of parasomnias comprises some of the most exceptional behavior disorders because complex and apparently purposeful, goal-directed behavior is associated with a deep sleeping brain. Parasomnias occur during REM sleep, any of the four stages of non-REM sleep, and during transitions between sleep and wakefulness. Sleepwalking, sleep terrors, and confusional arousals are associated with non-REM sleep, nightmares and sleep paralysis are associated with REM sleep, while sleep starts and sleep talking are associated with sleep-wake transitions. Sleep enuresis has been observed with all sleep types.

The International Classification of Sleep Disorders subdivides parasomnias into the following four groups (Table 1): Arousal disorders, sleep-wake transition disorders, rapid eye movement (REM) stage sleep parasomnias, and other parasomnias (American academy of Sleep Medicine, 2004).

When accompanied with excessive motor activity and other complex motor behaviors, these parasomnias may significantly affect the patient's quality of life and that of the bed partner. Motor behaviors may or may not be restricted to bed but can become dangerous when the subject ambulates or is agitated. The behaviors are inappropriate for the time of occurrence but may seem purposeful or goal directed. Therefore, appropriate diagnostic and therapeutic strategies are needed (Young, 2008).

2. Causes of parasomnias

Parasomnias occur due to abnormal transitions between the three primary states of being wake, rapid eye movement (REM) sleep, and non rapid eye movement (NREM) sleep. These different states may overlap or intrude into one another, and it is the overlap of wakefulness and NREM sleep that gives rise to confusional arousals, and the intrusion of REM sleep into waking that produces REM sleep behaviour disorder (Matwiyoff et al, 2010).

Parasomnias may have genetic basis, but occurrence is usually triggered by heavy physical activity, febrile illness, sleep deprivation, excessive caffeine drinks, hypnotics, and emotional stress. Intake of alcohol increased occurrence of confusional arousal, night terror, and sleepwalking, while heavy intake of caffeinated drink increased occurrence of sleep walking in a population study (Oluwole, 2010).

Arousal disorders
Confusional arousals Sleepwalking Sleep terrors
Sleep-wake transition disorders
Rhythmic movement disorder Sleep starts Sleep-talking Nocturnal leg cramps
Parasomnias usually associated with REM sleep
Nightmares Sleep paralysis Impaired sleep-related penile erections Sleep-related painful erections REM sleep sinus arrest REM sleep behavior disorder
Other parasomnias
Sleep bruxism Sleep enuresis Sleep-related abnormal swallowing syndrome Nocturnal paroxysmal dystonia Sudden unexplained nocturnal death syndrome Primary snoring Infant sleep apnea Congenital central hypoventilation syndrome Sudden infant death syndrome Benign neonatal sleep myoclonus

Table 1. The International Classification of Sleep Disorders classification of parasomnias

Heredity was described for many forms of parasomnias but detailed genetic studies are lacking. The composition of non-REM and REM sleep was shown to have genetic roots. Especially the amount of slow-wave sleep was recently shown to be genetically predisposed by a specific gene, the retinoid acid receptor beta encoding gene (Young, 2008; Maret et al., 2005).

3. Diagnosis of parasomnias

All parasomnias more commonly affect persons who have breathing disorders during sleep. Polysomnography is appropriate for any patient with symptoms or signs of obstructive sleep apnea, such as daytime hypersomnolence, nocturnal hypoxia, loud snoring and increased neck circumference. REM behavior disorder often occurs concomitantly with degenerative neurologic illnesses that may require further evaluation. In adults, the onset of arousal disorders such as somnambulism and night terrors may reflect underlying neurologic disease. Thus, neurologic evaluation, including imaging of the central nervous system, may be indicated (Bornemann et al., 2006).

Diagnosis of parasomnias relies on a comprehensive clinical evaluation. Additional testing with polysomnogram and time-synchronized video recording may be indicated for cases that are associated with very frequent episodes, complaints of excessive sleepiness, unusual presentation, or injury to the individual or bed partner. A formal laboratory sleep study or polysomnogram with an expanded electroencephalographic montage can help distinguish among non-REM and REM parasomnias and nocturnal seizures. The latter may manifest clinically as arousals from sleep associated with vocalization and/or complex behaviours (Farid et al., 2004).

