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The FNDamentals of Functional Neurological Disorder

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<https://youtu.be/EZ3bEUGJPmE?feature=shared>

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Interprofessional Continuing Education

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Daniel Freedman, DO

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Objectives

- Discuss the modern conceptualization of functional disorders
- Review common presenting symptoms
- Discuss treatment options

Why does this matter?

- It's common – teen psychogenic nonepileptic seizures (PNES) – 59.5 per 100,000
 - Villagran et al., 2021, Epilepsia
- Increased standardized mortality ratio 2.5 times the general population
 - Comparable to refractory epilepsy
 - Nightscales et al., 2020, Neurology
- Median cost to diagnose PNES in Australian adults: \$19,000
 - Seneviratne et al., 2019, Epilepsia
- Annual cost of ED visits in the US for pediatric & adult FND: \$163 million
 - Higher than the annual cost of ED visits for refractory epilepsy
 - Stephen et al., 2020, JAMA Neurology

DSM 5 (2013)

- One or more symptoms of altered voluntary motor or sensory function
- Clinical findings that demonstrate incompatibility between the symptom and recognized neurologic or general medical conditions (e.g., Hoover's sign of functional limb weakness or a positive entrainment test for functional tremor)
- The symptom or deficit causes significant distress and psychosocial impairment or warrants medical evaluation
- Subtypes: seizures, weakness/paralysis, abnormal movement, anesthesia, special sensory, swallowing, speech, mixed

