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Idiopathic Intracranial Hypertension: An Updated Literature Review

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**Department of Neurosurgery, Vilnius University National Hospital; Faculty of Medicine, Utena University of Applied Sciences Summary. Idiopathic intracranial hypertension is defined as an increase in intracranial pressure that occurs without a known cause and is accompanied by symptoms of elevated intracranial pressure, such as headache and vision loss. We want to provide a comprehensive and up-to-date overview of idiopathic intracranial hypertension including the most recent research and treatment choices by analyzing scientific publications from PubMed, SpringerLink, and ScienceDirect databases. The fact that idiopathic intracranial hypertension has been referred to by several different names over the years is a reminder that the underlying mechanism is still unknown. The goal of the treatment of idiopathic intracranial hypertension is to lower intracranial pressure, relieve headaches, and save patients' vision. Acetazolamide still appears to be an acceptable first-line treatment, according to research. Cerebrospinal fluid diversion techniques, optic nerve sheath fenestration, bariatric surgery, and venous sinus stenting are all approved surgical procedures for medically resistant idiopathic intracranial hypertension. With low failure rates and a favourable complication profile, venous stenting delivers the highest results in pain relief and visual outcomes, therefore it should be considered the first-line surgical therapy option for medically refractory idiopathic intracranial hypertension.

Keywords: idiopathic intracranial hypertension, benign intracranial hypertension, pseudotumor cerebri, disorder of cerebrospinal fluid circulation.

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is a rare condition characterized by symptoms of increased intracranial pressure (ICP) of unknown cause in the absence of a focal brain lesion. Also widely known as primary intracranial hypertension (PIH), it was first described in the 1890s by the German physician Quincke and given the name "meningitis serosa", linking the condition to an excess production of cerebrospinal fluid (CSF) [1, 2].

In 1904, this condition was also described by Nonne, who named it "pseudotumor cerebri", presenting a series

of patients with signs of intracranial mass, but not having one, as it turned out on proper evaluation [3]. The term gained further popularity until, in 1955, Foley again renamed it "benign intracranial hypertension", although significant morbidities, namely permanent visual abnormalities (10–25%) and chronic headaches, caused this name to fall out of favour [4–7].

Friedman et al. attempted to clarify the terminology underlying the uncertainty of these names by proposing the term "pseudotumor cerebri syndrome" (PTCS), further dividing the patients into two groups: primary (with no identifiable cause, naming risk factors, such as female gender, post-pubertal status, obesity, or polycystic ovarian syndrome that do not directly result in ICP) and secondary (with an identifiable secondary cause, such as cerebral sinus venous thrombosis or oral drug use) (See Table 1) [8].

Despite numerous attempts, the underlying cause of this condition is yet to be discovered. Currently, the term "idiopathic intracranial hypertension" is most commonly used.

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METHODS

A systematic literature review on IIH was performed by analyzing English and non-English scientific publications from PubMed, SpringerLink, and ScienceDirect databases.

DEMOGRAPHICS

This disease disproportionately affects overweight women of childbearing age. In obese females of childbearing age with a mean age of 33 years, the estimated prevalence is approximately 12–20 per 100,000 people per year, with only 0.5–2 per 100,000 people per year in the general population. In healthy males, the global incidence is 0.3 per 100,000, but it also rises in obese males to 1.5 per 100,000 [9].

Pre-pubertal boys and girls are affected equally. The annual pediatric incidence is estimated to be 0.63 per 100,000. Polemikos et al. reported homozygous twins who both presented with IIH 3 years apart and were both obese [10]. However, obesity has not been found to correlate with an increased risk of PIH until around the time of puberty [11].

Due to the increasing incidence of obesity globally, the number of IIH cases is increasing, and a retrospective study shows that the rate of IIH in the general population has risen to 2.4 per 100,000 (in 2002–2014) compared with 1.0 per 100,000 (in 1990–2001) [12].

CLINICAL PRESENTATION

The symptomology of IIH tends to be quite variable, in most common cases headaches, neck pain and stiffness, pulsatile tinnitus, and visual changes, such as vision loss, transient visual obscurations, and binocular horizontal diplopia are present. Less common symptoms include dizziness as well as nausea, vomiting, and lethargy. Possible additional findings include back pain and paresthesia [13]. In younger patients, the symptoms of intracranial hypertension are less evident.

