Idiopathic Intracranial Hypertension Explained

A Guide for Patients and Families

Kyle M. Fargen

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For my wife, Christine, and for Chelsea, Eric, and Cooper. I love you all more than you know.

Preface

This book was written with the purpose of providing patients and families with a guide to help understand a very complex, painful, and scary disease. Our understanding of idiopathic intracranial hypertension (IIH) is surprisingly limited, especially given that we have known about it for decades. I have attempted to explain this disease, to the best of my current understanding, in a manner that can be appreciated by individuals with no medical knowledge. I am hopeful that this will help patients understand why they are suffering and explain the treatments so that they don't seem so scary when they are discussed.

Before going forward, it is important for the reader to understand my perspective on IIH because it colors the book and the way I present information. My personal interest in this disease stems from my training as an endovascular neurosurgeon and my interest in cutting-edge interventional therapies, most notably venous sinus stenting. As a surgeon, my experience is mostly with medically refractory IIH, meaning that most of the patients I see have severe symptoms that have persisted even when treated with medications. Also, as a surgeon, my skill set resides in doing procedures, not in managing headaches with medications. Those patients with mild-moderate IIH that are successfully treated with medications alone are not usually sent to me in referral. The patient population that I regularly treat tend to be people that are suffering with significantly impaired quality of life and therefore are willing to undergo aggressive surgical treatments to get better. As such, I tend to see the patients that are more severely affected by IIH, and my perspective on medical treatments (which tend not to help all that much in this population of patients) tends to be more cynical than other physicians that routinely see success managing patients with mild symptoms with medications alone.

Further, I tend to be more aggressive than the average physician in trying to improve quality of life. This means that, in my practice, I may be more likely to offer stenting or shunting to people than other neurosurgeons around the country. There are practice variations among different physicians in determining candidacy for different surgical procedures, in the work-up and diagnostic tests ordered, in medications used, and differences in surgical technique or devices used. I have developed my personal protocols for patient management based on what I think is

best; however, I am young in my career, am to some degree a product of the places I trained as a physician, and my practice is shaped by the environment and devices available at the hospital where I practice. It is likely that every neurosurgeon around the country has their own patient management strategies. In instances where I think what I do or use really matters, I'll point this out. But otherwise it's probably merely a matter of physician preference.

Finally, I do a fair amount of research studying the brain and the pressures within the skull and publish frequently in medical journals. My default writing style is "doctor speak," which I have done my best to try to hold at bay for this book. Also, I have academic interests in this disease that I discuss because I think they are fascinating, but in reality may not be that helpful to a given patient.

In summary, I think it is important to understand my personal biases in patient management and experience because everything you will read in this book derives from that perspective. Hopefully this book will help you or your loved one understand this perplexing disease and make it seem less frightening.

Winston Salem, NC, USA Kyle M. Fargen

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Thanks to Jennifer Aldridge for taking such great care of my patients and helping me with this book. Without you this book would not be possible!

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Chapter 1
Introduction

Idiopathic intracranial hypertension (IIH, or "pseudotumor cerebri," or "benign intracranial hypertension") is an uncommon condition that causes symptoms of severe headache and visual changes. It is most commonly seen in overweight women in their child-bearing years (usually 20–40 years) but occasionally is seen in other patient groups. The symptoms are a result of high pressure ("hypertension") in the cerebrospinal fluid (CSF) inside and around the brain. Patients with this condition usually present with progressive, chronic headaches. Often times patients will say they have had headaches for months or years that were tolerable but then seek medical attention finally because of progressive worsening to where they significantly affect quality of life. Rarely, patients may present with symptoms other than headache. Some patients have visual blurring or vision loss that occurs in the absence of headache. Some patients only have a loud whooshing sound in their ears that sounds like they are hearing their heartbeat at all times. Others present with dripping of CSF from their nose or ears due to the high pressure squeezing fluid out through small holes in the base of the skull. Others have no symptoms at all but are found to have swelling of the optic disc in the back of the eye when a doctor checks using an eye scope during a routine physical exam. IIH therefore may manifest as a spectrum of symptoms, ranging from mild headache or no headache with no visual complaints to severe, unrelenting headaches with severe visual loss.

