

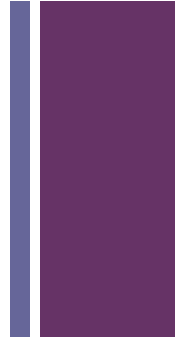


Sleep in Children and Adolescents with Williams Syndrome

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WSA Conference
July 13, 2018

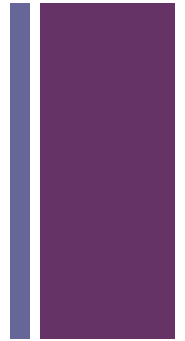


+ Overview



- What do we know about sleep in WS across development?
- Why might sleep be disrupted in WS?
- Ways to improve common sleep problems
- Questions

+ Before the Science...





+ Sleep in Williams Syndrome
Across Development

+ Sleep in Toddlers with WS

- No studies in infants < 15 months
 - Are sleep problems:
 - Characteristic of WS?
 - Related to difficulties with attention, sociability, anxiety etc.?
- Toddlers:
 - Parents 21x more likely to report that their child has a sleep problem
 - Shorter nighttime sleep duration (not naps)
 - Longer and more frequent nightwakings
 - Later bedtimes, longer to settle

+ Sleep in Toddlers with WS

■ Toddlers:

- No difference in parental involvement at bedtime BUT 8x more likely to sleep with a parent
- Parents of children with WS stronger beliefs about limit setting

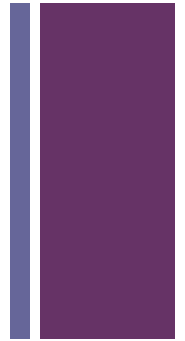
+ Sleep in Children with WS

■ Parents report:

- Difficulty falling asleep, staying still before sleep
- Restless sleep
- More frequent and prolonged night wakings

■ But not:

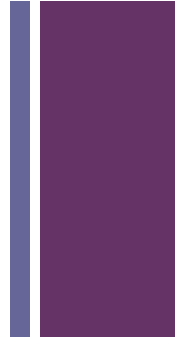
- Snoring/ breathing problems
- Daytime sleepiness
- Nap problems



+ Polysomnogram

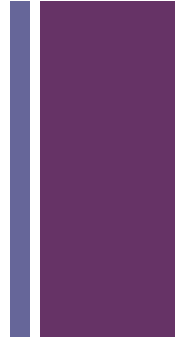
- What is it?
 - Overnight sleep study
- Awake about 15 minutes more
- Parents of children with WS, report strongly correlated with PSG

+ Sleep in Adolescents and Adults with WS



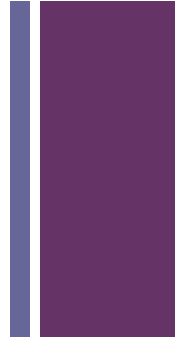
- Average 9 hours in bed, but 7.6 hours of sleep
- Reduced sleep efficiency 74.4%
- Time to fall asleep = 37.7 minutes
- Wake after falling asleep = 56.1 minutes
- No male/ female difference

+ Sleep in Adolescents and Adults with WS



- Self-report
 - Have problems with sleep 36.4%
 - Want to change something about sleep 76.7%
 - Daytime sleepiness 77.3%
 - Daytime tiredness 95.2%
 - Nocturia 90.5%
 - Snore 22.7%
 - Symptoms of restless legs 13.7%

+ Putting it all Together...

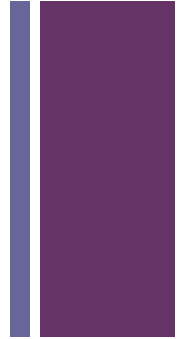


- Difficulty settling, falling asleep at all ages
- More frequent and longer night wakings at all ages
- More daytime sleepiness during adolescence/ adulthood but not in childhood
- Despite these complaints, sleep does tend to improve over time vs. other neurodevelopmental conditions



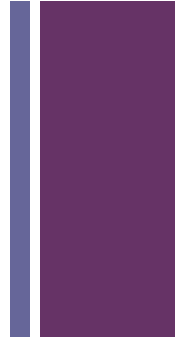
+ Reasons for Sleep Disruption
in WS

+ Melatonin



- Melatonin
 - Rises about 2 hrs before typical sleep onset
- Cortisol
 - Increase quickly upon waking
 - Lowest level in early hours of sleep
- In children with WS:
 - Overall melatonin levels same
 - Rise in melatonin at bedtime much shallower
 - Cortisol levels not as low at bedtime

+ Psychological



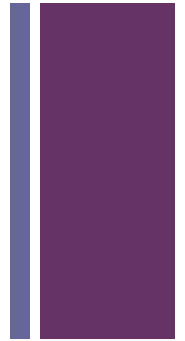
- Co-morbid conditions
 - Autism Spectrum Disorders

