

Treating Heart Failure in Adults with CHD Heart Liver Transplant

Announcer: Welcome to the Mayo Clinic Cardiovascular Continuing Medical Education podcast. Join us each week to discuss the most pressing topics in cardiology and gain valuable insights that can be directly applied to your practice.

Dr. Burchill - Welcome back to Interview with the Expert, a podcast series from Mayo Clinic Cardiovascular Education. I'm your host, Dr. Luke Burchill and I am leading the Adult Congenital Heart Failure Care Pathway here at Mayo Clinic. And joining me today is Dr. Julie Heimbach, Professor of Surgery and Director of the William von Liebig Center for Transplant. Thanks for joining us.

Dr. Heimbach - Oh, thank you very much. I'm delighted to be able to meet with you today.

Dr. Burchill - So Dr. Heimbach, I'm a cardiologist. You are our liver transplant expert. I'm going to start with the simple question, why does the heart affect the liver?

Dr. Heimbach - Well, you know, it's surprising how often this connection is missed. Actually, patients that we think, you know, maybe just have a heart problem, unexpectedly, when we look further, we recognize that there's involvement of the liver. And this has probably been one of the biggest advancements in caring for patients after their Fontan procedure as they, you know, progress through their teenage years. And we recognize that there can be injury to the liver and this is really primarily related to the continued increased pressure. So the venous return is not normal. The liver is subjected to sort of a continuously higher pressure environment that leads to an inflammatory process, a scarring process that can progress over time to fibrosis, advanced fibrosis, cirrhosis, and unfortunately even hepatocellular carcinoma, which is a, you know, a primary tumor of the liver that occurs usually in the setting of an inflammatory state and in the setting of cirrhosis in particular.

Dr. Burchill - And you mentioned the Fontan patients which is certainly probably the most common reason we end up collaborating and discussing patients at the time of transplant evaluation. Are you seeing many other patient groups, other people with say right heart disease coming in with changes in their liver?

Dr. Heimbach - Definitely. And this is probably the population that is now, you know, being missed because we're so tuned into the Fontan patients, but we don't always recognize this in other patient populations. And it's surprising sometimes even the right heart problems are missed and we are just treating the patient for their cirrhosis and then we understand that actually the liver is not the primary problem that there's actually a heart problem as well. So we definitely are seeing it particularly with, you know, valvular conditions or other congenital heart conditions or other reasons for the right heart to not be working as it should.

Dr. Burchill - And I agree we are seeing a lot of this. So for me now I see right heart disease, I wonder about the liver and what's interesting is how often that liver does look abnormal on the screening ultrasound. So the next question I have for you is what does an abnormal appearing liver actually mean or tell us when it comes to someone's need for evaluation for potential liver transplant?

Dr. Heimbach - Yeah, so this is, you know, I wish I could say well it's just a, you know, a blood test that we can check or there's a sort of a really clear, you know, evaluation that tells us a hundred percent that this is what needs to be done. So it definitely requires further evaluation and abnormal appearing liver on screening ultrasound is always a good for a start, but it, you know, we don't know for sure just based on that test that they do or do not need a liver. I think that any finding that suggests liver involvement needs to be followed up and then we would have to look at that with a variety of different techniques. And depending on the clinical scenario, you know, we can be reassured or we even have to proceed to more invasive tests like biopsy. But even biopsy is not always as, you know, as helpful as we would like it to be. So it is a huge challenge for us, but luckily we are building a, you know, increasing level of expertise with us and an ability to make that decision.

Dr. Burchill - So I was always trained to think about the liver and its synthetic function, so I'm looking at albumin, I'm looking at prothrombin time or INR. And then more recently incorporating things like the MELD score into my clinical practice and my thinking. So for the practicing cardiologist out there who may have identified, here is a patient with some problems with their liver, can you maybe provide just further information on what other tests are being used in common clinical practice that are informative for staging where that liver might be in terms of its function?

