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THE ROLE OF NEURODIAGNOSTICS IN FUNCTIONAL DISORDERS

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The role of neurodiagnostics in functional disorders is primarily to exclude organic diseases. However, new and novel forms of neuroimaging in recent years have provided us with some insight in our understanding of the underlying neurological substrates that may play a role in the symptoms of functional disorders. Somatoform disorders are common in the population with an approximate prevalence of 5% in the primary care. Among unselected patients attending neurology outpatient clinics in the United Kingdom, as many as 4% have been estimated to have conversion disorders.[27] A significant number of patients will remain seriously disabled as a result of these illnesses that are known world-wide and in all cultures.

The latest edition of the *Diagnostic and Statistical Manual of Mental Disorders* (DSM-IV)[2] clearly sets out the diagnostic criteria of the somatization disorder and the differential diagnosis of the somatoform disorders; these issues will not be reviewed in this section. It may be convenient to retain the use of the word *hysteria* to describe not a single disease but a broad functional syndrome encompassing dissociative, conversion, and somatizing symptoms mimicking primary dysfunction of the nervous system (pseudoneurologic symptoms). A thorough neurological assessment is the most important diagnostic step in patients with hysteria.[1]

CLINICAL PHENOMENOLOGY OF HYSTERIA

The common clinical presentations of hysteria can be classified on the basis of the dominant pseudo-neurologic symptom. This allows appropriate selection of the neurodiagnostic tools to exclude organic diseases.

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1. Disorders of motor functions: hysterical motor paralysis, loss of balance, incontinence, dysphagia, and movement disorders
2. Disorders of somatic sensations: hysterical sensory loss, hysterical pain, and neuralgia
3. Disorders of special senses: hysterical blindness, double vision or blurred vision, deafness, anosmia, and dizziness
4. Paroxysmal disorders of neurological function: hysterical seizures and fainting attacks
5. Derangements of cognition and language: hysterical fugue states, amnesia, dementia, and speech disorders
6. Disorders of autonomic and neuroendocrine functions: hysterical fever, hysterical vomiting, hysterical asthenia (fatigue), hysterical pregnancy, and lactation

NEUROLOGICAL ASSESSMENT OF A PATIENT WITH HYSTERICAL SYMPTOMS

Motor

Absence of objective abnormalities in the motor neuron pathways (e.g., reflex changes) is a useful pointer to a possible functional disorder in someone with persistent motor weakness. Hysterical paralysis may involve an arm, a leg, one side of the body, or both legs. Movements are characteristically slow, hesitant, and poorly sustained when the affected limb can be moved. It is often possible to demonstrate that the strength of voluntary movement is proportional or less than the resistance offered and when the resistance is suddenly withdrawn, there is no follow-through or rebound as is normally the case. The muscle tone and reflexes are generally normal, and severe muscle atrophy is lacking. The only differential diagnosis is acute upper motor neuron lesion that lacks the usual changes in reflexes and muscle tone. For weakness involving one arm or one side of body, MRI of brain is recommended; MRI of the cervical cord is required in addition if both arms are weak. MRI of brain as well as that of the entire spinal cord in a body coil may be the best approach for a patient with bilateral leg weakness without possible anatomic localization. For hysterical incontinence, MRI of brain and the lower spinal cord with adequate imaging of conus and cauda equina is necessary. Common diagnostic pitfalls of hysterical motor weakness are demyelination and vascular disease; intermittent symptoms of motor weakness with normal examination in between warrant exclusion of demyelination (Uhthoff's phenomenon) if the symptoms are lateralized and periodic paralysis or myasthenia gravis if the symptoms are generalized. Careful neurophysiological examination may be required in the latter cases with repetitive motor nerve stimulation study and EMG. In hysterical motor weakness, EMG shows reduced recruitment of motor units on attempted contraction of the weak muscles that improve on encouragement.

One needs to exclude obstructive uropathy as well as lesions in the upper motor neuron (e.g., multiple sclerosis) and lower motor neuron (e.g., cauda equina syndrome) pathways in patients presenting with urinary retention or difficulty in passing urine. Measurement of residual urinary volume may be useful to assess bladder atony. Formal assessment of urinary bladder function may be obtained by a cystometrogram that measures intravesicular pressure as a function of the volume of saline flowing into the catheterized bladder under gravity. However, patients with hysteric retention may not report sensation of bladder fullness and/or suppress motor function of the bladder, thus simulating a deafferented and/or motor denervated

bladder caused by organic pathology (e.g., sacral root lesions by diabetes, neurosyphilis, or a spinal cord lesion below T12 due to a tumor or developmental cord lesion). Although in organic cases, anesthesia in the saddle area with or without loss of anal sphincter control may be seen, this may not always be the rule (e.g., tethered cord syndrome). Full neuraxis (brain and spinal cord) imaging with MRI and electrophysiology (for peripheral nerve and root lesions) will be required in all cases of hysterical urinary retention to exclude an organic pathology.

