

Case report

doi:10.5633 / amm.2027.0121

A CASE OF A CHILD WITH BENIGN INTRACRANIAL HYPERTENSION AS A CONSEQUENCE OF A PROLONGED DIETARY MANAGEMENT

Marija Veselinović¹, Aleksandar Veselinović², Marija Trenkić¹, Dragan Veselinović² Suzana Branković³

¹University Clinical Center of Nis, Ophthalmology Clinic, Niš, Serbia

²Eye Clinic Veselinovic, Niš, Serbia

³Military Medical Center Karaburma, Beograd, Serbia

Contact: Marija Veselinovic

48 dr Zorana Djindjića Blvd., 18000 Niš, Serbia

E-mail: drmarija.veseliovic@gmail.com

Benign intracranial hypertension has been described as pseudotumour cerebri, 'false brain tumour', or idiopathic intracranial hypertension. Pseudotumour cerebri occurs in cases of increased intracranial pressure with no obvious reasons.

Subjective symptoms and the fundus finding indicate the condition with increased intracranial pressure.

Benign intracranial hypertension rarely occurs in children, so it requires urgent diagnostics and exclusion of multiple neurosurgical disorders.

This paper presents a case of a 14-year-old girl who followed a rigorous diet, had hormonal imbalance, a cessation of a menstrual period, and clinical manifestation of pseudotumour cerebri. The discontinuation of the diet and oral application of carbonic anhydrase inhibitor for several months contributed to the loss of subjective problems and recovery of clinically visible symptoms in the forms of diplopia and the fundus changes.

Key words: benign intracranial hypertension, pseudotumour cerebri, child, diet

SLUČAJ DETETA SA BENIGNOM INTRAKRANIČNOM HIPERTENZIJOM KAO POSLEDICA PRODUŽENOG ODRŽAVANJA DIJETE

Marija Veselinović¹, Aleksandar Veselinović², Marija Trenkić¹, Dragan Veselinović² Suzana Branković³

¹Univerzitetski klinički centar Niš, Klinika za oftalmologiju, Niš, Srbija

²Očna klinika Veselinović, Niš, Serbia

³Vojnomedicinski centar Karaburma, Beograd, Serbia

Kontakt: Marija Veselinovic

Bulevar dr Zorana Đinđića 48, 18000 Niš, Srbija

E-mail: drmarija.veseliovic@gmail.com

Benigna intrakranijalna hipertenzija je opisana kao pseudotumor mozga, „lažni tumor mozga“ ili idiopatska intrakranijalna hipertenzija. Pseudotumour cerebri se javlja u slučajevima povećanog intrakranijalnog pritiska bez očiglednih razloga.

Subjektivni simptomi i nalaz fundusa ukazuju na stanje sa povišenim intrakranijalnim pritiskom.

Benigna intrakranijalna hipertenzija se retko javlja kod dece, pa zahteva hitnu dijagnostiku i isključivanje višestrukih neurohirurških poremećaja.

U radu je prikazan slučaj devojčice od 14 godina koja je bila na rigoroznoj ishrani, imala je hormonski disbalans, prestanak menstruacije i kliničku manifestaciju pseudotumora mozga. Višemesečni prekid dijetе i oralna primena inhibitora karboanhidraze doprineli su gubitku subjektivnih tegoba i oporavku klinički vidljivih simptoma u vidu diplopije i promena fundusa.

Ključne reči : benigna intrakranijalna hipertenzija, pseudotumor mozga, dete, dijeta

INTRODUCTION:

Although benign intracranial hypertension is uncommon in pediatric clinical practice, it can be a diagnostic, as well as therapeutic challenge.

Symptoms associated with benign intracranial hypertension may be similar to those of a brain tumour, but the tumour cannot be detected. It is commonly seen in adult, obese women, 35-40 years of age. (1, 2, 3)

The exact mechanism of bilateral optic nerve edema occurrence in obese population, especially females, has not been explained yet. Benign intracranial hypertension in pediatric patient is uncommon, so it requires detailed examinations and ruling out potential intracranial changes that need neurosurgical treatment (4,5).

