Hydrocephalus:

An Owner's Manual

Edison McDaniels, MD

Hydrocephalus:
An Owner's Manual

Edison P. McDaniels II, MD
Board Certified in the Practice of Neurosurgery
by the
American Board of Neurological Surgeons

HYDROCEPHALUS: An Owner's Manual

Edition 1, March 2013

Disclaimer

This is a work of nonfiction. It is not an exhaustive treatment of hydrocephalus, but rather is a useful companion to the shunted patient. It is meant to be informational only. It cannot and should not replace appropriate consultation with a medical professional in general or a neurosurgeon in particular.

What You Won't Find Here

You will not find a bunch of charts and diagrams here. This is a very straight forward monograph, my attempt to pare hydrocephalus to the bones and make it as understandable as possible to the greatest number of folks. There are plenty of books out there that will show the anatomy and the various products we use to treat this unfortunate condition. What is here are the simple and most basic definitions and explanations of hydrocephalus—enough, I hope, to demystify it for the average patient and his/her family.

The reader is invited to seek out more info if desired.

WHY I WROTE THIS MONOGRAPH

I am a brain surgeon.

Several years ago, I was confronted with a young man in the emergency room who had earlier that morning been found unconscious by his college roommates. In fact, when I met him he was essentially comatose, that is, unresponsive in any meaningful way. Fortunately, one of his roommates recalled something about him having a *shunt*. With this piece of information, the emergency physician quickly called for a stat head CT and a diagnosis of *shunt malfunction* was made. I was called, took the patient to surgery for an emergent *shunt revision*, and he recovered and lived happily ever after.

Well almost. It turned out he was a college student and the ordeal left him rather exhausted, though *neurologically normal*, and he would spend several months recovering from his near death experience. His mother, who lived in a city 160 miles away, drove over immediately and was waiting for me when I came out of the operating room. I have seldom seen a mother so grateful as that woman—unless it be virtually every other mother I have ever dealt with as a neurosurgeon.

Largely because of their children, parents are special people.

The bond between parent and child is like no other. I have seen octogenarians break down while recalling the death of a forty-year-old son or daughter—never mind that the death occurred fifteen years before. Perhaps the only bond in all of nature that can never be fully broken, it continues beyond divorce, separation, abandonment, illness, and even death. At its best, the parent-child bond drives us to be our best, to meet our full potential. Even when it is missing, entire lives are predicated, even formulated, on the basis of such a loss.

Almost without exception, the parents I meet would gladly exchange places with their child in these moments of extreme stress. These parents feel helpless and at the mercy of the situation. I am often asked *What could I have done?* Or *Is there anything I can do to prevent this from happening again?*

I know the feeling.

I am a brain surgeon. I am also a parent. Several years ago, my oldest son died suddenly. In my years on this earth I have lost people close to me—a brother, a half-brother, both parents, several close friends—but all of their

deaths paled in comparison to losing a child of my own. It was and remains the single most difficult event of my life, the defining moment if you will.

A bond which cannot be broken.

Which brings me to this monograph.

Hydrocephalus—loosely defined as a build-up of fluid in the brain—is a life-threatening, fairly common, and relatively easily treated condition. Unfortunately, with existing medical technology, the treatment requires a lifelong diligence on the part of loved ones as well as the patient himself/herself. But that being said, the treatment is not onerous on a daily basis and the benefits are dramatic. Most patients with hydrocephalus live normal lives in virtually every respect. They play sports (even extreme ones), marry, have regular jobs, carry babies through labor and delivery, and die as an old man or woman (or at least we expect they will—the technology is only fifty or so years old and thus people shunted as young children are only now reaching late middle age). With the possible exception of the more remote parts of Alaska, if you live in the United States you almost certainly have at least one friend, acquaintance, student, or co-worker with a *vp shunt*—though you may not know it.

So why this monograph?

Because, to put it in the simplest terms possible, failure to recognize a shunt malfunction can be fatal. The boy I took care of above had failed to get out of bed for class one morning. When his roommates returned home for lunch, they found him unresponsive and still in bed. They called an ambulance and he was taken to my hospital, where a CT of the brain showed the problem. He received prompt medical attention—but only belatedly and it nearly cost him his life.

Had his roommates known the gravity of his failure to arise that morning, his brush with death would likely have been avoided. His mother recognized this fact. She knew how close to the edge he had come. She was one of those who asked *What could I have done? Is there anything I can do to prevent this from happening again?*

The advice I gave her became this monograph.

NEUROSURGERY 101

What is Hydrocephalus?

Loosely speaking, hydrocephalus is an abnormal build-up of spinal fluid within the head. This can happen for a number of different reasons. I will define hydrocephalus more fully in a moment, and we will look at a number of different ways it can come about, but first a quick look inside the box.

A Closed Box

The skull is, for all its complexity, essentially nothing more than a container, a closed box. Clinically speaking, that is looking at it from the neurosurgeon's point of view, it has only three important openings: the paired openings for the eyes and a third opening through which the spinal cord exits at the base of the skull.

