Fits and Funny turns

A Silwal

Paediatric Neurologist

Epilepsy

- Common neurological disorder
- Affects 0.5-1% during childhood
  - 63,400 – epilepsy Δ
- Different clinical manifestations
  - >40 different epilepsy, seizure type
- Clinical diagnosis

Versus

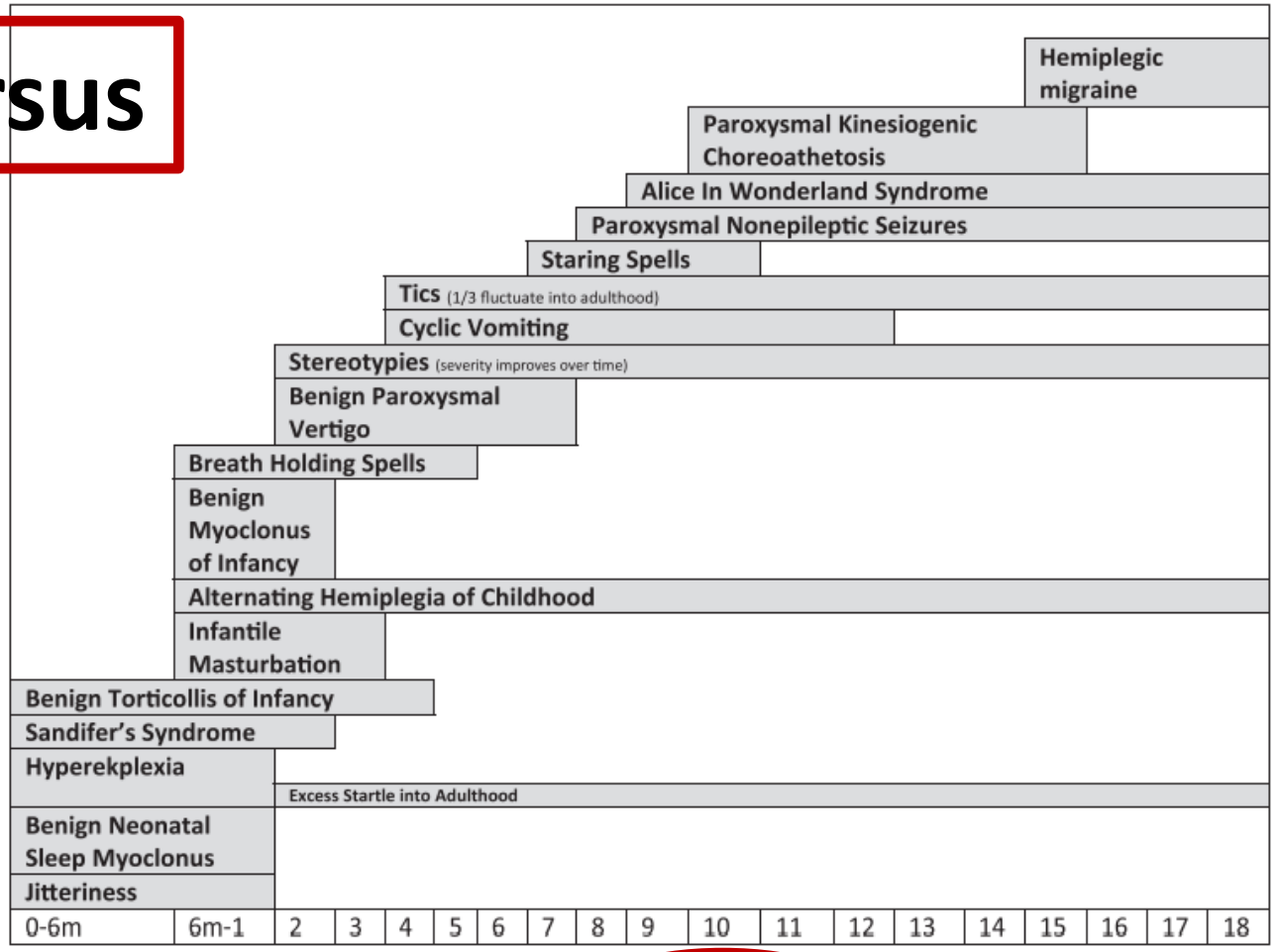


Figure 1. Age ranges (months and years) of seizure mimics, typical onset and resolution.

Stainman RS, Kossoff EH. Seizure mimics in children: An age-based approach. *Curr Probl Pediatr Adolesc Health Care*. 2020;50(12):100894. doi:10.1016/j.cppeds.2020.100894

Misdiagnosis

- Hindley et al – Epilepsy diagnosis only in 23% of children referred for fits, faints, funny turns
- Uldall et al - 39% of those referred to the Danish Epilepsy Centre did not have epilepsy
- Leicester enquiry misdiagnosed 618/1948 (31.7%) children as having epilepsy

Diagnosis in “fits, faints and funny turns” clinics

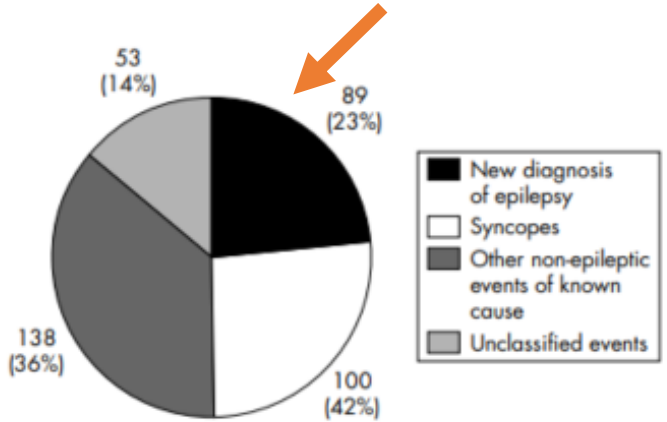


Figure 1 Categories of diagnoses (n = 380).

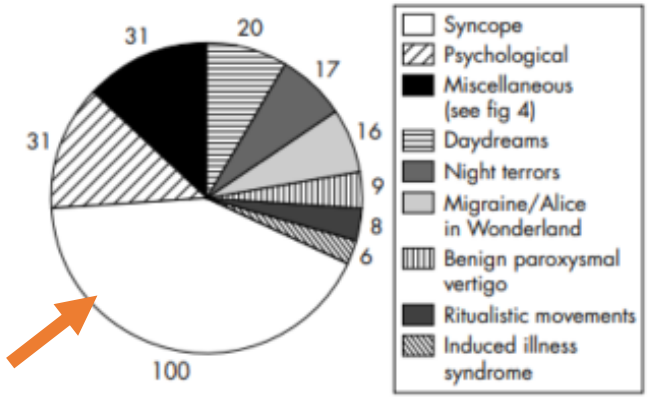


Figure 3 Non-epileptic diagnoses (n = 238).

