

Transcript of Hydrocephalus After Hemispherectomy And Other Resective Procedures

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Everyone, it is truly an honor to be here and have a chance to meet so many wonderful families and I am really thankful and appreciative to have the opportunity to speak to you. Actually, what a really, really important topic. Hydrocephalus following hemispherectomy as in any really any generally large epilepsy resection. No conflicts of interest.

So we're going to start off talking about basic anatomy and physiology, about the brain and how brain and spinal fluid physiology works in the brain. We're going to talk about something called the Monroe-Kellie Hypothesis which is an important paradigm from which we make decisions and how we think about pressure in the brain and hydrocephalus. We are going to talk about diagnosing hydrocephalus, management options, and complications and troubleshooting particularly shunt problems.

So here is the anatomy of the ventricular system: The fluid is made in the center of the brain in these pockets known as ventricles and there's a particular substance here called the choroid plexus and the Choroid Plexus' job is to produce brain and spinal fluid continuously at a rate of about every three minutes or so you may get one milliliter of brain fluid so it's very, very slow. This gets created in the center of the brain and it kind of follows this path from these ventricles down to this other ventricle, to another one and then it goes around the spinal cord and the brain and bathes the brain.

For each drop of brain fluid which we call CSF – CSF stands for cerebrospinal fluid – for each drop that is made, the same amount should be absorbed. Hydrocephalus is the imbalance of what's made and what's absorbed; so if you have too much brain fluid made and not enough absorbed you develop hydrocephalus. There are very few conditions that give you too much brain fluid being created but any of those can give you hydrocephalus. Hydrocephalus stands for too much fluid in the brain, too much brain fluid, too much CSF. These are the rates at which CSF is made in adults and kids.

So here's a closer look at the fluid spaces of the brain, the ventricles, and you can see this red structure, that is the choroid plexus and that's what creates most of the brain fluid – it does not create all of it but that's where most of it comes from and this will be relevant later on as I talk about one of the treatment options for hydrocephalus. And where is the fluid absorbed? It is actually on the surface of the brain and these areas called arachnoid villi and this is the primary absorption site. In hemispherectomies in general the type of hydrocephalus you develop is likely related to scarring of these arachnoid villi, so scarring and preventing the absorption of brain fluid. That generally tends to be a problem.

So, a little bit about CSF physiology: Alexander Monroe was a Scottish anatomist and he and his student George Kellie defined the Monroe-Kellie Doctrine and there are several principles. The first is that the skull is a closed box and it's generally inelastic. I say 'generally elastic' because in little babies their skulls are actually quite soft and they actually have the room to expand. But once that soft spot closes up the skull really is a closed box and when you think about it if you have a swelling in the brain or if you have any tumor or fluid buildup or infection or anything that takes up volume, initially the brain can compensate. It compensates by draining more brain fluid; it can sometimes decrease the amount of blood volume in the head but eventually it will amount to pressure on the

brain pressure on the brain and pressure on the brain is not good. So that's really in a nutshell what the Monroe-Kellie Doctrine is about.

This is a nice curve that shows that as you increase the volume of contents in the brain, the brain can compensate initially and it does so by draining more brain fluid, by decreasing the amount of blood in the brain. The blood I am talking about is the blood in the arteries and veins but eventually it gets to the point of decompensation where the brain can no longer compensate and then small increases in volume can lead to large increases in pressure in the head and that's generally when children or adults, people with hydrocephalus, become symptomatic.

Here's a picture of what it looks like: This is what normal ventricles look like and this is what it looks like when you have hydrocephalus. You see the fluid buildup inside the brain; you see this area in red or really the swelling and pressure that's on the brain.

So again, what is hydrocephalus? It is the imbalance between the fluid production and absorption of CSF and it can result in increased intracranial pressure.

Now, very important to know that increases in pressure may or may not be associated with a change in the ventricular size. Most of the time there is a change in the ventricular size except for children that have had multiple operations especially if you have had infections. The reason why is the Ependyma, the surface of the ventricular system scars in and once it scars in it has limited compliance so even though the fluid spaces may appear normal, it doesn't necessarily mean that there may not be a pressure problem – so that's important to keep in mind. Now, this gets a lot more attention than it should, it is actually not that frequent but it does happen, it is very important to know, especially in children that have had multiple shunt operations.

