

IIH UK 2018 Megan Bhari Patient Conference



Our 2018 Patient conference was held on 7th July at the Park Inn by Radisson, West Bromwich, Birmingham. It was our first full day conference which was attended by 75 IIH Patients and their families. Shelly Williamson, IIH UK Chair, welcomed everyone to the conference and after some housekeeping announced the first speaker. Dr Deepa Krishnakumar, Consultant Paediatric Neurologist at Cambridge University Hospitals.

Dr Krishnakumar talked about Paediatric IIH, its pathophysiology and how it is poorly understood. She said that there has been no paediatric randomised controlled trails for IIH and that accurate diagnosis is essential due to the risk of visual failure in 10% of children.



She spoke about Cambridge's experience in treating children with IIH and referenced a paper outlining the pitfalls of diagnosing IIH in childhood which you can find at <https://onlinelibrary.wiley.com/doi/full/10.1111/dmcn.12475> she covered topics such as the Modified Dandy Criteria and the associations/secondary causes of raised intracranial pressure. She went on to say that symptoms she sees in the paediatric population are: Nausea/vomiting. Headaches which are 'novel' in nature. Diplopia, visual obscuration, transient visual loss, blurring of vision, photophobia and 'shimmering of lights with coloured centers' Lethargy, neck pain, tiredness, dizziness, mood change, tinnitus (52%) , sleep and behaviour changes. Papilloedema, sixth nerve palsy(9-48%) and visual field defects. She said that visual field loss or decreased visual acuity has been reported in 13-27% of cases.

She explained that when a child attends her clinic her approach is start with general examination which includes an eye examination, blood pressure, BP, height and weight - calculation of the BMI, ear, nose and throat examination and to look for signs of medical conditions which can cause secondary intracranial hypertension e.g. hypothyroidism. If IIH is suspected further investigations will be done including: Full blood count, urea and electrolytes, bone function tests, plasma glucose and thyroid function tests, Vitamin D, MRI, MRV and eye tests; OCT, fundal photographs and lastly if there is confirmed optic disc swelling an LP will be performed.

Dr Krishnakumar then went onto talk about intracranial pressure (ICP), treatment, long term outcomes and described the pitfalls in diagnosis and management. She explained that normal ICP depends on age, posture and clinical condition of child and that lumbar opening pressures do not always represent intracranial steady-state pressures. Although elevated steady-state ICP has been reported in up to 93% of IIH patients, some patients show periods of lower or indeed normal ICP between periods of marked intracranial hypertension. Treatment options mentioned are weight reduction, acetazolamide and topiramate. Surgical procedures like CSF shunt insertion, optic nerve sheath fenestration and stenting of the transverse sinus if a stenosis is found were also mentioned. Regarding long term outcomes she said that she saw persistent headache recurrence in 6-22% of children. Pitfalls were; absence of headache. Poor recognition of other headache disorders e.g. migraine. Absence of papilloedema. Normal weight/BMI. Failure to trial off medications. Optic disc swelling -

consider other causes. Lumbar puncture – indication, correct method and prolonged pressure recording. No evidence to say that IIH in the absence of papilloedema is a threat to vision.

Lastly she talked about how discovering more about paediatric IIH is a work in progress and outlined the current progression. 1. The East of England IIH pathway is now on the NHS networks website. 2. The British Paediatric Neurology Association IIH special interest group which held its first meeting in November 2017 of which IIH UK are part of. She went onto say that there is still much work to be done including a Best Practice National Guideline for paediatric IIH and Randomised controlled Trial of medications in children.



Next to talk was Sharon Whyte, Neurology Specialist Nurse, Cambridge University Hospitals. Sharon talked about the new Eastern Region Referral Pathway for suspected IIH in children. The pathway was developed to enable a quicker and easier diagnosis of IIH in children in the Anglia area and it is hoped that it can be rolled out to other areas too. Sharon explained that the pathway begins before referral to the Paediatric IIH Clinic at Cambridge. When a child is thought to have IIH the referring hospital should firstly review the history of each child, they should document their medications, pubertal status, Tanner scale, weight, height and BMI. An MRI should be performed including orbits/optic nerves and pituitary fossa, also an MRV to rule out venous sinus thrombosis. Sharon went on to explain that blood tests should also be performed, including FBC, U&E, LFT, BFT, Ferritin, Glucose, TFT, Vitamin D level and Lipid profile. If, after all of these tests, it is thought that the child does have IIH, the next step is for the referring hospital to discuss the case with Dr Krishnakumar or the neurologist on call at Addenbrookes, Cambridge. An urgent referral letter is then sent to the IIH Clinic and another to the neuro-ophthalmology clinic. If no optic nerve swelling or grade 1-3 and vision is not affected then a decision on LP/CSF infusion study is made with an ophthalmology review in 2-4 weeks' time. If optic nerve swelling is grade 3/4 +/- retinal Hemorrhages or the vision is compromised and an infusion study not available/unable to be organised urgently at Addenbrooke's then consider an urgent LP to reduce the pressure with ophthalmology review within 2-4 weeks.