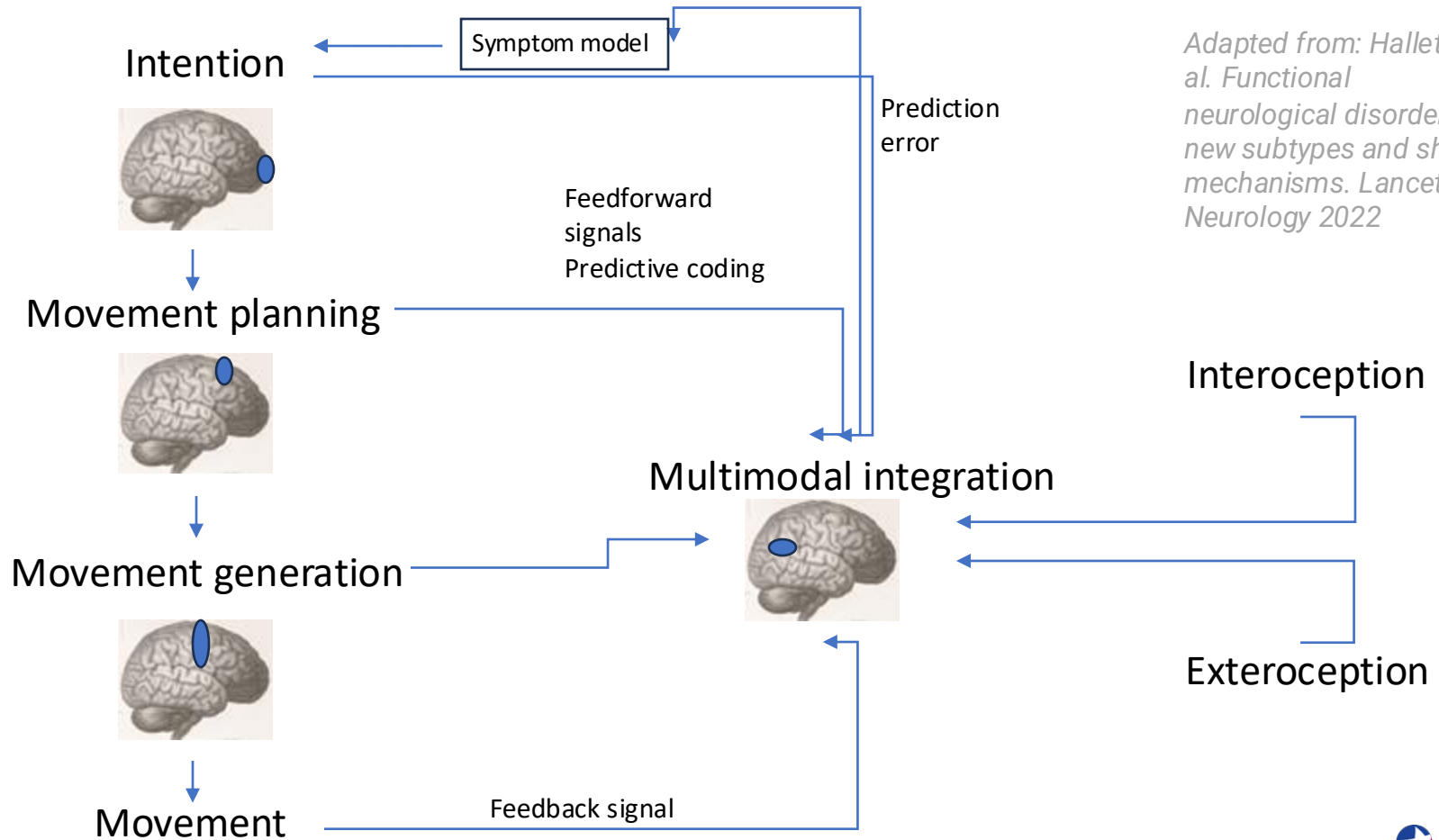
DSM-5-TR

- 2022 update
- From “Conversion Disorder (FND)”
- Now “FNSD (Conversion Disorder)”

What is it?

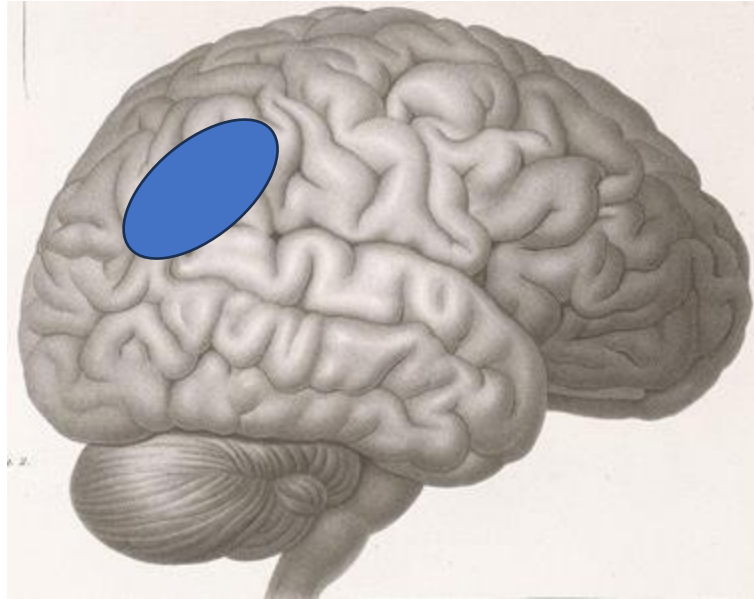
- Refers to symptoms that reflect inconsistent but altered functioning of the nervous system that are incongruent with other neurological disorders, often with influence from psychological risk factors, and occur without an identifiable pathology
- Incongruence
- Inconsistency:
 - Variability
 - Disproportionate disability
 - Suppression or change with distraction
 - Magnification with attention

Adapted from: Hallett et al. Functional neurological disorder: new subtypes and shared mechanisms. Lancet Neurology 2022



Agency

→ Illusory movement
← Motor intention



*Adapted from:
Desmurget **et al.**
Movement intention
after parietal cortex
stimulation in
humans. Science
2009*

Predictive Coding

- Brain as a prediction machine
- Evolved to maintain homeostasis
- Dependent on interoception



