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Discussion

Presenter: Dr Christian Pizarro

Dr Christian Pizarro *(Wilmington, Del)*. I'll entertain any questions. Thank you.

Dr Richard Ohye *(Ann Arbor, Mich)*. Thanks, Christian. Really fantastic results with a very, very difficult group of patients. I have a couple of questions based upon your slides. When we think about these—as a group of patients—they tend to be those kids that do well on prostaglandin and can be weaned and go home. There are those that can't be weaned from prostaglandin, and they usually fall into 2 categories, either because of cyanosis or because of cyanosis and heart failure. In those kids that fail because of cyanosis, we can usually shunt them and let them go home and come back and fight another day. I saw that none of your patients were like that. Was it just bad luck with your patients, or do you prefer this sort of more aggressive initial step during the neonatal period to decide whether you're going to go single or 2 ventricles?

Dr Pizarro. That's a great point, Rick, and perhaps a little bit of the former. The thing is you could see a lot of these patients had an organ dysfunction, and I think that our threshold to intervene was based not only in the fact that the patients remained blue, but the fact that they had actually a progressive rise in their creatinine, the urine output was not good, the lactate was trending up, and that really prompted us to intervene.

Dr Ohye. Okay. Thank you. On your slide that said, I think, it was initial stabilization intervention. I noticed that some of the patients actually decreased in their pulmonary blood flow with things like pulmonary banding and occlusion of the main pulmonary artery. It's a little bit counterintuitive, and I just thought maybe you could fill us in a little bit on that management strategy.

Dr Pizarro. You're absolutely right, and I think that this is something that I'm glad you pointed out on this particular slide, which I believe is probably one of the most important lessons that were learned in the management of these patients. I think that traditionally, we believed these patients are extremely blue and challenged just because of ineffective pulmonary blood flow, which I think is probably half

of the story. I think that it's been our experience that when we control, for example, severe pulmonary insufficiency in those patients with a ductus that is open and have a circular shunt. We ended up with Po_2s above 40, and I think that there is a component of maldistribution of cardiac output, which when we banded some of those branch pulmonary arteries, much like a hybrid, has resulted in a very prompt ability to resolve the hyperlactatemia, improve oxygen delivery, and those patients get better pretty quickly. So obviously something that needs to be tailored according to what the patient is telling us.

Dr Ohye. Okay, great. I also noticed in your conclusion slides where you said the criteria for poor outcome are probably not applicable in the current era. As you pointed out, most of your patients would have been predicted not to do well. So, I know there's only 1 death, but can you tell us anything about what you think are potential criteria for poor outcome?

Dr Pizarro. Only 1 death didn't help us to risk-stratify anything, but I think that overall, the GOS Score and the cardiothoracic ratios you pointed out, those patients—none of them would have survived in this particular cohort. I think that we ought to be very careful about making inferences about the potential prognosis on the basis of those old criteria. I think that nowadays, it seems to me that coexisting rhythm disturbances are incredibly challenging to manage and very destabilizing and perhaps something that should be included in any score. And the other thing is perhaps volume measurement with MRI, which I've seen that we've had patients with predictive volume less than 35% who have survived, but it seems like less than 30 is extremely difficult to get these patients through.

Dr Ohye. Thanks. I've had a couple patients where we patched out their tricuspid valve sort of in a supra-annular position and then went back—do that to allow for continued growth, and then went back and did a repair on their tricuspid valve. Have you had any experience with that?

Dr Pizarro. That's a great point. No, we haven't. We have entertained that a couple of times, and I think when we had discussion with the parents, they really had not been interested in pursuing an invention that might be potentially more morbid in the short-term and have some complications rather than taking a very simple, predictable intervention which would be a second stage. I think that's a great idea now that we have a good tool to repair those valves. It would be something that we should entertain, and I think that probably something that should be done sooner rather than later. So, we don't end up with a valve that has been subjected to a very limited amount of pulmonary blood flow and therefore has become small.

Dr Ohye. I think we probably have time for 1 more quick question before our 3 minutes are up. Your 1 patient where you did an RF perforation. I know that I've always been sort of afraid to balloon the pulmonary valve or do an RF perforation, getting sort of a circular shunt. So, what were your thoughts on that, and how did that patient do?

Dr Pizarro. Actually, the patient ended up in the cath lab without our knowledge. And there was a fair bit of enthusiasm. The patient didn't do well, and we ended up rescuing that patient. And that patient actually had a reintervention and I think that it didn't work as it was intended, and it was just probably an overoptimistic approach to this particular patient.

Dr Ohye. So still a bad idea?

Dr Pizarro. Yes.

Dr Ohye. Alright. Thanks, Christian. I know we're going to do a little bit of a live Q and A now, so I appreciate your time.

Dr Pizarro. Thank you for your comments.