Not your everyday epilepsy

The average person struggles to recognise common seizures. Here, epilepsy nurse Jane Burford sheds light on some unusual epilepsies which even specialists may not recognise.

Most people have never witnessed a seizure. A common misconception is that all seizures are tonic-clonic (‘grand mal’). Often, even complex partial seizures can appear unusual and may not be recognised as seizures.

I was recently at a large social event where someone had a complex partial seizure. No-one identified it as a seizure. Instead, an ambulance was called and the person had to spend the entire night in an accident and emergency department.

This article outlines some epilepsy types that are less known and recognised or considered rare or unusual.

Unusual seizure triggers

A seizure trigger is something that is likely to bring on a seizure. Common triggers include missing medications, physical or emotional stress, lack of sleep, poor diet and eating habits, hormonal changes in women, alcohol and drugs.

Among the most unusual and intriguing seizure disorders are the reflex epilepsies when seizures are provoked by complex actions or mental processes. Some people’s seizures only occur in response to triggers. These can include reading, writing, particular movements, decision-making, eating, tapping and touching, certain noises, immersion in hot water and even toothbrushing! The most common form of reflex epilepsy is photosensitive epilepsy, in which flickering lights and changing colours or geometric patterns can trigger seizures.

Reflex epilepsies only affect a small percentage of people with epilepsy and start at any age, with seizures mostly being generalised (although complex partial seizures may also occur). People with reflex epilepsies generally have normal development and normal findings on a neurological examination.

Reflex epilepsy is generally well controlled with low doses of medication. Of course, avoiding triggers is an effective way to prevent most of these seizures, but usually people require medication because their triggers are unavoidable in everyday life.

Unusual seizure types

Gelastic epilepsy is an epilepsy type where seizures are ‘gelastic’, from the Greek word for laughter. Very rare, it is slightly more common in boys than in girls. (Crying during seizures, called ‘dacyrstic’ seizures, also occurs).

The seizures generally start with sudden laughter, usually completely out of place. This normally lasts under one minute, followed by typical signs of a complex partial or focal seizure. These can include eyes and head moving to one side or the other, automatisms (such as lip-smacking, mumbling or fidgeting of the hands) and altered awareness. This period may last for seconds to many minutes. Sometimes older children may report a warning (aura) beforehand.

The episodes of laughter can be confused with behavioural or emotional disorders, which may delay diagnosis – particularly in young children.

Gelastic seizures are often difficult to control and rarely does seizure control last more than a few weeks or months at a time. Probably the best outcome is when a benign tumour in the hypothalamus causes the seizures. Successful surgery may improve a person’s seizure control plus any behavioural and learning problems.

Supplementary motor seizures (SMS) belong to a group of frontal lobe seizures frequently misdiagnosed as pseudoseizures, non-epileptic events or a sleep disorder. Abrupt and bizarre, they tend to occur during sleep several times a night, and often present with sudden rising of the arms in front of the body.

Many people have no alteration in consciousness or confusion afterwards. Consequently, seizures are often not the initial diagnosis. To compound the problem, no detectable electroencephalogram (EEG) changes may occur, or changes are obscured by muscle interference. Sometimes people may have secondary generalised tonic-clonic seizures after the SMS, which helps clarify diagnosis.

Anti-epileptic drugs can significantly improve these seizures.

Unusual happenings after a seizure

Todd’s Paralysis is a neurological condition with no known cause, where seizures are followed by temporary paralysis which resolves completely after 30 minutes to 36 hours. Paralysis may be partial or complete, but usually occurs on just one side of the body. Speech and vision may also be affected.

It is important to distinguish Todd’s paralysis from a stroke, which requires different treatment. There is no treatment for this condition; people must rest as comfortably as possible until the paralysis disappears. Sometimes brief paralysis occurs with seizures which is not Todd’s paralysis.

Rare epilepsy syndromes

An epilepsy syndrome is a condition which takes wide-ranging features into account such as seizure type, characteristic EEG, the expected course of the disorder, precipitating features, expected response to treatment, and genetic factors. Among the many epileptic syndromes are:

Rasmussen’s encephalitis - A rare disease usually affecting only one hemisphere of the brain. It occurs in children under 10 (more rarely in adolescents and adults), characterised by frequent and severe seizures, loss of motor skills and speech, paralysis on one side of the body (hemiparesis), brain inflammation (encephalitis), and mental deterioration. Scientists currently think Rasmussen’s encephalitis is an autoimmune disease in which
immune cells enter the brain, causing inflammation and damage.