Next to give their presentation was Dr Tara Walker, General Adult Psychiatrist, Worcestershire Health and Care NHS Trust. Dr Walker talked about Depression and its key features. She explained that depression affects 5% of the population annually, 1 in 4 women and 1 in 10 men. Risk factors include previous history and significant physical illness. She shared a couple of websites that anyone can use for self-help whilst suffering with low mood; Living life to the full www.lttf.com and Mood Gym <https://moodgym.com.au>. Dr Walker then spoke about other treatments for depression; a combination of antidepressant medication and psychological intervention and said that Gp's should refer patients with moderate to severe depression to psychiatric services. She then spoke about electroconvulsive therapy (ECT) and ended her presentation by saying the average episode of depression is 6-8 months but that as many as 80% may have a relapse. Some delegates asked questions and shared their experiences of living with depression whilst having a chronic illness, this was the first time that we've had speaker talk about Mental Wellbeing at our conference, mental health is an important subject and we were delighted to have Dr Walker with us this year.





After a short break Sister Julie Edwards, Headache Specialist Nurse, Birmingham spoke next. Her topic was Headache in IIH. She explained that Headache is the most common symptom affecting 94% of IIH Patients. Other symptoms reported are visual obscurations 72%, pulsatile tinnitus 60%, back pain 53%, dizziness 52%, neck pain 42%, Visual loss/blurring 32%, memory disturbance 20% and lastly double vision 18%. She explained

that headaches in IIH are very variable, but are a big issue for patients she also went onto say that Headaches in IIH are often complex and non-specialist doctors are not always well informed about how to recognise and deal with them, she also mentioned that headache in IIH can affect a patients quality of life.

Sister Edwards then went onto talk about the different types of headache that an IIH Patient can experience; migraine, high pressure, low pressure, menstrual related and medication over use. Delegates learnt that having an LP improved headache in 72% of patients but that it is short lived and that regular LP's are not recommended due to the potential side effects of LP (Discomfort, low pressure headache, back pain) IIH Improved with LP (72%) they also learnt that getting relief from an LP does not necessarily mean that the headache was caused by high pressure as 25% of patients that do not have IIH also get relief after having an LP.

She then talked about how a patient is assessed, how a detailed history about headache will be taken as this can help evaluate the type of headache. It is not uncommon for more than one type of headache to exist and some IIH patients may ALSO have migraine. She went onto talk about migraine and explained some symptoms of migraine; photophobia (intolerance of bright light) phonophobia (intolerance of loud noise), osmophobia (hypersensitivity to odors), pounding, vomiting and nausea. She explained that migraine is common, affecting 1 in 7 of us. Sister Edwards spoke about the phases of migraine; tiredness, yawning, increased urination and cravings and about the different auras that someone with migraine might experience; Visual - zig zag Lines, visual hallucinations, scotoma (central vision missing) hemianopia (loss of vision in half of visual field), broken vision. Sensory – pins and needles and/or numbness. Motor – one sided weakness. Speech – slurring, aphasia (inability to process language) and lastly Vertebrobasiliar – Vertigo and dizziness. She spoke at length about how migraines are treated then went onto speak about medication overuse headache. Medication overuse headache can occur in anyone using pain killers more than 2-3 times a week so anyone using pain killers more often than this that has a dull, continuous daily headache may be suffering with medication overuse headache and should mention this to their doctors to get it addressed.

Dr Ruchika Batra, neuro-ophthalmologist, University Hospitals, Birmingham. spoke at our patient conference in Scotland last year to give a 'Holistic Approach to Weight loss' talk. Delegates enjoyed it so much that we decided to invite her along again. As last year, everyone enjoyed her talk very much. Whilst we realise that weight is an emotive subject among the IIH community we felt it was important for IIH patients to understand why weight loss is important for those people with IIH and have problems controlling their weight.



