CHRISTOPHER MCLEOD: I'm Chris McLeod. I'm one of the cardiac electrophysiologists at Mayo Clinic. And I specialize in a variety of heart rhythm management and disorders with the focus being on ventricular arrhythmias but also on adults who develop arrhythmias and have a background history of congenital heart disease.

Even though I'm an electrophysiologist, I'm some also boarded in adult congenital heart disease. And I work closely with our team across the enterprise involving cardiac surgeons, adult congenital cardiologists, radiologists, interventional cardiologists, really a combined team to approach these arrhythmias in this complex group of patients in a more cohesive, complete way.

As one of the team members for our group that cares for adults with congenital heart disease, there are some really important differences, some nuances in the management of these patients, that really require a comprehensive team to look at why the patients are developing arrhythmias and how best to treat them.

As the audience probably knows, these patients are becoming more and more common in their practice because they're surviving to adulthood just because of the successes with cardiovascular surgery, cardiovascular care, arrhythmia management. But it is such a common presentation. Arrhythmias in congenital heart in adults with congenital heart disease is the most common reason why these patients will come to the hospital. And it not only signals that there are potentially scar-related arrhythmias from the prior surgeries, but it can very often herald the deterioration and the hemodynamics of that patient's cardiovascular repair or cardiovascular syndrome.

The cardiologists and the electrophysiologists that see these patients in practice-- we like to be thought of as a resource for those clinicians to call on for these complex patients. We're able to discuss these issues at a multidisciplinary session involving surgeons, interventionists, pediatric cardiologists, adult electrophysiologists, to come together with a cohesive plan, whether that's addressed on our Mayo Clinic campus or whether it's even addressed locally.

And there are some very important key differences in managing these patients. And I just think for the cardiologists and for the electrophysiologists that see these patients, it's important to go over a couple of things.

The primary issue here is the differences in recognizing arrhythmia. For the patient with congenital heart disease who are developing arrhythmias, it's going to be atrial arrhythmias or ventricular arrhythmias. And I think we just start off with the atrial arrhythmias to begin with. These are by far the more common arrhythmia in clinical practice for adults with congenital heart disease. And they can present very abruptly with an acute deterioration. And that very often is not missed. It's the more subtle, gradual presentation of worsening heart failure driven by these slow atrial tachycardias that result in these patients presenting with a deterioration in their exercise capacity. Their ventricular function has gone down. They're just not doing well. And you look at the ECG, and their heart rate is around 100, 105 beats per minute.

So the common mistake here is to assume that there's a sinus tachycardia related to poor hemodynamics, which it certainly could be, but to first of all exclude that this is not a re-entrant scar-related atrial flutter, or intra-atrial re-entrant tachycardia. These are the most common atrial arrhythmias. And if you follow all patients with congenital heart disease, as they get older at least 50, 60% of the world with congenital heart disease are going to develop one of these at some time in their life. And that atrial arrhythmia presents differently from how atrial fibrillation presents in the adult.

The scar-related atrial flutters typically have a much slower cycle length. The P waves are different. The ECG looks different. The presentation is really different. And so that would be the first thing, is just in these very common atrial arrhythmias to have your ears pricked up. Try not to miss. Think of all the time that you could be dealing with a patient who has a very slow atrial flutter that's mimicking sinus tachycardia.

As far as managing after you've identified one of these reentrant atrial flutters, a these scar-related arrhythmias, management is distinctly different. We don't use the class 1C medications that are standard practice for paroxysmal atrial arrhythmias in the young patient. These very often will slow down the flooding and make it more resistant, recalcitrant, and even conduct to the ventricle even quicker. And so they're dangerous. They're not suitable for this group of patients. And the Class III drugs or ablation is standard here. And if you are going to go ahead with intervention or ablation in your practice, very often to run this by a radiologist who specializes in congenital heart disease, as you may have to cross baffles and change your approach based on the anatomy.

