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NHS
Doncaster and Bassetlaw Teaching Hospitals
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# THE PEN

## Paediatric Education Newsletter


## 'Fits and Funny Movements' Issue

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## AN INFANT WITH FUNNY MOVEMENTS...

A 7-month-old boy presents with brief episodes of head drop and flexion of the body, followed by tonic extension of arms and legs. These last 1-2 secs and occur in clusters of 5-7 episodes, with several clusters per day. The movements are mostly seen on awakening or just before falling asleep. Birth history was uneventful and there is no family history of epilepsy. He has normal developmental milestones.

What is the diagnosis? What investigations will you do?

**Options**

**1) EEG**
**2) FBC, U+E, LFT, glucose, bone profile, magnesium**
**3) Lactate, amino acids, urine organic acids**
**4) Microarray comparative genome hybridization**
**5) All of the above**

**Answers below...**

## IDIOPATHIC INTRACRANIAL HYPERTENSION

**Papilloedema:**

A 14 year old girl is referred to you from the ophthalmologist with papilloedema. She has been having frontal headaches for the last 2 months which are worse when she lies flat or stoops down. Her MRI/V head is normal as is her neurological examination. Her LP is a champagne tap (go you!) with an opening pressure of 30cmH20.

**Questions (see page for answers):**

**1) What should you drain her CSF pressure to?**
**2) Does she have IIH?**
**3) True of false:**
**A: Prednisolone is the first line treatment for IIH**
**B: Treatment withdrawal can be attempted if asymptmatic for 4-6 months**
**C: Regular analgesia e.g. paracetamol or codeine may be useful**
**D: Acute visual impairment is an indication for discussion with neurology**

# ST VITUS'S DANCE
## CASE PRESENTATION

10 yr old boy, presented with sudden onset funny jerky movements of upper and lower limbs. Associated with dysarthria and ? tics. Otherwise fit & well with no history of trauma/fall.

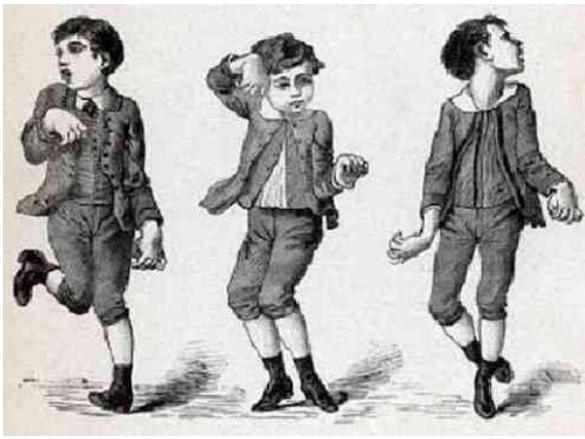
## An overview of movement disorders

- **Movement Disorders**
  - Pyrimidal syndromes
    - Spasticity
  - Cerebellar disorders
    - Ataxia
  - Basal Ganglia Disorders
    - Hyperkinetic
      - Essential Tremor
        - Rhythmic Oscillation of limbs
      - Chorea
        - Excessive irregular movement flowing from one limb to another
      - Tics
        - Stereotyped movements or vocalisations - may be temporarily suppressed
      - Myoclonus
        - Jerky movements
      - Dystonia
        - Sustained muscle spasms causing abnormal gait or movements.
    - Hypokinetic
      - Parkinsonism
      - Apraxia
      - Rigidity

## Syndenham Chorea

- APALS *
- Sydenham chorea
- SLE
- Telangiectasia

*(Anti-phospholipid antbody syndrome)



- Autoimmune / Inflammatory
- Drugs / Toxins
  - Alcohol
  - CO
  - Mercury
- Metabolic/ Endocrine
  - Hypo/hypercalcemia
  - Hyperthyroidism
  - Hypo/hypernatraemia
  - Hypoparathyroidism
  - Hypo/hyperglycaemia
  - Hypomagnesemia
- Chorea Causes
- Infectious
  - Encephalitis
  - Meningitis
  - Toxoplasmosis
- Hereditary
  - Huntington disease
  - PKU
  - Wilson's disease
  - Ataxia

### Symptoms

**Neurologic symptoms**
- Chorea, motor impersistence + interference with gross & fine motor control.

+

**Psychiatric symptoms**
Emotional lability, irritability, distractability, OCD. Occurs before/concomitant/after the onset of chorea.