The two openings for the eyes allow the optic nerves to pass thru the skull to the eyeballs themselves. The optic nerves are actually not nerves at all, but true extensions of the brain itself—which makes them the only part of the brain visible from outside the body under normal circumstances. Every time you go to the doctor and he looks in your eyes with that bright light (an ophthalmoscope), you are having your brain examined. What he or she is actually looking for is the shape and color of the optic nerve head, that is, that portion of the nerve just as it enters the eye. Pressure inside the head, we will talk about this below, gives the nerve head a different appearance depending on whether it is high or low, normal or abnormal, old or new, etc. That is, the pressure inside the head is propagated along the nerve and causes visible changes in the eye when that pressure is abnormal.

So, the skull is, for our purposes, a closed box. One important caveat: a closed box has a fixed volume (think of your dishwasher—you can only fit so much inside it). The skull cannot expand. That is, there is only a certain amount of space inside the skull.

Contents of the Box

As far as the neurosurgeon is concerned, there are only three things inside the skull that matter and take up any space: the brain itself, the blood (which normally flows through the brain in blood vessels but sometimes hemorrhages and then is found outside the blood vessels, where it takes up space needed by the brain), and cerebral spinal fluid (CSF).

Brain, blood, and CSF.

Each of these substances takes up a certain amount of space. The key thing to remember is this: *an increase in any one of these substances requires a decrease in the other two.*

Why?

Because the skull is a closed box. And a closed box has a fixed volume.

Think once again of your dishwasher. Say it holds plates, glasses, and pots, but only a limited number of each. Of course, you can put in more glasses if you desire, but only at the expense of fewer plates and pots.

Intracranial Pressure

Lets do a thought experiment. You are blowing up a balloon and watching it expand. It expands for two reasons: the walls are flexible and you are increasing the pressure inside the balloon.

Now imagine the walls are not flexible but semi-rigid. But you have superhuman strength and can still blow into the balloon and expand it, though now the balloon expands only very slowly. It enlarges in an attempt to keep the pressure inside the balloon equal to that outside. It will continue to enlarge until one of three things happens: the balloon pops, you can't generate enough pressure to expand the balloon further (in this case the balloon is at equilibrium inside and out), or the walls encounter something external that is more rigid and prevents their further expansion.

Now imagine you are blowing up the balloon inside a closed box. And that box is the skull.

Now imagine the balloon to be any one or more of the three substances—brain, blood, or CSF.

You will see from this little thought experiment that the interaction of the three substances (brain, blood, CSF) inside the closed box of the skull creates a certain pressure, called the intracranial pressure (ICP). Normally, the ICP is in equilibrium across the three substances, but what happens if one of the substances grows in volume?

Well, as we have seen above, the other two must decrease. Or the pressure, the ICP, will rise.

It is this rise in the ICP under adverse conditions that is so dangerous inside the head. Why? We'll get to that.

For now, just know that as a first approximation, if the ICP rises higher than the blood pressure, blood won't be able to flow into the head and the brain will fail. This is what happens when a person suddenly faints. The two common

reasons for fainting: the blood pressure suddenly falls or the ICP suddenly rises. Either way, the ICP eventually exceeds the blood pressure, blood flow to the brain fails, and the patient faints (if they are lucky, they will fall on flat ground and, with the brain and heart now level, blood flow to the brain resumes and the person wakes up).

Some causes of a sudden fall in blood pressure:

—A bad coughing fit, which actually reduces blood return to the heart and thus there is nothing to pump out to the brain;

—Sudden heart failure, such as a heart attack;

—A bad scare, in which case the blood vessels of the body sudden dilate wide open and the blood pressure falls as a result.

—Have you ever heard of micturition syncope? That's the fancy medical term for what happens when a person (usually an older man) is straining to pee and suddenly faints. Why does this happen? Because, as any man past middle age knows, the enlarged prostate gland partially obstructs the flow of urine out of the bladder. The greater the obstruction, the harder a man has to push to pee. This pushing, or straining, effectively increases the pressure inside the belly, which in turn decreases the return of blood to the heart (though only transiently). Just as with a bad coughing fit, there's nothing in the heart to pump and the brain fails. Result: a faint. Fortunately, the victim of such quickly awakens once he faints and thus stops straining to pee. Unfortunately, he may have cracked his head on the toilet...

Some causes of a sudden increase in the ICP:

—An acute obstruction to the flow of CSF, i.e., acute hydrocephalus (the usual cause of this is a shunt malfunction. A less common cause is a brain tumor);

—A spontaneous hemorrhage in the head, such as a ruptured aneurysm.

—A blow to the head, such as one might sustain from a simple fall (say, while straining to pee standing up).

A Recap

The skull is a closed box.

Only three important things take up space in the box: brain, blood, and CSF.

An increase in any one of these substances requires a compensatory decrease in the other two, or...

Or the ICP will rise, usually to the detriment of the patient.

NEUROSURGERY 102

Compartments

It turns out that the interior of the skull is divided into several areas, that is, it is *compartmentalized*. Essentially, there are three compartments: the *supratentorial* space, which is divided into right and left halves by a tough shelf of tissue called the falx; and the *infratentorial* space, which is not divided right from left but is separated from the supratentorial space by a second tough shelf of tissue call the tentorium.

The supratentorial space houses the cerebral hemispheres, within which are the right and left *ventricles*. We shall look at the ventricles in a moment, as they are crucial to the issue of hydrocephalus. The supratentorial space is located in the top half of the head, essentially above the level of the ear holes.

