

# CMEO BriefCase

## *Recognition of Narcolepsy in Your Patients*

*This program is supported by an independent  
medical education grant from Jazz Pharmaceuticals*

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# Learning Objective

Screen for narcolepsy based on patient presentation or patient or family/caregiver description of function and changes in quality of life (QoL).

# Patient Case: Raina

- 15-year-old Black female presents with her parents to PCP with complaints of being “tired all the time” and “not being able to stay awake,” starting when patient was age 8
- Her parents report she struggled with school because she would sleep during class but attributed it to “being a light sleeper at night” and “waking up frequently”; they realized something was wrong when the patient was found asleep by the bus driver after dropping the bus off at the garage
- Patient reports she has struggled socially in school for some time and reports depression symptoms as well as poor attentiveness; she wants to be more involved in sports but feels she gets too tired during the week to participate
- PHQ-9 was administered (PHQ-9 = 13) and she was diagnosed with depression and prescribed escitalopram 10 mg daily; patient was counseled on sleep hygiene and recommended therapy and was advised to exercise three times per week

PCP = primary care practitioner; PHQ = Patient Health Questionnaire

# Audience Response



Which part of Raina's presentation is seen in **ALL** patients with narcolepsy?

- A. Poor attentiveness
- B. Sleep disruption
- C. Poor school/work performance
- D. Excessive daytime sleepiness
- E. I don't know

# Audience Response



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# Narcolepsy: Patient Presentation

## Narcolepsy prevalence:

## Also seen with:

<b>C</b>	ataplexy	65%-75%	→	Hypotension, seizure
<b>H</b>	allucinations	33%-80%	→	Schizophrenia, nightmares, depression
<b>E</b>	xcessive daytime sleepiness (EDS)	100%	→	Periodic limb movements, OSA, IH, depression, insomnia, sleep deprivation
<b>S</b>	leep paralysis	25%-50%	→	Depression, nightmares, schizophrenia, psychiatric disorders
<b>S</b>	leep disruption	30%-95%	→	OSA, insomnia, sleep deprivation, periodic limb movements

IH = idiopathic hypersomnia; OSA = obstructive sleep apnea

Cheung J, Ruoff CM. Central nervous system hypersomnias. In: *Sleep and Neurologic Disease*. 2017. Roth T, et al. *J Clin Sleep Med*. 2013;9(9):955-965. Thorpy MJ, Dauvilliers Y. *Sleep Med*. 2015;16(1):9-18. Moturi S. *Psychiatry (Edgmont)*. 2009;6(6):38-44.



# Quality of Life in Children with Narcolepsy

## Pediatric QoL Study

- 18 narcolepsy, 15 idiopathic hypersomnia, 33 control
- Mean age: 13
- Patients with narcolepsy have worse QoL compared to healthy peers

Outcome	Narcolepsy/ IH	Control	P Value
Sleepiness (ESS)	<b>12.3</b>	<b>6.9</b>	< .001
BMI	<b>26.2</b>	<b>22.2</b>	.03
Injury history	<b>39%</b>	<b>15%</b>	.03
Physical activities	<b>30%</b>	<b>56%</b>	.04
Extracurricular activities	<b>25%</b>	<b>68%</b>	.001

Outcome	Narcolepsy/ IH	Control	P Value
Academic grades < C	<b>25%</b>	<b>12.5%</b>	.04
Physical functioning	<b>74.5</b>	<b>85.5</b>	.001
Social functioning	<b>75.8</b>	<b>87.8</b>	.01
School functioning	<b>65.9</b>	<b>78.1</b>	.007
Overall QoL	<b>73.4</b>	<b>83</b>	.001

BMI = body mass index; ESS = Epworth Sleepiness Scale; QoL = quality of life  
Avis KT, et al. *J Clin Sleep Med*. 2015;11(11):1281-1288.