The classic symptom of IIH is a pressure-type headache. Most people with IIH say that they feel a gnawing or throbbing ache behind their eyes. This feeling of "pressure" behind the eyes is in fact due to a literal buildup of pressure in the cerebrospinal fluid (CSF) behind the eyes. That fact alone is easy for patients and their families to understand and easy for doctors to communicate: "too high of pressure" causing your headaches. But the rest of IIH—the anatomy, the reasons for the pressure buildup, and the treatments—are not so simple. In fact, as someone who has spent countless hours trying to explain the condition and its treatments to patients and their families in my clinic, I will be the first to tell you that IIH is *not* easy. The anatomy, the normal physiology, and the reasons for the disorder are, let's just say

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complex. Not to mention the fact that we discuss separate and totally distinct "pressures" while discussing treatment options: arterial blood pressure, venous blood pressures, and intracranial pressures. And these have different units of measure that have to be converted mathematically! Talk about confusing.

Over the last few years, I have developed and repeatedly performed a 60-minute run-through of the main principles when seeing patients in my clinic. This summary, which I now have memorized like a speech, starts with the symptoms, then an explanation of pressure within the head, then to why we thought the pressure was high, followed by older treatment strategies based upon this hypothesis, then to our new understanding of the disease, and then finally to our treatment options knowing what we know now. This seems to work as a rough outline, but so much information is left out during this 60-minute discussion that would be of use to patients. Since I now have the ability to talk as much as I want, I am going to walk through an explanation of IIH from the beginning to the end, hitting on all the things that I think are important for patients and their families to understand. This book basically contains almost everything I know about this condition. Some of the information presented will be unnecessary. Much of it will be a little confusing. But hopefully by following this logical stepwise format, we will make a very confusing and scary disease something that is not so complex and not so scary.

Chapter 2
Basic Brain Anatomy and Physiology

The Brain and its Fluid

An explanation of IIH requires understanding a thing or two about how the brain looks and functions. I completely understand that most readers will have little or no medical background and that's OK—the point here is to establish some important anatomical or physiologic concepts because they will help us understand why we do what we do from a treatment standpoint. So let's begin by discussing some normal anatomy about the brain.

The brain sits within the skull and is surrounded by a membrane. This membrane is called the "dura mater," or just "dura." It is a tough membrane that houses the brain and its fluid. The same membrane extends down into the spine around the spinal canal where the spinal cord sits. The brain and spinal cord are surrounded by a clear fluid called cerebrospinal fluid (CSF, also known as "spinal fluid"). CSF is made by the brain continuously, travels through fluid chambers inside the brain, and then bathes the brain and spinal cord, serving as a protective fluid surrounding the nervous system. The dura keeps brain fluid from leaking out. The fluid chambers in the brain are called "ventricles" (note: there are chambers in the heart called ventricles too, but these are totally unrelated). There are two large ventricles called the "lateral ventricles," one on each side of the brain, and they communicate with a smaller midline "third ventricle" (Fig. 2.1). Under normal conditions, the structures of the brain are symmetric, so the two lateral ventricles appear like mirror images of each other.

CSF that leaves the brain's chambers (ventricles) then bathes the brain. Fluid extends forward and surrounds the optic nerve that goes to the eye. Fluid also extends down out of the skull and into the spine where it surrounds the spinal cord. In the lower spine, the spinal cord separates into a bunch of nerve roots that float in this fluid. Therefore fluid in the ventricles (within the chambers of the brain) is continuous with the fluid surrounding the brain, in the spine, and around the optic