The infratentorial space is at the back of the head, below the ears. A small space, it houses the highest priced real estate in the brain: the so-called *brainstem*, which controls important but mundane things like breathing, swallowing, pulse, and blood pressure (to name but a few; there is also an area here seemingly devoted to vomiting—called the *area postrema*—and pressure here produces, you guessed it, vomiting).

ICP Revisited

We have already seen how the pressure inside the head, the ICP, must be at equilibrium. What this means in reality, is that pressure across the compartments mentioned above must be at equilibrium. Since every high school student knows that items move from an area of higher pressure to an area of lower pressure (this is why storms move across the atmosphere and forecasters and ship captains pay inordinate attention to barometric pressure), it stands to reason that if the pressure rises in one compartment more than another, shifts may occur inside the head. That is, the brain (or part of it), might move from one compartment to another!

In clinical terms, this is called *herniation* and it is deadly.

Herniation

As a neurosurgeon, everything I do inside the head must take into account the possibility of herniation. The last thing I want is the brain shifting around. Fortunately, it turns out one can predict these shifts fairly easily. And if one can predict them, one can prevent them. Usually.

Herniations (think of them as unwanted shifts of brain substance) occur when the ICP goes out of equilibrium because of an increase in one or more of the three important substances mentioned above (brain, blood, or CSF).

Increases in brain substance are represented by brain tumors, of which there are many kinds (some cancerous, some not).

Increases in blood substance are represented by bleeding inside the head. There are many different types of such hemorrhages, some requiring emergency surgery to remove.

Increases in CSF are represented exclusively by hydrocephalus. In fact, the definition of hydrocephalus is an unwanted and pathologic build-up of CSF within the skull, either inside or outside of the brain. The remainder of this monograph deals with hydrocephalus, a few of its variants, and how it is treated by modern neurosurgical techniques. Please note that what follows is not an exhaustive discussion but is for informational purposes only. Nothing here is meant to supersede or replace consultation with a competent expert, usually a neurosurgeon.

The Ventricles

The brain floats.

In the normal course of things, the brain floats in the liquor cerebrospinalis, CSF. The CSF is produced in the ventricles, which are four cavities deep within the substance of the brain, usually rather small and inconsequential.

Three of the ventricles, the right and left *lateral ventricles* and the *IIIrd ventricle*, are located in the supratentorial space. The right and left lateral ventricles are offset to the right and left of the body's midline, and connect with the IIIrd ventricle through a small opening called the foramen of Munro. The foramen of Munro is the first choke point in the system. Choke points are areas small enough to be blocked, or at least partially obstructed, and so have the potential for trouble. The IIIrd ventricle is on the midline and is very close to the exact center of the head. It has a very small tail off of its back end, a narrow tube called the cerebral aquaduct (about the diameter of a pencil lead normally), through which every drop of CSF produced in the lateral and IIIrd ventricles must pass (a major choke point) on its way to the *IVth ventricle*, which is located on the midline in the infratentorial space. The IVth ventricle in turn opens into the wider spaces at the base of the brain through three openings, called foramina, which rarely cause problems.

CSF is actually absorbed into the venous system across the surface of the brain at the top of the head. Unfortunately, the absorption of CSF can fail

following hemorrhage or infection (which perhaps gums up the works and thus prevents the reabsorption). This failure of absorption, combined with the continued production of CSF in the ventricles, leads to one common form of hydrocephalus called communicating hydrocephalus.

TYPES OF HYDROCEPHALUS

The Kinds of Hydrocephalus

In theory, there are two major causes of hydrocephalus: over-production and under-absorption. In reality, over-production is rare, probably less than 1% of cases and generally caused by a tumor of the cells producing CSF.

Under-absorption is the culprit in all common forms of hydrocephalus. In general terms, there are two types:

Obstructive hydrocephalus & non-obstructive hydrocephalus.

Let us look at these two groups separately.

Obstructive Hydrocephalus

Obstructive hydrocephalus (also known as noncommunicating hydrocephalus) relates to the fact that one of the choke points has been obstructed and therefore CSF is building up behind it. Think of a hose attached to a faucet. Turn the faucet on and crimp the hose with a pair of pliers. The hose is the CSF pathway, the water the CSF itself. The crimp is the choke point being occluded or obstructed. Imagine what would happen if the water continues to flow into the hose and can't get out the other end: it balloons to the point of bursting. That is obstructive hydrocephalus—continued production of CSF in the face of an obstructed flow pathway. The CSF is being made but can't get out. In this situation, trouble is bound to develop, usually sooner rather than later. These patients can get very sick, very fast. Within hours.

This situation is commonly caused by an obstruction at the level of the cerebral aquaduct between the IIIrd and IVth ventricle. This is called *aquaductal stenosis* and is a very common form of hydrocephalus. It is more common in young people and children.

Obstructive hydrocephalus is usually caused by a structural lesion, such as a tumor creating a choke point (which eventually obstructs entirely) or a veil of tissue obstructing the aquaduct. If it is a tumor, surgery to remove the tumor may be possible. However, removal of the tumor does not guarantee resolution of the hydrocephalus.

