

Dr. Superina: You know, you started the podcast by saying that we are a big transplant center, but we try to show the world that these patients don't really need a transplant most of the time that they can be treated by non-transplant means.

Amanda Wilde (Host): Children can experience several conditions that lead to liver transplant. The goal at Ann and Robert H. Lurie Children's Hospital of Chicago is to improve quality of life and avoid transplant whenever possible. So let's learn more about the cutting edge treatment at the Liver Center at Lurie Children's with transplantation experts, Dr. Riccardo Superina and Dr. Caroline Lemoine. This is Precision, Perspectives on Children's Surgery. I'm Amanda Wilde, Dr. Superina and Dr. Lemoine are world renowned authorities in the field of pediatric liver, kidney and intestinal transplantation. They are two of only a handful of surgeons in the United States who perform highly specialized transplantation procedures in children.

Doctors, thank you for being here. Tell me about the Liver Center at Lurie Children's. Dr. Superina, you are the creator and leader of the Liver Center. What is your mission? And can you explain your role?

Riccardo Superina, MD (Guest): Thank you, Amanda. It's a pleasure to be here with you. I think that the concept of a center is one in which the center brings together healthcare providers from many different specialties that will then address the needs of a child with a variety of liver problems. So liver problems can vary, anything from malignancies to disorders of the bile ducts, to disorders of the circulation of the liver, and often what a patient may encounter is a bewildering array of appointments that they have to do, tests that they have to undergo.

So our mission with the center is twofold. One is to publicize the fact that we have a group of liver centered specialists here, and second is to form a network of specialists that can then facilitate the care of the child and to make it easy for the parents to connect with everybody who would be involved with this patient's care.

And it's not necessarily that every child needs to see every specialist, but there is a well oiled connection between, let's say the people who first take the call, the liver pediatricians, the imaging specialists and finally the surgeons who will plan the operation in conjunction with a liver specialized unit in the operating room, who works on liver day in and day out so that it's both speedy, efficient, safe, and effective.

Host: Dr. Lemoine, do you work on liver day in and day out?

Caroline Lemoine, MD (Guest): Yes, absolutely. And thank you also, Amanda, for having us here and have the chance to talk about our Center. So just like Dr. Superina, I'm also a pediatric surgeon and a transplant surgeon, which means we have specialized in both areas of surgery and definitely as part of the Liver Center, we see a lot of patients with various liver disorders, but it's actually not exclusive to the liver. It also encompasses the bile ducts, but also the pancreas, amongst other things. But yes, as part of the team, more and more, where you realize that it's a teamwork, right? And so it's good to have more than one brain in a specialty to think about things. And, being more than one surgeon allows you to go through very complex surgeries and have two sets of expert hands, and two sets of expert heads thinking about a problem and troubleshooting and coming up with creative solutions to those very complex problems.

Host: Dr. Superina, Dr. Lemoine just talked about how you bring in specialists from all kinds of areas. What might those areas of medicine be that you bring in for the various conditions that you treat?

Dr. Superina: Well, our partners in this Liver Center include primarily the pediatric hepatologists who are our counterparts, when it comes to the care of the liver. The medical imaging people in particular, the interventional radiologists, who are our partners in the surgical approaches, both, in the diagnostic area, as well as in treatment areas. For example, you know, I could give you a number of examples where we partner with our interventional radiology colleagues in planning an operation, and, delineating the anatomy and also in actually performing parts of the quote unquote surgery, to deal with any particular problem. We work with our surgical oncologists very closely for the treatment of patients with liver tumors.

And then there's the whole group of people who work with us in the intensive care unit, where we have a highly dependent relationship with them. And there are certain people in the ICU who have an affinity to the treatment of some of the things that we treat surgically. And then finally we have our APN's, our advanced practice nurses who work with us and who are also extremely specialized in this area.

So this is just a kind of a small example. We bring in endocrinologists sometimes, you know, for example, for some of the circulatory disorders of the liver, they may manifest themselves with a endocrinological problem. And so we have them come in and help us with the diagnosis and the treatment of the patient.

We're sort of like the center of the wheel and the spokes that go away from us are all the various subspecialties that we are affiliated with, and that we work with almost on a daily basis, but I think the biggest partners. Oh, and forgot to mention anesthesia, our operating room team is key to the success of these patients.