+

Rheumatic Fever Symptoms

### Treatment

**Antibiotics**
Eradicate GAS + Prophylaxis after

**Symptomatic**
D2 blocking agents
eg Diazepam, Valproate, Levetiracetam

**Immune Modulation**
Corticosteroids, IVIG/ PLasmapheresis

### INVESTIGATIONS

- Test for GAS (Group A Strep) Throat Swab, ASOT, Anti-DNAse B
- CRP/ESR
  Expect to be normal/not significant. But is a diagnostic criteria of RF and if elevated indicates other cause ie SLE/encephalitis)
- ECHO & ECG
  (To rule out carditis)
- MRI/LP
  - to be considered if atypical presentation Hemichorea/fevers

# Answers and explanations

## An infant with funny movements...

**Diagnosis:** Infantile spasms. **Correct answer:** 5 (All of the above)

Infantile spasms are commonly seen in West Syndrome. This is a severe infantile epilepsy syndrome with a characteristic age of onset (2-14 months), and pattern of seizures and EEG changes. There is high morbidity (intellectual impairment, ongoing epilepsy, etc.) associated with infantile spasms.

The spasms typically consist of a sudden truncal flexion with stiffening of arms and legs but there can also be extension of the back, arms and legs. It may also be subtle, such as a head nod. The spasms can occur in clusters and multiple times a day. The child may have regressed or plateaued in their development.

Confirmation of the diagnosis requires an EEG. Hypsarrhythmia - (disorganized activity with high voltage slow waves and multifocal spikes) is the characteristic EEG finding in infantile spasms.

### Infantile spasms management algorithm

**Initial Assessment & Diagnosis**

- History & examination (including OFC, Wood's light, BP, developmental status)
- Urgent EEG (within 3 days) capturing sleep and awake periods confirming 'hypsarrhythmia' or 'modified hypsarrhythmia'
- Referral to pediatric neurology

↓

**First Line Treatment**

- Treatment to be commenced as soon as possible after confirmatory EEG
- Discuss potential adverse reactions of treatment with family
- Start combined treatment of
  - prednisolone 10mg four times daily
  - vigabatrin 25mg/kg twice daily
- Monitor BP, urine glycosuria, UE
- Take varicella serology pre-treatment
- Gastric protection with steroid treatment

↓

**Further investigations**

- Repeat EEG at 2 weeks
- Diagnostic investigations if cause of IS unknown performed in the following order until cause identified:
  - MRI brain
  - CGH array, serum lactate, serum amino acids and urine organic acids. Ophthalmology assessment (may be helpful in identifying a diagnosis)
  - Discuss further investigations with pediatric neurology team and this may include an Early Epilepsy Gene Panel.

### Idiopathic intranial hypertension

Answers:

1) <20cmH20
Opening pressure is raised if ≥ 25cmH2O (≥ 28cmH2O if sedated/obese). If it is raised but ≤ 40cmH2O -> drain to <20cmH2O. If pressure >40cmH2O: calculate 2/3 x (pressure - 20) and drain by this much. Never drain by >30cmH20.

2) Yes. See box below for diagnostic criteria

3)
A = false
The first line treatment is acetazolamide and second line is furosemide. Overweight patients should be encouraged to lose weight. Prednisolone or topiramate are other options but are used only alongside acetazolamide. Prednisolone may be used in the short term whilst awaiting surgery when there is high risk of visual impairment.