Nonobstructive Hydrocephalus

Nonobstructive hydrocephalus (also known as communicating hydrocephalus) is the second common form of hydrocephalus. Not a structural lesion, there is no issue with the choke points. Consider our hose analogy above. In the normal course of events, water will flow out of the hose, where it is reabsorbed—that is re-circulated—back to the beginning. In nonobstructive hydrocephalus, the problem is that the CSF is not re-circulated, that is, it is not properly reabsorbed into the veins of the head. Think of these veins as filters. In this type of hydrocephalus, the filters are clogged with debris.

Usually this occurs after a bad infection, such as some types of meningitis, or a hemorrhage (bleeding) inside the head. Think of these things as muddying the water, or clogging the CSF.

The result is serious, though often nonobstructive hydrocephalus is better tolerated, perhaps because the obstruction in this case is rarely complete. The patient therefore still has some level of CSF reabsorption.

It may take days, or sometimes weeks to get sick when their shunt fails. Other patients just feel persistently lousy and never get acutely ill. These patients may have low grade headaches and mild cognitive problems (trouble thinking). This is sometimes referred to as *subacute hydrocephalus*, of which there are other causes as well.

Normal Pressure Hydrocephalus

A third type of hydrocephalus is more mysterious than the others. It is called normal pressure hydrocephalus. In this situation, the pressure is normal but the ventricles are still much enlarged. It is as if the set point for the entire system has been moved, but the brain still suffers.

—Most common in the elderly.

—Sometimes misdiagnosed as alzheimer's disease or senile dementia.

—Not a lethal condition, even when left untreated or a shunt malfunctions.

—Not an urgent matter and requires a careful evaluation since the risks of shunt placement may be higher in these patients (in fact, in certain situations this risk may outweigh the potential benefit of shunting—such a patient should not be shunted).

—Occasionally a similar condition is seen a month or more following aneurysmal subarachnoid hemorrhage (this is another form of subacute hydrocephalus; patients with such a history will very often benefit from shunt placement).

SIGNS AND SYMPTOMS - NEUROSURGERY 103

Intro

The benefits of shunting include treating the hydrocephalus and its attendant signs and symptoms. Symptoms are what a patient complains of (headache, nausea, vomiting, double vision, blurred or decreased vision, clumsiness, etc.) and signs are what a physician discovers on examination (decreased consciousness, abnormal eye movements including sunsetting and crossed eyes, enlarging head circumference in infants, etc.).

Consciousness

The most important sign of hydrocephalus is the patient's level of consciousness. The more advanced the hydrocephalus—that is, the greater the pressure in the head—the sleepier the patient becomes. In advanced cases, the patient can lapse into a coma, even death.

Headache

Headache is an important but too general symptom of hydrocephalus. In this context, it is rarely important in isolation, but gains more significance in the company of other symptoms, such as nausea and vomiting, double vision, and abnormal eye movements.

In a shunted patient, the combination of headache and one or more of these symptoms should not be ignored.

The Eyes

The eyes are very telling in shunt patients. Obviously abnormal eye movements—where such abnormal movements did not exist previously—must be investigated emergently.

In more subtle cases of shunt malfunction, the eyes may have a glassy appearance. Often the parents will notice this when others don't. Such a child

should be watched carefully.

Nausea & Vomiting

Much like headache, nausea and vomiting is too general to be of much use in isolation. It is more useful in the presence of other signs and symptoms, such as headache and/or double vision, which would suggest the need for emergent evaluation.

Nausea and vomiting associated with diarrhea, or without any other associations, are not likely related to the shunt, though if the symptoms persist beyond a few days evaluation by your doctor is advisable.

Belly Pain

While some shunt infections can cause belly pain, a non shunt problem (including a simple viral infection) is much more likely. This is not a cause for concern unless persistent, severe, and/or associated with vomiting or diarrhea, in which case see your physician. It is unlikely to be a shunt problem in any event.

Fever

Fever is too nonspecific to be of use when in isolation. Again, fever in the company of other findings (such as discussed under headache) may be more significant.

Irritability & Poor Feeding

These symptoms are seen in infants and perhaps others who lack the ability to communicate readily.

The patient becomes something other than himself, being more cranky, irritable, and not wanting to play. By poor feeding is meant a tendency not to take the bottle well or not wanting to eat. Some babies in this situation will spit up whatever they are fed, even small volumes. This can lead to dehydration and thus should be attended to with some urgency.

Seizures & Opisthotonus

Rarely is a seizure a sign of a shunt problem. This is true even in patients with an underlying seizure disorder. Many patients with shunts have underlying seizure disorders and the two are not necessarily related. The exception is in a patient with a sudden flurry of seizure activity and no discernable cause. In such a case, the shunt should be evaluated by a neurosurgeon.

There is one important notice that must be taken in regard to shunts and seizures. Opsithotonus is a condition loosely defined as extreme arching of the back and especially the neck. In opisthotonus, the patient's head is thrown back

forcibly and the appearance is one of extreme discomfort with the neck in extension. Attempts to straighten out the patient meet with great resistance and may be impossible. The patient may have a distant look in the eyes, or may be unconscious altogether. Opisthotonus may be intermittent or sustained.

