Transcript of Revision Surgeries After Failed Hemispherectomy: Difficult Disconnections
Presented by Dr. James Baumgartner, MD
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Thanks for the opportunity and we are going to try to get through this relatively quickly.

Disclosures are as listed.

Hemispherectomy is an old procedure, it was first used in 1938, the first big pediatric series was in 1950 and the seizures freedom rate with the initial operation was about 60%, some would be more than that but the procedure was associated with a lot of post-operative delayed complications and it was later abandoned. So the first approach was the first failure.

Then in the main picture here and the upper right hand corner, [TRS Mason 01:11] was a Montreal Neurological institute came up with the idea of leaving a lot of brain in the head but isolating it, disconnecting it from the remaining brain and it said it would be as good as the original hemispherectomy in terms of seizure control but much less complicated in terms of trouble related to the surgery. He proved that and his approach has been modified by additioning of subsequent surgeons to be much less resective and much more disconnective with improved results.

So if neurosurgery is the Olympic Games, then hemispherectomy is the marathon – it takes a long time, it's technically challenging, we have to cover a lot of ground and especially when patients have abnormal brain anatomy it becomes a very difficult operation to perform and we are working on trying to make things better.

This is a review article from 2014 summarizing the results of various kinds of hemispherectomies and what you can see is that it is 70-80% affective percent which means we are failing 20-30% of the time.

When a hemispherectomy doesn't work, why doesn't it work? The first part of it is improper selection of patients. So, what's been very helpful for us in epilepsy surgery in the last 10 years or so is that the genetics of epilepsy have been better characterized and there are people with [---02:57] in whom hemispherectomy is not a good idea, as we know it doesn't work. Then if you are confused by the EEG information you acquire you can choose to pursue the surgery in someone how is not a great candidate. So that's the selection error.

The technical failures, there are 4 of them: You don't properly disconnect the Corpus Callosum which connects the 2 hemispheres, you don't properly isolate the posterior, interior, mesial part of the frontal lobe, you don't get an adequate resection in the insular cortex or you don't adequately disconnect during the temporal lobe tissue.

The current rates of technical failure – technical failure rates run from about 10% to as high as 54% depending on the technique, the approach and age of the patient. So the younger you are the more likely is that the surgery will not be as complete as if you are older. You have something called Hemimegalencephaly that makes it one of the most difficult things that neurosurgeons try and do and in spite of trying hard as you can to do well, not succeed.

So we decided at our hospital to try and do something to leave the completeness of the disconnection at the first surgery and that involved using an MRS intraoperatively with a link called DTI sequences to see before we

finished the operation if we had done a good job or not. If we found that we hadn't done what we wanted to do then we would go back at the time of the first surgery and keep working until the MRI looked good. We found in 32 consecutive cases from 2002-2014 that there were 11 cases where we identified inadequate disconnection or resection at the time of surgery and that let us get to a better outcome. So, this just summarizes what I was talking about: You use the MR with the DTI test to assess the completeness of the resection and then fix it it's not.

This is the DTI following the first and then the second intraoperative MRI. The white arrow shows you a surface, a C shape band of tissue that is connect on the left in A and on the left side you can see that this C shaped area has been divided. So the DTI sequences allow you to look at the white matter tracks in real time and get to a better state of surgical technical completeness.

The other thing we've chosen to do is to record from the insular cortex which is deep grey matter that sometimes causes trouble and what you can see on the left hand side of this slide is that there are squiggles on the EEG that were recording on the left hand side of it and then after we'd done an adequate resection that quieted down, and we think that helps as well.

This is a seizure freedom following curative hemispherectomy where you can see for us is that using this sort of approach we got to the point where 2 years out have about 91% seizure freedom which is a big bump up from the 60% of the initial operation.

Then Monica asked me to talk about the Hospital for Sick Children in Toronto experience where they went back and looked at their own failed hemispherectomy where 8 out of 88 that underwent hemispherectomy at that hospital between 1995 and 2012 experienced seizure recurrence that was identified as having been a technically inadequate operation. They used the same DPI approach except they used it after the fact to analyze these patients. So the initial patients were operated young at a median age of 9.7 months and their seizure freedom following the first surgery averaged 32½ months, so your seizures can come back pretty late and be related to incomplete disconnection. They had a very comprehensive repeat approach to these patients which included video EEG monitoring and they really focused on the MRI with DTF sequences.