Headache is the most prevalent symptom at the time of IIH presentation and also the most common. Headaches, usually reported as pressure-like or throbbing, occur daily in about 90% of patients and have an overall median frequency of 12 days per month. They can be generalized or confined to the frontal or retro-orbital region [14]. Sometimes they are aggravated by Valsalva maneuvers [15]. Can be associated with neck, back, and radicular pain [16]. A recent review also found that patients reported associated symptoms of photophobia (70%), phonophobia (52%), and worsening pain with routine physical activity (50%) [13]. Patients may exhibit migrainous symptoms or have a history of migraine headaches. These patients may also have a history of drug overuse for persistent head-

Table 1. Causes of secondary pseudotumor cerebri syndrome

Medical conditions	Medications
Cerebral venous thrombosis	Minocycline/tetracy-
Hydrocephalus	cline/doxycycline
Pregnancy/eclampsia	Fluoroquinolone
Pregnancy/eclampsia Adrenal insufficiency (often on steroids) Hypoparathyroidism (in early stage of treatment) Behcet's disease Crohn's disease Refeeding syndrome Craniofacial syndromes Chiari malformation Traumatic brain injury Intracranial tumor Intracranial hemorrhage Meningitis/encephalitis	Fluoroquinolone Growth hormones Oral contraceptives (likely secondary due to venous thrombosis) Corticosteroids (especially during withdrawal) Cyclosporine A Cytarabine Lithium carbonate Nalidixic acid Retinoic acid
• Lyme disease	
Demyelinating disease/multi- ple sclerosis	
• Leukemia	
• Lymphoma	
Vitamin A (excess or deficiency)	
Vitamin D (deficiency)	

aches, as well as secondary headaches as a result of this overuse [13, 17-20].

Papilledema is identified in practically every patient on physical examination. However, a tiny percentage of patients, about 5%, do not have papilledema [21, 22].

Blurred vision sometimes accompanied by a narrowing of the visual field is a typical visual change symptom. Monocular or binocular partial or complete transient visual obscurations (TVOs) connected to changes in posture, lasting less than a minute, are also possible, with further complete visual recovery. They are not IIH specific as assumed to be caused by transient ischemia of the optic nerve head caused by disc edema, as a result of increased intracranial pressure [7, 23, 24].

In roughly 20% of patients, diplopia and visual field impairments are caused by cranial nerve VI (CN VI) palsy, which can be unilateral or bilateral and is caused by traction on the abducens nerve, resolving with the improvement of intracranial pressure [25]. Although cranial nerve VII (CN VII) involvement has been recorded, it is considered very uncommon [26].

Pulsatile tinnitus, usually unreported until asked directly, either unilateral or bilateral, is another symptom reported by more than half of patients. It can happen as a result of turbulence in the dural venous sinus stenosis, described as a whooshing sound or a heartbeat, resolving with the improvement of intracranial pressure [24, 27]. The return of tinnitus after drug weaning can occur before other symptoms or even papilledema [11].

The initial symptoms of PIH have been reported to be spontaneous CSF rhinorrhea and otorrhea. Symptoms of PIH do not show until the leak is fixed or disappears spontaneously [28].

Bershad et al. observed impairment in olfactory detection and a slight loss of smell identification [29]. In a similar report, five IIH patients admitted that they had a diminished sense of smell when questioned, and further direct testing revealed hyposmia in 41% of IIH patients. Acute presentations with a lowered olfactory detection threshold had the highest incidence of the impairment [30].

Cognitive decline in adult patients is a frequent report with IIH. Processing speed and reaction time were the most commonly affected regions. Except for one small retrospective study, working memory was not impaired [11].

There is a recent report of COVID-19 being associated with IIH [31].

DIAGNOSIS

Dandy originally summarized the criteria for diagnosing IIH in 1937, when he presented a series of patients with elevated intracranial pressure but no dilated ventricles on ventriculography [32]. Dr. J.L. Smith amended the criteria and renamed them the Modified Dandy Criteria in 1985 as a result of the widespread inpatient evaluation by computed tomography (CT). In 2002, Friedman and Jacobson updated the criteria once more. Friedman et al. once again amended this classification in 2014, categorizing IIH as a subset of pseudotumor cerebri syndrome (PTCS), which now includes both primary and secondary intracranial hypertension (See Table 2) [8].