# Parents Can Be Good Reporters of Their Children's Symptoms

Parents reported lower QoL in their children's lives

Outcome	Narcolepsy/IH Parent Report	Control Parent Report	P Value
Physical functioning	<b>62.5</b>	80.6	< .001
Emotional functioning	<b>58.9</b>	70.8	.007
Social functioning	<b>64.8</b>	81.2	.002
School functioning	<b>49.8</b>	77.7	< .001
Overall QoL	<b>59.5</b>	78	< .001

**Parent/caregiver report can be key in recognition of narcolepsy in children!**

# Impact of Narcolepsy on Children

## Child patients

- Lower HRQoL vs. control ( $p = .001$ )
- Lower vitality vs. control ( $p = .002$ )
- Lower sense of well-being vs. control ( $p = .002$ )
- Poorer self image vs. control ( $p = .03$ )



## Adolescent patients

- Lower QoL index vs. control ( $p = 0.008$ )
- Lower physical well-being vs. control ( $p < .001$ )
- Fewer friends vs. control ( $p = .001$ )
- Less leisure activities vs. control ( $p = .006$ )



HRQoL = health-related QoL

Inocente CO, et al. *CNS Neurosci Ther.* 2014;20(8):763-771.

# Patient Case: Raina

- Patient returns to clinic at age 18 with continued complaints of “being tired all of the time,” along with concerns regarding certain life events
- Recently had an auto accident when she nodded off at the wheel; she also was let go from her part-time job because she often would not wake up for work
- Patient was interviewed for sleep symptoms and reported pertinent issues:
  - Frequently waking up at night
  - Experiencing hallucinations when dozing back to sleep
  - Having weakness when laughing hard
- ESS and PHQ-9 was administered: ESS = 17/24, PHQ-9 = 7

# Audience Response



**Which domains of Raina's health-related quality of life appear to be most impacted by a possible narcolepsy diagnosis at this time?**

- A. Mental health and bodily pain
- B. Mental health and social functioning
- C. Physical role functioning and vitality
- D. Social functioning and feelings of control
- E. I don't know

# Audience Response

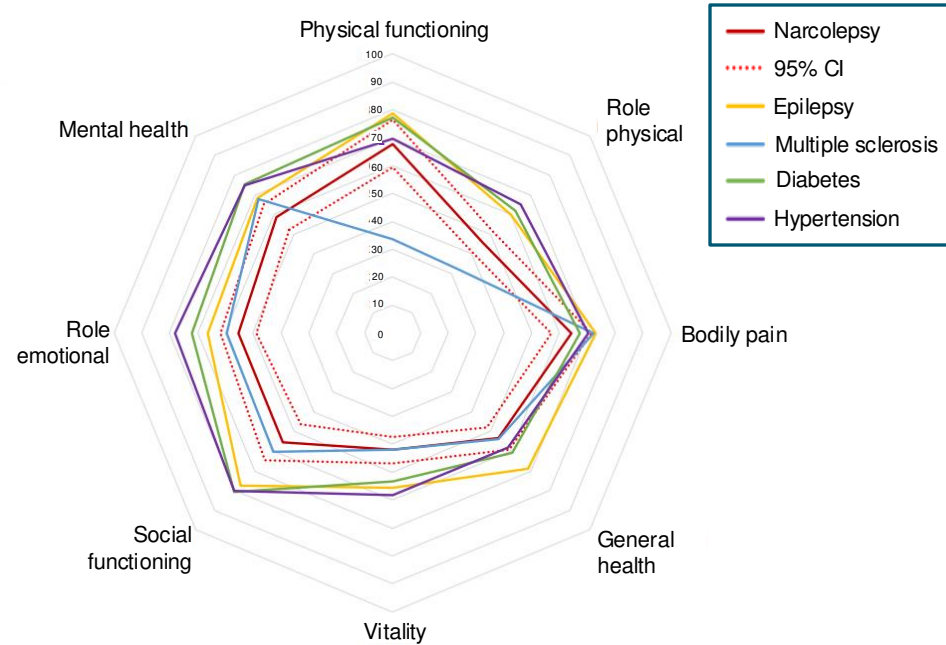
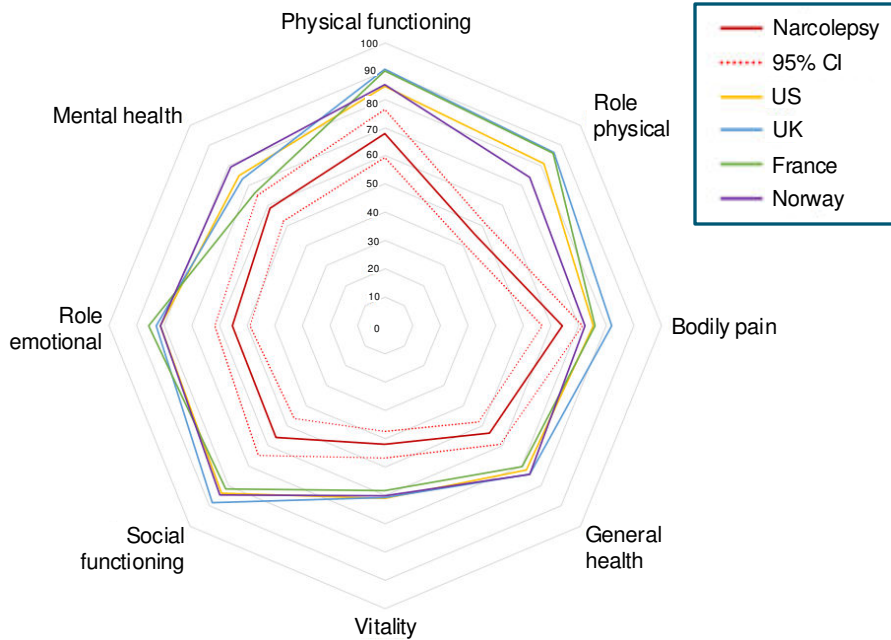