So unlike other hospitals where there might be the uncommon perhaps occasion when there's a liver operation, there may not be a liver specific anesthesiologist. It might be an anesthesiologist who does many other types of operation. Whereas when we operate on a patient with a complex liver disorder, we have a very specific group of anesthesiologists who partner with us and a very specific group of nurses who partner with us. So we don't just get whoever's on call that day for a specific room. It's a very tight group of people.

Dr. Lemoine: If I can just add on something to Dr. Superina, specifically for the patients that have a disorder called the Abernathy malformation, which has congenital portosystemic shunt's, where the blood is driven away from the liver, about 25% of 30% actually may be higher of those patients also have cardiac anomalies. So the cardiology team is also very involved in the care of those patients to make sure that it's safe from a cardiac standpoint to operate on them or to treat them. Sometimes we also have dermatology conditions also, so dermatology can be involved. And, one of the consequences of that shunting of the blood away from the liver can create disorders of the lungs. So it really takes a village, like a whole group of people to take care of those patients. And, at the center like Dr. Superina was saying, we involve the specialists based on the individual needs of each patient that comes and sees us. It's really tailored to the associated conditions that that patient has.

Host: So taking the Abernathy malformation and all these people you have involved. Dr. Superina, how do you then proceed with planning treatment with all these cooks in the kitchen?

Dr. Superina: Well, it depends on how the patient comes into our system. So for example, this is a very uncommon condition. And often it's the parents themselves, who've done the research, and reach out in particular to me, or they find their way to me through perhaps hepatology or pulmonology, but many of them reach out directly to me.

So then my job, and many of them come from different states. So we have a very broad scope of referrals, national as well as international. And so our first sort of stop is our International Center. I think it's has a different name now. But because it used to be only dealing with international patients, but now it deals with people from out of state as well, because many of the needs of patients

from out of state are the same as people from out of the country. So they make a lot of the preliminary arrangements. There's a lot of administrative details that have to be taken care of first. Then when the patient finally comes to see us, we will review the imaging generally with our radiological colleagues. And that is a very important element of planning for the care of these patients.

The anatomy can be quite different. In fact, we have a classification system that we're working on. The original classification system came up in 1994, believe it or not when I was working in Toronto and people still use that classification, but it's a very simplistic one and it doesn't really cover all of the various anatomical details.

So after we've reviewed the imaging, then we go ahead and usually have them seen as Dr. Lemoine said, if they have some sort of pulmonary disorder, the manifestations of the Abernathy's malformation are legion. There's so many ways that they can present. So once we've taken care of all of those details, then we go ahead and we plan the operation and informing the parents and the child of what comes ahead is an extremely important part.

We don't want them to be surprised or disappointed, and they have to stay in the city of Chicago for about three to four weeks at minimum. They know that they're going to be in the hospital for about two weeks. So there's a general pattern for the treatment of these congenital portosystemic shunts. Some of them can be treated just by our interventional colleagues. Then it's a very quick, and I would say relatively uncomplicated course, although what they do is very complicated, but they do it so often that in their hands, it's not as complicated. It's the other 75% of the cases who come to see us need an operation. And usually it's a two-stage operation. So then we have to tell the parents exactly what that's going to involve. So, getting the contact, getting the administrative details worked out, reviewing the imaging, getting the ancillary programs involved as needed and then planning the surgery, and then planning the outpatient care because many of these patients will be followed by us for years.

I mean, they keep coming back and back and back because we don't have the time to go into all the different things that can happen to these patients or what can happen. You know, some of them may have liver tumors that need to be addressed and we get our oncology service involved. We also have a very big research arm that is continuously publishing on this data in order to make other doctors aware that these cases do not need a transplant.

You know, you started the podcast by saying that we are a big transplant center, but we try to show the world that these patients don't really need a transplant most of the time that they can be treated by non-transplant means. And when

the patients come to see us for a second opinion and hear that, you can imagine their relief. But it's very rewarding for both us and for the kids and for the families, obviously.

Host: So some of the surgeries you do Dr. Lemoine, will help these children avoid liver transplantation?