In the absence of papilledema, the diagnosis of PTCS can be made using the same criteria as above, except for the first, and the presence of unilateral or bilateral abducens nerve palsy. In addition, the presence of certain MRI abnormalities can now be used to suggest the diagnosis in the absence of papilledema or sixth nerve palsy. A diagnosis of likely PTCS might be made if three of the following four abnormalities are present on MRI. These findings consist of:

- 1. partially empty sella,
- 2. flattening of the posterior aspect of the globe,
- 3. distension of the perioptic subarachnoid space with or without tortuous optic nerve, and
 - 4. transverse venous sinus stenosis [9].

Although these stricter criteria do not indicate that treatment should be withheld if a patient does not meet definite or likely criteria, it does raise the possibility that patients will be neglected or untreated, resulting in greater morbidity. The question of whether the criteria are fully applicable to pediatric patients remains open.

In the examination of patients with potential IIH, the role of imaging has changed dramatically. A brain MRI with gadolinium is the most appropriate test to rule out underlying disease, and it should show normal brain parenchyma with no signs of hydrocephalus, a space-occupying

Table 2. Diagnostic criteria for PTCS with papilledema

- (1) papilledema must be present
- (2) normal neurologic examination except for cranial nerve abnormalities
- (3) no parenchymal lesion on brain imaging
- (4) normal CSF composition
- (5) the opening pressure must be >25 cmH2O

lesion, or aberrant meningeal enhancement [15]. Furthermore, there is now considerable evidence that imaging of patients with IIH might, in fact, show various abnormalities, and that radiologists should search for subtle ones. A partially empty sella, flattening of the posterior globe, cupping of the optic disks, and distension of the optic nerve sheaths are among these findings. Hartmann et al compared MRIs of children with pediatric IIH with those of adults [33]. With the exception of meningoceles, they found all imaging abnormalities seen in adults in their cohort. Because remission is usually seen following treatment, bilateral transverse venous constriction is thought to be the outcome rather than the cause of intracranial hypertension [34].

Furthermore, with the discovery that a large percentage of patients have dural venous sinus stenosis, intracranial venography has become increasingly important [9]. Cerebral venous sinus thrombosis (CVST) can have symptoms very similar to IIH, with some overlap. As a result, to rule out CVST, venous imaging using magnetic resonance venography (MRV) should be performed. According to some, MRV should be ordered routinely as part of the IIH workup [35]. CVST is an uncommon condition and is usually recommended in atypical patients, such as nonobese individuals, prepubertal children, men, patients with progressive visual loss despite medical therapy, and high-risk patients for venous sinus thrombosis, such as women on oral contraceptives who smoke cigarettes [20].

When it comes to diagnosing increased intracranial pressure, an ophthalmologic exam is critical. Visual acuity, pupillary function, extraocular motility, colour vision evaluation, and optic nerve appearance during fundoscopy are all-important examination components. Abducens nerve palsy can occur unilaterally or bilaterally.

The most useful quantifiable test for evaluating visual function is visual field testing. Wall and George studied 50 patients with IIH who underwent automated and Goldmann perimetry and found that enlargement of the blind spot, nasal defects, and widespread depressions were the most common visual field defects. Localized nerve fiber bundle defect (in 60%) and enlarged blind spot with partial arcuate anomaly were the most prevalent visual field impairments reported in the Idiopathic Intracranial Hypertension Treatment Trial (IIHTT). Loss in the inferior hemifield was greater than loss in the superior hemifield [7, 13, 20, 36].

The hallmark of IIH is papilledema, which is primarily assessed clinically. A formal ophthalmologic examination, such as ocular ultrasound, is recommended to ensure

that papilledema is not mistaken for pseudo papilledema, such as drusen or congenital anomalous discs, or other unilateral causes of optic disc enlargement, such as optic neuritis or anterior ischemic optic neuropathy. In addition, there have been several reports of unilateral papilledema associated with IIH [20, 37]. In real papilledema, fluorescein angiography can also be used to confirm disk leakage and differentiate between disc edema caused by high intracranial pressure and optic nerve drusen, or conditions when both diseases coexist. The Frisén scale is used to formally grade papilledema, however optical coherence tomography of the retinal nerve fiber layer (OCT RNFL) has recently become an important tool for objectively quantifying it. The thickness of the retinal nerve fiber layer (RNFL) in particular may be related to the severity of papilledema [20].

