

Parasomnias

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PARASOMNIAS - **undesirable behavioral (physical & mental) phenomena** that occur mainly (or exclusively) during sleep - disorders of AROUSAL, PARTIAL AROUSAL, and SLEEP-STAGE TRANSITION.

- feature of many of parasomnias is **abnormal muscle activation behaviors** during sleep.
- main presenting complaint is related to behavior itself (i.e. parasomnias do not cause insomnia or daytime sleepiness).

I. AROUSAL disorders - behaviors related to **sleep stages 3-4** (occur during first third of night when deep sleep predominates):

1. Confusional Arousals
2. Sleepwalking (s. Somnambulism)
3. Sleep Terrors (s. Pavor Nocturnus)

- usually affect **children & adolescents**; resolve during adulthood (sometimes may carry over into adulthood or arise secondary to medications).

II. SLEEP-WAKE TRANSITION disorders - occur at **sleep onset**, during transition from wakefulness to sleep (some also occur during wakefulness):

1. Rhythmic Movement Disorder (nocturnal head banging [jactatio capitis nocturna], body rocking, head rolling)
2. Hypnic Jerks (s. Sleep Starts)
3. Sleep Talking (s. Somniloquy)
4. Nocturnal Leg Cramps

- often affect **healthy individuals**.

III. Parasomnias associated with REM Sleep - occur later in night, when **REM sleep** predominates:

1. Nightmares
2. Sleep Paralysis
3. REM Sleep Behavior Disorder (RBD)

4. Impaired Sleep-Related Penile Erections
5. Sleep-Related Painful Erections
6. REM Sleep-Related Sinus Arrest

IV. OTHER parasomnias

1. Sleep Bruxism
2. Sleep Enuresis
3. Sleep-Related Abnormal Swallowing
4. Nocturnal Paroxysmal Dystonia
5. Sudden Unexplained Nocturnal Death Syndrome
6. Primary Snoring
7. Infant Sleep Apnea
8. Congenital Central Hypoventilation Syndrome
9. Sudden Infant Death Syndrome
10. Benign Neonatal Sleep Myoclonus
11. Other Parasomnia NOS

AROUSAL DISORDERS

- **impaired ability to arouse fully from slow-wave sleep** → confusion and amnesia during and following arousal.

- related to **sleep stages 3-4** - occur during first third of night (when deep sleep predominates).
 - peak INCIDENCE at **ages 4-10 years** (esp. boys) - amount of slow-wave sleep is at its peak
 - increased PREVALENCE in first-degree relatives - genetic factors may play role.
 - almost always self-limited - **resolve during adulthood** (sometimes may carry over into adulthood* or arise secondary to medications).
 - * underlying psychologic disorder or alcoholism should be considered.
 - risk factors: Tourette syndrome, post-traumatic stress disorder, violent & abusive families.
 - may be precipitated / aggravated by stress, sleep deprivation, fever, sleep apnea, noise.
- N.B. most affected children do not have severe psychopathology!

CLINICAL FEATURES

- exist as continuum (with considerable overlap):

1. **Confusional arousals** - low levels of motoric and autonomic activation.
2. **Sleepwalking** - motoric activation with little autonomic activation; affects 10% children.
3. **Sleep terrors** - pronounced motoric, autonomic, and emotional activation; affects 1-6% children.

SLEEP TERRORS (s. PAVOR NOCTURNUS)

- abrupt **fearful agitation** with decreased responsiveness (to parents or surroundings) and inconsolability.

- accompanied by loud, high-pitched **screaming** and attempts to leave bed or room.
- patients may fall down stairs and may react violently to attempts to restrain them.
- accompanied by **autonomic changes** (tachycardia, hyperventilation, sweating, pupillary dilation).
- after several minutes patient calms down and returns to sleep.
- **morning recall is absent** (some patients have vague recollection of terrifying situation); vs. **nightmares** (occur in REM sleep) – clear graphically remembered dreams.

SLEEPWALKING (s. SOMNAMBULISM)

- **complex automatisms** during sleep (typically getting out of bed and walking).

- some patients sit or stand in bed without getting out of bed.
- some patients prepare food and eat.
- subgroup of patients (usually young adult men) perform acts that are destructive or harmful to themselves, e.g., breaking furniture, throwing objects, climbing out or walking through window → **injuries**.
- although sleepwalkers often respond inappropriately, they sometimes follow instructions to return to bed.

N.B. somnambulists walk with open eyes and avoid obstacles; difficult to arouse, but when awakened cannot recall episodes.

CONFUSIONAL AROUSALS (s. SLEEP DRUNKENNESS, SEVERE SLEEP INERTIA)

- confusion and disorientation, with inappropriate behavior (talking / moaning / agitation) during or following arousals from sleep, typically from slow-wave sleep in the first part of the night, but also upon awakening in the morning.