B = true
If the symptoms restart then restart treatment and consider repeating LP.

**Box 4: Diagnostic criteria of idiopathic intracranial hypertension (IIH)**

REQUIRED of the diagnosis of IIH

A. Papilloedema
B. Normal neurological examination except for cranial nerve abnormalities
C. Neuroimaging: Normal brain parenchyma without evidence of hydrocephalus, mass or structural lesion, and no abnormal meningeal enhancement on MRI.
D. Normal CSF composition
E. Elevated opening lumber puncture pressure (≥25CM CSF in not sedated and not obese) and (≥28CM CSF in sedated and obese children

[**Definite IIH** if A-E are satisfied] [**Probable IIH** if A-D are satisfied but CSF pressure is below defining criteria]

In the absence of Papilloedema the diagnosis of IIH syndrome can be made if B-E from above are satisfied, and in addition the patient has unilateral or bilateral Abducens nerve palsy.

If Papilloedema is absent, B-E are satisfied, IIH can be **suggested** if additional at least 3 of following neuroimaging criteria are satisfied

i. Empty Sella
ii. Flattening of posterior aspect of the globe
iii. Distention of perioptic subarachnoid space with or without tortuous optic nerve
iv. Transverse venous sinus stenosis

C = false
Patients should avoid excessive use of paracetamol, ibuprofen and codeine as these may contribute to medication overuse headache. If there is another co-morbid headache type e.g. migraine then prophylactic medications should be considered.

D = true
Do an urgent therapeutic LP to relieve pressure and discuss with neurology. Other indications for neurology discussion: refractory to 1st/2nd line treatments and opening pressures remain raised on 3rd LP, diagnostic uncertainty, evidence of cerebral venous sinus stenosis on MRV (may need surgical intervention).

# 'FIRST FIT' - WHAT SHOULD WE DO?

## Febrile convulsions

If child > 18m + fully recovered within an hour + you are sure of the diagnosis + source of fever known-> discharge with safety netting if parents happy

Recurrent febrile convulsions - only discharge if back to normal, definite diagnosis + source of fever known

SIMPLE FEBRILE CONVULSION
Seizure occurring in a child aged from 6 months to 5 years , precipitated by a fever arising from infection outside the nervous system, in a child who is neurologically normal and without complex features.

COMPLEX FEBRILE CONVULSION
Duration greater than 15 minutes, multiple (ie >than 1 convulsion per episode of fever), partial or focal.

Give relevant leaflets + first aid advice for if the seizure recurs + advice on managing pyrexia

Look for source of fever, if still febrile, consider anti-pyretics (although no evidence this reduces risk of further convulsions)

## Afebrile convulsions

Check for any injuries that may have been sustained during the seizure.

Precipitating factors: head injuries inc. NAI / toxins / drugs / infections / intercurrent illness / pregnancy?

FIRST SEIZURE
May be discharged if back to normal + normal neurology + no concerning features in hx/ex
- First fit clinic Paediatric OPD F/U must be arranged
-Give them a 'first fit' advice leaflet*

SEIZURE IN CHILD WITH KNOWN EPILEPSY
May not require admission if their 'usual seizure' + now back to normal + parents happy. Update the treating consultant.

Routine investigations: BM on everyone, ECG if first fit

Other Ix if indicated: FBC, Septic screen , U&E, Ca, Mg , toxicology, anti-epileptic levels, CT head

Taken from SCH guidelines
*RCPCH leaflet available from: https://www.rcpch.ac.uk/resources/safety-netting-information-following-first-seizure-without-fever-children-young-people#downloadBox

# EXTRA RESOURCES AND JOURNAL ARTICLES

For extra reading, and lots of useful guidelines and review articles follow the link below:

https://docs.google.com/document/d/1tJwyXGAlUG5PN95yn3lyf4XWU_cZ1kXUNXnGTuVMOI4/edit?usp=sharing