The best approach, both in diagnosing and treating the children with benign intracranial hypertension, is a multidisciplinary one, including an ophthalmologist, pediatrician, pediatric neurologist, nutritionist, radiologist, and neurosurgeon.(6)

Having in mind the fact that there is no obvious reason that causes intracranial pressure increase, this syndrome has also been described as idiopathic intracranial hypertension. Increased intracranial pressure causes papilla stagnans, optic nerve head swelling, identical to one of a brain tumour.

Within the clinical manifestation of pseudotumour cerebri, there are characteristic symptoms, such as headache, blurred vision, tinnitus, diplopia, and nausea. (7,8).

The signs of optic disc edema may last for months, even years, but in most cases they withdraw spontaneously. Apart from characteristic ophthalmic manifestation of papilla stagnans, optic disc edema, the degree of initial edema and its monitoring can be done by using optical coherence tomography. Visual acuity is preserved in most cases, or minimally reduced, and patients often experience visual obscurations daily. (8,9).

Enlargement of the blind spot, as well as peripheral isopter constriction, can be registered in the visual field, and fundus fluorescein angiography reveals intense fluorescein leakage in optic disc area.

Medical therapy by administration of acetazolamide can greatly improve condition in children with pseudotumour cerebri. It has been proved that administration of acetazolamide reduces cerebrospinal fluid production and results in symptom improvements in 47-67% cases. (10).

Case report:

A week before coming for ophthalmic examination, patient P.M., 15 years of age, had blurred vision in both eyes lasting a few minutes after getting up from the bed. These disturbances were associated with strong, throbbing headaches in the frontal region that did not go away after taking pain killers. Seven days after the initial problems she noticed double vision and left eye turning inwards. She came for an ophthalmic examination and was recommended to be admitted to hospital for further examination and treatment (Figure 1).



Figure 1. Paresis of external rectus lateralis muscle of the left eye with the presence of paralytic convergent strabismus

Visual acuity in both eyes was 1.0, IOP values: 14mmHg, and there was a bilateral manifestation of papilla stagnans on the fundus, with prominence of 1 dpt.

Fluorescein angiography showed a finding characteristic of papilla stagnans. An enlargement of the blind spot and reduced peripheral contrast sensitivity were recorded in the visual field of both eyes. Visual evoked potentials finding was regular in both eyes. **(Figure 2,3).**

