Functional Neurologic Disorders

Andrew N. Pfeffer, MD
Associate Professor of Emergency Medicine
Program Director, Emergency Medicine Residency
University of Kansas

Conflicts of Interest/Disclosures

- None :-)

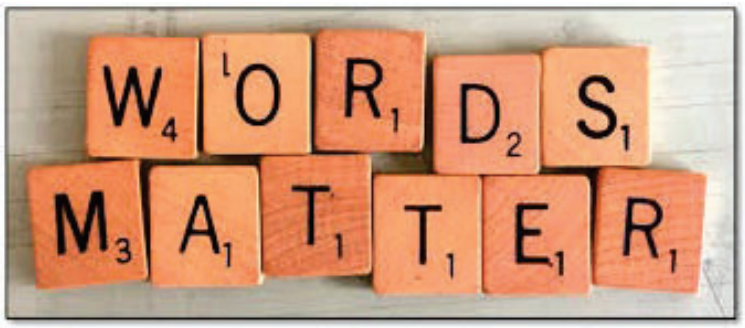
Objectives

- Discuss and explain key concepts related to the work-up and diagnosis of functional neurological disorders (FNDs) in the Emergency Department, as well as Delusional Parasitosis (DP)
- Describe the importance of a holistic approach in the evaluation and management of patients with FNDs and DP
- Develop a patient-centered approach to communication and shared-decision making for patients with FNDs and DP

The Why...

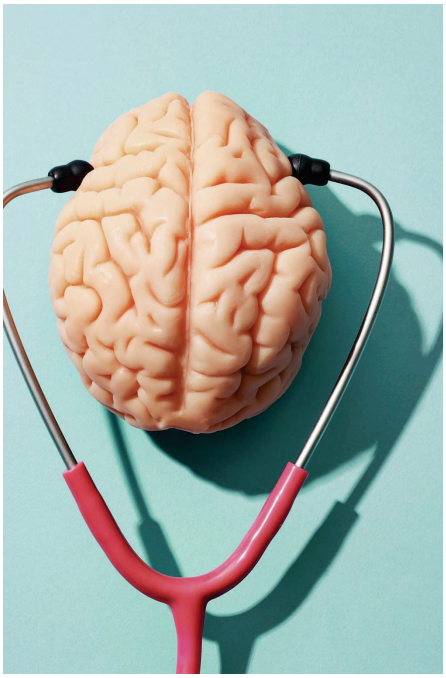
- To Be Better
- Because we all have stories
  - Some good...
  - Some not so good...

Functional Seizures (FS)



- Psychogenic Nonepileptic Seizures
- Pseudoseizures
- Stress Seizure

Tilahun BBS, Bautista JF. Psychogenic nonepileptic seizure: An empathetic, practical approach. Cleve Clin J Med. 2022 May 2;89(5):252-259. doi: 10.3949/ccjm.89a.21109. PMID: 35500924



Functional Seizures: What it is...

- Paroxysmal events that resemble an epileptic seizure in signs and symptoms, but is not caused by abnormal epileptiform electrical activity in the brain

Asadi-Pooya AA, Brigo F, Tolchin B, Valente KD. Functional seizures are not less important than epilepsy. Epilepsy Behav Rep. 2021 Oct 21;16:100495. doi: 10.1016/j.ebr.2021.100495. PMID: 34805820; PMCID: PMC8585631

Functional Seizures: What it is **Not…**

- **Not** intentional
- **Not** conscious
- **Not** factitious or malingering

Janneck L, Muncie M. Functional Seizures. EMRAP Corependium. May 2023

Functional Seizures: Stats

- Incidence: 3-5 per 100k
  - Prevalence: 2-108.5 per 100k
- Age: Most common between ages 15-35
  - 3/4 women*
    - Or not…
- 1 in 4 patients in EMU found to have functional seizures

Tilahun BBS, Bautista JF. Psychogenic nonepileptic seizure: An empathetic, practical approach. Cleve Clin J Med. 2022 May 2;89(5):252-259. doi: 10.3949/ccjm.89a.21109. PMID: 35500924

Lehn A, Watson E, Ryan EG, Jones M, Cheah V, Dionisio S. Psychogenic nonepileptic seizures treated as epileptic seizures in the emergency department. Epilepsia. 2021 Oct;62(10):2416-2425. doi: 10.1111/epi.17038. Epub 2021 Aug 15. PMID: 34396517.