4. Epidemiology

Generally parasomnias, particularly those that are associated with non-REM sleep are commoner in childhood, but studies showed that non- REM parasomnias are not uncommon in adults. Parasomnias have been reported in approximately 4% of the adult population (Ohayon et al., 2000).

Prevalence of sleepwalking, which consists of a series of complex behaviours that are initiated during slow wave sleep and result in walking during sleep, varies from 10 per 1,000 to 145 per 1,000. In a population of adults prevalence of sleep walking was 20 per 1,000 (Guilleminault et al., 2003).

Sleep terrors, which are characterized by sudden arousal from slow wave sleep with a piercing scream or cry, accompanied by autonomic and behavioral manifestations of intense fear, are a common parasomnia in childhood. Its prevalence in children varies from 30% to 398 per 1,000, but prevalence of 22 per 1,000 was found in an adult population (Kales et al., 1980).

Nightmares are frightening dreams that usually awaken the sleeper from REM sleep. Between 10 and 20% of children experience nightmares that disturb their parents while 50% of adults have occasional nightmares and 1% have one or more nightmares per week.

Sleep paralysis consists of a period of inability to perform voluntary movements at sleep onset, hypnagogic or predormital form, or upon awakening, either during the night or in the morning, hypnopompic or postdormital. Lifetime prevalence of isolated sleep paralysis in the general population in Germany and Italy was shown to be 62 per 1,000.

Sleep enuresis is characterized by recurrent involuntary micturition that occurs during sleep. In children prevalence of sleep enuresis could be up to 250 per 1,000. In adults prevalence of nocturnal enuresis varies from 2 to 38 per 1,000.

Sleepwalking occurs more frequently in children with an estimated prevalence of up to 40 per cent in this age group. Prevalence among adults is about 4 per cent.
Prevalence of RBD is estimated to be about 0.5 per cent13. REM sleep behaviour disorder tends to affect older adults, with a mean age of onset of 50 to 60 years, predominantly affecting males.

5. Clinical features and symptoms

The disorders that are primarily discussed in this chapter are confusional arousals, sleep terror disorder, sleepwalking disorder, nightmare disorder and REM sleep behavior disorder.

5.1 Confusional arousals

Arousal disorders, including sleepwalking, sleep terrors, and confusional arousals, are the most common forms of parasomnias. They are predominantly associated with arousals from slow-wave sleep, which in turn occur most prominently in the first third of the night. They can present as one disorder or any combination of the three forms mentioned. Awakening the person during the arousal type of parasomnia is difficult; the affected individual usually will not remember the event on awakening in morning. Confusional arousals can occur throughout the night but are seen most commonly during the first half of the major sleep period when NREM density is highest. Confusional arousals are estimated to affect 4 percent of adults. It is characterized by abrupt awakenings with apparent confusion, diminished vigilance, disorientation and occasional violent or inappropriate behaviour (Farid et al, 2004).
Confusional arousal typically appears in young children up to the age of five years. Polysomnographic recordings of affected individual show clear association of confusional arousal episodes with slow-wave sleep mainly in the first part of the night. Confusional arousals usually are not harmful to the patient and are usually self-limited. Usually, there is no indication to intervene during the episodes of confusional arousal (Young, 2008).

5.2 Sleep terror

Sleep terror (pavor nocturnus) is an abrupt, terrifying arousal from sleep, usually in preadolescent boys although it may occur in adults as well. It is distinct from sleep panic attacks. These emerge when normal wake and NREM state boundaries become destabilized and elements of the waking state intrude into NREM sleep. Sleep terrors are believed to be a reaction to a frightening image that results in agitated arousal and sympathetic nervous activation. Polysomnographic recordings of these events have shown that they are associated with 2 abnormalities during the first sleep cycle: abnormally low electroencephalogram (EEG) power and frequent, brief, nonbehavioral EEG-defined arousals.
Symptoms are fear, sweating, tachycardia, and confusion for several minutes, with amnesia for the event. Demystification of these conditions and reassurance, particularly for parents of pediatric patients, is an important aspect of clinical intervention. Patients rarely remember the events in detail, but if actively probed after 4 years of age, they often report vague memories of having to act—run away, escape, or defend themselves—against monsters, animals, snakes, spiders, ants, intruders, or other threats. Children may report

feeling complete isolation and fear. Parents often describe terrified facial expressions, mumbling, shouting, and inability to be consoled.

