The Anatomy of a Gelastic (or Laughing) Seizure

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ABSTRACT: *Also known as laughing seizures, and usually associated with the presence of a Hypothalamic Hamartoma (HH), gelastic seizures are often mistaken for stereotypical laughter or crying. Experienced by day, or nocturnally, they are usually preceded by an aura of deep (often euphoric) feelings of déjà vu. Patients can be heard to make involuntary and out of context expressive vocalisations strikingly similar to laughing, coughing, barking, crying, or even cooing; there can even be sudden and involuntary outbursts of emotion, action or activity. Quite often gelastic seizures are followed by signs of partial (now reclassified as focal onset[1]) seizures.*

KEYWORDS: crying seizures, dacrystic; déjà vu; dyskinesia; epilepsy; epilepsy syndromes; focal emotional seizures; GE; gelastic epilepsy; gelastic seizures; gelastic syncope; GS; HH; Hypothalamic Hamartoma; laughing seizures; nystagmus; paroxysm; Pallister-Hall Syndrome; photopsia; PHS; pituitary dysfunction; seizures; temporal lobe epilepsy.

CONTENTS:

1. Definition:
GELASTIC (jell-lass-tic) - of or pertaining to laughter; from the Greek gelastikos, 'able to laugh'.

A rare syndromal related type of neurophysical event, a gelastic seizure (GS) could best be described as a paroxysm; i.e., a sudden and involuntary outburst of emotion, action or activity.

Typically associated with the presence of a Hypothalamic Hamartoma (HH) - a benign, non-cancerous, non-malignant and usually not life-threatening tumour - gelastic seizures are often mistaken for unprovoked stereotypical laughter, or crying.

Gelastic seizures are one of the central diagnostic features of Pallister-Hall Syndrome (PHS) - an extremely rare pleiotropic genetic disorder and multiple-anomaly congenital syndrome.

Experienced by day, or nocturnally (at night), gelastic seizures are generally preceded by an aura (sensation) of deep (often euphoric) feelings of déjà vu (a feeling of familiarity).

Patients can be heard to make involuntary and out of context expressive vocalisations strikingly similar to laughing, coughing, barking, grunting, crying (dacrystic, i.e., stereotypical crying), or even cooing; there can even be sudden and involuntary outbursts of emotion, action or activity; quite often these are followed by signs of 'partial' ('focal') seizures.

When observed, patients may look like they are smirking, giggling, laughing, crying, mumbling, disorientated, or in a state of altered awareness.

2. Seizure Classification:
The International League Against Epilepsy has reclassified gelastic and dacrystic seizures as ‘focal emotional’, i.e. “this seizure type begins with spontaneous fear, anxiety, or less often joy. There may be involuntary laughing or crying, each of which might or might not be accompanied by a subjective emotion.”[1, 2]

3. General:
The medical designation 'seizure' is a misnomer and more indicative of grand mal and petit mal epilepsy where patients drop, fall and writhe.

As well, the connotation that the patient laughs or cries per se is misleading.

But regardless of which is the more applicable designation, 'seizure' or 'paroxysm', the severity, impact and implications of the episode are in no way diminished.

During a gelastic seizure, a patient can be heard to make involuntary and out of context expressive vocalisations strikingly similar to laughing, coughing, barking, crying, or even cooing.

For patients, the outward and tell-tale emotional signs of the gelastic seizure are the result of the abdomen rapidly expanding and contracting; i.e., the diaphragm oscillates - the abdomen vibrates – nausea ensues.