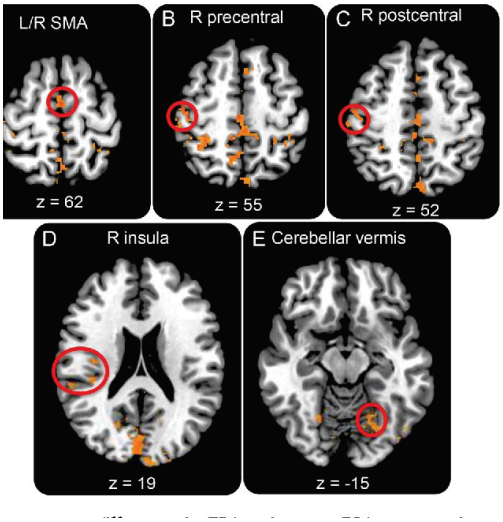
Functional Seizures: Some more stats…

- 10-22% of pts w/ functional seizures have epilepsy
- 12% of people with epilepsy also have functional seizures
- Mortality Rate is 2.5 higher than general population
  - Similar rates to those with epilepsy
  - And, is 8.3% higher in those <30 y/o

Janneck L, Muncie M. Functional Seizures. EMRAP Corependium. May 2023

Functional Seizures: Pathophysiology

- Studies show abnormalities in limbic structures
  - Includes amygdala, hippocampus, prefrontal cortex
- Childhood trauma is a significant risk factor
- Association with Comorbid Conditions
  - Migraine (50%)
  - Asthma (33%)
  - Depression (36-80%)
- A form of dissociation?



Tilahun BBS, Bautista JF. Psychogenic nonepileptic seizure: An empathetic, practical approach. Cleve Clin J Med. 2022 May 2;89(5):252-259. doi: 10.3949/ccjm.89a.21109. PMID: 35500924

FIGURE 1 Decreased functional connectivity between the right temporoparietal junction and bilateral sensorimotor regions in patients with functional movement disorder*. *Reproduced from Maurer CW, LaFaver K, Ameli R, Epstein SA, Hallett M, Horovitz SG. Impaired self-agency in functional movement disorders: A resting-state fMRI study. Neurology. 2016;87(6):564-570. https://n.neurology.org/

Functional Seizures: Clinical Features

Which is Which?

Functional Seizures: Clinical Features

TABLE 2 Clinical features distinguishing functional from epileptic seizures[38,72-74]

Clinical sign	Notes	Reliability[a]
Highly suggestive of functional seizures		
Closed eyelids during ictal peak	Patients may actively resist eyelid opening.	+++
Prolonged duration	Most epileptic seizures will stop spontaneously in 2 min or less. Particularly useful if it resolves spontaneously after prolonged duration, without significant postictal period. Caution: patients with status epilepticus will have prolonged seizure activity.	++
Fluctuating course	Movements may wax and wane in intensity or stop and start.	++
Ictal awareness/memory of seizure	Only relevant for generalized seizures (abnormal movements of all four limbs). Caution: frontal lobe seizures can involve bizarre movements with retained awareness. Loss of awareness is standard for most functional seizures.	++
Ictal/postictal weeping	Relatively specific for functional seizures, although low sensitivity. May also have other signs of emotional distress.	++
Asynchronous limb movements	Caution: can also be present in frontal lobe seizures.	++
Side to side head shaking	May rarely be present in epileptic seizures. Good differentiator for generalized shaking events only.	++
Response to stimuli during ictal period	Only applies to generalized shaking attacks.	++
Highly suggestive of epileptic seizures		
Figure of four sign	One arm flexed at elbow, other arm extended at the elbow, usually present just before secondary generalization.	+++
Guttural cry/scream	During tonic phase, typically at seizure onset.	++
Prolonged rigid phase with cessation of respiration	Based on authors' experience.	++
Postictal stertorous breathing	Low-pitched sound from back of throat, like sound from nasal congestion or snoring.	+++
Unhelpful features common to both		
Tongue biting Injury (although severe burns and shoulder dislocation should prompt consideration of epilepsy) Urinary incontinence Attack appearing from sleep/no witnesses to seizure Presence of aura or postictal confusion Breath holding High serum lactate after an event[71]		

+++ = highly reliable; ++ = reliable; + = suggestive

[a]Reliability determined based on available clinical data[73,75-77] and author consensus.

Finkelstein, S. A., Cortel-leblanc, M. A., Cortel-leblanc, A., & Stone, J. (2021). Functional Neurological Disorder in the Emergency Department. *Academic Emergency Medicine: A Global Journal of Emergency Care (AEM)*. Advance online publication. https://doi.org/10.1111/acem.14263

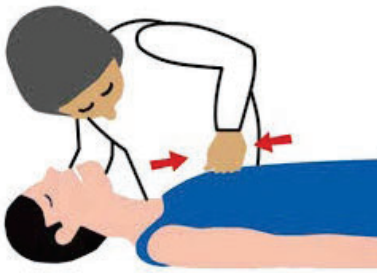
Functional Seizures: Diagnosis

- This is hard, and many times you can't
  - That's ok.
- VEEG and Neurology
  - But remember, EEG is NOT perfect either...

