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A Short Tour of Congenital Heart Disease Associated PAH (CHD-PAH)

Announcer:
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Dr. Krasuski:
A short tour of congenital heart disease associated pulmonary arterial hypertension. So, let's talk a little bit about adult congenital heart disease. Congenital heart disease affects slightly less than 1% of live births in the U.S., and this excludes bicuspid aortic valve and mitral valve prolapse. Pulmonary hypertension is a common complication of congenital heart disease, somewhere between five and 10% of all patients, particularly if the congenital heart disease is significant and unrepaired. With newer and improved diagnostic techniques and evolving medical, catheter-based, and surgical interventions, there are now over 1.4 million adults with congenital heart disease living in the United States. And roughly about a decade and a half ago, we reached a point where there were more adults than children. Unfortunately, most physicians get minimal exposure to congenital heart patients during their training.

And ideally, all adult congenital heart disease patients should be seen by an ACHD specialist at least once during their lifetime. Complex patients need follow-up every six to 12 months in an accredited ACHD center. Specific aspects of pulmonary hypertension in congenital heart disease include higher flow and more downstream shunt lesions that are more likely to cause pulmonary arterial hypertension. Like any case of pulmonary hypertension, ACHD-related pulmonary hypertension can be either pulmonary venous or pulmonary arterial and emphasizes why catheterization is so important for the diagnosis. We use the same hemodynamic criteria for diagnosis as other group one disease. Differentiation of etiology dramatically impacts management, and ideally catheterization should be performed by a board-certified ACHD physician.

Now there have been a lot of advances that I've mentioned in congenital heart disease, and this has led to a much larger patient population of adults with congenital heart disease. If you were born in the 1960s, you can see your chances of dying within the first year were about 50%. Your chances to survive to adulthood with congenital heart disease was only about 15%. Whereas if you're born in 2010, your chances of surviving to adulthood is now probably close to 90%. So, there are 1.4 million adults with congenital heart disease. Many patients are only palliated for their disease. Their lesions can recur, and the palliative methods can cause problems. And even the simple lesions, we consider shunt lesions to be some of the simplest lesions, they can result in pulmonary hypertension, arrhythmias, and heart failure even after successful repair, and that's why it's so important for these patients to be closely followed. Epidemiologic studies are limited. There's varied complexity of these patients, and in the United States at least there's no centralized database. Although there's definitely work being done towards that process, and hopefully we'll have one in the next decade.

So, in patients with congenital heart disease and pulmonary hypertension, we can classify them into four distinct groups of patients. In group one, we have Eisenmenger syndrome. These are patients who develop increased pulmonary vascular resistance with unrepaired shunts. When they develop progressive increase in their pulmonary vascular resistance, they get right-to-left shunting, cyanosis and its various complications. Second group is persistent systemic-to-pulmonary shunts. These can be either repairable or unrepairable lesions. And the third group are small, coincidental defects, in which case it's simply pulmonary arterial hypertension and an incidental

defect such as a small atrial septal defect. And finally in group four are patients who've undergone defect correction, and many years later potentially can develop pulmonary hypertension. For patients with shunts repaired and unrepaired, they need to be screened at an ACHD center for the development of pulmonary arterial hypertension. And then finally recommendation-wise, postoperative PH screening should occur in subgroup four, patients who've undergone defect correction who don't have pulmonary arterial hypertension at the time of their surgical repair. But this workup should include clinical, echocardiographic and ECG screening during follow-up visits, starting at three to six months after the defect correction, and then ideally yearly follow-up afterwards in long-term cardiovascular clinics.

So, let's briefly discuss the manifestations and common physical exam findings in patients with pulmonary arterial hypertension. So here is a patient with a milder form of pulmonary arterial hypertension. This patient will likely have an elevated jugular venous pressure. It's important to look at the JVP and compare it to the carotid pulse. In most of these patients, you will see an A and a B wave. You'll also be able to push on the liver and assess for hepatojugular reflux. Occasionally there can be no decline in the jugular venous pressure with inspiration or a Kussmaul's sign, suggesting some element of either constriction or restriction. They can have relatively clear lung fields if they have no concomitant lung disease. On auscultation, you'll hear an increased pulmonary closure sound, oftentimes a narrow splitting of the second heart sound. They will have both pulmonary regurgitation and/or tricuspid regurgitation and often a right-sided S4, which is heard just below the right clavicle. And by palpation, they can have a prominent P2 and a right ventricular lift. And then depending on the degree of pulmonary hypertension, they may have some degree of cyanosis.

So here is the progression of pulmonary arterial hypertension related to congenital heart disease to Eisenmenger syndrome. You start with a left-to-right shunt. You get increased pulmonary blood flow and pressure. This leads to pulmonary vascular remodeling, an increase in the pulmonary vascular resistance, and eventually when that PA pressure exceeds the pressure in the systemic bed, you can reverse the shunt, and now you're chanting right-to-left. You get cyanosis and Eisenmenger syndrome related to the cyanotic complications. So, in Eisenmenger syndrome, the physical exam becomes more extreme. For those patients, they're almost always going to have an elevated jugular venous pressure. They may have headaches, seizure, stroke related to their increased thrombotic potential, related to their congenital heart disease, and polycythemia. They again are going to have often clear lung fields, a very increased P2. Sometimes a single S2 only is heard. Just the P2's heard. A high-pitched pulmonary regurgitation. TR murmur is again heard. Right-sided S4 is very common. They're going to have very commonly a palpable P2 sound and a right ventricular lift. These patients oftentimes going to be tachycardic as well at baseline. They are going to be cyanosed, but in addition they'll have clubbing. Their nail bed angle will change, and they'll have large, clubbed fingers and sometimes toes. If you see a patient and they have a right hand that is not cyanosed, but they have a left hand and both feet or just both feet, that is called differential cyanosis. Very classic for patent ductus arteriosus with development of Eisenmenger syndrome. They'll have hepatomegaly, often a pulsatile liver. They're prone towards gallstones and development of ascites. They also develop kind of this patchiness on their skin we call livedo reticularis. They're at increased risk for bleeding. They have complications such as leg cramps, abdominal cramps related to hyperviscosity, and peripheral edema and also predilection to gout.

So, what are the unique considerations in the congenital heart disease-related pulmonary hypertension patient? First of all, we want to ask the question is there too much or too little pulmonary blood flow? We don't often think about this, but in a congenital patient, there is often a important balance between ensuring adequate blood flow and preventing overcirculation. So, in the infant, oftentimes we'll band the pulmonary arteries to reduce the risk for overcirculation. And likewise, if they're undercirculated, we'll create a shunt so that patient can make it through their early years and allow their lungs to develop. If there's a defect present, is it correctable? If the defect is correctable, what's the best method in which to correct it? For some patients, a defect can be corrected through a surgical approach only, and in other patients we have transcatheter options. And I will say that defects that we thought were only correctable through surgical means, we are now approaching percutaneously in the little over two decades that I've been practicing. We have advanced medical therapies, but have they been adequately tested, and do they work in some of the patients with congenital heart disease?

Announcer:
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