Quite often those witnessing and/or affected by gelastic seizures exhibit, experience, describe and report instances of:

> Abdominal (belly or stomach) pain or discomfort
> Abnormal muscle contraction such as eyes moving rapidly from side-to-side (nystagmus)
> Abnormal sensations such as numbness or tingling
> Attention deficit hyperactivity disorder (ADHD)
> Automatisms (fidgeting, lip-smacking, mumbling)
> Autistic behaviour
> Behavioural issues or problems
> Blackouts
> Convulsions (dry heaving)
> Crying (spontaneous without direct cause)
> Déjà vu (a feeling of familiarity)
> Dilated pupils
> EHS (Exploding Head Syndrome)
> Fidgeting of the hands
> Hallucinations often associated with the aura / déjà vu ("I've been here, or done this before") phase, i.e. seeing, smelling and hearing things not there - a state of altered awareness
> Headaches and migraines
> Head, or body rocking involuntarily from side-to-side (dyskinesia)
> Inattentiveness
> Insomnia (the inability to, or return to sleep)
> Laughter (uncontrollable and sometimes described as empty, or hollow)
> Learning difficulties
> Lip-smacking
> Mood swings
> Mumbling
> Nausea (feeling sick)
> Partial ('focal') seizures
> Photopsia (the presence of flashes of light, or floaters in the vision)
> Rapid heartbeat and pulse
> Retching (dry heaving)
> Staring spells
> Stomach ache
> Sweating
> Visual disturbances
> Vomiting

Often mistaken for stereotypical laughter or crying, gelastic seizures can be sporadic and experienced by day, or nocturnally (i.e. during the night); they can occur intermittently or frequently in more severe cases.

Nocturnal gelastic seizures can appear symptomatic of, and mistaken for night terrors.

A young child whom experiences a gelastic seizure nocturnally may associate his or her 'laughter' on waking with a funny dream; e.g., being in a circus - whereas a baby will gurgle, fret, or coo.

Young children will often be observed involuntarily rocking their head, or themselves side-to-side.

During the progression of a gelastic seizure a person might appear to be in a euphoric state where one is said to experience a sense of extreme well-being, delight, bliss, or mirth; but nothing could be further from the truth.

Gelastic seizures can be very intense and appear very violent when at their worst!

Parents are usually distraught and find it particularly difficult to cope when their children are affected as they feel totally helpless and useless.

4. Sequence of a Gelastic Seizure:

The phases that characterize the sequence of a gelastic seizure are, once experienced, usually quite consistent and predictable.

Where young children are concerned, parents and those closest to the child will become more able to distinguish the paroxysm from normal emotional and responsive behaviour.

Usually the patient might experience a sensation of fullness behind the nose or eyes, or see flashing lights (a condition known as photopsia) - the aura - which is indicative of the impending onset of the gelastic seizure. It is at this time that some patients describe feeling "funny in the head or stomach."

A patient whom experiences, and has learnt to identify the onset (aura) of a gelastic seizure, will usually (where possible) remove him or herself abruptly from a group situation for fear of embarrassment, or consequences.

Often coupled with the déjà vu phase, young children have been known to run (seemingly for no apparent reason) to a consistent and familiar location where they feel safe - seeking the comfort of a parent or a favourite toy.

Between the initial aural sensation and the onset of the seizure itself, there is usually a period of between 5 to 15 seconds during which time the patient may be seen to rub his or her nose, squint their eyes, take a big deep breath, followed by a number of rapid short breaths.

As the seizure progresses the patient, still fully conscious (unless asleep), experiences deep (often euphoric) feelings of déjà vu where he or she senses and feels an intense awareness of familiarity with his or her surroundings.

During the déjà vu phase, which can last from about 15 seconds, up to a full minute or more, the patient can experience and feel a sensation akin to jumping on a trampoline – vertigo (light-headedness, spinning, or whirling).

The patient can be observed lip-smacking, appear dazed and become confused with his or her surroundings, feel as if he or she is somewhere else at another time, and may wander about in a bewildered state.

Observers might notice the patient's pupils are fixed and dilated; eyes can be involuntarily moving from side-to-side (a condition known as nystagmus).

It is at this time that the oscillating (vibration) of the patient's abdomen will result in the involuntary emission of any one or more of those tell-tale sounds characteristically associated with a gelastic seizure: laugh, cough, bark, cry or even a coo.