So those are the important management differences for the atrial arrhythmia itself. But probably the most difficult element in managing this common arrhythmia is going to be what do we do with anticoagulation. What do we do with blood thinning? Is the risk of stroke from thromboembolic disease in a young patient with congenital heart disease with atrial flutter, the same as their counterpart with a normal heart. By and large it isn't. By and large the left atria are normal.

By and large the thromboembolic risk is substantially different. This really is an important one because adults with congenital heart disease have around a 10- to 100-fold time higher incidence of stroke than patients with acquired heart disease or even normal hearts. So it's a really big issue for them. But the patterns and the predictors do not follow the same algorithms. They haven't been enrolled in the same trials. We can't use the same data. We can't use the same risk stratification. And so it really is a difficult one. That's definitely something that we struggle with on a daily basis here. That's often something that we discuss as a group.

So those would be the three things in dealing with those most common atrial arrhythmias. It's recognition of the slow scar-related atrial flutter. It's the salient differences in management avoiding the class I drugs, using the class III drugs or ablation. And then the most difficult is what do we do about blood thinners for this group of patients who we know has a high incidence of stroke.

Let's move on then to device management in these patients. So for the cardiologists and an electrophysiologist to see these patients, saying once again we're just happy to act as a resource for these difficult questions. The questions that come up is how to pace these patients. Should it be transvenous? Should it be epicardial? What do I do about anticoagulation? Should we place a lead in a Fontan circulation? These are all difficult questions, which just as a center which does a lot of this, we commonly discuss this as a group at a multidisciplinary conference where we come up with what we think is a unified approach because there is not historically a lot of patients here to refer to in the trials.

There are some very important elements, though, that I'm going to identify for the clinician. And these would be as follows. The first would be know the anatomy. But because based on prior surgery, you may not have access to the chamber that you're targeting with your lead. The second would be is there any shunting at an intracardiac level. If there's any shunting at an intracardiac level, clots that form, thrombus that form on the leads, are prone to paradoxical embolus going across into the systemic circulation and resulting in stroke. So two very important I guess no-brainer things, but you really do have to think of them every time you go and see one of these patients. Can I get to that chamber? And should it be epicardial or should it be transvenous?

Once access is considered, once the anatomy is considered, is it even appropriate for that patient to get a defibrillator? A very difficult question for a wide spectrum of these disorders-- we know that still one of the most common reasons why these patients die is sudden death. Sudden death still drives the mortality in the majority of the complex congenital heart disease syndromes. But who gets a defibrillator and who doesn't? Does dysfunction in the right ventricle mean the same as dysfunction in the left ventricle or the systemic ventricle? They're very difficult questions. And they aren't specific trials that address this with big numbers where we have a concrete yes or no answer like we do in patients with acquired heart disease.

So another reason why I'm suggesting we're happy to act as a resource even to bounce those difficult questions off. But I certainly would take any kind of event really seriously in someone with repaired congenital heart disease, especially if the repair is in the ventricle or especially if there's any kind of ventricular dysfunction if they have an event. So some kind of unexplained, lightheaded spell or syncope we take very seriously-- aggressive monitoring until we show we're not dealing with a malignant life-threatening ventricular arrhythmia.

Which device as far as defibrillators is definitely a difficult one? Do we use a single coil with a different anatomy? Is a dual coil better? Should we be using more subcutaneous ICDs? These are all difficult questions, which we'd be happy to provide consultative service even if it's just a phone call or an email to bounce that off.

As far as other managements of these life-threatening arrhythmias, ventricular tachycardia ablation in this group of patients is one of the only ablations that we can sometimes do that is curative for ventricular arrhythmias. This is distinctly different from ischemic cardiomyopathies, dilated cardiomyopathies, whereby the substrate is still probably evolving in those patients, where we have young patients with relatively normal or preservation of ventricular function, and we have discrete scar-related ventricular arrhythmias.

