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Released: 04/15/2022
Valid until: 04/15/2023
Time needed to complete: 2h 00m

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Pulmonary Hypertension in an Adult With Complex Congenital Heart Disease

Dr. Krasuski:
Hello, I'm Dr. Richard Krasuski from Duke University, and today I'd like to talk to you about a case of pulmonary hypertension in an adult with congenital heart disease.

So I call this a complicated return to care. It's a 34-year-old woman with complex congenital heart disease who presented to urgent care complaining of progressive dyspnea on exertion. She was born with transposition of the great arteries, a ventricular septal defect, a small atrial septal defect, and pulmonic stenosis. She had been followed by pediatric cardiology until age 26 but had not seen a physician in the eight years between due to insurance issues. And this is not an uncommon problem we face in adult congenital heart disease, where people are lost to care in their 20s, often due to a loss of insurance in between.

So, here is basically her anatomic picture. She had d-transposition of the great arteries with pulmonic stenosis, so if you see the pulmonary artery is narrowed in the outflow tract, and you see her great vessels there are coming off the opposite chambers. So you have the right atrium connecting to the right ventricle and going out the aorta, and then you have the left atrium connecting to the left ventricle and going out to the pulmonary artery. So left unrepaired, there would be mixing of blood, and there would be a need to obviously get the blood from the venous circulation out to the lungs and get the pulmonary venous circulation out to the body. Now, transposition of the great arteries accounts for five to 7% of all congenital heart disease, and often, there are associated lesions. So up to 50% will have ventricular septal defects, up to 1/4 will have outflow tract obstructions, which affect the left ventricle, and 5% will have coarctation of the aorta.

So this patient underwent an atrial septostomy at birth, and then a Rastelli procedure at age three. Now, because of the issues with the Rastelli procedure, she had three subsequent operations, which were described as conduit revisions. For about three years, she'd experienced dyspnea with activity, first, when she was exercising, and then eventually, with pretty much any activity. Her physical examination was notable for an oxygen saturation of 95%, a jugular venous pressure of 11 with very prominent V-waves, a three out of six pansystolic murmur, clear lungs, and 2+ peripheral edema.

So just to illustrate what the Rastelli procedure is, this was developed back in 1967 at the Mayo Clinic. The aortic outflow is routed through the ventricular septal defect via patch. The pulmonic valve opening is oversewn, and then a conduit is placed between the right ventricle and the pulmonary artery. So as an effect of this surgery, you restore the left ventricle as a systemic ventricle, unlike the atrial switch procedures, the Mustard or the Senning, which in those cases, basically, you created an atrial baffle, so the blood from the venous circulation was taken out to the lungs via the left ventricle, and then blood was returned from the pulmonary venous circulation through the right ventricle and out to the aorta with the Mustard or the Senning. And the problem there is, you develop systemic ventricular dysfunction, and you can also develop AV valve regurgitation of the systemic AV valve because it's a tricuspid valve. But with this correction, you actually have the correct ventricles pumping out to the great vessels. But this problem with this particular surgery is the need for frequent conduit interventions.

So, here is the patient's imaging. So first, she underwent a chest x-ray, which showed an enlarged cardiac silhouette, very prominent pulmonary arteries, and some postoperative changes as are often seen in adults who undergo congenital heart surgery. She underwent a V/Q scan, which was a low probability of pulmonary embolism, and then she underwent a series of echocardiograms, including

transthoracic and transesophageal studies. Now, this showed normal left ventricular size and systolic function, a dilated right ventricle with mildly reduced systolic function, and mild tricuspid regurgitation with the right ventricular systolic pressure of 65. And on this particular diagram, on this echo here, you can see, on the right side of the screen in the upper right, the flap, and this is basically the prior patch that is dehisced now and is moving back and forth. And if you look at the color image on the lower right, you can see the flow going from the systemic side over to the pulmonary side, so this is a rather prominent shunt, going from one side to the other.

So an MRI confirmed that there was a partially dehisced VSD patch. The Qp:Qs was calculated at 1.3:1, so in other words, there was 30% more blood going out to the pulmonary vasculature than was going to the body, and by MRI we could tell there was a moderately dilated right ventricle. She underwent a cardiac catheterization that showed a right atrial pressure of eight, a right ventricular pressure of 70/8, a pulmonary artery pressure of 70/30, with a mean pulmonary artery pressure of 45, and a wedge pressure of eight, so there's really no gradient across her pulmonic valve, but pretty significant pulmonary hypertension. The Qp was 1.7, Qs was five, so that gives you a Qp:Qs ratio of 1.4:1, and the pulmonary vascular resistance calculated to 5.3 Wood Units.

So the question here is what would you recommend? How would you treat this patient? Would you refer them for reoperation to repair the shunt? Would you refer the patient for percutaneous attempt to close the shunt? Would you initiate advanced medical therapy for the increased pulmonary vascular resistance, or would you make no changes and arrange for close observation? So in this patient, we decided to treat their pulmonary hypertension. So the question is, when can shunts be safely closed?

