DR LORI BLAUWET: Hello, my name is Dr. Lori Blauwet, I am a cardiologist at the Mayo Clinic and currently serve as the director of the Cardiac Sarcoidosis Clinic.

DR SURAJ KAPA: And my name is Dr. Suraj Kapa, a cardiac electrophysiologist at the Mayo Clinic with a special interest in cardiac sarcoid.

DR LORI BLAUWET: Together, we would like to share some information with you about sarcoidosis in general and, in particular, about sarcoidosis in the heart. Sarcoidosis is a rare disease. It most commonly affects the lungs, but it can affect any organ in the body, including the brain, the nerves, the eyes, the skin, and of course, the heart.

No one knows what causes sarcoidosis. Many of us who are experts in this disease think that sarcoidosis is caused by an interplay between some sort of infectious, environmental, or occupational exposure, and genetic predisposition.

Some potential triggers associated with the development of sarcoidosis include bacteria, and viruses, chemicals, such as insecticides and pesticides, environmental exposures, such as mold and occupations, such as working with metal or as a firefighter.

Sarcoidosis usually affects young adults, particularly people between the ages of 30 to 50, although sarcoidosis can also affect children and older adults as well. Sarcoidosis sometimes, but not always, runs in families. In general, sarcoidosis occurs more often in women than in men but, interestingly, cardiac sarcoidosis occurs more often in men than women, for reasons which remain unclear but are under investigation.

Most people who have sarcoidosis in their hearts have sarcoidosis in other organs. It is particularly difficult to diagnose cardiac sarcoidosis when it is present only in the heart. Sarcoidosis is caused by a build-up of a certain type of cell called granuloma. These granulomas can grow in either small or large clusters anywhere in the body, including in the heart. When the granulomas grow in the heart, they may occur in any part of the heart, including the heart muscle, the heart valves, or the sac around the heart called the pericardium.

The granulomas are not evenly distributed throughout the heart because they grow in clusters. So we call this a patchy distribution of granulomas throughout the heart.

DR SURAJ KAPA: The symptoms associated with cardiac sarcoidosis can be highly variable. Many patients will simply present with a vague sense of tiredness, shortness of breath when they exert themselves, perhaps some symptoms of heart failure such as fluid overload. Other individuals can be found to have either a very slow heart rate, may have episodes of passing out, which can occur either because of a slow heart rate or a heart rate that's too fast, or actually develop symptoms of extra heartbeats, or so-called palpitations.

The problem is with cardiac sarcoidosis that, in fact, the vast majority of patients who have cardiac involvement, especially in the early stages, can even be asymptomatic. The question is, how long it takes for the disease to progress to such an extent that it involves the heart enough to result in symptoms attributable to the heart.

As a result, because of this highly variable symptomatology associated with cardiac sarcoidosis, it requires some degree of suspicion of the possibility of the disease, to look for it. And this gets into the complexity of both how to diagnose it, whether by imaging or labs, which I will go on to Dr. Blauwet who will explain how we try to go about this.

DR LORI BLAUWET: Sarcoidosis can be very difficult to diagnose. And sarcoidosis in the heart can be particularly difficult to diagnose. Some laboratory tests may be helpful, including angiotensin converting enzyme, or ACE, or inflammatory markers. If these test results are positive, this may support a diagnosis of sarcoidosis. If the test results are negative, however, that does not rule out the possibility that a patient might have cardiac sarcoidosis.

Imaging tests are very helpful in terms of diagnosing cardiac sarcoidosis. A chest X-ray can reveal abnormalities in the lung, which are common in patients with sarcoidosis, but also can show the size and shape of the heart.

An echocardiogram, or cardiac ultrasound, is very helpful for showing the size of the heart chambers, how well the heart pump functions, and how well the heart valves work.

A cardiac MRI can also be very helpful in terms of describing the size and function of the heart. A cardiac MRI is particularly helpful, however, in demonstrating areas where there may be scarring in the heart due to the deposition of granulomas.

A cardiac PET scan is also very helpful in showing where there may be areas of scarring in the heart. In addition, a PET scan can show where there may be areas of inflammation in the heart. This information is very important because it helps us to direct our treatment, particularly if we're trying to decide whether or not immunosuppressive medications are warranted for treating a particular patient.

In our institution, the PET scan is combined with a CT scan, and we scan the body from the top of the eyes down to the thighs so that we can look for evidence of inflammation anywhere in this area. As I mentioned previously, sarcoidosis can affect any organ in the body. Being able to view all of these organs, in addition to the heart, helps us to determine the extent of the involvement of sarcoidosis and can also help diagnosis in terms of targeting areas that may be helpful for obtaining a biopsy, which can, under the microscope, potentially show evidence of granulomas which is diagnostic for this disease.