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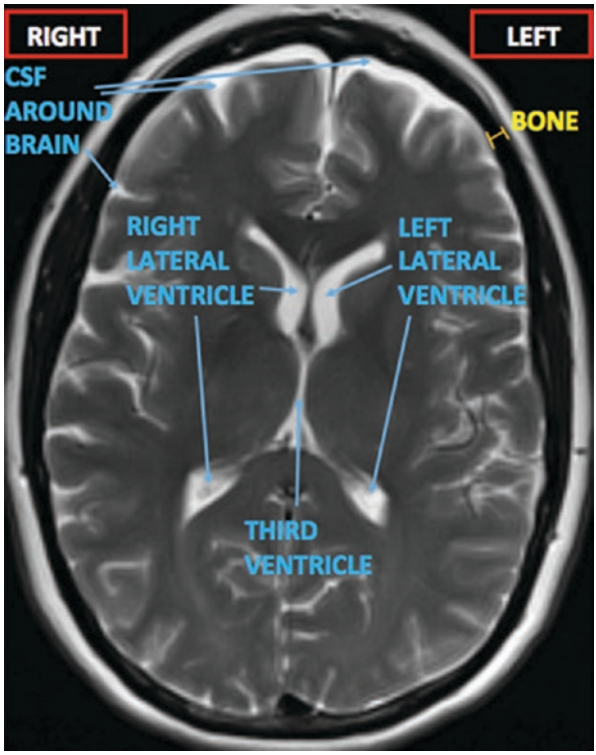

Fig. 2.1 Brain scan with cerebrospinal fluid (CSF) appearing bright. The brain tissue is grey. Everything inside and around the brain that appears bright is CSF. CSF is inside the lateral ventricles and the third ventricle, as well outside and around the brain

nerves. Fluid can move freely within these spaces. Another way to put it: if you mixed a few drops of blue food coloring into the fluid, in a short while the fluid inside and around the brain, outside the optic nerve, and in the spine would all be colored blue, as it would mix throughout all of this fluid freely.

Key Points

1. The brain and spinal cord are surrounded by clear fluid called cerebrospinal fluid, or CSF.
2. CSF is contained within a membrane called the dura, which surrounds the fluid spaces.
3. The large fluid chambers in the brain are called ventricles.
4. CSF in the ventricles mixes freely with CSF outside the brain, around the optic nerves to the eye, and down into the spinal canal around the spinal cord.

The Flow of Brain Fluid

Cerebrospinal fluid (CSF) is made inside the brain, circulates through the ventricles of the brain, then leaves the brain, and eventually surrounds the brain. Most of the CSF gets reabsorbed into the blood stream outside the brain. At any given time, there is about 150 ml of fluid inside and surrounding our brain and spinal cord. To put this into perspective, 150 ml is about 2/3 of a cup or about ten tablespoons. Our brains make about 450 ml per day, which means that we produce (and reabsorb) the entire volume of CSF within and around our brains three times per day. For this reason, fluid drained off (by a spinal tap, for instance) tends to be replaced in a very short period of time (a few hours).

The best way to think of the CSF around the brain and spinal cord is to use the "sink" analogy. I think this analogy simplifies the CSF space into an easily understandable example for most people and helps to explain how our brains make, and then reabsorb, fluid. In the sink analogy, there is a (1) faucet that produces fluid, (2) a drain at the bottom that drains fluid from the sink, and (3) some amount of standing water in the sink (Fig. 2.2). The faucet of the sink is similar to the small organs that make CSF within our brains. In people, the faucet is always "on," because these small little organs are constantly producing CSF. Therefore, in our analogy, consider that the faucet is always producing the same amount of water. This water flow into the sink never stops.