Functional Seizures - Treatment Do's and Don'ts

Do:

- ABC-NGT
- Consider broad work-up and Neurology consultation
- Compassion, Empathy, and Clarity
- Normalize and destigmatize the condition
- Emphasize treatment options and a path forward



Functional Seizures: Treatment Do's and Don'ts

DON'Ts

- Anchor
- Noxious stimuli
- Escalation to meds or intubation (if possible)
- Anti-epileptics, which may exacerbate functional seizures

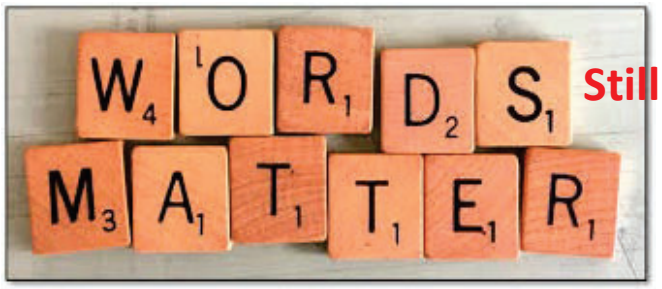
Tilahun BBS, Bautista JF. Psychogenic nonepileptic seizure: An empathetic, practical approach. Cleve Clin J Med. 2022 May 2;89(5):252-259. doi: 10.3949/ccjm.89a.21109. PMID: 35500924

Functional Seizures: Dispo

Home

Admission...

Functional Neurological Disorder:



NOT:

- Conversion
- Psychogenic
- Psycho-somatic
- Pseudo-anything
- Hysteria...

Functional Neurological Disorder: What it is...

An INVOLUNTARY change in motor or sensory function that is inconsistent with patterns of known neurologic diseases or other medical conditions, AND significantly impacts the patient's ability to function

Peeling JL, Muzio MR. Functional Neurologic Disorder. 2023 May 8. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan–. PMID: 31855394.

Functional Neurological Disorder: What it is **NOT**...

Intentional

Conscious

Factitious

Malingering

Streitz M, Hanlin E. Conversion Disorders. EMRAP Corependium.. August 2022

Functional Neurological Disorder: Stats

Incidence = 4-12 per 100k

Prevalence = 0.4-4%

7-15% of stroke mimics are found to have FND

9% of acute neurological admissions

Finkelstein, S. A., Cortel-leblanc, M. A., Cortel-leblanc, A., & Stone, J. (2021). Functional Neurological Disorder in the Emergency Department. *Academic Emergency Medicine: A Global Journal of Emergency Care (AEM)*. Advance online publication. https://doi.org/10.1111/acem.14263

Functional Neurological Disorder: Pathophys

Neurobiological Model

- Functional neuroimaging shows differences/dysfunction of certain brain networks
  - Attention
  - Perception
  - Sense of agency
- Dysregulation of attention

Psychiatric

Streitz M, Hanlin E. Conversion Disorders. EMRAP Corependium.. August 2022

Functional Neurological Disorder:

Clinical Features + Dx

IS IT A STROKE!?!?! ?!?!?!?!?!?

Or another organic neurological emergency?

Functional Neurological Disorder:

Clinical Features + Dx

Clinical sign	Description	Reliability[a]
Hoover's sign[20,35-37,39]	Weakness of voluntary hip extension that resolves with voluntary contralateral resisted hip flexion. Difficult to detect in bilateral leg weakness.	+++
Platysma overactivation[40]	Contraction of one side of the platysma, creating the effect of a facial droop.	++
Hip abductor sign[37]	Return of strength to hip abduction in the weak leg with contralateral hip abduction against resistance	++
Give-way/collapsing weakness[35,41,42]	Strength is initially normal and then collapses with resistance.	++
Dragging monoplegic leg[20,35]	Plegic leg is dragged behind body often with hip internal or external rotation and without hip circumduction.	++
Drift without pronation[35,43]	Isolated downward arm-drift without associated pronation.	+
Global pattern of weakness[35,44]	Equal weakness of both flexor and extensor muscles, both proximally and distally.	+
Motor inconsistencies[45]	Inability to produce one movement, while using the same muscles to produce a different movement. For example, a patient may have difficulty dorsiflexing while supine, but be able to stand on heels without difficulty.	+

+++ = highly reliable; ++ = reliable; + = suggestive.

