


Idiopathic Intracranial Hypertension: A Case Study of Patient Engagement in the Treatment of a Chronic Disease

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Abstract

Idiopathic intracranial hypertension is a rare neurological disorder characterized by increased intracranial pressure, which can lead to visual loss and headaches. While medical therapy exists, weight loss is the only disease-modifying treatment. Weight loss is the only therapy that leads to sustained resolution of papilledema. Involving the patient in their disease management through patient engagement is a way to improve disease outcomes, and strengthen the therapeutic relationship. This feature discusses an overview of the disease, a patient's experience, and a physician's perspective.

Keywords

communication, clinician–patient relationship, challenges, medical decision making, patient/relationship-centered skills, patient engagement, patient expectations, patient perspectives/narratives

Introduction

Idiopathic Intracranial Hypertension

Idiopathic intracranial hypertension (IIH), also known as pseudotumor cerebri, is a neurological disorder characterized by high intracranial pressure (ICP) leading to visual loss and headaches (1). First reported by Quincke in 1893, the condition has been widely recognized as signs and symptoms of intracranial hypertension without a known cause, effectively making it a diagnosis of exclusion (2). Symptoms occur when cerebrospinal fluid (CSF) builds up in the space around the brain and spinal cord, causing pressure on the brain and optic nerve (1). IIH typically presents with headache, visual disturbances including transient visual obscuration and diplopia, nausea, vomiting, dizziness, and/or tinnitus (2). It is diagnosed by meeting the Modified-Modified Dandy Criteria listed in Table 1 (3). The annual incidence of IIH is 0.9/100,000 persons, making it fairly rare, however, it is more common in overweight women of reproductive age (1). In obese 20 - 44-year-old women (20% over ideal body weight), the incidence was increased to 19/100,000 (4). Untreated, IIH can lead to visual impairment or blindness as a result of optic atrophy (5). The mainstay of medical management is acetazolamide (a carbonic anhydrase inhibitor), which decreases CSF production (5). Topiramate may also be used as it is a carbonic anhydrase inhibitor, although evidence for this medication is less robust (6). The use of corticosteroid

therapy is controversial as there is a risk for rebound intracranial hypertension after withdrawal. However, steroids have been used for transient reduction of ICP for patients awaiting surgical intervention (5). Acetazolamide is a category C drug with a potential risk for teratotoxicity in the first trimester (5,7). This is an important consideration as pregnancy increases the risk of IIH above the already elevated incidence in women of reproductive age, making management more difficult in this context (8).

A sustained resolution of papilledema and symptoms can be achieved with weight loss, whether by counseling, interventional programs, or surgical therapy if necessary (5,9). A prospective cohort study demonstrated a reduction in ICP, improvement of papilledema, headache, and visual symptoms with weight loss (10). Three months later these improvements were sustained in patients that maintained their weight loss (10). Those that have achieved remission

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Table 1. Modified-Modified Dandy Criteria for IIH (Friedman et al. (3)).

1. Symptoms and signs of increased intracranial pressure (eg, papilledema)
2. No localizing neurological abnormalities (except abducens and facial nerve palsies)
3. Increased CSF opening pressure with normal CSF analysis
4. Neuroimaging reveals no structural cause or hydrocephalus
5. No other causes of intracranial pressure were found through the workup

Abbreviations: IIH, idiopathic intracranial hypertension; CSF, cerebrospinal fluid.

can experience a recurrence of symptoms with regaining as little as 6% of their body weight (11), emphasizing the importance of maintaining weight loss in the management of this illness (12).

Patient Engagement in the Management of IIH

Given the chronic nature of the condition and difficulties in treatment, it is important to actively involve patients in the management of their disease. Patient engagement describes active collaboration between patients, family members, caregivers, and health care professionals to help improve understanding of patient experiences, preferences, and needs (13). The Health Quality Ontario Patient Engagement Framework in Canada was created as a guide to implement and evaluate patient engagement in the health care system (13). This framework includes multiple domains, with aims to engage patients in personal care and health decisions, program and service design, and policy, strategy, and governance (13). While there are opportunities for the management of chronic diseases across all domains, the first is the primary focus of this article. This domain includes increasing patient activation and improving capacity building to help patients manage their health conditions, a notion included in other definitions of patient engagement (13–15). Struggling with refractory headaches and an unexpected pregnancy while suffering from IIH that resulted in a protracted disease course, LM was invited to write about her perspective to support patient and clinician education in the management of IIH.

Discussion

LM's Journey with IIH

In 2014, I began to notice my body was covered in bruises after continually running into furniture. An optometrist diagnosed swelling of my optic nerves and immediately sent me to the emergency room for further testing. As I sat listening to the doctor explain all the potential causes for the swelling, I was terrified and wondered if I was going blind? Subsequently, I met with an ophthalmologist and a neurologist and was diagnosed with IIH in January 2015 at the age of 34.