- episode can be triggered by a forced awakening and may even cause violent behavior during sleep or amnesia of the episode.
- it is not considered a disorder in DSM-V.

DIAGNOSIS

- can usually be made based on **clinical criteria**.
- if diagnosis is uncertain → **polysomnography** with simultaneous **video monitoring** and several **additional EEG channels** (onset of episodes during stage 3-4 sleep).

N.B. standard polysomnography is generally insufficient for definitive diagnosis!

psychogenic dissociative episodes – EEG shows waking activity;

RBD – EEG shows REM sleep activity;

nocturnal epilepsy – EEG shows seizure activity.

MANAGEMENT

Treatment is required if behaviors are **potentially injurious** or **excessively disruptive to family**.

- confusional arousals rarely require treatment.
1. Good **sleep hygiene!**
 2. Ground floor bedrooms, window locks, absence of sharp objects or toys on bedroom floor.
 3. Bedtime **DIAZEPAM, IMPRAMINE, CLONAZEPAM**.
 4. **Hypnosis** (or other behavioral treatments) may be helpful for some patients.

SLEEP-WAKE TRANSITION DISORDERS

- occur at **sleep onset**, during transition from wakefulness to sleep (some also occur during wakefulness).

- often affect **healthy individuals**.

HYPNIC JERKS (s. SLEEP STARTS)

- myoclonic jerks at sleep onset.
- do not recur periodically.
- simultaneously run through most of body.
- sometimes associated with sensation / illusion of falling.
- normal phenomenon (experienced by most people on occasion).

RHYTHMIC MOVEMENT DISORDER

1. Nocturnal head banging (s. *jactatio capitis nocturna*)
2. Head / body rocking
3. Head / body rolling

SLEEP TALKING (s. SOMNILOQUY)

- talking during sleep (usually during arousals); little clinical significance.

NOCTURNAL LEG CRAMPS

- painful cramps of foot / calf that produce awakenings. see Mov3 p.

PARASOMNIAS ASSOCIATED WITH REM SLEEP

- occur later in night, when **REM sleep** predominates:

REM SLEEP BEHAVIORAL DISORDER (RBD)

- 1) intermittent **loss of normal muscle atonia** during REM sleep.
 - 2) **motor automatisms** (vocalizations, nocturnal shouting, movements, violent behavior, jumping out of bed) - patients appear to act out their dreams (**DREAM-ENACTING BEHAVIOR**).
 - episodes usually last few minutes.
 - patients can easily be awakened from episode without confusion and with vivid imagery reported (vs. sleep terrors).
 - timing and duration of episodes parallel distribution of REM sleep across night.
 - causes **frequent injuries** to patient or bed partner!
 - "**Jekyll and Hyde syndrome**" - **quiet peaceable behavior** during day + **violence** during REM sleep.
 - resemble *oneiric activity* in animals following selective **lesions of pontine nuclei controlling motor atonia**.
- chronic disorder with few if any remissions.
 - occurs mainly in **older persons** (vs. other parasomnias!); men > women.
 - **associated disorders**: Parkinson's disease, Alzheimer's disease, multi-infarct dementia, multiple system atrophy, olivopontocerebellar degeneration, narcolepsy, focal brain stem lesions.
 - N.B. 50% patients have no major abnormality on neurological examination or MRI (but substantial proportion of patients **develop Parkinson's disease** over next several years!!!)
 - **polysomnography** - during REM sleep atonia is disrupted by periods of increased tone and increased phasic muscle activity (even without apparent disruptive behavior).

- principal differential diagnosis - nocturnal seizures.

TREATMENT

- a) bedtime doses of **CLONAZEPAM!!!** – almost universally effective.
- b) alternatives - **IMPRAMINE, LEVODOPA-CARBIDOPA, DIAZEPAM, TEMAZEPAM, CLONIDINE, CARBAMAZEPINE.**

NIGHTMARES

- **FRIGHTENING DREAMS** during **REM sleep** (vs. sleep terrors – NREM sleep).
 - motor activity is limited, vocalization is much less intense (than in sleep terrors).
 - patients are *easily fully aroused* with *vivid dream recall*; child can be comforted by parent.
 - occasional nightmare is normal, but persistent or frequent nightmares warrant evaluation.
 - affect children more frequently than adults.
 - precipitating factors:
 - 1) frightening experiences (e.g. scary stories, television violence), particularly in 3-4 year olds who cannot readily differentiate fantasy from reality.
 - 2) fever
 - 3) excess fatigue
 - 4) alcohol ingestion.

