ALEXANDER EGBE: Greetings. I'm Dr. Alexander Egbe, Assistant Professor of Medicine and Pediatrics at Mayo Clinic. Today, we'll be discussing atrial and ventricular arrhythmia in complex congenital heart disease. I'm joined by my colleagues Dr. Bryan Cannon, Associate Professor of Pediatrics, and Dr. Christopher McLeod, Associate Professor of Medicine, who specialize in electrophysiology. Welcome.

CHRISTOPHER MCLEOD: Thanks, Alexander.

ALEXANDER EGBE: So let's start off with Dr. McLeod Please can you talk to us about medical management of a atrial arrhythmia in congenital heart disease?

CHRISTOPHER MCLEOD: Yeah, it really is an important one. There is so many this thing differences in the rhythm management itself just because the mechanisms are different, that I think important for every general cardiologist, every intern internist to realize that we don't just use standard medications. I'm going to speak a little bit about the mechanisms, because it gets into why the medicines themselves are different. Because these are typically young patients. They typically symptomatic.

They have fast arrhythmias. And if you think of the noncongenital heart disease population, that group of people, that group of patients who are young with otherwise normal hearts, we usually use class 1 C drugs or even beta blockade, AV nodal blockers for those. And this distinctly sort of the approach that shouldn't be used, or doesn't really work very well for this group of patients. So because these are re-entrant scar related arrhythmias most of the time in the complex repair congenital heart disease patient, we want to increase re-polarization and break these reentrant flutters.

So that's the class three drugs. It's your sotalol, and the dofetilide, maybe amiodarone, depending on the patient. It's never usually our first drug. But it's those [INAUDIBLE] drugs that are specifically studied, specifically proven to be of useful and definitely safer in this group of patients who very often have ventricular scar. I'm sure the audience knows that anyone with ventricular scar-- if you're only going to use a class one drug like propafenone or flecainide, you can precipitate dangerous arrhythmias.

So that's the standard approach for an adult with normal heart disease. The class one drugs when started with normal hearts. But with complex congenital heart disease, we really have to steer away from those.

It's an important difference. Bryan looks after a generally a younger population than me. Is there anything different from your side there?

BRYAN CANNON: It's basically the same principles and the same philosophies, but you just have to realize that a lot of times there's complex interactions. Sometimes there's sinus node dysfunction and other things that you have to take in to a role whenever you're picking the anti-arrhythmic medications that you choose. And the standard things, like you said, that may relate to an adult after a mild cardio infarction not don't necessarily relate to a complex congenital heart disease. It may have multiple scars, multiple circuits a flutter, or sinus node dysfunction, or other things that may cause problems.

CHRISTOPHER MCLEOD: Yeah. It's an important one. The other thing that I just want to make sure that the fellows who are watching this, it's just understand for their training, for atrial flutter, these re-entrant atrial flutters, the atrial cycle length, how long it takes for a revolution of these, is typically much slower than the regular [INAUDIBLE] flutter, the typical flutter. If it's a slower atrial cycle length, it gives the AV node longer to recover, and thereby you can conduct quicker paradoxically down to the ventricles.

So slow atrial cycle lengths, slow atrial flutters have paradoxically this faster ventricular response. And that's why AV nodal blockers, metoprolol, [INAUDIBLE], they really don't work in this group of patients. We almost always have to go into an anti-arrhythmic strategy. It may be drugs or it may be ablation, but just making sure that flutter, which in some patients you can [INAUDIBLE] control. It usually isn't the case in this group of patients.

BRYAN CANNON: Yeah, kind of dovetailing off that a little bit, I think that one of the biggest challenges in this population is actually picking up patients that have arrhythmias. When we talk about the standard atrial flutter, that's a circuit using the entire atrium. So it's pretty easy to see in an EKG, because you'll see that typical sawtooth pattern.

However, the kind of flutter that we see in congenital heart disease patients is something that we call scar flutter, or more properly turned to intra-atrial reentrant tachycardia. This is very different and it can actually fool you on an ACG because there may be periods where you have an isoelectric baseline or a flat baseline, and then you'll P waves coming through. So you have to very carefully look in the T wave, you have to look for patterns to look for intra-atrial reentrant tachycardia or you'll miss it.