Dr Batra's presentation started with the symptoms of IIH and the emotions that IIH patients suffer as a result. She talked about how being positive towards the idea of weight loss is a big step in realising that losing weight has many other benefits and not just to help alleviate IIH symptoms. She made clear the fact that she realises that not all people with IIH are overweight but the fact remains that around 90% of people with IIH are obese females, researchers are still unsure as to why this is but research has shown that weight loss can alleviate papilloedema and headache in IIH patients and has even 'cured' some people with IIH which is why IIH patients are told to lose weight. One of the slides from her presentation was titled 'Diets make you fat and crazy' which was met with a ripple of agreement from those attending. She went on to explain why diets can be ineffective for weight loss, that dieting consistently leads to weight gain, that dieting intensifies cravings, it leads to food obsession and emotional distress and can also lead to binge eating. So how do we solve this crisis? Ruchika took us on a history lesson from cavemen to Elizabethan times to the 1950's to the present day to help us find out. There was lots of interaction with the audience throughout this part of her presentation and she set them this task. 100% of delegates chose the meal plan on the right as the best plan to help them lose weight. We will return to this later.



Ruchika then went on to talk about the different food groups and why they are important to include them all. Delegates learnt that carbs are an important source of energy, B vitamins and fiber. Protein is important for growth and maintenance of body structure and they make you feel full for longer. Dairy products are an important source of vitamins, minerals and protein. Fruit and vegetables provide vitamins and minerals (antioxidants) and increases fiber intake. Fats are a source of fat soluble vitamins; the 3 main sources of fat are saturated from animal products, polyunsaturated fats from vegetable oils and seed and monounsaturated fats from olive oil, nuts. Portion size was discussed which led onto 'should we cut calories'? She explained that when we cut calories our bodies enter starvation mode in which fewer calories are burnt but once we increase our calorie intake the starvation mode persists causing us to gain weight, it's a vicious circle.

Ruchika then talked about what happens when we eat sugar, whether it is sugar from carbohydrates, refined or artificial. She explained that sugar causes an insulin spike and it is insulin that makes us fat. Delegates learnt that sugar is 8 times more addictive than cocaine and the only way to tackle it is to cut out sugar all together, even the artificial ones. Most importantly delegates learnt about the importance of set mealtimes. Breakfast, lunch and dinner. Always sit at a table, eat your meals as if you are being watched by your boss/future mother-in law. Eat slowly as this give your stomachs stretch receptors time to work. From the first mouthful it takes around 20 minutes for those receptors to work, eating slowing allows this process to happen before you have over eaten. Also think about the food you are eating and enjoy it, always drink water whilst eating as this too will stop you eating too much.

Her presentation was rounded off with a review of the 'Pick a meal plan' task that she mentioned earlier. Both plans were 1940 calories each. One was for 3 meals a day consisting of breakfast, lunch and dinner. The other was more familiar to anyone on a diet; it consisted of breakfast, snack, lunch, snack, dinner, and snack. Both meals have identical calories and delegates were very surprised to hear that the plan of 3 meals a day was inductive to losing weight. The reason for this is that every time you eat your sugar level rises which in turn makes your insulin rise and it is insulin that makes fat. So in conclusion

delegates decided that it was better to eat 3 home cooked healthy meals (yes, including the fats and sugars) 3 times a day than it is to eat processed 'diet' foods that are inductive to snacking.



First up after lunch was Miss Susan Mollan, neuro-ophthalmologist, University Hospitals Birmingham who talked about the new IIH Guidelines.

Over a period of 25 months a specialist interest group including neurology, neurosurgery, neuroradiology, ophthalmology, nursing, primary care doctors and patient representatives from IIH UK met. An initial UK survey of attitudes and practice in IIH was sent to a wide group of physicians and surgeons who investigate and manage IIH regularly. A comprehensive systematic literature review was performed to assemble the foundations of the statements. An international panel along with four national professional bodies, namely the Association of British Neurologists, British Association for the Study of Headache, the Society of British Neurological Surgeons and the Royal College of Ophthalmologists critically reviewed the statements.

Over this period of time 20 questions were constructed: one based on the diagnostic principles for optimal investigation of papilloedema and 21 for the management of IIH. Three main principles were identified: (1) to treat the underlying disease; (2) to protect the vision; and (3) to minimise the headache morbidity. Statements presented provide insight to uncertainties in IIH where research opportunities exist. In collaboration with many different specialists, professions and patient representatives, we have developed guidance statements for the investigation and management of adult IIH.

