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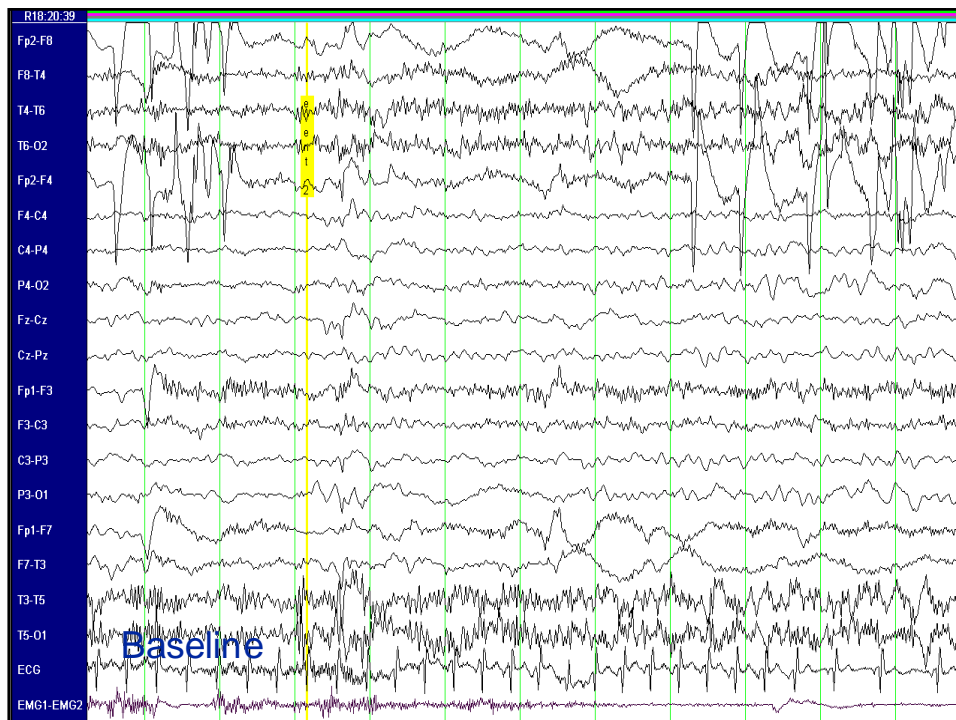
Clinical Features and Differential Diagnosis of Syncope and Apnoea in Children and Infants

