Discussion to: Outcomes of surgical management of Ebstein’s anomaly and tricuspid valve dysplasia in critically ill neonates and infants

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Disclosures:

None
Dr. Christopher Knott-Craig (Memphis, TN):

Well, I want to congratulate the group from Columbia and the co-authors on a very difficult group of patients to deal with. The audience should be well familiar that neonatal Epstein's requiring surgery carries a mortality three times higher than that of a Norwood operation, which is a step 5 case, and it still even in your hands, had a mortality of 22% or at least in the manuscript that you submitted to me. There were a couple of things that I thought were interesting and worth talking about. One was the Starnes operation was offered to children that were really sick very soon after birth, and that makes good sense. In our own institution, we've kind of changed a little, and so the patients that you identify as having a circular shunt or having severe pulmonary regurgitation will take them straight from the delivery suite to the operating room, like at the main pulmonary artery and place bilateral pulmonary artery bands, maintaining the circulation with prostaglandins, give them 5 or 6 days to recover, avoiding cardiopulmonary bypass initially, and then either do a Starnes repair or do a two ventricle repair. So that was the one thing.

The second thing I thought was interesting is that for Epstein's anomaly, if I understood your manuscript correctly, the one-year mortality for Epstein's anomaly was 44%, and the majority of those patients had a Starnes repair. I think two-thirds of them had a Starnes repair. So I wanted to ask two questions: A, have you considered postponing the Starnes-- obviously recognizing that the patients need an operation right away, but maybe postponing the Starnes down the road, the way I've sort of suggested, A, and B is if you could maybe comment on the incidence of heart block, which was 22% in your patient population and maybe a comment on even though the patients had a single ventricle pathway, the one-year mortality was still 44%.

Dr. V. Reed LaSala (New York, NY):

Thank you very much for your questions and comments. So, regarding the approach that you've taken in your work, I definitely appreciate that. And I've read about it. That's not something that we have done, but it's definitely something that we could consider doing in the future, doing those patients as postponing the Starnes or two ventricle repairs as much as possible and doing a stage 1 procedure early on. For your second question regarding the patients with circular shunt, could you remind me of the question? I'm sorry.

Dr. Knott-Craig:

How many patients, in fact, had a circular shunt in your cohort of—

Dr. LaSala:
There were three. Yeah.

Dr. Knott-Craig:

So, were those the same three patients that had preoperative ECMO?

Dr. LaSala:

It was not the same three patients, no. But yeah, two of them died; one of them lived. Yeah, they all had a Starnes.

Dr. Knott-Craig:

And then just last, if you could just comment on the incidence of heart block in 22%.

Dr. LaSala:

Sure. Yes. We did have a couple of patients with complete heart block. Some of those patients had an annuloplasty for tricuspid valve repair, so that is a possible etiology. And of course, the Starnes patch is placed in an area that's near the AV node. So that's a possible etiology as well.

Dr. Knott-Craig:

Yeah. So just one last comment, if you don't mind, Jose and Luciana De Silva have published very nicely, changing the tricuspid valve repair to be outside the annulus of the tricuspid valve along the ligament of Todaro, and then including the coronary sinus as you come down. And that is pretty much eliminated in the experience and in our experience eliminated complete heart block. And so the question is, are you still placing the Starnes patch on the inside and leaving the coronary sinus on the atrial side, or are you coming across the coronary sinus and leaving it on the atrial side, but using the ligament of Todaro to avoid the conduction tissue or doing the conventional way which has been described?

Dr. LaSala:

We're still doing the former thing that you mentioned. We're not using the ligament of Todaro.

Dr. Knott-Craig:
That may be something worth thinking about.

Dr. LaSala:

Definitely.

Dr. Knott-Craig:

Thank you very much.

Dr. LaSala:

Thank you.

Unidentified Speaker 1:

Congratulations on your great presentation. Yeah. As you said, it's hard to evaluate the results; the Ebstein anomaly in the neonatal period with a small series. In Pittsburgh, we also had a small series. We have 11 patients with two of them went home without medical treatment and the other had operation. And when we did the repair, we did later on. We tried to delay and did with 40 days, one, the other one, about 35. So, they're not really in neonatal period anymore. But we think in the patient, we had a hard time, and the patient had ulnar atresia. Most of them require ECMO support and all of them we did the Starnes procedure. And actually, I didn't do any. Victor did most of them and Myra did one too. And so, if you have pulmonary regurgitation, severe with circular shunt, also we think it's important to do the Starnes procedure. And I hope with the Starnes procedure, we can improve the results in those more difficult cases. And in some other ways, I think we can do the repair if you can postpone a little bit, you can do the biventricular, I mean primary recon repair or other repair at neonate. But for those you need a good function of the right ventricle, and the main issue is the tricuspid regurgitation that is so severe that is not allowed. It is hard to develop good systolic pressure, and in that case, you might be able to keep the patient prostaglandin or a stent, and then do the repair early, but not as early as in a few days after they were born. So that would be my comment. And I will ask you when you did the primary repair, did you do it early on as a neonate or wait a little bit?

Dr. LaSala:

So, thank you very much for your comments and questions. The patient that we did the cone repair on was actually at nine days of age. So, they were very
young. But they did survive, and it worked out. We have taken a very similar approach to what you're describing, where the neonates who are critically ill with circular shunt, we will do a Starnes, and then come back once the tissue is better to do the cone repair. And I think that's the direction that we're moving in.

Unidentified Speaker 1:

Yeah. All right. Thank you very much.

Dr. LaSala:

Thank you.