DR SURAJ KAPA: The complexity of understanding sarcoidosis, as Dr. Blauwet was alluding to, lies in the number of ways in which it can manifest. She talked about the structural manifestations, things you see on echocardiography or ultrasound of the heart, MRI in terms of looking for scarring, but now we're going to segue into the electrical elements.

As we explained earlier, the symptoms associated with sarcoidosis might be highly variable. And some of these may be associated with changes in the heart rate or the heart rhythm. The interesting thing is, historically, the majority of patients at the time they're diagnosed as having possible, probable, or definite cardiac sarcoidosis present with abnormal electrical findings.

As a result of this, a close collaboration is important between the electricians, or electrophysiologists, as well as the clinical cardiologists, a.k.a. The expert diagnosticians. One of the preliminary tests we give in any evaluation on the electrical elements of cardiac sarcoidosis, is something called an ECG, or electrocardiogram. The electrocardiogram gives us information about how the heart is activating. Specifically, it can tell us do the chambers of the heart talk to one another consistently. How is the heart activating? Are there blocks in any of the normal electrical highways that we sometimes see in cardiac sarcoidosis?

The problem with an ECG, however, it only shows us about a 10 second moment in time. And sometimes, if somebody is having symptoms such as what we call palpitations, or feeling of skipped heartbeats, or fast heartbeats, or slow heart rhythms a single ECG may not be enough.

So then we get into more prolonged monitoring and there are a number of different options for this but one of the primary tests we obtain in our cardiac sarcoidosis practice is at least a 24-hour Holter monitor.

The Holter monitor is a monitor that you wear that has patches attached to the chest, similar to an electrocardiogram, and we look for several things on this Holter monitor.

Number one, we look for, over the course of 24 hours, are there any episodes where the heart rhythm suddenly becomes too slow, where the activity of the heart speaking to itself doesn't happen. We also look for rhythms that might be too fast, rhythms that may be occurring in the upper chamber, such as atrial fibrillation, which is commonly seen in patients with cardiac sarcoidosis, or perhaps more ventricular arrhythmias, or ventricular tachycardia. The ventricles are the bottom chamber of the heart and when they start going fast on all their lonesome and not in relationship to the upper chambers, this can sometimes be associated people getting dizzy, actually passing out, or in some cases, sudden death. And thus, this in particular is something we really want to identify.

The last thing we look for on the Holter monitor is evidence of extra beats, or so-called PVCs, or premature ventricular contractions. Depending on where the patching involvement occurs in the heart, we might identify that there might be extra heartbeats that might be resulting from the active inflammation or the scar that's left behind.

We are going to be segueing later on a little bit into prognosis and how to deal with the possibility of risk in many patients with cardiac sarcoidosis. Well, one of the diagnostic elements that underlies this to see how the electrical system of the heart might or may not be involved in a specific patient with cardiac sarcoidosis, is a more invasive study called an electrophysiology study.

Electrophysiology studies help us understand OK, maybe the Holter looks OK. Maybe the ECG looks relatively OK. But, looking at a more close level at the heart, is there evidence of any disease in the conduction system. If we stimulate or stress the heart using our stimulation tools, can this cause us to identify problems that might happen in the future?

Finally, and this is the close alliance that we as electrophysiologist have with our colleagues in the interventional lab, we have started working together to perform something we call voltage guided biopsies. As Dr. Blauwet alluded to, the cardiac sarcoidosis can be extremely patchy. And in order to offer a definitive diagnosis of the disease, we need some sort of evidence of what we call pathology. In other words, taking a piece of tissue and being able to look at it under a microscope and say yes, this is cardiac sarcoidosis. Or sarcoidosis elsewhere with inflammation of the heart suggesting that the sarcoidosis that exists elsewhere also involves the heart.

The difficulty is honing in on exactly what is involved. Because if you take a piece of normal tissue, it's going to look normal. As a result of this, we do this procedure in some patients where they have no other targets for biopsy outside of the heart called a voltage guided biopsy, where we essentially use our tools that we use with electrophysiology to identify abnormalities in the heart, and then hone in on those as areas we should be obtaining tissue from in order to optimize our ability to diagnose.

