Disturbi funzionali in Neurologia

Pseudocrisi

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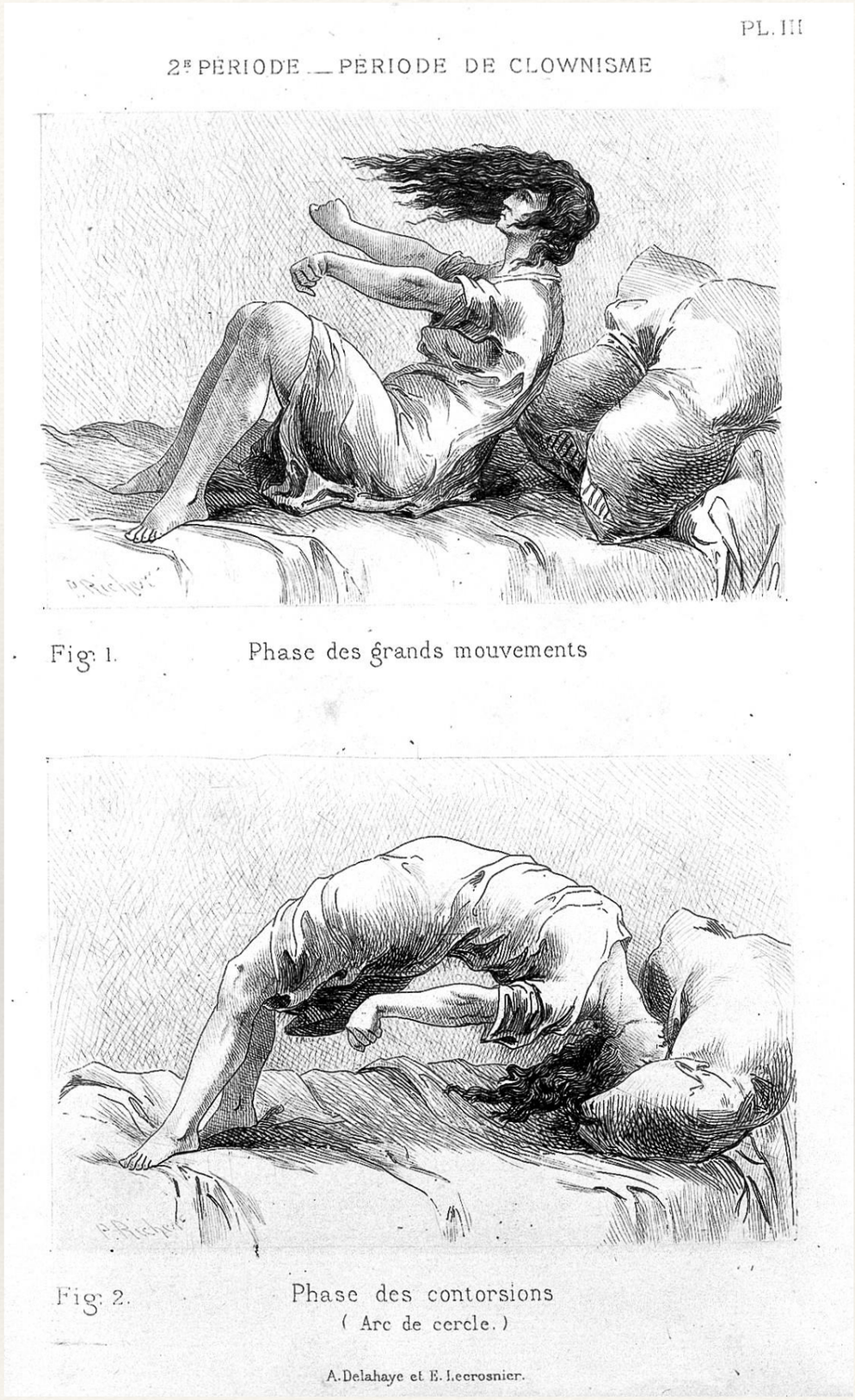


Paul Richer, Études cliniques sur l'hystéro-épilepsie ou grande hystérie, Paris 1881)

Pseudoseizures: an old term

- *"Events that appear to be epileptic seizures but, in fact, do not represent the manifestation of abnormal excessive synchronous cortical activity, which defines epileptic seizures".*

- They are not a variation of epilepsy but are of psychiatric origin.