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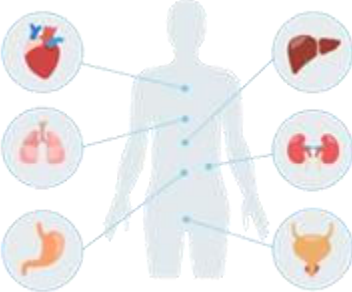
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- D. Social functioning and feelings of control
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# Health-Related QoL in Narcolepsy: Meta-Analysis

- Short Form 36 meta-analysis (n = 4,600)
  - Mean age 40.8 (95% CI: 37.12-44.46), 54.31% female
  - Lower HRQoL in numerous domains compared to general population; multiple chronic diseases including multiple sclerosis and epilepsy



# Impacts on Quality of Life: Adults

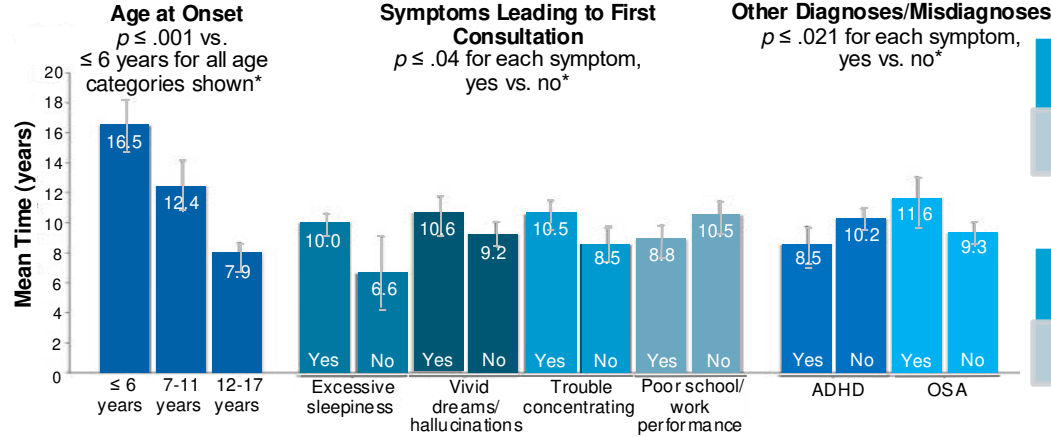
- Patients are at **2x** to **4x** greater risk of\*:
  - HRQoL impairment
  - Prevalence of long-term disability 
  - Increased absenteeism
  - Decreased presenteeism
  - Hospitalization, emergency department visits 
  - Increased traditional health care professional visits
  - Comorbidities 

\* $p < .05$   
Flores NM, et al. *J Clin Sleep Med*. 2016;12(3):401-407.