Finally, the diagnosis requires a lumbar puncture to determine the opening pressure and a spinal fluid examination. The presence of normal CSF contents and an opening pressure higher than 250 mmH2O in adults and higher than 280 mmH2O in children and adolescents (250 mmH2O CSF if the child is not sedated and not obese) [8, 35] is diagnostic. The lumbar puncture should be performed in the lateral decubitus position with legs relaxed and head extended for an accurate measurement. The general tendency is to withdraw a substantial amount of CSF, resulting in pressure close to normal. A study by Aylward and Way states that a significant reduction of intracranial pressure from baseline increases the probability of post-LP headache. With the evacuation of approximately 0.91 mL of CSF, intracranial pressure changes by 1 cmH2O in pediatric patients [11, 38]. Opening pressure shows a negative linear relationship with CSF protein, with no correlation to BMI, age, or gender [39].

It is important to note that IIH is an excluding diagnosis.

TREATMENT

The clinical history of IIH is highly diverse; many individuals respond well to treatment and may even have spontaneous remission, while others may have a fulminant disease. Relapse is a common occurrence. The primary goal of therapy is to preserve vision, as well as reduce symptoms, especially those associated with headache. There are currently no clear guidelines for the treatment of IIH because there isn't enough evidence or agreement on the best therapeutic strategies. Intracranial pressure is reduced as a major part of the treatment. The type of treatment depends on the severity of the symptoms. After lumbar puncture, which lowers intracranial pressure to normal level, some patients improve symptoms.

Conservative approach

A low-sodium diet and weight loss are two conservative methods to reduce or eliminate symptoms. Reversal of symptoms requires a weight loss of about 6%, although this usually is difficult to achieve [40].

Medical management

Since the first recorded successful use for IIH in the 1970s, acetazolamide, a powerful carbonic anhydrase inhibitor that reduces CSF production, has become the drug of choice [41]. Unfortunately, acetazolamide has many side effects, and many patients may need to use topiramate instead. Fatigue, paresthesia, and kidney stones are among the most prevalent side effects of acetazolamide [42].

In the Idiopathic Intracranial Hypertension Study, acetazolamide combined with weight loss was found to result in modest improvements in visual field compared with diet alone. The acetazolamide diet group performed considerably better than the placebo diet group in terms of papilledema grade [0.7 dB (95% CI 0.99 to 0.41)], CSF pressure [59.9 mmH2O (95% CI 96.4 to 23.4), P=0.002] and quality of life scores. This had no noticeable impact on the severity of headaches [43]. As a consequence, the Longitudinal Idiopathic Intracranial Hypertension Trial was performed to see if the favourable benefits of acetazolamide in improving visual function continued in people already participating in IIHTT up to 12 months. The second study also suggested that acetazolamide may still have a beneficial impact on headache scores, as the effect was significant (P=0.01) [44].

Topiramate is used as an alternative or supplemental treatment in many cases. The advantage of this drug is that it prevents migraine, suppresses appetite (which helps in weight loss), and has a mild carbonic anhydrase inhibitor effect [45].

If necessary, Lasix can be added under certain circumstances. Corticosteroids may be used to temporarily reduce blood pressure, but long-term use is not indicated, and discontinuation may result in a rebound effect. Repeated lumbar punctures can also be used to lower intracranial pressure, however many patients become tired of this procedure quickly. In addition, a CSF leak can occur, making it difficult to determine the exact origin of the following headaches [9].

Surgical interventions

When rapid or progressive vision loss occurs despite optimum medical therapy, or when medical therapy is not tolerated or fails, more severe methods should be considered. Depending on local preferences, the procedure used may vary.