Hysterical gait can be diagnosed clinically. In walking, a patient with hysterical weakness tends to hesitate and advance the weaker leg in a bizarre manner (tremulous, ataxic, or both). The patient does not lift the foot from floor while walking, and unlike the characteristic circumduction of the paralyzed leg in organic hemiparesis, patients with hysteric hemiparesis often drag or push the leg along. The hysterical paraplegic cannot drag his/her legs and depends entirely on bilateral support; some patients may remain helpless in bed or bound to a wheelchair with secondary changes in the muscles (contracture). In a study of 60 cases of hysterical gait, the hemiparetic and monoplegic forms were twice as frequent as the quadriparetic form.[21] Sudden falls without protective body movements are common; the balance is inconsistent, and one may see postures of extreme dystonia, wild veering to one or the other side, and gyrating body movements. One interesting aspect of hysterical gait is that in most cases, leg movements in the bed are unimpaired. *Astasia-abasia* is a condition in which patients, although unable to either stand or walk, demonstrate normal motor function of their legs while lying or sitting in bed. Althougth astasia-abasia without any other neurological features is invariably hysterical in the author's experience, this form of gait disorder may be seen rarely in frontal lobe disease and normal pressure hydrocephalus (gait apraxia or Bruns' ataxia). A CT scan (or MRI) of the brain is therefore recommended in patients with astasia-abasia.

Globus hystericus is a common pseudoneurologic symptom in which swallowing is affected. Patients often complain of a choking sensation or painful "lump" in the throat that may be intermittent or persistent. The results of barium swallow of the esophagus, video-fluoroscopy of swallowing, and vocal cord examination are normal.

Both tremors and choreiform movements are known in hysterical movement disorders. Hysterical tremors are usually gross, unpredictable, irregular, and affect one limb more severely. If the examiner restrains the affected limb, the tremors move to another part of the body (migratory tremor). Recently, it was shown that unlike organic limb tremors, patients with hysteric tremors often demonstrate an increase in the tremor amplitude when the affected extremity was loaded with additional weights. It was proposed that the increasing amplitude in hysterical limb tremors was caused by co-activation of the muscles in order to maintain the oscillation (the co-activation sign of psychogenic tremor[9]). Hysterical chorea is often associated with violent swaying of the top half of the body. All forms of hysterical movement disorders reduce by fatigue and may disappear with distraction. It is important to consider drug-induced (neuroleptic) movement disorders, Wilson's disease, rheumatic chorea, and Huntington's disease in the differential diagnosis. Paroxysmal dystonic posturing, however, is more likely to be organic; disorders of calcium metabolism (pseudohypoparathyroidism) and paroxysmal dystonic choreoathetosis must be considered as possibilities. A CT scan of brain is often helpful if Huntington's disease or pseudohypoparathyroidism are considerations in the differential diagnosis.

Sensory

Although relatively uncommon, hysterical sensory loss may present as hemianesthesia, facial anesthesia, or sensory loss affecting hands and/or feet. Paresthetic sensory symptoms (numbness and/tingling) are more common. The pattern of sensory impairment does not follow the normal anatomical distribution of the peripheral nerves, nerve root, and the ascending sensory tracts. Often, patients may complain of a sharp cut-off of sensations below a line in one or more limbs affecting all modalities equally. This is in contrast with peripheral nerve lesions that show a gradient in which the area of sensory loss may differ between the nerve fiber types. In hysterical sensory loss affecting one half of the face, sensations may be cut off at the hairline above and over the angle of the jaw at the side that would not be in keeping with a trigeminal nerve pathology. Hysterical hemisensory loss has a sharp midline cut, involving precisely one half of the body with loss of all modalities of sensations. When present, midline loss of vibratory sensation over one half of the head is a characteristic feature of hysterical sensory syndrome. Touch, pain, smell, taste, and vision may all be affected on the same side in hysterical hemisensory loss, which is anatomically improbable. In organic hemisensory syndrome, loss of sensations will follow the anatomy of the ascending leminiscal system. Thus, in a spinal cord hemisensory syndrome, loss of temperature and pain is contralateral to the side of loss of propioception and vibration (e.g., Brown-Séquard syndrome). Crossed sensory disturbance is also a characteristic feature of the brainstem sensory syndrome, i.e., loss of pain and temperature sensation on one side of the face and opposite side of the body. Higher cerebral lesions (e.g., anterior parietal lesion of Verger-Déjerine[1]) typically abolish discriminative sensory functions of the opposite leg, arm, and side of face without impairment of the primary modalities of sensations unless the lesion is deep and destructive. Since demyelination of the central nervous system may manifest as somatic hemisensory syndrome, MRI scan of brain may be required in these cases. However, organic hemisensory syndrome due to a single lesion will never affect somatic sensations (e.g., pain) and special sensations (e.g., vision) on the same side.