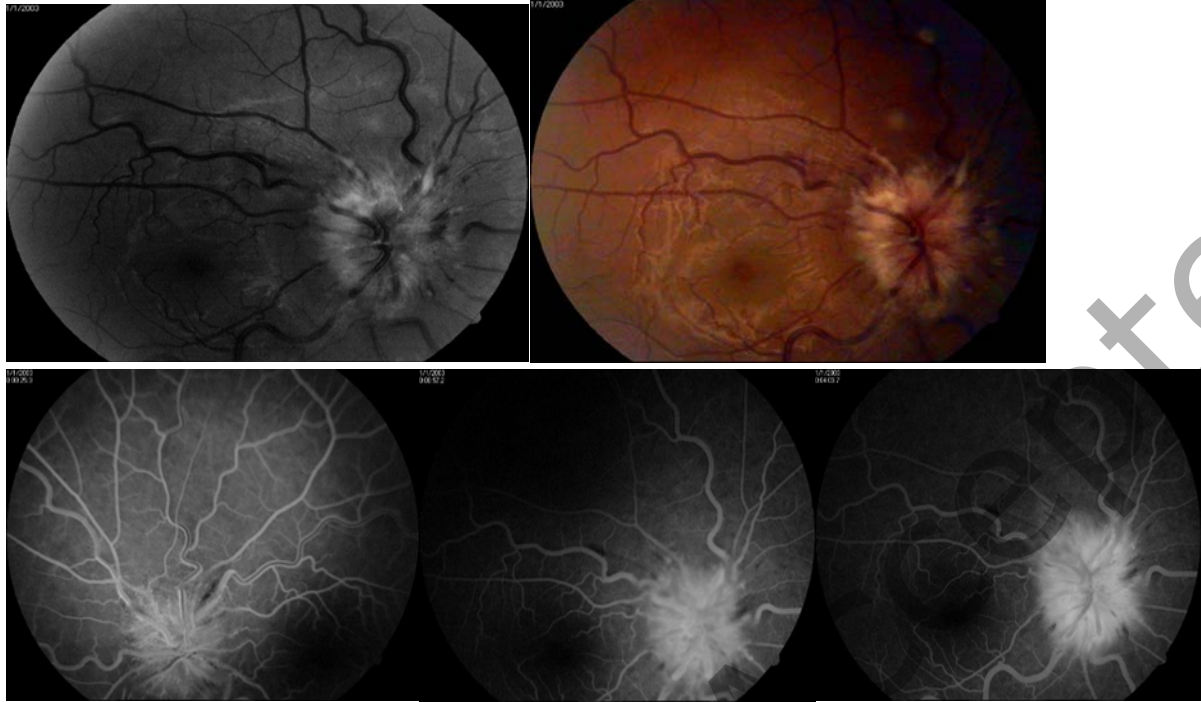


Figure 2 FAG of the right eye

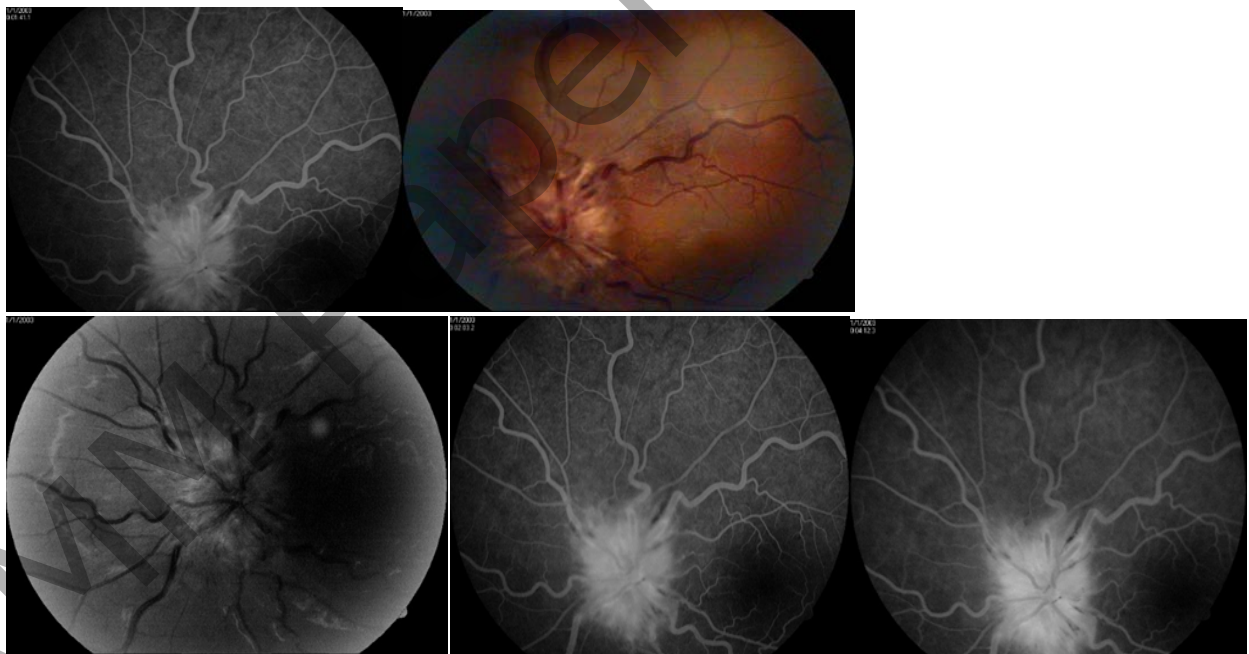


Figure 3. FAG of the left eye

Optical coherence tomography (OCT) imaging of the both eyes indicated bilateral nerve fiber thickening and in all quadrants of retinal fibril nerve layer (RNFL) of the optic disc, with average RNFL thickness of 247mic in right and 242 mic in left eye.(Figure 4).

