FIT, FAINT OR FUNNY TURN – IS THIS EPILEPSY?

Donald Macgregor, Consultant Paediatrician
Lancaster Acute Hospitals NHS Trust

ALL THAT TWITCHES IS NOT EPILEPSEY

In day to day practice, the doctor in general practice, Accident and Emergency or Child Health is commonly confronted with a child who has had some sort of "funny turn" as an isolated or possibly recurring event. An important question (often left unsaid) which needs to be asked is "Is this epilepsy?" This article will aim to identify what is and is not epilepsy in children, the most helpful factors in the history and clinical examination as well as what investigations, if any, need to be considered.

IS THIS EPILEPSY?

Epilepsy is common – there are more than 80,000 children in the UK whose lives are or have been affected by epilepsy (which is much more common than cystic fibrosis or diabetes). A careful and detailed history may be all that is required for diagnosis, though this is often easier said than done. The event may have occurred at school, or elsewhere, so that the parent and child will have the information second- or third-hand. An accurate eye-witness account is invaluable. What was the child like before the event? What happened during it? What happened afterwards? Younger siblings are often able to mimic the event better than they can describe it and if the episodes are frequent then the use of a camcorder may be worthwhile. Many conditions produce episodic disturbance that mimic epilepsy (see Table 1). Even in a specialised paediatric service I noted that 54/200 consecutive referrals (from GP or consultant) with presumed diagnoses of epilepsy had other conditions. Childhood is an impressionable time and the wrong diagnosis can label or stigmatise a child for life (see Figure 1). It can also mean needless investigations and longterm ineffective treatment for some children and, conversely, inadequate treatment for those who would benefit. Thirty percent of referrals to specialist epilepsy services with "intractable seizure disorders" don't have epilepsy. The clinical history must take note of family history, pregnancy and the perinatal period, developmental milestones, school progress and previous illness or injury. Clinical examination should include head circumference and auscultation for intracranial bruits. A careful search for any of the stigmata of the neurocutaneous syndromes such as tuberous sclerosis, Sturge-Weber Syndrome or neurofibromatosis may be rewarding – the use of a Wood's light will help (see Figure 2).

NO IMPAIRMENT OF CONSCIOUSNESS	IMPAIRMENT OR LOC
tics, rhythmic motor habits	syncope
rigors	cyanotic breathholding attacks
jitteriness (newborn)	reflex anoxic seizures
daydreams	delirium
hypnagogic/sleep myoclonus	night terrors
non-epileptic (pseudoseizures)	migraine
Munchausen by proxy	narcolepsy
gastro-oesophageal reflux	cardiac dysrhythmias

Table 1 – conditions often mistaken for epilepsy

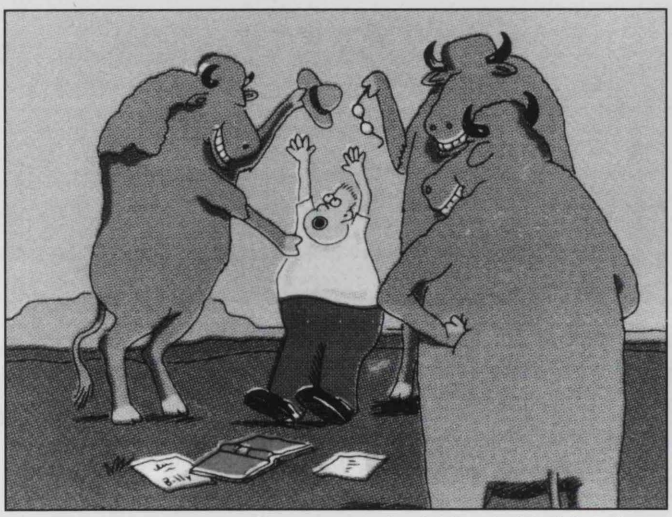

Fig. 1 – An impressionable moment in the childhood of Buffalo Bill (by Gary Larson)