Hysterical pain may involve any part of the body. Regional pain syndromes are common in hysteria, e.g., headache (generalized or localized), abdominal pain, facial pain, and chronic low back pain. Typically, a patient with hysterical pain is unable to give a clear description of the character of his/her symptoms of pain, often resorting to metaphorical description (electric shock like head pain splitting the skull or facial pain resembling the sensation of the flesh being cut by a butcher's knife) without providing concise information regarding the localization, frequency, relieving or aggravating factors of the pain. Hysterical pain is generally persistent and unrelieving, although it seldom interferes with night time sleep. Cutaneous sensory loss is frequently associated over the area of pain. A number of patients may become analgesic abusers and, in some instances, may experience drug-induced organic pain symptoms (e.g., codeine-induced headache). Sometimes, the painful episodes are described as intermittent and neuralgic, although a careful history in these cases will often identify features that would be inconsistent with organic neuralgic syndromes such as trigeminal neuralgia. Before considering any painful syndrome as functional, it is important to exclude any other diseases that could account for the pain syndrome. Neuralgic facial pain, even if atypical, requires the exclusion of underlying demyelination and structural pathology in the posterior fossa. For other painful symptoms, one must exclude the pain due to tumor and infection (e.g., abscess). Abdominal migraine, familial Mediterranean fever, and acute intermittent

porphyria must be considered in the differential diagnosis of any unexplained, intermittent abdominal pain in a young patient.

Special Senses

Hysterical blindness is often sudden in onset. It affects one or both the eyes with or without other pseudoneurologic symptoms such as motor weakness. Clinically, a patient with hysterical blindness demonstrates normal pupillary responses to light and preserved optokinetic nystagmus. Cortical blindness, posterior leukoencephalopathy, and variants of Balint's syndrome are the main diagnostic considerations; hence, MRI scan is necessary. Presence of normal pattern reversal visual evoked response (VER) from the affected eye confirms the normal integrity of the nerve conduction in the visual pathway. The functional nature of the symptom may also be supported by a positive menace reflex when the patient blinks in response to a sudden visual threat to the affected eye. When left unattended, a patient with hysterical blindness is often able to reach for an object and does not bump into an obstacle on the way. The isolated occurrence of convergence eye spasm with pupillary constriction (*spasm of the near triad*) is usually hysterical.[22] In this condition, both eyes suddenly converge on attempted fixation straight ahead. True (organic) causes of spasm of convergence are extremely rare; Guildford and others[17] found only 14 cases in the literature. More recently, paroxysmal convergence spasm was reported in a single case of multiple sclerosis where MRI revealed a brainstem demyelinating plaque in the region of the medial longitudinal fasciculus.[29]

Hysterical loss of smell (anosmia) can be distinguished easily from the disease of the olfactory nerve by applying substances with pungent or irritative odor (liquor ammonia) to the nostrils. These substances stimulate the trigeminal nerve, and a patient with organic anosmia would still be able to identify the smell whereas a patient with hysterical anosmia would deny this, despite having reflex tears from the irritation. Hysterical deafness is best evaluated by brainstem auditory evoked response (BAER), which reveals normal cochlear nerve potential and retrocochlear conduction in functional deafness.

