**Transcript Details**
This is a transcript of an educational program. Details about the program and additional media formats for the program are accessible by visiting: https://reachmd.com/programs/project-oncology/cardiac-complications-in-sickle-cell-disease-emerging-evidence-and-risks/35929/

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Cardiac Complications in Sickle Cell Disease: Emerging Evidence and Risks

**Announcer:**
Welcome to *Project Oncology* on ReachMD. On this episode, we'll hear from Dr. Parul Rai, who will be discussing cardiac injury in patients with sickle cell disease. Dr. Rai is a physician in the Department of Hematology at St. Jude's Hospital in Memphis, Tenessee.

**Dr. Rai:**
Cardiopulmonary complications are one of the leading causes of death in adults with sickle cell disease. We've also seen that some changes start happening in the heart in even children—structural changes and functional changes in the heart. And early on it's in response to the anemia that we see in sickle cell disease, and these are called adaptive changes. In response to their anemia, their heart gets bigger, so there's dilation of the heart that happens, but over time we see that these adaptive changes kind of become maladaptive, meaning that now the heart is not able to function properly. It's not able to relax properly, which we call diastolic dysfunction. There is some fibrosis that we see in the heart muscle, and there are some publications, including our studies at St. Jude, that we published showing that this fibrosis is seen even in children. So this injury starts early on. We don't know how quickly it progresses, but we do know that cardiac injury over time in adults has a lot of morbidity and lot of mortality associated with it.

So the early changes that we see—we call it the pan chamber dilation, meaning all the heart chambers, like in the atria and ventricles, all four of them, are dilated in this population. We probably see a little bit more left atrial dilation than you would see with anemia, which is one of the early signs that this is not all related to anemia; something else is also happening. The other changes that we see is, as I mentioned, some myocardial fibrosis. That is fibrosis in the heart muscle. And over time you can also develop this condition called diastolic dysfunction, meaning the heart muscle is not able to relax, so when the heart is not able to relax properly, it doesn't fill with blood, and it's not able to push the blood out. So that is a problem called diastolic dysfunction. Then we also have noted that this population is high risk of developing arrhythmias, so could it be because of the fibrosis that they have a lot of arrhythmias. It's unclear, but sudden cardiac death, which most likely could be from these arrhythmias, is something that we see in this patient population at a very young age, in young adults, and that is one of the causes of early deaths as well.

So there are no accepted guidelines for cardiac screening of patients who don't have symptoms—who are asymptomatic. There are conflicting recommendations. While the American Society of Hematology does not recommend getting screening echocardiograms, the American Thoracic Society recommends getting echocardiograms at regular in adults to look for primary hypertension. So because we're not doing any screening tests right now, as it's not recommended, the ideal thing would be to make sure you're asking all the right questions. And we're also working towards finding what on the echocardiograms—if we do get echocardiograms in children, let's say—what the most sensitive markers would be. How can we identify really early injury? And then follow those patients closely to make sure that they are not developing full-blown injury, they're not coming to the hospital in heart failure. So that's something else that can also be done: work on developing more sensitive markers of early cardiac injury.

**Announcer:**
That was Dr. Parul Rai talking about addressing cardiac complications in patients with sickle cell disease. To access this and other episodes in our series, visit Project Oncology on ReachMD.com, where you can Be Part of the Knowledge. Thanks for listening!


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