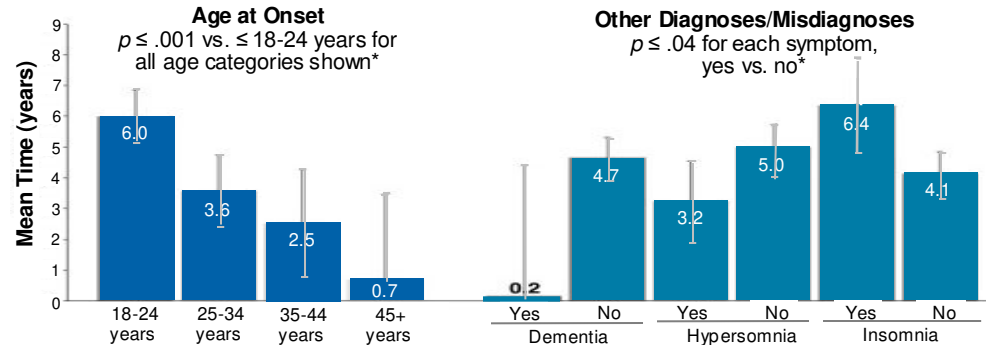


# Patient Journey: Nexus Narcolepsy Registry

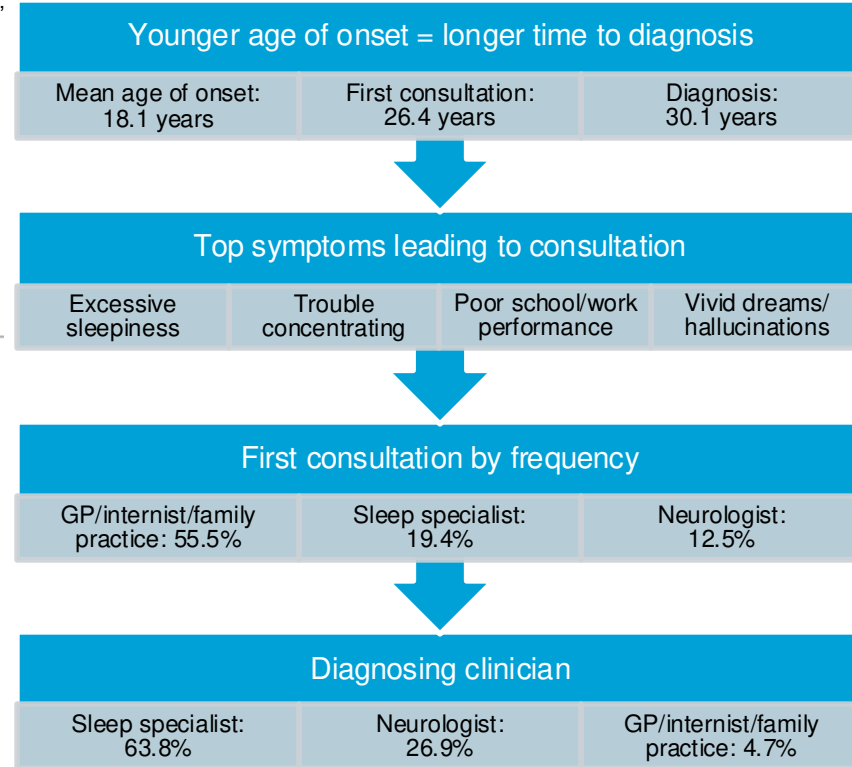
## Pediatric Symptom Onset



## Adult Symptom Onset



## Overall Findings



# Misdiagnosis of Narcolepsy and Common Comorbidities

% of Participants Reporting	Participants Reporting:					
	Misdiagnosed Conditions			Correctly Diagnosed Comorbid Conditions		
	Pediatric Symptom Onset (n = 541)	Adult Symptom Onset (n = 374)	P Value*	Pediatric Symptom Onset (n = 541)	Adult Symptom Onset (n = 374)	P Value*
Depression	34.2	29.1	.108	32.0	35.3	.296
<b>ADHD</b>	19.2	12.3	<b>.005</b>	8.9	11.0	.294
Anxiety disorder	17.6	15.2	.354	27.5	25.7	.529
Insomnia	15.7	13.4	.326	7.0	5.9	.493
<b>Bipolar disorder</b>	14.2	9.1	<b>.019</b>	3.0	3.7	.512
Hypersomnia	14.0	14.4	.868	8.7	11.0	.251
<b>OSA</b>	10.9	14.2	.138	9.6	14.2	<b>.033</b>
<b>Epilepsy</b>	7.8	3.7	<b>.013</b>	1.3	0.5	.253
<b>Schizophrenia</b>	5.4	2.1	<b>.015</b>	0.2	0.0	.405
Dementia	2.2	2.4	.852	0.0	0.0	—

**59.3% reported receiving at least one misdiagnosis before being diagnosed with narcolepsy**

\*Pediatric onset vs. adult onset

ADHD = attention-deficit/hyperactivity disorder

Ohayon MM, et al. *Sleep Med.* 2021;84:405-414.

# Screening for Narcolepsy: Clinical Interview



## Listen carefully, and evaluate for:

### Cataplexy

- Do your knees ever buckle or give out? Do you experience muscle weakness when you are in certain situations? Do you notice your child falling down or head dropping?

### Hallucinations

- Do you see, feel, or hear things that are not there when you are drifting into or out of sleep?

### EDS

- How often do you feel tired or drowsy? How often do you need to nap during the day? Are there ever times you are not feeling tired?

### Sleep paralysis

- Do you ever wake up and cannot move? Can you describe that sensation?

### Sleep disruption

- How often do you wake up during the night? Is it difficult for you to wake up in the morning?

**Patients often have trouble recognizing or describing symptoms;  
be sure to use patient-friendly language!**

# Screening Tools: Epworth Sleepiness Scale

## Epworth Sleepiness Scale

Name: \_\_\_\_\_ Today's date: \_\_\_\_\_

Your age (Yrs): \_\_\_\_\_ Your sex (Male = M, Female = F): \_\_\_\_\_

How likely are you to doze off or fall asleep in the following situations, in contrast to feeling just tired?

This refers to your usual way of life in recent times.

