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Management of CHD-PAH - General Considerations

Announcer:
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Dr. Krasuski:
Management of the CHD-PAH patient, what are the general considerations? All patients with pulmonary arterial hypertension should receive lifelong tertiary care in congenital heart disease patients. Patients and their families should be made aware of the major risks of pregnancy. The mortality for some of these patients, like those with Eisenmenger Syndrome, can be as high as one in three. And there's considerable morbidity for the patient and for the fetus. Effective contraception should be provided to patients with complex congenital heart disease and pulmonary hypertension. You want to encourage regular exercise and maintenance of an active lifestyle. Periodic six-minute walk distance or CPET are recommended to provide prognostic information and to guide management. Immunization against influenza, COVID-19, and pneumococcus is important. And it's also important to provide psychosocial supports for these patients.

Now, this is a complex algorithm, but I want to mention that expert referral should be at the top of this. So ideally, a patient with congenital heart disease-related pulmonary hypertension or Eisenmenger Syndrome should be managed in a specialty center. If they have Eisenmenger Syndrome, a structural intervention is contraindicated, and you should assess functional status and initiate medical therapy if they have any symptoms. If they have a systemic to pulmonary shunt, if their PVR is normal or slightly increased, these defects can be safely repaired. If they have a moderately elevated pulmonary vascular resistance, this ideally needs to be managed in a super special specialized center because the decision making here can be very challenging. If their PVR is severely elevated, generally we say that repair is contraindicated. We want to treat them medically and we want to reassess them. Some of these patients may respond to medical therapies and become correctable at a later stage. For patients with small defects, we generally, again, do not recommend intervention. We want to assess their functional class or we want to treat them if they have any symptoms.

Once we've established that a patient needs to be treated, we want to assess their function class. We generally become more aggressive for patients with more advanced disease. For Eisenmenger Syndrome the recommendations are still a first line endothelial receptor antagonist and then stepped-up therapy should their symptoms progress. For patients with a repaired or unrepaired shunt, we often start with first line combination medical therapies and then step these up again, depending on clinical deterioration. If they have worsened right heart failure or decreased functional capacity, a decrease in their six-minute walk distance or a lack of increase in that, an increased NT-proBNP or worsening RV function, these are all indications to become more aggressive in treatment.

Organ transplantation requires extensive discussion and collaboration between many specialists. Transplant options for advanced complex congenital heart disease and pulmonary hypertension are much more complicated. Some of these patients will require lung transplantation. Some of these patients will require heart-lung transplantation. For some of these patients, we may decide to do a lung transplant and a cardiac defect repair. In general, if we have a patient who can be done with less than 60 minutes of pump time, we'll try to repair the defect and then do a lung transplant. But if the repair is more involved, often we'll just do a heart-lung transplant. Congenital

heart disease still accounts for a large population of heart-lung transplantation, over a third of the data. This is back from 2011, and it's only growing since. Support options are becoming available as bridges to transplantation. And I think that's a very exciting area to be in at this point.

Announcer:
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