

**RAMIN**

Endoscopic Third Ventriculostomy and sometimes associated with that is Choroid Plexus

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Cauterization, I know it's a mouthful. So we tend to shorten it to ETVCP. It is a method of treating Hydrocephalus that does not require placement of a ventriculoperitoneal shunt, which is the traditional method of treating Hydrocephalus.

The benefits of that procedure over the traditional shunt are number one, the need for having hardware kept in the body over a long period of time. So a shunt requires you to have that shunt hardware, which involves a tube in the brain, connection valve, and then a tube that goes from the head all the way down to the belly or other areas of the body, to try and drain the fluid. That typically stays in for life. Anytime you have hardware like that you risk infection. So this procedure obviates the need for any of those hardware implantations. That's a big, big part of it.

The other benefit overall is that if it works, if you can create a bypass route for that baby or that child's brain to absorb their own fluid, over time they create their own equilibrium.

Whereas with a shunt, it can malfunction. And if it malfunctions, because the patient has now become dependent on that tubular system, even slight changes in pressure are not really tolerated well. And so the need for tubing and the risk of infection, as well as not having a potential for a malfunction, are two main reasons that it's beneficial.

The procedure of the ETVCP has actually been around for a long, long time. The old method of doing the procedure used to be done in an open manner. With surgeons actually going in physically with instruments through the brain with a microscope, or not, and removing the choroid plexus sometimes, to help prevent CSF production. Or making the hole in the bottom of the third ventricle, and performing a third ventriculostomy in an open manner. So the procedures themselves have been around as long as neurosurgery has really been around.

However, the ability to do this through the endoscope really changed that procedure in a good way. Because it allows you to do it through a very small hole. And so instead of having to do a big open brain surgery, you can do it through a camera, through a small hole. The next generation of the way that it came about actually should be credited to Dr. Ben Warf, who is a pediatric neurosurgeon working out of Boston Children's Hospital now. He basically went to an area of Uganda where sub-Saharan Africa has a very high population of Hydrocephalus. And usually it's from infection reasons, post-meningitis, post-ventriculitis infections. And he saw that

in those cases where he has to implant shunts, those kids are basically going off to their villages. If they have an infection or malfunction they're probably not going to survive.

So he looked at doing a procedure that could potentially bypass the need for shunting in a lot of the patients. And because the population was so high he was able to do a lot of those procedures and had great follow up, and recorded all his findings. And the data that came out of there really allowed for a relook at this procedure for kids that are younger. And in the past we tended to not do this for kids younger than three years, definitely not younger than one year. And what Dr. Warf showed, and since then other people have replicated this, is that you can get away with doing this on young, young kids, even newborns. But the more we look into it, the more we've researched it, there are certain patient populations that are better for having this done. And there's those that tend to not tolerate it as well, they just don't have the absorption capacity to do it.

So the procedure has evolved to becoming more and more minimally invasive. But at the same time, the technology that allowed that to happen is what made it possible to do in the developing world. At the very beginning when this was started in Uganda, nobody really learned how to do it, other than the people that were in Uganda. And one of Dr. Warf's, actually two of the surgeons that he trained in Uganda to help to continue this legacy after he left, started training other surgeons in Africa. Because obviously this is one central location, very difficult to get to when you're far away. We had patients come from Sudan. And it took them a week travel just to get there. But they felt like this is something that they need to train others.

So they started training other neurosurgeons, even other general surgeons to do this. But really when the rest of the developed world started to become interested, the first criteria was you have to be trained by someone who has been trained there. And so a lot of people went directly to Uganda to train. So most people, young surgeons most of the time, would go to Uganda for about two to three weeks, and do as many cases as they could during that time. A lot of it is technical training. The procedure itself is not necessarily a difficult one. But the instrumentation is different than what we're used to. The scope is a flexible scope that requires manual positioning changes. And all these things take a little bit of time to learn. But once they got a few cases under their belt in Uganda, they came back and they continued to do them. They were sort of allowed to do them.

Now that there's been a generation of folks that have already been trained, those people are

now training the next generation. So now really it's part of the neurosurgical armamentarium. Most of the time you learn the most in your pediatric fellowship. You go to a place that has someone that was trained. And they teach you how to do that throughout your training. I was one of the first residents to be able to go out there. And I spent about two months there. And so I was able to do a lot of those cases with the surgeons out there. And was able to learn it. And then when I went to fellowship, I sort of took that training to my fellowship. And I taught the surgeons at my fellowship location at Stanford. And so now they're doing it over there, as well. And they're teaching the next generation through the residency program. And so since I've been here I've been teaching our residents how to do it as well.

So the procedure itself usually takes about 90 minutes, 60 minutes to 90 minutes. So it's not a very long procedure. There's minimal head shave. The incision is about a centimeter and a half by a centimeter and a half. It's almost always a little boomerang incision. Because you use the same incision for putting a shunt in. So it just sets it up in case you ever need to put a shunt. It's in the right frontal region. You go down and you can access the brain through either a drill if the skull is solid, or through the edge of the fontanel, the soft spot, if the skull is soft. And you enter through with the endoscope, which is a flexible instrument that has a tip that can move around.

Once you're in the ventricle everything else is through that scope. Through the scope you can walk your way into the third ventricle, which is a fluid cavity that separates the top of the brain from the brain stem region. And typically speaking on these patients, the floor, or the very bottom of the third ventricle, which is where you want to puncture, is bowed down from the pressure building up, like a water balloon blowing up in the brain. And it thins it out so you can puncture through it very easily. There are instruments that are made to go through the endoscope, that help facilitate the opening of that floor. Once you're done with that, you exit out of the third ventricle. And you come back into the big fluid space of the lateral ventricle. That's where the choroid plexus lives. And you go around and using a monopole or a cautery device within the ventricle, you coagulate the choroid plexus to make it shrink up and sort of die off.

