Psychosocial Aspects of Narcolepsy

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Objectives

• Become familiar with the history of the psychology-narcolepsy relationship
• Be able to identify common psychosocial comorbidities with narcolepsy
• Understand disruptions of quality of life in narcolepsy
• Learn solutions and future steps to address these difficulties
Fortuyn, 2010
The Beginnings of Psychology and Narcolepsy

• First clinical reports of Cataplexy (Germany, 1877) and Narcolepsy (France, 1880)
  – Neurology and psychiatry one discipline
  – Hysteria is the new diagnosis

• Case study of famous pedophile (Westphal, 1877)
  – Excessive daytime sleepiness documented
  – Maybe Narcolepsy associated with hiding/shame of vices
    > Repressed homosexuality
Westphal
First description of 'eigenartige Zufälle'.

1877

Loewenfeld
Motor inhibition required for diagnosis.

1902

Redlich
First case-series. Rule out hysteria. Cataplexy required.

1915

Thiele & Bernhardt
Organic syndrome, with localized 'brain complex'.

1933

1881

Gélineau
Describes secondary narcolepsy, due to e.g. hysteria.

1910

Lhermitte
Narcolepsy can be expression of hysteria.

1920

Myers
Treatment with hypnosis. Psychoanalytical approach.

1928

Wilson
Narcolepsy is a syndrome; psychogenic cause is possible.

1942

Simmel
Sleep symbolizes passive experience of death. Applies dream model of Freud.
1944

1946

Simmel
Sleep symbolizes passive experience of death. Applies dream model of Freud.

1953

Bernadetti
Focal brain and endocrine syndrome.

1960

Vogel
Discovery of sleep-onset REM. Alleged ‘proof’ of psychoanalytical model.

1973

Knecht
First description of canine narcolepsy.

1975

Bourguignon
Narcolepsy as a specific response to psychological conflicts.

2000

Nishino et al.
Hypocretin deficiency in human narcolepsy.

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Narcolepsy and Psychology Since 2000

- No link with Schizophrenia or Depression (Vourdas, 2002)
  - 45 adults w/Narcolepsy, 50 controls. UK
  - Schizophrenia's presence overstated in early research
- 15-37% with mod-severe Depression (Dauvilliers, 2009)
  - 600+ adults. France
- 53% with Anxiety or Panic (Fortuyn, 2011)
  - 60 adults with NC. Netherlands
- High prevalence (25%) of mod-severe Depression (Vignatelli, 2011)
  - 54 adults w/Narcolepsy. Italy
    - Longer duration of Narcolepsy, less depression
- 25% with clinical depressive symptoms (Inocente, 2014)
  - 88 children w/Narcolepsy. France
    - Especially girls over 10y/o
    - Longer period between symptoms and diagnosis = higher Depression
- Anxiety and Depression higher in Narcolepsy than general population (Alasim, 2019)
  - Saudi Arabia, 74 adults w/Narcolepsy
Mood and Narcolepsy

• Ohayon, 2013
  – 320 adults, phone interview
  – 17% -MDD
  – 21% -Social Anxiety
  – 5% -Generalized Anxiety
  – 11% -PTSD
  – 12.5% -Panic disorder
• Higher than general pop
• More prevalent in women

• BOND Study (Black, 2017)
  – 9000+ adults with Narcolepsy
  – 46500+ controls
  • 13% Anxiety
  • 24% Mood Disorder
  • Significantly higher than controls
  • No difference b/w NC and N-C
Mood and Narcolepsy

- 50%+ of patients w/Narcolepsy and Depression had Depression first (Lee, 2017)
  - Shared pathophysiology related to hypocretin deficiency?
- Higher incidence of Anxiety and Depression in Females age 12-17 with Narcolepsy (Chen, 2020)
  - Taiwan, Cohort study, 478 youth/adults
  - Those with Anxiety more likely to develop Narcolepsy
    > Gap between symptoms and diagnosis
Why Mood and Narcolepsy?

• Hypocretin has strong connection with limbic system
  – Deficiency of hypocretin
    > Induces imbalance in neurotransmitters
      – Effects vigilance and mood regulation
    > Stimulates Hypothalamus-Pituitary-Adrenal axis
      – Activates body’s stress response
• Similar findings to patients with amygdala lesions
  – Impaired startle response to unpleasant stimuli (Khatami, 2007)
  – Pronounced amygdala regulation to humorous pictures (Schwartz, 2008)
    > Amygdala involved in pathophysiology of narcolepsy
    > May have abnormal emotional processing
Why Mood and Narcolepsy?

- Premorbid Depression/Anxiety (Black, 2017; Ruoff, 2013)
  - May impact misdiagnosis/diagnostic delays
  - May be result of misdiagnosis/diagnostic delays
- Response to symptoms
  - Perceived loss of control
    > Cataplectic events
    > Irresistible attacks of sleep
  - Fragmented perception of reality
  - Hallucinations
  - Invalidation from providers, society
  - Impaired school, relationships, occupation
Why Mood and Narcolepsy?