Hindley D, Ali A, Robson C. Diagnoses made in a secondary care "fits, faints, and funny turns" clinic. *Arch Dis Child*. 2006;91(3):214-218. doi:10.1136/adc.2004.062455

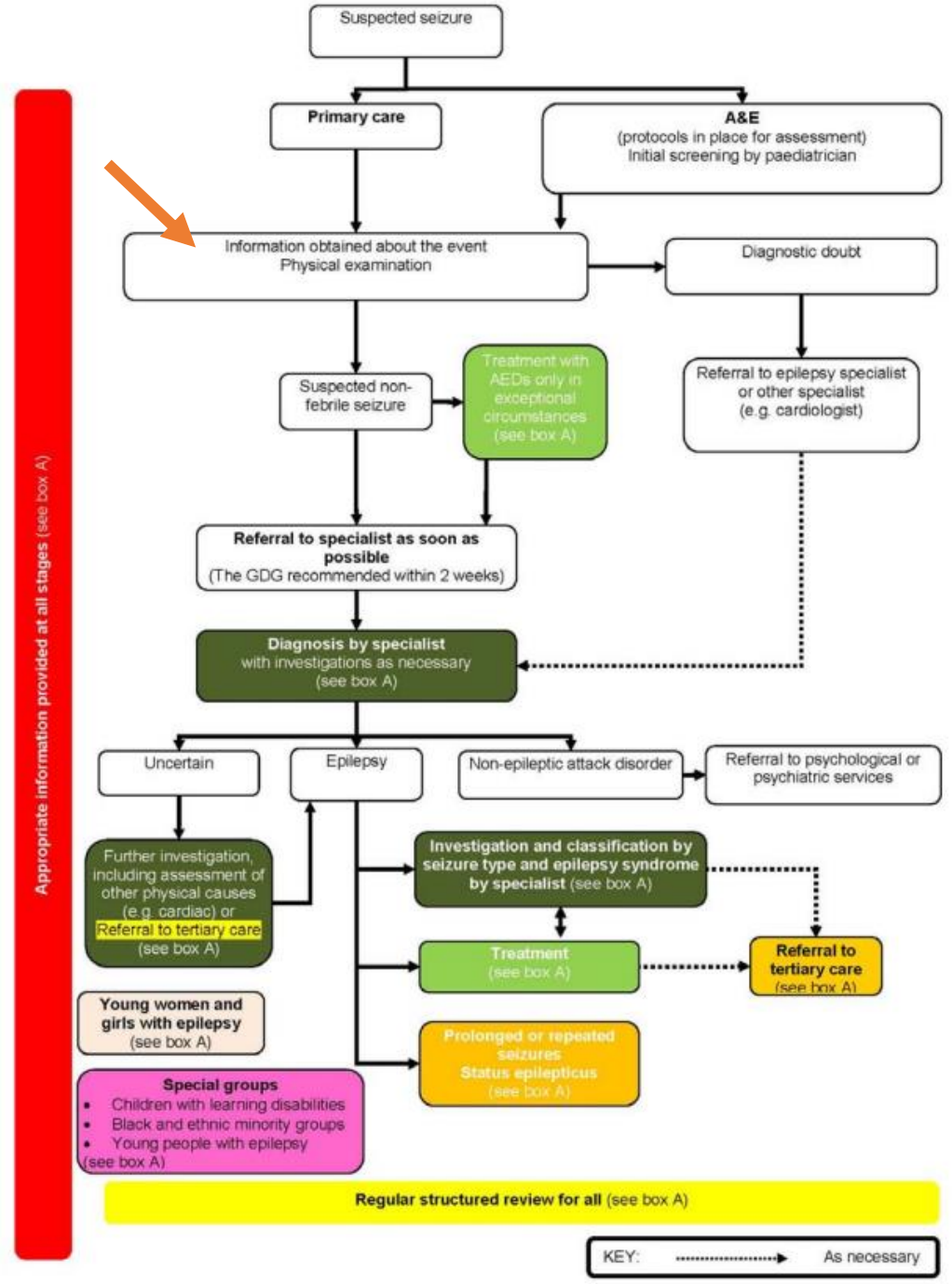
Impact of wrong diagnosis

- Unnecessary investigation
- Wrong treatment / SE of treatment
- Impact on physical, social well being
- Cost / Medico-legal implications

Outline care algorithm for children

Detailed event (ictal) history – **Semiology of events**

Pre - During – Post



Epilepsies: diagnosis and management (CG137 NICE guideline appendix C)

Pre event

- Timing, where
- What was child doing - awake/asleep/eating, playing video games

Any warning (aura) - abdominal discomfort, fear, unpleasant sensations, seeing lights, duration

Triggers:

- Only in sleep
- Feeds related
- Situational/boredom
- Exercise or movement
- Unwell or sick prior episode (fever)
- With excitement or emotion
- Following unpleasant stimuli
- Flashing lights / Alcohol / drug use or medication change

Table 3 Localising value of auras

Aura	Localisation
Somatosensory	
Marching sensory aura	Primary sensory cortex
Tingling, numbness	Primary or secondary somatosensory cortex, supplementary motor area, insula
Pain/ warmth	Secondary somatosensory cortex, insula
Visual aura	
Elementary (lights, shapes)	Primary visual cortex
Illusions/ distortions	Temporo-parieto-occipital
Complex visual hallucinations	Temporal
Auditory	
Elementary	Heschl's gyrus, superior temporal
Illusions/distortion	Lateral temporal, insula
Other	
Gustatory	Insula, mesiotemporal
Olfactory	Insula, mesiotemporal, orbitofrontal
Vestibular	Posterior temporal, parietal
Autonomic	Insula, amygdala, cingulate

Chowdhury FA, Silva R, Whatley B, *et al*
Localisation in focal epilepsy: a practical guide.
Practical Neurology 2021;**21**:481-491

Event description

What were the movements

- Clonic – sustained rhythmical jerking
- Tonic - muscles stiffen and tense
- Atonic - sudden loss of muscle tone
- Myoclonus – sudden lightening like jerk, lasts milliseconds
- Spasm - sudden flexion, extension or both of proximal and truncal muscles (1-2 seconds)
- Absences – vacant stares, brief
- Automatisms – purposeless, stereotypical movements – chewing, picking at clothes

- State of awareness
- Ability to talk, understand
- Change in colour
- Eyes: open, closed, uprolled, deviated
- Arms / legs: which side
- Associated features: frothing, tongue bite, change in breathing pattern, incontinence

Length of event, frequency

Post Event

Duration - how long before child returned to normal activity
  - Response to voice/touch
  - Able to talk or communicate

- Memory for events
- Weakness or numbness or headache
- Tired, sleepy

Scenarios

First Jerking episode

8 year old

Pre-event:
- Standing in assembly at school
- Early morning during heat wave
- Darkening of vision, felt clammy

Trigger:
- Was well when went from home

Event:
- Fell down to the side
- Few body shakes
- Eyes closed
- Lasted 1 min

Post event:
- Tired and sleepy 15-20 min

Syncopal event

2 year old

Pre-event:
- Having temperature and runny nose, given calpol

Trigger:
- Febrile illness

Event:
- Eye rolling
- 2-3 minutes of body jerks
- Blue on face with frothing

Post event:
- Tired and sleepy 15-20 min

Febrile seizure

6 year old

Pre-event:
- Was out with parents on weekend, forest trail, eating lunch

Trigger:
- Was well the previous night, though slept late

Event:
- Jerking of whole body suddenly
- Eyes deviated to one side
- Frothing, gurgling noises
- Lasted 3 min