So a few facts about hydrocephalus: It is most common thing that a pediatric neurosurgeon may treat. It affects about one or two out of every thousand babies. The type of hydrocephalus that we are talking about is generally post-epilepsy surgery so it's slightly different. There is no cure for it. There is no effective medical therapy. The only effective treatments are surgical and it is very important to know that hydrocephalus is also a lifelong condition; it's not the type of thing you get treated once and say, 'I am in the clear and I don't have to think about this diagnosis again'. Especially as you think about it, when a child gets treated when they're very young we need to follow up with them as they grow up to get into adulthood because you can face different challenges and sometimes you need revision operations for your hydrocephalus.

So in a nutshell, I touched upon this, hydrocephalus is either increased brain fluid production, obstruction of those fluid pathways can give you hydrocephalus or lastly due to impaired absorption.

I'm not going to go into the surgical techniques much but the anatomic hemispherectomy where an entire half of the brain is resected – this operation is associated with the greatest amount of blood loss. It actually has the highest risk of hydrocephalus than other operations that involve removing less brain but it is the most certain procedure to ensure disconnection and it's generally indicated for situations nowadays when a functional hemispherectomy has not worked.

This is what can happen after an anatomic hemispherectomy: As you can see the fluid spaces of the brain are quite large and you see that white signal in that image on the left side around the fluid spaces, that's a sign that pressure is building and now that fluid is sort of defusing across the membranes into the brain. Children like this

can present in two ways. You may catch this in a child that is asymptomatic, who has no symptoms and you may just find this on imaging, you may find it in a child that has headaches, nausea and vomiting and in fact some children present with recurrence of their seizures because of hydrocephalus. So, especially if you've gone many, many years without seizures and all of a sudden you're developing seizures, one of the things to look at is does your child have hydrocephalus because this can trigger seizures as well.

So in general when we talk about epilepsy surgery the problem of hydrocephalus is due to impaired absorption and the reason why is blood products go in and can scar those Arachnoid Villi that are responsible for absorption of brain fluid.

So we have evolved in our hemispherectomy techniques; we have several different techniques. The functional hemispherectomy, the vertical hemispherectomy, peri-insular, transylvian, nowadays there's a endoscopic method of a hemispherectomy as well where you just go in with a very small camera and a lot of these techniques have generally evolved because we want to do a smaller operation, a smaller craniotomy, hopefully reduce some of the operating room time, decrease blood loss, perhaps decrease infection. If you do a shorter operation there are many studies that show they have a lower chance of developing infection. More importantly, decreasing the risk of hydrocephalus.

So these are my pearls for decreasing the probability of developing hydrocephalus. Some neurosurgeons may not agree but in general disconnected hemispherectomies have a lower chance of developing hydrocephalus as opposed to anatomic hemispherectomies. You want to decrease the blood loss as much as you can during surgery and that's very important for these operations. Having a surgeon that works with or is a pediatric trained neurosurgeon because pediatric neurosurgeons are very attuned to blood loss. Kids, especially babies, don't have a lot of blood volume so a little bit of blood loss can actually be quite a bit. So it's very important to be very meticulous when you're doing this operation. You want to limit the amount of blood that actually pools in the fluid spaces of the brain because that can lead to hydrocephalus; you want to aggressively drain brain fluid after surgery. So after a hemispherectomy generally a drain is left and that's to get rid of all that debris, all those blood products out of the head. It has several benefits: It helps with post-operative fevers but it also very importantly, when you clear up that fluid (we don't have great studies to show this) but we do think that that decreases the probability of developing hydrocephalus.

There is one last thing I left on here which I'll talk about very briefly at the end but the Choroid Plexus is where that brain fluid gets made and one of the things that I have been doing is cauterizing or burning some of that tissue that creates brain fluid during the operation and we have a wonderful opportunity here as we do the surgery to have access to that Choroid Plexus and that can really decrease the amount of brain fluid being made and hopefully that the brain can absorb what is truly being made. In my opinion, it's really decreased the amount of hydrocephalus after surgery but the final results aren't out yet and we need to have more experience to be able to say this definitively.

