Is it Papilloedema?

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Aims

- Children/young people
- A bit about hypoplasia / NFL
- Why is it a problem?
- Background
- Mild optic disc changes
- Marked disc changes
- Testing for papilloedema
Optic nerve hypoplasia

Failure of development of optic nerve

OR

Abnormal loss of neurones early in pregnancy
Cupped optic discs (non-glaucomatous)

Late prenatal or perinatal cause: expanded occipital horns, loss of white/gray matter posteriorly. “Cupped” or atrophic optic discs
Optic atrophy
Look at nerve fibre layer – biomicroscopy and fundus photography
Why do we have a problem?

Pseudopapilloedema and headache are common
Raised ICP in children is unusual
Poor evidence base for investigation
No single gold standard test
Many cases left with various degrees of uncertainty
Anxiety

- Doctor
- Optometrist
- Family

Opportunistic screening without evidence base.
Background

Papilloedema a sign of raised intracranial pressure

- Idiopathic intracranial hypertension
- Obstructive
  - Space-occupying lesion
- Communicating
  - Endocrine
  - Drugs
  - Systemic disorder
Causes of ↑ ICP

Endocrine disorder
- Hyperthyroidism
- Hypothyroidism
- Vitamin D deficiency
- Hypoparathyroidism
- Adrenal insufficiency
- Hyperadrenalism
- Head trauma

Systemic
- Infections
  - Otitis media/mastoiditis
  - Sinusitis
- Venous sinus thrombosis 1°, 2°
- Systemic disorders
  - SLE
  - Protein malnutrition
  - Guillain-Barre syndrome
  - Iron deficiency anaemia
  - Leukaemia
  - Hypercoaguable states

Drugs
- Tetracycline
- Corticosteroid withdrawal
  - Inc topical for eczema
- Nitrofurantion
- Nalidixic acid
- Oral contraceptive
- Isoretinoin
- Thyroid replacement
- Growth hormone replacement
- Vasopressin
- Phenytoin
- Indomethacin
- Cyclosporin
Symptoms of ↑ ICP

Headache
- Characteristic

Vomiting without nausea

Visual disturbance
- Characteristic obscurations
- Diplopia
- less specific: photopsias / photophobia
- Vision loss

Back pain

Tinnitus
- pulsatile
*Useful* Symptoms of ↑ICP

**Headache**
- **Not** useful unless characteristic (too common)
  - Especially postural – worse on lying, better on standing
  - Worse on wakening, less during the day
  - Opposite of tension headache / migraine
(Vomiting without nausea)

**Visual disturbance**
- Characteristic obscurations
  - Momentary bilateral complete vision loss, not on standing up (this is postural hypotension)
- Diplopia – *if unequivocal manifest VI palsy present*
- Vision loss – *if profound disc swelling, unequivocal atrophy and/or haemorrhages/exudates*
Idiopathic intracranial hypertension

- Raised ICP with
  - Normal imaging
  - Normal CSF composition
  - Normal consciousness
  - Normal neurology
    - Except papilloedema/VI palsy
  - >70% not obese in children
Intracranial pressure

Upper limit of normal
- 20cm CSF – older child
- 13.5 cm H₂O < 5 years old
- 7.5 cm H₂O < 2 years old

Most under GA or sedated
- Effect on ICP not studied.

X mmHg = 1.36X cmH₂O
X cmH₂O = 0.736X mmHg
Papilloedema

In papilloedema there is an expanded subarachnoid space just behind the globe.

Raised ICP and papilloedema in an astronaut after prolonged weightlessness.
Is it MILD Papilloedema?
Papilloedema or pseudopapilloedema?
Is it MILD Papillodema?

Checklist
<table>
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<th>Pseudopapilledema with buried drusen</th>
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<td>Sharp peripapillary nerve fiber</td>
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Papilloedema or pseudopapilloedema?
Spontaneous venous pulsation

Video
SVP: Direct ophthalmoscopy

Very useful for SVP as greater magnification than biomicroscopy, but not done like this, and here child is too young except for red reflex check!
Absent SVP in pseudopapilloedema

25% vs 75% in normals

Bilateral pseudopapilloedema: elevated discs but no surrounding oedema

Ekdawi, Brodsky Rochester BJ O 201
Non-mydriatic fundus camera:

children >3y.o. find very easy, allows assessment and communication

Orthoptist or technician using non-mydriatic (no drops needed) fundus camera
OCT

- OCT of optic disc not useful
  - Too much variability in population
- RNFLA – retinal nerve fibre layer analysis
  - Useful in diagnosis and follow-up
  - Measures oedema around disc
  - Use same programme as glaucoma, but looking for ↑ not ↓ thickness
Developing papilloedema
Causes of pseudopapilloedema

- Hypermetropia
- Small discs
- Disc drusen
- Dysplastic discs

Exposed disc drusen
Disc drusen
Buried drusen

Daughter and Father

Exposed drusen
With age
Tests for Disc Drusen?
Tests for Disc Drusen
Disc drusen:
Red free with
Autofluorescence

Fong Bristol
Arch Dis
Child 2010
FAF : autofluoreseence with cSLO

488nm excitation
Barrier/emission >500-520nm

Disc drusen
- emission 520-520nm
U/S set to low gain confirms buried Disc drusen (here, in both eyes surprisingly)
Is it SEVERE papilloedema?
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Papilloedema
Severe Papilloedema: PRATs

Focal arrest in axonal transport: same as cotton wool spot
Severe Papilloedema:
Haemorrhage
Severe Papilloedema:
Haemorrhage

Macular star pointing to disc
Severe Papilloedema:

Haemorrhage

Haemorrhages and axonal transport arrest imply rapid ongoing permanent damage to vision.

There is usually a reversible element to vision loss if treated urgently.

Vessel obscuration on disc
Different questions depending on how obvious are the signs

Mild signs
- Is it normal or not?
- Is it pseudopapilloedema?

Obvious signs
- Could it be something different?
  - Eg ischemic optic neuropathy?
“It can’t be papilloedema”
"It can’t be papilloedema"

Vision loss without papilloedema, due to pre-existent obvious optic atrophy, in patients with known past history of raised intracranial pressure and papilloedema.
Comment

Most important diagnostic test is fundus examination

- Use photo as ancillary and help communication
- Direct ophthalmoscope for SVP
- Checklist

Next most important is history

- Symptoms
- Neurology
Referral
If there is papilloedema an urgent referral (I.E. SPEAK WITH THE HOSPITAL THAT DAY) is required.
Referral

- intended to be guidance about which conditions require emergency or urgent referral.
- You should follow relevant local protocols for referral.
- If a patient presents with a condition requiring an emergency referral you may wish to seek advice from the on-call ophthalmologist.
Headaches in general – common and worth learning about them
Need good source of information about headaches

- Eg International Headache society website
- PS non-specific headaches are very common in children (and adults)