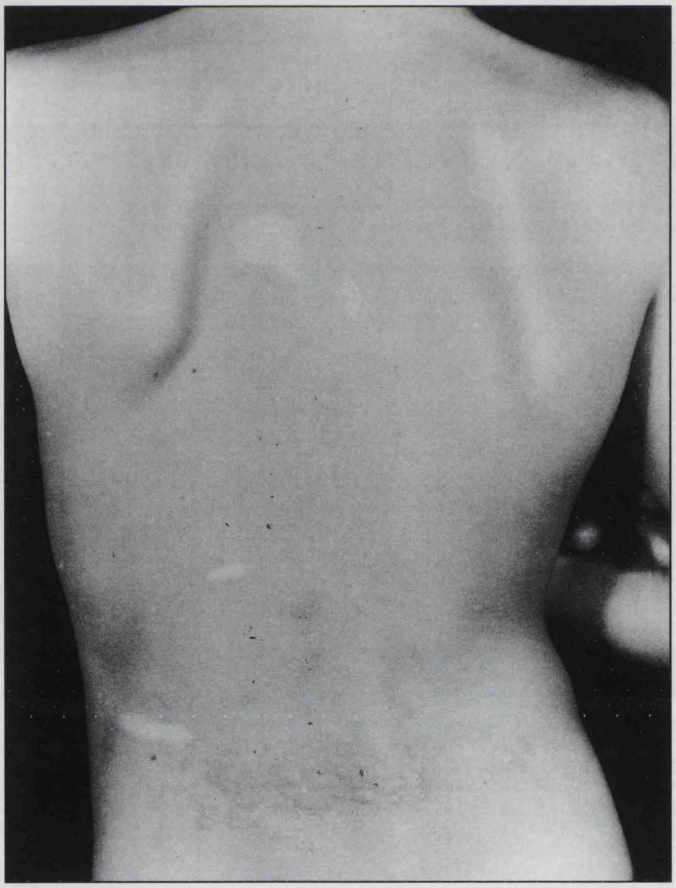

Fig. 2 – Depigmented "ash-leaf" macules and the Shagreen patch in the lumbosacral region confirm the diagnosis of tuberous sclerosis in this child with epilepsy and learning difficulties

SPECIFIC DIAGNOSES

Syncope Perhaps the commonest error is to label syncopal episodes or faints as epilepsy. The onset is typically gradual with some premonition, muscle weakness and sweating. Witnesses will note pallor. Conversely, if there is no pallor and the loss of consciousness (LOC) lasts more than a few seconds then it is probably not a simple faint.

Cyanotic breathholding attacks occur in toddlers and pre-school children. Typically, the child becomes angry or hurt then cries out and holds his breath in expiration followed by cyanosis, LOC and collapse. This can be followed by some tonic/clonic movements.

Reflex anoxic seizures/pallid syncopal attacks are more dramatic and often follow minor injury or fright. There is rapid LOC, pallor, stiffening and opisthotonus accompanied by asystole which if persisting will produce convulsive movements. If doubts persist about this diagnosis, then an episode can be reproduced by ocular compression plus EEG. Antiepileptic drugs (AED) will have no effect on this condition.

Night terrors affect most children (and even adults) at some time, but unlike most nocturnal seizures they tend to manifest in the first few hours of deep non-REM sleep. Typically, the child produces a few strangled words then cries or screams before sitting up wide-eyed and terrified. He does not recognise his carers at that time but settles back to sleep.

Migraine may be confused with epilepsy, especially basilar artery variants with brief LOC due to vertebrobasilar ischaemia and of course the two conditions may co-exist. Consistent precipitating factors with persisting headache, nausea, vomiting and prostration after the event suggest the diagnosis.

Cardiac dysrhythmias can be associated with impaired consciousness or LOC and although supraventricular tachyarrhythmias are more common, one area that has provoked much interest in recent years is the prolonged Q-T interval syndromes (with susceptibility to ventricular tachyarrhythmias). Any child who presents with "fits" following exercise should have an ECG to check for this[1].

Tics are usually random in time and constant in place with a predilection for the head and neck musculature. There is often a family history and the child can suppress the movements for a time but as one boy told me, "It's like the worst itch in the world, eventually you've just got to scratch".

Rhythmic motor habits (stereotypies) such as head banging, hand flapping or rocking are common in toddlers and also older children with multiple learning difficulties. These are gratification and/or ritualistic behaviours and may be accompanied by a vacant stare (this can lead to confusion with epilepsy).