Paroxysmal Events

Hysterical seizures are the most common functional paroxysmal events. Not all non-epileptic events resembling epileptic attacks are hysterical seizures, since a proportion of the non-epileptic attack disorders may be associated with depression, psychoses, hyperventilation, or malingering. Dissociative convulsion is another name for the non-epileptic seizure in hysteria but is somewhat misleading because it may inappropriately imply that hysterical seizures are always dissociative. The term *pseudoseizure* is probably the best descriptor of the hysterical seizures. Witnessing the event may offer clues because hysterical pseudoseizures may have several features that are seldom seen in epileptic seizures. Unusual features suggesting possible pseudoseizure include lack of aura, initiating cry, retention of consciousness during an attack associated with motor movements affecting both sides of the body, presence of pelvic thrashing and flailing limb movements, side-to-side head movements, facial grimacing, body squirming, hurtful falls ("carpet burns" are rather common), hitting at furniture or a wall, striking at or resisting those trying to help, longer duration of attacks, absence of post-ictal drowsiness or confusion, initiation of the attack by suggestion, and termination by strong sensory stimulation.[1] Typically, serum creatine kinase and prolactin levels in the blood taken immediately after the attack are normal. The absence of any epileptiform changes in the EEG recorded at the time of

the attack is a reliable evidence that the attack is non-epileptic, although some partial epileptic seizures, especially those originating in the frontal lobe, may not have EEG signatures for the ictal event.[3] Interictal EEG is more difficult to interpret not only because 3% of healthy people may have "epileptiform" changes in the EEG,[11] but because epileptic and non-epileptic pseudoseizures may coexist in a given patient. A video recording of the ictal and post-ictal activity is an important diagnostic tool for pseudoseizures. Simultaneous video and EEG recording (video-telemetry) is the only neurodiagnostic technique that offers a direct correlation between the observed seizure activity ("the attack") and the ongoing brain electrical activity, allowing the physician to identify whether or not the witnessed event was epileptic.

It is important to remember that frontal lobe epilepsy, catalepsy, and rare partial seizures are occasionally misdiagnosed as "hysterical." There are techniques to provoke seizures, although their use in clinical practice may raise ethical issues. Suggestive techniques using a placebo injection or hypnosis have been successful in generating hysterical pseudoseizures, but their use without local ethical approval cannot be recommended. In addition, suggestibility of a seizure, in itself, is not a definitive diagnostic tool because both epileptic and non-epileptic seizures may be provoked using these techniques. Patients with hysterical pseudoseizures are no more suggestible than those with epileptic seizures.

Hysterical fainting attacks, like hysterical pseudoseizures, are very common. Hysterical faints are dramatic, and like pseudoseizures, these attacks usually take place in front of an audience and may be suggestible. There is no change in the color of the skin, blood pressure, or heart rate that typically accompanies vasodepressor syncope ("vasovagal attack"). Hysterical faints and pseudoseizures may occur together. One must, however, distinguish hysteric faints from orthostatic intolerance and panic attacks. Strong sensory stimulation may reverse fainting due to hysteria, but this in itself should not be used as the diagnostic marker of hysterical faints. Mass hysterical faints have been reported in regiments and school marching bands, although in some instances, these faints might have been feigned and therefore cannot be classified as hysteric by definition.

Cognition and Speech

In hysterical trances or fugues, the patient wanders about for hours or days without subsequent recollection of the event. During this period, the patient is able to carry out complex acts and may appear unaware of any difficulty. This is rare. Hysterical fugues may simulate temporal lobe epilepsy or confusional state. There is no test than can reliably distinguish hysterical fugue. Witnessing an episode of hysterical fugue is of assistance because unlike patients with temporal lobe epilepsy or confusional state, a patient in hysterical fugue retains a certain level of alertness and orientation. Patients may be able to recall the events that occurred during the episode under the influence of hypnosis, suggestion, or sedation with amobarbital (Amytal).[28] Hysterical amnesia is typically seen in a female patient brought to the hospital often found wandering about aimlessly. This must be distinguished from the cases of selective amnesia reported by males often involved in a crime and feigning amnesia for the criminal act (see Ganser syndrome below). The difficulty to recall in hysterical amnesia is always anterograde. Hysterical amnesia improves gradually after periods of rest, encouragement, and emotional support.[34] Dissociative amnesia (loss of memory with ability to recall important personal information) occurs after head injury. Rare cases of temporal lobe epilepsy have been attributed to dissociative fugue in which a person may wander away from home. Epileptic events and

transient global amnesia are the differential diagnoses of true hysteric amnesia. An EEG study may be helpful (but not diagnostic) in epileptic amnesia and fugue states. *Ganser syndrome* is not hysterical and is best considered as an example of malingering. Characterised by amnesia, disturbance of consciousness, and hallucinations, patients with Ganser syndrome typically offer approximate answers (*Vorbeireden*) to the questions asked and behave in an insane or demented fashion. This syndrome was first reported by Ganser in a small group of prisoners and has since been reported in other convicts.[34]