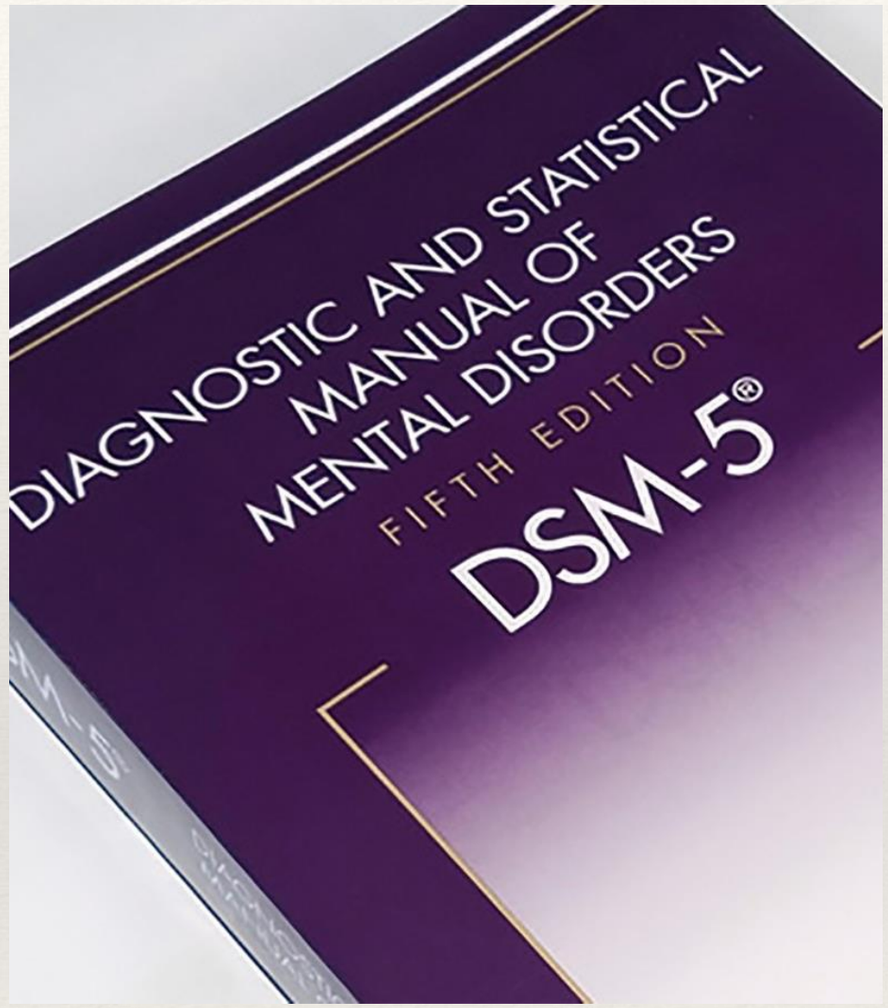
- Other terms used in the past include hysterical seizures, psychogenic seizures, and others. The most standard current terminology is ***psychogenic non epileptic seizures (PNES).***

- ***Huff and Murr, in StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021.***

PNES: a psychiatric disorder

- *According to the DSM 5, PNES are a subgroup of conversion disorders, while the ICD 10 classifies PNES as **dissociative disorders**.*
- **Psychiatric comorbidities:**
- personality disorders
- post-traumatic stress disorders,
- anxiety
- major depressive disorders
- **RISK FACTORS**
- childhood history of abuse
- female gender
- Trauma, brain injury, surgical procedures
- learning disability

- *Anzillotti et al, Frontiers of Neurol, Jun 2020*



"a disruption and/or discontinuity in the normal integration of consciousness, memory, identity, emotion, perception, body representation, motor control, and behavior"