Pseudoseizures may be fabricated by children with epilepsy, particularly in adolescent girls who achieve a secondary gain such as increased attention or, as is often the case, a lower expectation of their potential achievement by peers and parents due to their apparent disability. Distinguishing these events from genuine seizures is sometimes easy, often difficult and at times downright impossible. Ambulatory EEG monitoring (± video telemetry) may be helpful. In young adult females, these episodes can be an abreactive response to sexual abuse in early childhood and in these cases the "seizure" may have overtures of the original abuse with pelvic thrusting etc [2].

Munchausen by proxy may present as factitious epilepsy where an adult, usually the child's mother, gives a factitious history and may also fabricate clinical signs. Recurrent apnoeas, cyanotic attacks, seizures and even sudden death (suffocation) have all been described in this condition. The involved carer usually has some sort of paramedical background and there is extremely complex family psychopathology [3].

Gastroesophageal reflux can produce dystonic or opisthotonic posturing (Sandifer's Syndrome), presumably as a reflex stimulation of the spinal accessory nerve and/or preserved primitive reflexes. This is particularly common in children with tetraplegic or bulbar cerebral palsy[4].

WHAT TYPE OF SEIZURE? [5]

Seizures may be classified as partial, arising from a focal discharge, or generalised, where most of the brain is involved (partial seizures can become secondarily generalised).

GENERALISED	• absence • tonic-clonic • tonic • clonic • myoclonic • atonic
PARTIAL	• simple (no LOC) • complex (impaired or LOC)

Table 2 – Seizure type and classification[5]. This classification abandons the time-honoured terms such as grand mal and petit mal and they should no longer be used.

A **simple partial seizure** arises in a focal area of the brain usually affecting just one modality (motor, sensory, autonomic) and there is no impairment or LOC. **Complex partial seizures** cause some impairment of consciousness but can be missed or more often misinterpreted as "strange behaviour". The child may stare or appear vague, and these events can be confused with typical absence episodes (previously called petit mal) though the latter are of much shorter duration (less than 20 seconds). There are often accompanying simple automatisms such as lip smacking, chewing, picking movements or more complicated ones where clothes are divested, which can easily be misinterpreted.

Generalised seizures are classified descriptively.

Tonic seizures show stiffening of all the body musculature and are often accompanied by an expiratory cry as the child keels over like a felled tree. During a **clonic seizure** there are regular jerking limb and facial movements (about one per second). Tonic-clonic seizures were previously referred to as grand mal. **Atonic seizures** are frequent in some of the epilepsy syndromes such as Lennox-Gastaut. There is a sudden loss of all muscle tone, so that the child collapses headfirst and there is repetitive head and facial injury. Many of these children should wear protective helmets. **Typical absence seizures**, previously called petit mal, are brief (5-15 seconds) blank, *motionless* spells with rapid recovery. They can often be reproduced in the consulting room by overbreathing, in order to confirm the diagnosis. We have all experienced the sudden contractions of **myoclonic jerks** in

sleep (especially when physically exhausted). These often affect the upper limbs in juvenile myoclonic epilepsy particularly just after wakening – "the flying cornflakes syndrome"[6].

WHAT TYPE OF EPILEPSY?

Classification of the seizure type is essential in order to define the epilepsy syndrome, which may help to guide prognosis and management. These syndromes are characterised by both clinical and EEG features such as seizure type, age of onset, family history and neurodevelopmental progress in addition to the ictal or interictal EEG. A description of the epilepsy syndromes of childhood is beyond the scope of this article but particularly important ones include:

Febrile seizures occur in 4% of children between three months and five years in the absence of acute or chronic brain disorder. Most are brief generalised events with no significant sequelae. The use of domiciliary rectal Diazepam preparations can be helpful for children with frequent or prolonged seizures.

West's Syndrome is uncommon and usually occurs at between three and six months of age. Parents describe the child "switching off" and also exhibiting flexor or extensor spasms. The majority of these occur in children with an established primary brain disorder such as tuberous sclerosis or cerebral malformation. The EEG is very characteristic and can help to monitor response with therapy (Figure 3). It appears that prompt intervention with ACTH or corticosteroids leads to a better outcome.

