

DEFINING AND DIAGNOSING INVOLUNTARY EMOTIONAL EXPRESSION DISORDER

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FOCUS POINTS

- Involuntary episodes of emotional expression (crying, laughing, or related emotional displays) can accompany a variety of neurodegenerative diseases and neurological injuries.
- An extensive literature is devoted to disorders of emotional expression, but inconsistent use of terminology has led to debate and confusion.
- The authors propose the term involuntary emotional expression disorder (IEED) as a medically accurate and unifying term, as well as diagnostic criteria for this disorder.
- When arriving at a differential diagnosis of IEED, the most important step is distinguishing crying as part of IEED versus crying in the context of a depressive episode.
- The proposed IEED terminology and diagnostic criteria will promote treatment and facilitate research by providing a clear definition for clinicians and investigators.

ABSTRACT

Uncontrollable episodes of emotional expression occur in a variety of neurological conditions. This emotional disinhibition syndrome is characterized by episodes of crying or laughing that are unrelated to or out of proportion to the eliciting stimulus. This syndrome is common among patients with amyotrophic lateral sclerosis, multiple sclerosis, stroke, and traumatic brain injury and a variety of terms and definitions have been used to describe it. The confusing nomenclature has been a barrier to understanding, diagnosis, and treatment of this disorder. The authors propose a unifying term, involuntary emotional expression disorder (IEED), and provide diagnostic criteria for this disorder.

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Introduction

Involuntary displays of emotion occur in numerous neurological disorders. A classic example of this phenomenon is pseudobulbar palsy resulting from bilateral damage to the descending pyramidal tracts, producing cranial nerve dysfunction as well as uncontrollable crying or laughing that is unrelated to or out of proportion to the patient’s underlying mood. Often referred to as pseudobulbar affect (PBA),^{1,2} this syndrome occurs in amyotrophic lateral sclerosis (ALS),³⁻⁵ multiple sclerosis (MS),⁶⁻⁹ stroke,¹⁰⁻¹⁶ traumatic brain injury (TBI),¹⁷⁻¹⁹ Alzheimer’s disease,^{20,21} Parkinson’s disease,²² and brain tumors,²³ among other disorders. Recent evidence suggests a link between the core symptoms of crying and laughing and a broader class of emotional disinhibition symptoms.²⁴ The authors of this report suggest the term involuntary emotional expression disorder (IEED) as an inclusive terminology. IEED is relevant to patients with neurological disease or injury, resulting in episodic and involuntary bouts of uncontrollable emotional expression. Diagnostic criteria to assist in recognition, diagnosis, and management of this debilitating condition are proposed.

Historical Background

An extensive literature is devoted to disorders of emotional expression, but inconsistent use of terminology has led to debate and confusion.^{8-11,25,26} The clinical syndromes related to IEED have been described using many different terms including PBA, pathological laughing and crying (PLC), pathological weeping, emotional or affective lability, emotionalism, emotional incontinence, pathologic emotionality or affect, and emotional dyscontrol (Table 1).^{2,8,9,26,27} These terms are sometimes used to denote a specific or distinct condition and at other times are used as synonyms.^{1,9} PLC has been defined as crying or laughing that is incongruous to the eliciting stimulus,^{9,25} or as out of proportion to any provocation.^{10,12,28,29} PBA is also a term with multiple definitions. One definition for PBA, affective disinhibition similar to that seen as a result of pseudobulbar palsy, is derived from its theoretical underlying neuropathologic state.²⁶ PBA has been alternatively defined as either mood-incongruent or mood-congruent but out of proportion to the circumstances.³⁰⁻³² Some definitions of PBA are more inclusive and have identified the syndrome with affective and emotional lability.²

A distinction often has been drawn between mood (defined as an internal emotional state) and affect (defined as the outward physical manifestations of emotion) in the literature devoted to PBA. This use is confounded, however, by labeling some mood disorders (eg, bipolar illness, major depression) as affective disorders.³³⁻³⁶ The application of this distinction has been inconsistent and a cause of further confusion.

Ideally, terminology for a condition assists in recognition, diagnosis, and management; is not pejorative

to patients (such as “emotional incontinence”); and is medically accurate and phenomenologically descriptive. A useful terminology communicates essential elements of the syndrome. None of the existing terms for the syndrome described have fulfilled all these criteria, and only the seldom-used *International Classification of Diseases*, 9th Edition code 301.3,³⁷ which includes emotional instability (excessive) and pathological emotionality, shares the referent of this syndrome. The term IEED fulfills most of these desirable elements; is consistent with nomenclature conventions such as those of the *Diagnostic and Statistical Manual of Mental Disorders*, Fourth Edition;³⁸ and avoids the confusion associated with the distinction between mood and affect. IEED can provide a basis for diagnostic coding and for reimbursement of health services.