DR LORI BLAUWET: I want to talk briefly about the relationship between coronary artery disease and cardiac sarcoidosis. Sarcoidosis in general can be a great mimicker and sarcoidosis in the heart can mimic problems associated with coronary artery disease, or blockages in the heart arteries. For that reason, in every patient in whom we suspect cardiac sarcoidosis, the incidents or probability that they have coronary artery disease, must be evaluated either by a cardiac stress test, or a coronary angiogram to look at the heart arteries directly.

Now we'll move on to talking a bit about treatment. First of all, it is important to know that there is no cure for sarcoidosis in general, and no cure for cardiac sarcoidosis.

However, there are medications and treatments that can be used to alleviate symptoms and try to prevent the progression of the disease. If a patient has active inflammation in the heart, we generally treat with immunosuppressive medications. The most common immunosuppressive medication that is used around the world to treat sarcoidosis is Prednisone, or corticosteroid.

In our practice, we do treat patients with Prednisone, but, because long-term Prednisone therapy can result in many side effects, we generally also treat patients with other immunosuppressive medications to try to reduce the duration of steroid therapy.

We generally start with oral medications or pills that can suppress the immune system. If the oral medications do not adequately suppress the immune system, then we may treat patients with intravenous medications. In our practice, we monitor the response to treatment, primarily by obtaining cereal PET scans.

As you recall, I mentioned that the PET scan is the best test to evaluate for active inflammation. If there is no inflammation in the heart, there is not necessarily any reason to continue long-term treatment with immunosuppressive medications.

As mentioned earlier, one of the complications of sarcoidosis in the heart is that the granulomas in the heart muscle can cause the heart muscle to become weak, and a patient may develop symptoms of heart failure. In this case, medications that are used in general to treat heart failure may be helpful, including beta blockers or ACE inhibitors or diuretics.

DR SURAJ KAPA: So, as we talked about earlier, the manifestations of cardiac sarcoidosis may be vague, and may involve almost any element of the heart. Dr. Baluwet talked about the heart failure elements, when the heart function is reduced. But as I alluded to earlier, we also worry about electrical elements, electrical abnormalities. In many patients who have cardiac sarcoidosis, one of the first presentations might be a rhythm called complete heart block, or heart block in general.

This finding is one in which the upper and lower chambers of the heart are atria and ventricles, which are supposed to talk to one another sequentially. Stop talking to one another in a one-to-one fashion. A lot of times, this can just occur with normal aging or because of any multitude of other factors. But cardiac sarcoidosis is one of those things that can cause this.

Many patients, when they see this lack of communication between the upper and bottom chambers of the heart, might receive a pacemaker in order to put the two sides back in sync.

However, one of the key things we worry about in cardiac sarcoidosis is not just the slow rhythms, but the dangerous fast rhythms, those that can cause a patient to suddenly pass out or, in some cases, suddenly die. And the problem is, we do not want to wait for the first episode to happen, because the first episode could be the last.

Because of that, we talked earlier about the importance of getting tests such as ECGs, Holter monitors which might show us some evidence of these admirable rhythms. But in many cases, electrophysiology studies. In electrophysiology studies as I talked about earlier, we will try to stimulate the heart out of rhythm. And if we find, based on specific criteria, that we can fairly easily stimulate the heart out of rhythm, we might identify a patient who merits an implantable cardioverter defibrillator.

Now, pretty much all cardiac defibrillators can potentially act as pacemakers. But a pacemaker is not necessarily a defibrillator. They serve very different functions. So nicety kind of covers both sides of the coin.

There's a lot of study going on in terms of who would benefit most from defibrillators and cardiac sarcoidosis. Our early guidelines suggested that just having a diagnosis was enough. More recent guidelines suggest that we look at results of the electrophysiology study. Whether there's enough involvement of the heart to cause the heart function to go lower, looking at all elements of scar on the MRI, that might suggest what we call substrate or areas of short circuit that may cause the abnormal rhythms.

Because we're doing this in many cases for primary prevention, while we hope you never have an episode, the device never has to go off, you never have an abnormal rhythm. We're trying to prevent you from potentially dying from the first one.

Now, if a patient is having abnormal rhythms, any particular type of abnormal rhythm-- less dangerous rhythms, such as atrial fibrillation, or PVCs, or premature ventricular contractions, which are early extra beats to the bottom chamber of the heart, which might be symptomatic. Or, in more severe cases, actual ventricular arrhythmias where the bottom chamber is just going off to the races on its own, potentially causing somebody repeatedly pass out or the defibrillator to have to shock them.

Then we start talking about how can we stop these abnormal rhythms from happening. And there are two avenues to that. One is using medications, such as antiarrhythmic drugs. And the decision-making on specific antiarrhythmic drugs is complex and requires close discussion with an individual with expertise in this area.