Opsithotonus is not a seizure, though at times it is referred to confusingly as a 'brainstem seizure' or brainstem spell. It is an ominous discovery, indicating one of the herniations (brain shifts) alluded to above. Its importance cannot be overstated. Patients with opisthotonus are at a life-threatening moment and must be evaluated and perhaps operated on immediately. I once ran down three flights of stairs with a young child in my arms, his neck arched in opisthotonus all the way. The elevator had failed.

Opsithotonus is not a seizure, but to the uninitiated it can look like a seizure. In a patient with shunted hydrocephalus and no history of seizures, the report of a seizure should never be taken at face value. Given even the slightest bit of arching, one should assume a life-threatening shunt malfunction and act accordingly.

THE SHUNT ITSELF

What is a Shunt?

The shunt itself is a silastic tube about the diameter of a wet spaghetti noodle. It has two ends, proximal and distal, and one valve. There is usually a reservoir which can be needled to gain CSF as well. Often, though not always, the reservoir and the valve are located together.

The proximal end of the shunt is the head end, with the tip usually in the ventricle (one of the cavities in which CSF is produced).

The distal end of the shunt is usually in the belly, or peritoneum. This is not truly in the stomach, but in the space around the stomach and intestines, where the catheter floats among these structures. The CSF exits the distal end of the shunt and is reabsorbed into the bloodstream, where it circulates back to the head and is reused.

The valve is the crucial and significant mechanical part of the shunt. It is placed inline between the proximal and distal tubing, usually under the skin of the skull behind the hairline. It is the 'unsightly bump' seen atop the head of an infant, the lump visible after the head has been shaved for a shunt revision.

The function of the valve is to provide a certain resistance to flow of CSF so that overdrainage does not occur. Without the valve, it is possible that too much CSF would drain, creating its own set of problems.

There are a number of valves from different manufactures. In general, they can be divided into programmable and nonprogrammable valves. Programmable valves can be adjusted with a magnet held over the scalp. The advantage of this: the valve setting can be adjusted without requiring an operation; nonprogrammable valves do not have this option—if the valve setting is wrong, surgery is required to replace the valve. Note that programmable valves may have to be reset after an MRI. It is at least conceivable that a programmable valve could be reset in other situations of modern life, though this is apparently rare.

All valves are subject to mechanical failure and tend to be a choke point in the shunt system.

Risks & Benefits of Surgery

Most of the time, shunt placement is done without a hitch. However, the procedure is not without certain risks. For most individuals, the benefits far outweigh the risks.

The benefits of shunting include treating the hydrocephalus and its attendant signs and symptoms. Symptoms are what a patient complains of (headache, nausea, vomiting, double vision, blurred or decreased vision, clumsiness, etc.) and signs are what a physician discovers on examination (decreased consciousness, abnormal eye movements including sunsetting and crossed eyes, enlarging head circumference in infants, etc.). Another benefit not to be overlooked is the opportunity for an individual to reach their full potential. Even in non-life threatening cases of hydrocephalus, the hydrocephalus can cause memory disturbance and cognitive decline.

With the above said, it should be recognized there are many risks of shunting. The biggest risk is infection, perhaps as high as ten percent. This includes superficial wound infection, infection of the shunt itself, ventriculitis (an infection of the ventricles where CSF is produced), and peritonitis (infection of the belly cavity). Most cases of infection occur within thirty days of surgery, though the risk is elevated for as long as six months. Most cases of infection will require removal of the shunt, perhaps with a prolonged hospitalization.

Other risks include intracranial hemorrhage (bleeding), stroke, and injury to the bowel. The need for revision is always out there as well, usually because of infection or obstruction, occasionally because of misplacement of the distal catheter outside of the peritoneum.

In preterm infants, there is a risk the peritoneum will not have enough absorptive capacity. For this reason, shunting is avoided in these cases.

Ventriculoperitoneal shunts can cause or enlarge inguinal hernias, much more commonly in boys. Small boys should be checked for hernia in the months following shunt placement. If possible, hernias should be repaired prior to shunting.

With a ventriculoatrial catheter, in which the distal catheter is placed in the heart, there is a potential for clotting of the end of the distal catheter. Also, if the shunt becomes infected, the infection can track directly into the blood stream —a decided disadvantage. For this reason, the usual shunt of choice today is a ventriculoperitoneal shunt.

Rarely, ventricular shunts are placed to other parts of the body, the most common being a ventriculopleural shunt. These are exceptional cases, with their own attendant risks.

Shunt Failure v. Infection

When a shunt fails, the signs and symptoms of hydrocephalus return.

The usual cause of shunt failure is either obstruction or infection.

A shunt can fail in the absence of infection. The usual cause for this is obstruction, usually the proximal end or the valve. In such a case, the surgeon

will perform a revision, replacing just that part which is obstructed, if possible.

In the case of shunt infection, the shunt will usually (but not always) shows evidence of malfunction as well. Such cases can be very complicated to treat, especially if the patient is shunt dependent. Generally, these are treated with IV antibiotics in combination with revision. Usually the entire shunt system has to be pulled and replaced. If the infection is significant and the patient is shunt dependent, the entire shunt is removed and an external ventricular drain (which drains CSF to a bag at the bedside) is placed. This may be required for a week or more, during which the patient must remain hospitalized.