As the déjà vu phase culminates the patient will usually be compelled to lie, sit down, or in some cases just collapse.

Where a gelastic seizure occurs nocturnally (during the night), the patient may awaken from his or her sleep with an acute stomach ache similar to that suffered during bouts of diarrhoea; he or she may not even remember experiencing the paroxysm itself, but the consequences of it are set in motion.

When the patient experiences a gelastic seizure during the day, he or she can experience symptoms of nausea, an acute stomach ache, retching (dry heaving), convulse, or in extreme cases vomit gastric fluids.

After the gelastic seizure has run its course the patient will usually be quite exhausted and drowsy, with recovery, depending on the severity of the paroxysm, taking up to an hour or more.

Sometimes, patients can lapse into a state of altered awareness or unconsciousness.

Gelastic seizures recurring in quick succession can be quite violent, daunting, frightening and traumatic to the patient, and those around them, resulting in elevated adrenal levels, panic attacks and heightened irritability.

The patient can appear to be in a shock like state: pale, sweaty and shivering with uncontrollable shaking and trembling.

Adults who experience a gelastic seizure can appear and act as if they are intoxicated, so it is advisable for patients to advise family and friends of their condition to avoid any panic, misunderstandings, unnecessary confrontations, embarrassment, or legal consequences.

Patients who cannot control their gelastic seizures should not drive, operate machinery, swim, or be in a position where he or she is responsible for the safety of others.

Where adults are concerned, it is advisable for patients to carry a medical card with them outlining their condition.

5. Prevalence:
Beginning at any age, gelastic seizures are a rare form of epilepsy that statistically affects boys more than girls.

6. Diagnosis:
The evaluation and diagnosis of gelastic seizures in infants and young children particularly can be problematic as symptoms can be difficult to differentiate from general laughter, crying, behaviour or emotional development.

Where they occur infrequently, gelastic seizures are usually difficult to evidence in Electroencephalographic (EEG) studies.

To rule out the possibility of any diagnostic confusion with behavioural and emotional disorders it is advisable that episodes be video recorded where possible with attention paid to focussing on the patient's actions and facial expressions.

As gelastic seizures are often associated with the presence of a Hypothalamic Hamartoma (HH), medical imaging by Magnetic Resonance Imaging (MRI) is recommended; particularly when a child exhibits signs of precocious puberty.

7. Seizure Triggers:
Trauma, stress and emotional upheaval, as well as salty, or salted foods and some food additives (artificial preservatives) have been noted to trigger nausea and retching (dry heaving).

When dining out be mindful of condiments like mustards, pickles, tomato and barbeque sauces as are often used to garnish take-away foods.

8. Treatment - Medication:
Many anti-epileptic medications have side effects and the results of their use vary from person-to-person.

The claims by some that gelastic seizures cannot be controlled by medication should be ignored; it is a matter of finding which medication, dose, or combination and striking the right balance.

It should also be noted that some patients can be adversely affected when taking their medication with certain foods and drinks; particularly reactive with some seizure medications are citrus based drinks and/or foods and some fruits like mangoes.

A particularly reliable medication in the treatment of gelastic seizures is the anti-epileptic prescription drug Trileptal (Oxcarbazepine), but its use is affected by citrus-based drinks and/or foods, as well as artificial preservatives.

Other medications used in the treatment of gelastic seizures include: Carbium/Hexal/Tegretol (Carbamazepine), Epilim (Sodium Valproate), Frisium (Clobazam), Keppra (Levetiracetam), Lamictal (Lamotrigine), Topamax (Topiramate) and Vimpat (Lasocamide).

9. Self Help:
From my own experience - and I guess with age - I have discovered that as soon as I get an aura, or inkling of a gelastic seizure coming, I can stave off, or prevent it from manifesting by simply not opening my mouth and breathing gently through my nose for about 3 to 5 minutes.