Previous Diagnostic Criteria

The core clinical feature of IEED is involuntary outbursts of crying and/or laughing. Historically, the syndrome characterized by such outbursts has been described with variable definitional criteria. Wilson²⁸ first described the syndrome as “exaggerated, forced, involuntary, uncontrollable laughing or weeping,” and coined the term PLC. In addition, he identified three critical aspects of the emotional outbursts, including: (1) the unbalanced relationship between stimulus and response, noting that “stimuli are often inadequate and inappropriate”; (2) the disconnection between mood

TABLE 1

Historical Terms for Involuntary Emotional Expression Disorder

Affective lability
Emotionalism
Emotional dyscontrol
Emotional incontinence
Emotional lability
Excessive emotionality
Forced laughter or crying
Inappropriate hilarity
Pathological affect
Pathologic emotionality
Pathological emotionalism
Pathological laughing and crying
Pathological weeping
Pseudobulbar affect
Pseudobulbar crying

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and affect (in this case the external of mood), describing that “visible emotion does not necessarily correspond to the patient’s real feelings”; and (3) the stereotyped nature of the outbursts, discussed as an “invariability of emotional response, whatever the stimulus.”^{28,29}

Subsequently, Poeck²⁵ proposed the only previously existing diagnostic criteria for PLC. The proposed four criteria for pathological laughing and crying are: (1) response to nonspecific stimuli; (2) absence of a corresponding change in affect (here referring to internal state) or a lack of relationship between affective change or the observed expression; (3) absence of voluntary control of the extent and duration of the facial expressions; and (4) absence of corresponding change in mood lasting beyond the actual laughing or crying, and lack of relief after an event.²⁵ Poeck speculated that the mood-incongruity characteristic of PLC was due to the impact of the underlying condition on motor pathways involving the muscles of facial expression.²⁵ The Poeck criteria have been criticized for the required absence of correspondence between mood and affect.⁸

Phenomenology

The emotional outbursts that characterize IEED share a set of common features.²⁶ They are involuntary, with patients having no control over the timing or duration of outbursts. The outbursts are episodic in nature, with a return to a baseline state upon conclusion. Outbursts are stereotyped, although single individuals may have episodes of both laughing and crying. They are mood incongruent or have an intensity that is out of proportion to the stimulus.

Episodes of IEED may be accompanied by signs of pseudobulbar palsy, the neurological condition from which the term PBA was derived. These signs include hyperactive jaw and facial reflexes, and dysarthria.³⁹ Episodes of laughing and crying also can occur in the absence of signs of pseudobulbar palsy,⁴⁰ and not all patients have pyramidal tract lesions typically associated with pseudobulbar palsy.^{15,41}

Autonomic, respiratory, and vocal changes also may occur in IEED. Bulbar involvement in any disorder is likely to be accompanied by deterioration of the voice⁴² and difficulties in phonation, mastication, articulation, and respiration. Each of these symptoms potentially may be present in patients with IEED.⁴³ The presence or absence of autonomic symptoms in IEED, however, may depend on the type and stage of underlying neuropathology.

Mood changes, especially episodic anger, also may be associated with IEED. In a recent study, the presence of poststroke anger proneness was significantly correlated with IEED, but not with poststroke depression.²⁴ No relationship was detected between excessive or inappropriate crying or poststroke anger proneness and scores on the Beck Depression Inventory, suggesting that both pathological crying and episodes

of anger occur independently of depression. In a separate study, the same authors showed that patients with poststroke IEED were unable to control anger or aggression.¹⁶ They noted that the distribution of lesions was similar between patients with poststroke IEED and proneness to anger or aggression, although the small number of study subjects precluded statistical analysis of the lesion pattern.¹⁶ The Center for Neurologic Study-Lability Scale (CNS-LS), a diagnostic tool specifically developed to assess PBA/PLC symptoms, includes an auxiliary subscale to assess labile frustration, anger, and impatience.⁴⁴ The symptoms of PBA/PLC and loss of control of anger often co-occur in studies using this instrument.⁴⁴ Together, these reports suggest that some forms of episodic anger may be supportive of a diagnosis of IEED.