 - Attention problems
 - Related only to long term sleep deprivation
 - Timing of stimulant medications

 - Anxiety

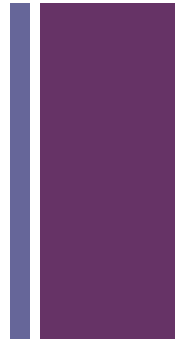
+ Sleep Disorders on PSG

- Periodic Limb Movement Disorder
 - May be higher in children with WS, mixed results
- Obstructive Sleep Apnea
 - Snoring
- Nighttime seizures



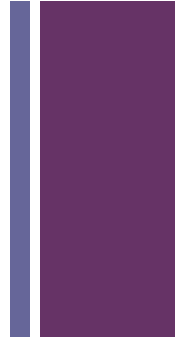
+ Other Sleep Stealers

- Asthma/ allergies (longer time to fall asleep)
- Cardiac problems (shorter sleep duration)
- Reflux
- Eczema
- Teething/ other pain
- Sleep habits





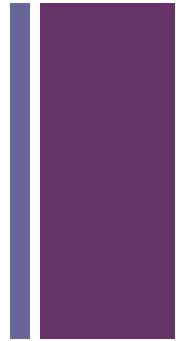
How to Improve Sleep



- There are very few “shoulds”
- The exception is that safe sleep should be a priority

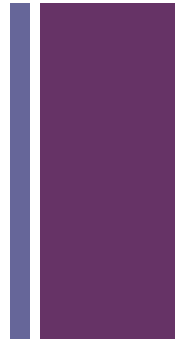
+ Safe sleep in Infants

- Separate sleep surface, same room
- Infants on their backs
- Firm surface / mattress (no soft bedding)
 - No chair or couch sleeping
- No secondhand smoke
- No pillows, lovies, blankets, or crib bumpers
- No bottle-propping (leaning bottle against a pillow or other support)



+ First Things First

- Consistent schedule
 - No more than 2 hours later on non-school days
- Bedtime routine
- Sleep promoting conditions
 - Dark
 - Blackout shades
 - Cool
 - Quiet
 - Sound machine
 - Avoid electronics
 - Avoid caffeine

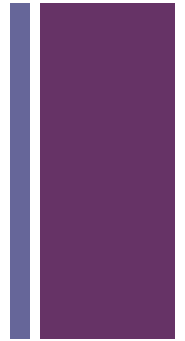


+ Bedtime Routine

- Same every night
- 3-4 activities (e.g., bath, pjs, stories, song)
 - For babies, start with feeding
 - Nothing too upsetting
 - Include typical requests after lights out (snack, tuck in)
 - Avoid electronics
- Short and sweet
 - Heading towards the bedroom
- Minimize attention after lights out

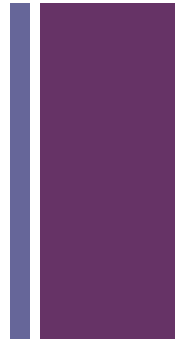
+ Also...

- Visual supports
 - Social stories
 - Visual schedule
 - Motivation (If/ Then)
- Transitions
 - Multiple warnings
 - Visual timers
- Sensory strategies
 - Assess sensory profile with OT
 - Create specific calming environment and routine



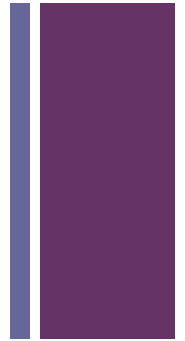
+ Positive Sleep Associations

- Night wakings are normal
- It's getting back to sleep independently that can be difficult
- What is needed to fall asleep is what is needed to return to sleep during normal night wakings
- Switching associations
 - Whatever is present at bedtime should be there all night
 - Increase independence where possible
- All at once or slowly = same results



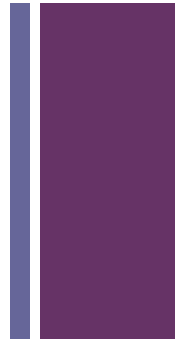
+ Bedtime Problems

- Bedtime stalling
 - Frequent requests
 - Tantrums
 - “I’m scared”; “I hate my bed”
 - Refusing to sleep in own room
- Incorporate requests into routine
- Bedtime pass
- Assess/ address anxiety