**Angelman Syndrome** – A complex genetic disorder characterised by developmental delay, intellectual disability, severe speech impairment, hyperactivity, a short attention-span and problems with movement and balance. Most (90%) children also have recurrent seizures of different types, and a small head size (microcephaly). Children with this syndrome are often happy and excitable, and exhibit hand-flapping movements. Most have difficulty sleeping and need less sleep than usual. For people affected, intellectual disability, speech impairment, and seizures are life-long.

**Ohtahara Syndrome** – A neurological disorder characterised by epileptic seizures. This disorder affects newborns, usually within the first three months of life (most often within the first 10 days), commonly resulting from metabolic disorders or structural damage in the brain. Most infants with the disorder show significant underdevelopment of part or all of the brain. Doctors have observed that more boys are affected than girls.

**Landau Kleffner Syndrome (LKS)** – A rare, childhood neurological disorder characterised by the sudden or gradual development of the inability to understand or express language in speech, and an abnormal EEG. This disorder usually occurs between ages 5 and 7. Typically, children develop normally but then lose language skills for no apparent reason. While many have seizures, some do not. The disorder may be misdiagnosed as autism, pervasive developmental disorder, hearing impairment, learning disability, auditory/verbal processing disorder, attention deficit disorder, mental retardation, childhood schizophrenia, or emotional or behavioural problems.

**Infantile Spasms (IS)** – These seizures occur in *West Syndrome*, an epilepsy syndrome of infancy and childhood. West Syndrome is characterised by infantile spasms, developmental regression, and chaotic brain waves on EEG, typically starting at 4-8 months old. The seizures primarily consist of a sudden lurching forward of the body with stiffening of the arms and legs; some children arch their backs as they extend their arms and legs. Spasms tend to occur upon awakening or after feeding, often in clusters of up to 100 spasms at a time. Infantile spasms usually stop by age five, but may be followed by other seizure types. Many underlying disorders – such as birth injury, metabolic disorders, and genetic disorders – can give rise to spasms, making it important to identify the underlying cause. Sometimes none can be found.

**References:**
- [http://professionals.epilepsy.com/page/syndromes.html](http://professionals.epilepsy.com/page/syndromes.html)
- [http://www.fbai-epilepsy.org/Visitors/Centre/ft/syndromes.cfm](http://www.fbai-epilepsy.org/Visitors/Centre/ft/syndromes.cfm)
- [http://www.indianpediatrics.net/sep2002/sep-879-880.htm](http://www.indianpediatrics.net/sep2002/sep-879-880.htm)
- [http://www.epilepsy.org.uk/info/gelastic.html](http://www.epilepsy.org.uk/info/gelastic.html)

**More Information:**
- [http://www.epilepsy.org.au](http://www.epilepsy.org.au)
- [http://www.epilepsy.com](http://www.epilepsy.com)
- **Epilepsy Clubs and Support Groups:**

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**Case Study: What’s so funny?**

**A personal experience, by George Helon**

It first happened when I was about 5 years old; I was awakened during the night “laughing”. On and off until I was a teenager, these mirthful, yet troublesome episodes were confined to the nocturnal and private realm of sleep.

Then puberty kicked in and unbeknownst to me, my life was to be irreversibly and shockingly turned upside-down!

Out of the blue I started feeling really weird in the head from time to time. I began – much to my horror and embarrassment – to experience frequent and physically draining episodes of déjà vu and laughing.

“What! You think something is humorous? What’s so funny?” My religious education teacher would bellow.

Suddenly I was branded the class idiot because I would uncontrollably “laugh” out loud when things were serious or demanding of utmost attention. “A disruptive influence who doesn’t do what he is told…The class clown who interrupts others and talks too much…”

I avoided group activities, kept to myself and became withdrawn for fear of not only embarrassing my peers but more so myself. The downside of my self-imposed exile was that when my school years ended, I had no friends, no social networking skills and worst of all, not a clue that medically anything was really wrong with me.

With age, these seemingly random episodes of euphoria and nirvana struck more frequently with the effects becoming more physically intense and emotionally bothersome.

To cut a long story short, in 2001, 20 years after the episodes first commenced, after having a CT scan for a recurring earache, I was diagnosed with a Hypothalamic Hamaritoma – a pituitary tumour – one of the most significant features of an extremely rare disorder known as Pallister Hall Syndrome (PHS). Among the many features of this condition are Gelastic or “laughing” seizures.

**Note:** Since diagnosis, George trialled different medications and is now seizure free taking only one anti-epileptic drug. He has an aura about once a year. He feels more confident and emotionally stable and has educated himself about PHS and his seizures. This has enabled him to dedicate himself to helping and educating others about PHS. He believes education in schools and the community is crucial to improving understanding about seizures and epilepsy, particularly unusual or rare conditions.