Neurobiology

As previously indicated, IEED can occur in many brain disorders and is not etiologically specific. Lesions causing IEED are widely distributed in the brain, but most involve the structures of a neural network involving the frontal lobes, limbic system, brainstem and cerebellum, or the interconnecting white matter tracts of this network.^{15,45-48} This neural network is thought to modulate the complex function of emotional motor expression.^{1,15}

A variety of neurotransmitters have roles in the neural circuitry of emotion, and disorders affecting transmitters may contribute to IEED. Glutamate, serotonin, acetylcholine, norepinephrine, dopamine, and sigma-system receptors all may influence IEED.^{1,41,49}

Diagnostic Criteria for IEED

Diagnostic criteria facilitate the identification and differentiation of disorders with overlapping phenomenology. They provide a standard for use in evidence-based medicine with the aim of reducing practice variance in diagnosis and treatment. They can facilitate the monitoring of response to treatment and support research by identifying a population with a shared syndrome.

The proposed diagnostic criteria for IEED are outlined in Table 2. For a diagnosis of IEED, the patient must exhibit episodes of involuntary crying, laughing, or related emotional displays that are the result of a brain injury or disease. The presence of episodes represents a change from the patient’s emotional behavior as it was prior to disease or injury onset. Episodes may be incongruent with the person’s mood, but are most often congruent with mood but exaggerated. Accordingly, they appear unrelated to or in excess of the eliciting stimulus. Although the number of episodes experienced by the patient may vary greatly, the presence of episodes usually produces clinically significant distress or impairment in social or occupational functioning. The symptoms are

not the result of another neurological or psychiatric disorder (eg, gelastic or dacrytic epilepsy, facial dystonia, facial or vocal tics, facial dyskinesias, mania, depression, panic disorder, psychosis, personality disorders), or the physiological effect of a substance.

In addition, a number of observations can be considered supportive of a diagnosis of IEED. Patients may experience accompanying autonomic changes, as well as pseudobulbar palsy signs such as increased jaw jerk, exaggerated gag reflex, tongue weakness, dysarthria, and dysphagia. Patients also may exhibit episodic anger or a proneness to anger.

IEED can be characterized by a number of descriptive qualities that are not necessary for diagnosis. The episodes are stereotyped. They have a similar appearance

across disorders and within patients. The frequency, length, and types of episodes all may occur in a stereotyped fashion. Episodes typically are brief, lasting on the order of seconds to minutes. The frequency and severity of episodes vary among patients but are characteristically similar within individual patients.

Episodes may be seen in ALS, MS, Alzheimer's disease, vascular dementia, frontotemporal dementia, brain tumors, Parkinson's disease, progressive supranuclear palsy, stroke, TBI, and a variety of other neurologic disorders (Table 3).²⁶

Differential Diagnosis of IEED

The most critical differential diagnostic step in identifying IEED is distinguishing pathologic crying from depression (Table 4). Crying may be a sign of sadness in depression and grief syndromes, whereas the pathologic crying of IEED is in contrast to the underlying mood state or greatly in excess of the experienced mood. While patients with IEED may be depressed, the emotional expression is disproportionate to the changes in mood. The most useful distinction between IEED and depression is duration. Depression is a tonic disorder lasting weeks or months, while IEED is a phasic disorder, with episodes occurring in a brief and stereotyped fashion. Although the episodes of crying associated with both IEED and depression may be phasic, depressed patients also evidence thoughts of helplessness, hopelessness, and guilt. These are not characteristic of IEED. Sleep and appetite disturbances are features of depression and not of IEED.³⁸

TABLE 2

Proposed Diagnostic Criteria for Involuntary Emotional Expression Disorder

Necessary Elements

Episodes of involuntary or exaggerated emotional expression that result from a brain disorder; including episodes of laughing, crying, or related emotional displays

- Episodes represent a change from the person's usual emotional reactivity
- Episodes may be incongruent with the person's mood or in excess of the corresponding mood state
- Episodes are independent or in excess of any provoking stimulus

The disturbance causes clinically significant distress or impairment in social or occupational functioning

The symptoms are not better accounted for by another neurologic or psychiatric disorder (eg, gelastic or dacrytic epilepsy, facial dystonia, facial or vocal tics, facial dyskinesias, mania, depression, panic disorder, psychosis)

The symptoms are not the direct physiological effect of a substance (eg, drug of abuse or medication)