I grew up in Oshawa, Ontario and I led a relatively healthy and active life playing competitive soccer. After I graduated high school, I attended Queen's University completing degrees in physical and health education and psychology.

As I reached my late 20s and early 30s, I started to experience health issues that I attributed to the stress of life as an adult. It was at this time I started to experience chronic debilitating headaches but believed them to be a result of stress, dehydration, and insomnia. When I finally received the IIH diagnosis from my neurologist, I was unprepared for the discovery of such a rare chronic condition. Other symptoms I experienced, which I now understand were related to IIH include the complete loss of vision upon standing, an occasional "whooshing" in my ears, nausea, vomiting, and fatigue. After my diagnosis, I began a long and tenuous experience with medication, specifically acetazolamide which, outside of the symptoms of IIH, led me to experience extreme nausea. The nausea was so severe that I experienced a loss of appetite, daily vomiting, and a rapid weight loss of 20 pounds.

I was hospitalized in June 2015 due to severe dehydration and intolerance to the medication. The treatment that was saving my eyesight was making me so ill that I could not work or function most days. My dose of acetazolamide was reduced in an attempt to minimize the side effects of the medication and a new prescription, topiramate, was introduced. Over the course of my treatment, I have received three therapeutic lumbar punctures, which all provided immediate but short-term relief. After months of struggle, I finally met with a neuro-ophthalmologist who understood the condition and was able to find a medication dose I was able to tolerate.

Through my journey, I have been encouraged to lose weight as a means to reduce the symptoms. While easy in theory, working out and sustaining any long-term weight loss while experiencing daily headaches, overwhelming fatigue, and nausea has been a challenge. I have felt frustrated by weeks of wellness followed by sudden days of illness. The unpredictability of IIH and its symptoms have led to many days of crippling anxiety and depression. The stress of working full-time while managing IIH treatments and symptoms, as well as the financial burden caused by unpaid sick leave, has been overwhelming. Employers can be less sympathetic to those with invisible illnesses. I have also experienced strain on a number of my personal relationships. The unpredictable nature of this condition has forced me to miss work, family events, and break social commitments with friends.

Early on in my experience, I learned that finding a support system was incredibly important to my overall wellness. Facebook groups for IIH provided a nonjudgemental forum to ask questions and learn what does and does not work for other people. Discovering that I was not alone in my struggle has aided in maintaining a positive outlook for the future. Other supports providing me with some physical and emotional relief include cranial-sacral massage, talk therapy, a healthy diet, and exercise when I am able. Also, an open and honest relationship with my health practitioners has been crucial in feeling empowered to live my life outside of IIH.

In 2012, my spouse and I began the process of attempting to start a family. After six years and multiple failed interventions, we believed a baby was not in our future. To our surprise and delight, I became pregnant in October 2018. What would normally be a joyous time for most parents-to-be was frightening for us as I was on acetazolamide at the time and we were concerned about how a pregnancy would impact my condition? Weight gain in any capacity at this point had triggered a relapse of symptoms and vision loss, so how would we navigate pregnancy weight gain?

We devised a plan with our neuro-ophthalmologist and began working with a high-risk obstetrician. Our doctors worked collaboratively to support my health and that of my unborn baby. Monthly check-ups, support of a dietician, a neurologist, a lumbar puncture, the use of acetazolamide only in the second trimester of pregnancy, and only gaining nine pounds through the duration of the pregnancy supported the healthy birth of our baby girl in June 2019. With tremendous help from our diverse support system, we successfully navigated all of these complications and realized our dream of starting a family.

Living with a chronic condition is challenging, but it does not define me or my life. Goals that I believed to be unattainable at the beginning of this journey are now accomplishments, and I have found hope for myself and others with this condition. Through this experience, I have discovered a renewed commitment to my health and well-being. My life looks different than I believed it would be, but I have learned to advocate for myself, trust my instincts when it comes to my health and now I can clearly see an exciting future.

Physician Perspective

IIH is a common condition in a neuro-ophthalmologist's practice. Treating IIH can be challenging despite the various medical and surgical therapies available. Common problems encountered include intolerance to medication, inability to lose weight or further weight gain, noncompliance, and unexpected pregnancy while the patient is being treated. Weight loss is one of the foremost strategies in the management of IIH. Losing as little as 6% body weight has been proposed to be effective (12). However, the inability to lose weight remains to be one of the major shortcomings in disease management. With chronic headaches and concomitant psychiatric illnesses such as anxiety and depression, patients may find it difficult to get into a routine exercise regimen and lose self-motivation. Enrolling in a weight reduction program and working with a dietician should be recommended given the difficulties patients with IIH often encounter. Bariatric surgery has been proposed for patients with severe obesity where conservative management for weight loss failed (16), with class IV evidence of its efficacy in IIH. It is important to note that nutritional deficiencies following bariatric surgeries can rarely result in optic neuropathies, nyctalopia, ophthalmoparesis, and nystagmus (17).