Now this is a temporary thing, because the body is really good at healing. And it knows and it wants that to come back. So typically speaking this decreases production of CSF for about six months. After six months the body tends to restart the same production of CSF, even though that choroid plexus has been coagulated. So what you're really doing is you're doing sort of a

belt and suspenders. You're making the hole for the bypass of the fluid to be absorbed differently. You're also decreasing overall production in the beginning to give them a new equilibrium.

So the procedure did really take off in Uganda and became a mainstay of the method that they treated Hydrocephalus with. Since Dr. Warf kind of left Uganda and came back and took a position at Boston Children's, he started doing the same procedure on the patients that we had here in the United States. And found that there is very similar success rates in certain patient populations. And because we're a little more sophisticated with our imaging, and our follow up techniques, and an overall psychological testing and everything, we've now started to study this much more in-depth through a network called the Hydrocephalus Clinical Research Network, the HCRN.

And that's a multi-center network that we hope to one day be a part of once we're a little bit bigger. And it studies individual questions within the field of Hydrocephalus in pediatrics. Now it's actually doing the same thing in adults, which is great. But it takes a specific question and it throws it within the whole network. And so suddenly instead of one center doing a project, you've now got 14 centers doing the same project. Your numbers increase dramatically. So the confidence with which you can come up with data and results exponentially increase, and are much faster. So since this procedure has been performed in the US, multiple centers have now become very interested. And so the HCRN basically took it over and said, all right, we're going to start studying this in a much more rigorous manner. So that's where a lot of these outcome data are coming out as saying, all right, this patient population is probably not good to offer it at all, this patient population's on the fence, this one is a great one you should offer it in all these.

The patients that tend to do the best are definitely the ones that are a little bit older. So there's been age data coming out at newborn, three months, six months, nine months, 12 months. If they're older than a year they tend to do very well. And the procedure has probably about a 75% success rate. And the kids that don't have old infections, or old hemorrhages, it tends to have a better success rate. In kids who have obstruction as an anatomical feature that prevents the fluid from flowing in the right direction, those kids tend to have a very good success rate, upwards of 80%. The kids in which it doesn't do as well are the kids that are born premature. Because we think that most likely their absorption pathways haven't fully developed. And because they're born premature, they don't ever completely fully develop, or

develop much later. So if they have Hydrocephalus as an early infant, by the time they develop a good enough absorption capacity they probably have needed something done already. And those are the kids that we tend to put shunts in earlier.

But a lot of the work that's been done on these kids does allow you to do this procedure even before doing a shunt. So the good thing is, these kids have an open skull and so pressure builds up slowly. And unlike the kids whose skull is completely closed, the brain has some capacity to mitigate that pressure through the skull. Because of that you can offer the procedure in the right patient population. And we tend to at least if they're full gestational age or at least three months of age even with hemorrhages, sometimes we'll still talk to the family about offering it as a procedure to do before doing a full shunt.

Unfortunately if you do a shunt first usually the fluid space is collapsed down around the shunt. And you don't have the opportunity to do the endoscopic procedure from a safety standpoint. You need the big ventricles as a pathway to be able to go in and do the procedure safely.

I think what I'd like to see happen is I'd like to see us continue on this evidence based pathway. The HCRN is doing great work right now. They have a head to head follow up study on shunted kids versus those undergoing endoscopic surgery. And they're going to look at their neuropsychological outcome at a later date, which is a great way of looking at it. Because to be honest, survival is not what we're after. If a mom and dad come to me, they want their kid to be in a normal school. They want them to be able to play sports. They want them to do everything that other kids are doing. And so if we can show that the outcome of this surgery at a later date 5 years, 10 years, college years, is the same, or otherwise equivalent to shunting, that really supports continuing to do it on this patient population. But if it comes out and says, you know what, in these patients even though it seemed to have worked, by the time they're 5 or 10 years old they have a lower neuropsychological outcome. Then we might go back and say, hey, maybe we need to revisit this. Do we need to do something else? Do we need to alter our methods?

So I would love to see us continue down this evidence based path. Because it really is the best care we can provide our patients. Instead of one person thinking one way is the right way and another person thinking a different way. Having the HCRN do these huge studies with numbers that are significantly better than in the past, and be able to give us good data is really the best way to go.

I think from a physician standpoint, any time we do surgery on these kids with Hydrocephalus and we put shunts in, this is not just our problem. It's not only the worry and the burden of the family, especially the child as they grow older, but it's also the burden on the pediatricians and their overall pediatric health. Any time they have a headache or start vomiting, or anything like that, especially if they're young and can't communicate yet, those are ER visits, and pediatrician calls, and mom worry, and any of those things could be a shunt malfunction potentially. So I think having something like this really lessens the burden overall on the healthcare system.

If you look at the costs of Hydrocephalus to the United States, there was a study that came out in the mid 2000s that estimated a greater than \$2 billion overall annual cost of Hydrocephalus in the United States, in pediatrics. And this is not just the cost of a surgery or a hospitalization. This is the cost of infections, malfunctions, ER visits, imaging of the head, the potential for radiation in places that don't have our MRI capabilities. So there's a much larger downstream effect of not having shunts in the world, that we hope to be able to continue doing this procedure.