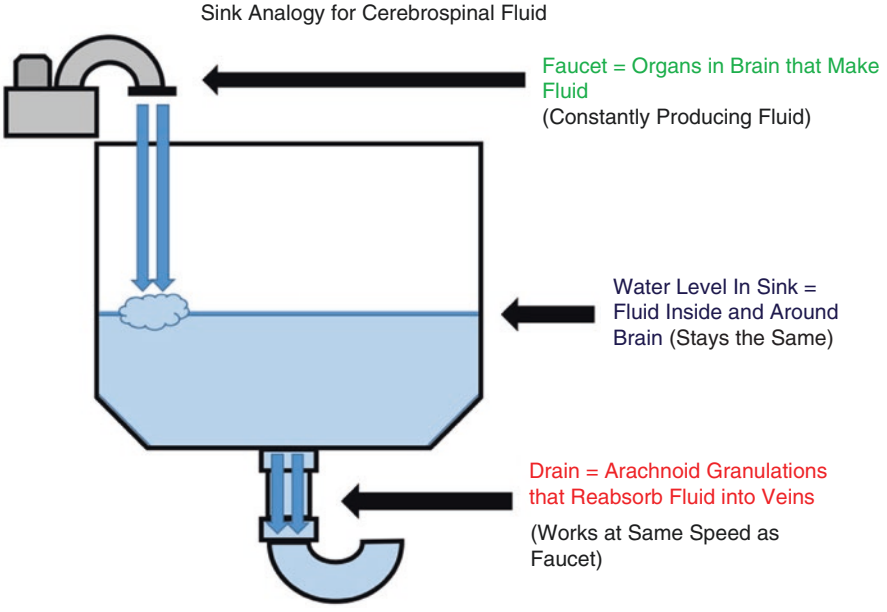

Fig. 2.2 The sink analogy of cerebrospinal fluid (CSF) production and reabsorption. In a normal situation, the water level does not change because the faucet and the drain are working at the same speed

The second key element of the sink analogy is the drain at the bottom of the sink. We have small organs that reabsorb CSF back into the large veins outside the brain. The little organs represent the drain in the analogy. The drain functions only as well as the pipes that lead out of the sink, and the drain holes can be blocked by debris within the sink.

Lastly, the standing water in the sink is similar to the CSF sitting within the ventricles of our brains and in the space around our brain and spinal cords. This amounts to about 150 ml of fluid at any given time (25 mL in the ventricles, the rest around the brain and in the spinal column).

In a normal situation, the water level is constant in the sink. A steady water level in the sink means that the faucet and the drain are working at the same speed (fluid is being produced at the same speed at which it is being reabsorbed). Therefore, in a normal situation, our brains produce CSF constantly, reabsorb CSF constantly, and have a constant volume of CSF inside and around our brains.

In abnormal situations, where either the drain holes are blocked or if the pipes leading out of the sink are partially clogged, water won't move through the drain as quickly. However the faucet is unaware of this and will continue to pour out water at the same speed. This will cause the water level in the sink to rise. For example, a patient with bleeding in the brain may have blood "block the drain holes" which causes a buildup of CSF within the brain, causing increased pressure within the brain and enlargement of the ventricles (fluid chambers) in the brain.

Key Points

1. The brain constantly makes CSF. This fluid constantly gets reabsorbed. CSF drained off (by a spinal tap, for instance) is replaced by the brain over a few hours.
2. The easiest way to think about the flow of CSF is to think about your brain like a "sink." The faucet is always on and therefore always producing fluid, and the drain is always draining water out at the same speed as the faucet is making it, so in normal situations the water level is at a constant level in the sink.
3. Abnormal situations occur when the drain doesn't work as well as it should. A drain that stops draining fluid out of the sink will cause the water level to rise in the sink. This leads to a buildup of fluid (and pressure) inside the brain.

The Reabsorption of Brain Fluid Is into Veins

In our sink analogy, the water that leaves the sink through the drain has to go somewhere. In our brains, the CSF that is reabsorbed from the fluid space around the brain travels through small organelles called arachnoid villi and then into the big veins outside the brain. These veins are large and surrounded by the membrane

outside the brain (the "dura") and therefore are called the "venous sinuses" or "dural venous sinuses." The venous sinuses are not the same as the air-filled spaces of the skull that are also called "sinuses" (sinus infections are of the air-filled spaces in the skull, not the veins).

CSF leaving the brain travels through the arachnoid granulations into the venous sinuses and then mixes with the blood in these veins. As fluid gets reabsorbed, it dissolves into the blood and becomes one with it. This blood is then carried back to the heart through the large veins in the neck and chest. *The fact that CSF gets reabsorbed into the veins outside the brain is critical for understanding why IIH occurs.* So in our analogy of the sink, imagine that the drain at the bottom of the sink leads to the veins outside the brain. In the next few sections, I'll describe the anatomy of the brain veins and those leading back to the heart. Afterward, I'll do my best to explain how this fluid gets reabsorbed and why there is a reabsorption problem in patients with IIH.

Key Points

1. Cerebrospinal fluid (CSF) gets reabsorbed into the bloodstream by traveling from the space around the brain, through tiny organelles called arachnoid villi, and then into the large veins outside the brain.
2. The big veins outside the brain are covered in membrane called the dura and therefore are called "dural venous sinuses."