If the patient is not shunt dependent (as in normal pressure hydrocephalus), the infected shunt is removed, the patient is treated with IV antibiotics, and the shunt is replaced at a later date. No external ventricular drain is necessary.

Note that essentially all children and most adults are and should be considered shunt dependent from the moment a shunt is placed. In the absence of information one way or the other, the neurosurgeon will assume shunt dependence.

A MOTHER'S COMPANION

Imaging

Generally, a CT is a better study to look at shunt function than is an MRI.

When evaluating shunt function in the ER, the size and shape of the ventricles on CT are of most significance. Being able to compare a CT obtained in the ER under symptomatic conditions to a past CT obtained for simple surveillance (during a period when the hydrocephalus was asymptomatic and the patient was well) can make all the difference in the world (and possibly prevent unnecessary surgery). Such a comparison will often make the diagnosis of a shunt malfunction much easier.

It is reasonable to obtain a surveillance head CT (without contrast; your doctor will know the meaning of this) every three years during childhood through the growth spurt at puberty. In adults, surveillance CT is obtained following a shunt revision but is rarely necessary otherwise. In reality, shunted patients are occasionally seen for headache in the ER and often a CT scan will be obtained at that time. These studies are often normal and reassuring to the physician and the family.

Most patients with a shunt and a headache will not have a malfunction and do not need a CT scan. This is a clinical decision.

It is worth remembering that a CT is a static image of one moment in time. It does not replace a careful history and physical examination.

A shunt series is often helpful. This is a plain set of regular xrays, including at least two views of the skull, chest, and abdomen. The purpose is to follow the shunt tubing throughout its course, looking for kinks, disconnections, or tubing out of place (such as when the abdominal catheter has pulled up out of the belly and is coiled under the skin—where it won't work properly).

Medic Alert

Once a child starts kindergarten, a medic alert tag is recommended, ex. *Obstructive hydrocephalus with VP shunt*.

Traveling

Carrying a digital copy of the latest surveillance CT when traveling will help in the evaluation of a person with a shunt issue. Check with your local hospital or physician as to how to obtain a copy of the CT scan on CD or flash drive.

The Special Case of Infancy

Throughout this monograph, we have assumed the skull is a closed box with a fixed volume. However, this is not the case in infancy, up to about eighteen months of age. During this time, the head is rapidly expanding with normal growth and the skull is composed of several plates of bone, which interact with each other along what are called suture lines. These sutures are very pliable and capable of separating, allowing for expansion of the skull and the underlying brain.

Therefore, the volume of the space inside the skull is not fixed in infants. The box is open.

What this means in practical terms is that hydrocephalus in infants is manifested by an enlarging head circumference, which is why your pediatrician measures the head circumference at the well baby checks.

One can also feel along the sutures, which separate and leave gaps in hydrocephalus.

The anterior fontanelle, the soft spot on the top of a baby's head, becomes very tense or taut from the underlying pressure.

An important finding at the time the diagnosis is made, as well as in the presence of a shunt malfunction, is that of *sunsetting*. This occurs when pressure inside the head affects that part of the brain which controls eye movements. The eyes are forced to look down, sometimes very forcibly. This is an ominous finding and requires emergent evaluation and intervention to prevent permanent brain injury or death.

One more thing. Although infants can get pretty sick from hydrocephalus, only in the most extreme cases do they suffer acutely life-threatening problems with a shunt malfunction (because the head just expands a little more; in untreated infantile hydrocephalus, the head is capable of truly monstrous expansion). Adults, on the other hand, with a fixed amount of space, very often have acutely life-threatening issues with shunt malfunction and hydrocephalus. There is little room for compensation in adults, in whom the box is always closed.

IIIrd Ventriculostomy

In the presence of obstructive hydrocephalus (but not communicating or non-obstructive hydrocephalus, and not normal pressure hydrocephalus) there is sometimes an alternative to shunting: IIIrd ventriculostomy.

In this case the surgeon creates an artificial opening between the obstructed ventricle and the CSF space at the base of the brain. This effectively

bypasses the obstruction to the flow of CSF and allows it to be reabsorbed through regular pathways.

IIIrd ventriculostomy is performed through an endoscope by a qualified neurosurgeon and is only available when the anatomy is favorable. There are potentially significant complications, but the benefit if it works is that lifelong shunting can be avoided.

My Hydrocephalus Log

My neurosurgeon is:

Recommend that every adult family member keep the neurosurgeon's card in their wallet or purse, or on their iphone. Give the card to college roommates, landlords, and place in employee health records as well.

Type of hydrocephalus: Obstructive (non-communicating)
Non-obstructive (communicating)
Normal pressure hydrocephalus (NPH)

The original cause of my hydrocephalus:
Congenital (born with it)
Spina bifida
Aquaductal stenosis
Hemorrhage
Meningitis
Post-trauma

Date of most recent shunt:
Type:
VP
VA
Manufacturer:
Programmable:
Current setting:

Date of first shunt:

Number of revisions:

Have I ever had a shunt removed because of infection:
Date:

About The Author

Edison McDaniels is a writer, wordsmith, novelist, and physician living in the American midwest. His writing tends to involve ordinary people in extraordinary circumstances and is often informed by medicine. His stories showcase historical fiction and the supernatural, especially ghosts. He received honorable mention in The Seventeenth Edition of the Year's Best Fantasy and Horror (2003), and has been published in Paradox Magazine, The Summerset Review (available online), The Armchair Aesthete, On The Premises Magazine, and others. Several of his short stories can be found online.