5.3 Sleepwalking

Among arousal parasomnias, sleepwalking (somnambulism) is the most common. Sleepwalking (somnambulism) includes ambulation or other intricate behaviors while still asleep, with amnesia for the event. Sleepwalking is a complex behavior that ranges from limited and noninjurious activities to dangerous activities associated with injuries to self or others. Up to 40% of normal children have experienced at least one episode of sleepwalking and 2% to 3% of children experience it at least once a month (Klackenberg, 1971).

It affects mostly children aged 6-12 years, and episodes occur during stage 3 or stage 4 sleep in the first third of the night and in REM sleep in the later sleep hours. Despite widespread prevalence of these disorders and the recognition that they may arise from incomplete arousal, their pathophysiology is not well understood. Evidence for a strong genetic background of sleepwalking was shown in epidemiological surveys as in twin studies. Further evidence for heredity of sleepwalking is documented by the 10-fold increased prevalence of sleepwalking in relatives of patients suffering from sleepwalking. Sleepwalking in elderly people may be a feature of dementia. Idiosyncratic reactions to drugs (eg, marijuana, alcohol) and medical conditions (eg, partial complex seizures) may be causative factors in adults. During an episode of sleepwalking, a person may appear agitated or calm and behaviour may range from simple ambulation with a "glassy stare" to more complex activities such as driving. Sleepwalking may be preceded by confusional arousals or sleep terrors.

Depending on the degree of confusion, bedroom location, furniture, and strength of the subject, sleepwalking may lead to accidents and self-injury. Safety precautions should be taken for sleepwalking. These include removing dangerous objects, placing heavy drapes on glass doors and windows, and special locks on doors. Sleepwalking episodes occur in slow-wave sleep, during which time the individual is not easily arousable. Family members may gently guide the person back to the bed; strong stimuli to awaken the patient may cause resistance or aggression and are not recommended. Sleep terror and sleepwalking episodes are disturbing to parents but prepubertal sleepwalking is usually self-limited. Adult-onset sleepwalking with complicated patterns of sleepwalking, however, may contain a psychiatric component. These patients may benefit from psychotherapy, relaxation, or hypnosis (Farid et al, 2004).

5.4 Nightmares

Nightmares are vivid nocturnal events that cause feelings of fear and terror, with or without feeling anxiety. In most cases, a person having a nightmare will be abruptly awakened from REM sleep and is able to give a detailed account of what he dreamt about. Also, the person having a nightmare has difficulty returning to sleep. Episodes typically occur in the latter half of the night. Following the awakening, the individual becomes fully alert and profoundly anxious. There is vivid recall of the preceding dream as well as difficulty returning to sleep. Compared to sleep terrors, there is less autonomic activation, and tachycardia and tachypnea, if present, are not as severe. Episodes can be precipitated by

illness, traumatic experiences, and alcohol and medication use, such as antidepressants and beta-antagonist antihypertensive agents.

Nightmares affect 20 to 39 percent of children between five and 12 years of age. Contrary to popular belief, frequent nightmares in children do not suggest underlying psychopathology. Nightmares and night terrors in children are usually disturbing to parents and family members; therefore, proper diagnosis and education of family members are important components of management. It is essential to control the environment by removing dangerous objects and providing barriers to prevent escape from a safe sleeping environment. Reassurance and support are often the only therapy required because these disorders rarely, if ever, reflect underlying illness and usually disappear with maturity. Pharmacologic intervention is not usually indicated; in fact, it should be discouraged because it may contribute to further sleep disruption. Behavioral methods for treatment of frequent nightmares are effective in older children.