A couple of clues is if you see a P wave axis change on the surface ECG, or a patient who's normally bradycardiac in the 40's or 50's, has a heart rate in the 90's or 100's, because as Chris mentioned, these tend to be slower flutters. So half the battle is actually picking these up. In addition, if you take a look on Holter monitors, if you see no heart rate variability, or you see a standard heart rate, those may be clues that you have intra-atrial reentrant tachycardia. And certainly, it's easy to miss on an ECG.

CHRISTOPHER MCLEOD: Bryan just pulled up a very important point, and we should have probably brought it in earlier. But really, these patients can present with, as he was suggesting, heart rates in the 90s or 100s. Just because the patient has congenital heart disease doesn't mean that they should have a faster heart rate.

They can be very well compensated. Heart rates are generally on the low side. Someone with a heart rate of 90 or 100, you really need to be suspicious that there's not an underlying intra-atrial reentrant flutter going on.

Another subtle clue would be patients doing well, not aware that they're in flutter, but ventricular function has gone down. And they've been just too long in a slow atrial flutter, not enough heart rate variability, and the ventricular function has decreased.

ALEXANDER EGBE: Thank you very much for this insight. Doctor Cannon, can you discuss the role of [INAUDIBLE] for management of atrial arrhythmia?

BRYAN CANNON: Sure. I think it plays a very important role. I think that that's an important thing for two reasons. First of all, as you know, all of the medications have side effects. Our most effective medication that we can use is typically amiodarone. But as Chris mentioned, amiodarone has a litany of side effects.

And it's not something we like to leave patients on long term. In addition, most of the other medications have some type of side effect that may be undesirable. The second thing is it's just on medications alone, the recurrence rate tends to be very high. So a lot of times, we'll tend towards taking them to the catheterization lab. Of all the procedures that we do, I think that these can be the most challenging.

They take a long time. You need somebody with expertise, with knowledge about the basic underlying anatomy. Also ablation techniques, because the ablations that we do in these patients can be more complex because of scars, because of the thickening of the atri and the ventricles. And it can make these ablations very complex.

Having said that, if you have a center that has expertise in doing ablations in congenital heart disease patients, we can have a pretty good success rate. And that would give us the advantage of that we can avoid them having symptoms. We can hopefully help them with their ventricular function or other issues that they may be having, and may potentially be able to create a situation where we can have them off medications rather than having them on that long term.

The other thing we have to realize is that by doing one ablation, it's not one and done. These are patients who would typically have a recurrence. And if you take a look, about half of patients will have a recurrence within five years.

So it's a high recurrent situation. So you have to be diligent, even after you performed an ablation to make sure that the ablation has been successful. Chris, anything you want to add about that?

CHRISTOPHER MCLEOD: Yeah, there's one important point. The one thing I guess for people to realize is atrial arrhythmias in their own right, aren't really that dangerous. Obviously you don't want ventricular function to decline because that's not going to be good.

But they're not malignant like some of the ventricular arrhythmias. They're not associated with sudden death. Whereas you're starting a medication like sotalol, like dofetilide, and the risk of that medication, although it's small, the risk is sudden death. So starting a 21-year-old on with reentrant flutter for a medication that can cause sudden death, for a rhythm that doesn't cause sudden death, really, you have to weigh up carefully with the family the pros and cons of that kind of approach. And it's very often that presented with lifelong suppressive medication as an option that has that risk of malignant life threatening arrhythmias, very often they would lean towards ablations so they can avoid that in the long term.

There's one more thing about the ablation strategies. Our approach here as a group has been to do preemptive ablation at the time of ablation for a particular clinical flutter. So the patient would come into the lab with a flutter, or would induce the flutter, confirm that it's the flutter that's been causing trouble, that they've needed cardio versions for, that's affected their ventricular function or symptoms. Get rid of that flutter. And then carefully map the rest of the atrium or the atria to look for other areas that can potentially form flutter in the future, sort of a prophylactic strategy, so that we don't have to come back a few years from now and go over that.

And so taking out of the circuit, taking out sort of the electrical milieu as it were, substrate which really can be troublesome in the future. So a preemptive strategy-- and it does-- it just makes the ablations just that much longer. But when you tell patients it's rather to have one long ablation then to come back for several successive ablations.

BRYAN CANNON: And one of the things I'd like to add is, is that typically, we think of atrial arrhythmia as is causing symptoms, and ventricular arrhythmias as causing sudden death. But in some of the congenital heart disease patients, it may actually be that the atrial arrhythmias are associated with sudden death. So we can't say atrial arrhythmias, eh, we don't have to worry about them. We don't have to be that concerned about them. Just because they don't cause problems.