Detecting hydrocephalus: The babies have a window, the baby's soft spot here and one of the signs of too much fluid in the brain is a bulging, intense soft spot. The soft spot is called the fontanelle. So every time you see your pediatric neurosurgeon when your baby is very little they're going to feel the top of your child's head and that can give us an indication if there's pressure or not. In really young babies, we follow them on this head circumference chart and you can see that this child is crossing percentiles or from the third percentile to well over the 99 percentile, this is a sign of pressure building up in the brain. It doesn't have to be fluid, it doesn't have to be

hydrocephalus, anything that takes up volume can do this. A tumor can do this, infection, pus; any of those things can do this so that is what we look at.

So symptoms and signs of hydrocephalus in infants: Abnormal increases in the head size, drowsiness, irritability, poor feeding or vomiting. Really, really young babies have breath holding spells and their heart rate changes. Actually heart rate changes can happen even in older children. But those are typically the way hydrocephalus manifests. In older children they generally complain of headaches, they may have learning problems, again nausea, vomiting, drowsiness, difficulty with balance, difficulty with coordination, swelling of the optic nerve. So oftentimes your neurosurgeon may send your child to an ophthalmologist to look at the swelling at the back of the eye because that gives us a window into the brain to tell us if there is pressure in the brain or not. And sometimes an inability to look upwards.

So these are what the scans look like. Typically when you have hydrocephalus this is what a CT scan looks like and you can see there is fluid buildup in the brain. One of the two options - and we're going to talk very briefly now about shunts - is where you put this drain. You can put it inside the fluid space and this is typically where shunts go in the ventricular system (called a ventricular shunt) or your surgeon may choose to put it in the cavity.

This is an example of a post stroke hemispherectomy. So a lot of this tissue was missing because of the stroke. So again, different neurosurgeons have different opinions on it, I personally prefer the ventricular shunts because shunts are made to be in the ventricular system, they have support there and typically it's hard to leave a shunt in the cavity without causing it to either lean against one side of the brain or not but this is this is highly controversial. Some surgeons may prefer to just put the shunt on the cavity side just because if there's any bleeding or complications relating to putting the shunt in that side of the brain is already disconnected. So that may be a reason to put it in the cavity but in my experience ventricular shunts tend to work longer.

So what is the natural history of untreated hydrocephalus? Untreated hydrocephalus can actually become quite severe and about half the children, if it is not treated, don't survive. The other half, if it is not treated, they get to a stage called arrested hydrocephalus. This doesn't necessarily mean it's a good thing, that means the fluid spaces aren't getting any bigger but it usually comes at an intellectual price and only about 15% of these kids may have a normal cognitive and neurological findings. This is all-comers with hydrocephalus not particularly about the epilepsy population.

What are shunts? This is technology that has been developed in the 1950's and it's largely unchanged since then. Many, many efforts to try to design better shunts and more effective shunts and shunts that don't block but as it stands today unfortunately there is not a medical device out there that fails as often as a VP shunt. So we have a love/hate relationship with shunts. We love them because they can work; we hate them because they can fail. Many, many new versions have come out with very little improvement in outcomes.

So a shunt typically has three components. There is a ventricular catheter. This goes in the ventricle or in the brain in the fluids spaces, there's a valve that kind of dictates when the shunt drains or not, and there's a distal catheter that generally tunnels down under the skin, generally into the belly but there's other sites to put a shunt as well. So these are the three main components.

This is what it looks like when it's implanted. The most common place is in the abdomen, it is called a ventriculoperitoneal shunt. Other common options are in the chest that's called it a ventriculopleural shunt. This

is generally not a good option for kids under the age of four because their chest isn't large enough to absorb brain fluid but after the age of four this becomes a viable good second option. The other option is putting it in the heart – the ventriculoatrial shunt. This may be a good option if child is very young and the ventriculoperitoneal shunt is not an option but the problem with putting it in the heart is that as the baby grows that the position of that shunt moves and you may need revision operations just to keep it in the same location.

There are many types of valves: There are ones that work based on differences in pressure between the head and the belly, there's ones that work based on very constant flow rate and regardless of pressure they drain the same amount, there's programmable shunts, there's anti-siphon devices, there are more shunt valves than you want to know about, but the bottom line is there is no one shunt valve that's better than the rest. We've done a shunt design trial, one of the important studies that came out in pediatric neurosurgery, where we can look at all of these four different types of shunts and over time they all pretty much fail at the same rate. So there's not a single one. So what happens in a lot of times it's based on the preference of your neurosurgeon. There's certain instances where you may choose one valve or another but none of this is evidence based and generally in medicine when there's a lack of medical data anyone is an expert because who's to say you're wrong with whatever you say? So that's important to keep in mind.