Progressive loss of memory and other cognitive skills (dementia) as symptoms of hysteria often occur in the setting of an ongoing personal or family stress in the sufferer. There may be a family history of dementia (Alzheimer's disease) in some of these cases, and an underlying depression may be contributory (pseudodementia of depression). An important clue for the diagnosis of hysterical dementia is a complete loss of memory for all previous life experiences in patients who are still able to take care of themselves. While it is usually a family member or a friend (rather than the patient) who notices the first symptoms of organic dementia, it is always the patient who complains of loss of memory in pseudodementia and hysterical dementia. In very severe degrees of hysterical dementia, the patient may regress to a helpless infantile state, losing the ability to walk, talk, or eat with fingers; make inarticulate noise in response to conversational speech; and become incontinent. Known as hysterical puerilism or hysterical infantilism, this condition is exceptionally rare, and when it occurs, one will easily be able to find evidence of gain and a history of unstable premorbid personality in the absence of objective neurological signs of organic pathology (e.g., released reflexes).

Hysterical dysarthria is uncommon and should not be diagnosed until organic causes (especially cerebellar and neuromuscular) have been excluded with appropriate tests. Hysterical aphonia, in contrast, is common.[4] Patients will require similar assessment as in hysterical dysphagia. Loss of language function as a manifestation of hysteria has been known in clinical practice. In these cases, there is a dissociation between the ability to speak and the ability to write, i.e., the patient is usually able to write normally but is unable to speak, which would be unusual with cortical aphasias. One must, however, be extremely cautious before attributing loss of language function to hysteria and exclude intra- and interhemispheric dissociation (disconnection) syndromes (e.g., pure word deafness, alexia without agraphia). Cerebral neuroimaging (MRI) is essential in the evaluation of any speech and language disorder.

Autonomic and Neuroendocrine

Patients with hysterical fever consider themselves to be pyrexial, yet most of them will have a normal temperature when their oral or axillary temperature is measured with a thermometer. Although occasional cases of psychogenic fever have been documented in the literature, patients with hysterical fever are essentially apyrexial and need to be reassured that their body temperature is normal. Hysterical vomiting is often combined with hysterical pain of the abdomen and is not uncommonly encountered in young adults and women. Vomiting may be intermittent or, more typically, occur after meals. One must distinguish hysterical vomiting from vomiting associated with bulimia and anorexia nervosa, hypothalamic tumor, hyperemesis of pregnancy, hepatobiliary disease, and, rarely, from vomiting due to ketoacidosis.

Hysterical asthenia must be distinguished from post-viral chronic fatigue syndrome. Typically, hysterical asthenia is associated with generalized body and limb

weakness that is seldom seen in patients with chronic fatigue syndrome. In addition, the severity of fatigue in hysterical asthenia is unvarying and is improved by rest or sleep, whereas fatigue in chronic fatigue syndrome usually fluctuates in its severity and is unrelieved by rest or sleep. Hysterical pregnancy (pseudopregnancy) and lactation have also been reported in the medical literature; women with hysterical pregnancy report fetal movements and uterine contractions.[32] External appearance of hysterical pregnancy may be very similar to true pregnancy with the exception that the umbilicus in hysterical pregnancy is inverted, i.e., remains below the level of the enlarged abdomen. Examination will reveal the absence of fetal heart sounds and body parts in pseudopregnancy that can be further confirmed by an abdominal ultrasound.

INVESTIGATIONS

These have already been discussed above. Briefly, the choice of investigation reflects the likely organic cause that shares similar features as the hysterical pseudoneurologic syndrome. Thus, a patient with suspected hysteria will require a combination of appropriate imaging (CT/MRI), neurophysiology (evoked responses, EEG, and EMG), hematological (e.g., serum autoantibody screen), and biochemical investigations (e.g., for porphyria) depending on the clinical circumstances and the differential diagnosis (Table 1). It is important to emphasize that the use of provocative measures (suggestion, hypnosis, or amobarbital interview), although effective in certain situations, requires prior institutional ethical approval. There is a long list of systemic disorders that can affect the brain (Table 2), and it is worth remembering that a more generalized systemic disease process, e.g., metabolic (hypothyroidism or porphyria) and vascular (systemic lupus erythematosus), may manifest as a functional pseudoneurologic syndrome. Finally, relatively uncommon neurologic disorders may be mistaken as hysterical (e.g., Balint's syndrome, Kleine-Levin syndrome, pure word deafness, nocturnal frontal lobe epilepsy, and paroxysmal movement disorders), and one must not hesitate to seek opinion from the more experienced in unfamiliar cases.