[a]Reliability determined based on available clinical data[34] and author consensus.

Finkelstein, S. A., Cortel-leblanc, M. A., Cortel-leblanc, A., & Stone, J. (2021). Functional Neurological Disorder in the Emergency Department. *Academic Emergency Medicine: A Global Journal of Emergency Care (AEM)*. Advance online publication. https://doi.org/10.1111/acem.14263

Functional Neurological Disorder: Work-up + Tx

- ABC NGT
- Thorough H&P
- Neuro Consult +/- Stroke Alert
- TPA?
- Other Meds?
  - Haldol
  - Seroquel
  - Versed

Functional Neurological Disorder- Treatment

Compassion, Empathy, and Clarity

Normalize + Destigmatize + Name it

Don't Anchor – Don't Dismiss

Emphasize treatment options and a path forward

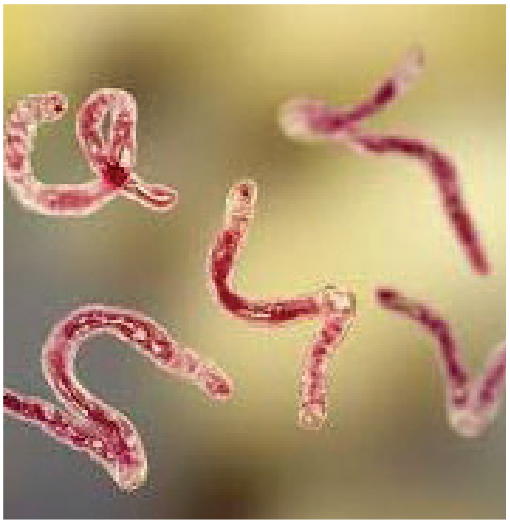
Functional Neurological Disorder: Dispo

Home...

Admission...

Delusional Parasitosis: What is it?

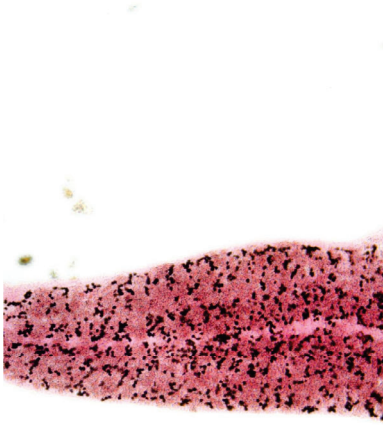
- Ekbom Syndrome
  - Fixed false belief of parasitic infection
  - Normal functioning outside of belief
- Morgellons Syndrome
  - Specific variant with fibers coming from skin



McLellan, Susan. Perspectives: Delusional Parasitosis. CDC Yellow Book. 2024

Delusional Parasitosis: Pathophysiology/Differential

- Primary Psychiatric
- Secondary
  - Is it actually a parasite?
  - Trauma
  - Dementia
  - CVA
  - Thyroid or nutritional deficiencies
  - Encephalitis
  - Syphilis and HIV
  - Hepatitis
  - Eosinophilia
  - SUD
  - Cipro?



Gold A, Roit Z, Llovera I. Pitfalls and Pearls in Delusional Parasitosis. Clin Pract Cases Emerg Med. 2019 Oct 14;3(4):387-389. doi: 10.5811/cpcem.2019.8.44619. PMID: 31763595; PMCID: PMC6861047.

Delusional Parasitosis: Diagnosis

Don't anchor on initial diagnosis — Conduct a thorough history and physical examination

Consider red flags such as age and travel history — Conduct necessary labs, imaging, lumbar puncture, and consultation

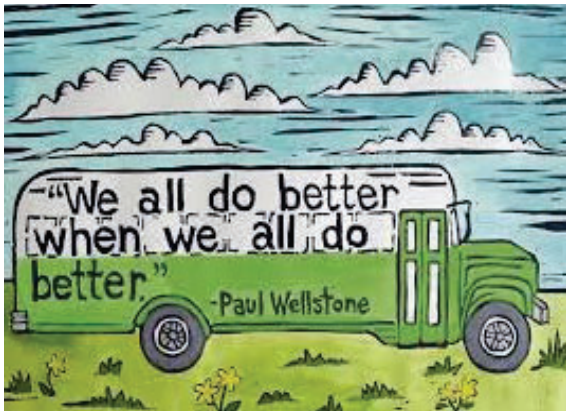
Treatment and disposition should be carefully considered

Delusional Parasitosis: Treatment and Disposition

- Focus on the realness of the symptoms, and goal for symptoms control
  - The pain
  - The suffering
- Reassurance, Destigmatize, Provide a Path Forward
- Anti-Psychotics:
  - Zyprexa
  - Risperdal
- Referral:
  - PCP
  - Psychiatry
  - Dermatology if needed

Guàrdia A, González-Rodríguez A, Betriu M, Estrada F, Seeman MV, Uribe IP, Labad J, Vidal DP. Case series of delusional parasitosis in an emergency department: Sociodemographic features and clinical outcomes. Eur Psychiatry. 2021 Aug 13;64(Suppl 1):S517. doi: 10.1192/j.eurpsy.2021.1384. PMCID: PMC9475867.