Supportive Observations

Autonomic changes (eg, flushing of face)

Pseudobulbar palsy signs (eg, increased jaw jerk, exaggerated gag reflex, tongue weakness, dysarthria, dysphagia)

Proneness to anger or episodes of anger

Descriptive Characteristics

Episodes are sudden in onset

Episodes typically are brief (seconds to minutes)

Episodes vary in severity among patients (each episode is of similar severity within individual patients)

Episodes are stereotyped. Stereotypical features include not only the appearance of episodes across disorders, but also within patients. Stereotyped attributes can include frequency, length, and types of episodes

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TABLE 3

Disorders Which Involuntary Emotional Expression Disorder can Accompany

Alzheimer's disease

Amyotrophic lateral sclerosis

Brain tumors

Frontotemporal dementia

Multiple sclerosis

Neurosyphilis

Normal pressure hydrocephalus

Olivopontocerebellar degeneration

Parkinson's disease

Progressive supranuclear palsy

Stroke

Traumatic brain injury

Vascular dementia

Wilson's disease

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IEED also must be differentiated from a few uncommon neurological disorders. Rare cases of crying or laughing in the course of seizures have been described.^{45,50,51} In these cases there are likely to be accompanying signs of epilepsy, such as impaired consciousness, during the episodes of laughing or crying. IEED must be distinguished from facial movement

disorders that may produce grimacing, such as tics and dystonia. These cases lack the other features of IEED described above.

Assessment Scales

The appropriate assessment of IEED includes evaluation of the underlying disorder and review of specific findings pertinent to IEED. Physical examination includes a screen for signs consistent with pseudobulbar palsy.

Rating scales can assist in quantifying the severity of IEED, but are not used in diagnosis. They should be applied after the presence of the syndrome has been established using the draft criteria proposed here. Although multiple scales exist,^{12,44,52,53} two have been used most extensively in IEED research. Developed by Robinson and colleagues,¹² the Pathological Laughter And Crying Scale (PLACS) is an interviewer-administered questionnaire that has been validated in stroke,¹² Alzheimer’s disease,²⁰ and TBI.¹⁹ More recently, the seven-item CNS-LS has been utilized in clinical trials for IEED.^{31,54} This self-administered scale was developed for use in ALS patients⁴⁴ and also has been validated in MS.⁵⁵ Both the PLACS and the CNS-LS rely heavily on questions regarding the extent of patient control over episodes and the appropriateness of the displays of emotion. Although administration of these scales is unnecessary for diagnosis, they offer an opportunity to evaluate severity for the purpose of documenting clinical progress, monitoring treatment efficacy, and facilitating research.

Conclusion

Episodes of involuntary emotional expression often result from disease or injury to the brain. Historically, the terminology used to describe or define this kind of emotional disinhibition has involved symptoms (PLC), affected brain regions (PBA), or broader psychological concepts (affective lability). Inconsistent usage of terms over time has led to confusion and likely has contributed to low rates of diagnosis, treatment, and study. The inclusive terminology of IEED will serve to organize a historically confusing suite of terms and to improve communication about this disorder among patients, caregivers, and physicians. Moreover, the IEED terminology and the criteria proposed for its recognition will promote treatment and will facilitate research by providing a clear definition for epidemiologic, pathologic, and therapeutic studies.

TABLE 4

Features Distinguishing Involuntary Emotional Expression Disorder from Depression

Clinical Feature	IEED	Depression
Emotional expression	Crying, laughing or both	Crying, tearfulness
Emotional experience	Independent of the expressed emotion or excessive	Mood congruent with sadness
Duration	Brief (minutes)	Tonic mood state lasting days to months
Voluntary control	None to minimal	Often can be modulated by the situation
Stereotypy	Episodes are stereotyped	Crying episodes are variable
Neurological disorder	Always present	May or may not be present
Pseudobulbar signs	May be present	No relationship
Anger	May accompany episodes	No relationship
Provocative stimulus	Usually specific to each patient and may be minimal compared to the emotional response; in some cases no provocation is apparent	Crying may be provoked by specific mood-related situations
Accompanying thought content	No specific relationships; anger and frustration commonly accompany the episodes	Worthlessness, helplessness, hopelessness, guilt, thoughts of death
Fatigue	No relationship	Common
Changes in appetite	No relationship	Anorexia common
Sleep disorder	No relationship	May be present
Diurnal variation	Not present	May be present
Anhedonia	Not present	Common

IEED=involuntary emotional expression disorder.

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