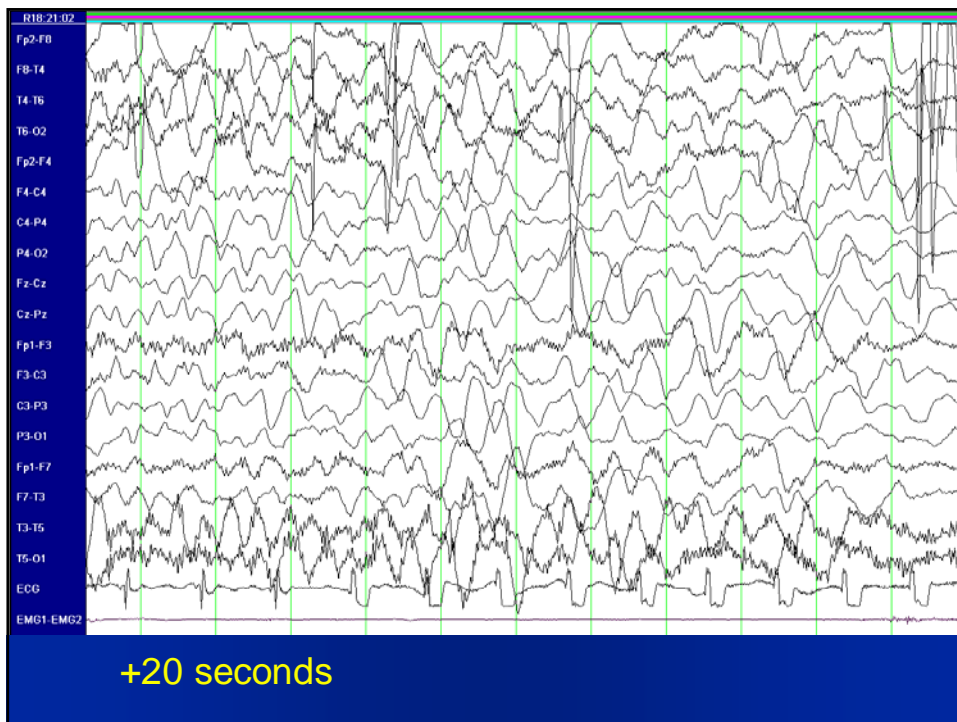
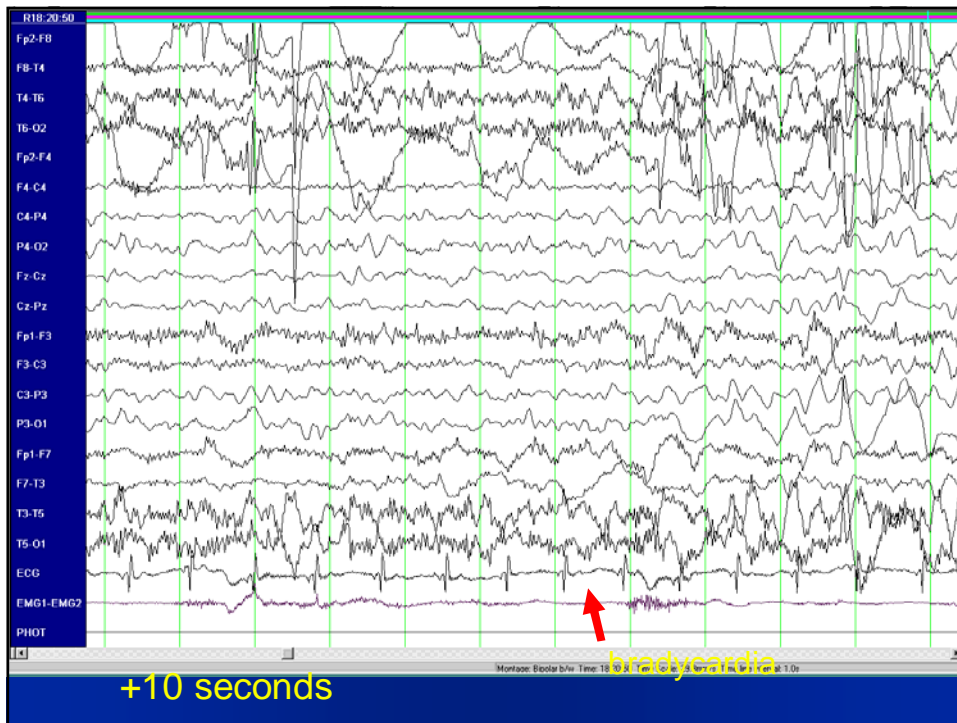
- Breath-holding events
- Apparent life threatening events
- Apnoea and posturing : Raised intracranial pressure
- Mimicking syncope : Pseudoseizures