Shunt complications: Three main types. There's mechanical failure – shunts are mainly a piece of equipment; they can block, they can break, they can kink, they can move, they can obstruct. Any of those things can happen so that's why it's very important to have long term follow up once you have a shunt.

Infection used to be a larger problem in the past; the infection rates are a bit lower. There are hydrocephalus guidelines nowadays that most hospitals are following in terms of the procedure in which they put in the shunt and the infection rates have gone from about 10-15% to closer to 5%.

Functional failure: That means that these are shunts that are working but maybe perhaps not draining enough or draining too much. Three origins of shunt problems: It could be the shunt, it could be the patient, it could be the surgeon.

Complications in the early period: Hematoma. This is a blood clot. These are low risks but as you are penetrating the brain with a catheter the potential for having bleeding. As you are passing a catheter into the belly there is a potential for bowel injury, thoracic injuries – injuries to the chest, the organs of the chest as you pass the shunt down. Infection when it occurs, most commonly it's upfront. Usually in the first three months, if not in the first six months, after six months it's pretty rare to have a shunt infection - it's really, really rare. So it's not impossible but that's usually the time period when a shunt infection happens.

The most common reason for a failed shunt is a proximal shunt obstruction. One of the most common reasons for proximal shunt obstructions is over shunting and you get the ventricle to completely collapse around the catheter, it blocks off the catheter and the brain and choroid plexus can get stuck into the holes of the catheter and that can be a very common reason for shunt obstruction.

So what do you do when you suspect a shunt problem? Generally you're coming to the emergency room. There are two sets of imaging that's always necessary to be able to fully diagnose a shunt problem: One is some sort of brain imaging, so either a CT scan if your child is really sick, most hospitals now can do fast MRI scans which

limits the amount of radiation and it's almost as quick as a CT scan. But you need brain imaging and you need shunt x-rays because you need to also make sure that the shunt is in continuity. That's where you usually start.

If you are suspecting infection look at fever, abdominal pain, there are blood markers that you can look at and depending on the problem you may need to get abdominal x-rays or abdominal ultrasound, chest x-rays if you have a ventriculopleural shunt. Sometimes you need to tap the shunt to take some fluid off to make sure it's working or send it off for testing. There are nuclear studies and sometimes ICP monitors. There are still a certain proportion of children that no matter what you do you may have to end up going to the operating room to explore their shunt and now I will briefly talk about that.

So we have three types of shunt patient. It is very difficult in a venue like this to talk about every scenario, but these are the three common types of shunt patients. One is a child, they come in, the fluid spaces look normal and they are clearly sick by something else. They have gastritis, they ate something, they have diarrhea because they're sick for a different reason and just because they have a shunt the emergency room physician may consult a neurosurgeon but it is very clear that there is something else going but it's not the shunt. Those are easy.

Also easy are the kids that have really large fluid spaces in the brain and they're very sick - horrible headaches, vomiting nonstop and a scan that looks really bad. So those are the kids that are also - you know there is a shunt problem and we need to fix it now.

Unfortunately nature has it that that the most common situation is a child that is somewhere in between, where the imaging doesn't look any different, the child may have headaches. You can get headaches for many different reasons, there are probably a thousand different causes of headaches, and the you're in the situation where you don't know it's the shunt or not and that's OK and that's a very common situation to be in and sometimes it takes time to diagnose it. Sometimes it is not easy to say right away in the emergency room that you have a shunt problem or not. Sometimes you need to admit the child, watch them, see what happens with their symptoms or headaches, if they are vomiting or so on, make sure it stops, and every once in a while like I said you still need to take a child to the operating rooms explore the shunt and make sure that both sides of the shunt are working – the proximal end, the distal end, and explore it that way.