Dr. Lemoine: Absolutely. Actually like Dr. Superina was saying, and you were also mentioning when the patients and the families come to us first, often, they'll have been told that they would need a liver transplantation for otherwise their liver is functioning just fine. The only problem for the Abernathy malformation or the main problem is that the blood flow that is supposed to feed the liver and making it grow and be healthy, is actually hidden and derived into the systemic circulation as we call it, which it goes back straight to the heart without being filtered by the liver.

And they're told by other teams that they need a transplant because there's one vein that's lacking and that is called the portal vein that feeds most of the blood into the liver. And what we do here with the help of all of our colleagues, especially in radiology is we unmask the presence of that vein. And if, we are able and we are able in the vast majority of cases to restore some portal venous flow to the liver, then the liver will receive all the nutrients that it's supposed to receive. And therefore you can avoid liver transplantation. And for families and providers and other institutions who look up on the internet, or sometimes they try to look up the scientific publications, there is a lot of publications on liver transplantation. So one might assume that this is the treatment for that condition because that's what other people have done.

But actually in our experience of I think the latest number is probably somewhere close to 80 patients and believe it or not, that is one of the biggest experience in the world. And certainly the biggest in North America, only one of our patients ultimately needed a liver transplant and that patient was very special. I mean, they are all different than special obviously, but that patient had another condition, which in itself needed a liver transplantation so that the child was not transplanted because of his Abernathy malformation, but for the other condition of the liver. So yes, in the vast majority of cases, those children do not need a liver transplantation because when you get a liver transplantation, it's curing your life for other diseases, but it also exposes you to taking medication for the rest of your life and other conditions and complications that can arise.

So if a child or a patient of any age can keep his or her liver and avoid a tremendously big surgery, like a liver transplant and all the conditions that go with it, then the medications, then that's definitely the best thing that a child can

get.

Host: Dr. Superina, how has this procedure advanced over the years? It sounds like from what Dr. Lemoine is saying that you can do things now you may not have been able to do a couple of decades ago and avoid liver transplant that you might've had to do in past situations.

Dr. Superina: What's changed is our approach to this whole problem. When I first started doing these operations, in the mid nineties, the first two patients actually got a transplant and one survived and one didn't and there was very little known about this whole condition. And then these patients came to see me very sporadically. It wasn't like there was a constant stream of patients because it was really a relatively unknown problem. It was more of an anatomical curiosity, but then after we published this first small series, with a classification system, people started writing to me saying, you know, what do you think we should do about this patient?

What do we think we should do about that? And some of these patients started coming themselves. I mean, it's amazing how literate some of the families are, who have kids with these conditions and they just want to know what to do about them. And very often, the answer from other physicians and surgeons is that, well, we can't do anything about it.

And if we're going to do something about it, you're going to need to have a liver transplant. So that was sort of the state of the art about 30 years ago, and then gradually, in the first decade of the 2000s, we started approaching these patients with one stage procedure. We didn't really know very much about how the body was going to react to having the sudden change in hemodynamics. And then we evolved from a one-stage procedure to a two-stage procedure. Then we started doing the interventional piece before the actual surgery so that we would know how the body would react to the sudden closure of this very abnormal communication. And so, it's evolved and then of course we started learning that some of the conditions that are associated with the Abernathy malformations, such as the failure to be able to exchange oxygen in the lungs, something that you wouldn't expect have happened from a disorder of the liver, but it can affect the lungs and the heart.

We found out that these things were actually reversible so that it became more imperative to have these patients treated earlier rather than later. And then it became known that a lot of these kids suffered from neurocognitive problems. Some of them couldn't concentrate, they couldn't go to school, they couldn't sleep properly. And that's because of the very high ammonia levels that are associated with these conditions. So we've learned a tremendous amount and

one of the things we do a lot of right now is to actually do research on this subject so that can bring to light these problems, unmask them, and then offer solutions that are not dependent on transplantation.

We know that many of the conditions that are associated with the shunt are reversible. So relatively unknown condition, 30 years ago. And now, to the point where we have international conferences simply on this one subject.

Host: Which shows that the body of information is greatly increased. Dr. Superina, you mentioned the research arm of the pediatric liver center. So, in the context of what you just said, what do you think the future holds?