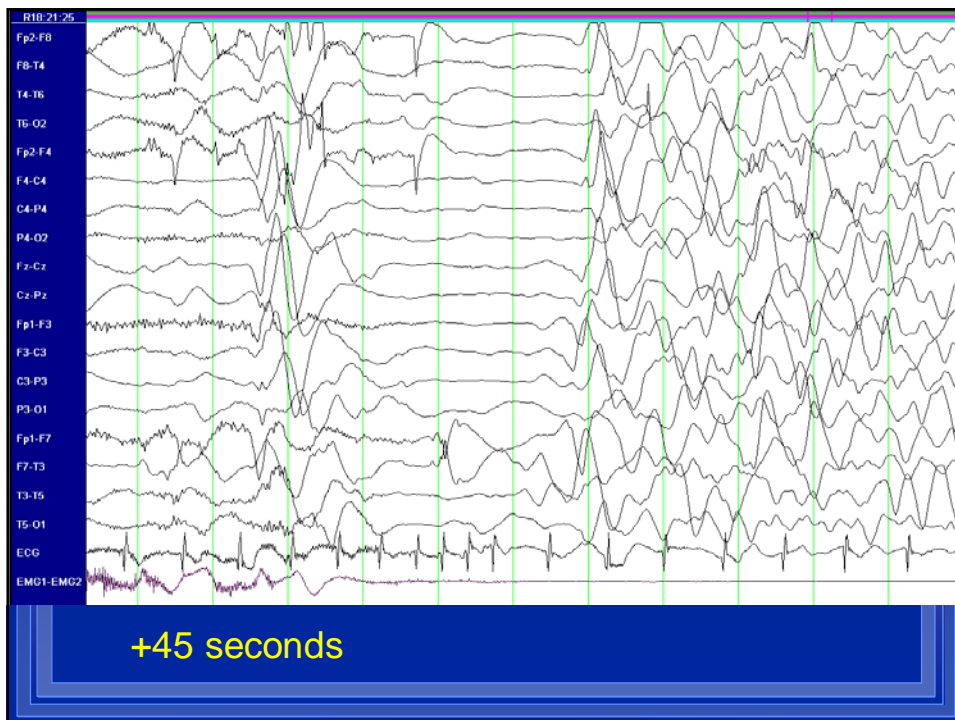
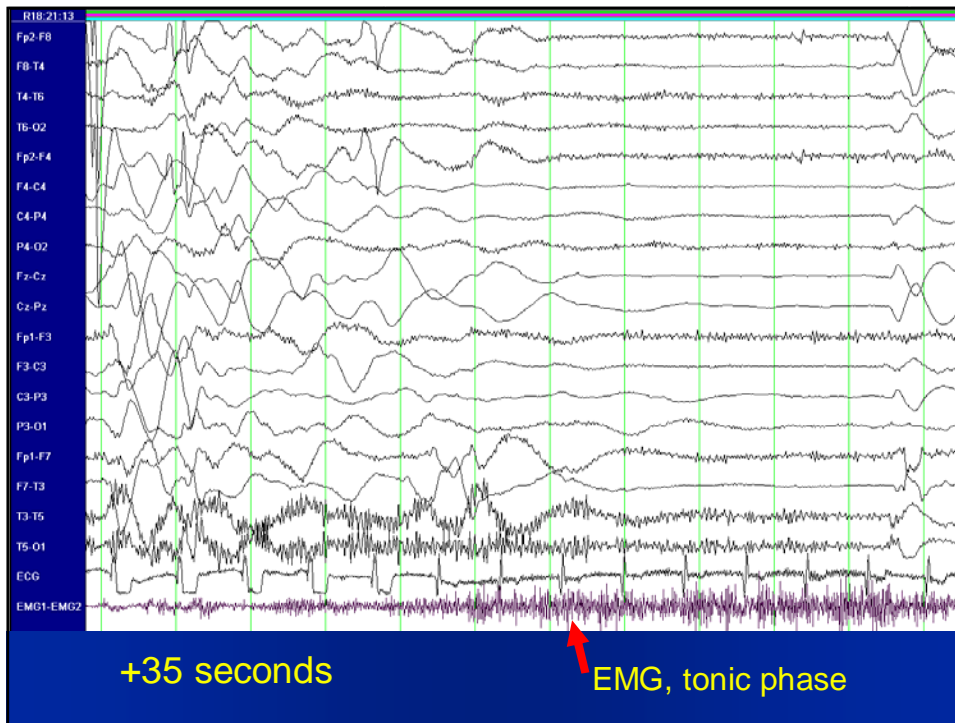
Infantile Syncope

Transient loss of consciousness and postural tone caused by a global reduction of blood flow to the brain.

Syncopal attacks illustrate well the dictum that the differential diagnosis of epilepsy depends much more on what precedes or follows an attack than on what occurs during it.