- Delayed diagnosis
  - Long delays and misdiagnosis common (Kryger, 2002)
    - Frequent comorbidities with psychiatric conditions (Morse, 2018)
      - Debilitating and not adequately treated
    - Delays M=15 yrs, 60+ yrs in some cases! (Thorpy, 2014)
      - Trending down
      - Lack of recognizing symptoms, misdiagnosis
- Delays and misdiagnosis
  - HUGE impact upon:
    - Mood
    - Social functioning
    - Health care utilization
    - Employment
    - QOL
Psychosocial QOL

- Lower HRQOL than population (Daniels, 2001)
  - 300 adults in UK
  - Impaired education, home-life, work, and social-life due to symptoms
- Lower HRQOL than population (Ervik, 2006)
  - 100 Norwegian adolescents and adults
  - Impaired social functioning, general health perception
- Lower HRQOL than population (Vignatelli, 2011; Inocente, 2014)
  - 54 adults in Italy
  - 5 year follow up, QOL stable and impaired
    > Worse if depressed
Psychosocial QOL

- More financial and social challenges than population (Jennum, 2009)
  - 459 Danish adults
  - Higher unemployment, lower income, higher health system contacts, use of public supported medication
  - Socioeconomic consequences for individual and society

- Lower QOL than population (Ozaki, 2011)
  - 185 adults in Japan
  - Impaired autonomy in controlling job schedule, increased divorce, break up due to symptoms, forced to relocate or dismissed from job, negative perception of support from others

- Early diagnosis can improve occupational prognosis (Ingravallo, 2012)
  - 100 adults with NC in Italy
  - Dx < 30, improved occupational prognosis, general health perception
    - Less unemployment, work changes, work absences
    - Less costs (missing work, travel, out of pocket pay)
Living with Narcolepsy is a Daily Struggle.\textsuperscript{1,3,4} 
It Impacts Family, Relationships, Work and School.\textsuperscript{1,3}

Many in the survey found it difficult to live a normal life.\textsuperscript{1}

86% reported that narcolepsy is a life-changing disease.\textsuperscript{1}

4 in 5 said that living with narcolepsy is a daily struggle.\textsuperscript{1}

76% said that narcolepsy has affected important moments in their lives.\textsuperscript{1}

37% reported having failed a class at school or withdrew from a class.\textsuperscript{1}

Nearly All People Surveyed Agree 
There is More To Be Done.\textsuperscript{1}

Managing narcolepsy is a lifelong journey. There is a clear need for improved communication and more education.\textsuperscript{1,2}

Even with treatment, people living with narcolepsy continue to feel the impact of their symptoms.\textsuperscript{1,2}

More than half surveyed (54%) said narcolepsy controls their lives, instead of them controlling narcolepsy.\textsuperscript{1}

40% said they do not usually discuss with their healthcare professionals how narcolepsy affects their daily lives.\textsuperscript{1}

94% felt that new treatment options are needed.\textsuperscript{1}

88% felt that people in general do not understand how disruptive narcolepsy is an day-to-day life.\textsuperscript{1}

94% felt that more education is needed.\textsuperscript{1}

REFERENCES:

To learn more, visit www.KnowNarcolepsy.com

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Culbertson, 2005. (27 adults in Australia)

- Main findings:
  > Health professionals need to understand narcolepsy’s disruption to normal social functioning
  > Importance of looking at disorder’s impact on social interactions
  > Examine experience in youth
    - Delayed diagnosis, mood, and different social experiences during adolescence
Qualitative QOL- Interpersonal Relationships

- “[she] never accepted that it was there, and she never realized what the issues are, she was never aware of the sort of struggle I was having…”
- “…31 years and he still hasn’t got it straight, he will walk in and give me a surprise and walk off and I’m standing there grabbing hold of something and he has walked off. He never even thinks to look.”
- “It probably forced me to talk to them more. They’ve understood it. We’ve talked about a lot of things that we may not have if I hadn’t have had it. I’ve been forced to talk to them.”
- “With friends [falling asleep] doesn’t bother me any more. We might even laugh about it. In a situation with strangers around I am so on edge that I will fall asleep. I get really nervous that I will go to sleep, really nervous… I am so afraid to fall asleep.”
- “It’s very, very difficult. People invite me out for meals occasionally… It’s just awful… They always get a nice warm room ready, they always get you a big meal… and it’s just so conducive to go to sleep. Its so embarrassing… and that dreadful feeling of trying to stay awake, ‘Oh I mustn’t go to sleep, I must try and stay awake, I must listen to what they’re saying, I must talk to them and be sociable; and all the time I’m dropping off. It’s just awful.”
Qualitative QOL- Trustworthiness/Perception

• “you don’t tell people any more than they need to know… sometimes it causes problems… I do hide a great deal.”

• “…It’s a bit useless trying to explain because they don’t hear you or want to hear you, they think you’re making a fuss about nothing actually.”