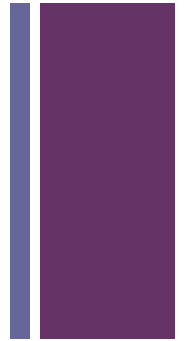
+ Bedtime Problems

- Bedtime stalling
 - Incorporate requests into routine
 - Bedtime pass
- Takes longer than 30 minutes to fall asleep
 - Bedtime closer to when currently falling asleep
 - Reminder: Calm body, quiet voice
 - Tailored calming routine with sensory strategies
 - Include recommendations from OT
- Assess/ address anxiety



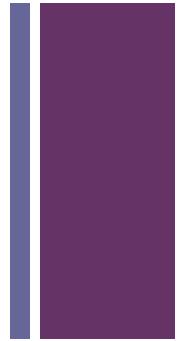
+ Bedtime Pass

- Make actual passes together!
- Start close to where you are (3-4)
- Passes traded for reasonable request
- Once passes are used, no more requests
- When ready to move forward
 - Decrease number of passes
 - Trade in the am for something
 - Preferred breakfast
 - Motivating activity

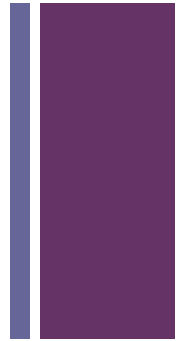


+ Night Wakings

- Make sure falling asleep independently
- Consider changing goals
 - Increase independence/ maintain safety
- Wake strategies
 - Good morning light
 - Clear expectations (visual)
 - Basket of activities
 - Avoid electronics/ other reinforcers



+ Key Points



- Consistency
 - A consistent small step is better than trying a big step
- Minimize talking
- Start where you are
 - Always begin where success is most likely
 - Many roads end at the same place
- Extinction burst

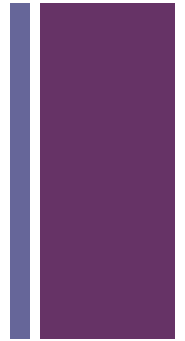
+ Other Interventions

- Evidence does not support:
 - Handouts
 - Weighted blanket
 - Aromatherapy

- Evidence does support:
 - Behavioral interventions
 - Melatonin
 - Medications

- Some support for:
 - Sleep restriction
 - Meditative movement (e.g. yoga, tai chi)
 - Sensory strategies

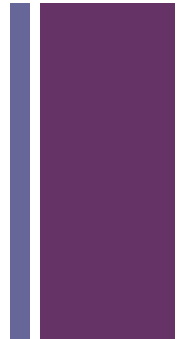
Adkins et al. 2012, Escalona et al. 2001; Frasier; Griggs-Damberger & Ralls 2013; Malow et al. 2012, Piravej et al. 2009; Souders et al. 2017; Wang et al. 2016



+ Sleep Medications Used in WS

- Recruited from WSA Conference
 - N=513
 - 25% have taken meds
- Age first used (6 months-45 years)
- Melatonin
 - 91% helpful or somewhat helpful
 - Fewest side effects 6%
 - Theoretical impact on puberty
- Benadryl 29%
- Quetiapine 8%

Martens et al. (2017). *Journal of Developmental and Behavioral Pediatrics*.

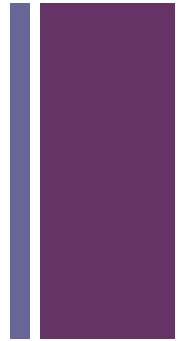


+ Melatonin

- 16 treatment studies support short term effectiveness in NDC
 - SOL
 - WASO
 - TST
- Most studies 3-5 mg (range 0.5-15mg)
 - Titrate weekly
- 30 minutes before bedtime (range 30-45 minutes)
- Theoretical impact on puberty, seizures (both directions)

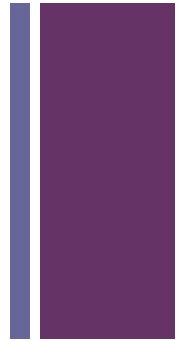
+ Melatonin

- Long term study
 - N=44
 - Combined NDC
 - Q. 3 month follow up for 3.8 years
 - Parent report
 - No adverse events or tolerance



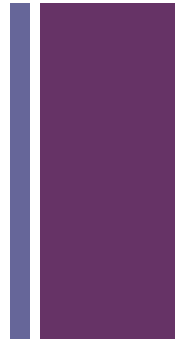
+ When to See a Sleep Specialist

- Snoring, gasping, difficulty breathing during sleep
- Complaints of funny feelings in the legs
- Need help making a change
- At your limits of sleep deprivation



+ Summary

- Sleep problems common in children with WS across development
 - Especially difficulty falling asleep and night wakings
 - Tend to persist but may improve
- A consistent bedtime and bedtime routine are key
- Whatever is needed to fall asleep is needed to return to sleep during normal night wakings
- Multiple approaches can help to improve sleep
- Help is out there if you need it



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Questions?

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