IIH is a disease that prevails in women in the childbearing age group. Patients with known IIH can have a recurrence of papilledema when they get pregnant and management can become more challenging, requiring frequent lumbar punctures (18). The IIH treatment trial has provided evidence that acetazolamide therapy in the setting of weight loss is effective in IIH patients with mild visual loss (19). I advise my patients to first bring the condition under control with medical therapy, a low-sodium diet, and adequate weight loss and to plan pregnancies only after they are able to discontinue acetazolamide. However, this may not always align with a patient's family planning goals, especially because the duration of acetazolamide therapy varies with individuals based on the response to the medication, severity of the disease, ability to achieve weight loss, and patient compliance. So, it is important to discuss these goals with patients in the early course of the disease, to understand their preferences, and to bring the condition under control quickly. Teratogenesis from the use of acetazolamide has been reported in rodent studies (20), but the potential teratogenic effect of this medication in humans in the first trimester of pregnancy is not clearly known. Limb and dental deformities have been reported in humans with the use of acetazolamide during the first trimester (7). Although there are larger case series that support the safety of acetazolamide in pregnancy (21,22), caution must be taken for the liberal use of acetazolamide in the first trimester.

There are no randomized trials that have studied the teratotoxic effect of acetazolamide in the first trimester of pregnancy. However, based on two case series, acetazolamide can probably be recommended to pregnant patients after informed consent (21,22). Due to the possible risk of fetal deformities and for fear of miscarriage, pregnant IIH patients with persisting papilledema often decline acetazolamide and may require frequent therapeutic lumbar punctures to alleviate symptoms and preserve vision. To avoid this quandary, I routinely emphasize to all female patients to avoid getting pregnant while on the acetazolamide, and also give written instructions with the prescription.

Management of IIH often requires a strong collaborative effort from both the physician and the patient, as it is considered a life-long condition. A thorough understanding of the underlying disease, adherence to treatment, and assistance from support groups can help patients cope with the headaches and psychosocial challenges associated with this condition. Patient education brochures can be obtained from reliable sources such as the North American Neuro-Ophthalmology Society website (www.nanosweb.org).

Patient Engagement in the Management of IIH

LM's presentation to medical attention was characteristic of IIH, however, her illness trajectory provides insight into multiple complications associated with IIH. Patient engagement continues to be recognized as a crucial component of high-quality health care services (23). Involving patients in the

management of their condition enforces patient-centered care and improves clinical outcomes by involving multiple care perspectives (24). There is increasing attention directed toward patient engagement in the context of chronic conditions, not only in research, but also by clinicians that manage these diseases. Patients who take an active role in their disease management enhance their experience and prompt clinicians to be more responsive to deliver better care (25). Studies in other chronic conditions, such as type 2 diabetes (T2DM), highlight the importance of grounding care in individual patient experiences (25). Effective patient engagement invites the patient to be an active partner in the management of their disease, while ensuring care and disease management align with a patient's goals. This can lead to a stronger therapeutic relationship, increased patient activation, capacity building, and better health outcomes (13).

LM's story is an example of this, discussing the individualized care tailored to her unique circumstances. With the help of ANES and a treatment plan built with her individual goals in mind, LM was able to take steps to manage her own disease. She specifically highlighted the importance of an open and honest relationship with her clinicians contributing to a feeling of empowerment, demonstrating that patient engagement improves patient activation and impacts behavior. Some patients may lack this feeling of empowerment, or feel as though have insufficient knowledge. The goal of this article was to highlight the importance of patient contributions, regardless of education level, socioeconomic status, or disease course. While LM was invited to share her experience due to her background in health education, it is her experience as a patient which was important to share to help educate others.

Weight loss, while an essential component of the management of IHH, is often a difficult matter to address between clinicians and patients. In a study of primary care, overweight patients wished for weight management assistance involving diet advice, goal setting, and exercise recommendations, although stated these points were often not discussed (26). In those with T2DM, individualized exercise prescriptions were shown to result in maximal health benefits (13). This provides an interesting comparison to IHH management, where exercise and weight loss are also effective treatments, and this form of patient engagement may be similarly beneficial (13). Weight loss is multifactorial and may be more difficult given a patient's financial means, health status, access to healthy foods, and/or psychosocial supports. These factors may make it more difficult for a patient to take similar steps to be involved in their care. Empathetic assistance from clinicians can support a patient's experience and engage them to meet their goals.

Conclusion

The experiences described by authors ANES and LM exemplify the importance of patient engagement to support

successful outcomes and improve patient activation. The lessons learned can help future clinicians including various specialists as well as primary care providers and patients navigate the management of chronic conditions such as IHH, and help stakeholders better understand one another's perspectives. Ultimately, creating a meaningful therapeutic relationship that values and involves the patient will improve outcomes, and support clinicians and patients in their goals.

Patient Consent

The patient is an active author in this article and wrote their own perspective and story.


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