Dr. Heimbach - Yeah, I do think you've hit on, you know, some really important things looking at the, you know, the synthetic job of the liver. The liver has so many different jobs but producing the proteins, you know, so albumin is good only it is an acute phase reactant so anytime of you know, illness it's going to go down. So that's where we have some limits to the albumin. But in a stable outpatient, that's a good test. The INR is a really good test, but it can be impacted of course if the patient is taking anticoagulation so that you really can't use it in that scenario. But in elevated INR in the absence of anticoagulation in a well-nourished patient I think is an excellent early screening test. The ultrasound, as we've already mentioned, is quite valuable because it tells us what the echogenicity of the liver appears and if there's any masses and of course ascites can help us although that can also be present in the setting of heart failure. So that's why there needs to be an interpretation. The other problem with ultrasound is it's operator dependent. So you do need a degree of expertise to perform that test. We then can go to cross-sectional imaging which helps us look for signs of portal hypertension. We can use either CT or MRI. The problem with MRIs, if the patient has implantable things that restrict the use of MRI, then that, you know, is a problem for us. CT requires the use of IV contrast so in the setting of renal dysfunction we have to be quite cautious with that. But those tests are really valuable to us, especially in doing the screening for any worrisome lesions that could be hepatocellular carcinoma which would be really critical to identify prior to proceeding to transplant. It doesn't preclude transplant, we just have to know about them and potentially treat them or be sure they're

within criteria. So it is a complicated set of assessments that we perform but the basic labs bilirubin is helpful to add in there with INR and albumin, ultrasound is a good test to get. The rest I would probably defer further. You mentioned the MELD score and the value of the MELD score is it is a score that is developed in patients with chronic liver disease that is quite predictive of their risk of, you know, further liver problems or essentially it's death from liver disease in the next three months and originally developed for patients that were being considered for TIPS but now is being used for patients as a prioritization for wait listing for liver transplant. The problem with MELD is the creatinine is part of the system. So if patients have intrinsic renal disease the MELD score will be impacted by that. And the INR is also part of that. And again, if the patient is taking anticoagulation that will impact that scoring system. But a score above 12, we should definitely get anybody's attention.

Dr. Burchill - And I think what I'm hoping as a key message just from this brief dialogue is that this is something to call on your colleagues for. I think that, you know, even for those of us that are physicians that started as physicians before we sub-specialized, there's so much happening. I think specifically within the liver world and risk stratification and understanding disease processes, prognosis, I know that we rely heavily on that collaboration with the GI, hepatology and if necessary, the liver transplant team. So for those listening out there, call a friend, phone a friend is the key message.

Dr. Heimbach - Yeah, I think collaboration in this particular assessment and decision making process is really valuable. We definitely couldn't make these determinations on our own within the field of GI hepatology and transplant. So we definitely find the collaboration with our cardiology colleagues are essential in sorting out what is the next best step for these patients.

Dr. Burchill - And so let's imagine we're talking about someone who we really understand, they have both end stage heart disease and end stage liver disease. And these are the patients that we're talking about combined organ transplantation, patient in both heart and liver. Maybe if we start really in simple terms is this very common? Is it something that's really available to many people?

Dr. Heimbach - Well that's a great question and it sounds, you know, really amazing to think about doing two organs at the same time, especially heart and liver. But first of all, luckily it's not that common and the expertise tends to, I would say lodge in a small number of centers. But I think in those centers we've become relative, you know, I don't want to say it's routine, but it's definitely something we feel pretty confident about. At the centers that are doing it, it's definitely something that the patients do well when they're well selected and we plan things and we screen for the things that we know we need to screen for. It can be done and it can be done safely with outstanding outcomes.

Dr. Burchill - Okay. And when I mention heart-liver, the most common response that I get is or the question that I get is, "Oh so which do they get first? And how do they stay alive without one while we're waiting for the other." So I'm going to let you answer as to exactly what heart-liver transplant really means particularly in terms of sequence and timing.

Dr. Heimbach - Yeah, so the key part is that it's typically done, you know, what we typically do is a simultaneous heart-liver transplant. You know, if the patient has advanced heart failure and you know their liver is cirrhotic, we need to do them at the same time. Sometimes they do, like for a patient with an inherited condition called amyloidosis, it would be possible to say do the heart and then, you know, a couple months later do the liver because the liver is really a metabolic problem. It doesn't have an, you know, intrinsic failure. So that would be possible in that scenario. But otherwise when both organs are, you know, experiencing dysfunction, we have to switch them at the same time. And typically we would have the patient undergo general anesthesia, being completely asleep. We would proceed with dissection for the heart, put the patient on cardiopulmonary bypass, do the heart transplant, come off bypass, do the liver transplant. That would sort of be the most standard. There is a group that does unblock, meaning they put both organs in connected to each other all at one time, that's not common. The first scenario is most common. And another way that we've actually found that is important for a select group of patients who are what we call highly sensitized, meaning they have antibodies against other people's cellular markers. What we know about the liver is it's extremely good at managing those HLA antibodies and we see those antibodies disappear very rapidly when we do liver transplant. And that's why we don't really need to worry about the cross-match in most liver transplant patients. And the highly sensitized heart-liver candidate we actually do put the liver in first and then put the patient on bypass and then we put the heart in and we recirculate. So that's another option. Typically the first option would be most common.