You can read the open access document here:

<https://jnnp.bmj.com/content/early/2018/08/31/jnnp-2017-317440>



Next up to speak was Dr Alexandra Sinclair – IIH UK Patron, Consultant Neurologist, University Hospitals Birmingham. NIHR Clinician Scientist, Institute of Metabolism and Systems Research, The University of Birmingham. Dr Sinclair began her presentation by talking about the importance of each participant in clinical trials and how they are working towards developing novel therapies for IIH, she told the audience how Hospital Statistic Data showed that IIH incidence and admissions were climbing proving the need for better therapies. Dr Sinclair then went onto give a 'Research Round up' on the various trials past and present.



Firstly she spoke about the IIH:Pressure trial (full name IIH:Pressure: The acute and chronic effects of gut neuropeptides on intracranial pressure regulation) which is currently recruiting IIH Patients. This trial aims to use a new method of pressure monitoring to measure the brain pressure continuously without lumbar puncture and to test if a new drug (Exanatide) can change brain pressure. It is a randomised, double-

blind, placebo-controlled trial in patients with IIH and so those who are accepted onto the trial will be put into one of two groups which will be decided at random. The primary outcome of the trial is ICP and they will use a novel approach to measuring ICP with implanted telemetric probes which allow continuous, non-invasive recording of ICP once implanted, without the need for lumbar punctures. Secondary outcome measures include ocular biomarkers, headache and quality of life.

Patients will have a 1 month run-in period before surgery to implant the probes, and then a 24 hour baseline stay where they will be randomised to take drug or placebo. ICP will be recorded continuously during the visit. The participants will then take the active drug or placebo for 3 months before a final study visit. This study has two parts. The first part of the study includes participants having telemetric intra-cranial pressure sensors fitted. Participants are randomised to one of two groups.

Those in the first group receive exenatide through skin injections twice daily for 12 weeks. Those in the second group receive a placebo (a dummy medication) also given through skin injections twice daily for 12 weeks. Participants are followed up at two and 12 weeks with intracranial pressure recording (ICP) which is a non-invasive monitor, as well as blood tests, headaches scores and cognitive (mental) testing). At 12 weeks, participants are assessed for their quality of life, and clinical measurements. The second part of the study randomly allocates participants to receiving one of five medications for two weeks (with one week washout between them).



Dr Sinclair then spoke about the IIH:WT, she said that they have recruited enough people with IIH but a small number of people without IIH are still required. (please get in touch if you would like more information). The IIH weight trial is a randomised controlled trial of bariatric surgery versus a community weight loss program for the sustained treatment of IIH. The aim of this trial is to assess if weight loss through bariatric surgery and/or dietetic intervention is an effective sustainable treatment for IIH, with sustained reduction of Intracranial Pressure (ICP), visual symptoms and headaches.

She went on to explain that they conducted a randomised controlled parallel arm trial where participants were randomised 1:1 to a bariatric surgery pathway or to a community based Weight Watchers diet programme. Sixty four participants (32 to each arm) were randomised.

Suitable patients were identified at NHS Trusts across the country. Participants randomised to the bariatric surgery arm were referred to their local bariatric surgery pathway according to NICE guidelines; participants randomised to the Weight Watchers arm were enrolled in their local Weight Watchers group where they were given vouchers at baseline, 3, and 6 months that exempt them from paying for consecutive and specified weeks of their local Weight Watchers. They were also enrolled into their initial meeting. Attendance at the groups is being monitored through participant self-reporting.

Dr Sinclair explained that the primary outcome measure is the change in intracranial pressure between baseline and 12 months. There will also be a battery of secondary and exploratory outcome measures that will be used to further investigate the treatment and pathogenesis of IIH and participants will be followed up for 5 years. Namely; visual tests, cognitive tests and sleep apnoea tests.



Dr Sinclair spoke briefly about the IIH:Life Registry and how it is slowly being rolled out to neurological hospitals across the UK. The first phase of the registry was rolled out in Birmingham and Moorfields London for patients newly diagnosed with IIH. When it is fully up and running, forty UK hospitals will be participating. She explained how the database comprises of two parts – one for Neurologist/Neuro Ophthalmologist entries and the other for patient input. The medical professionals will be entering data which will include: visual test results, lumbar puncture opening pressures, medication (including dose), height, weight etc. The patient will be asked about their pain levels and asked to complete an annual Quality of Life survey.

She stated that she hoped that the registry would be able to indicate the optimal treatment for: 1. Sight Preservation 2. Managing Headaches 3. Improving IIH Sufferers' Quality of Life.