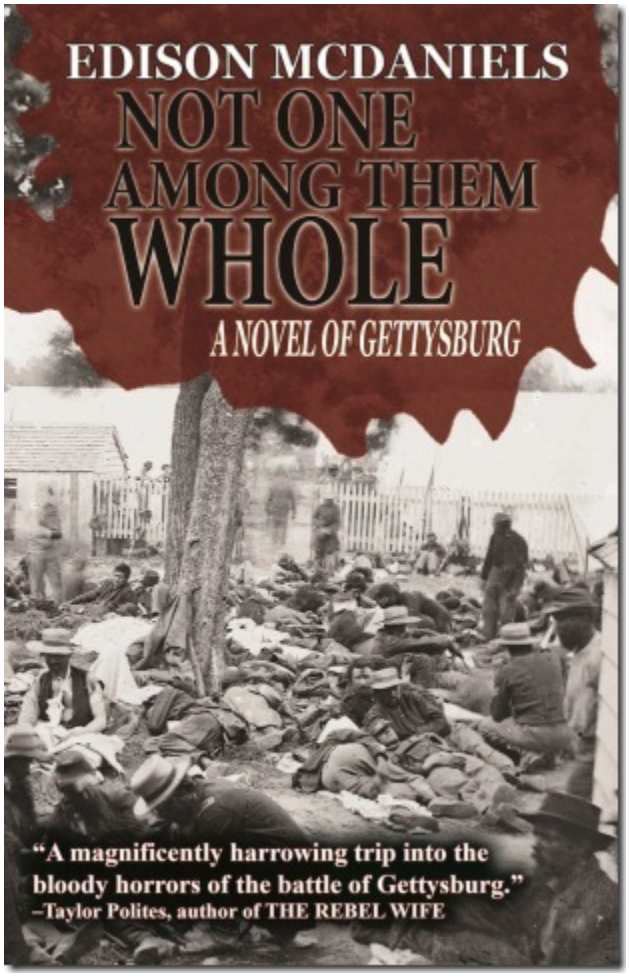
He is also a graduate of Stanford University and is a neurosurgeon. He is board certified in the practice of adult and pediatric neurosurgery, with over 7,000 operations to his credit.

He and his wife collect historical etchings and attend at least 1-2 baseball games a week between April and October, more if the Minnesota Twins are in town.

His novels include NOT ONE AMONG THEM WHOLE, THE BURDEN, and the forth coming THE MATRIARCH OF RUINS. His latest novella, BLADE MAN, is available as an eBook.

Hydrocephalus: An Owner's Manual is his first nonfiction work for the masses. However, he blogs about hydrocephalus and all things neurosurgery at www.surgeonwriter.com.


www.surgeonwriter.com
NEUROSURGERY101—TheBlog
Damn
Interesting



Amazon 5.0 out of 5 stars

Engaging, Heart-Breaking and Absolutely Fantastic February 13, 2013

By D. Buxman TOP 1000 REVIEWER VINE™ VOICE

Format:Kindle Edition

I'm not a Civil War buff. I've always enjoyed history, but I've never been crazy about historical fiction. NOT ONE AMONG THEM WHOLE was the most pleasant surprise I've had in the past year. I was hooked from the first chapter. The writing is crisp, the dialogue is engaging and the plot lines are intricately woven and wonderfully timed. I would give this book 6 stars if I could. Edison McDaniels has a rare gift for honing the experiences of an epic battle into fine points of individual struggle and sacrifice. The descriptions of primitive surgical techniques nearly caused me physical discomfort at times, but I kept coming back for more since I truly cared for the characters. Although I finished the book a few days ago, I still find myself thinking about it. This is a terrific book that I will likely read again. I can think of no higher praise.

Amazon 5.0 out of 5 stars
A must for Civil War buffs!, January 22, 2013
By **P. B. Sharp**
(TOP 500 REVIEWER) (VINE VOICE) (REAL NAME)
This review is from: NOT ONE AMONG THEM WHOLE: A Novel of Gettysburg (Kindle Edition)

Gettysburg, 1863. Once a meadow of golden wheat, now a playing field of horror, where men were programmed to play a part, to play a position. If their part was to die, they did. If their part was to heal others, they did. North and South came together in an unholy skirmish in which men were drained and bled and left somewhere on that playing field shattered in body and in mind. It was dog eat dog to the men in blue and grey- kill or be killed. "The only real commodity was suffering and the only true coin was death."

The surgeons working in unspeakable conditions, perhaps in an old church or in an abandoned shed had to resort to ingenuity, such as making a splint from the bone of a dead horse, or creating a tube into a soldier's shattered windpipe by wrapping a wire tightly around the neck of a bottle, breaking the bottle and gently inserting the wire in place. Author McDaniels is a surgeon and he takes you there, takes you into the heat of battle and introduces you to the horrors that were Gettysburg.