Ablation across isthmuses that are serving to support that ventricular tachycardia can be curative. That's data that we have found in our cohort, also in Boston and also in the Netherlands. So that kind of ablation probably should be done at a complex ablation center where a lot of adult congenital ablation is done because the goal for some of those patients is to avoid ICD implantation even though they have a ventricular arrhythmia. If you cannot get rid of the arrhythmia, then it makes sense to consider ICD-- not ideal in a young patient, so ablation very high up there on our priority list when dealing with these malignant VTs in a young group of patients.

Regardless of the specifics of the atrial arrhythmias and the ventricular arrhythmias in the adult with congenital heart disease, with the presentation being different and the management different, I distinctly want to highlight for the cardiologists and the electrophysiologist the importance of having that patient reviewed comprehensively at a center that does good hemodynamic assessment of their patients. So often we've seen the patient with scar-related atrial arrhythmias or recalcitrant ventricular arrhythmias. And there's a failing pulmonary valve. Or there is a failing hemodynamic issue. And so we really cannot treat these patients in isolation with just ablating atrial arrhythmia or the ventricular arrhythmia. We really need to have them seen in a complex congenital center.

As the audience probably knows, there are only a few of these around the country. And we are more than happy to partner with the local cardiologist, with the local electrophysiologist, to go through that assessment to make sure that nothing is being missed when the patient is presenting with an arrhythmia, and then also partnering with them for comprehensive continued care as they need more following down the road.

As the audience probably knows, these are part of the American Heart Association HRS/ACC guidelines. These patients do need to be seen in a complex comprehensive center where all of these disciplines are available from genetics to surgery to electrophysiology to hepatology. It really is an important comprehensive assessment that needs to be done and needs to be a partnering process with the local cardiologists or EP.

So as the cardiologist, electrophysiologist, or the clinician knows, that each one of these congenital heart syndromes is not the same, that one Tetralogy of Fallot is not the same as another Tetralogy of Fallot, and the same within atrial septal defect, same with any of these defects. And so an individualized, tailored approach is essential for each one of these patients.

And so I'm making the case that in partnering with the local clinician, we really want to provide a comprehensive approach to this. The Fontan who's developing cirrhosis and liver failure needs to see a hepatologist that sees a lot of Fontan patients. And the Eisenmenger patients with pulmonary hypertension need to see pulmonary hypertension specialists that see a lot of Eisenmenger patients. These are distinct entities-- different from the primary pulmonary hypertension, different from the other forms of cirrhosis. And that goes across the board for genomics. It goes across the board for electrophysiology, for the overall cardiovascular management of these patients. And that comprehensive approach is key also in keeping with the guidelines, but really key for these patients, making sure that every single one of these elements is addressed.

One other important consideration for the clinician who sees these patients-- if your patient is really interested in moving forward our understanding and our care and our management of these patients, we try to enroll all of these patients in a database so that we can reflect on this in the next five years and say, "You know what? Our 500 patients with Tetralogy of Fallot-- this particular management strategy is most appropriate.

And so for the patient who is interested in that, for the clinician who wants to be involved in moving forward this process, that kind of research, clinical study, is what we specialize in across our Mayo enterprise. It's key for this kind of patient-- this adult congenital patient where there are not a lot of them. We need those numbers to improve research and improve our understanding and improve care.

In addition to that, I do feel, though, that we're at the cutting edge of understanding what's novel, what's appropriate, what we shouldn't be doing for these patients. And so I'd like to say that our team is up to date on all of those approaches. In addition, if you have a group of interventionalists, electrophysiologists, surgeons that do this on a regular basis, then the complex patient is done in a methodical, careful way by a team that's a well-oiled machine used to dealing with these patients rather than seeing these patients once every year or once every two years.

I'm proud to say that all of our centers do a really good job of taking our time in a comprehensive way, looking carefully at their arrhythmia and looking making sure that nothing is being missed, making sure that all the other bases are covered from hemodynamics to genomics, making sure then that the management strategy is the most current and up to date.