Benign Rolandic epilepsy comprises 15-20% of all children with epilepsy and occurs predominantly in boys. There are partial tonic-clonic movements of face, lips and oropharyngeal muscles with resulting drooling and dysarthria. Most episodes occur at the sleep/wake interval and may be associated with nocturnal tonic-clonic events.

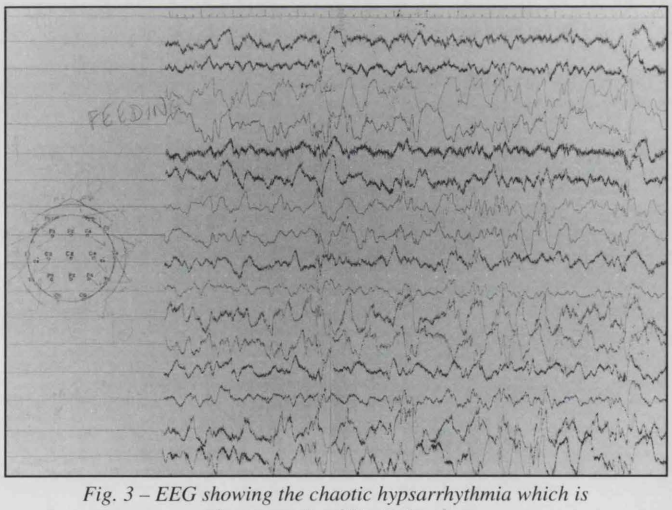

Fig. 3 – EEG showing the chaotic hypsarrhythmia which is pathognomonic of West's Syndrome

DIAGNOSIS MADE: THE NEXT STEPS

Right, so we've decided that a child has some sort of epilepsy. Should we do an EEG? Should we do a brain scan? Should we do anything else?

The EEG

This is essential once the diagnosis of epilepsy has been made, but actually rarely confirms epilepsy except perhaps in typical absence epilepsy (Figure 4) or West's Syndrome (Figure 3). It may be normal in 50% of subjects with epilepsy. Conversely, 10-15% of those with no clinical features of epilepsy will have non-specific abnormalities and some of these may even show epileptiform activity. This investigation, however, will aid classification of the particular epilepsy syndrome and may show changes that increase suspicion of a structural lesion, such as focal slow waves[7].

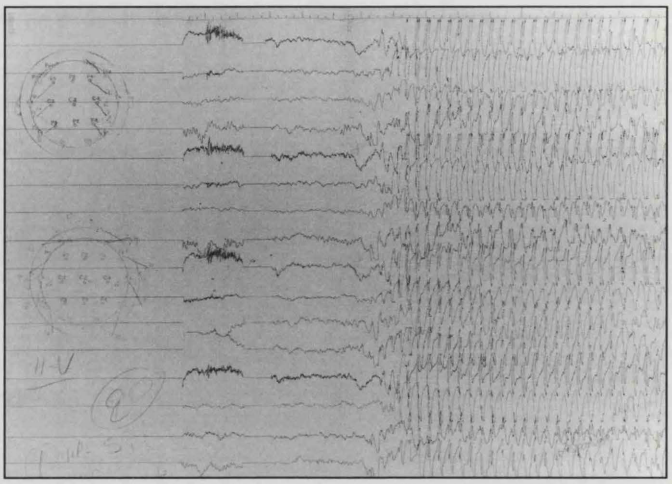

Fig. 4 – EEG showing the generalised 3 per second spike/wave complexes of typical absence epilepsy

The brain scan

Once the diagnosis of epilepsy is made, parents in particular are often anxious that there may be an underlying brain tumour and will assume the need for a brain scan. Less than 2% of children with epilepsy have a brain tumour and it is extremely rare for epilepsy to be the presenting feature of one – I have seen this once in 16 years of practice. My own practice is to request imaging in the following situations:

- partial seizures in children with neurological deficit
- partial seizures not controlled by AED
- neurocutaneous syndromes
- simple partial seizures
- focal slow waves on EEG
- anxious parents or anxious doctor!

The next question is "CT or MRI?" The greater availability of CT answers that question and this modality is perfectly adequate for most situations. The CT scan is often superior at detecting calcification (Figure 5), but MRI is capable of detecting more subtle structural lesions such as the hippocampal sclerosis of temporal lobe epilepsy or migrational and dysplastic changes.