There is a really neat study that was done out of Birmingham, Alabama, where they looked at 205 consecutive children who presented to the hospital with possible shunt problems and they commonly asked the families, 'do you think it's the shunt or not'? So families, I must say, they were really, really good at saying that it wasn't the shunt. So when families thought, 'no, this doesn't sound like it's the shunt', they had about a 90% accuracy. But when it came to them being accurate with thinking it is the shunt, slightly less than 50%. 40% of the time they were correct. The other thing that they found is that the more experienced parents, the more you have gone through this, the more accurately you were at saying whether it was or wasn't the shunt, which makes sense.

There is The Shunt Book believe it or not, so for all your difficult shunt questions these are the two people to ask. One of them I trained under, Dr Drake in Toronto and Christian Sainte-Rose but there are many, many different situations and it's very difficult in a lecture like this to go over everything I know about shunts.

There is a really neat study that was done by a post-hemispherectomy hydrocephalus work group and they looked at hemispherectomies in particular and they found that 23% of patients who underwent hemispherectomy between

1986 and 2011 developed hydrocephalus. In my experience it's been even less so I don't know if that's just more recent data, but that seems like a good number to quote.

And then timing range: So, half the time you figure out your child has hydrocephalus in hospital right after surgery. They have the drain that is draining the extra fluid and they realize, no we can't remove this drain because the child needs the drainage and that way you know this child needs treatment for hydrocephalus. But half the time it could happen years later and in this study it was 8½ years later was the longest; I've seen a child that was 10 years later, so it can happen down the road as well. Obviously the further out you go the less likely that you're going to require a shunt.

This is really small, but this tells you the most common symptoms with which a child after hemispherectomy may present, requiring a shunt. The most common is failure to remove that drain after surgery. That's about 61% of the time. There is imaging changes about 80% of the time. Headaches, vomiting. These are the common causes. The bottom line is kids can present differently with their need to have a shunt.

The hemispherectomy technique was very important. With anatomic hemispherectomy the hydrocephalus rate is 30%, with the functional hemispherectomies it is closer to 20%. There are some other risk factors but none of these studies are perfect. One thing to know about medical studies is that they all have limitations and just because they have found certain factors, these factors need to be restudied and the more times we can confirm the same findings the more reliable these findings are. Just because they haven't looked at other factors that does not mean there are not other factors that may related to the development of hydrocephalus.

Very quickly I want to talk about another type of treatment for hydrocephalus. This is particularly in the event that you have an obstruction in the fluid pathways of the brain. Nowadays we can go in with a very tiny camera right in and create an internal connection in the brain that will allow the fluid to drain and the nice thing whenever you can do this procedure is that it is shuntless, there is no hardware that needs to be left behind. Generally hydrocephalus after hemispherectomy is treated with shunts but hydrocephalus and shunts should not be synonymous. What we try to teach our residents is not to, as soon as you hear the word hydrocephalus, associate it with a shunt. There are different ways to treat it.

This is a procedure that we are doing at UCLA and certain other places around the country. This is the Choroid Plexus, this is what it looks like during surgery and we can go in and we can cauterize or burn some of that tissue that is creating brain fluid and this is what we're doing. So while you are doing a hemispherectomy you have a wonderful opportunity as are doing your disconnection and you have access to the Choroid Plexus. So one of the things that is very simple to do is to cauterize some of that Choroid Plexus and that helps decrease the amount of brain fluid production. Like I said before hydrocephalus is all about the balance of how much fluid is made and how much is absorbed and it you can tip the balance in a way that you know the less fluid that's being made can be absorbed perhaps you can get with away with avoiding hydrocephalus.

So, prevention of hydrocephalus although it's not always possible in my mind is best for a strategy. Hydrocephalus actually seems like a simple problem but it is actually deceptively very complex. The other thing to keep in mind is although shunt failures tend to present similarly in children and for those families that have kids that have had multiple operations generally you can tell this is the shunt or if it's not the shunt, but it's important to keep in mind that it that doesn't always have to present the same way, it can present differently, and that's part of the challenge.

The last thing is hydrocephalus is a long term lifelong condition and you need lifelong follow up. So even for that treatment where we talked about potentially doing a procedure with a camera and not leaving any hardware behind that doesn't mean your child is cured. They may develop hydrocephalus after that so that's very important to keep in mind.

Thank you.

