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Key Words: Ebstein anomaly, cone reconstruction, cone repair, cardiac magnetic resonance imaging, surgery

Discussion

Presenter: Mr Ralph M. L. Neijenhuis



Dr Emile A. Bacha (New York, NY).

Thank you very much. Mr Neijenhuis, that was an excellent presentation and really an excellent study showing significantly improved reverse remodeling of both ventricles, although at different times after the cone procedure. This is especially important

because magnetic resonance imaging (MRI), as you explained, is the only modality that gives us valid right ventricle (RV) function data.

So for those of us who do the cone procedure and believe in it, it is a very gratifying study. As a quick aside question: Do you call this remodeling or reverse remodeling? I thought this would be reverse remodeling.



Mr Ralph M. L. Neijenhuis (London, United Kingdom). I think that's quite difficult to say. Could you clarify what you mean by reverse remodeling instead of remodeling?

Dr Bacha. I thought the positive changes that you observed and ventricles were called reverse remodeling,

but I don't want to spend too much time on this. My first question relates to the degree of residual tricuspid regurgitation (TR) that you have and how does that correlate with improved RV and then left ventricle (LV) function? Did you find that if you had the best repair, so to speak, one that has the least TR postoperatively, resulted in the most improved LV and then RV function, or was there no correlation? This alludes to the fact that this is a complicated operation. The results can be mild plus TR or even moderate TR postoperatively. And so the question is: Do you get the same amount of improvement with moderate TR versus mild TR postoperatively, understanding that the vast majority of patients start off with wide-open TR?

Mr Neijenhuis. We are still doing the final analysis, but what I can say is that in our cohort we had some patients who still had a paravalvular leak or had more severe TR at later follow-up on MRI. These patients tended to do worse and that's what explains our findings and still having this remaining TR. Of these patients, 1 underwent reoperation, actually. But we do not have more recent MRI of that patient.

Dr Bacha. So what you're saying is: It is important to get as good as possible technical result with as little TR as possible postoperatively because that will give you the best improvement in ventricular function. It was very interesting to look at the differential results between LV and RV with the LV improving almost immediately or early on, and

the RV initially having a decline in function, and then improving. I'm sure that the patients who get postoperatively complications or those who even die, die from the initial decline in RV function that you demonstrated so well. Can you further elucidate for us the timing of this RV improvement? When would you expect to see it—after a year, after 6 months, after a year and a half? When do you see the RV start to improve?

Mr Neijenhuis. The previous study conducted at our center investigated this as well. They included what we called back then midterm results—so that was with a mean follow-up of around a year. In that work, we did show the improvement in LV form and function, but not in the RV form yet. To give you a specific answer would be quite difficult for me, because I'm the research fellow, and I think Professor Victor Tsang has a better answer for you. But yes, it does seem to happen at a later moment—after a couple years.

Dr Bacha. My final question to you is related to age at repair. There are patients who present late—adults, older adults, aged 50 years. Let's say aged 50 years where you wonder whether you're doing a good thing by repairing versus replacing, or whether they should go directly to transplant or not. Does your study inform us on that, and does it give us sort of the sweet spot in terms of ideal age for the younger patients? So that's a 2-part question: Is there an age where you think changes are irreversible and you don't see an improvement? And, For the younger patients, the children, is there a sweet spot where you think you get the most improvement, let's say around age 4 or 5 years?

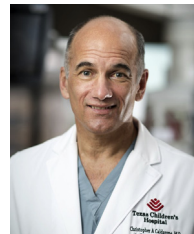
Mr Neijenhuis. Looking at our data, we of course have included a lot of adult patients but really most of our patients are in their teens and 20s with some of them being late. Also, more recently we saw a change in our center of doing less and less of the adult patients, and so they will be treated with different surgery, instead of the cone reconstruction. As for the younger patients (and we do have quite a lot of the really young patients in our cohort, and they get the cone reconstruction, the biventricular repair), I would still find it difficult to say, in my position, when I would suggest doing a cone reconstruction and when not. We have some very young patients who are doing incredibly well. Then, there are also patients who have really dilated hearts preoperatively and for them, it's still more difficult to get by after surgery.

Dr Bacha. Maybe I can ask Dr Starnes: What's your ideal age for a cone repair in an asymptomatic child?



Dr Vaughn A. Starnes (*Los Angeles, Calif*). We usually approach that at around age 4 to 5 years if we can get that far. Mr Neijenhuis: Did the volume of atrialized ventricle enter into your calculations about improvement of end-diastolic volume and ejection fraction in your analysis?

Mr Neijenhuis. The volumes of the atrialized component were not used for the analysis of the indexed volumes (and ejection fraction) on the cardiac MRI. For the general form of the ventricle, the atrialized component was not used preoperatively. We only used the part apical of the tricuspid valve for the area of the RV. So postoperatively, the atrialized part of the RV was included in the figures, partly explaining the increase as well. I hope that answers your question.



Dr Christopher A. Caldarone (*Houston, Tex*). This is a terrific study and the MRI data are really exciting. And I recognize that it's probably hard to get this follow-up data on a big cohort of patients like this. Really only a third of the patients had postoperative MRI data. This doesn't take away from

what you're trying to do, but I think you're going to need to do a bit of an analysis to prove that that third of patients with postoperative MRI data is representative of the group as a whole. I suspect the reason some got in some didn't is travel considerations and your referral system with patients coming from a long distance. So I understand why that limitation may exist, but when it comes to publishing this work, you're going to need to prove that that subset is representative of the whole group.

Mr Neijenhuis. Thank you, Dr Caldarone. You're right. We looked at the differences in the group preoperatively, so did the whole MRI group have a lot of differences from the total cohort? And there we did not find anything really significant, but of course with the need for patients to go back to our center, which can be quite the travel, you can think of possible biases of patients being worse off only coming back for example or patients living in the region. I think it's a great point, and I think we should include it in the manuscript.