Tourette's Syndrome & Associated Daytime and Sleep Behaviors: Evaluation and Treatment Options

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Lack of sleep can make you do strange things!

Goals of Talk

• Link between daytime behaviors and nighttime sleep behaviors
• Overview of developmental sleep issues
• Review associated childhood sleep and neurobehavioral problems
• Commonsense diagnosis, management ... medication side effects etc.
Daytime and Nighttime Behaviors: Nature of the Problem

- “At least one sleep problem is found among 37% of school age children”
  
  Judith Owens, MD, MPH

“Sleep is the other side of the behavioral coin in that sleep disorders may mimic or worsen psychiatric disorders.”

Ronald E. Dahl, MD

What the Neuroscience tells us about sleep

- Sleep is a mandatory, cyclical process—important to health
- Internal, biological clock drives the sleep/wake cycle
- Sleep is a complex, critical function
- Sleep is important for consolidating information for learning
- Sleep deprivation has major consequences

The Drive to Sleep is Regulated by Biology

NIGHT OWL OR MORNING LARK?
Sleep and Physiology

Sleep Deprivation Consequences

• Inadequate sleep can cause:

> Performance
> Concentration
> Reaction Times
> Consolidation of Information for Learning

• Inadequate sleep can cause:

> Accidents and Injury
> Lapses in Memory
> Behavior Problems
> Lability in Mood
> Changes in metabolism (↑obesity?)
Common Sleep Problems Lead to Serious Consequences

- Disruption of sleep (poor sleep)
- Inappropriate timing of sleep

Daytime sleepiness

Night time Sleep Problems

- RLS
- RMD
- Parasomnias
- Delayed Phase Syndrome
- Behavioral Insomnia
- Snoring, Sleep Apnea

Sleep Deprivation

- Must be taken seriously: sleep needs vary but range from 12-13 hours for 3-6 years old to 9.25 to 9.5 for adolescence.

- Impairment the same whether: drunk, sleep deprived (<4-6hrs.) 2nd to insomnia, sleep apnea etc.

- Asthma, GERD, allergies, ear-tubes and snoring are related to sleep disordered breathing
Irritability and Sleep in Toddlers

- Total sleep time 12 - 14 hours
- Most give up 2nd nap at about one year
- Developmental issues: separation anxiety, night time fears, independent sleeping, ability to self-soothe
- Awakenings, sleep terrors: Sleep problems common (20-40%)

- Importance of bedtime routines can decrease daytime irritability and tantrums

Sleep in Pre-Schoolers

- Sleep cycles: REM/Non-REM 90 minutes
- Total sleep time: 11-13 hours
- By age 4-5, many children give up regular daytime naps
- Sleep problems that are present are at risk to become chronic

Middle Childhood (6-12 years old)

- Total sleep time: 9-11 hours (10 - 11 hours in 6-7 year olds; 9-11 hours in early adolescence)
- Sleep pattern more stable, night-to-night consistency
- Naps rare, not sleep in the daytime
- School and lifestyle influences, bedtimes, insufficient sleep
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**Sleep Changes in Adolescence**

Did You Know?
- 85% of teens get less than the minimum requirement of 8 1/2 hours of sleep
- Less Sleep = More Time
- Shortened sleep impairs learning, performance, health and safety
- 55% of fall-asleep crashes involve drivers 25 years of age or younger
- 51% of adolescents who drive report that they have driven drowsy in the past year
  - 16% of 11th graders and 20% of 12th graders drive drowsy once a week or more

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**Sleep Changes in Adolescence**

- Sleep deprivation during the week with makeup sleep on the weekend
- Sleep deprivation leads to decreased attention and focus as well as depression, emotionality lability, decreased attention and concentration.

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**Common Pediatric Sleep Disorders**

- **Insomnia**: wants to sleep but cannot
- **Parasomnias** (abnormal sleep behaviors)
  - REM-associated (nightmares, RBD)
  - Bruxism
- **Sleep Apnea**: sleepy during day, snores (throat obstruction) problem of sleep quality
- **Restless Legs Syndrome**: leg discomfort, relieved by movement, symptoms day and night
- **Rhythmic Movement Disorder** (RMD)
- **Circadian Rhythm Disorders** (Delayed Phase Sleep Disorder)
Comorbid Neurobehavior and Sleep Problems in Children

Insomnia, Parasomnias, Restless Legs Syndrome

ODD Oppositional Defiant Disorder

Obsessive Compulsive Disorder

Tourette's Syndrome

PANDAS

ADHD

ASD/Asperger's Syndrome

Motor Problems

Sleep Problems

Cognitive/Language Problems

Dyslexia/Learning Problems

Relationship of TS & ADHD to Sleep Problems

- Restless Legs/Periodic Limb Movements in Sleep
- Sleep Disordered Breathing/Apnea
- Narcolepsy
- Rhythmic Movement Disorder
- Disorders of Partial Arousal (Sleep Walking, Sleep Terrors, Confusional Arousals)
- INSOMNIA