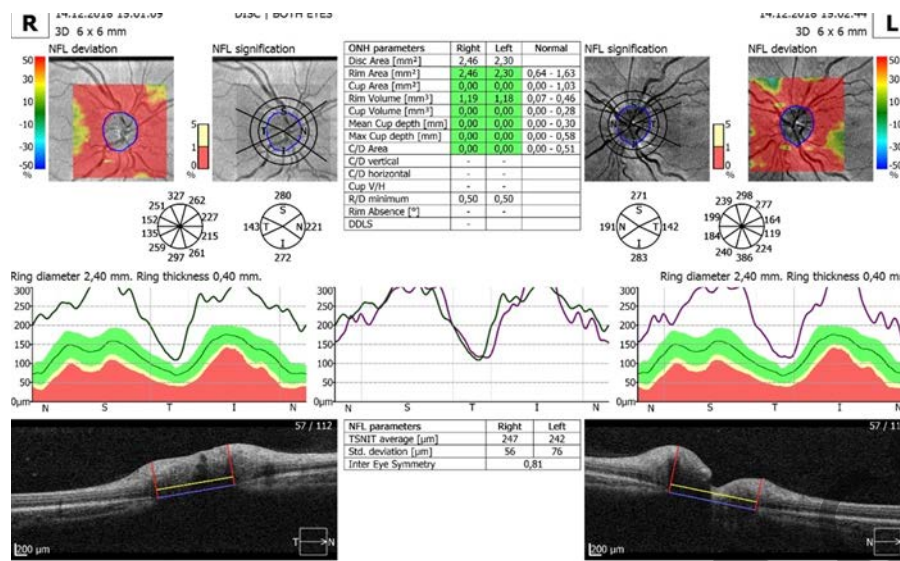


Figure 4. OCT imaging of optic disc , both eyes

Two weeks prior the onset of ophthalmic symptoms, our patient had a common cold with a raised body temperature of 38°C. Five years ago she suffered from infectious mononucleosis, followed by elevated transaminase levels.

A year before the onset of the headaches she started a diet, having 80 kg of body mass at the time. She was taking weight loss products for a months, then she pursued a rigorous diet for 11 months attaining the weight of 47 kg. During all this time she was subjected to physical exertion, since she had actively trained handball, with everyday training sessions. In that period she felt exhausted and suffered mental stress and anxiety, so she discontinued her diet after a year and went back to normal eating.

Since the very beginning of her strict diet she lost her menstrual cycle for a year, and it got regulated after diet discontinuation. She also had regular gynecological examinations in that period and the findings were normal. This condition was explained as "a diet caused hormonal imbalance". Before starting her diet, she had severe headaches during the menstrual period and they were reduced only after taking Mannitol saline and Novalgetol ampoules.

After hospitalization, magnetic resonance imaging of the brain and angiography were done, revealing no intracranial pathological changes that would explain the presence of the papilledema in both eyes. (Figure 5)