[Applause]

Questions from the audience:

Question: I just want to ask about possibility of intermittent hydro? So my daughter suddenly had increased seizures, she vomiting, she was nauseous so we took her to the hospital, they did an MRI, and it looked stable. They even put an intracranial pressure monitor on her and by that time her symptoms has subsided and it was normal and this was just 3 weeks ago and she's had 3 normal weeks but she had this sudden acute change of nausea, vomiting, dizziness, seizures. Could intermittent hydro cause that?

Dr. Fallah: Yes. So, intermittent hydro is the most challenging to diagnose because like you said, the imaging is pretty much always unchanged, usually by the time the neurosurgeon walks into the room the symptoms have usually resolved. You say, 'I swear he was vomiting all this time, but now he's stopped'! There is no perfect test. You can do nuclear studies where you put a nuclear dye in and watch it go down the shunt. That's great, it can show that the shunt is patent but that doesn't mean it's always patent you're just getting one study at one time. You can get a CT scan or an MRI show the fluid spaces are the same, again that just gives you one snap shot in time. You can even tap the shunt and it can be working at the time but it doesn't mean that it's always working. ICP monitors are good because they can give you a reading over a long period of time but the problem is when you put an ICP monitor on and the child's restricted to the bed and it is not what they do on a normal day to day basis it's a very artificial environment to keep the child in. So, unfortunately there is no good test and those are the kids that go in and out of the hospital but at some point we may have to go to surgery, explore the shunt and sometimes just replacing the shunt.

She doesn't have a shunt?

Question/Comment: Almost 4 years after surgery she suddenly had an acute change in her seizure activity about a month ago, I am given her all this emergency meds I have never had to do, she is vomiting, nauseous, she is going unconscious, all these things. I admit her to the hospital, she does that the first 3 or 4 days by the 6th day they put the ICP monitor on her head, she seems to be coming out of whatever she was in. The ICP monitor is fine. Now she's been good for 3 weeks....

Dr. Fallah: The way we think about developing hydro before you have a shunt...developing hydro is generally not the type of thing that waxes and wanes, there is usually a linear progression forward. When you think about it, if your pressure in the head just builds up it doesn't intermittently go away. I haven't heard of a case, it doesn't mean it doesn't exist but I haven't heard of a case like that but again, you can get other pieces of information. Ophthalmology can look into the eye; make sure there is no papilledema or pressure in the eyes and sometimes just monitoring it over time to see if it's something that will go away.

There are ways to also temporarily divert the fluid away without committing to a shunt. You can put a drain; you can put a lumbar drain to transiently divert the fluid away to see if those symptoms get better. But again, it's difficult when the symptoms come on or off because when you do it she may not have any.

Question: You know, the problem with repeated CT scans is the exposure to radiation, I know we do fast MRIs now, I just want you to discuss that.

Dr. Fallah: Radiation has harmful effects to the developing, growing brain of a child especially the younger they are they're more at risk. So kids, really under the age of 3, even the next milestone, kids under 10, you really want to limit them and especially with shunt problems because some of these children come in and out of the emergency room a lot and it is very prudent for us to try out best to limit CTs. Most pediatric hospitals have special protocols where the amount of radiation is actually pretty low. So our Neuroradiologist tells us that our pediatric CT scan gives us the same amount of radiation as a shunt series or shunt x-ray. So my general advice is as much as we can, is to try to do the fast MRIs and most hospitals are able to do this. So this is a very quick MRI, doesn't require any radiation, takes about 20 seconds to do. So provided the technologist feels comfortable. There is a certain age group that very tricky – toddlers, it is hard to keep them in any one spot for too long. So generally fast MRIs whenever we can but if your child is sick and if they're are vomiting and we need to make a decision right away a CT scan is still a good option so I would not hold off getting a CT scan if your child is actually really, really sick and we need to know an answer right away because that still seems to be in urgent situations, the best the best way forward.

Question: What is the max life of a VP shunt?

Dr. Fallah: With shunts there is no max. I say a shunt can last 2 hours or it can last a lifetime. Neurosurgeons in previous generations to me they would say that their first shunt case in the morning would also be the last one of the day where they had to revise the original shunt from the beginning so there is a history of shunts not working very long but there are certain kids that have had one operation that's been it and there's no life time to a shunt and we would never revise it if the child is doing well.

For more information on hydrocephalus after hemispherectomy and other resection procedures, go to www.brainrecoveryproject.org