Clusters Syndrome

- Reliable estimates suggest that nearly 50% of patients with TS have some degree of obsessive-compulsive features.
- No linkage studies for TD/OCD and ADHD, however, clinically these disorders run together

CoMorbidity - ADHD + Tics/ Tourette's

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Decreased sleep quality and increased sleep related movements in patients with Tourette's syndrome, S. Corhs et al., 2000
Sleep Related Movement Disorders

- RLS Restless Legs Syndrome
- PLMD: Periodic Limb Movement Disorder
- Sleep related leg cramps
- Sleep related bruxism
- Sleep Related Rhythmic Movement disorder
- Sleep Related Rhythmic Movement disorder due to medical condition or substance abuse
Movements in Sleep

Restless Legs Syndrome (RLS)
- Urge to move legs, accompanied by painful or uncomfortable sensations
- Urge to move legs, sensations worse with inactivity
- Urge to move legs, sensations worse at night
- Urge to move legs discomfort at rest, relieved by movement
- Severity: mild to incapacitating
- Affects 5-10% of population, women more
- Familial, often begins in childhood
- Still not well known by patients or doctors
- Common, easily diagnosed, and treatable

2003 International RLS Consensus

Dx of RLS in Children
- *Must meet all 4 essential adult criteria and
- *Must be able to describe leg sensations in own words
  OR
- *Must meet all 4 essential adult criteria and
  *Must meet 2 of 3 following:
    Sleep disturbance for age
    Parent or sibling has definite RLS
    PLMS index of > 5/hr Sleep.
**RLS/PLMD Etiology**

- **Primary**
  - Genetic component >50% of cases
  - Autosomal dominant
  - Linkage analysis to 3 loci (12q (French Canadian), 14q (Italian), 9p (American)
  - Younger age onset ("early onset" < 30)

- **Secondary**
  - Iron deficiency
    - Tyrosine hydroxylation is rate limiting enzyme for dopamine synthesis
    - Serum ferritin < 50 mcg/L; CSF basal ganglia iron stores reduced (80% of Childhood Onset associated with FE deficiency!)

**Confused with RLS**

- Leg Cramps (electrolyte disturbance, neuromuscular disorder)
- Anxiety, Stress, Akathisia (Drug effect)
- Arthritis, (JRA)
- Exercise
- Leg pains (growing pains)
- Osgood-Schlatters: tenderness of tibial tuberosity increased by activity
- Chondromalacia patella: worse on knee flexion, movement

**RLS/PLMD and ADHD**

- Clinical samples show RLS/PLMS more prevalent in children with ADHD
  - Children with ADHD as compared to non-ADHD controls have more symptoms of RLS/PLMD
    - Up to 44% of ADHD subjects have RLS symptoms
    - Up to 26% of RLS subjects have ADHD symptoms
- Same treatment for PLMD and RLS Dopamine agonists
RLS/PLMD Treatment

- Regular Sleep Schedule
- Adequate sleep for age
- Restrict or eliminate caffeine
- Treat anemia with iron supplementation for 6 weeks to 3 months
- Increase physical exercise
- Relaxation, massage and biofeedback
- Heat cold packs for pain

RLS/PLMD Pharmacological Treatment

- Dopaminergic agents
  - Levodopa/carbidopa (25/100)
  - Pramipexole, ropinrole
- Gapapentin
- Benzodiazepine (Clonazepam)
- Alpha 2 agonists (Clonidine, Guanfacine)

Drugs That Worsen RLS

- Antihistamines: Benadryl, Antivert
- Antidepressants: amitriptyline (Elavil), fluoxetine (Prozac), Paxil, Zoloft, Celexa
- Major Tranquilizers: haldol, Zyprexa
- Anti-nausea: Reglan (metoclopramide)
Insomnia: Acute or Chronic

- Insomnia: inability to get to sleep, stay asleep, wakes up early, with daytime impairment
- Acute Insomnia (<4 weeks): stress, illness
- Chronic Insomnia: >4 weeks, often years
- Treat basic problem: >60% psychological
- Psychotherapy and behavioral therapy better than medicines, not easy to treat
- Psychologists, Psychiatrists, PCP

Insomnia

- Adjustment Insomnia
- Psychophysiological Insomnia
- Paradoxical Insomnia
- Idiopathic Insomnia
- Inadequate Sleep Hygiene
- Behavioral Insomnia of Childhood
- Insomnia due to substance abuse
- Insomnia due to a medical condition

Factors Affecting the Development of Insomnia

- Conditioning
- Substance Abuse
- Performance Anxiety
- Poor sleep hygiene
- Personality
- Sleep-Wake cycle
- Circadian Rhythm
- Coping Mechanisms
- Situational
- Environmental
- Medical
- Psychiatric
- Prescription Medication
**Trial of Melatonin?**

- Melatonin is a pineal hormone that regulates sleep wake cycle.
- Recent literature suggests trial of evening melatonin may help sleep problems in children with SOI.
- Consider starting with 3-7 mg of Melatonin approximately 45 minutes -1 hour before bedtime.