The EEG monitoring and MEG analysis confirmed that the seizures were still coming from the hemisphere that they thought was the problematic hemisphere, then they did the MRI with DTI sequences and they found 3 different patterns of technical failure. One was incomplete division of the corpus callosum which is exactly what I showed you from our experience on that prior MR image. Incomplete disconnection of the temporal lobe structures and then in 2 cases there was incomplete disconnection of both the corpus callosum and the temporal lobe structures. So they went back and re-operated, mean age for the second circuit was 6.8 years and they used the same incision but they did a more limited opening and they used computer guidance to help them get to the place where there was the problem so they could take care of it. The operations were much shorter than the typical hemispherectomy, lasting 2-4 hours and only one child required blood transfusion. They did very well, 5 of the patients became completely seizure free and the other 3 patients were nearly seizure free after the second operation and their conclusion was that reoperation is an option for consideration for patients with recurrent seizures following hemispherectomy.

[---09:24] here and Jocelyn was a case that we learned a lot from a caused us to modify our approach. She was a good candidate for a hemispherectomy, she had seizures coming from one hemisphere and this is her MRI Scan. You can see it's on the right hand side of the slide, that the right half of the brain is a little bit larger than the left

half of the brain so she is a Hemimegalencephaly baby.

We did a modified functional hemispherectomy and the seizures recurred. We did a repeat modified functional hemispherectomy and the seizures recurred and then we did an anatomic hemispherectomy and the seizures recurred and it was very disappointing to the family, it very disappointing to the neurologist, it was very disappointing to the neurosurgery team. We really dug deep and changed how we approached patients and Jocelyn really was the impetus behind this series that I talked about earlier.

The other issue with repeat hemispherectomy is that surgical landmarks can be difficult to identify in a kid with Hemimegalencephaly at the first go but at the second time round it is really, really hard to know where things are and where you are and it is also very difficult to keep the spinal fluid from leaking out of their heads after you have been in several times because you just can't close things up as well as you would like to. So it is very much more difficult afterwards and you really have to plan carefully and the availability of navigation, whether you get computer guidance to tell you where you are in the head really is absolutely critical to doing a good job. So you want to really do a good repeat phase I evaluation , that's also more difficult for the neurologist after a significant amount of brain has been removed so you have to get everybody on board working together and you use whatever diagnostic sequence you need to pin it down and then you go.

This next slide – revised hemispherectomy surgery 2013. So you can see on the upper half of the left side of the slide there's a little area that's orange and that is based on a study called SISCOM and what you can see is there's a really tiny little area of brain that's causing all this trouble for Jocelyn. And we finally got in. The picture on the right half of the slide is after surgery, so it was a very small amount of tissue that was removed. It was an area of cortical dysplasia and happily she has been seizure free since.

Conclusions: Anybody can fail. Every neurosurgeon who does hemispherectomies has failures and you have to really get everybody in the room who is taking care of this patient to work together to get to a plan to be successful but I think that the take away point from all of this is that repeat hemispherectomy is often an option and the outcomes can be very good with that.

This is Jocelyn with her 'I train neurosurgeons', t-shirt and absolutely does and has and we're fortunate to have families that are willing to stay in the game and keep working to get to the good seizure end point. If there are any questions I will try and answer them.

Thank you very much, I really appreciate how much you really wanted to stay with us Dr Baumgartner so I hope you are having a great time in England. I have a difficult question I think for the neurosurgeons - I don't know Dr [---13:12] is still in here and Dr [---13:14] is in the back. Brain shift after anatomical hemispherectomy historically in the published research has been a significant issue. I know it has not been reported in over 30 years but we have quite a few children in our community who have had a shift after hemispherectomy. Research and literature says 1cm is bad, our son shifted 3cm and started to herniate across the falx, so I know a lot of families want to know how far can you shift before it becomes an issue?

I wouldn't put a number on how far the shift goes before you really need to worry, I think it is much more important to look at the patient and see, in a team fashion, to look things over to try to decide whether you think that the shift that we are seeing is something that is causing trouble or is something that you can ignore. I have seen some children who have had a lot of shift who have not really had any issues related to it, I have also seen

some children with a little bit of shift who did have issues related to it. I think you have to factor in the hydrocephalus if it is there and do a multidisciplinary approach to figure it out but I don't have any hard or fast numbers on how much is too much or how much is too little.

Should a child post anatomical be monitored on a regular basis, maybe with an MRI maybe 2 a year to check for shift and shift progression?

Again, what I am more worried about is the delayed presentation of hydrocephalus after an anatomic or a functional hemispherectomy because they have higher rates of hydrocephalus post op and that is something that you can really do something about if it shows up. The shift thing, I haven't ever operated on the shift just because it is there, I have operated on the shift if there are symptoms related to it. So I think it is more important to examine the patient and talk to the neurologist and see if you think it is causing problems rather than just serve a knee jerk, 'this is too much shift, we've got to do something'.

Anyone in the audience have any question for Dr Baumgartner.

Thank you very much - we're going to hang up on you!

OK thanks, bye bye.

For more information on revision surgeries after failed hemispherectomy, go to www.brainrecoveryproject.org