BroadcastMed | Brugada Syndrome vs Pattern: What You Should Know

RAHJIV GULATI: Hello, I'm Rahjiv Gulati from the Division of Cardiovascular Diseases, Mayo Clinic, Rochester, Minnesota. Joined today by my colleague, Dr. Suraj Kapa, who is also a cardiologist, but who specializes in electrophysiology. And we're going to talk today about Brugada syndrome.

Saraj, welcome.

RAHJIV GULATI: Thank you, Dr. Gulati.

RAHJIV GULATI: Perhaps you can start by telling us, what is Brugada syndrome?

SURAJ KAPA: So that's a great question. And one I think that is fairly commonly misunderstood. It's important for people to understand that Brugada has been recognized for decades now as a syndrome. And what it is is an abnormality in the ion channels of the heart, which results in abnormalities in how the normal heart cells activate. Which, in turn, can result in patients suddenly dying.

Such that they can just be walking down the street, and experience abrupt-onset syncope, or experience actual sudden death, which they don't wake up from, unless there is somebody around to resuscitate them.

It's estimated that about 0.1% to 2.6% of the population carry the genetic characteristics that may put them at risk for having Brugada.

But it's important to discern between a Brugada pattern and the Brugada syndrome. Because many patients might have a electrocardiogram that shows Brugada, but does not necessarily mean they have the syndrome, or at risk of sudden death. Though all of that may have these genetic abnormalities that cause the problems in the ion channels.

And when we look at Brugada syndrome, we come to understand that it's a much more multifaceted disease than just saying it's a genetic mutation, or it's an abnormal electrocardiogram, but really putting all of those factors together into the diagnosis.

RAHJIV GULATI: Very interesting. Thank you. So me, as a non-EP guy, we worry about missing the Brugada pattern on the ECG, but also worry about overcalling it sometimes. Maybe you could elaborate on some of the challenges of making the diagnosis.

SURAJ KAPA: So that's a wonderful question, as well. The truth is that Brugada can be extraordinarily challenging from both estimations. There are other syndromes and other diseases that can look like a Brugada pattern on an electrocardiogram.

When we talk about the Brugada pattern, we talk about abnormalities in V1 and V2 on the electrocardiogram. The two precordial leads. The reason for that is when we look at the heart, and where Brugada affects the heart, it's principally around the right ventricle, along the right ventricular outflow tract, where we see the abnormality in terms of the substrate of the heart.

And that, in turn, creates abnormalities in those two precordial leads that are looking at the anterior surface of the heart. Once we look at that, we realize that there are other types of diseases that can show similar abnormalities. Arrhythmogenic right ventricular dysplasia can look very similar, and even a typical right bundle branch block can be mistaken for that.

So you can see a baseline electrocardiogram, which is suggestive of the abnormality, but actually isn't Brugada. The flip side though, as you were alluding to, is also underdiagnosis. And the problem there is not every single electrocardiogram from the same person will necessarily reveal that Brugada pattern.

And thus, we need to be cautious to review all electrocardiograms, especially under conditions of stress. Because sometimes, specific stressors, fevers, use of tricyclic antidepressants, anesthesia exposure, surgery, can result in revealing the Brugada pattern on an electrocardiogram that would not otherwise be present at baseline.

And thus it's important to consider patients who may come in with nondescript symptoms, syncope, otherwise normal electrocardiogram, or seizures even. because that can also be secondary to ventricular arrhythmias, where you lack appropriate blood flow to the brain.

And then, even though they have a normal electrocardiogram, maybe do provocative maneuvers in order to bring it out.

SURAJ KAPA: That's really informative. Thank you. So just let's work through this. And so say I see an ECG in a convincing story from a patient, and there is a concern for the Brugada pattern. Walk us through what you do next. How do we provoke and risk stratify who needs invasive treatment. Perhaps you can help with that.

SURAJ KAPA: OK. So there's a couple of things we need to think about. Say I get a patient in clinic, walking in. Their physician was concerned. They saw the Brugada pattern on the electrocardiogram.