Brain Veins

Like most organs in the body, arteries carry blood from the heart to the brain. Blood that leaves the brain travels in veins back to the heart. Because the brain needs a lot of blood to function normally, 15–20% of the total amount of blood that your heart pumps goes to the brain. This means that a lot of blood is being pumped to the brain through arteries and a lot of blood is traveling through the veins from the brain back to the heart.

The brain is covered in small veins that collect the blood and drain into the large veins outside the brain, called the *venous sinuses*. Remember that these veins are within the membrane outside the brain, called the dura, and therefore are often called "dural venous sinuses." As mentioned previously, these sinuses are different than the air-filled sinuses (holes) in the skull. The dural venous sinuses are inside the skull, outside the brain. In some people, part of the sinuses may appear to be missing, which may be entirely normal. The word "aplasia" (or "aplastic") usually refers to a normal variant where part of the sinus is missing. You may also see the word "hypoplastic," which is medical jargon for "small."

The main venous sinus in the center of the head is the superior sagittal sinus. This big vein collects blood from both sides of the brain (Fig. 2.3). As it travels down toward the base of the skull, the superior sagittal sinus splits into two separate veins at a site called the "torcula", with one traveling on each side of the head by the ear. These

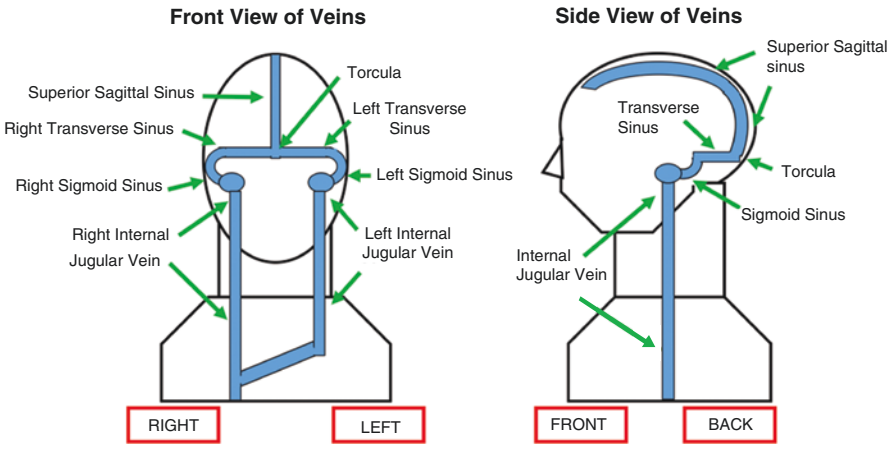

Fig. 2.3 Major veins of the brain and neck, as viewed from the front and the side. A single superior sagittal sinus splits at the torcula into the right and left transverse sinuses, which then travel downward to become the internal jugular veins. In this image, the transverse sinuses are co-dominant (both large and about the same size). On the side view, the right and left transverse sinuses, sigmoid sinuses, and internal jugular veins overlap as they are at the same location on both sides

are called the *transverse sinuses.* The size of the two transverse sinuses can vary. In some people, the two transverse sinuses are the same size (or "co-dominant," meaning both are large). In most people, the right transverse sinus is larger ("dominant"), and the left transverse sinus is small ("non-dominant"). Sometimes, the left transverse sinus is larger, and the right transverse sinus is smaller (Fig. 2.4). The transverse sinus is very important for the treatment of IIH as it is a common site of narrowing.

The transverse sinus on each side takes a turn and becomes the sigmoid sinus. The two sigmoid sinuses then leave the skull by the ear on each side, where the vein becomes the internal jugular vein. The internal jugular vein (or just "jugular vein," or "IJ" for short) travels from the skull down to the chest where it joins the bigger veins by the heart.