Optic nerve sheath fenestration (ONSF)

The optic nerve sheath is fenestrated or punctured by a medial (most common) or lateral orbital approach or a superomedial lid crease incision to produce a window or slits to relieve CSF pressure. Optic nerve sheath fenestration is

generally safe and successful. The procedure is usually done bilaterally, but it can also be done unilaterally. In a recent review that included 23 ONSF studies using different procedures, such as medial or lateral transconjunctival, superomedial lid slit, or endoscopic approaches, a total of 1365 ONSF procedures were performed in 818 patients. Despite initially successful ONSF, 118 of 700 patients (16.9%) required an additional intervention, such as shunting or stenting, to obtain symptom control, most commonly headache. Finally, 145 patients out of 724 had 161 problems, most of which were minor and transient; only 16 (2.2%) of the complications were classified as serious. Complications may include retinal vascular occlusion and/or traumatic optic neuropathy, irreversible vision loss, diplopia, and pupil irregularities, as well as other complications [20].

CSF diversion procedures

Ventriculoperitoneal shunts and lumboperitoneal shunts are two options for CSF diversion. Because the ventricles are not enlarged in IIH, the latter is often preferred as it is an extracranial treatment. After analyzing 28 studies which provided data on about 609 patients with a mean age of 32.7, CSF diversion was the first surgical technique performed to treat IIH in 350 of 391 patients (89.5%).

Thirteen of the 28 studies specifically examined individuals who received a ventriculoperitoneal shunt. These studies included 142 patients, 80.3% of whom were women (106/132), with a mean age of 32.4 and a follow-up period of 32.7 months. Visual acuity and visual fields improved in 57.8% (37/64) and 67.3% (33/49) of participants, respectively. Headaches improved in 90.8% (89/98) of patients, and papilledema disappeared in 90.9% (30/33) of cases.

Nine studies included 157 patients who had lumbarperitoneal shunts for IIH, 90% of whom were women (125/139), with a mean age of 32.1 years and an average follow-up of 38.5 months. Visual acuity and visual fields were restored in 70.1% (41/58) and 72.3% (47/65) of cases, respectively. Headaches in patients improved in 97.9% (113/123) of cases, and papilledema decreased in 86.6% (71/82) of cases.

In more recent trials, neuronavigation is used to make ventricular catheter insertion easier. After CSF diversion, only 1.4% (6 of 423) required additional surgical intervention. Shunt infection was the most common severe complication, accounting for 9.4% of all severe complications. Shunt obstruction was the cause of most revisions [46].

Venous stenting

Stenting of the dural venous sinuses is a more recent option, used primarily in adults. Venous sinus stenting was assessed in 47 studies including a total of 825 patients. Headache resolved or reduced in 72.1%, and papilledema resolved in 87.1%. On the treatment side, the most com-

mon side effect is a temporary headache or partial hearing loss. Perforation of the venous guidewire, cerebral bleeding, and transient unsteadiness are uncommon. With the exception of early studies, all of them used dual antiplatelet drug before the procedure and heparinization during the procedure [46]. Patsalides et al. recently examined pre-CSF and 3-month post-CSF opening pressure of 50 IIH patients who underwent venous sinus stenting. This study found that venous sinus stenting, in addition to acetazolamide and weight loss, is an effective treatment to significantly reduce ICP [47].

Bariatric surgery

There is also evidence that bariatric surgery may help with IIH due to weight loss and possibly decreased cerebral venous pressure. Bariatric surgery is performed to achieve significant long-term weight loss, resulting in a reduction in ICP and, as a result, improvement in the signs and symptoms of IIH. Laparoscopic gastric bypass (LGB) and laparoscopic adjustable gastric banding (LABG) are the two main procedures performed. However, due to the time it takes for bariatric surgery to take effect, it would not be a suitable first-line option in a more severe case. Band migration, anastomosis leak, gastric stenosis, dumping syndrome, and starvation are among complications linked with bariatric surgery [20].

Based on data from 11 bariatric surgery studies in which a total of 50 patients were evaluated, visual fields of 10 of the 12 patients in the small bariatric surgery cohort improved, and visual acuity was restored in all 5 participants tested, 42 of 45 patients who underwent bariatric surgery reported improvement in headache, 4 studies reported data on 12 individuals, all of whom had their ICP normalized [46].

There is no consensus on the best surgical intervention to treat IIH at this time. Surgical procedures are more costly than medical treatment.