Like any other area in clinical science, it is important to make an accurate diagnosis of hysteria. Misdiagnosis between organic and hysterical neurological syndrome may be potentially serious. Patients with hysterical neurologic syndrome may

TABLE 1. Neurodiagnostics of Hysteria

Clinical
Full neurological examination
Use of suggestion, hypnosis, and amobarbital interview*
(*may uncover symptoms)
Neuroimaging
Anatomical (CT/MRI): to exclude structural pathology, demyelination, and vascular lesion
Functional (SPECT, PET): only as research tools
Neurophysiology
EEG and video-telemetry
Evoked responses (VER and BAER)
EMG
Other tests
Blood (hemoglobin, autoantibody screen, calcium, thyroid function)
Screen for porphyria
Test for pregnancy in women
Serum copper and ceruloplasmin*
(*only for young onset movement disorders)

TABLE 2. Common Systemic Disorders Associated with Functional Symptoms

Metabolic	*Vascular*
Anemia	Systemic lupus erythematosus
Hypoglycemia	Thrombotic thrombocytopenic purpura
Acute intermittent porphyria	Rheumatic heart disease
Electrolyte imbalance	*Inflammatory*
Vitamin deficiencies (e.g., B_1, B_{12})	Sepsis
Liver, renal, pulmonary, and cardiac failure	Infective endocarditis
Wilson's disease	Malignancy
Endocrine	*Toxic*
Thyroid diseases	Drug overdose
Parathyroid diseases	Alcohol
Hypopituitarism	Neurotoxins (e.g., ciguatera fish toxin)
Adrenocortical insufficiency	

be mistakenly subjected to extensive, expensive, and unwarranted investigations with invasive tests or multiple specialist referrals and offered potentially dangerous therapy. A good example is pseudoseizure. There have been many instances in which patients with pseudoseizures have been treated with multiple anti-epileptic drug therapy and at times anesthetized and ventilated on a mistaken diagnosis of convulsive status epilepticus. The author had the experience of being called to see a pregnant patient with respiratory depression after large doses of intravenous diazepam were given without success to treat her pseudostatus epilepticus. Hysterical syndromes (pseudoseizures, pain, and neuralgia) rapidly become refractory to the standard medical therapy that is generally successful in the vast majority of comparable organic syndromes. On the other hand, it is all too easy to dismiss patients who do not conform to the physician's expectations for organic disease as hysterical. Hysteria was the favorite diagnosis in a patient, usually a woman, displaying unfamiliar symptoms and having normal laboratory test results. Previous follow-up studies at the academic centers of patients diagnosed as hysterics revealed a staggering misdiagnosis rate between 20 and 40%.[13,26,30] The most common errors were failure to identify diseases like systemic lupus erythematosus, epileptic partial seizures, and neurological diseases such as demyelination. The rate of misdiagnosis, however, has fallen sharply with the use of appropriate technology and a careful clinical assessment. A recent follow-up study of 56 patients with conversion disorder (exclusive of pseudoseizures) identified only two cases with a missed neurological diagnosis (multiple sclerosis and cerebrovascular lesion), suggesting a current misdiagnosis rate of 3.6% (2/56)[7] that is very similar to the proportion of patients (4%) with conversion disorder seeking neurology opinion.[27]

PSYCHOLOGICAL ASSESSMENT

Once a diagnosis of hysteria is established, it is useful to have a psychological assessment of the patient. The psychological interview may uncover important conflicts or needs that might have been suppressed. Interpretation of psychological stressors precipitating the hysterical pseudoneurologic syndrome may be complicated because acute psychological stress can also precipitate organic neurological syndromes like multiple sclerosis.[14] History of sexual or physical abuse may be present in patients and should be enquired about with due sensitivity and care. However, it would be incorrect to assume that the neurologic complaints developing after a major psychological trauma is invariably functional. As an example, epileptic seizures may arise from brain injury sustained as a result of childhood physical abuse.