Key Points

1. The brain requires a lot of blood. This blood travels to the brain from the heart in arteries and leaves the brain in veins to get back to the heart.
2. The large veins outside the brain are within the membrane outside the brain called the dura and are called the "dural venous sinuses" or just "venous sinuses."
3. The large vein in the middle of the head is the superior sagittal sinus. This vein splits into the two transverse sinuses at a site called the "torcula."
4. In most people, the right transverse sinus is large and the left transverse sinus is small. However everyone is different. In some people, the two are equal in size; in others, the left side is bigger.
5. Blood traveling through the transverse sinus next travels through the sigmoid sinus and then leaves the skull and drains back to the heart in the internal jugular vein.

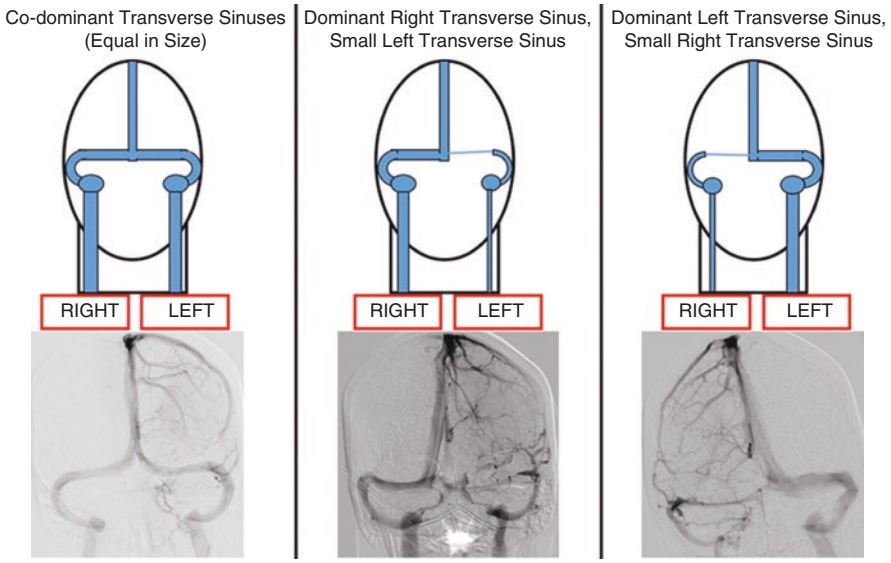

Fig. 2.4 Examples of different sizes of the transverse sinuses. On the left, the right and left transverse sinuses are both large and roughly equal in size. An angiogram of an example patient with co-dominant sinuses is shown below. In the middle is a right transverse sinus dominant illustration with angiogram below. On the right is a left-side dominant example

Blood Pressure: Arteries Vs. Veins

When you go to the doctor and have your blood pressure checked, the doctor is checking the blood pressure in your arteries. A normal arterial blood pressure is somewhere around 130 over 80 and is measured in a unit of measure called millimeters of mercury (*mmHg,* as "Hg" is the abbreviation for mercury). By convention, blood pressures, in either the arteries or veins, are measured in this unit of measure, mmHg. So in a normal individual, arterial blood systolic pressure is somewhere around 130 mmHg.

Veins, on the other hand, have dramatically lower pressures. Even in the biggest veins by the heart, the pressures in the veins are usually 8 mmHg or less. So while arteries have high pressures (130 mmHg) because of the blood being pumped from the heart, when blood is traveling back to the heart, its pressure is dramatically lower (8 mmHg).

Because arteries and veins deal with blood of wildly different pressures, they are designed differently. Arteries bound with each heartbeat because of the high pressure, which generates a "pulse" that you can feel. The walls of arteries tend to be more rigid, meaning that they are harder to compress. For example, feel the radial pulse at your wrist by the base of your thumb. Feel how the artery bounds with each heartbeat. Now push on the artery to make the pulses stop. You have to push pretty hard to make the pulse go away.

Veins, in contrast, are soft and easily compressible. Vein walls are thin because they don't deal with high pressures. Veins, consequently, are easy to squish. Note how easy it is to compress the veins that stick out of your hand. The large veins outside the brain (venous sinuses), just like the veins in your hand, are also easy to squish. This will be important when we talk about IIH, because the transverse sinuses may get compressed which can lead to IIH symptoms. In addition, veins in the abdomen may be compressed by excess body fat which can also lead to IIH symptoms.