Dr. Burchill - So I'll just highlight again for the listener, if you are seeing highly sensitized patients more common with those adults with congenital heart disease because of their exposure to different tissue, different substrate, high levels of circulating antibodies, we do have heart after liver transplant now as an option for a select number of people. And I think that's really important for increasing the accessibility of transplant for those highly sensitized patients with some good early results reported by our center for those that have undergone heart after liver transplant. So that's exciting sort of new work that's just happening now.

Dr. Heimbach - And just to clarify, it's all still at the same operation, it's just the order by which the organs are placed into the patient and blood is then circulated through those organs, what is all one operation essentially.

Dr. Burchill - Yeah. And maybe people are always interested in the waiting list and how it works but does it become even more complicated for people who are listed for both heart and liver transplant? How does that work?

Dr. Heimbach - Yeah, that's a really key thing to speak about and you know, we may have seen in the press, especially recently, that you know, there's a lot of ongoing change in organ allocation. Currently it is possible to receive two organs and they would come from the same donor, to clarify that, and the process by which people are prioritized on the wait list remains, you know, still not whether the heart is this priority and the liver just follows with the heart or the liver is the priority and the heart follows the liver. Both of those scenarios are possible but

most commonly the patients, you know, most urgent status is the heart and the liver would be allocated with the heart under a specific set of criteria that relate to the distance around the donor hospital and from, you know, the recipient hospital to the donor hospital. But typically the way that the patient is prioritized on the list has to do with their, you know, most urgent condition, which is in most cases you know, the illness that they're experiencing from their heart. So their priority status or their position on the wait list is determined by the degree of heart dysfunction or heart failure that they have which is a very specific set of criteria. And then of course other factors such as their size and of course their blood type needs to be compatible. And in rare cases when the liver is the most affected and the sickest organ, then we could be prioritized according to how much liver failure they're experiencing which again is already something we talked about calculated by the MELD score and then the heart would be required to be following the liver in the allocation sequence.

Dr. Burchill - Thank you. Well this has been a fantastic conversation. I'll just finish with one last question and it's about those patients that have had a transplant, maybe they've had a heart-liver transplant. What is the follow-up of those patients from a liver perspective?

Dr. Heimbach - Yeah, so that always gets to, you know, if the patient has two organs, you know, or do we have two cooks in the kitchen? This again requires continued collaboration. Luckily, the care of a post-transplant heart patient and a post-transplant liver patient, they have a lot of similarities. And so for example, the anti-rejection medicine, you know, really similar and we screen for many of the same infections and use a lot of the same medications to prevent, you know, conditions like ulcers and other things that can happen after transplant. So we really find that the care is quite similar. There are specific conditions that can happen and so of course we need to continue to see those patients together. So they typically would be coming to see their heart transplant team and then there would also be visits scheduled with the liver transplant team. But we definitely let, you know, one team sort of run the show. We've already organized who that would be and it's typically that we let the heart team manage the anti-rejection medicine and then if there's a problem with the liver, we, you know, need to step in. Otherwise we just, you know, schedule our visits around the heart visits. And typically the hospital stay and their recovery is not that different for a heart-liver patient compared to a heart transplant patient alone. Many similarities and really how long people need to stay in the hospital or other things like that really are more determined by how strong they're coming into the surgery, most of all. And if there are any setbacks, you know, following the surgery that require extra procedures to be done, that can lengthen the hospital stay. But otherwise the follow-up is very similar.

Dr. Burchill - Thank you Dr. Heimbach. This has been a wonderful conversation. I'll just summarize, firstly, think about the liver, particularly for right heart disease. We know the Fontan associated liver disease patients are a key group but we're also seeing this in other patients who have significant right-sided valvular heart disease. So think about the liver, phone a friend. This is a multidisciplinary collaboration that leads to best outcomes for those that have evidence of liver disease in their setting of their heart failure. Heart and liver transplant is an option for a small but important number of people and we are finding that it may be very helpful, particularly for those that are highly sensitized in a few select centers that are offering heart after liver transplant. And we just heard about some follow up, as again, being a collaborative model of

care. So I hope that you've enjoyed this information and thanks again to Dr. Heimbach for your time.