Medical literature is replete with the suggestions that the hysterical patients are characterized by a distinctive personality. Many writers have commented on the rather youthful, coquettish, or seductive manner of the patients and their constant demands for attention and approval. Others have equated hysteria with susceptibility to suggestions and hypnosis. Indeed, older texts never fail to mention the "stigmata of hysteria" characterized by corneal anesthesia, absence of gag reflex, and circumscribed areas of tenderness over the vertex, chest, and abdomen often suggestible by the examiner. Another common misperception is that patients with hysteria have "*la belle indifference*." The term *la belle indifference* was coined by Janet, one of Charcot's pupils, to describe a lack of concern regarding important bodily dysfunction such as hemiparesis.[21] However, while every clinician would recall seeing a few patients with these characteristics, overall, these features no longer apply to the patient population with hysteria. Indeed, *la belle indifference* is often absent during the presentation of a hysteric syndrome, and conversely, it may occur in seriously ill patients with organic diseases who may be stoical. Hemineglect is probably the best clinical example of *la belle indifference*, and it was Anton who drew attention to this syndrome of parietal lobe lesion in which a patient with contralateral hemiparesis may remain indifferent to the motor weakness (Anton-Babinski syndrome).[1] More consistently, primary or secondary gain may underlie the presenting symptoms in hysterical patients. It is also not uncommon that there may be another member of the family or a friend who might have physical symptoms similar to the one in a hysteric patient. Rarely, identical hysterical pseudoneurologic syndrome may develop simultaneously or in tandem among two close members of the same family (spouses or siblings[8]); this phenomenon is often referred to as dissociative *folie a deux*.

NEUROANATOMIC SUBSTRATE OF HYSTERIA: CONTRIBUTION FROM FUNCTIONAL NEUROIMAGING

Neurological symptoms are common in dissociation, conversion, and somatization disorders that constitute the spectrum of hysteric pseudoneurologic syndromes. In the absence of any objective, organic pathology, pseudoneurologic symptoms are presumed to be psychogenic or "in the mind." Since the neuroscientists still do not know what precisely constitutes the mind, it has been difficult to develop a neuroanatomic paradigm for the functional disorders. Words like "dissociation," "conversion," and "somatization" are less meaningful from a neurologist's perspective in the absence of any possible neuroanatomic template corresponding to these terms.

Historically, the term *conversion* was used by Freud and his students to describe a process by which the psychic energy of an unresolved mental conflict in a patient was transformed (*conversion reaction*) into a specific pseudoneurological syndrome (hemiparesis, blindness, etc.).[15] This is still a useful concept, although Freud's reasoning for the underlying mental conflict (unfulfilled sexual urges in women) is far less convincing. In this paradigm, hysteric symptoms appear when psychological stressors influence the function of a specific neurological network either negatively (e.g., hysterical paralysis and blindness) or positively (hysterical tremors and pseudoseizures). Advances in the functional brain imaging in the past few years (PET and SPECT) have shed some light on this issue. It has now been shown conclusively that hysteric motor paralysis is different from the feigned loss of movement (factitious or malingering disorder). In a PET study, two men with hysterical motor weakness of their left arm were compared with two other individuals with feigned loss of left finger movements and six normal controls. Comparing the

brain activation during movement of the left hand relative to the resting state, the authors found that hysteric patients exhibited relative hypoactivity of their *ipsilateral* (left) dorsolateral prefrontal cortex; in contrast, feigners exhibited hypofunction of their contralateral (right) prefrontal cortex, as would be expected.[31] In a single case study of a patient with long standing leg paralysis imaged with PET, two distinct areas of the contralateral prefrontal cortex (anterior cingulate and orbitofrontal cortex) were activated,[19] a pattern that the workers were able to reproduce in a susceptible individual by inducing hypnotic paralysis.[18] More recently, patients with unilateral hysterical sensorimotor loss were studied, both during their initial presentation and after their recovery, by using cerebral SPECT scan for regional blood flow changes. The authors of this study found a consistent decrease in the regional cerebral blood flow in the thalamus and basal ganglia contralateral to the deficit with resolution of these changes after recovery.[35]

Taken together, the results of the functional neuroimaging[18,25,31,35] suggest that the neuroanatomic substrate for the motor deficit in hysteria might underlie either a failure to generate motor programs in the striato-thalamo-cortical circuits and/or a failure to execute normally generated motor programs at the higher cortical level.

However, the responsible cortical areas may not necessarily be frontal in every case of hysteria. A failure of the auditory cortex has been implicated in functional hearing loss. In a single case study of a female patient with hysteric deafness using event-related potential (ERP), a reduction in the amplitude of the cortical potential was noted.[16] This observation was interpreted to suggest that the processing auditory brain area in the temporal cortex was repressed in hysterical deafness. Clinical experience also indicates that occipital cortical suppression is the likely explanation for hysterical blindness. Indeed, cortical neurologic syndromes like Balint's are often mistaken as hysteria. However, cortical hypoactivation may only possibly offer explanations for the negative symptoms of hysteria. The complexity increases if one were to explain the "positive" neurological symptoms (e.g., hysterical tremors) on the basis of a specific, dysfunctional neural network.