And, in some cases, we may even go on to ablation, which is a more invasive procedure where we go in to identify the areas of short circuits and try to intervene upon them by, essentially, burning or causing alteration in those cells to prevent them from acting up. And depending on the rhythm, depending on the situation, the likelihood of success may be highly variable. And thus, going to a center which has high volume and an individual with expertise in a specific type of ablation you might need is so critical.

And really this delves into prognosis. The issue is that we don't want to see you progress. We don't want to see the patient with cardiac sarcoidosis to get worse. We want to arrest or stop the progression of it when we first see them. And part of this is avoiding the potential ways it can cause complications. In the most severe manifestations, that would be death.

But, in other manifestations, it might be heart failure worsening and getting to a point where we start talking about transplants, which doctor Blauwet will talk about it in a little bit. But we also want to make sure that these rhythm abnormalities don't happen, that we have the right safety nets, the right things in place to avoid the worst possible situations that might happen from cardiac sarcoidosis.

The prognosis in cardiac sarcoidosis, unfortunately, is not overall very well-established. In some patients, it can rapidly progress. In other patients, it can have an incident episode of it and do fairly well. That's why here, we are so focused on doing studies and trying to engage in collaborative discussions and potentially trials into how to best arrest the course of this disease and avoid these worst possible outcomes and decide when certain interventions are needed which might be different from other types of heart disease.

DR LORI BLAUWET: The prognosis for patients with cardiac sarcoidosis is extremely varied. Some patients initially have inflammation, it's treated, they don't have any rhythm problems, and they can live long and healthy lives and sarcoidosis never comes back.

In other cases, no matter how aggressive we are with our treatments, the sarcoidosis continues to be active and cause a multitude of problems. In general, sarcoidosis tends to be slower or more smoldering disease than some other types of heart disease, but not always.

In cases where patients have active sarcoidosis and the sarcoidosis is treated, the patients get better, the patients aren't necessarily cured as I said earlier because the sarcoidosis can come back or become more active. For this reason, patients with sarcoidosis, particularly cardiac sarcoidosis, require ongoing serial monitoring to assess for progression, or regression, or recurrence of the disease.

We generally provide serial monitoring with clinical examinations, imaging tests, and heart rhythm tests as needed depending on that particular patient. Sarcoidosis prognosis also depends on the extent of sarcoidosis throughout the entire body. In some patients, sarcoidosis only settles in the heart. But, as I said earlier, in other patients, sarcoidosis can be in many organs in the body. For that reason, we, in our Cardiac Sarcoidosis Clinic, work very closely with our colleagues in other clinics here at the Mayo Clinic, including the ophthalmologist, the rheumatologist, the pulmonologist, the gastroenterologist, the nephrologists, and neurologists. So I think it is this multi-disciplinary environment that we are able to engage in here that can help us to optimally provide care and recommendations for patients with sarcoidosis.

In some instances, despite our best efforts, a patient with cardiac sarcoidosis may experience either severe heart pump failure or severe heart rhythms that cannot be controlled, even with our best practices. In these cases, patients may require implantation of a heart pump or even a heart transplant.

DR SURAJ KAPA: It has been an amazing opportunity to be able to talk to everybody about cardiac sarcoidosis. And the reality is here at the Mayo Clinic, we realize the complexity of care associated with this. This is truly an individualized, personalized practice we have to consider.

As a result of that, Dr. Blauwet, myself, and others have been engaged in trying to understand how we can better look at the individual patient, not just as an individual, but amongst other patients similar to them. So we can both learn about how to better take care of the individual, as well as to better inform them of what they should go on to expect.

And sometimes, it's to get engaged in one element of their care. Sometimes there are resources that may be needed to help identify the process, such as voltage guided biopsies, or an additional commentary on the best treatment, such as when steroids have not been effective in the initial treatment of an individual sarcoidosis.

So the goal of our Cardiac Sarcoidosis Clinic is to offer this multi-disciplinary perspective on how to best approach the management of an individual's cardiac sarcoidosis, but also to learn more about the disease at a population level. For more information about either how to contact us, or for more information about cardiac sarcoidosis, please see the information on your screen. Thank you for listening to us, and thank you Dr. Blauwet for your expertise.

DR LORI BLAUWET: Thank you Dr. Kapa. And as Dr. Kapa said, please do not hesitate to contact us if you have any questions or wish to be seen here in our Cardiac Sarcoidosis Clinic.