Even if you haven't done some of these things recently try to work out how they would have affected you.

Use the following scale to choose the **most appropriate number** for each situation:

- 0 = would **never** doze
- 1 = **slight chance** of dozing
- 2 = **moderate chance** of dozing
- 3 = **high chance** of dozing

*It is important that you answer each question as best you can.*

Situation	Chance of Dozing (0-3)
Sitting and reading _____	—
Watching TV _____	—
Sitting, inactive in a public place (e.g. a theatre or a meeting) _____	—
As a passenger in a car for an hour without a break _____	—
Lying down to rest in the afternoon when circumstances permit _____	—
Sitting and talking to someone _____	—
Sitting quietly after a lunch without alcohol _____	—
In a car, while stopped for a few minutes in the traffic _____	—

You must quantify sleepiness in all patients with sleep complaints!

0-5: Lower normal daytime sleepiness

6-10: Higher normal daytime sleepiness

11-12: Mild excessive daytime sleepiness

13-15: Moderate excessive daytime sleepiness

16-24: Severe excessive daytime sleepiness

# Patient Case: Raina

- Patient returns to clinic at age 18 with continued complaints of “being tired all of the time,” along with concerns regarding certain life events

What should have been done in the first patient interview with Raina?

How can the ESS help us rule out other diagnoses?

What can help us differentiate comorbidities (such as depression) from narcolepsy?

When do we refer our patient?

# SMART Goals

Specific, Measurable, Attainable, Relevant, Timely

- Screen all patients with complaints of sleep dysfunction and EDS for symptoms associated with narcolepsy.
- Recognize narcolepsy and comorbidities associated with narcolepsy when considering patient presentation and patient/caregiver description of QoL during office visits.
- Conduct effective clinical interviews that quantify EDS, determine narcolepsy-specific sleep symptoms, and use patient-friendly language for every patient presenting with tiredness.

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2

Differential Diagnosis for  
Narcolepsy

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Choosing and Optimizing  
Therapy in Narcolepsy

[www.cmeoutfitters.com/sleep-disorders-hub/](http://www.cmeoutfitters.com/sleep-disorders-hub/)

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