Dr. Superina: Well, we do mostly outcomes research on the surgical side. That is we study the effects of the surgery. We do some prospective studies. For example, now we're doing a prospective study on the effects of shunt ligation on neurocognitive function. We also do a lot of research on the condition called portal vein thrombosis, where we are following these kids for years and trying to understand why some of them failed to grow properly and start to grow immediately after portal vein recirculation has started. There's research on cancer. We have, one of the things that I failed to mention is that we have an internationally known pathologist now who's working with us on many of these projects. So there's going to be a better understanding from a pathological standpoint.

And the Liver Center also focuses on things like biliary atresia, that is a disorder with which babies are born with and is the single largest cause of liver transplant. And there are among us, the hepatologists in particular who are doing some basic science work and trying to understand the pathogenesis of this disorder and some of the things that can be done to ameliorate it.

There's research in the ability to reverse some of the metabolic diseases that afflict children like progressive familial intrahepatic cholestasis, alagille syndrome. So there's a wide ranging amount of research that's done and one of our missions is to enlighten people.

And There are many, many conferences and venues in which some of this research is highlighted. And of course funding is a huge problem. So we're always trying to get funding for these and philanthropic gifts, also help to a great extent. So there's a lot to do, with this. And, there's a lot more that can be done.

Certainly, in the last 30 years, I think we've shown that there have been great advances in some of the things that we do particularly well here, and hopefully

we'll continue to do well. And of course education is probably the single most important thing. So education, not just the families, but bringing a younger generation of caregivers and educating them to be able to continue this work into the future.

Host: So education is a big part of the mission of the center as well. Dr. Lemoine, any thoughts you want to add about the future of treatments or the studies that you're doing?

Dr. Lemoine: One of the conditions that we treat is called portal vein thrombosis, and for that condition, we need to create a bridge between the veins of the bowel that feed the liver. It's that portal vein that is no longer present in those children. And to bypass that obstruction, we have to use other veins of the body, like a vein of the neck or a vein in the abdomen. But wouldn't it be great if we could have those veins handy and not have to take a vein in the neck or otherwise, if we could grow those veins, it would be a great thing. And that, could be something that could be done to help, specifically that condition or also in the setting of transplantation.

But, I think otherwise the research is trying to understand more diseases such as the Abernathy malformation, right? It's a developmental disease. We don't know why some children develop it than some others don't, and how some develop complications or not, or how the body adapts to reverting, to quote on quote the normal circulation of the liver and how that happens very suddenly in just a few days, and how the body needs to adapt.

I think there's a lot of body of research that needs to occur in that very important moment, those few days when the circulation changes and to expand a bit on the education, you were mentioning a lot of not only families, but trainees, that come to our institution have never heard about either the Abernathy malformation or very minimally about, the condition of extra hepatic portal vein obstruction that's alongside portal vein thrombosis.

And that reflects also the lack of knowledge in providers currently in practice and the importance of educating the doctors of tomorrow so that they know of the condition and they know how to treat it. They know to refer it to the right person, to do those operations also and also to teach the physicians who are currently in practice because if it's not available in their center or even if, they are aware of it, but not that much, maybe again, transplantation will be recommended because that's what's available in the center where those providers are practicing. So teaching them about the alternatives will help children ultimately get the right surgery for their disorder.

Host: So education for professionals and patients is always an evolving undertaking along with research and treatment.

Dr. Lemoine: Correct. That's very well said.

Dr. Superina: I think education of people like her is key. Like she will carry on this tradition. And so, you know, we need more people like her, pediatric surgeons who have a special interest in the liver so that they can go on carrying on this work. And, she's the future. And other people like her will be the future of this field.

Host: Well, Dr. Superina and Dr. Lemoine, thank you for sharing your expertise and for your continued work to advance treatments and outlook for children with liver conditions.

Dr. Superina: Thank you anytime. Pleasure speaking with you.

Caroline Lemoine, MD (Guest): Yeah, thank

Dr. Lemoine: you for the invitation.

Host: It was great having you. Visit Luriechildrens.org/livercenter for more information or make an appointment by calling 1-800-KIDSDOC. I'm Amanda Wilde. Thanks for listening.