Post event:
- Tired and sleepy 15-20 min

First afebrile seizure

Assessment

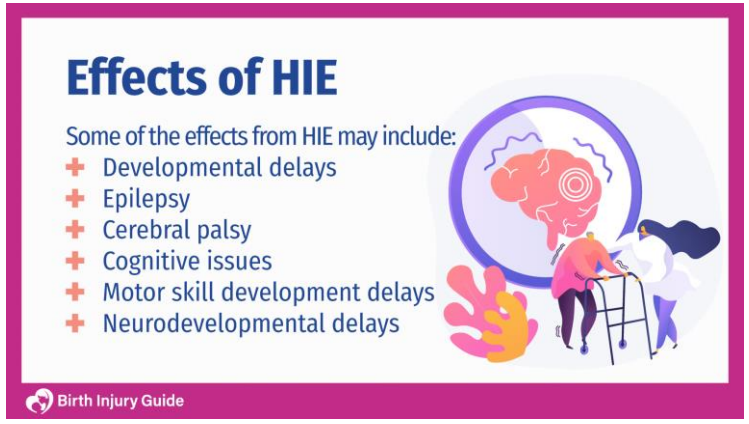
Other history:

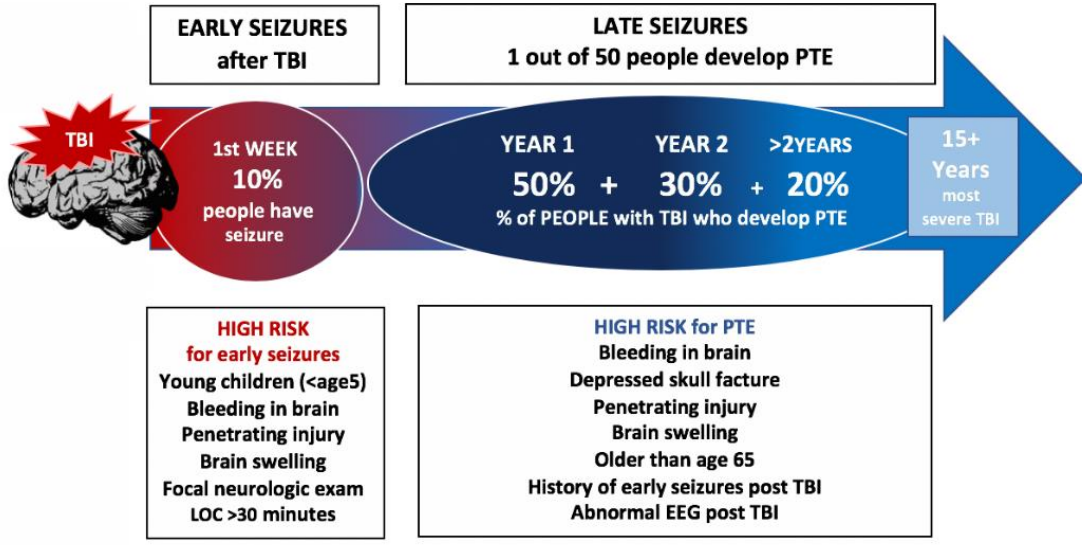
- Perinatal history (HIE /stroke)
- Significant past history (head injury / infection)
- Developmental history
- Learning and Schooling
- Family history - consanguinity

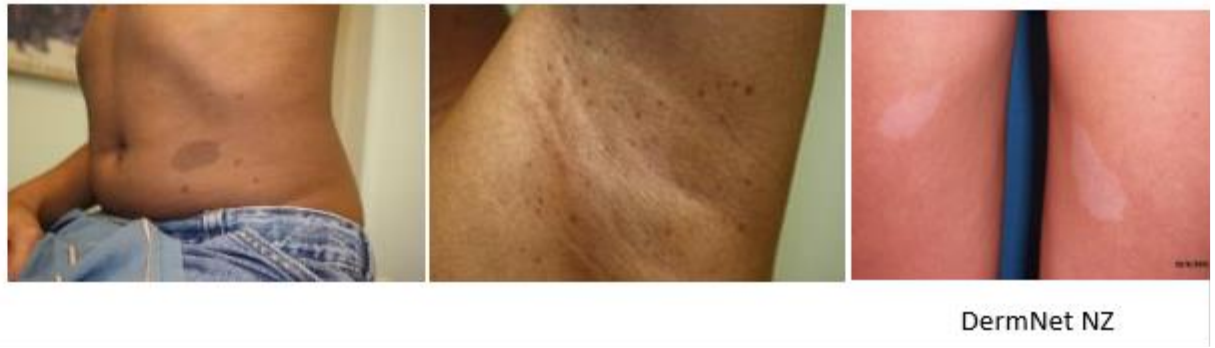
Examination:

- Vitals – temp, HR, RR, BP, Sats
- Growth parameters – weight, HC
- Skin markings, Distinctive features
- Developmental stage
- Signs of meningeal irritation
- Signs of raised ICP
- Systemic examination (inc cardiac)
- Neurology examination

- Video of the event

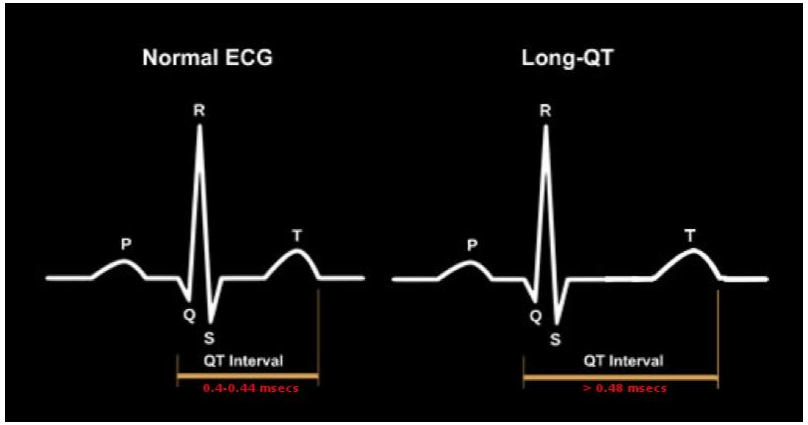






Investigation

- FBC, UEs, Blood glucose and gas, Ca, Mg, clotting
- CRP, blood cultures, urine dip
- Lumbar puncture
- Metabolic – lactate, ammonia, amino acids, urine organic acids
- Urine toxicology
- ECG
- Imaging
    - Acute symptomatic seizure – CT scan
    - MRI : (specialists)
        - epilepsy <2 years
        - focal onset on history, examination or EEG (unless clear evidence of self-limited focal epilepsy)
        - Seizure continue despite first-line medication



EEG

- Variable specificity (78% to 98%) and sensitivity (26% to 56%)
  - Positive (epileptiform discharges) – support (caution: false +)
  - Negative - cannot exclude