But actually in the congenital heart disease, as Chris mentioned, because you can conduct rapidly to the ventricles, because these tend to be slower, they're already tend to be problems with function, they actually can result in sudden death. So that's why we have to be vigilant for both atrial and ventricular arrhythmias. And either aggressively treat them with medical therapy and or ablation there.

ALEXANDER EGBE: Thank you very much for that exhaustive review of atrial [INAUDIBLE] management. So now we're going to shift gears a little bit now and talk about ventricle arrhythmias. Dr. McLeod, can you discuss the different strategies for ventricular arrhythmia and congenital heart disease?

CHRISTOPHER MCLEOD: It's clearly a very, very important one. And Bryan is alluding to that the atrial arrhythmias can be malignant, which is a real thing in this population. But still, probably the most common cause of death is going to be sudden death.

Most of that is going to be from ventricular arrhythmias, ventricular tachycardia. We'd really break them up into two categories. It's going to be the one which is scar related, very similar to the atrial flutters or atrial tachycardias we've been discussing. The ventricle has been opened. These are scar related re-entry patterns.

And the treatment for that is very similar in some ways. You're going to use a class three anti-arrhythmic drug in some ways to prevent that. And you're also going to consider ablation. Because these are often discrete isthmuses. And it's one of the cases for ventricular tachycardia that is potentially curable.

And this is distinctly different from the ischemic cardiomyopathy patient, from the dilated cardiomyopathy patient, where they have progression, relentless progression of disease over time. And so where a catheter ablation in that group is to reduce shocks. And even though it may reduce shocks in adults with congenital heart disease repaired ventricular disease, it also can cure that patient of that substrate. It's not necessarily a reason not to put in a defibrillator. And often can be a very difficult decision in a young patient, where you've been able to get rid of the VT.

Do they still need a defibrillator or not? That kind of decision, I think needs to be taken carefully by the adult congenital specialist, the electrophysiologist, the family, the patient. And then that particular scar related reentry, still the defibrillators as therapy for ventricular tachycardia is important because it's such a big risk factor, or big cause for sudden death or mortality at full stop in patients, in adults with congenital heart disease. The other type of VT is a little different. The other type of VT is sort of the big, bad, right ventricle that's dilated, that's failing, or the left ventricle for the left side lesion that's failing.

Here, we don't have a discrete scar related reentrant pattern to just go and ablate. The right ventricle, which was not built to pump to the systemic circulation and CCTGA for instance, is now failing, its dilated. There's a lot of diseased substrate throughout, and we can't ablate that all. And then almost certainly defibrillators are crucial.

Risk stratification, how to identify those patients, we'll speak about in a minute. But it's a hard one. And class three anti-arrhythmics and ablation, they're going to be to avoid shocks from the defibrillator. So it's important to be seen in the center where that difference can be at least understood, dissected.

Does the patient need a defibrillator? Do they need an ablation? Do we need adjunctive medical therapy to avoid shocks from defibrillators? The fibrillator issue is a whole nother one altogether. But we'll get to that.

ALEXANDER EGBE: Thank you very. Now that you've mentioned sudden death Dr. Cannon, can you talk about sudden death risk stratification and how you determine candidacy for the different types of defibrillators?

BRYAN CANNON: I agree completely with Chris. I think this is an incredibly important issue. And the more that we're learning about congenital heart disease, the first thing that we're learning is pretty much if you make a scar on the heart with an incision to fix a congenital heart disease, at some point you're going to have arrhythmias, either atrial or ventricular. And before, we used to think that the risk of ventricular arrhythmias and sudden death was relatively low.

But as we're studying larger patient populations as they get older, we found that that's a real risk. And it's really something that you have to be concerned about. So if you have a patient that has ventricular arrhythmias, that has syncope, that has palpitations, you need to take that very, very seriously. So when we talk about the treatment for that, although medications and ablation can have some role predominantly and decrease in the arrhythmia burden, when you really talk about sudden cardiac death, the only preventative therapy for sudden cardiac death, or the best one that we have is an ICD. The problem with these is that a lot of times in patients who have structural congenital heart disease, we can't put in ICDs in the standard fashion.