The novel is built around vignettes. McDaniel's fine Civil War novel is not the world of Robert E. Lee or Ulysses S. Grant or even Abraham Lincoln. McDaniel's Gettysburg is a microcosm, a seething world of its own from which no player escapes. The reader follows the characters as they play their parts in the murky tapestry of civil war. There will be a price to pay, not only death but indignity, the exposure of the souls of those who survived. Gettysburg is a very level playing field.

Plying his trade of undertaker, Jupiter Jones, an elderly black man who had graduated from being a snake oil salesman to a salesman of another sort- making the dead look decent. Jupiter is perhaps the most interesting character in the book. Wearing an old green bowler atop his head and always accompanied by a small monkey named Archimedes, Jupiter would set about transforming the cadaver beneath his hands into something that almost -at least Jupiter thought so-resembled life. Pumping some sort of arsenic mixture into the body, Jupiter would affectionately pinch the corpse's cheeks like one would pinch a child, tuck a wad of chewing tobacco inside if the dead soldier's yellow-stained hands revealed tobacco use.

Snake oil, a concoction garnered long ago from trees in Africa was applied to the open eyes, plumping them out. Jupiter waxed philosophical, talking to the doctors laboring beside him, talking to the bodies. "The dead talk to me" he remarks even though "nobody gets out of life alive." Although the doctors work with the living, and Jupiter with the dead, somehow the macabre rituals are the same.

The surgeons, laboring under unspeakable conditions, using the altar of an old church as an operating table, can only push their weary bodies so far. Author- surgeon McDaniels does not spare the reader. Throughout the novel the horrendous wounds and their infinite variety and the procedures of the embattled surgeons are described in great detail. Perhaps the most interesting surgical operation concerns the trepanning of a young soldier who had been shot in the head. Surgeon Hardy had made the appalling discovery that the barely- alive boy was his own son but Hardy could not force himself to operate, so Doctor Boyd, although addicted to laudanum, performs the very tricky drilling into the boy's skull. What makes the medical narrative so authoritative is, of course, due to the author's expertise. But the information has to be honed to fit the appalling conditions, where instruments were never sterilized, where doctors could not wash their hands, where the floor of the operating theater was slimey with mud and gore. Doctor McDaniels will take you there.

On the battle field two soldiers fall together side by side, one a Yankee, the other a Rebel. The two former enemies are bonded together in misery and indeed they do bond in extremis, giving each other comfort by reciting the Beatitudes. The Yankee boy, Ezra Coffin, manages after great effort as each physical move is agony to get the Rebel's gun, thinking that he can kill them both to put them out of their misery. But there is only one ball left in the gun's chamber...

What goes around comes around, even in war, even in tragedy. Good springs from evil and author McDaniels will bring his novel to a satisfying close. Perhaps, when you finish the book you will find yourself thinking of Gettysburg not so much as a shrine but a place where humans suffered horribly and where the healed America rose- I will say it- like a phoenix from the ashes.

4.0 out of 5 Stars
IndieReader Review Feb 22, 2013
Verdict: *A vivid, engrossing story of one battle, told from the*

perspective of the soldiers that fought it

In "Not One Among Them Whole," author Edison McDaniels, takes us by the hand and leads us through the horrors of battle as witnessed by a handful of individuals. It serves as a timely reminder for the video game generation that might see war as a glorious endeavor from which heroes emerge triumphant and whole. There are no heroes in McDaniels' book, only people coping as best they can with desperate situations. There's no black and white, only a grimy grey through which the novel's characters grope for relief from the relentless horror that envelops them.

The story unfolds during and immediately after the Battle of Gettysburg, which lasted for three long days. The statistics are grim: 8,000 died on the battlefield with another 27,000 wounded. McDaniels set himself a difficult task in attempting to humanize those numbers; he has risen to the occasion. The characters in the novel are individuals with their own stories, brought together through the vagaries of a war that, in its time, was the most destructive the world had seen.

Two soldiers—one from each army—introduce us to the battle and it's horrific aftermath. McDaniels describes, in sometimes gruesome detail, just what it might be like to lie in a field full of dead, wounded, and dying men—not to mention the scavengers (both animal and human). Even the weather, a supposedly disinterested element, seems to conspire against the soldiers and add to their misery.

The book, though, really revolves around those not intimately

involved in the fighting, especially the surgeons who do their best to save the wounded. Those wounds are described in clinical detail. And the descriptions of surgery carried out in primitive conditions, before germ theory and universal hand washing, are equally detailed and not for the squeamish. (The surprise is not that so many died, but that anyone survived.) These surgeons are also wounded, by their past and present lives, and their individual tales are woven through and around the story of the two wounded soldiers.

This relentless misery, one of the book's most salient characteristics, makes it difficult reading at times. Some relief is provided by snake oil salesman Jupiter Jones, who has gathered a crew of misfits to help sell his miracle cure: Jupiter's Oil. But even that is short lived and the story soon returns to the battlefield.

Despite the death and decay that permeate the novel, it is a compelling read, largely because of the skill with which McDaniels unfolds his characters' stories, day by day, minute by minute. Equally skillful is the manner in which he brings together all the characters and crafts dramas within dramas against the backdrop of the American Civil War and this one important battle. So compelling are those stories, the war fades into the background as the fate of individuals hang in the balance.

Reviewed by Brid Nowlan for IndieReader