Impact on the Child and Family

Making the diagnosis is just the start. One should never underestimate the psychosocial consequences for the child and family[8].

"Dear S,

You know you said that I could always talk to you, well I really need advice and a shoulder to cry on. I rowed with my mum because I assumed that she was having a dig at me about my epilepsy, which I know she wouldn't do. I have so many emotions and feel so confused since I found out I am epileptic. My life is a pile of shit and I really wish I was dead. I'm sorry to burden you.

N *xx*"

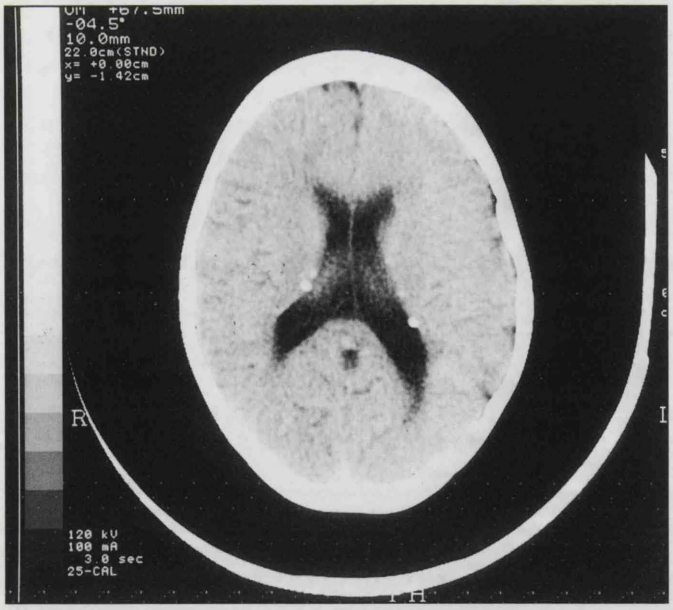

Fig. 5 – CT scan showing the characteristic calcified periventricular tubers of tuberous sclerosis

This note was brought along to my clinic by a new patient who had been told that she "just had epilepsy". Older children particularly may show low self esteem and/or social isolation. The perception of epilepsy as a life-threatening condition can provoke maladaptive responses within the family where overprotection or ineffective discipline may be an issue. Many of these issues are common to other chronic illnesses in children and need to be addressed proactively so that there is a clear understanding of the child's condition the prognosis and the rationale for treatment.

Communication is the key:

- with the family

(1) written information on the type of epilepsy and the medication

(2) management of seizure episode

(3) voluntary epilepsy associations

(4) use Nurse Specialists (if available)

- with school health (teacher, nurse, doctor)
- with the family practitioner

In my experience it can be a major task to avoid halftruths and misinformation. On one occasion a seven-year-old child had been wrongly "labelled" as having epilepsy when in fact it seemed that she was having pallid syncopal attacks (reflex anoxic seizures). An EEG with ocular compression produced prolonged asystole and so the nature of the condition was explained to the mother. A few days later there was a 'phone call from an almost hysterical headteacher who wanted this child with a life-threatening cardiac condition to be excluded from class until she had some sort of pacemaker fitted. It turned out that the mother had told the school that these episodes "were just a kind of heart attack" and that the child's heart "stopped for a few minutes"! Although most children "grow out" of their epilepsy, the issues of education and employment (including career guidance) must be considered. Ignorance, prejudice, intolerance and superstition still beset the child (and family) with epilepsy. The 1990s were designated as the "Decade of the Brain" by George Bush (then President of the United States) and endorsed by the World Federation of Neurology[9]. The diagnosis and subsequent management of epilepsy in childhood is rightly influenced by many factors unique to the growing and developing child. Thankfully, there has also been a realisation that in addition to determining the basic cellular mechanisms or the molecular genetics of epilepsy, there is a need for a more holistic approach with education and communication at the core[10].

SUMMARY: IS THIS EPILEPSY?

1 Take detailed clinical history (eye witness/video)

2 Be aware of other "faints and funny turns"

3 If in doubt don't diagnose epilepsy (review history)

4 Do not use the EEG to diagnose or to exclude epilepsy

5 Be aware of the impact on the child and family

6 Establish effective communication

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