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ACHD: Basic Lesion Types

Announcer:
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Dr. Krasuski:
So let's talk about the basic lesion types in adult congenital heart disease. So, there are many different lesions present in pulmonary hypertension and congenital heart disease but very few have been included in prospective studies. On this slide, you can see there are different defects that, and the ones in red here are the ones that predispose towards pulmonary hypertension. In congenital heart disease, we break down lesions into simple, moderate, and complex lesions. In each of these various lesions, we have specific disorders that are more prone to the development of pulmonary hypertension, but it's important to recognize the studies that have been done have focused on two main groups the Eisenmenger group here, and these are in the complex group. And then the simple shunt lesions and slightly more complicated Primo defects. But we have a lot of defects here that have not yet been well studied with advanced medical therapies. And I think that's a real gap in our current knowledge.

So, shunt lesions are the most common causes of pulmonary hypertension. Patients with repaired and unrepaired defects can develop pulmonary hypertension. Up to 10% of those who do not have defect correction will develop pulmonary hypertension. And as many as two to 5% of patients with corrected lesions will develop pulmonary hypertension. These patients will have often subtle symptoms, increasing dyspnea, declining exercise capacity, and an annual you'll see an progressive increase in PVR, if you perform invasive assessment. 25 to 50%, a congenital heart disease related pulmonary hypertension patients, if they're not treated can progress to Eisenmenger syndrome. So again, earlier recognition and treatment appears to be helpful as in other forms of pulmonary hypertension.

So again, breaking down the subgroups in group one, you have Eisenmenger syndrome. These are cyanotic patients, they get secondary erythrocytosis. They get multisystemic disorders. In group two, you have the shunt lesions. There are correctable shunt lesions who have pulmonary hypertension. Oftentimes these are people that are just over circulated, if you correct the lesion and or treat their pulmonary hypertension medically, they will improve. There are also patients that are non-correctable. These patients have moderate to large defects. They haven't quite reached Eisenmenger syndrome but they may be heading in that direction. These patients are not cyanotic at rest, but if you ambulate them, sometimes they'll develop cyanosis.

So, these are a little bit more challenging when the Eisenmenger syndrome patient walks through your office door, it's pretty obvious they're cyanotic, they're clubbed. It doesn't take a rocket scientist to know that they have pulmonary hypertension but a patient who has milder disease, you may only see them get cyanotic when you actually push them exercise wise. In group three, you have patients with pulmonary arterial hypertension and small coincidental shunts. These patients in general are not going to get better with defect, closure and often defect closure is contraindicated. They may be using that defect as a pop-up valve for their right side. So, if they get sick or the exercise, they may shun more but it's a compensatory mechanism to increase their cardiac output at the expense of dropping their saturation. And group four of the patients after defect correction, this pulmonary arterial hypertension can develop immediately or many years potentially after their correction. And it can occur in the absence of significant postoperative hemodynamic lesions. Now, the first

thing you want to do whenever you assess one of these patients is make sure they don't have postoperative hemodynamic lesions. Because again, this may be somebody that's correctable reversible in that case, but we now know that this is a very aggressive phenotype that needs to be rapidly identified and rapidly treated.

Morbidity and mortality are higher in patients who have congenital heart disease-related pulmonary hypertension compared to patients without. So, this is data from the province of Quebec almost 40,000 patients with congenital heart disease, a little over 2000, a diagnosis of pH. So that meant that about one in 17 patients had congenital heart disease-related pulmonary arterial hypertension. In those patients, the mortality was twofold higher. Their morbidity hospitalizations was threefold higher and when they were hospitalized, their hospital days were threefold higher. So, this is a sick group of patients that needs very close medical attention.

Announcer:
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