- Requested after second epileptic seizure (after first by specialists)
- Inform seizure type and epilepsy syndrome
- Help to assess risk of recurrence

- Improve diagnostic yield
  - Sleep induced /deprived EEG

Do

- EEG requested to support a diagnosis of epilepsy where suspicion of epileptic seizure

Don't

- EEG should not be used to exclude a diagnosis of epilepsy

Management

- Depends on the cause

EPILEPSY IMITATORS

1. OVERVIEW
2. SYNCOPE AND ANOXIC SEIZURES
   a. Vasovagal syncope
   b. Reflex anoxic seizures
   c. Breath-holding attacks
   d. Hyperventilation syncope
   e. Compulsive valsalva
   f. Neurological syncope
   g. Imposed upper airways obstruction
   h. Orthostatic intolerance
   i. Long QT and cardiac syncope
   j. Hyper-cyanotic spells
3. BEHAVIORAL, PSYCHOLOGICAL AND PSYCHIATRIC DISORDERS
   a. Daydreaming /inattention
   b. Self gratification
   c. Eidetic imagery
   d. Tantrums and rage reactions
   e. Out of body experiences
   f. Panic attacks
   g. Dissociative states
   h. Non-epileptic seizures
   i. Hallucinations in psychiatric disorders
   j. Fabricated / factitious illness
4. SLEEP RELATED CONDITIONS
   a. Sleep related rhythmic movement disorders
   b. Hypnogogic jerks
   c. Parasomnias
   d. REM sleep disorders
   e. Benign neonatal sleep myoclonus
   f. Periodic leg movements
   g. Narcolepsy-cataplexy
5. PAROXYSMAL MOVEMENT DISORDERS
   a. Tics
   b. Stereotypies
   c. Paroxysmal kinesigenic dyskinesia
   d. Paroxysmal nonkinesigenic dyskinesia

EpilepsyDiagnosis.org Diagnostic Manual

Syncope and anoxic seizures

- Transient loss of consciousness, abrupt blood flow and oxygen supply to brain
- Can be collapse, stiffening or tonic-clonic movements - not due to epileptic discharges
- e.g – Reflex anoxic – early infancy following painful stimuli, good prognosis

Benign neonatal sleep myoclonus

- Movements seen in neonatal period and thereafter for months, in sleep
- Brief flurries of jerking of one limb or other
- Normal examination and development

Tics

- Involuntary, sudden, repetitive movements
- common in childhood
- compulsive urge and suppressibility

Review videos
ECG
Reassurance

Not Epilepsy
Do not trial anti-epileptic drug
If unsure - refer

Acute symptomatic seizure

- Secondary to acute CNS insult
- may be metabolic, structural, infectious, or inflammation
- E.g – within a week of stroke, meningitis, HIE
- Unlikely to be recurrent

Febrile seizures

- Seizure in child 6 m – 6 y with fever
- Simple (GTC, <15 minutes, not recur within same febrile illness) versus Complex
- Management
  - Investigate underlying cause
  - Antipyretics; Seizure first aid
  - Risk of recurrence

First afebrile seizure

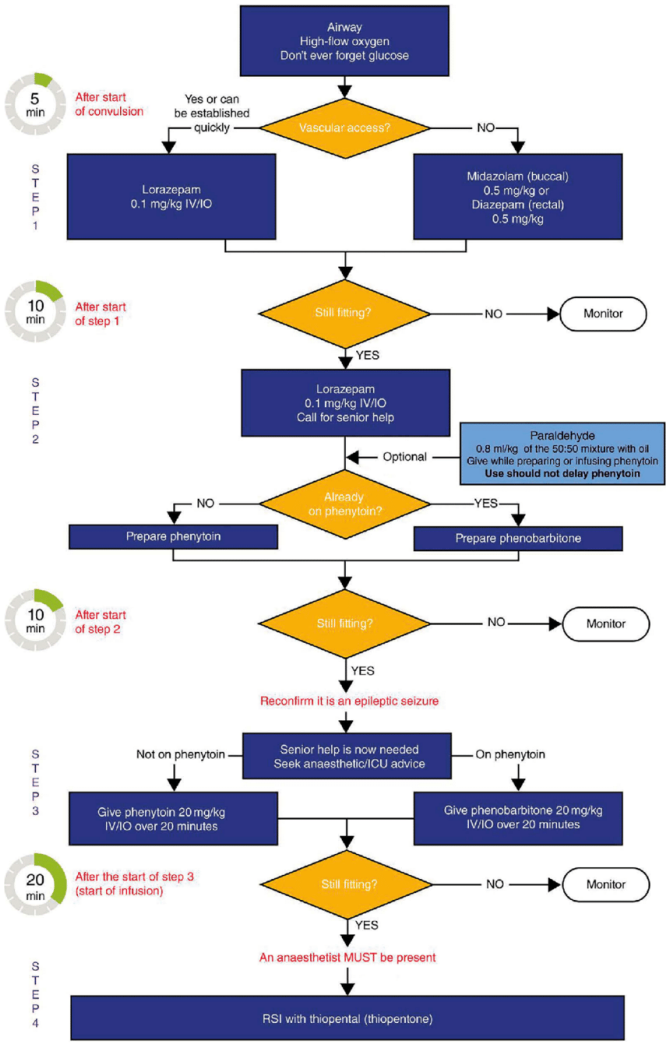
- Seizure first aid
- Information leaflet

Risk of recurrence after a first tonic–clonic seizure
- 36–52%
- meta-analysis 2 year risk of recurrence ~ 40%

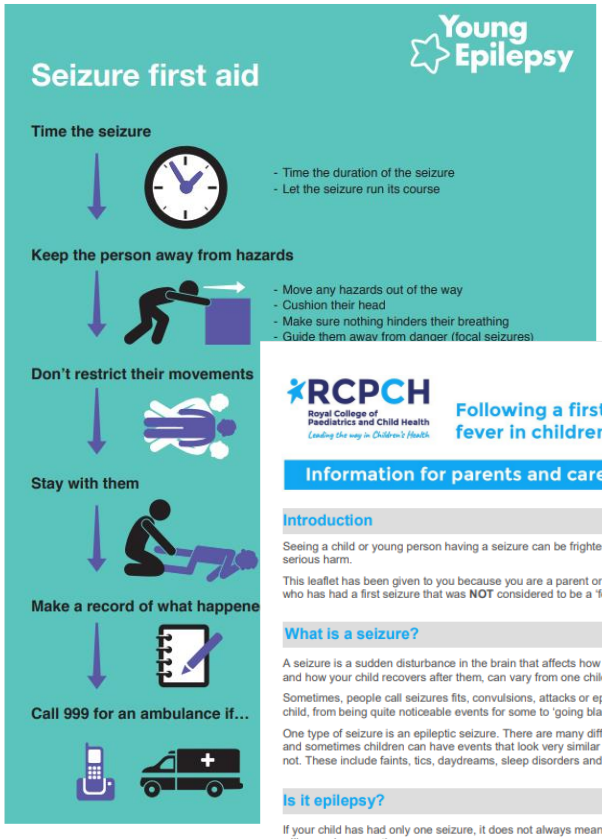
Predictors of recurrent seizures
- remote symptomatic etiology
- abnormal neurologic examination
- Intellectual disability
- epileptiform discharge on EEG
- focal onset seizures

