



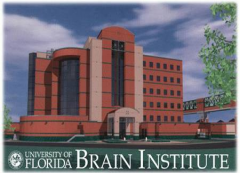
# Hypersomnias of Central Origin



# Hypersomnias of Central Origin



- Narcolepsy
  - With cataplexy
  - Without cataplexy
- Recurrent Hypersomnia
- Idiopathic Hypersomnia
  - Long sleep time ( $> 10$  hours)
  - Short sleep time ( $< 10$  hours)
- Behaviorally induced insufficient sleep syndrome
- Hypersomnia due to a medical condition
- Hypersomnia due to drug or substance
- Hypersomnia not due to substance (non-organic)
- Physiological (organic) Hypersomnolence



# Narcolepsy with Cataplexy



- A. The patient has a complaint of excessive daytime sleepiness occurring almost daily for at least 3 months
- B. A definite history of cataplexy, defined as a sudden and transient episodes of loss of muscle tone triggered by emotions, is present

*Note: To be labeled as cataplexy, these episodes must be triggered by strong emotions-most reliably laughing or joking-and must be generally bilateral and brief (less than 2 minutes). Consciousness is preserved, at least at the beginning of the episode. Observe cataplexy with transient reversible loss of deep tendon reflexes is a very strong, but rare, diagnostic finding.*

- C. The diagnosis of narcolepsy with cataplexy should be confirmed by nocturnal polysomnography followed by MSLT; the mean sleep latency on MSLT is less than or equal to 8 minutes and 2 or more sleep onset REM periods are observed. Alternatively, hypocretin-1 levels in the CSF or less than or equal to 110 pg/ml or one third of the mean normal control values

*Note: the presence of 2 or more sleep onset REM periods during MSLT is a specific finding, whereas a mean sleep latency of less than 8 minutes can be found in up to 30% of the normal population*



# Narcolepsy with Cataplexy



## Key Points

- Affects 0.02% to 0.2% in the United States and western European populations
- Onset almost always after age 5 years old and most typically between ages 15 and 25
- Most present with excessive daytime sleepiness initially
- Risk of a first degree relative developing narcolepsy with cataplexy is only 1-2%
- Ranges in mild sensation of weakness to complete postural collapse
- Sleep paralysis, hypnogogic hallucinations, and nocturnal sleep disruption are common
- Differential diagnosis includes idiopathic hypersomnia, narcolepsy without cataplexy, recurrent hypersomnia and conversion disorder
- TX:
  - EDS + Cataplexy: Xyrem
  - EDS: Nuvigil, Provigil, methylphenidate/stimulants, protriptyline
  - Cataplexy: antidepressants



# Narcolepsy withOUT Cataplexy



- A. The patient has a complaint of excessive daytime sleepiness occurring almost daily for at least 3 months
- B. Typical cataplexy is not present, although doubtful or atypical cataplexy like episodes may be reported
- C. The diagnosis of narcolepsy with cataplexy should be confirmed by nocturnal polysomnography followed by MSLT; the mean sleep latency on MSLT is less than or equal to 8 minutes and 2 or more sleep onset REM periods are observed.

*Note: the presence of 2 or more sleep onset REM periods during MSLT is a specific finding, whereas a mean sleep latency of less than 8 minutes can be found in up to 30% of the normal population*



# Narcolepsy withOUT Cataplexy



## Key Points

- Hypocretin-1 levels in the CSF are normal
- Incidence: 0.02% to 0.2% in the United States and western European populations
- Onset almost always after age 5 years old and most typically between ages 15 and 25
- Ranges in mild sensation of weakness to complete postural collapse
- Sleep paralysis, hypnagogic hallucinations, and nocturnal sleep disruption are common
- Differential diagnosis includes idiopathic hypersomnia, narcolepsy with cataplexy, recurrent hypersomnia and conversion disorder
- EDS: Nuvigil, Provigil, methylphenidate/stimulants, protriptyline



# Recurrent Hypersomnia Kleine-Levin Syndrome



- A. Recurrent bouts of excessive sleepiness of 2 days to 4 weeks in duration occurring weeks or months apart
- B. Episodes recur at least once a year
- C. The patient has normal alertness, cognitive functioning, and behavior between attacks

## Key Points

- A. May have flu like symptoms prior to 1<sup>st</sup> episode
- B. Occurs in adolescents, mostly male (4:1)
- C. During sleepy phase may sleep 18-20 hrs
- D. Behavioral abnormalities may be present with binge eating, hypersexuality, irritability and aggressiveness
- E. PSG shows decreased sleep efficiency and increased wake time
- F. EEG during the episodes, bisynchronous, generalized, moderate to high-voltage 5-7 Hz waves lasting 0.5-2 seconds and
- G. MSLT shows reduced sleep latencies but highly variable depending on patient cooperation
- H. EDS: Nuvigil, Provigil, methylphenidate, protriptyline



# Idiopathic hypersomnia with long sleep time



- A. The patient has a complaint of excessive daytime sleepiness occurring almost daily for at least 3 months
- B. The patient has prolonged nocturnal sleep time for more than 10 hours documented by interview, actigraphy, or sleep logs. Waking up in the morning or at the end of naps is almost always laborious.
- C. Nocturnal polysomnography has excluded other causes of EDS
- D. The polysomnogram demonstrates short sleep latency and a major sleep period that is prolonged to more than 10 hours in duration
- E. If an MSLT is performed following overnight polysomnography, a mean sleep latency of less than 8 minutes is found and fewer than 2 sleep onset REM periods were recorded





# Idiopathic hypersomnia without long sleep time



- A. The patient has a complaint of excessive daytime sleepiness occurring almost daily for at least 3 months
- B. The patient has normal nocturnal sleep time for greater than 6 hours but less than 10 hours documented by interview, actigraphy, or sleep logs. Waking up in the morning or at the end of naps is almost always laborious.
- C. Nocturnal polysomnography has excluded other causes of EDS
- D. The polysomnogram demonstrates a major sleep period that is normal with greater than 6 hours but less than 10 hours
- E. If an MSLT is performed following overnight polysomnography, a mean sleep latency of less than 8 minutes is found and fewer than 2 sleep onset REM periods were recorded



# Idiopathic Hypersomnia



## Key Points

- Head trauma should not be considered as a possible cause of the sleepiness
- Rarely seen before adolescence
- EDS: Nuvigil, Provigil, methylphenidate, protriptyline



# Behaviorally Induced Insufficient Sleep Syndrome



- A. Patient has complaint of EDS or, in prepubertal children, a complaint of behavioral abnormalities suggesting sleepiness. The abnormal sleep pattern is present almost daily for at least 3 months
- B. The patient's habitual sleep episode, established using history, sleep log, or actigraphy, is usually shorter than expected from age-adjusted normative data
- C. When the habitual sleep schedule is not maintained (weekends or vacation), patients will sleep longer than usual
- D. When PSG is performed, sleep latency is less than 10 minutes and sleep efficiency  $> 90\%$ . During the MSLT, a short mean sleep latency of  $< 8$  minutes (with or w/o multiple SOREMPs) may be observed