It seems unlikely that we would be able to rapidly develop an acceptable neuroanatomic model for hysteria, although the results of recent research using functional neuroimaging appear both interesting and promising. Nevertheless, a neurobiologic role of the cerebral cortex in hysterical symptoms has been consistently supported by the observation that hysterical pseudoparalysis is more frequent on left-sided limbs and may be facilitated by co-existing brain injury.[11] A possible role of the right hemisphere in hysterical pseudoparalysis has been postulated.[33] In this respect, patients with Anton-Babinski syndrome of hemineglect due to parietal lobe lesion provide an intriguing contrast. According to Hécaen's statistics,[20] unilateral hemineglect (asomatognosia) occurs predominantly with left-sided motor weakness, being seven times more frequent in right (non-dominant) parietal lesions than left parietal ones. This is strikingly similar to the observation that left-sided pseudoparalysis and hemianaesthesia are more common in hysteria. More recent observations indicate that patients with right parietal lesions show not only contralateral (left-sided) neglect, but also a degree of ipsilateral (right-sided) neglect, suggesting that in terms of spatial attention, right parietal lobe is truly dominant.[36] As already noted, *la belle indifference* is comparable to the sensory inattention and hemineglect observed in patients with the parietal lobe pathology. One of Janet's patients[21] was brought to his attention because of her profound *belle indifference* to a long standing left-sided hysterical hemianesthesia after she had seen a physician for a small area

of sensory loss on her right hand following a traumatic right median nerve injury. Hemineglect (asomatognosia) is the consequence of a neural network failure affected by the sensorimotor deficit, loss of the stored engrams of the body scheme, as well as a conceptual negation of paralysis resulting from the parietal lobe lesion. Hysterical syndromes may be considered to represent a reverse model of asomatognosia in which body perception may be heightened and stored engrams of body image become dissociated with the conceptual negation of normal body function. Under normal circumstances, the human parietal lobe is responsible for the supramodal integration of ascending thalamic input. Additionally, the parietal lobe receives large association fiber connections with the frontal, occipital, and temporal lobes of the same hemisphere and, through the middle part of the corpus callosum, with corresponding parts of the opposite hemisphere. Thus, one may hypothesize that a functional failure of this parietal integrator (preferentially right-sided) in hysteria could dissociate the cortical[25,31] and the subcortical[35] areas with consequent downstream metabolic changes (hypoactivity) that have already been identified in the functional neuroimaging data. It is possible to test this hypothesis by $^{1}H_{1}$-proton magnetic resonance spectroscopy that is likely to become a valuable neuroimaging tool for future research in this area.

It also remains unexplained why hysteria is at least three times more common in women and has an increased prevalence among populations with low socioeconomic and educational status (e.g., rural environment).[10] However, neuroimaging of the literate versus illiterate brain has offered some insight regarding the role of social forces like education in modulating the adult brain function. In an elegant PET study of normal Portuguese women, it was found that the illiterate women had inferior ability in the language processing task (repetition of nonsense words) associated with the failure to activate the anterior cingulate cortex despite having equal intelligence and social function as the literate women.[5] This observation confirmed the view that learning to read and write during childhood may fundamentally change the functional neuroanatomy of the brain. It is thus conceivable that a psychologic stressor may precipitate a hysteric syndrome only if the underlying brain was already "primed" for such an event, perhaps as a result of early life events, abuse, or the effect of social forces like education and economic affluence.

CONCLUSION

Professional neurological examination and the selective use of appropriate investigations (see Table 1) are the key requisites for the assessment of pseudoneurologic syndromes. Currently, functional neuroimaging and ERP should be regarded as research tools in evaluating hysteria, and their application for routine diagnosis cannot be advised. However, functional neuroimaging has the potential to reduce the morbidity in hysteria by improving the understanding of the illness.[19]

Human behavior is the outcome of a complex interplay between brain structures and social forces; hysteria is one of the best examples. Freud[15] wrote that "the hysteric acts in his paralysis and other manifestations as if anatomy were non-existent" (i.e., *neglect*) "or as if he had no knowledge of it" (i.e., *denial*). Both neglect and denial of body schemes are highly specialized, supramodal functions of the human parietal lobe. A possible role of the right (non-dominant) parietal integrator in the hysterical manifestations will provoke researchers. Critchley had remarked that to establish a formula for normal parietal lobe function will prove to be "a vain and meaningless pursuit."[8] Only time will tell if this remark may also apply to our journey to understand hysteria that has only just begun.

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