Idiopathic intracranial hypertension (IIH), previously known as pseudotumor cerebri or benign intracranial hypertension, is a rare condition of increased intracranial pressure without any identifiable pathology. The clinical definition and association of this unique condition have evolved with time and the advances in neuroimaging making both the diagnosis and management challenging. Despite intervention, the clinical course of IIH is often prolonged and recurring with potential complications of distressing headache and blindness. The overall (children and adult) annual incidence of IIH is estimated to be 1-3 per 100,000 population, however the epidemiological data on childhood IIH to date is sparse and limited to hospital based retrospective case series. The principle objective of this BPSU study is to determine a contemporary national annual incidence of IIH in children. Furthermore, up to date clinical information will be collected to devise current best practice to guide clinicians in future management of paediatric IIH cases.

This BPSU surveillance will be undertaken for 13 months commencing in July 2007 with the study first appearing on the orange card circulated at the end of July 2007. Please report any new cases seen within that month.

The case definition is any child aged 1 to 16 years (not including 17th birthday) who fulfils at least two of the key features and all of the three essential criteria:

At least TWO Key Features:
1. Symptoms of raised intracranial pressure (such as headache, nausea, vomiting or irritability) and/or visual symptoms of diplopia, blurring vision or transient visual loss
2. Papilloedema, unilateral or bilateral
3. Raised opening cerebrospinal fluid pressure above 20 cm by lumbar puncture

And all THREE Essential Criteria:
1. Normal level of consciousness
2. Cranial imaging (including CT or MRI and CT or MR venography) does not reveal a structural cause such as ventricular dilatation, cerebral mass, vascular lesion or sinus venous thrombosis*, to explain the presenting symptoms or signs of raised intracranial pressure.
3. Normal cerebrospinal fluid contents (for atraumatic tap: white cell count < 6 x 10^6/L, protein < 0.4 g/L and glucose CSF/plasma ratio > 0.5)

Excluding: *Sinus venous thrombosis whose neuroimaging appearances can be difficult to distinguish from venous obstruction related to raised intracranial pressure. Please report if in doubt or if case was excluded due to sinus venous thrombosis.

Caution: Optic nerve head Drusen (a degenerative condition consists of hyaline deposits within the optic nerve head which results in an apparent elevation or swelling of the optic disc) can mimic papilloedema. However papilloedema and optic nerve head Drusen can occur concurrently, their differentiation can be made by optic ultrasound and/or orbital CT scan. If in doubt, please report suspected cases.

Paediatricians who have reported a case that meets the above case definition will be sent (1) a short questionnaire seeking demographic, clinical and initial management details and (2) a brief follow up questionnaire a year after initial diagnosis seeking data on subsequent management and the clinical outcome. All research study data sought are obtainable from medical records and there would be no additional investigation, intervention or direct contact with cases or their families.

This study, which has MREC, North East Wales NHS R&D and PIAG approval and is being run at the Paediatric Department, Wrexham Maelor Hospital in collaboration with the BPSU.

If you would like any advice regarding the eligibility of a particular case for inclusion in this study, or any other information about the study please contact:

Principal investigator: Dr Yim-Yee Matthews, Wrexham Maelor Hospital
Tel: 01978 291100 and E-mail: YIM-YEE.MATTHEWS@new-tr.wales.nhs.uk

Co-Investigators: Miss Fiona Dean, Dr Krystyna Matyka and Dr Karen McLachlan (University Hospitals of Coventry & Warwickshire), Dr William Whitehouse (Queens Medical Centre, Nottingham), Dr Cathy White (Morriston Hospital, Swansea), Professor Colin Kennedy (Southampton University Hospitals), Mr Guirish Solanki (Birmingham Children’s Hospital)