5.5 Rapid Eye Movement (REM) sleep behavior disorder

REM sleep is characterized by a paucity of muscle activity with near complete somatic muscular atonia. REM sleep behaviour disorder is characterized by the intermittent loss of REM atonia due to disinhibition of normally inhibitory mid-brain projections to spinal motor neurons. This, in conjunction with an active dream state, results in behavioural release and the apparent "acting out of dreams". Abnormal behaviours include sleep talking, yelling, limb movement, and complex motor activities. Patients with REM sleep behaviour disorder arouse from sleep to full alertness often with complete recall of fearful dream content, which may involve being chased or attacked. The motor behaviour exhibited tends to correlate with dream content. REM sleep periods typically occur in the latter half of the night. The most common symptom at time of presentation is injury of the patient or bed partner. As a result of the behaviors, bed partners often simply move to another bed or room. Also, patients and families may have a sense of guilt or shame regarding the behaviors, even though the behaviors may not be consistent with patients' personalities. This is particularly true when sexual behaviors are involved. Sleep disruption and daytime sleepiness are often part of the history. REM sleep behaviour disorder tends to be a disease that occurs in older men, although women and people of all ages may be affected. The reason for the strong predominance toward men, with an approximately nine-to-one men-to-women ratio, is not clearly known. The average age of onset is between 52.4 years to 60.9 years. Unlike those who experience sleep terrors, the victim will recall vivid dreams. The frequency of these episodes varies from once every few weeks to several times a night. Episodes tend to occur 90 min or more after sleep onset, when the first REM period typically begins. (Mahowald et al., 2005).

REM sleep behavior disorder has been linked to a number of other neurological conditions; thus, a careful review of systems and a physical examination are crucial. Polysomnographic monitoring in patients with REM sleep behavior disorder reveals increased tonic and/or phasic electromyographic activity, often accompanied by muscle twitching, extremity flailing, or vocalization during REM sleep. REM sleep behavior disorder is often associated with a growing number of underlying neurologic disorders, and may be induced by numerous medications, particularly selective serotonin reuptake inhibitors (Boeve et al., 2004).

REM sleep behavior disorder can be controlled with medication. Clonazepam is the mainstay in the treatment of REM sleep behavior disorder and leads to either a complete or partial response in approximately 90% of cases. Before it is prescribed, the potential benefits of treatment should be weighed against the possible side effects. Other medications have been tried when clonazepam is not effective or is poorly tolerated. Discussions related to safety are very important, because precautionary measures may prevent serious injury to the patient or family members (Schenck et al., 2002).

5.6 Other parasomnias

Ten disorders are classified under this category (Table 1). The most common are sleep bruxism, sleep enuresis, and primary snoring.

Sleep bruxism is the third most common parasomnia and it can be bothersome to the bed partner. Bruxism is not a dangerous disorder. However, it can cause permanent damage to the teeth and uncomfortable jaw pain, headaches, or ear pain. Approximately 8.2% of people experience it at least once a week. Sleep apnea and anxiety disorders are the most prominent risk factors for bruxism. Bruxism could be a reflex to open the airway after an apneic or hypopneic event. Bruxism may improve with treatment of sleep apnea with continuous positive airway pressure. Sleep bruxism does not have a definite cure. The goals of treatment are to reduce pain, prevent permanent damage to the teeth, and reduce clenching as much as possible. Stress reduction, relaxation, biofeedback, hypnosis and improvement of sleep hygiene have been tried with no persistent or significant improvement. To prevent damage to the teeth, mouth guards or appliances (splints) have been used since the 1930s to treat teeth grinding, clenching, and TMJ disorders. A splint may help protect the teeth from the pressure of clenching. Pharmacologic interventions are indicated for short-term management of patients who experience complications of sleep bruxism, including pain in the temporomandibular joint. Benzodiazepines could be effective because of their muscle-relaxing and anti-anxiety properties. Additionally, they increase the arousal threshold that could precede teeth grinding. (Farid et al., 2004)

Sleep enuresis, more commonly known as bedwetting, refers to the lack of ability to maintain urinary control during sleep. This recurrent involuntary urination is also called nocturnal enuresis, which is characterized by at least two occurrences per month in 3 to 6 years old infants and at least one occurrence per month for older children. Sleep enuresis is observed in 10% of children at the age of 6. The prevalence decreases with age. Approximately 77% of children had enuresis when their parents were enuretic, whereas 44% of children with one parent who was enuretic developed enuresis. Simple behavior modifications can be very effective treatments for children with enuretic episodes. For example, intake of liquids and dietary bladder irritants such as citrus products should be discouraged before bedtime. Taking note of when the enuresis actually occurs, and waking and taking the child to toilet before that hour, can also be very helpful Matthias et al., 2002).