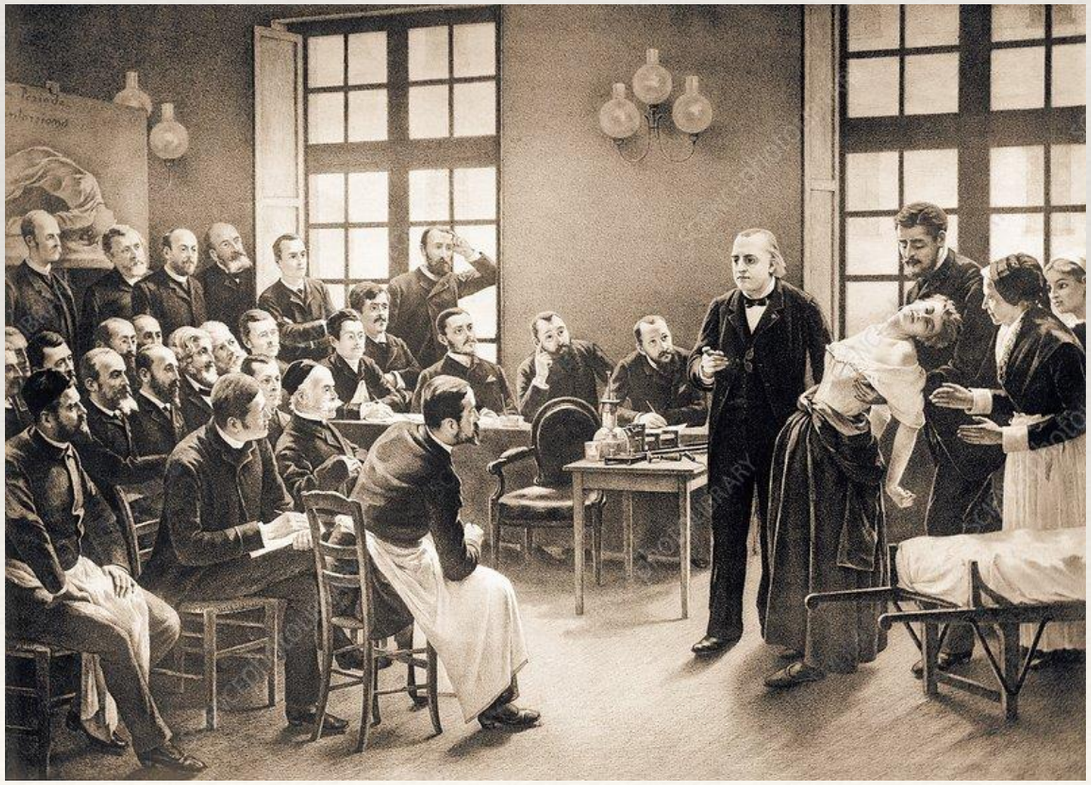
Epidemiology

- Incidence: 1.4–4.9/100,000/year,
- Prevalence: between 2 and 33 per 100,000.
- Age onset: young adulthood, although the disorder can occur at any age
- *5-10% of the outpatients of epilepsy clinics*
- *20–40% of the inpatients of epilepsy monitoring units*

PNES: Classification

All these classifications can be simply reconfigured in terms of “motor” vs. “non-motor” PNES or PNES with or without “unresponsiveness”.

Anzellotti et al, Frontiers of Neurol, Jun 2020



Charcot demonstrating Hysteria, 1886

Gröppel et al.	1. Major motor 2. Minor motor or trembling 3. Atonic psychogenic seizures
Selwa et al.	1. Catatonic 2. Trashing 3. Automatisms 4. Tremor 5. Intermittent 6. Subjective
Seneviratne et al.	1. Rhytmic motor 2. Hypermotor 3. Complex motor 4. Dialeptic 5. Non-epileptic auras 6. Mixed
Hubsh et al.	1. Dystonic attack with primitive gestural activity 2. Paucikinetic attack (with preserved responsiveness) 3. Pseudosyncope 4. Hyperkinetic prolonged attack with hyperventilation and auras 5. Axial dystonic prolonged attack
Dhiman et al.	1. Hypermotor 2. Akinetic 3. Focal motor 4. PNES with “subjective symptoms
Magaudda et al.	1. Abnormal motor 2. Affective emotional behavior phenomena 3. Dialeptic coma-like state 4. Aura 5. Mixed

PNES as a conversion disorder (C.D.)

- "patients who show difficulty in expressing conflicts verbally, sometimes express distress somatically."
  Stone et al, 2011 (about C.D.)