Based on the American College of Cardiology guidelines for adult congenital heart disease, if you have any mixing at the atrial or ventricular level, the recommendation is to put in an ICD in a way that's not transvenus. Especially patients that have complex congenital heart disease or single ventricles, we typically, except under very special circumstances, don't put things in the standard transvenus fashion. So if we can't put things in through the veins, that puts us in a little bit of a difficult situation.

So that leaves us a couple of different possibilities. The first possibility is that we can put on an epicardial ICD. So basically what we can do is going to the pericardial sac, put the coil and the leads on the epicardial surface, and then tunnel that to the abdomen. That way there's nothing in the vasculature that would increase the risk of stroke or pulmonary embolism.

The second method that we can do that is there's entirely subcutaneous ICD that goes underneath the skin. We have to meet a specific set of criteria in order to implant those devices, as there are specific requirements in order for you to receive that device. But with these newer devices, patients who previously may not have been a candidate for an ICD, maybe a very good candidate. And we may actually be able to provide life saving therapy for ventricular arrhythmias through these devices.

CHRISTOPHER MCLEOD: One addendum to that, it really is hard to know which patients should be getting a defibrillator and who you can avoid putting one in. You really don't want to be putting in defibrillators in younger patients. As you know, just over the years, there just so many problems that can happen with the device, with infection, with the leads. And so risk stratification is really crucial. And as the audience knows, very well described for ischemic heart disease, non-ischemic heart disease, dilated cardiomyopathies, but very poorly understood for most of congenital heart disease.

The poster child of this is obviously [INAUDIBLE], real risk of sudden death from reentrant ventricular arrhythmias, from near the VSD patch, the [INAUDIBLE] tract, pulmonary valve replacement. Lots of isthmuses there that can lead to rapid VT and sudden death. So there, there are some fairly well described risk stratification factors, and also some algorithms which can be used. By and large though, when confronted with a patient with [INAUDIBLE], just as Bryan was suggesting, someone who's had a light headed spell, someone who's got some palpitations, some nonsustaining VT on a [INAUDIBLE], left ventricle is looking bad, QRS is really, really extended-- all of these are things that we really worry about in that group of patients. But it hasn't been well-characterized for most of the rest of complex congenital heart disease.

The general cardiologist should know though that there's some high risk syndromes. So certainly the de-transposition is one of the higher risk syndromes, CCTGA to some degree as well. And then beyond that, we don't have really any good tools or risk stratification tools if someone, for instance, has a single ventricle that's not being studied.

BRYAN CANNON: And I think if you take a look at those, it's very complex. And there are all kinds of factors that play a role in deciding to put an ICD in. But I think that if you take a look across congenital heart disease, the main factor, just as in adults, is if you have poor systemic ventricular function. Patients who have poor systemic ventricular function are at a higher risk for sudden death and arrhythmias. And those are patients you have to very seriously consider putting in ICDs earlier.

We don't like to put these devices in, because similar to as all patients with congenital heart disease will develop arrhythmias, all ICDs that we put in at some point will fail. The leads fail. They can be difficult placement. It can be difficult to maintain them. The batteries wear out.

There can be problems with recalls of the leads in the devices. So we take this decision very seriously. But I think that we have to look at these patients in a broader spectrum and say OK, if we have known arrhythmias, if you have poor ventricular function, if you have symptomatic palpitations or syncope, those are patients that you really need to strongly consider upgrading your therapy and considering an ICD in those patients.

But like Chris said, it's a difficult decision. So it's something you want to have-- somebody who has a lot of expertise, current knowledge of literature because this is a moving target that changes frequently. And also somebody who has the expertise who can actually put it in once you actually make that decision.

ALEXANDER EGBE: Dr. McCleod can you talk about Maze and the role of preemptive [INAUDIBLE] for ventricle arrhythmias?

CHRISTOPHER MCLEOD: It's an important one for the cardiologist who is sending these patients to the operating room. Very often, atrial fibrillation, in this group of patients is not the same as the normal heart acquired heart disease. And you may not need to isolate the veins. You really need to have a discussion with your surgeon.

You should be an expert in anti-arrhythmia surgery. Very often, it's just an isolated right sided Maze that is necessary. But that needs to be looked at carefully. There's also very good evidence that cryoablation of the outflow tract at the time of pulmonary valve replacement for tetralogy of flow is not pro-arrhythmic, and more than likely prevents VT in the future. Two important things.

ALEXANDER EGBE: Thank you Dr. McLeod and Dr. Cannon for this very important insight. And thank you for joining us on the heart.org on Medscape.