• “You get half way through a sentence and you don’t know how you’re going to end it. People don’t want you to feel bad and they tend to go, ‘Oh I’m like that too’. But they’re not like it to the extent that we are.”
Qualitative QOL- Work

• “…certainly I would never have been employed if I told them that I had narcolepsy. I had made up my mind that I was going to be very open about this. I realized pretty rapidly that wasn’t going to be very sensible.”

• “I think it would be harder for me to maintain a sense of well-being if I wasn’t working… working, I feel, is my one contact with normal.”

• I want to have a life, I want to have respect and do something for myself. I’m sick, OK I understand that, but this is not a disease I can’t manage. I could have a very well oriented career with this disease and be able to be successful…”
Qualitative QOL- Invisible/Having Support

• “...(it’s) frustrating that you don’t see it. I know that people don’t believe me. I would expect them not to believe; it’s too bizarre.”

• “I know there are other illnesses that are so much more grave and life threatening than mine, but in many of these, the public knowledge is so much better; understanding and acceptance are there…”

• “…I was the first narcoleptic that she’d spoken to and she was the first one I’d spoken to, I spent 2/3 of a day with her and I didn’t want to leave.”

• “Very wonderful to know I am normal for what I am and so is the other person.”
Qualitative QOL- FDA

- 2013 FDA public meeting
  1. Biggest daily struggles
     > Excessive daytime sleepiness
     > Chronic sleep deprivation
     > Brain fog
  2. Factors impacting psychological comorbidities
     > Cataplexy
     > Hallucinations
     > Sleep paralysis
     > Uncontrollable symptoms/unpredictable loss of control
  3. QOL difficulties
     > Social, emotional, and financial tolls for individuals and their families
     > Difficulty maintaining jobs, attending school, caring for home, engaging in social activities, maintaining relationships
4. Frustration from inappropriately being labeled as lazy, careless, or incapable
   > By colleagues, health care professionals and public
5. Medication has drastically improved symptoms for many
   > Some given up due to intolerable side effects or tolerance
6. Non-pharmacological treatment can help, but difficult to sustain
   > Scheduled naps, diet modification, physical activity
7. Challenge of proper diagnosis and treatment
8. Lack of supports in school, work, and community

"Sleepiness is NOT laziness."
Why Narcolepsy and Poor QOL

- Misdiagnosis and inappropriate treatment
- Stigma, incorrectly labeled as lazy
- Poor psychosocial adjustment
  - Low self-esteem, Mood
- Lack of supports
What did all of these studies recommend?

• Increased knowledge of Hypersomnia/Narcolepsy
  – In school, work, and healthcare
• Early diagnosis and treatment
  – Can reduce disease burden and socioeconomic impact
  – Better management of symptoms of Narcolepsy and Depression in kids
• Clinicians should assesses for symptoms of Narcolepsy
  – High comorbidities risks leaving patients under/misdiagnosed
  – Females with Anxiety
  – Sleepiness endorsed
    > Part of differential diagnosis (Mood vs Narcolepsy vs both)
      – Sleepiness or poor response to treatment
    > More detailed questions
• Screen for comorbidities of Narcolepsy
Find more effective treatments for comorbid psychiatric symptoms
- Multidisciplinary approach offers therapeutic benefits

Intervention process should be in context of patient and not prevent regular follow up
- Patient-specific, measurable interventions
  - Currently lack interventions w/multicenter studies or validation process

CBT should have elements common to any psych treatment
- Relaxation, reframing, behavioral modification
- Treatment overlap for Hypersomnia/Depression

CBT-N should start from clinical formulation based on symptoms and effects on social life
Clinical decision tree in CBT-N

Narcoleptic Symptoms and Maintenance Factors

Predisposing and Precipitating Factors in Cataplexy
- Stimulus Control
- Systematic Desensitization
- Progressive Relaxation

Associated factors and accompanying symptoms (sleep paralysis and hypnagogic hallucinations)
- Lucid Dreaming with self-awareness, Imagery Rehearsal Therapy

Excessive Daytime Sleepiness (EDS)
- Sleep Satiation

Cognitive precipitating factors (dysfunctional beliefs & dysfunctional coping strategies)
- Cognitive Behavioral Therapy
- Scheduled Naps

Fig. 2 – CBT flow chart for treating narcolepsy-cataplexy syndrome.

Agudelo, 2014
Support

- Patients with Narcolepsy may need social support and counseling
- Inclusion of family, caregivers, and support groups (for clinicians and individuals w/Narcolepsy)
  - Of utmost importance
  - Assists in obtaining information, personal issues, social difficulties, treatment efficiency, adherence
Summary

• Narcolepsy can have a significant impact on mood and QOL
• CBT
  – Treating comorbidities
    > Coping with emotion
    > Reducing Anxiety triggers
    > Improving self-care
• Behavioral modification
  – Scheduling naps
• Adherence to drug therapy
• Educating others/finding support