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**Good Sleep Hygiene: Basics**

- Regular times for sleeping and awakening
- Maintain bedroom dark, quiet, cool
- Use bed only for sleep
- Avoid late daytime naps, bedtime routine
- Avoid at night: alcohol, caffeine, nicotine
- Sleep around 9 hours every night
- Prudent exercise and eating
- Avoid stressful situations at bed time transitions

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**Things that go bump in the Night**

Parasomnias:

Episodic nocturnal behaviors, often involve disorientation, autonomic, skeletal muscle disturbances; related CNS immaturity
Parasomnias

- Disorders of arousal from NREM sleep
  - Confusional arousals
  - Sleepwalking
  - Sleep Terrors
- Parasomnias usually associated with REM sleep
  - REM Sleep behavior disorder
  - Recurrent isolated sleep paralysis
  - Nightmare disorder
- Other parasomnias
  - Sleep enuresis
  - Sleep related groaning
  - Exploding head syndrome
  - Sleep related hallucinations
  - Sleep related eating disorder (SRED)
  - Parasomnia, unspecificied, related to medical or substance abuse

Somnambulism: Sleep Walking

- Prevalence: up to 15% of population; most between 6 and 16 years
- Age onset: usually 4 - 6 years
- Duration: one third for 5 years; 12% for 10
- Quiet vs. agitated; displacement from bed, often complex behaviors; SAFETY issues

Pavor Nocturnus: Night Terrors

- Arousal from slow wave sleep first or second episode
- Incidence: 1-3%; 10% of sleepwalkers; peaks at age 5-7 years
- Male > female
- Age onset: 4-12 years
- Frequency: often highest at onset; often higher (> once a week) with younger onset
- Usual disappearance by adolescence
Sleep-Related Breathing Disturbances

- OSA is characterized by prolonged partial upper airway obstruction, intermittent complete or partial obstruction, or both prolonged or intermittent obstruction that disrupts normal ventilation during sleep.
- Prevalence of 2-4% and is more common on children born premature.
- Higher incidence among Hispanics and African Americans.
- Equally prevalent among boys and girls.

Obstructive Sleep Apnea

What is OSA?
- Cessation of airflow with ongoing respiratory effort.
Causes of Sleep Apnea

- Age (tonicity decreases)
- Smaller-than-normal jaw
- Enlarged tonsils
- Lateral pharyngeal walls close in
- Large tongue
- Tongue moves posterior which displaces the soft palate
- Tissues that partially block the entrance to the airway

### TABLE 60-3. SYMPTOMS AND FEATURES OFTEN FOUND IN OBSTRUCTIVE SLEEP APNEA SYNDROME

<table>
<thead>
<tr>
<th>Nocturnal</th>
<th>Daytime</th>
</tr>
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<tbody>
<tr>
<td>Apnea</td>
<td>Mouth breathing</td>
</tr>
<tr>
<td>Snoring</td>
<td>Hyperventilation (excessive daytime sleepiness)</td>
</tr>
<tr>
<td>Pauses with breathing at night</td>
<td>Poor school performance</td>
</tr>
<tr>
<td>Frequent awakenings from sleep,</td>
<td>Abnormal daytime behavior</td>
</tr>
<tr>
<td>restless sleep</td>
<td>Aggression</td>
</tr>
<tr>
<td>Nightmares</td>
<td>Hyperactivity</td>
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<td>Nocturnal enuresis</td>
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<td>Nocturnal diaphoresis</td>
<td>Short attention span</td>
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<tr>
<td>Cynosis*</td>
<td>Morning headache</td>
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<tr>
<td>Near sudden infant death syndrome*</td>
<td>Weight problem (failure to thrive or obesity)</td>
</tr>
<tr>
<td></td>
<td>Frequent upper respiratory tract infections</td>
</tr>
<tr>
<td></td>
<td>Chronic rhinorrhea</td>
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<tr>
<td></td>
<td>Dysphagia</td>
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<tr>
<td></td>
<td>School and learning problems</td>
</tr>
<tr>
<td></td>
<td>Feeding difficulties*</td>
</tr>
</tbody>
</table>