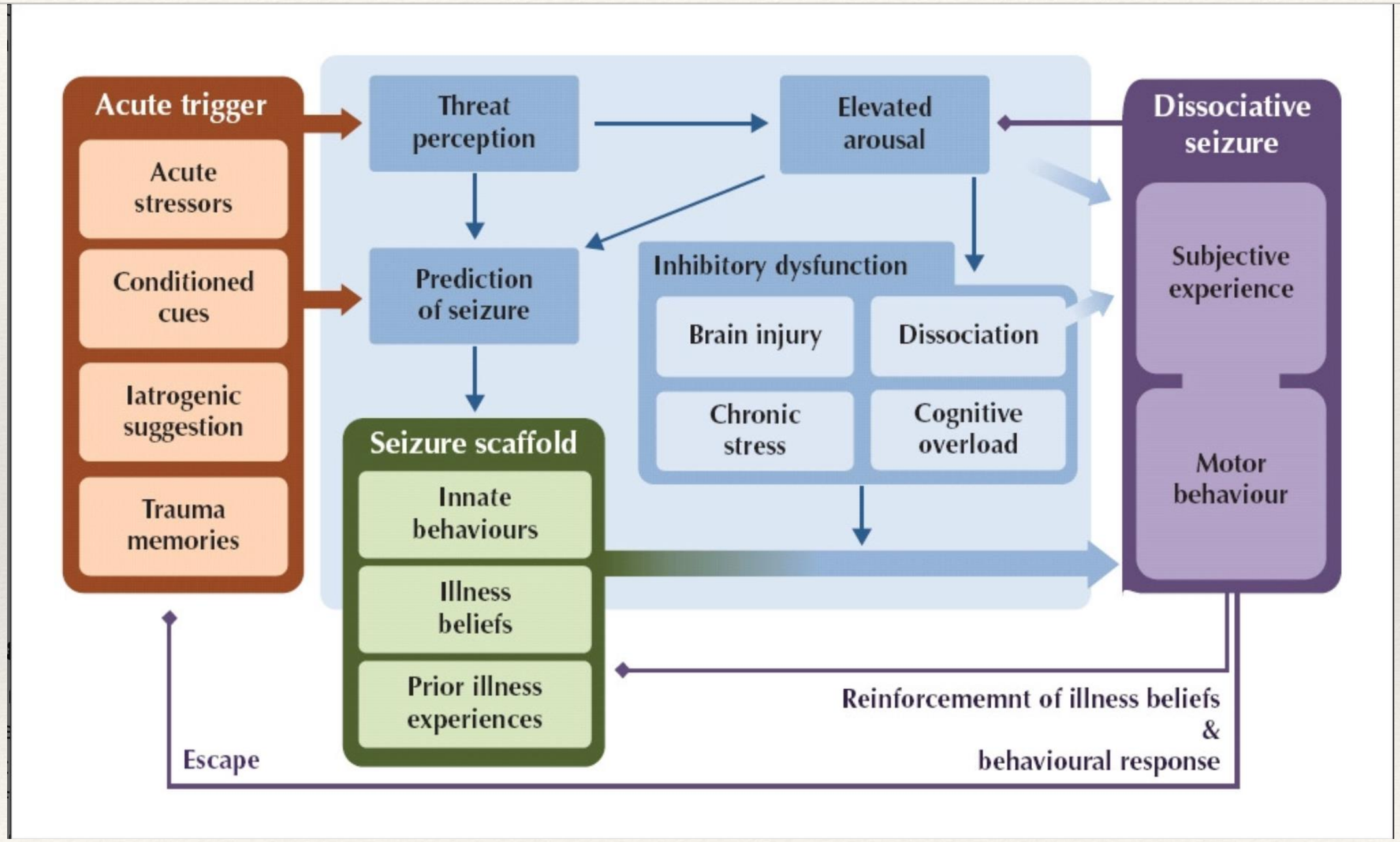
- People who experience PNES often exhibit a variety of dissociative symptoms

PNES Pathophysiology

Freudian model	PNES is a physical manifestation of emotional stress
Moore and Baker model	PNES results from learned behavior and activated via operant conditioning
Dissociative models by Bowman and Baslet	PNES results from dissociated memories or mental functions that are set in motion by a traumatic event (Bowman) PNES is an acute dissociative response to a threat or a state of high arousal (Baslet)
Integrated Cognitive Model (ICM) by Brown and Rewber	PNES results from an altered stimulus that in physiological conditions would have caused the activation of memory networks; the response to the stimulus depends on the familiarity with it.

Anzellotti et al, Frontiers of Neurol, Jun 2020

Integrated Cognitive Model (ICM)



Popkirov et al, Epileptic Disorders, 2019

PNES: implications for patient and physician

- PNES patients commonly experience delays in the diagnosis and/or receive inappropriate treatment.
- Antiepileptic drugs may exacerbate the disorder
- Risk Factors (es. brain injury) may conduct to misdiagnosis
- ***Epilepsy in clinical history: increase diagnosis' difficulty and may conduct to "pseudo-refractory epilepsy"***
- ***About 10% of patients with PNES also exhibit genuine epileptic seizures (Labiner, Epilepsia, 2019)***