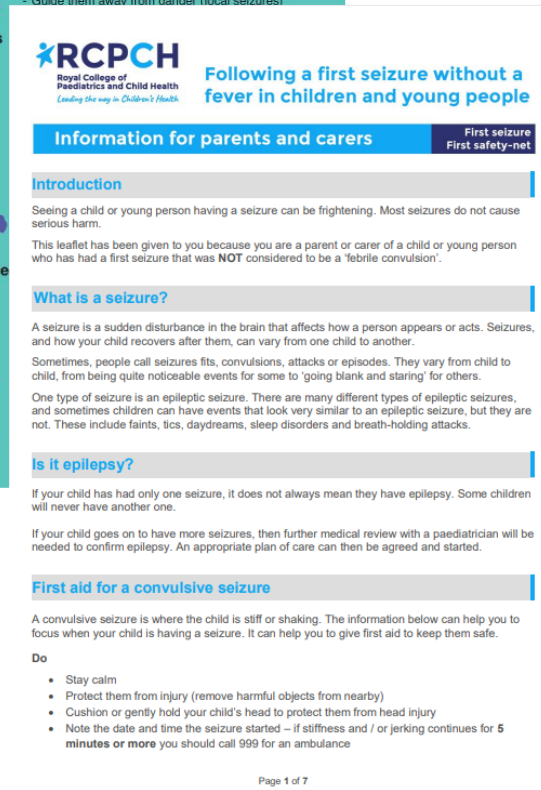
Hamiwka, L.D., Singh, N., Niosi, J. and Wirrell, E.C. (2007), Diagnostic Inaccuracy in Children Referred with "First Seizure": Role for a First Seizure Clinic. Epilepsia, 48: 1062-1066.

- Emergency medication
- Referral to first fit clinic



Advanced Paediatric Life Support-Status Epilepticus Algorithm




RCPCH
Royal College of Paediatrics and Child Health
Leading the way in Children's Health

Following a first seizure without a fever in children and young people

Information for parents and carers | First seizure First safety-net

Introduction

Seeing a child or young person having a seizure can be frightening. Most seizures do not cause serious harm.

This leaflet has been given to you because you are a parent or carer of a child or young person who has had a first seizure that was **NOT** considered to be a 'febrile convulsion'.

What is a seizure?

A seizure is a sudden disturbance in the brain that affects how a person appears or acts. Seizures, and how your child recovers after them, can vary from one child to another.

Sometimes, people call seizures fits, convulsions, attacks or episodes. They vary from child to child, from being quite noticeable events for some to 'going blank and staring' for others.

One type of seizure is an epileptic seizure. There are many different types of epileptic seizures, and sometimes children can have events that look very similar to an epileptic seizure, but they are not. These include faints, tics, daydreams, sleep disorders and breath-holding attacks.

Is it epilepsy?

If your child has had only one seizure, it does not always mean they have epilepsy. Some children will never have another one.

If your child goes on to have more seizures, then further medical review with a paediatrician will be needed to confirm epilepsy. An appropriate plan of care can then be agreed and started.

First aid for a convulsive seizure

A convulsive seizure is where the child is stiff or shaking. The information below can help you to focus when your child is having a seizure. It can help you to give first aid to keep them safe.

Do

- Stay calm
- Protect them from injury (remove harmful objects from nearby)
- Cushion or gently hold your child's head to protect them from head injury
- Note the date and time the seizure started – if stiffness and / or jerking continues for **5 minutes or more** you should call 999 for an ambulance

Page 1 of 7

Epilepsy

1.1 Definition of epilepsy

2004

An epilepsy is defined as a neurological condition characterised by recurrent epileptic seizures unprovoked by any immediately identifiable cause. An epileptic seizure is the clinical manifestation of an abnormal and excessive discharge of a set of neurons in the brain [1].

Epilepsy should be viewed as a symptom of an underlying neurological disorder and not as a single disease entity. The term 'epilepsies' is used in the title of the guideline to reflect this.

Practical Definition of Epilepsy

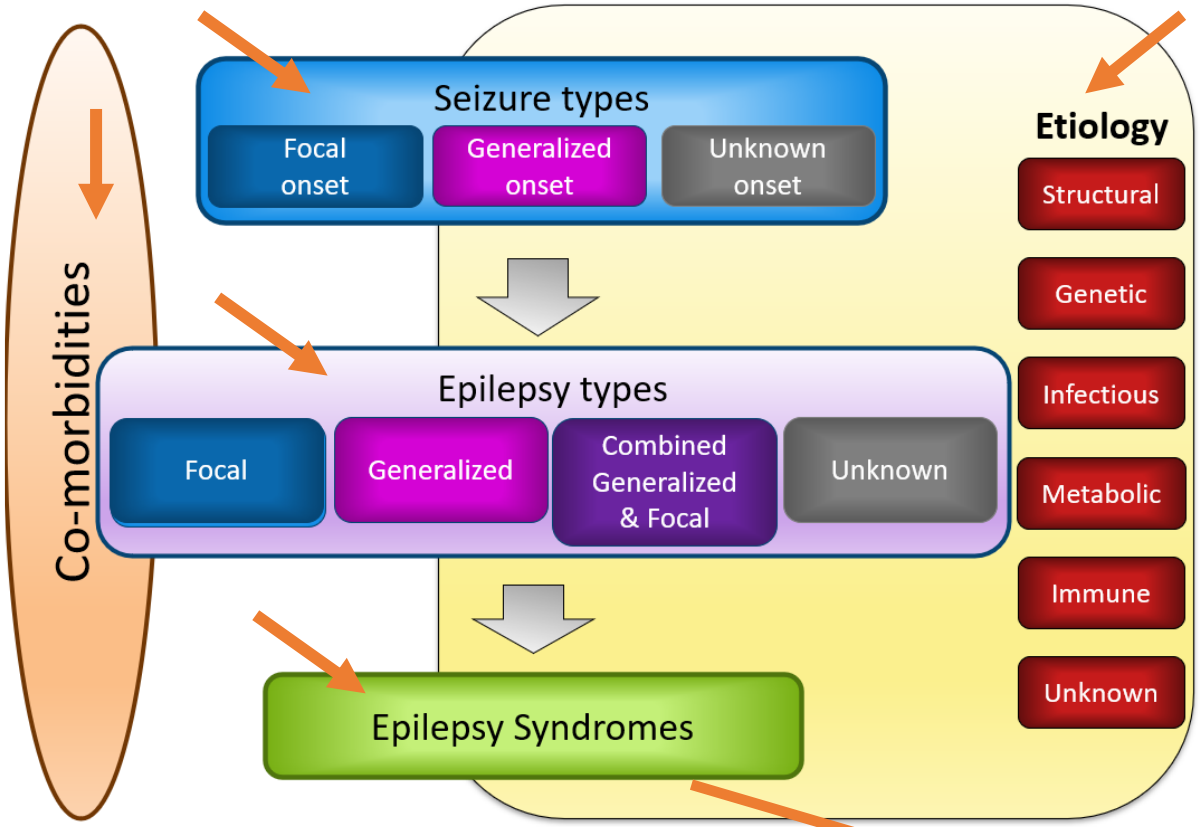
Table 2. Operational (practical) clinical definition of epilepsy