Breath-Holding Spells (Cyanotic and Pallid Infantile Syncope)

Paediatrics 39;4;563-581, 1967

- Incidence : 4.6%
- Family history : 23%
- Types : cyanotic : 62%
: pallid : 19%
: mixture : 19%

Subsequent data : cyanotic : pallid : both : 5:3:2

Breath-Holding Spells (Cyanotic and Pallid Infantile Syncope)

Paediatrics 39;4;563-581, 1967

- Onset : Neonate to 4years, 7% in first 2months, almost all by 3years
- Peak Incidence : 6/12 - 18/12 (76%)
- Pattern : 85% attacks free by 5
- Frequency variation
- Precipitating factors
- Frequent sequel : syncopal attacks

Prospective study of children with cyanotic and pallid breath holding spells

- **Aim** : Determination of natural history of severe breath holding spells
- **Results** : 95 children
 - Median presentation : 6-12 months with **15% < 6 months.**
 - Median frequency of spells : weekly with **30% = or >1 /day.**
 - Median age at peak frequency : 12-18 months.
 - Of those in remission for 12 months last spell occurred at median age of 37-42 months.
 - Of those with current spells oldest age at time of latest spell was 7.
 - **Hypoxic convulsions : 15% of participants.**
 - **Positive family history : 34%.**
 - **Syncope : 18%.**

Pediatrics 107 (2), 265-9, 2001

Mechanisms

- Autonomic
- Family history
- Role of Iron

Cyanotic Breath-Holding Attacks Prolonged Expiratory Apnoea

1. Crying → hypocapnoeic cerebral vasoconstriction
2. Apnoea → hypoxaemia
3. Valsalva → increased intrathoracic pressure and reduced venous return
4. Reduced cardiac output
5. Impaired cerebral circulation (1.4)

Maybe intrapulmonary shunting as cyanosis occurs at 90% oxygen saturation

Predominantly sympathetically mediated response

Pallid Breath-Holding Attacks Reflex Anoxic Seizures

- Precipitated by little crying and loss of consciousness is rapid. Major clinical observation is pallor and main precipitant is pain.
- Loss of consciousness is preceded by seconds of asystole. Cascade of events is primarily **parasympathetically mediated**.
- Exaggerated oculocardiac reflex.
 - Ocular compression: hypersensitive cardiac inhibitory reflex, mediated by vagus nerve
- Pacemaker implantation has been used for severe spells associated with seizures, life threatening bradycardia or asystole

— *Pediatrics* 108,3, 698-72, 2001

Family Pedigree of Children with Severe Breath-Holding Spells (SBHS)

(*J Pediatr* 1997;130:647)

- 57 probands (44 cyanotic, 13 pallid) studied whose families comprised 1683 individuals
- 27% of proband parents and 21% of proband siblings had current or prior SBHS
- Father to son transmission was observed in 7 instances

Autosomal dominant trait with reduced penetrance

Breath holding spells in 91 children and response to treatment with iron

Arch Dis Child 1999 ;81: 261

- Age : 6-40 months, cyanotic (60), anoxic/mixed (31) followed median 45 months
 - Frequency : 49 < 10/month; 22 :10-30/month; 20 : > 30/month.
- Iron deficient : 63/91 treated 3 months
- **Significant difference for correction of breath holding spells between treated and not (84.1% vs 21.4%)**
- Iron deficiency may play a role in autonomic dysregulation (via catecholamine metabolism)
 - Improvement in autonomic function (heart rate variability) has been demonstrated following iron therapy
 - (*J Child Neurol* 2002;17: 337-40)

Anoxic-epileptic seizures : Observational study of epileptic seizures induced by syncope

Arch Dis Childh 2005, 90 (12) 1283-7

- 27 children (1972-2002)
- Age of onset of syncope 5 days – 10 years, median 8 months
- Frequency of syncope : 2 in total to 40/day
- **Types of syncope:**
 - Reflex anoxic seizure (9). Asystole : range 17-31 seconds
 - Prolonged expiratory apnoea /cyanotic BHS (6)
 - Infantile syncope of mixed origin (9)
 - Vasovagal (2) : ear piercing, hair grooming
 - Compulsive valsalva (1), boy with autism

Anoxic-epileptic seizures : Observational study of epileptic seizures induced by syncope

Arch Dis Childh 2005, 90 (12) 1283-7

- Median age of onset anoxic epileptic seizure : 17 months, total of one to 3/day
 - Epileptic component ; bilateral clonic, median duration 5 minutes, range : 0.5-40 minutes
 - epileptic component > 10 mins (12), > 30 minutes (4)
- i.e. status epilepsy is frequent complication**
- 3 independent diagnoses of epilepsy
 - Management of epileptic component : diazepam/anticonvulsants

Infants with Apparent Life – Threatening Events

Infantile Apnoea

Infants with apparent life threatening event.

