Case Study: Pediatric Pseudotumor Cerebri (Idiopathic Intracranial Hypertension)

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Idiopathic pseudotumor cerebri (iPTC) cerebri is a condition in which patients present with symptoms and signs of increased intracranial pressure, without evidence of a mass, lesion or cerebrospinal fluid (CSF) obstruction on neuroimaging. These patients have a lumbar puncture that documents high CSF pressure with normal CSF composition. In those patients where an etiology is identified, the term “idiopathic” is dropped, and the patient is said to have pseudotumor cerebri (PTC) secondary to the identified etiology.

The demographic characteristics of pediatric pseudotumor cerebri are different than the typical adult patient. In adults, iPTC has been characterized in obese women of childbearing age. However, in children, iPTC is rare and the clinical profile has been suggested to be very different. In the prepubertal child, there seems to be less of a gender predilection as well as a lack of a clear association with obesity. On the other hand, after the onset of puberty pediatric iPTC cases tend to align with the demographic characteristics of adult patients with iPTC. There are a few studies that have analyzed the clinical differences between adult and pediatric cases of iPTC. We reviewed our own cases of pediatric PTC to determine their demographic and clinical characteristics. The following case studies outline the profile of two obese pediatric patients, one a teen female and the other a young male, who both presented with similar symptoms.
Pseudotumor Cerebri Case Study 1

**Patient:** 17-year-old obese female

**Presentation:** Admitted through the Emergency Department complaining of headaches and blurred vision, which was worse in the right eye. She also had mild/moderate neck pain radiated or extended to the right arm. She was referred to ophthalmology due to the vision complaints.

**Eye Exam:** On examination her visual acuity in the right eye was 20/40 and the left eye was 20/20. Visual field testing of her peripheral vision revealed enlargement of the physiologic blind spot in both eyes. She also had marked swelling of both optic nerves. The patient was admitted for an MRI and MRV and a lumbar puncture was performed. The pressure in her spinal fluid was found to be high measuring 38 cm of H20. She was diagnosed with increased intracranial pressure or pseudotumor.

**Treatment and Outcome:** Acetazolamide was started and over the next six months her headaches, visual acuity and visual field testing improved. The swelling of the optic nerves resolved. She continues to take acetazolamide to control symptoms and is monitored with routine eye and neurologic exams.

1a and 1b: Right and left eyes respectively, showing swollen optic disc, indicating pseudotumor cerebri.

2a and 2b: Right and left eyes respectively, showing near normal optic discs post treatment at Nationwide Children's Pediatric Pseudotumor Cerebri Clinic.

Pseudotumor Cerebri Case Study 2

**Patient:** 6-year-old obese male

**Presentation:** Admitted through the emergency department complaining of headache and visual loss.

**Examination:** On examination it was noted that he was extremely obese for his age, at 82 pounds, which was over the 95th percentile for his age. He had marked optic disc edema and almost complete bilateral visual loss. His cranial MRI and MR venogram were normal, and a lumbar puncture confirmed markedly elevated cerebrospinal fluid pressure.

**Treatment and Outcome:** The patient was prescribed the medication, acetazolamide and underwent bilateral optic nerve sheath fenestration procedures without significant improvement in his vision. However, after placement of a ventriculoperitoneal shunt he regained near normal visual function, and avoided any permanent visual impairment. One year later he had only occasional headaches and his vision remained normal.

Setting the Pace for Comprehensive Pediatric Care

The recognition of idiopathic intracranial hypertension (pseudotumor cerebri) in children has increased dramatically in recent years and Nationwide Children's is leading the response to this trend with the development of a multidisciplinary clinic for the specialized diagnosis, treatment, and ongoing care of these unique children.

Managed by board-certified child neurologists and pediatric ophthalmologists who have a special interest in the disorder, the Idiopathic Intracranial Hypertension (Pseudotumor Cerebri) Clinic at Nationwide Children's is the first in the country to provide comprehensive care solely for children.

Additional members of the care team may include neuro-ophthalmologists, pediatric neurosurgeons, endocrinologists, radiologists, and other specialists.
Tracking Data

One important development of the Intracranial Hypertension Clinic at Nationwide Children’s is data collection and analysis of patients. For example, all patients seen by Ophthalmology from August 2005 to July 2008 who were given the principal diagnosis of pseudotumor cerebri were considered for inclusion in the study (N = 131). Patients were excluded from the analyses if they were seen only for a diagnostic visual field or the patient was seen only for a single visit. The study was approved by the Institutional Review Board of Nationwide Children’s Hospital and was conducted in accordance with HIPAA guidelines.