This has been looked at, and the consensus papers that have been written about this topic say, that if you have an indexed pulmonary vascular resistance that is less than four Woods Units, if your Qp:Qs is greater than or equal to 1.5 and the patient's symptomatic, that is the patient you want to ideally close. If the PVR is greater than eight Woods Units, if you're on the left side of this color diagram here and you have significant pulmonary vascular disease, those are patients you should not consider closure. And then the gray zone is the group in between. Some of the ways you can assess this. Basically, of course, you want to ideally send this patient to a tertiary care center, where they have experience with managing these patients. You may want to try a trial of balloon occlusion to see how the patient tolerates occlusion of the defect. You may want to do vasodilator testing. If they have a drop in the pulmonary vascular resistance greater than 20%, or the PVR drops, and it's now less than six Woods Units, with vasodilator testing, that's somebody you can probably repair safely.

There is also the treat and repair approach, in which you can try advanced medical therapies for pulmonary hypertension and reassess the patient. Again, close follow-up should always be followed, if you, especially, if you decide to close the patient, you really want to make sure you reassess what the hemodynamics do over time. And then partial defect closure has been attempted in some of these patients, where you actually will leave a fenestration, so they can have a pop-off valve for the right side.

So in this patient, we started medical therapy with first, macitentan, and then tadalafil was added later. Her six-minute walk distance improved from 310 to 350 meters. Her function class improved as well. And then on repeat catheterization, her right atrial pressure was now six, RV pressure was 50/6, PA pressure 50/24 with a mean of 30, and her wedge pressure is now six. Her Qp calculated at 9.5, Qs was six, that gave her a Qp:Qs of 1.6:1. And her PVR was now 2.5 Woods Units, so she was definitely now into that range where we would consider closure.

So the question now is, what should you do? Should you refer them for reoperation to repair the shunt? Should you, again, refer the patient for a repeat intervention to try to close the shunt? Should we add a third drug in this patient to treat the pulmonary vascular resistance, or should we make no further changes and arrange for close observation? So again, we discussed these options. With five prior sternotomies, surgery was considered to be a prohibitive risk. Device closure was initially attempted with a transcatheter device, a closure device, but was unfortunately unsuccessful. We could not get the device to seat properly. Transplant evaluation was pursued, but the patient was thought to be prohibitively alloimmunized, and so she was not a candidate for transplantation. And finally, the decision was made to attempt coverage of the defect with a covered stent, so we were going to place this into the pulmonary artery.

And so we printed a 3D model, and you can see this is the model here, the aorta coming around, and you can see the pulmonary artery here in pink. Here is now in the catheterization suite. We're across into the systemic ventricle, doing an injection here. You can see a wire that is now seated into the pulmonary artery. So what we did is we went into the pulmonary outflow tract. We placed a covered stent. We actually placed two separate covered stents. And so now the defect was fully covered, but unfortunately, this also resulted in covering of this conduit, and so we had to place a transcatheter pulmonary valve.

But this was very successful, three months after the procedure she's been maintained on macitentan and tadalafil, and now her six-minute walk distance has improved from 350 meters to 410 meters. Her New York Heart Association function class improved from two to one, and a repeat echocardiogram showed near normal RV size. And now the estimated RV systolic pressure is 40, with 13/8 millimeters of gradience across the pulmonic valve.

So what would you now recommend? Should we continue both the drugs? Should we discontinue both drugs? Should we discontinue tadalafil or should we discontinue macitentan? So what we did in this patient basically, is we continued the drugs, and we followed them very closely, with a plan that we would try to discontinue therapy, if pressures in function class improved and continue to improve. So, just a brief mention of pulmonary hypertension in transposition patients. This usually occurs after late repair, greater than age two. Usually with atrial switches, it's more common, if a shunt is present before repair. So if the patient was not repaired early in life, and if they had a large left to right shunt, that's a patient that develops pulmonary hypertension. You need to exclude pulmonary venous pathway obstruction, if there's a baffle obstruction on the pulmonary venous side. It can often present with very similar elevated pressures. That's something that can be treated either surgically or percutaneously, but once they develop, pulmonary hypertension's associated with a very high mortality.

So, putting this all together for a treatment algorithm for patients with congenital heart disease and pulmonary arterial hypertension. Ideally, this is somebody that should be evaluated in an expert center and confirmation of pulmonary arterial disease associated with congenital heart disease should be confirmed. There are, in fact, three separate groupings for these patients. We typically discuss four groupings, but patients with pulmonary arterial hypertension and small defects, or those who have had their defects repaired, are treated very similarly to group one patients. And, in fact, the ones with closed defects appear to have slightly worse outcomes, and so those are people that we want to obviously treat aggressively. There's Eisenmenger Syndrome, which we didn't discuss today, but again, if they're symptomatic, these are folks that ought to be treated with advanced medical therapies. And the group that we focused on is in the middle. Those are the folks with systemic to pulmonary shunts that have not been repaired. If their pulmonary vascular resistance is severely elevated, it means an indexed PVR greater than eight, that is somebody that should not be repaired. They should be treated with medical therapy. They can possibly be reassessed at a later time, like our patient was, to see whether they then become candidates for repair. If they're moderately elevated, this is, again, somebody, where you want to collect additional data. You want to look for vasodilator challenge data, balloon occlusion, et cetera, to decide who to treat.

And finally, if they're in the green zone here, if their index PVR is less than four Woods Units, that's somebody that would benefit from closure and should go ahead and close the defect. Should they develop pulmonary hypertension afterwards, that is somebody, if they're symptomatic, should be treated with advanced medical therapies. So we've discussed today, a very complicated case of a patient that developed pulmonary hypertension with congenital heart disease. These are the various groupings here that are present in the slide.

Again, consultation referral is extremely important in these patients and has been shown to improve long term clinical outcomes in these patients as well. I thank you for your attention.