Signs that favor PNEE	Evidence from primary studies	Specificity (%)
Long duration	Good	-
Fluctuating course	Good	96
Asynchronous movements	Good (excluding FLE)	93-100
Pelvic thrusting	Good (excluding FLE)	92-100
Side-to-side head or body movements	Good (convulsive events only)	92-100
Closed eyes	Good	74-100
Ictal crying	Good	100
Memory recall	Good	90-96
Signs that favor seizure	Evidence	Specificity
Occurrence from sleep	Good	100
Postictal confusion	Good	84-88
Stertorous breathing	Good	100

*Adapted from:
LaFrance WC Jr, et al.
Minimum requirements
for the diagnosis of
PNES: a staged
approach: a report
from the International
League Against
Epilepsy Nonepileptic
Seizures Task Force.
Epilepsia. 2013*

Positive Diagnostic Signs

Sign	Finding
Hoover	Hip extension weakness that returns to normal with contralateral hip flexion against resistance
Hip abductor sign	Abduction weakness returns to normal with contralateral hip abduction against resistance
Global weakness	Affecting extensors and flexors equally
Dragging gait	Forefoot remains in contact with the ground, typically with hip externally or internally rotated
Inconsistency	E.g., weakness of plantar flexion while laying supine but able to walk on tiptoes

Positive Diagnostic Signs

Sign	Finding
Tremor entrainment test	Copy rhythmical movement with contralateral limb; affected hand entrains to rhythm of unaffected hand or stops completely
Fixed dystonic posture	Characteristically of hand (flexion of fingers, wrist, elbow) or ankle (plantar and dorsiflexion)
Functional hemifacial overactivity	Orbicularis oculus or oris overcontraction, jaw deviation, platysma activation
Distraction during standing	Patients with +Romberg are asked to guess numbers written on their back or carry out complex task (e.g., use a cellphone)

Diagnostic Pitfalls

- Making assumptions based on a patient's psychological history or psychological stress
- Failure to consider an additional medical or neurological cause
 - Occam's razor vs Hickam's dictum
- Not basing diagnosis on the presence of positive diagnostic signs
- Bizarre \neq FND
- Reliance on negative tests

Diagnostic Pitfalls

- Making assumptions based on demographics
- Not keeping FND on the differential
- Lack of awareness of positive FND findings
- Overattributing mildly abnormal results (e.g., nonspecific WM lesions)

Don'ts

Don't:

- Assume new symptoms are related to FND.
- Use stress- or anxiety-focused language as this often deters family from treatment and doesn't fit with our current conceptualization of FND.
- Use language that implies the child is faking their symptoms. It is okay to highlight that the child can learn to control the symptoms; however, they do not currently feel in control.
- Imply that this is a “good” diagnosis. This can make the patient and family feel invalidated in their experience.

How to Explain the Diagnosis

- Use positive clinical signs
- Explain the signs to the patient & family
- Explain hardware-software
- Discuss the neuroscience
- Highlight prognosis: 77-80% recovery
 - Raper et al., 2019, Arch Dis Child
- Use teach back to verify understanding
- Refer to additional resources (e.g., neurosymptoms.org, fndhope.org, fndaction.org.uk)

Critical Aspects of Treatment

- Quick diagnosis and referral to specialty clinic, PT, psych, etc.
- Parental and patient understanding and acceptance of both diagnosis and treatment
- Identify reinforcing factors for patient and/or family
- Treat comorbidities (e.g., epilepsy, POTS, migraine, anxiety, depression)
- Collaboration with other providers
- Follow-up!

Dell Children's PNES Clinic

- 330 referrals
 - Texas, Louisiana, Arkansas, Florida, Hawaii, North Carolina
 - Declined referrals from MN, OH, OK, WA
- 2-month follow-up data for 34 patients
 - 79% reported improvement
 - 26% complete remission
 - 91% did not return to the ED for PNES

Treatment

- Most evidence for CBT
- Three targeted treatment areas
 - Patient control of symptoms
 - Control of reaction from systems
 - Return to previous activities

Psychological Treatment

- Goals of treatment are to
 - Improve events (frequency, duration, intensity)
 - Increase patient's sense of self-agency
 - Decrease fear and inevitability surrounding events
 - Increase patient's functioning