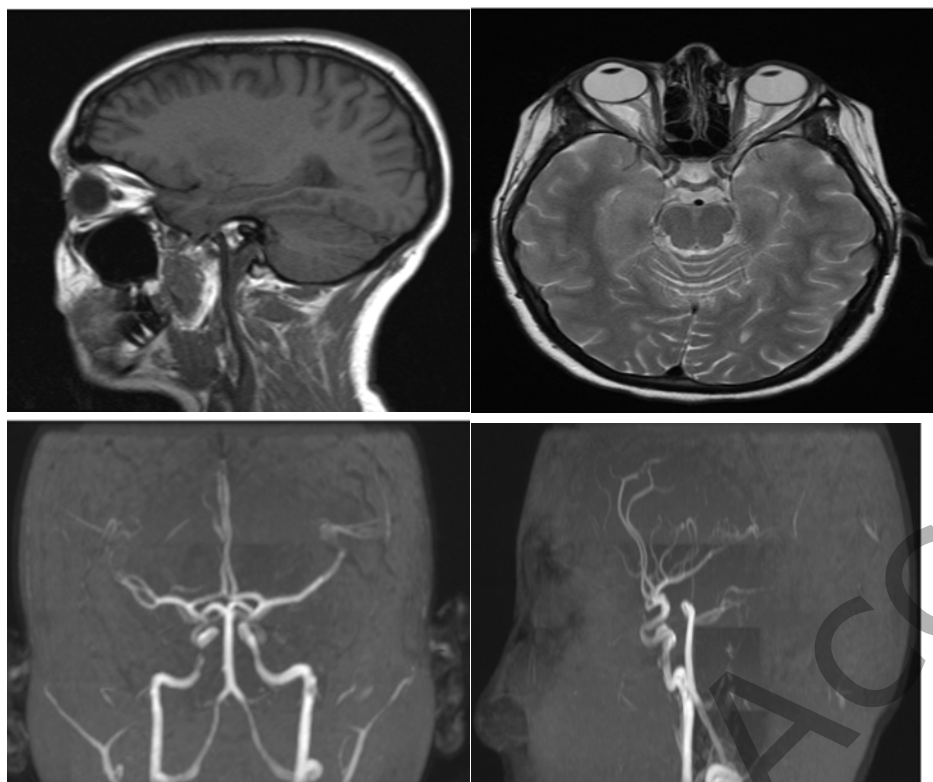


Figure 5 MRI and Angiography finding

After intravenous application of Mannitol saline, 250ml in three days, intensive headaches have stopped, and the patient was discharged with medication therapy of Huma-Zolamide 250mg 2x1. After another follow-up visit the patient denied having any subjective problems regarding transient visual acuity disturbances; double vision disappeared, with minimal paresis symptoms of extraocular lateral rectus muscle of the left eye. (Figure 6) Papilledema in both eyes was significantly reduced. Normal physical activity was recommended to the patient, as well as avoiding any kind of dieting, and further therapy by using lower doses of carbonic anhydrase inhibitors.



Figure 6 The beginning of the treatment

After 6 months

After six months diplopia completely disappeared and the function of extraocular lateral rectus muscle of the left eye was back to normal. Along with the assistance of a nutritionist and pediatrician our patient now weighs 62 kg, there are no subjective visual disturbances, and she normally performs all physical activities.

The ocular fundus finding is completely regular, without signs of papilla stagnans (optic disc edema). The finding of Optical Coherence Tomography also shows normal values of optic disc thickness, without the signs of edema. (Figure 7.)