I talk to them. The first key point in my mind is to say, OK, does this person have symptoms or not? By symptoms, I mean, have they had episodes where they've suddenly passed out just walking down the street, or doing something that would not otherwise be associated with them having any issues with feeling faint.

Are they having seizures or other nondescript symptoms that might be suggestive of an abnormal ventricular rhythm?

Once I establish that they have symptoms, and I look at their electrocardiogram, and it is very suggestive of a Brugada pattern, a lot of my trip is done at that point. Because what we found is those patients who have symptoms and the Brugada pattern are quite high risk for further arrhythmic events.

And you can put one and one together to make two, which would be the ICD. Without doing further strategies, such as EP studies or echocardiograms.

The difficulty becomes in those patients who have maybe a suggestive pattern on the electrocardiogram of Brugada, but it's not classically that. Or the patients who have the classic pattern, but don't have the symptoms.

And that's where really the problem right arises. In the patients who have symptoms, but the Brugada pattern is suggestive, but not quite there, we can take them to the electrophysiology laboratory, and provide them sodium channel blockers, such as Flecainide, or Procainamide challenge in order to try and elicit the Brugada pattern.

But even before that, one thing that physicians commonly forget, is if you just move V1 and V2 one interspace higher, you can actually elicit a classic Brugada pattern that goes along with actually having the syndrome at that point.

The other side of it is, you can actually turn around and say, well, this patient is asymptomatic. I can't elicit a clear history of anything that would suggest them ever having had an ventricular arrhythmia. And then we're in a little bit of a pickle, because what do we do with them? Because, frankly, we don't want them to have their first incident being walking down the street, and suddenly dying, and not having anybody around to resuscitate them.

So a lot of people over the last 20 years have looked into the role of electrophysiology studies where we try to provoke the ventricular arrhythmias in the EP lab. And while one large group has suggested that, yes, if you can provoke the arrhythmia during an EP study, then this is Brugada syndrome. You put in an ICD.

Unfortunately, we've seen exactly the opposite in other large scale studies, where from other large groups who have said it doesn't matter.

RAHJIV GULATI: And we're talking asymptomatic patients here.

SURAJ KAPA: Exactly. Never had any symptoms. Never had anything suggestive of a ventricular arrhythmia in the past.

RAHJIV GULATI: So it's an area that is not resolved yet. Do we here at Mayo have a particular belief in one version, which will be EP testing, plus or minus IDT versus a conservative, watchful, waiting strategy. Do you have any personal thoughts on that matter?

SURAJ KAPA: So there's several factors that go into it. One of the things is genetic analysis. We do recognize that there are certain genetic mutations. The most common genetic mutation is in the SCN5A sodium channel that might be associated with higher risk of an arrhythmic event in population studies. And maybe that could help.

Unfortunately, the lack of a positive genetic test does not necessarily mean that they don't have a genetic mutation.

RAHJIV GULATI: Right.

SURAJ KAPA: And that's the difficulty. And also, a genetic mutation, and this is research we've published out of here on multiple different subjects, does not mean that that is a pathogenic mutation, or the mutation causing the problem.

There is such thing called background noise. So the problem becomes it can help but it's not perfect. The next step is-- and this gets back to everything in medicine-- talk to the patients. Talk to them about what the potential risks are. Some might say, look, doc, this is the biggest fear in my life. I want everything done to reassure me as much as possible. And you might say, OK, let's do an electrophysiology study. If we don't provoke anything, all the data is suggestive that that is a very good suggestive--

RAHJIV GULATI: It's a low risk.

SURAJ KAPA: --negative prediction of you having a arrhythmic event in the future. But the problem is what to do with the positive electrophysiology study at that point.

The other thing we always talk about is talking to the patient about how can we monitor you? Maybe that weird symptom you had, that feeling of lightheadedness or palpitations, that you otherwise can't describe might have been suggestive of ventricular arrhythmia.

And one option that's available to us today is implantable loop recorders, where we can actually place a very tiny device right underneath the skin, that can record for up to three years.

And not only for symptoms. We'll have preset criteria to identify ventricular arrhythmias.

RAHJIV GULATI: Fascinating. Thanks. You're identifying the low risk seems like a nice option, and can be reassuring. But trying to figure out those who are at high risk remains a challenge for us.