Anzellotti et al, Frontiers of Neurol, Jun 2020

Seizures and PNES: differential diagnosis

Anzellotti et al, Frontiers of Neurol, Jun 2020

	PNES	SEIZURES
Aura	Less frequent	More frequent
Length of ictal events	> 10 minutes	<70 s (<2 min for tonic-clonic seizures)
Seizure pattern	Non-stereotypical, less organized spatial patterns, variable rhythmicity, and amplitude of movements	Stereotypical and organized progression
Clinical findings	Asynchronous limb movements, out-of-phase clonic activity, rhythmic shaking movements with episodes of inactivity, side-to-side head movements, pelvic movements, dystonic body posturing, closure of the eyes during the event	Bilateral adduction and external rotations of limbs followed by tonic extension of all four limbs, then the production of diffuse clonic jerking movements before the ictal offset
Vocalization	Present not only at the beginning of the event, can fluctuate, persist and be present, with different pitch intensities, throughout the whole course of the ictal episode	At the beginning of the seizures
Subjective symptoms	Less frequent	More frequent
Urinary incontinence	Less frequent	More frequent
Occurrennce at night	Less frequent	More frequent
Ictal self injury	Less frequent	More frequent

PNES: Diagnosis

- Anamnesis
- Home video of suspect episodes
- Video EEG monitoring
- Supplementary diagnostic procedures (Prolactine, BDNF: no conclusive)
- Neuroimaging
- Neuropsycological findings

PNES: Diagnosis

NeuroImage: Clinical 16 (2017) 210–221

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journal homepage: www.elsevier.com/locate/ynicl

Neuroimaging studies in patients with psychogenic non-epileptic seizures: A systematic meta-review

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- *The findings … suggest that alterations in functional connectivity in brain regions associated with attention and regulatory processes, memory, emotion processing and sensory and motor function may be compromised in patients with PNES.*
- Limits: small number of studies; small size of the samples; difficulty of defining "PNES-clusters"; the MRI studies cannot infer any causal relationship between the brain imaging results and PNES.
- *Although the results presented appear inconclusive, they are consistent with an association between structural and functional brain abnormalities and PNES*

PNES Management

- *Communication of the diagnosis*
- Terms like “hysterical seizures” and “pseudoseizures” should be avoided, preferring “attacks” or “seizures”
- (Plug et al, Epilepsia, 2009)

- *Acute therapeutic intervention*
- psychiatric comorbidities management
- early tapering and discontinuation of ASD (consider use of lamotrigine or valproate)

- *Long term intervention*
- Cognitive-behavioral therapy currently exhibits the most robust experimental and clinical evidence of efficacy.
- (Goldstein et al, Neurology, 2010)

Am. J. EEG Technol.
16:23-29, 1976

The Hysterical Seizure

Susan M. Johnson and James A. Lewis

University of Colorado Medical Center, Denver, Colorado 80220

A common problem for neurologists has been the differential diagnosis of “real” versus “hysterical” fits (10,13,14). Before discussing this, a review of hysteria is in order, particularly since there are certain terms that are easily misunderstood. Although many of the definitions may overlap, some of them can be specific.

First, the term hysteria itself: in most literature this term has come to mean the presence of “gross, often dramatic, somatic symptoms, unexplained on an organic basis” (2). Of all the mental diseases, hysteria has probably most occupied the interest of medical writers since antiquity. From the earliest times it has been associated with a disturbance in sexuality. The term itself is derived from the Greek word *hystera*, meaning uterus. Veith, in her book *Hysteria: The History of a Disease*, feels that it is a term that defies definition or concrete understanding (12). Nevertheless, there are certain clinical characteristics that have persisted through the years. First is its marked predominance in women. Second is the way the patient adapts to her environment prior to the appearance of symptoms and the way the symptoms are presented to the observer.

Many authors feel that hysteria and hysterical personality must be differentiated, for patients with the somatic complaints of hysteria do not necessarily exhibit the facets of a hysterical personality. According to Alarcon, the hysterical personality is characterized by: (a) emotional lability, in which the patient exhibits outbursts precipitated by insufficient cause which change rapidly and are superficial; (b) dependency; (c) excitability; (d) egocentrism or narcissism, in which the patient demonstrates a strong tendency to satisfy his own needs regardless of others; (e) seductiveness — a constant display of charm to impress and amuse others; (f) suggestibility; and (g) histrionic behavior, which implies a dramatic air of artificial superiority devised to gain sympathy, attention, or even admiration (2).

- *Di tutte le cose che ebbi modo di osservare durante il mio soggiorno presso Charcot, nessuna mi colpì tanto quanto le sue ultime ricerche sull'isteria….egli dimostrò che i fenomeni isterici sono qualcosa di autentico e conforme a uno scopo, che l'isteria è molto frequente negli uomini, che paralisi e contratture isteriche possono essere provocate dalla suggestione ipnotica e che questi prodotti artificiali hanno, fin nei minimi dettagli, le stesse caratteristiche degli attacchi isterici spontanei che spesso vengono provocati da un trauma."*
- *S. Freud*