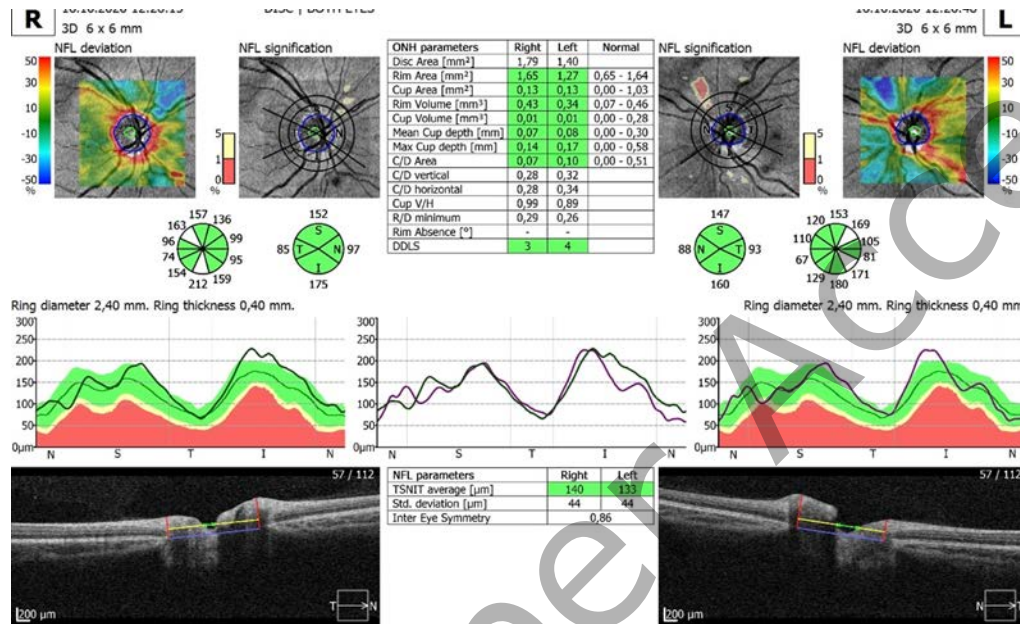


Figure 7. Improvement of OCT imaging of optic disc , after therapy

Discussion and conclusion

The most common symptom of benign intracranial hypertension in children is headache, present in about 91% cases. A headache is in the form of a throbbing, intermittent, and diffuse pain, which gets worse during the day. Retrobulbar pain may also be present, accompanied with the neck and back pain. A headache is sometimes associated with nausea and vomiting. (8,9).

Apart from headache, patients report the following symptoms as well: changes in visual acuity, transient visual obscurations, and double vision. Visual acuity loss is not usually significant, and it has been reported in about 6-20% cases. Blurry vision accompanied with different types of scotomas have been documented in 91% of patients. (10,11).

One of the common symptoms in children with benign intracranial hypertension is Abducens nerve palsy, present with diplopia in 46-60% of patients.

The dominant symptom in our patient was headache and diplopia caused by abducens nerve palsy, while the presence of scotomas has not been registered, except for a slightly enlarged blind spot as a result of optic disc edema.

One of the most common diagnostic signs in these patients is also bilateral optic disc edema, which refers to the ophthalmoscopic papilla stagnans. Optic disc edema is typically bilateral, but not so rarely it can be unilateral as well. Some authors report the absence of optic disc edema in 48% of their patients with benign intracranial hypertension in children. (12,13). Most authors agree that the absence of optic nerve head edema does not exclude the disease in the presence of other symptoms. (14).

Children with suspected benign intracranial hypertension should undergo a detailed examination by a pediatric neurologist.

Risk factors for pseudotumour cerebri are numerous and most commonly include obesity, hormonal imbalance, certain medications, lack of vitamins, and associated systemic diseases. Obesity as a risk factor occurs in 4-21 per 100.000 obese women. (15,16,17)

The incidence is significantly lower in children, ranging from 0.5 to 0.9 cases per 100.000 children. Obesity has been reported as one of common causes in children as well. In available literature we have not found the description of pediatric benign intracranial hypertension as a consequence of sudden weight loss due to a rigorous diet.

Therapeutic approach in underlying disease treatment includes body weight management, discontinuation of medication that may cause intracranial pressure elevation, and prescription of therapy that can alleviate the symptoms and speed the recovery of the patients. (18.19).

Medical treatment includes application of acetazolamide that can greatly result in improvement of the conditions in children with pseudotumour cerebri. It has been proved that acetazolamide use reduces the rate of cerebrospinal fluid production and improves the symptoms of the disease in 47-67% of patients. (20).

Timely diagnosis and treatment initiation in our patient resulted in disappearance of subjective difficulties and optic disc edema withdrawal. The patient has been followed for a few years and there are no signs of the disease recurrence.

We have not found description of pediatric benign intracranial hypertension as a consequence of sudden weight loss due to rigorous diet in available literature. There is a higher incidence of women up to 44 years of age in the group with pseudotumour cerebri. Therapeutic approach in underlying disease treatment includes body weight management, discontinuation of medication that may cause intracranial pressure elevation, and prescription of therapy that can alleviate the symptoms and speed the recovery of the patients.

Medical treatment includes application of acetazolamide that can greatly result in improvement of the conditions in children with pseudotumour cerebri. It has been proved that acetazolamide use reduces the rate of cerebrospinal fluid production and improves the symptoms of the disease in 47-67% of patients.

Conclusion

Benign intracranial hypertension is uncommon in pediatric patients and requires multidisciplinary approach in diagnostics and treatment.

The case of a 14-year-old girl who followed a rigorous diet indicates that benign intracranial hypertension is possible in that age, and such a procedure has to be done under the control of appropriate professionals and a pediatrician.

Major loss of body weight, disturbance and the loss of menstrual cycle, along with physical exertion, resulted in hormone dysfunction leading to increased intracranial pressure and manifestation of benign intracranial hypertension.