Key Points

1. Blood pressure is measured in millimeters of mercury, or "mmHg." Both artery and vein blood pressures are measured in this way.
2. Arterial blood is high pressure and pulsatile. The pressure in veins is dramatically lower in pressure and, even in the biggest veins by the heart, is very low (10 mmHg).
3. Arteries are rigid and hard to compress, while veins are soft and easy to compress.
4. The venous sinuses outside the brain and veins in the abdomen are therefore susceptible to being compressed.

Cerebrospinal Fluid Reabsorption Is Dependent on Venous Sinus Pressures

Thus far, we've hit on some important basic brain anatomy and physiology principles. We've established that cerebrospinal fluid (CSF) is made by the brain constantly, travels through the chambers of the brain (ventricles), and then gets reabsorbed into the venous sinuses outside the brain. Our brains constantly make CSF, and therefore we constantly reabsorb CSF. CSF that gets reabsorbed does so through tiny organs (arachnoid granulations) into the venous sinuses, where the fluid mixes with venous blood. This blood then travels through the superior sagittal sinus, through transverse and sigmoid sinuses, and then through the internal jugular veins back to the heart.

So how exactly does CSF get reabsorbed? CSF reabsorption back into the blood stream occurs via a pressure-dependent mechanism, such that the pressure of the CSF has to be slightly higher than the pressure in the venous sinuses outside the brain for fluid to be reabsorbed. *Understanding this point is critical in understanding why IIH occurs.* Old studies performed in animals suggest that CSF pressure has to be roughly 3 mmHg higher than the pressure in the venous sinuses to allow fluid to move through the arachnoid granulations into the veins. This means that higher pressures in the dural sinuses will cause the CSF pressure inside the brain to be higher. This is confusing, so let's think of a hypothetical patient with our sink

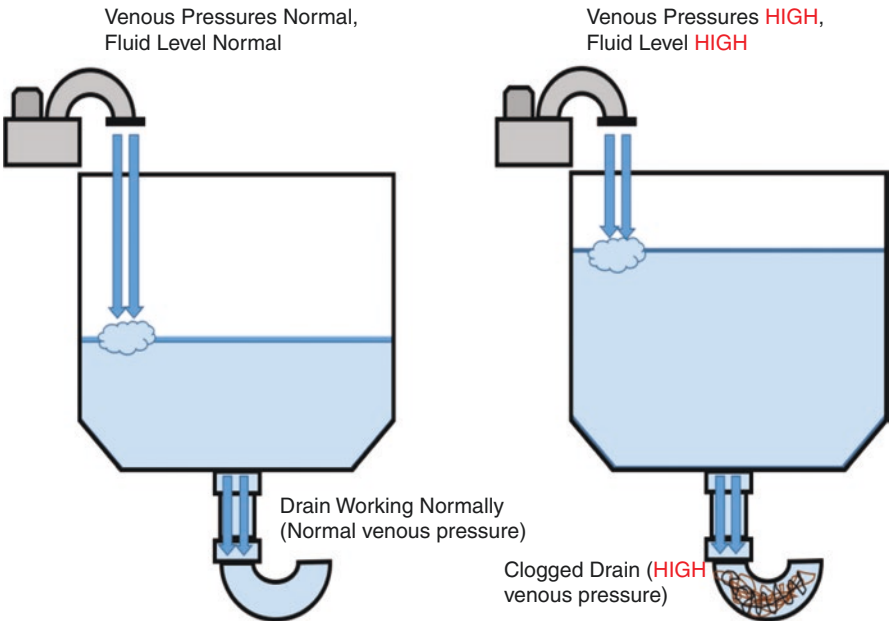

Fig. 2.5 High venous pressures are like a clogged drain. Water doesn't travel as well through the drain, causing the water level to rise, and the pressure in the water to be higher

analogy, where the venous pressure is analogous to a bunch of hair blocking the drain in the sink (Fig. 2.5). The higher the pressure in the veins, the more hair is clogging the drain pipes. Remember that the faucet is always on.