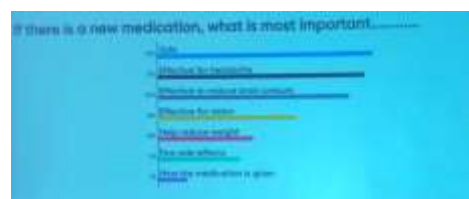
Dr Sinclair and her team will be looking to see which treatments have better outcomes and will look at how different hospitals treat IIH. The team anticipates that the data will help develop national evidence based standard for IIH management and provide a standard for care across the country. The IIH Life Registry will improve understanding of IIH as very little has been published on long-term disease outcomes and disability in IIH and will improve patient care and safety. It will identify patients at risk of poor outcomes and counsel patients and guide future research design. By involving patients the research will ensure patients' views and perception of care, play a central role in future IIH management.



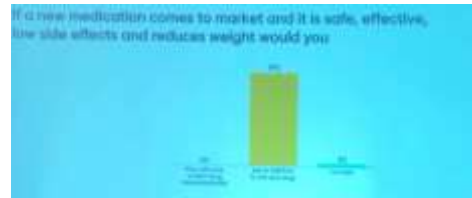
Next was the IIH:DT. The IIH Drug Trial is a randomised controlled trial of the drug AZD4017 vs placebo for treatment of IIH. It is a double blind placebo controlled randomised (1:1) phase II trial to assess the efficacy, safety, and tolerability of AZD4017 400mg given twice daily for 12 weeks in the treatment of IIH. Women aged 18-55 with active papilloedema, ICP>25 and normal brain imaging were recruited at the Queen Elizabeth Hospital, Birmingham, Walton Centre, Liverpool and Southern General Hospital, Glasgow. Participants were randomised to AZD4017 400mg twice daily for 12 weeks or placebo. Thirty one participants (against a target of 15 to each arm) were randomised. The aim of the IIH:DT trial is to assess if AZD4017 is an effective and safe treatment for IIH, with reduction of ICP, visual symptoms and headaches. The primary outcome measure is the change in intracranial pressure (ICP) between baseline and 12 weeks. There are a battery of secondary and exploratory outcome measures that will be analyzed to further investigate the safety and efficacy of AZD4017. Participants were followed up at Weeks 1, 2, 3, 4, 6, 8, 10, 12, and 16. The IIH:DT follow up has now finished recruiting and the results will be made available once the data has been analyzed.

Dr Sinclair's ended her presentation with audience participation by way of a live interactive session. Delegates were asked to go to a link that she provided and rank in order of importance the following question: 'If there is a new medication, what is most important'. The options that patients and carers ranked in order of importance were:

- 1, That a new medication is safe 22%.
- 2, That it is effective for headache 21%
- 3, That it is effective at reducing brain pressure 20%.
- 4, That it helps vision 14%.
- 5, That it help reduce weight 10%.
- 6, That it has few side effects 9% and lastly
- 7, How the medication is given 3%.



The next question was 'If a new medication comes to market and it is safe, effective and has low side effects would you? A, stay with your current drug 0% B, ask to switch to the new drug (97%) C, not sure 3%. As you can see almost all delegates said they would like to switch to the new drug. Shelly then thanked Dr Sinclair and spent 10 minutes answering questions from the audience.



Ben Thomas from Bbraun gave a talk all about shunts. He discussed the mechanics of shunts and valves and delegates were impressed to learn about how their shunts worked. It was the first time we had a guest speaker talk about shunts and his presentation was very well received.

The last talk of the day was about our 18 month long collaboration with the James Lind Alliance to find the IIH Top questions that research has not yet addressed. Our Research Rep Krystal Hemmings was the patient lead for this project but as she could not make this year's conference this talk was to be given by IIH UK Assistant Research Rep, Amanda Denton. Unfortunately Amanda was too ill to travel so Dr Sinclair, as clinical lead, kindly stepped in to do this presentation. Firstly she spoke about why we decided to undertake this Priority Setting Partnership. IIH is currently under-researched and treatment strategies vary widely across the UK. A Cochrane review showed insufficient evidence to recommend or reject any current treatments for treating people with IIH and in those with severely affected vision, surgery may be indicated.



She went onto say that for the majority it can be a chronic condition with increased healthcare costs, impacting quality of life and the ability to work. She reiterated the need for relevant and focused research that results in the best outcomes for individuals with IIH and those treating them.

You can read the IIH PSP report here: <https://www.iih.org.uk/images/IIH-PSP-top-10-report-final.pdf>

A selection of photographs from the day.

