



## Highlights from the CSF Disorders Day, Queen Elizabeth Hospital Birmingham, 2015

- Dr Alex Sinclair welcomed the attendees to the CSF Disorders Symposium and stated that the meeting would be run on a bi-annual basis and hoped that that it would forge discussion and collaboration for future research on IIH.
- As well as representatives from IIH UK, the audience consisted of eminent Professors, Neurologists, Neurosurgeons, Radiologists, Headache Specialists and Scientists, some of whom had travelled from the USA.
- A consultant who manages one of the largest cohorts of IIH patients in the UK and is actively involved in a portfolio of IIH research studies stated that in an ideal world it would be advantageous for photographs to be taken of patients' optic disks at A&E when they are referred from Optometrists or present with headaches. This would enable experienced Ophthalmologists to examine the photographs as, on some occasions, people were automatically presumed to have IIH when, in reality, other factors contributed to their papilloedema.
- Permanent vision loss is the major morbidity associated with IIH. An early, hospital-based study that followed 57 patients for 5-41 years found that 24% developed blindness or severe visual impairment. Community and clinic-based studies have found a lower rate of severe visual loss of 6-14%.
- Optic Nerve Sheath Fenestration (ONSF) – whilst the procedure saves sight, it can on occasion, lead to significant complications including blindness in 1–2% of people. The risk of blindness increases further if additional ONSF is undertaken.
- Lumbar punctures are only a snap-shot in time and results are totally dependent on the patient's anxiety level, position and whether they were holding their breath at the time. It was stated that there could be elevated results giving a false positive indication of IIH but that exactly the same results could be obtained in a normal person if they were anxious and/or holding their breath too. It was stated that in an ideal world, ICP monitoring would be available for every patient to confirm an IIH diagnosis.
- IIH without Papilloedema (IIHWOP) was discussed and the consultants present agreed that it did exist but was rare and hard to diagnose. Referral was made to Friedman and Jacobson's proposal of an alternative set of criteria, derived from Smith's modified Dandy criteria.
- Why a randomised trial of surgical intervention in IIH could never happen. Because of shunt failures and infections, a trial for example on a particular LP shunt couldn't be evaluated as, more than likely, the shunt would fail and the patient may end up with a VP shunt or other surgery. Neurosurgeons spoke about shunts and preferences and one said that, because of poor failure rates with LP shunts, he would only insert a VP.
- The attendees discussed and agreed that the treatment of IIH has two major goals: the alleviation of headache and the preservation of vision and both were equally important for patients' quality of life. They also agreed that any future trial should work for an outcome to address both these factors.

- IIH UK Chair spoke to Consultant Neurologist Headache specialists from the National of Neurology London and Sheffield and stated that pain management for IIH was not being fully addressed for many patients. She offered IIH UK's help to push out surveys to followers of the Charity and support group members to collate any data that specialists required to use as pre-trial data or collate information. Sandra stated that many members of the support group had been diagnosed with other medical conditions such as Fibromyalgia and PCOS and that IIH UK could provide clinicians with numbers and details through surveys.
- Clinicians spoke about the differences and similarities of CSF disorders Hydrocephalus and IIH. International work is being undertaken to look at CSF modelling and flow using magnetic resonance elastography (MRE), a new way to image the body. It works by combining MRI imaging with sound waves to create a visual map (elastogram) showing the stiffness of body tissues. Current research is looking at how CSF flows and scientists believe it is pulled out of the choroid plexus. Bio-engineers are trying to understand the diseased brain and the efficacy of current therapies using elastography.
- IIH statistics - In the last decade, 24,600 people have been diagnosed with IIH in the UK. The rate of incidence in England in 2011 was 6.21 per 100,000. When analysed further, the figures show that IIH affects 1 in 100,000 people but the rate of incidence increases to 1-4 per 100,000 of women of child bearing age and increases to 19 per 100,000 with a BMI >30. The highest rates of incidence of IIH in the UK are in North East England and the London area.
- A scientist is looking at the cellular and molecular pathways involved in the pathophysiology of IIH (functional changes that accompany the condition). The research is building at cellular level the work of the choroid plexus (the area in the brain that produces CSF) to examine how CSF is secreted. The research will be looking at how existing medications (like Diamox) work on the cellular choroid plexus and then on exciting new molecules which have been found.
- At the School of Immunity & Infection, investigations are being made to look at peripheral adaptive immune cells with the healthy central nervous system. The focus is on examining whether cerebrospinal fluid and brain derived lymphocytes could potentiate neuroinflammatory disease. Bloods from trial volunteers will be tested for inflammatory markers in the IIH Weight Trial, currently recruiting in Birmingham.
- Should IIH be treated at specialist centres? This is something that IIH UK Chair Sandra Doughty will be discussing further with clinicians.
- IIH UK would like to collect data regarding patients' IIH diagnosing LP as we believe that general patient experience is poor.
- Clinicians were advised that, through our patient group, IIH UK could undertake questionnaires and surveys on their behalf to enable them to obtain data for any aspect of IIH research.

Feedback from attendees of the symposium was for further meetings to discuss future IIH research.