OTHER PARASOMNIAS

HYPNOGENIC (s. NOCTURNAL) PAROXYSMAL DYSTONIA

- rare disorder of **sudden, brief, highly stereotyped DYSTONIC POSTURING and BALLISTIC CHOREOATHETOID MOVEMENTS**, arising from **NREM sleep**.
 - violent movements are *usually bilateral*, but they may predominate on one side and *patient is already awake* with eyes open during episode.
 - occur 10-40 times per night.
 - *resemble partial seizures* originating in supplementary motor area (also arise from NREM sleep).
 - major risk factors - lesions of supplementary motor area or mediobasal regions of frontal lobe.
 - **polysomnography with video-EEG monitoring** - abrupt onset from sleep with stereotyped behavior.
 - may respond to **CARBAMAZEPINE** and other antiepileptic agents.
 - course is variable;
 - spontaneous remissions may occur;
 - others have numerous nightly episodes that are refractory to all treatments.

SLEEP BRUXISM

- involuntary stereotyped repetitive forceful **TEETH GRINDING** during sleep or during arousals from sleep → disfiguring **destruction of tooth enamel and dentinum**.
 - patient is usually unaware of problem.
 - affects 10-20% of population.
 - typical age of onset is 17-20 years; spontaneous remission by age 40.
 - pathophysiology:

- a) dental abnormalities (e.g. malocclusion).
- b) central neural mechanisms.
- c) psychological factors
- treatment is dictated by risk of dental injury - **rubber tooth guard**; effective pharmacologic therapy has not been described.

SLEEP ENURESIS

- **BEDWETTING** during *slow-wave sleep* (more specifically, during arousal from slow-wave sleep) beyond expected age for bladder control; see also Psy41 p.

PRIMARY ENURESIS – bladder control was never achieved.

SECONDARY ENURESIS – bedwetting in patient who have been fully continent for 6-12 months.

- normal before age 5-6 yrs; spontaneously improves at puberty.
- PREVALENCE of primary enuresis - 30% at age 4 yr, 10% at age 6 yr, 3% at age 12 yr, **1% at age 18 yr**; rare in adulthood.
- boys > girls.
- etiology: see also 2590 p. (UROGENITAL)
 - 1) most cases of PRIMARY ENURESIS - *delay in neuromuscular maturation* (treatment is not recommended - *resolves spontaneously* before 6 yrs; spontaneous resolution rate after age 6 is 15%/yr).
 - 2) individual or family *psychopathology* – more common in SECONDARY ENURESIS.
 - 3) 1-2% cases have *organic etiology* (usually UTI; rare causes - congenital anomalies, sacral nerve disorders, diabetes mellitus or insipidus, pelvic mass) – more common in SECONDARY ENURESIS; accompanied by *daytime symptoms*!
- age threshold for treatment initiation depends on parental and patient concern (e.g. embarrassment) about problem (usually > 6 yrs):
 - a) **treat primary disorder** (e.g. psychotherapy).
 - b) **enuresis buzzer alarms** (triggered by few drops of urine) are *most effective treatment*!; requires *several weeks for success*; alarm should not be discontinued until 3 wk beyond last wetting episode.
 - c) **do not consume fluids** 2-3 h before going to bed (caffeinated beverages should be strictly limited); **awaken child to urinate** after 1-2 hours of sleep.
 - d) **bladder training exercises** (child must hold urine during day for progressively longer periods of time).
 - e) **behavioral therapy** - child assumes active role (urinating before going to bed, recording wet and dry nights, changing wet clothing and bedding himself); positive reinforcement for dry nights (e.g. star calendar, other age-appropriate rewards); child is counseled about etiology and prognosis to remove blame and guilt.
 - f) **symptomatic pharmacotherapy** (if unresponsive to counseling and alarms) for short-term relief to prevent embarrassment (camp, overnights, vacations):
 - 1) after-school **IMIPRAMINE** in less than antidepressant dose - response usually occurs in first week of treatment; after no enuresis for ≥ 1 mo, drug is tapered over 2-4 wk and discontinued; relapses are very common (H: 3-mo course).
 - 2) **oral DESMOPRESSIN** - effective for short term (4-6 wk); use cautiously (risk of hyponatremia! – better interrupt during acute illnesses).
N.B. since 2007-12-04 **intranasal DESMOPRESSIN** is no longer indicated for primary nocturnal enuresis (risk for developing severe hyponatremia → seizures → death)
 - 3) **OXYBUTYNIN**

Because of time-limited nature of disorder, use **drug holidays** at least q 6 months to assess need for continuing pharmacologic intervention.

BIBLIOGRAPHY see p. S40

Viktor's NotesSM for the Neurosurgery Resident
Please visit website at www.NeurosurgeryResident.net