Epilepsy is a disease of the brain defined by any of the following conditions

1. At least two unprovoked (or reflex) seizures occurring >24 h apart
2. One unprovoked (or reflex) seizure and a probability of further seizures similar to the general recurrence risk (at least 60%) after two unprovoked seizures, occurring over the next 10 years
3. Diagnosis of an epilepsy syndrome

Epilepsia, 55(4):475–482, 2014
doi: 10.1111/epi.12550

Scheffer IE, Berkovic S, Capovilla G, et al. ILAE classification of the epilepsies: Position paper of the ILAE Commission for Classification and Terminology. *Epilepsia*. 2017;58(4):512-521. doi:10.1111/epi.13709



ILAE 2017 Classification of Seizure Types Expanded Version [1]

Focal Onset

Aware | Impaired Awareness

Motor Onset
- automatisms
- atonic [2]
- clonic
- epileptic spasms [2]
- hyperkinetic
- myoclonic
- tonic

Nonmotor Onset
- autonomic
- behavior arrest
- cognitive
- emotional
- sensory

focal to bilateral tonic–clonic

Generalized Onset

Motor
- tonic–clonic
- clonic
- tonic
- myoclonic
- myoclonic–tonic–clonic
- myoclonic–atonic
- atonic
- epileptic spasms

Nonmotor (absence)
- typical
- atypical
- myoclonic
- eyelid myoclonia

Unknown Onset

Motor
- tonic–clonic
- epileptic spasms

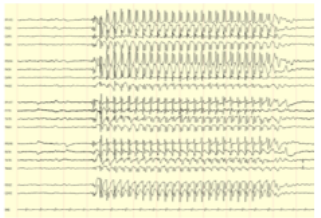
Nonmotor
- behavior arrest

Unclassified [3]

Classification example : Generalized epilepsy with absence seizures with no identified cause, likely CAE with no cognitive concerns

Childhood absence epilepsy

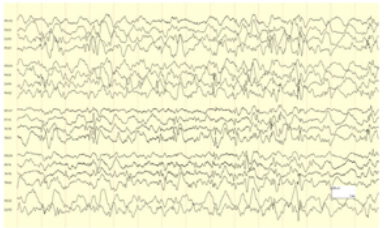
- 2 to 12 years (peak 5-6 years)
- Multiple, brief daily absences
- 2.5 - 3.5 Hz generalized spike-and-wave



- Normal past history, development and examination
- high likelihood of seizures spontaneously remitting
- Rx - Ethosuximide

West syndrome

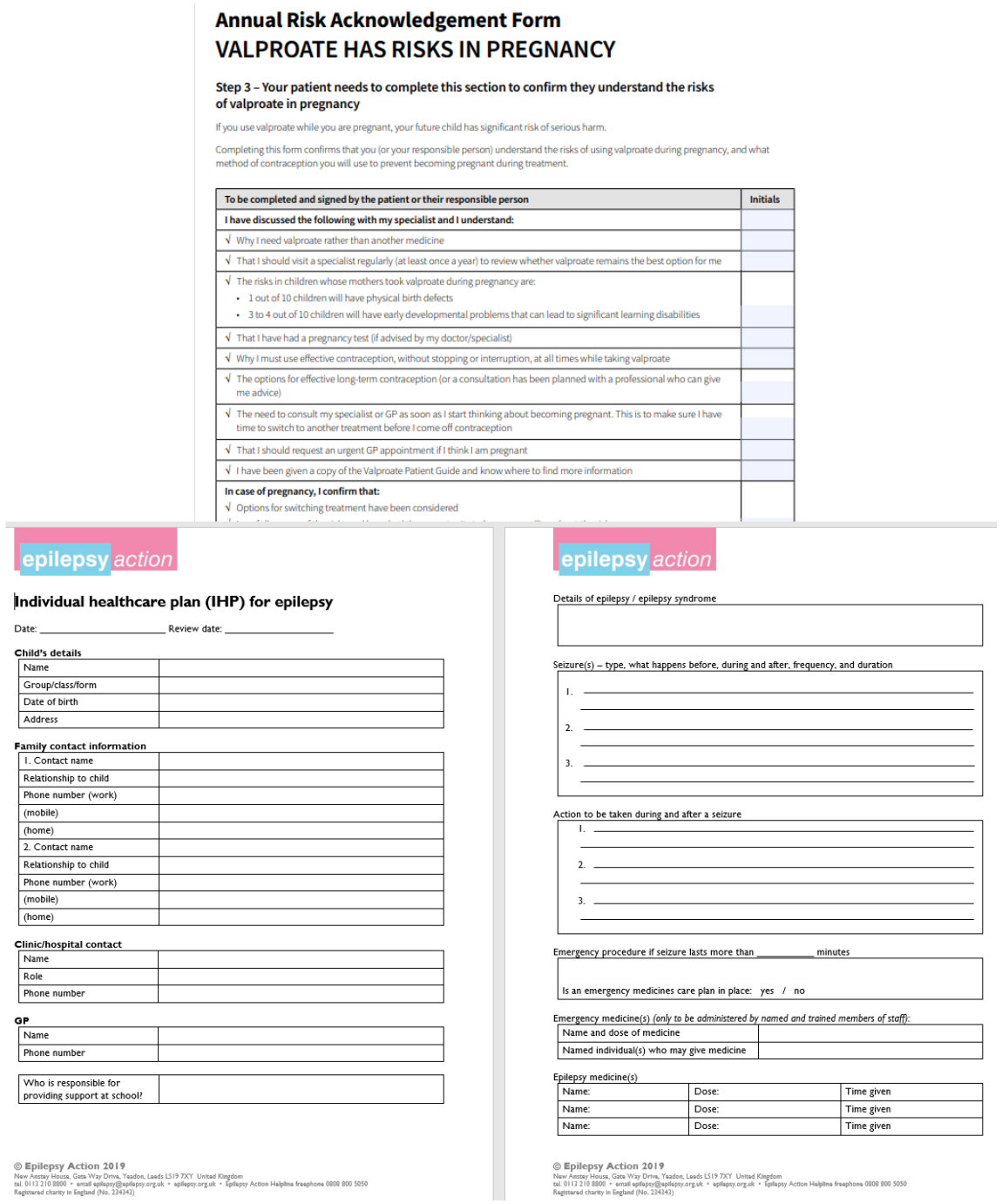
- Epileptic spasms between 3 and 12 months of age
- Hypsarrhythmia on EEG



- May or may not have underlying cause (HIE, syndromic – Down's)
- Global developmental delay with or without regression at seizure onset
- Rx – Steroids and/or Vigabatrin

Management

- Confirmed diagnosis
- Anti-epileptic therapy
  - Is it needed – do not give a trial of AED
  - Choice of AED
    - Sodium Valproate – wide spectrum, MHRA safety advise
    - Lamotrigine – focal onset seizures
    - Carbamazepine – narrow spectrum
    - Discussion of risks and benefits
  - Principles of starting – go low and slow
- Safety advise, SUDEP discussion
- Comprehensive health care plan
  - 95% of the children had a significant difficultly in at least one area of learning or behaviour
- Regular follow up
- Tertiary referral



Annual Risk Acknowledgement Form
VALPROATE HAS RISKS IN PREGNANCY

Step 3 – Your patient needs to complete this section to confirm they understand the risks of valproate in pregnancy

If you use valproate while you are pregnant, your future child has significant risk of serious harm.