- Syndrome predominantly affecting children < 1 year, peak incidence one week to 2 months. **Not a diagnosis**
- Constellation of symptoms : apnoea, change in colour, change in muscle tone, coughing or gagging. Episode is frightening!
- Broad spectrum of diagnoses. Gastrointestinal, **neurological**, respiratory, cardiac, metabolic, **child abuse**, sepsis, unknown
- **Medline analysis 1980-2002 : seizures 8%**
 - (Arch Dis Childh 2004; 89: 1043-48)

**Infants with apparent life threatening event (ALTE)
Description and Evaluation of Event**

- **Condition of child** : *awake or sleep, position of infant, location of child in relation to parents, pillows, blankets*
- **Activity and noise at time of event** : *feeding, coughing, choking, vomiting, stridor, collateral evidence of seizure*
- **Breathing efforts** : *none, shallow, gasping, increased*
- **Colour** : *pallor, red, purple, blue, distribution*
- **Movement and tone** : *still, staring, posturing, rigid, floppy*
- **Duration of event, resuscitation, time to normality**
- **Examination**

Infants with apparent life threatening event.

- Investigation and management based on assessment
- Importance of association and causation
- Overall mortality rate :4%
- The majority of sudden infant death occurs in children without a history of apparent life - threatening events.

Reflux Associated Apnoea in Infants : Evidence for a Laryngeal Chemoreflex

Am J Med 1997;103(5A), 120-124

- Mucosa covering interarytenoid space contains receptors stimulated by fluid igniting laryngeal chemoreflex.
- Aspiration preventative reflex response : swallowing, cessation of breathing, airway constricture and coughing.
- In certain infants a hyperactive reflex may cause episodic prolonged apnoea
- Has been implicated in apnoea of prematurity, gastric – regurgitation – related apnoea and apnoea associated with respiratory infection

Apnoeic attacks as an isolated manifestation of epileptic seizures in infants

(Eur J Paed Neurol 1998; 2: 187-191)

(Ann neurol 7: 577-582, 1980)

- Prolonged apnoeic spells without bradycardia should suggest seizure
- Prolonged apnoea with bradycardia suggests non epileptic phenomenon
- Multiple seizure locations demonstrated, commonest temporal (right or left) but central, occipital, parietal and frontal also recorded

Message

- **Differential Diagnosis of Acute Life Threatening Event is broad!**
- **Apnoea as a manifestation of seizure and presenting as an acute life threatening event is an important consideration**
- **Monitoring can be critical. Dual pathology maybe seen.**

Raised Intracranial Pressure

- Later stages : irregularity of respiratory rhythm, apnoea, decerebrate posturing, slowing of pulse and raised blood pressure
- Cushing reflex
 - Simultaneous onset of tachycardia and hypertension : indicator of impaired cerebral perfusion
 - *Br J Anaesth 2005,96(6), 791-9*
- Precipitants in patient with raised pressure: overhydration, narcotics, seizure
- Important in patient with shunt
- Compliance curve (pressure/volume) is exponential

Clinical Features of Syncope and Apnoea in Children and Infants

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Clinical Features

Two broad diagnoses on video-EEG: (n=110)

1. Attacks of collapse or limpness (37)
2. Attacks with prominent motor activity (73)

Duration of recorded attacks.

< 1 min: 5

1-30 mins: 97

>30 mins: 8

A gradual onset of event with fluctuating course and long ictus is unusual in epilepsy.

(Neurology 1991; 41: 1643-1646) 1

The Swoon

- Prolonged motionless collapse is not a prominent clinical feature of a seizure.
- May elicit response : resistance to eye opening
: visual fixations.
- Recording of alpha rhythm on EEG.

Issues of management

- Clear diagnosis: see event, home video
- Telemetry: show the video – need to capture event of concern
- Need to have family in agreement – description of event analysis
- Importance of avoiding anticonvulsants
- Avoidance of acute medication: is child safe?
- Need for team strategies as sudden loss of consciousness causes havoc at home and school

**Not all who stop breathing or moving
are seizing!**

**Some are fainting, some feigning and
some in big trouble!**

Think broadly