SURAJ KAPA: True. Exactly.

RAHJIV GULATI: So, again, as a non-EP guy here, you mentioned that the problem is local. It's confined to the region in the right side of the heart. Well, just ablate that area.

SURAJ KAPA: So that's a wonderful question, as well. So this is only something that's recently been come to be understood. When we look at the Brugada syndrome, people have looked at what the pathology is of the heart in these patients. Why is the electrocardiogram so localized in that area?

Those original pathology studies demonstrated that there is some fatty changes within the epicardial surface of the anterior aspect of the right ventricular outflow tracts. And this led to many people saying, hey, exactly, if this is so localized, why can't we take care of it by just addressing the substrate? Say somebody is having recurrent ICD shocks for ventricular arrhythmias. Or say somebody is having recurrent symptomatic VT, though not necessarily ultimately resulting in shock. Can we do something about this?

And the truth is, at least a couple of investigators have demonstrated that by going in and performing what we call an ablation, where they burn throughout the area of abnormal substrate, particularly focused on the epicardial surface, that they can not only prevent future ICD shocks, but in fact the Brugada pattern disappears on the electrocardiogram, and is no longer provocable.

However, of course, these are in small numbers of patients, and larger scale studies are necessary in order to understand what we're actually seeing.

RAHJIV GULATI: So it's fascinating. So ablation hasn't yet replaced-- we're not confident enough to say, well, we can maybe replace the ICD, it will be as an adjunctive thing for certain subsets?

SURAJ KAPA: Exactly, and I think that's an important point. Because the fact is, for these patients, the ablation might work. But say the ablation doesn't. Then they may still have a ventricular arrhythmia that may lead to their sudden death.

And thus, in those high-risk patients, those patients who are judged to have sufficient risk to merit being concerned about the ventricular arrhythmia burden and the risk of sudden death, you still need to put in the ICD.

RAHJIV GULATI: Fascinating. So what else is on the horizon for Brugada?

SURAJ KAPA: So one of the things that's on the horizon is how do we treat these patients with the defibrillator? Nowadays, we have subcu ICDs.

One of the reasons that we're having all of this rigmarole about what do we do? Do we put the ICD in or not? Is because putting in a transvenous ICD is not a simple thing that is not with any sort of risk. These are young patients.

When we look at that epidemiology we were talking about earlier. If you go to South Asia, for instance, it's estimated that 50% of all unexplained sudden deaths in structurally normal hearts, in patients less than 50, are due to Brugada syndrome.

And similarly, in European populations, it may be as high as anywhere from 10% to 30%. So if you think about that, you don't want to miss those.

The problem though is these are young people. If you put a transvenous ICD, where you have a ICD lead going through the venous system and into the heart itself, these leads scar in place over time. If they ever have an infection 20 years down the road, it's not a trivial process of taking this wire out.

And there's risks of inappropriate shocks in patients who can get their heart rate well above 200, because they're otherwise healthy. So you want to recognize that the problem of putting an ICD willy-nilly in patients who don't need it necessarily, and will never use it.

The subcutaneous ICD may change that to some extent, because the truth is that it's not endovascular. So their risk of scarring in place isn't there inside the heart, and thus removing it can be much more simple.

The negative being that you're still at risk potentially of inappropriate shocks. And not everybody might be a candidate for subcutaneous ICD. Thus, you still need to think about that risk stratification, and we need to start thinking about better ways of understanding how to risk stratify.

Some of the things we're looking at include can we understand the substrate better in advance of putting in ICD? Say the electrophysiology study is negative. Say they have clear MRI abnormality in that right ventricular outflow tract. Is the substrate enough to push us one way or the other?

Are there other provocative maneuvers that might help us? Should all of these patients be getting loop recorder? These are things we need to understand better, and this is really what's on the horizon, to follow these patients.

RAHJIV GULATI: Well, that's fascinating. Well, I have learned an awful lot, as always, from you, Suraj. I appreciate you sharing your knowledge.

Thanks to everyone for joining us. It's been a real pleasure for me. And I hope it's helped some of you understand this fascinating condition.