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Key Words: Fontan palliation, single-ventricle palliation, Fontan failure, Fontan takedown

Discussion

Presenter: Dr Supreet P. Marathe



Dr Damien J. LaPar (New York, NY).

Dr Marathe and his colleagues from Australia and New Zealand present a 43-year experience with a very complicated patient cohort: those with Fontan failure who ultimately undergo Fontan takedown. The authors' centers certainly have an extensive surgical as

well as published experience with the Fontan operation, including more than 1500 patients, and they have a leading Fontan registry that has once again served an important purpose in capturing and providing longitudinal outcomes for patients undergoing single-ventricle palliation. For this

analysis, the authors have examined the group of patients that have vexed almost every congenital heart surgeon in their practice to provide some insight into outcomes following surgical strategies for Fontan failure.

The authors have analyzed 36 patients who underwent Fontan takedown, which represents an overall incidence of 2.3%. The key findings of this analysis include the incidents of Fontan takedown over time that is nearly unchanged by surgical era, acceptable pre-Fontan hemodynamics, a median time to takedown of 26 days, and a high mortality rate of 44%. Certainly, these characteristics represent the various clinical challenges that patients represent for all surgeons. Based on these findings and their analysis, I do have a couple questions for Dr Marathe, which I'll ask one at a time.

First, based on your data, you report that the most common indications for Fontan takedown are low cardiac output and intractable pleural effusions, both of which often present in the immediate and early postoperative periods. However, the majority of takedowns in your series occurred between 3 weeks and 6 months following Fontan. Considering that many have argued that outcomes for failing Fontan are improved with as early takedown as possible, how do you think the timing of Fontan takedown in this series ultimately impacted the long-term fate for these patients?



Dr Supreet P. Marathe (Brisbane, Australia).

Thank you, Dr LaPar, for your question. I would like to point out that the group of patients who had a takedown between 3 weeks and 6 months had more or less a common theme. All of them presented with intractable effusions, and they under-

went multiple interventions—multiple chest tubes and pleurodesis, and they were sat on, and we did not take the Fontan down and then they ultimately died. So that was the most important take-home message for us, that we probably should not wait on these patients. The fact that they are developing these effusions points toward subtle markers that something is not right. Even if it looks fine on echo, it looks optimum on cath, the numbers are all okay, but the circulation is probably not right. It probably indicates that we should not shy away from taking down the Fontan, and as we can see, there are many other options after we take down the Fontan which we can provide them with.

Dr LaPar. Thank you. Also, related to the high frequency of low cardiac output in these patients, what is your center's experience? I didn't see it reported in your series for fenestrations at the time of Fontan.

Dr Marathe. We did not identify the presence of a fenestration to be predictive of takedowns. There was no difference regards to fenestration between the ones who died or had a transplant and the ones who did not.

Dr LaPar. Okay, great. My second question centers upon those patients undergoing takedown: Your late Fontan

failures for protein-losing enteropathy or plastic bronchitis. In this experience, while it's a relatively small number of patients, did the Fontan take down in these patients ultimately improve those symptoms?

Dr Marathe. Yes, they did.

Dr LaPar. Your data also demonstrate that 22% of the Fontan takedown patients ultimately underwent either re-Fontan or one and a half or 2-ventricle repair after takedown, whereas 31% of the patients remain with an intermediate circulation. So, my question is: After Fontan takedown, what is your center's approach or protocol to re-evaluating these patients for either surgical repair or potential re-Fontan? Do you repeat the cath at a certain time interval, or are there certain hemodynamics or characteristics that you look for that might push you toward operating on these patients?

Dr Marathe. So, because there are several member institutes, I would not say that there's a common protocol with regards to evaluation. But, in general, if we talk about the philosophy, we should definitely be doing the cath if there's something obvious. I think we should completely re-evaluate whether these patients really need to be along the single-ventricle pathway, because as we know, around the world, aggressive biventricular repairs are being pursued. We only had 2 biventricular repairs and 1 one-and-a-half ventricle repair in our series, but someone who had a single-ventricle palliation in the previous era might probably be okay with a biventricular circulation in the current era. So, we would also like to do magnetic resonance imaging to evaluate whether these patients are actually suitable for a biventricular repair; I think that should be the first step. The second step: If not, maybe we should consider them for a re-Fontan, and there are many ways; obviously, we can do aggressive atrioventricular valve repairs, we can optimize the pulmonary arteries, and there are many ways where we can make them good Fontan candidates if you don't find any obvious reason. The third option is, if we still are forced to stay with an intermediate circulation, that is where we wait.

Dr LaPar. My final question is: Do your data, after analyzing it, provide you or your group any insight into which patients ultimately might do best? That is, survive or ultimately achieve a re-Fontan or a complete or partial repair after the Fontan takedown?

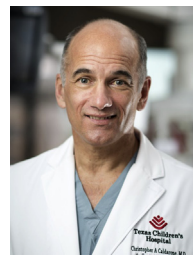
Dr Marathe. So, the aim was really this, we wanted to find out is there a common theme in them? Is there a basic diagnosis? Is there a predominant ventricular morphology which points toward these problems? But there's none, and the only difference is the timing of the takedown. Now, all this points to the fact that are there other things that we haven't looked at that we need to think about. They possibly could be subtle technical imperfections where the numbers are okay, but the flows are not optimal. So, what is the role of flow studies and whether we should consider doing them in these patients.



Dr Vaughn A. Starnes (*Los Angeles, Calif*). Did you notice any era differences about when people took Fontans down? It's a study over 43 years; did you notice in the last decade that people took them down quicker? In the earlier decades, did people go on to die?

Dr Marathe. We did not find such a difference, no. The *P* value is trending toward significance, but not actually significant, though.

Dr. Starnes. Thank you.



Dr Christopher A. Caldarone (*Houston, Tex*). The rationale for an early take down in a patient with low cardiac output syndrome might be different than one with prolonged pleural effusions. I didn't see that your analysis was stratified that way, but the message that you need to take down quickly may be more urgent in a low cardiac output state than it is in prolonged pleural effusions because, as you know, many patients have prolonged pleural effusions that resolve. So, how do you reconcile those 2 aspects of your analysis?

Dr Marathe. Absolutely. I cannot agree more. The ones who have early takedown, that is really a forced decision. We really don't have any other option. They have such low cardiac output, high inotrope requirement, it's really a last-ditch effort where we have to take them down. But this other group that we are talking about, we think they are fine, and we can potentially sit on them, but what our study shows is that we should probably not, and the most prudent way to go about them is to actually take down the Fontan.