These are difficult encounters...



References:

- Finkelstein, S. A., Cortel-leblanc, M. A., Cortel-leblanc, A., & Stone, J. (2021). Functional Neurological Disorder in the Emergency Department. *Academic Emergency Medicine: A Global Journal of Emergency Care (AEM)*. Advance online publication. https://doi.org/10.1111/acem.14263
- Tilahun BBS, Bautista JF. Psychogenic nonepileptic seizure: An empathetic, practical approach. Cleve Clin J Med. 2022 May 2;89(5):252-259. doi: 10.3949/ccjm.89a.21109. PMID: 35500924
- *Helman, A. Reid, A. Koblic, P. Ep 132 Emergency Approach to Seizures and Seizure Management. Emergency Medicine Cases. November, 2019. https://emergencymedicinecases.com/approach-seizures-management. Accessed*
- Asadi-Pooya AA, Brigo F, Tolchin B, Valente KD. Functional seizures are not less important than epilepsy. Epilepsy Behav Rep. 2021 Oct 21;16:100495. doi: 10.1016/j.ebr.2021.100495. PMID: 34805820; PMCID: PMC8585631.
- Chen, J., Zeng, Z., Fang, Z. *et al.* Risk factors for thrombolysis-related intracranial hemorrhage: a systematic review and meta-analysis. *Thrombosis J* **21**, 27 (2023). https://doi.org/10.1186/s12959-023-00467-6
- Streitz M, Hanlin E. Conversion Disorders. EMRAP Corependium.. August 2022
- Janneck L, Muncie M. Functional Seizures. EMRAP Corependium. May 2023

References:

- Ramamurthy S, Steven Brown L, Agostini M, Alick Lindstorm S, Dave H, Dieppa M, Ding K, Doyle A, Hays R, Harvey J, Perven G, Podkorytova I, Zepeda R, Das RR. Emergency department visits and readmissions in patients with psychogenic nonepileptic seizures (PNES) at a safety net hospital. Epilepsy Behav. 2021 Sep;122:108225. doi: 10.1016/j.yebeh.2021.108225. Epub 2021 Aug 2. PMID: 34352667.
- Meehan WJ, Badreshia S, Mackley CL. Successful Treatment of Delusions of Parasitosis With Olanzapine. *Arch Dermatol.* 2006;142(3):352–355. doi:10.1001/archderm.142.3.352
- Guàrdia A, González-Rodríguez A, Betriu M, Estrada F, Seeman MV, Uribe IP, Labad J, Vidal DP. Case series of delusional parasitosis in an emergency department: Sociodemographic features and clinical outcomes. Eur Psychiatry. 2021 Aug 13;64(Suppl 1):S517. doi: 10.1192/j.eurpsy.2021.1384. PMCID: PMC9475867.
- Gold A, Roit Z, Llovera I. Pitfalls and Pearls in Delusional Parasitosis. Clin Pract Cases Emerg Med. 2019 Oct 14;3(4):387-389. doi: 10.5811/cpcem.2019.8.44619. PMID: 31763595; PMCID: PMC6861047.
- McLellan, Susan. Perspectives: Delusional Parasitosis. CDC Yellow Book. 2024
- Asadi-Pooya AA, Brigo F, Tolchin B, Valente KD. Functional seizures are not less important than epilepsy. Epilepsy Behav Rep. 2021 Oct 21;16:100495. doi: 10.1016/j.ebr.2021.100495. PMID: 34805820; PMCID: PMC8585631
- Peeling JL, Muzio MR. Functional Neurologic Disorder. 2023 May 8. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan–. PMID: 31855394.
- Lehn A, Watson E, Ryan EG, Jones M, Cheah V, Dionisio S. Psychogenic nonepileptic seizures treated as epileptic seizures in the emergency department. Epilepsia. 2021 Oct;62(10):2416-2425. doi: 10.1111/epi.17038. Epub 2021 Aug 15. PMID: 34396517.