Discussion to: Impact of pulsatile pulmonary blood flow on cardiopulmonary exercise performance after the Fontan procedure

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Discussion to: Impact of pulsatile pulmonary blood flow on cardiopulmonary exercise performance after the Fontan procedure

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Dr. Edward Hickey (Houston, TX):

Good morning, everybody. Ed Hickey from Texas Children's and previously Toronto where we had a pretty sizeable cohort of adults who had previously undergone Bjork-modified Fontans in the '80s and '90s. And anecdotally, it was clear to us actually at the time that they really did very well in the long-term, and you have now shown that in what's a pretty elegant analysis and very well-written manuscript, so congratulations to you and your coauthors. I have three general questions for you. There's long been a recognition that some sort of contractional pulsatility in the Fontan circuit would be beneficial. In fact, it's my understanding in the very, very, very early Fontan procedures where valves were incorporated in the IVC and PA, there was a thinking that the right atrial contraction might contribute to pulmonary blood flow. That was quickly shown not to be the case. But in the Bjork modification, you are necessarily incorporating the ventricle in the circuit. It's unlike any other Fontan, which excludes the ventricle. So conceptually, that will help with pulsatility with pulmonary blood flow. So, this also will make sense. How can you tease out
though, the relative contribution of that ventricle and how it's contributing to pulmonary blood flow versus the fact that these really are the very best Fontan patients? These are the tricuspid atresias who have had very little in the way of prior palliation, most of them PA bounds or nothing versus many of the other groups, the TCPC, in particular. These have a very high prevalence of RV-dominant ventricles, multiple complex prior palliations. They're not really apples and apples, are they?

Mr. Lukas Klemm (Munich, Germany):

Thank you very much for your questions. Yes, you are completely right. This is probably the best subset of patients we have in this patient collective. If you look at our cohort, we had mostly patients with tricuspid atresia with a nominate position of the arteries as well as some patients with double-inlet left ventricle also with a known position of the arteries. When we now look at the function of the right ventricle and the rudimentary right ventricle to the systolic function, we also have to keep in mind the pulmonary artery size and the pulmonary vascular resistance. And in the previous studies, we could see that not so much the size of the right ventricle is the deciding factor but more likely the size of the pulmonary arteries and the pulmonary vascular resistance. We've seen some effect of, let's say, RV growth in the long run, but in initial phase, the RV size didn't have much of an effect on the pulsatility of those patients.

Dr. Hickey:

The revision rates in Bjork-modified Fontans are not insignificant at all. In fact, they can be pretty horrible revisions. The pericardial backflow is sitting right behind the sternum and it's just a sea of blood waiting for you. Once you are inside the chest, what made you strategize between putting a valve in that RA-RV connection versus taking it completely down to a TCPC? And then those that you did take down to a TCPC, how did you manage the right ventricular cavity and its outflow to the PAs?

Mr. Klemm:

That's a very good question. Thank you. First of all, when we look at the initial Fontan-Bjork procedure, he used in the beginning homografts and had problem with the calcification rate and also reoperation. And we had switched to providing the back wall just for a direct connection of the RA to the RV and used also Goretx for the initial procedures. We are now thinking if something like kind of a return of the Fontan-Bjorks, we are looking into decellularized homografts as well, if this might be an option in the long run. Regarding your question for the takedown or a conversion to a TCPC, in initial Fontan-Bjork, we had to close the ASD as well as the VSD, and those patients, as far as I can remember, we opened up the ASD again and close down the pulmonary artery for the TCPC procedure.
Dr. Hickey:

Okay. So, you left a blind ending RV cavity.

Mr. Klemm:

This was on one patient, actually.

Dr. Hickey:

Okay. Just one final question, just very briefly. So most contemporary modern young surgeons are probably completely unfamiliar with what a Björk modification is because the de facto norm nowadays is, at the age of three or four, you do an extracardiac or lateral tunnel Fontan. So, in 2023, now, when you have an infant with tricuspid atresia in a reasonable-sized RV cavity, are you actually pursuing and advocating for a Björk modification in the current era?

Mr. Klemm:

At the moment, at this day, we are still using the staged TCPC procedure. But due to the data we've shown, we are thinking about changing the procedure for this subset of patients which are highly selected, but we are not there yet. We are still discussing.

Dr. Hickey:

Okay. Thank you. Congratulations.

Mr. Klemm:

Good.

Unidentified Speaker 1:

What do you guys think? Are you guys going to try this after seeing this data? Yeah. Raise your hand if you're going to try it.

Unidentified Speaker 2:
Can I ask a question? How do you know - and this is because I've never done that procedure - which are going to be pulsatile, and which are not?

Mr. Klemm:

You don't. Also, it's difficult to say. If you see no impact on the VSD closures and we see no impact on the RV cavity, most likely those patients who have pulsatility have the factors of low pulmonary artery pressure and equally sized and sufficiently sized pulmonary arteries.

Unidentified Speaker 2:

Yeah. So, I guess that's my—

Mr. Klemm:

[crosstalk] have to look at those factors.

Unidentified Speaker 2:

Yeah. That's my point, that you don't know. So, the vast majority of them weren't pulsatile, right? The only group that showed better are the ones that are pulsatile. So, until you can figure that out, I don't see how it would be an improvement. Go ahead.

Dr. Christian Brizard (Melbourne, Australia):

Christian Brizard from Melbourne. It's the same debate as biventricular repair versus Fontan, the quality of life versus the risk. Here you are. It seems that you're promoting the quality of life of the very few that have survived. We know from the Fontan registry in Australia and New Zealand, where we have more than 200,000 patients now, that the Björk prognostic compared to TCPC is very, very different. So, you're trying to introduce a solution that has proven to have generated a much higher mortality risk in the long term for the very small benefit in terms of exercise ability. Reinventing the wheel after 20 years of developing the modern Fontan concept is a bit controversial, I would think. And also, the work from the Fontan registry demonstrates that the most efficient way to have an effect on the exercise ability in Fontan is the physical training, the diet, especially on tricuspid atresia. What do you think?

Mr. Klemm:
Let's take it back. We look at the pressures, and we look at difference between TCPC and the Fontan-Björk. We see that TCPC had much lower pulmonary artery pressures. However, on the other side, when we look at the exercise data, we've seen better exercise data in patients with the Fontan-Björk, and this is probably mostly due to the-- in the pulsatile group, to the better fusion or adaptive fusion into the pulmonary vascular bed as well. And I don't think that we can change it for all patients. We only can describe what we've seen in the past. Those patients with the right physiology in very selected cases can have a proficiently good quality of life with the Fontan-Björk. However, we're still having patients with a high rate of tachyarrhythmias of the reintervention rate due to the stenosis of the RV connection as well. So yes, I agree. It's very controversial. And we don't know if we're going to change our setup at the moment, but it was very, very interesting to look at the data and see the difference in those patients when we just look at the exercise capacity.

Unidentified Speaker 1:

All right. Very quick question, multiple choice.

Unidentified Speaker 3:

Yeah. Do you think that is related more to the pulsatility or the low CVP that we achieve with the valve in the tricuspid position? In the [inaudible] normally after [inaudible], now we are pushing to recover the right ventricle, and then we achieve a lower CVP. The pressure in this inferior vena cava is much less than when you have the Fontan procedure. And this can explain the better result, especially with the PLE. Thank you.

Mr. Klemm:

To be honest, I don't know. Honestly, I really don't know. If you look at the data of 11 patients, to answer this question is just-- the sample set is too small.

Unidentified Speaker 1:

Okay. We're going to move on.

[applause]