Of the primary adverse reactions, one is the feeling of light-headedness at the least, and another is experiencing something akin to a drunken stupor at worst.

10. Adverse Drug Reactions:
If consumed when taking Trileptal (Oxcarbazepine), citrus based products – or foods containing same - have been known to cause adverse reactions, e.g. fruit; fruit juice; fruit cake; wedding cake.

Additionally, foods containing some food preservatives have been known to cause an effect, which could be best described as a drunken stupor.

11. Support Services:

Proudly Sponsored by MedicReady (R), the Facebook Gelastic Seizure Support Hub was established (on 13 March 2013) to encourage, and to raise and advance the public awareness of gelastic seizures by: informing and educating the public, gelastic seizure patients, their families, carers, attending specialists, medical practitioners, allied health professionals and other interested persons about this rare syndromal related neurophysical event, its causes, diagnostic features, clinical characteristics and treatment options:

Gelastic Seizure Support Hub https://www.facebook.com/Gelastic.Seizures/

Additional support can also be found through the Pallister-Hall Syndrome (PHS) Support Hub founded on 9 September 2013.

Pallister-Hall Syndrome /PHS/ Support Hub https://www.facebook.com/Pallister.Hall.Syndrome/

12. Author Biography:

From Toowoomba, Queensland, Australia, I was diagnosed with Pallister-Hall Syndrome (PHS) in June of 2001 (hypothalamic hamartoma (HH), syndactyly, polydactyly, bifid epiglottis, gelastic seizures (GS), etc.) by an Endocrinologist whilst I was an in-patient at the Princess Alexandra Hospital in Brisbane, Queensland; the diagnosis of PHS was confirmed in December of the same year by research geneticist Dr Leslie Biesecker whom I met at the National Institutes of Health (NIH): Bethesda, Maryland in the United States of America.

At 55 years of age, and contrary to the opinions of some medical specialists, I am still very much alive and well - and not a Dodo bird!

When I was finally diagnosed with Pallister-Hall Syndrome (PHS) I started to put my skills as a researcher and historian to good use in my hunt for information and knowledge. I even sold almost everything I had to attend at the NIH in the United States of America.

Never in my wildest dreams did I think I would ever be doing medical research; it was all Dutch to me as it probably is to you.

Like any person new to something I found myself stumbling in the dark in my thirst for knowledge to understand my various medical conditions.

Every time I got new materials from somewhere I found I couldn't understand the medical terminology and jargon, which to me seemed to be gibberish; that which goes in one ear and out of the other - whoosh, over the head!

Anyway, my perseverance paid off and what you see here is what I know, something some of the so-called specialists wouldn't have a clue about. In fact, some years ago a top Queensland geneticist was rattling off to my parents all about PHS, HHs and gelastic seizures (GSs) without realizing she was reading verbatim an article of mine that was on the internet at that time.

On 28 November 2019 I was granted membership of the Genetic Support Network of Victoria's (GSNV) Speakers Bureau and am now available for public speaking engagements.

I am not a doctor or medical professional, but I am happy to share what I have experienced first-hand and what I know.

13. References:

1. Long Summary: 2017 Revised Classification of Seizures. International League Against Epilepsy: p2, 20 Dec 2016.
2. Scheffer IE, et al. ILAE classification of the epilepsies: Position paper of the ILAE Commission for Classification and Terminology. *EPILEPSIA*, 58 (4): 512-521, Apr 2017.

14. Disclaimer:
The content of this article has been compiled as a public service for the benefit of patients, their families, carers, treating specialists, medical practitioners, and other interested persons in the hope that gelastic seizures, their peculiarities, effects and diagnostic features can be more widely recognized, known about and better understood; the primary objective being to alleviate the fears, the stress, the frustration and the hopelessness that is often experienced by those desperately searching for information in their pursuit of knowledge and a diagnosis.

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