Daytime Sleepiness in Children: Conceptual Framework

**Daytime Sleepiness**

- Behavioral Problems: Irritability, Low Frustration, Tolerance
- Mood Disturbance: Emotional Lability
- Performance Deficits:
  - Difficulties in school
  - Short term memory
  - Attention and Concentration

OSA
Diagnostic Sleep Study: Polysomnogram

- Required to diagnose sleep apnea
- Not usually required to diagnose RLS
- Not usually required for many circadian rhythm disorders: advanced or delayed sleep phases
- May be used to diagnose PLMD, and many parasomnias
- Sleep studies require considerable skill and expertise

PSG Hookup

Treatment Options: T&A

- Tonsillectomy & Adenoidectomy is the “gold standard” to treat OSA in for children
- Positive pressure is for refractory or severe cases not improved by surgery or Adults...
- Stents the airway open and is 100% effective
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**Treatment**

- Tonsillectomy & adenoidectomy

<table>
<thead>
<tr>
<th>Author</th>
<th>Procedure</th>
<th>N</th>
<th>Patients</th>
<th>% Cure</th>
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<td>T &amp; A</td>
<td>55</td>
<td>Child</td>
<td>94</td>
</tr>
<tr>
<td>Kadosh³⁷</td>
<td>T &amp; A</td>
<td>31</td>
<td>Obese</td>
<td>All</td>
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<tr>
<td>Suss³⁸</td>
<td>T &amp; A</td>
<td>25</td>
<td>Child</td>
<td>95</td>
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<tr>
<td>Wiet³⁹</td>
<td>T &amp; A UPPP</td>
<td>48</td>
<td>Comp</td>
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<tr>
<td>Fradko⁴⁰</td>
<td>T &amp; A</td>
<td>32</td>
<td>Child</td>
<td>Most</td>
</tr>
</tbody>
</table>

T & A = tonsillectomy and adenoidectomy; UPPP = uvulopalatopharyngoplasty.

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Talk to Your Child’s Doctor if Any of the Following Symptoms are Observed:

- A newborn or infant is extremely and consistently fussy
- A child is having problems breathing or breathing is noisy
- A child snores, especially if the snoring is loud
- Unusual nighttime awakenings
- Difficulty falling asleep and maintaining sleep, especially if you see daytime sleepiness and/or behavioral problems

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**Tips for Helping School-Age Children Sleep Well**

- Introduce healthy sleep habits, disease prevention and health promotion
- Continue to emphasize the need for a regular and consistent sleep schedule and bedtime routine
- The child’s bedroom should be conducive to sleep: dark, cool and quiet. TV's and computers should be off and out of the bedroom
- Set limits
- Avoid caffeine
- Watch for signs of chronic difficulty sleeping, loud snoring, difficulty breathing, unusual nighttime awakenings and frequent daytime sleepiness
**Childhood Behavior and Sleep Problems: Summary**

- Tics, Tourette’s syndrome is commonly associated with ADHD, OCD and anxiety in children.
- The motor restlessness and compulsive movements as well as obsessive worries or intrusive thoughts can lead to RMD and/or insomnia.
- Treating the daytime behaviors with therapy or medication may improve the night time sleep problems.
- Good sleep hygiene and a routine, “wind-down routine” are the basis of good sleep for life.

**THE END**

- Thank you and have a good night sleep!

**Websites**

- Restless Legs Syndrome Foundation: www.rls.org
- The Movement Disorder Society: www.wemove.org
- National Sleep Foundation: www.sleepfoundation.org
- American Academy of Sleep Medicine: sleepeducation.com
- Village Sleep Lab www.villagesleeplab.com (Dr. Albino, coming in late February, 2007)
Childhood Sleep Questions

- **Bed** (only for sleep?)
- **Environment** (good temperature, decreased light etc?)
- **Activity** (Wind down or wind up?)
- **Rest** (Naps during the day?)

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Keep a Sleep Diary of the Child's Sleep Habits and Patterns

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Good Sleep Habits

- Regular relaxing routine to unwind at night just before bedtime
- Avoid all products containing caffeine (soda and chocolate)
- Avoid smoking and smokeless tobacco
- Avoid use of alcohols
- No stimulating activities in the evening
- Promote calm family atmosphere prior to bedtime
**Good Sleep Habits**

- Do not allow falling asleep while watching television or video
- Establish regular exercise routine and healthy diet
- Avoid late afternoon and evening bright light
- Go to bed later on weekend, awaking within two hours of usual weekday wake time

- Avoid napping - short nap only limited to 30 to 45 minutes

American Academy of Sleep Medicine, 2002