Patient criteria for the diagnosis of iPTC included those who presented with signs or symptoms of increased intracranial hypertension, normal neuroimaging, and normal cerebrospinal fluid studies except for an elevated opening pressure, and no other identifiable cause. If the data indicated characteristics that offered another etiology, it was considered to be a case of secondary pseudotumor cerebri. The iPTC patients were divided into a pre-pubertal group and pubescent/ post-pubescent group based on age. For the Intracranial Hypertension Clinic study, the onset of puberty was 11 years old for girls and 13 years old for boys. Each patient’s BMI was calculated and similarly plotted on age-matched percentile charts. Patients were defined as obese if their BMI was greater than 95 percent, as overweight if their BMI was between 85 and 94 percent, and as normal if their BMI was under 85 percent. Evaluation of treatment response was based on follow-up symptoms, optic nerve edema, and/or CSF pressures on repeat lumbar puncture.

Diagnosis and Treatment

Pseudotumor cerebri is suspected after a child with headaches is found to have optic disc edema. The diagnosis is then confirmed after neuroimaging studies eliminate a brain tumor or intracranial venous thrombosis and after a lumbar puncture confirms the elevated pressure. Once the diagnosis is confirmed, the child’s visual function is closely monitored, and any underlying risk factors are eliminated. Visual loss from pseudotumor cerebri can be either rapid or insidious, and the severity of headache or other symptoms does not reliably correlate with the risk of visual loss. Physicians encourage early referral of any children with optic nerve edema or other signs of pseudotumor cerebri.

Several medicines are available to reduce the intracranial pressure, and if these are ineffective, surgical procedures to lower the intracranial pressure or to prevent pressure damage of the optic nerve are employed. The medications are designed to lower the pressure of the CSF and decrease the swelling in the optic nerve. The three major groups of medications used include carbonic anhydrase inhibitors, steroids, and diuretics.

If the patient does not respond to medications and vision worsens, surgery is often recommended to reduce the pressure around the optic nerve and brain. There are two types of surgery available, an optic nerve sheath fenestration or a spinal fluid shunting.

Optic Nerve Sheath Fenestration

An optic nerve sheath fenestration cuts a window in the membrane that surrounds the optic nerve. This allows fluid to escape and relieves the pressure on the nerve itself. In addition, with time the sheath will scar down in the area directly behind the eye and prevent further episodes of high pressure from causing damage to the optic nerve. This stabilizes and in many cases, improves vision. The procedure may be done on one or both eyes.

Spinal Fluid Shunting

Spinal fluid shunting is a surgery that inserts a thin tube called a shunt into the brain or spinal cord that helps direct or drain excess fluid and lower intracranial pressure. This tube is usually directed into the abdomen where the fluid is reabsorbed into the body. This procedure may need to be revised over time to ensure that the shunt keeps working properly.

If the pediatric patient is obese, weight loss is recommended for the patient’s overall health, as well as to help decrease the symptoms of pseudotumor.

Nationwide Children’s supports research into this unique problem, and The Research Institute at Nationwide Children’s Hospital works with academic partner, The Ohio State University College of Medicine. There is much research to be done regarding these patients. Nationwide Children’s will now be able to capture data and monitor this population with the opening of the Intracranial Hypertension Clinic. Specifics, such as genetic predisposition will be tracked. If this is a congenital disease, it will be noted if parents show signs and symptoms as well. There are a variety of factors that will be monitored on these patients in the months and years to come so that physicians can better understand and treat this unique disorder.
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At Nationwide Children’s, we are creating the future of pediatric health care. We consider every detail. Every decision. Every aspect of the care we provide. From the child who comes to us with Duchenne muscular dystrophy, spinal muscular atrophy, or refractory epilepsy. To those with a sprain, a broken bone, or a fever. Here, the future health and potential of all children is being shaped. Here, our doctors are revolutionizing your child’s health and the health of future generations. Learn more at NationwideChildrens.org.

Referrals and Consultations
Like all of the specialized Neurosciences clinics at Nationwide Children’s, the Intracranial Hypertension Clinic accepts referrals from across the country.

Online: NationwideChildrens.org
Fax: (614) 722-4000
Phone: (614) 722-6200 or 1(877) 722-6220

Physician Direct Connect Line for 24-hour urgent physician consultations:
(614) 355-0221 or 1(877) 355-0221

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