The discontinuation of the diet, normalization of body weight and adequate dosage application of carbonic anhydrase inhibitors have resulted in headaches disappearance, cessation of visual acuity transient impairment, and the recovery of Abducens nerve paresis that caused double vision manifestation.

Literature:

1. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. *Neurology*. 2002;59:1492–1495.
2. Digre KB, Corbett JJ. Idiopathic intracranial hypertension (pseudotumor cerebri): a reappraisal. *The Neurologist*. 2001;7:2–67.
3. Corbett JJ, Thompson HS. The rational management of idiopathic intracranial hypertension. *Arch Neurol*. 1989;46:1049–1051. [PubMed]
4. Karahalios DG, Rekate HL, Khayata MH, Apostolides PJ. Elevated intracranial venous pressure as a universal mechanism in pseudotumor cerebri of varying etiologies. *Neurology*. 1996;46:198–202.
5. Babikian P, Corbett J, Bell W. Idiopathic intracranial hypertension in children: the Iowa experience. *J Child Neurol*. 1994;9(2):144–9.
6. Friedman DI, Jacobson DM. Idiopathic intracranial hypertension. *J Neuroophthalmol*. 2004;24(2):138–45. 1. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. *Neurology*. 2002;59(10):1492–5.
7. Ko MW, Liu GT. Pediatric idiopathic intracranial hypertension (pseudotumor cerebri). *Horm Res Paediatr*. 2010;74(6):381–9. [PubMed] [Google Scholar]
8. Yuval Cohen; Muhammad Mahajnah; Rana Hanna; Michael Idel; Beatrice Tiosano . OCT Imaging of Papilledema in Pediatric Idiopathic Intracranial Hypertension .*Investigative Ophthalmology & Visual Science* September 2016, Vol.57, 4546. doi:
9. Aylward SC, Waslo CS, Au JN, Tanne E. Manifestations of Pediatric Intracranial Hypertension From the Intracranial Hypertension Registry. *Pediatr Neurol*. 2016;61:76–82.
10. Celebisoy N, Gokcay F, Sirin H, Akyurekli O. Treatment of idiopathic intracranial hypertension: topiramate vs acetazolamide, an open label study. *Acta Neurol Scand* 2007;116:5:322–327.
11. McKiernan SP, DiFazio MP. Index of suspicion. Case 3. Diagnosis: Infantile pseudotumor cerebri. *Pediatr Rev*. 2001;22(6):211–5. [PubMed] [Google Scholar]
12. Rangwala LM, Liu GT. Pediatric idiopathic intracranial hypertension. *Surv Ophthalmol*. 2007;52(6):597–617.
13. Tibussek D, Distelmaier F, von Kries R, Mayatepek E. Pseudotumor cerebri in childhood and adolescence -- results of a Germany-wide ESPED-survey. *Klin Padiatr*. 2013;225(2):81–5
14. Rogers DL. A review of pediatric idiopathic intracranial hypertension. *Pediatr Clin North Am*. 2014;61(3):579–90.
15. Rook BS, Phillips PH. Pediatric pseudotumor cerebri. *Curr Opin Ophthalmol*. 2016;27(5):416–9.
16. Kesler A, Fattal-Valevski A. Idiopathic intracranial hypertension in the pediatric population. *J Child Neurol*. 2002;17(10):745–8.

17. Safwat Abouhashem, Ahmed A. M. Gad, Mohamed El-Malkey & Esam A. Daoud Idiopathic intracranial hypertension in pediatric and adolescent patients. Egyptian Journal of Neurosurgery volume 34, Article number: 32 (2019)
18. Balcer LJ, Liu GT, Forman S, Pun K, Volpe NJ, Galetta SL, et al. Idiopathic intracranial hypertension: relation of age and obesity in children. Neurology. 1999;52(4):870–2.
19. Mercille G, Ospina LH. Pediatric idiopathic intracranial hypertension: A review. Ped Review 2007;28:e77-86.
20. Friedman DI. Pseudotumor cerebri. Neurol Clin North Am 2004;22:99-131.