COMPLICATIONS

Visual impairment is the most common complication of IIH. Increased intracranial pressure causing papilledema results in gradual vision loss due to the optic disc atrophy. Visual fields are usually impaired first, and colour vision and acuity follow later [48]. In 15–67% of cases, recurrence is documented. Relapse can happen years after the initial episode and is often linked with recent rapid weight gain. The number of pregnancies effects the frequency of recurrences [49]. Overall quality of life is also lower in patients with IIH compared to controls due to higher anxiety and depression rates [50]. Mild cognitive impairment appears to be one of the possible comorbidities associated with IIH. While memory appears to be unaffected, other factors such as nonverbal memory, executive function, visual spatial processing, attention, motor abilities, problem

solving, and information processing speed showed statistically significant decline. Furthermore, these findings persisted even after intracranial pressures were reduced and headaches were no longer present [46].

As for surgical interventions, venous stenting seems to have the best overall risk profile, with only 9.4% of patients reporting any complications, followed by optic nerve sheath fenestration, which has a complication rate of 20%. However, it is worth noting that ONSF has the lowest rate of serious complications (2.2%), followed by VSS (2.3%). Compared to alternative treatment choices, bariatric surgery had the highest complication rate, with 55% of patients experiencing post-operative problems [46].

CONCLUSIONS

In summary, since it was originally documented more than a century ago, our understanding of idiopathic intracranial hypertension has grown, especially with the use of imaging technologies like MRI and OCT for diagnosis and follow-up. However, with numerous unsubstantiated theories, the pathophysiology remains a mystery. Although the prognosis for these patients is generally favourable, this condition can adversely affect the patient's quality of life, causing substantial pain, cognitive dysfunction, and permanent vision loss, therefore early detection and treatment are critical. The disorder progresses differently in each patient, with some achieving full recovery while others experience persistent or recurrent symptoms. Treatment choices vary widely, although the goal is always to reduce intracranial pressure. Nevertheless, weight loss has been shown to be an essential part of treatment, and recent evidence supports the use of acetazolamide as a first-line treatment in addition to weight loss, especially in situations of modest vision loss. Several surgical methods have demonstrated promising results in terms of vision recovery and clinical improvement. It is important to understand that all surgical alternatives come with risks; as a result, they are commonly used in circumstances where conventional therapy has failed or has been ineffective and vision loss has further progressed. The field is still open for future clinical research to determine the underlying mechanism and possibly develop more effective treatments.

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IDIOPATINĖ INTRAKRANIJINĖ HIPERTENZIJA: ATNAUJINTA LITERATŪROS APŽVALGA

Santrauka

Idiopatinė intrakranijinė hipertenzija apibrėžiama kaip intrakranijinio slėgio padidėjimas, atsirandantis be žinomos priežasties ir lydimas padidėjusio intrakranijinio spaudimo simptomų, tokių kaip galvos skausmas ir regėjimo praradimas. Norime pateikti jums išsamią ir naujausią idiopatinės intrakranijinės hipertenzijos apžvalgą, analizuojančią naujausius tyrimus ir gydymo būdus, pagal mokslines publikacijas iš "PubMed", "SpringerLink" ir "ScienceDirect" duomenų bazių. Tai, kad idiopatinė intrakranijinė hipertenzija, metams bėgant, buvo pavadinta keliais skirtingais pavadinimais, duoda suprasti, kad pagrindinis mechanizmas vis dar nežinomas. Idiopatinės intrakranijinės hipertenzijos

gydymo tikslas – sumažinti intrakranijinį spaudimą, palengvinti galvos skausmą ir išsaugoti pacientų regėjimą. Remiantis tyrimais, acetazolamidas vis dar yra priimtinas pirmosios eilės gydymas. Smegenų skysčio nukreipimo metodai, regos nervo apvalkalo fenestracija, bariatrinė chirurgija ir venų sinusų stentavimas yra patvirtintos mediciniškai atsparios idiopatinės intrakranijinės hipertenzijos chirurginės procedūros. Esant mažam nesėkmių dažniui ir palankiam komplikacijų profiliui, venų stentavimas duoda aukščiausius skausmo malšinimo ir regėjimo išsaugojimo rezultatus, todėl turėtų būti laikomas pirmos eilės chirurginiu būdu, gydant atsparią idiopatinę intrakranijinę hipertenziją.

Raktažodžiai: idiopatinė intrakranijinė hipertenzija, gerybinė intrakranijinė hipertenzija, pseudotumor cerebri, likvoro cirkuliacijos sutrikimas.

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