Psychological treatments such as encouragement of self-reliance, participation in management, inculcation of self-respect and responsibility are also recommended by many experts. Physical punishments and coercion, on the other hand, are considered to be the most counterproductive measures and should be avoided at all costs.

Using devices such as bedwetting alarms and moisture alarms, combined with bladder muscle exercises, dietary changes, retention control training etc can also be helpful remedies in treating sleep enuresis. Education, encouragement, and patience are prudent approaches for younger children. For older children who may be embarrassed by the occurrences, and who may be affected by the emotional concerns, more aggressive treatment is recommended. Biofeedback, including enuresis alarms, arousal training and desmopressin have been tried with prominent success rates, although they are associated with high relapse rates. Hypnotherapy and imipramine have been somewhat helpful in the management Schenck et al., 1996).

Primary snoring is reported in 40% to 50% of people over the age of 65 and approximately 25% of the middle-age group. Snoring is usually a symptom of sleep disordered breathing. Oral appliances and otolaryngologic procedures, including velopharyngeal surgery, can effectively resolve snoring. Most of the studies on oral appliances are conducted for treatment of obstructive sleep apnea syndrome, with no clear data on primary snoring. They have decreased the frequency of snoring by 50%.

6. Treatment options

The primary therapy for disorders of arousal is reassurance and prevention. For most, the disease course is usually benign and tends to resolve spontaneously with time. It is essential that both the patient and bed partner be educated about safety precautions for the home and bedroom environment, such as reducing or eliminating potential sources of injury (e.g., relocating the bedroom to a room on the ground floor, securing doors, using heavy draperies over the windows, removing mirrors, and keeping the floor free of objects that the sleepwalker might potentially trip over). Bed partners should be counseled not to attempt to stimulate the patient during an episode as this may trigger violent behaviour.

A trial of sleep extension or scheduled awakening may be considered. With scheduled awakening, the patient is awakened just before the typical time of the parasomnia episode and thereafter allowed to return to sleep.

Relaxation training and guided imagery may be helpful strategies for some patients, especially those with disorders of arousal or rhythm movement disorders.

When the events are frequent or particularly dramatic, medication with a long- or medium-acting benzodiazepine, such as clonazepam, at bedtime is effective therapy in most cases of non-REM disorders of arousal and REM sleep behavior disorder. In non-REM disorders, pharmacologic agents that have been used with some success include paroxetine and trazodone and low-dose benzodiazepines. Typically, medication should be used in combination with nonpharmacologic treatments after such techniques have been tried and found to be ineffective and only when the sleep disorder is affecting daytime function.

7. Conclusion

Although parasomnias can be distressing and it is important to recognize that parasomnias are diagnosable and treatable in the vast majority of patients. With recent understanding of the sleep stages and transition of these stages, many of the parasomnias are readily diagnosable and treatable.

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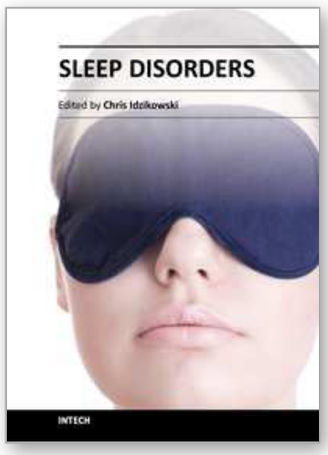

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For progress to be maintained in a clinical field like sleep medicine, unimpeded, unrestricted access to data and the advances in clinical practice should be available. The reason why this book is exciting is that it breaks down the barriers to dissemination of information, providing scientists, physicians, researchers and interested individuals with a valuable insight into the latest diverse developments within the study of sleep disorders. This book is a collection of chapters, which can be viewed as independent units dealing with different aspects and issues connected to sleep disorders, having in common that they reflect leading edge ideas, reflections and observations. The authors take into account the medical and social aspects of sleep-related disorders, concentrating on different focus groups, from adults to pregnant women, adolescents, children and professional workers.

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