Completing this form confirms that you (or your responsible person) understand the risks of using valproate during pregnancy, and what method of contraception you will use to prevent becoming pregnant during treatment.

To be completed and signed by the patient or their responsible person	Initials
I have discussed the following with my specialist and I understand:	
√ Why I need valproate rather than another medicine	
√ That I should visit a specialist regularly (at least once a year) to review whether valproate remains the best option for me	
√ The risks in children whose mothers took valproate during pregnancy are: • 1 out of 10 children will have physical birth defects • 3 to 4 out of 10 children will have early developmental problems that can lead to significant learning disabilities	
√ That I have had a pregnancy test (if advised by my doctor/specialist)	
√ Why I must use effective contraception, without stopping or interruption, at all times while taking valproate	
√ The options for effective long-term contraception (or a consultation has been planned with a professional who can give me advice)	
√ The need to consult my specialist or GP as soon as I start thinking about becoming pregnant. This is to make sure I have time to switch to another treatment before I come off contraception	
√ That I should request an urgent GP appointment if I think I am pregnant	
√ I have been given a copy of the Valproate Patient Guide and know where to find more information	
In case of pregnancy, I confirm that:	
√ Options for switching treatment have been considered	

epilepsy action

Individual healthcare plan (IHP) for epilepsy

Date: _____ Review date: _____

Child's details

Name	
Group/class/form	
Date of birth	
Address	

Family contact information

1. Contact name	
Relationship to child	
Phone number (work)	
(mobile)	
(home)	
2. Contact name	
Relationship to child	
Phone number (work)	
(mobile)	
(home)	

Clinic/hospital contact

Name	
Role	
Phone number	

GP

Name	
Phone number	

Who is responsible for providing support at school?	

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Registered charity in England (No. 234343)

epilepsy action

Details of epilepsy / epilepsy syndrome

Seizure(s) – type, what happens before, during and after, frequency, and duration

1. _____
2. _____
3. _____

Action to be taken during and after a seizure

1. _____
2. _____
3. _____

Emergency procedure if seizure lasts more than _____ minutes

Is an emergency medicines care plan in place: yes / no

Emergency medicine(s) *(only to be administered by named and trained members of staff)*:

Name and dose of medicine	
Named individual(s) who may give medicine	

Epilepsy medicine(s)

Name:	Dose:	Time given
Name:	Dose:	Time given
Name:	Dose:	Time given

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Summary

- Fits and funny turns are common presentation in children
- Epilepsy is a common neurological disorder characterised by recurring seizures and can be easily misdiagnosed
- History taking and assessment is key to differentiate epilepsy from non-epileptic movements
- Those with suspected epileptic seizure should be timely referred and appropriately managed

References

- Guidelines // International League Against Epilepsy (ilae.org)
- EpilepsyDiagnosis.org
- Epilepsy action www.epilepsy.org.uk
- Young epilepsy www.youngepilepsy.org.uk
- Epilepsies: diagnosis and management www.nice.org.uk/guidance/cg137
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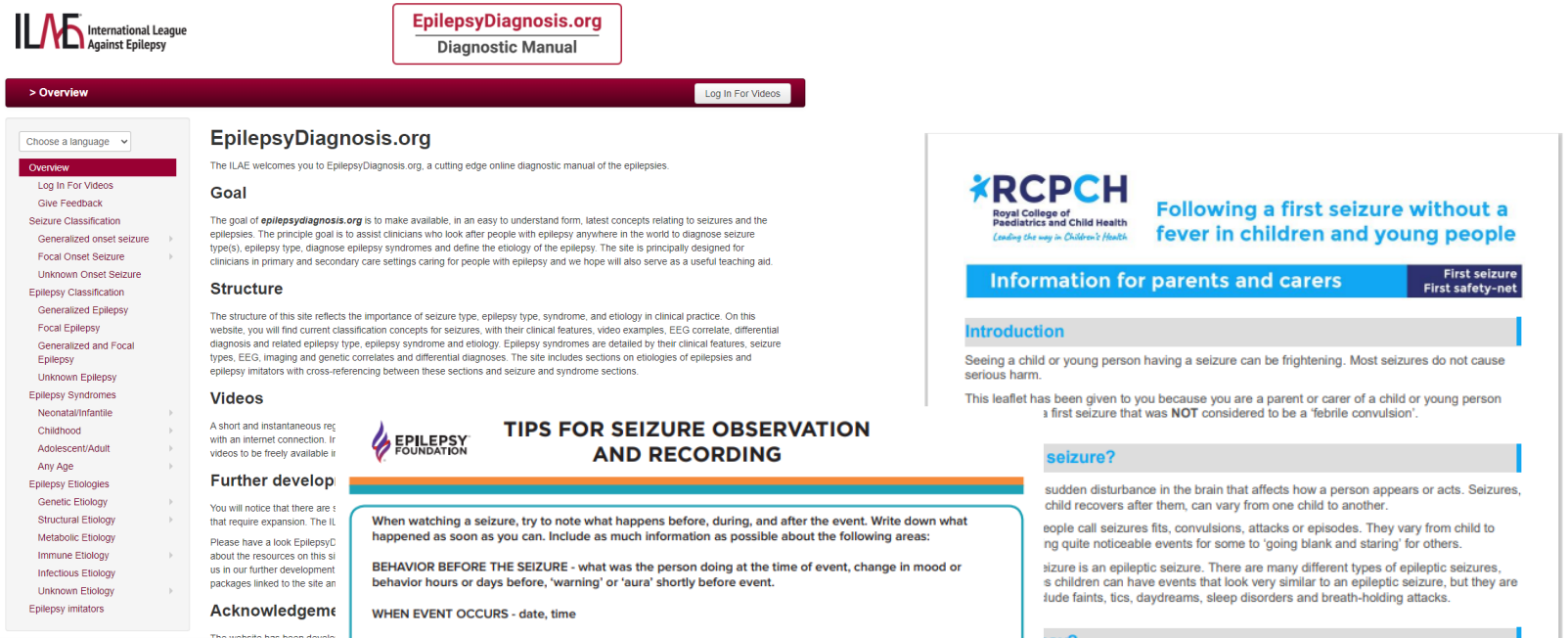
Resources