School as Treatment

- Withdrawal from school
 - Reinforces patient's symptoms are scary and harmful
 - Implies that patient is not able to learn strategies to gain control
 - Exacerbates risk factors for PNEE (e.g. anxiety, depression)
 - Decreases quality of life

School as Treatment

- Returning to school
 - Reinforces that patient is safe during events
 - Sends message to patient that we are confident they will improve
 - Can help address comorbidities
 - Increases quality of life

Key Points

- It's common, it's real
- Female predominance ranges from 60-90%
- 23% with persistent symptoms into adulthood
- Some of the lowest QOL measures in neurology
- Neurologists and pediatricians can help
- Treatment involves symptom-based brain retraining therapy, treating comorbidities, addressing social factors

Resources

- Join FNDS
- Patient advocacy: FNDHope
- Patient information: neurosymptoms.org

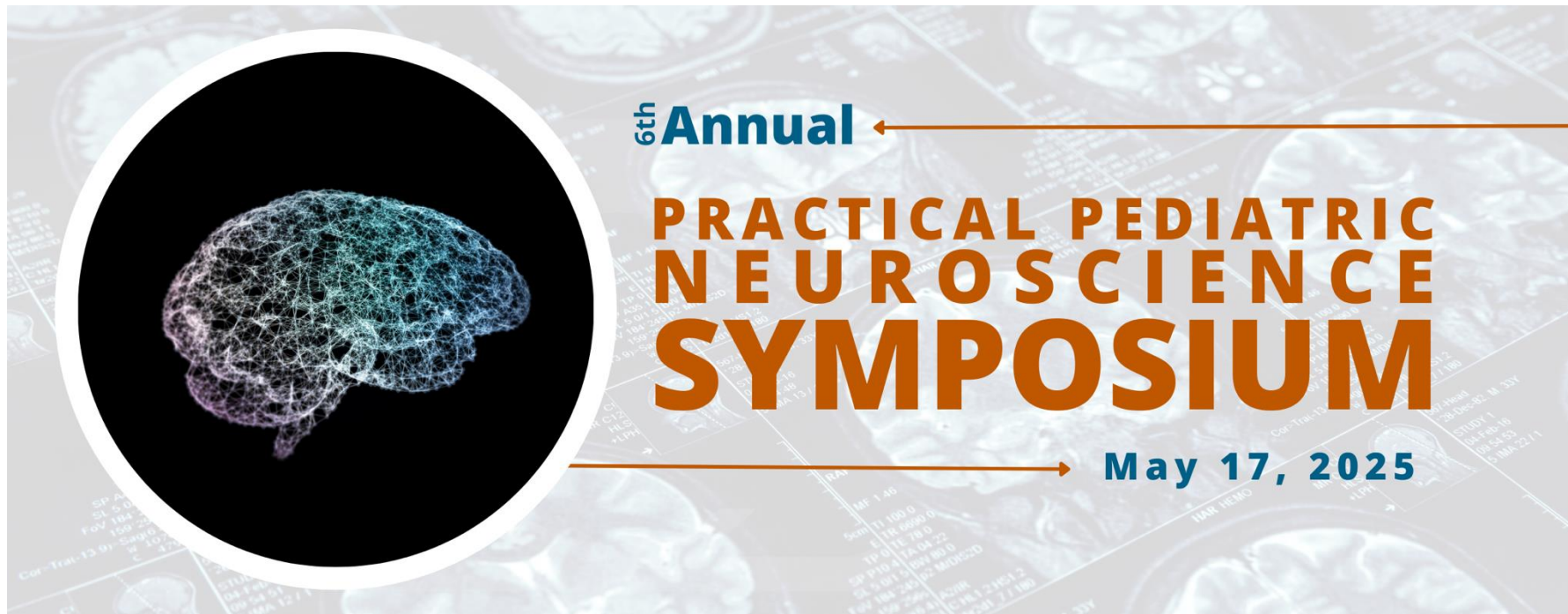




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