If an average person has a pressure of 10 mmHg in the venous sinuses, CSF will build up in and around the brain until the pressure reaches a value slightly higher than 10, to approximately 13 mmHg. Once this pressure is reached, CSF can get reabsorbed, so now the pressure will stabilize. In our analogy, the water level will rise to a point where enough pressure is in the sink to force water through the hair clogging the drain pipes. Once the water level rises far enough, the water level will stabilize, because the water being produced by the faucet equals the rate at which water is going through the drain.

If we take that person and then increase their venous pressure to 15 mmHg suddenly, no CSF will be reabsorbed until the CSF builds up under pressure to an even higher pressure. The faucet keeps producing CSF, and the drain now allows no water to escape (pressure not high enough yet), so the water level rises. Once the CSF slowly builds up under pressure until the pressure is 18 mmHg, the fluid can now be reabsorbed again, and the CSF pressure stabilizes at 18 mmHg.

Similarly, if we now suddenly decrease the venous pressure in the sinuses to 5 mmHg, CSF is rapidly reabsorbed into the veins until the CSF pressures reaches 8 mmHg, at which point reabsorption slows considerably, and eventually a new steady state is reached.

Therefore, *the pressure in the CSF is dependent on the pressure in the venous sinuses outside the brain* (Fig. 2.6). If the pressure in the veins goes up, CSF pressure will go up. If the venous pressure goes down, CSF pressure goes down. In a normal situation, the drain in the sink has no hair clogging its pipes and is functioning at full speed. In patients with IIH, the drain pipes are clogged, and the reabsorption of CSF only occurs at high pressures, leading to a steady state where CSF pressures are constantly high.

Key Points

1. Cerebrospinal fluid (CSF) reabsorption occurs via a pressure-dependent mechanism into the dural venous sinuses.
2. CSF pressure has to be roughly 3 mmHg higher than the pressure in the venous sinuses to allow fluid to move through the arachnoid granulations into the veins.
3. The pressure in the CSF is dependent on the pressure in the venous sinuses outside the brain. If venous pressures increase, CSF pressures must also increase. If venous pressures decrease, CSF pressures will decrease.

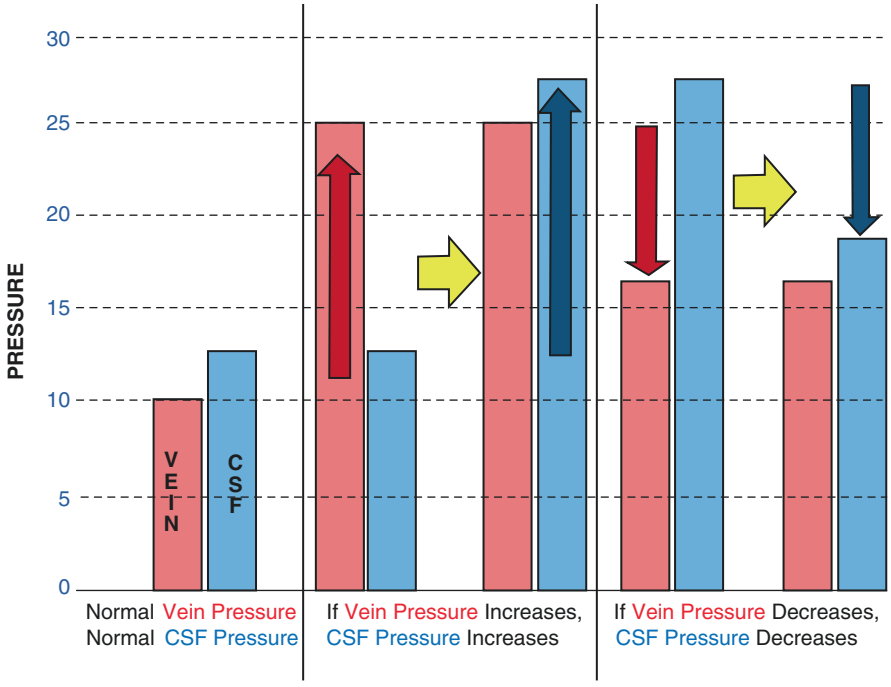

Fig. 2.6 CSF pressure (blue) is dependent on vein pressure (red). If vein pressures increases, CSF pressure will increase accordingly. If vein pressure decreases, CSF pressure will decrease accordingly