ILAE International League Against Epilepsy
EpilepsyDiagnosis.org
Diagnostic Manual
> Overview
Log In For Videos
Choose a language
Overview
Log In For Videos
Give Feedback
Seizure Classification
Generalized onset seizure
Focal Onset Seizure
Unknown Onset Seizure
Epilepsy Classification
Generalized Epilepsy
Focal Epilepsy
Generalized and Focal Epilepsy
Unknown Epilepsy
Epilepsy Syndromes
Neonatal/Infantile
Childhood
Adolescent/Adult
Any Age
Epilepsy Etiologies
Genetic Etiology
Structural Etiology
Metabolic Etiology
Immune Etiology
Infectious Etiology
Unknown Etiology
Epilepsy imitators
EpilepsyDiagnosis.org
The ILAE welcomes you to EpilepsyDiagnosis.org, a cutting edge online diagnostic manual of the epilepsies.
Goal
The goal of epilepsydiagnosis.org is to make available, in an easy to understand form, latest concepts relating to seizures and the epilepsies. The principle goal is to assist clinicians who look after people with epilepsy anywhere in the world to diagnose seizure type(s), epilepsy type, diagnose epilepsy syndromes and define the etiology of the epilepsy. The site is principally designed for clinicians in primary and secondary care settings caring for people with epilepsy and we hope will also serve as a useful teaching aid.
Structure
The structure of this site reflects the importance of seizure type, epilepsy type, syndrome, and etiology in clinical practice. On this website, you will find current classification concepts for seizures, with their clinical features, video examples, EEG correlate, differential diagnosis and related epilepsy type, epilepsy syndrome and etiology. Epilepsy syndromes are detailed by their clinical features, seizure types, EEG, imaging and genetic correlates and differential diagnoses. The site includes sections on etiologies of epilepsies and epilepsy imitators with cross-referencing between these sections and seizure and syndrome sections.
Videos
Further develop
Acknowledgeme

RCPCH
Royal College of Paediatrics and Child Health
Following a first seizure without a fever in children and young people
Information for parents and carers
First seizure
First safety-net
Introduction
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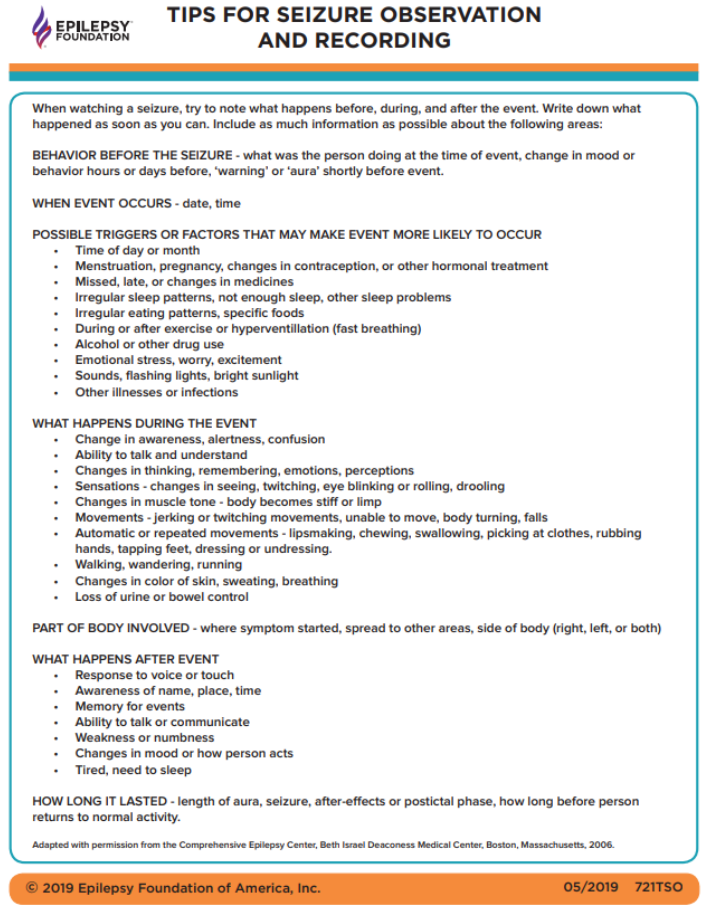
eople call seizures fits, convulsions, attacks or episodes. They vary from child to
ng quite noticeable events for some to 'going blank and staring' for others.
izure is an epileptic seizure. There are many different types of epileptic seizures,
s children can have events that look very similar to an epileptic seizure, but they are
lude faints, tics, daydreams, sleep disorders and breath-holding attacks.
sy?
s had only one seizure, it does not always mean they have epilepsy. Some children
e another one.
es on to have more seizures, then further medical review with a paediatrician will be
firm epilepsy. An appropriate plan of care can then be agreed and started.
or a convulsive seizure
eizure is where the child is stiff or shaking. The information below can help you to
ur child is having a seizure. It can help you to give first aid to keep them safe.
lm
them from injury (remove harmful objects from nearby)
n or gently hold your child's head to protect them from head injury
e date and time the seizure started – if stiffness and / or jerking continues for 5
s or more you should call 999 for an ambulance
Page 1 of 7


EPILEPSY FOUNDATION
TIPS FOR SEIZURE OBSERVATION AND RECORDING
When watching a seizure, try to note what happens before, during, and after the event. Write down what happened as soon as you can. Include as much information as possible about the following areas:
BEHAVIOR BEFORE THE SEIZURE - what was the person doing at the time of event, change in mood or behavior hours or days before, 'warning' or 'aura' shortly before event.
WHEN EVENT OCCURS - date, time
POSSIBLE TRIGGERS OR FACTORS THAT MAY MAKE EVENT MORE LIKELY TO OCCUR
Time of day or month
Menstruation, pregnancy, changes in contraception, or other hormonal treatment
Missed, late, or changes in medicines
Irregular sleep patterns, not enough sleep, other sleep problems
Irregular eating patterns, specific foods
During or after exercise or hyperventillation (fast breathing)
Alcohol or other drug use
Emotional stress, worry, excitement
Sounds, flashing lights, bright sunlight
Other illnesses or infections
WHAT HAPPENS DURING THE EVENT
Change in awareness, alertness, confusion
Ability to talk and understand
Changes in thinking, remembering, emotions, perceptions
Sensations - changes in seeing, twitching, eye blinking or rolling, drooling
Changes in muscle tone - body becomes stiff or limp
Movements - jerking or twitching movements, unable to move, body turning, falls
Automatic or repeated movements - lipsmaking, chewing, swallowing, picking at clothes, rubbing hands, tapping feet, dressing or undressing.
Walking, wandering, running
Changes in color of skin, sweating, breathing
Loss of urine or bowel control
PART OF BODY INVOLVED - where symptom started, spread to other areas, side of body (right, left, or both)
WHAT HAPPENS AFTER EVENT
Response to voice or touch
Awareness of name, place, time
Memory for events
Ability to talk or communicate
Weakness or numbness
Changes in mood or how person acts
Tired, need to sleep
HOW LONG IT LASTED - length of aura, seizure, after-effects or postictal phase, how long before person returns to normal activity.
Adapted with permission from the Comprehensive Epilepsy Center, Beth Israel Deaconess Medical Center, Boston, Massachusetts, 2006.
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