Discussion to: Outcomes of Pediatric Heart Transplantation in Children with Selected Genetic Syndromes

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Dr. Robert Dabal (Birmingham, AL):

Thanks to you Dr. Roy and Dr. Mitchell, the planning committee of the AATS, for this opportunity to discuss this paper. Dr. Alsoufi and his team have executed an interesting analysis that correlates pediatric transplant outcomes in the UNOS and PHIS databases in regard to congenital patients with genetic syndromes undergoing transplantation. Given the scarcity of suitable donors this is an important discussion as we are shepherds of these organs and have a responsibility to ensure that patient factors don't limit the longevity of these grafts. Based on your presentation, Dr. Alsoufi, I have several questions. The first is a question about the heterogeneity of the groups. So not all syndromes are the same so I am curious if you broke down the analysis by syndrome and I know there are limitations based on the sample size but I’m also curious why heterotaxy syndrome was excluded because these really can be the most technically challenging transplants that we do as congenital heart surgeons. And it seems like there were a significant number of these patients that underwent transplantation.

Next, I'd like to know the breakdown-- actually, you said the breakdown of patients who underwent transplant for cardiomyopathy versus end-stage congenital heart disease, and it's pretty clear that the patients with end-stage congenital heart disease fare worse than patients who have cardiomyopathy. And so, I'm curious what your thoughts are and how this impacted the data.
Next, I find it interesting that the five-year survival was similar in both groups despite the fact that the short-term results in the syndromic patients were worse. So, you had a higher mortality, you had almost double the rate of dialysis and a statistically longer length of stay. So, I'm wondering why with worse short-term results for the syndromic patients, why do we see no difference in their five-year survival?

And then my last question is just a philosophical question that I'd like to get your thoughts on. So your analysis is excellent and it's very creative I think to link the two databases, but it seems to me that there would be added benefit if you could link this to the STS database, and I'm not sure if there's presently a way for us to do that. But it seems like as a society, if we want to go forward in looking at longer-term outcomes, if we could figure out a way to link UNOS and PHIS and STS results, I think that we would probably have more granular data. We certainly could find out more about the perioperative course and any complications if we could somehow link the STS database to these sorts of studies. So, thanks again for this opportunity to discuss this important paper.

Dr. Bahaaldin Alsoufi (Louisville, AL):

Well thank you, and I hope you stay here because I will not remember all these questions.

Dr. Dabal:

I'm sorry, a lot of questions.

Dr. Alsoufi:

The heterotaxy, it's easy. The heterotaxy, I definitely wanted to look at it. The issue is that the ICD code has changed and you can only have reliable heterotaxy with the ICD 10, not 9, so that would have pushed our study to about 2015, 2016. So, I wanted to have wider range, more patients, so I had to exclude heterotaxy. However, understanding the limitation, we have examined that, and we sent that abstract already, studying only heterotaxy. The bottom line, the same. No change. But I could not merge it because otherwise the timeline-- and it's just again because of the coding for heterotaxy, ICD 9 and 10 were different. But you are right, it's interesting that I was able to find within that study period 150 heterotaxy patients who underwent transplants. So again, hopefully we'll present that in an upcoming meeting. We have already sent the abstract. But bottom line, they also do the same.

I remember your last question and I completely agree with you about the value of linking to other databases. I wish Jeff or Ram are here to comment more on it.
but I know there have been talks about it and then that will definitely be more helpful because I think we can get also more granular data from this STS and obviously then link that to get the long-term outcomes from UNOS. I am not sure I have-- I agree with you that the early outcomes are very different, like 19 days and 21 days statistically different. I don't know if from the clinical perspective that are really different. Mortality 3-4%. That was not statistically different. But dialysis, even 8 and 5% was not necessarily statistically different. Might be clearly more but I'm not sure it's significant clinically enough to say whether they should do worse. I'm missing one question, I think.

Dr. Dabal:

So, the last question was just with regard to individual syndromes, and I'm [crosstalk] the numbers are a limitation, but they're not the same and so I'm curious if—

Dr. Alsoufi:

Yeah, and we tried as much as possible to look at it, and I showed it here. You might think, okay, well do the DiGeorge syndrome patients do slightly worse? I don't know but statistically was not significant, but we looked at that as you can see.

Dr. Dabal:

Great. Nice presentation. Thank you.

Unidentified Speaker 1:

We have time for one quick question.

Dr. Jonathan Chen (Philadelphia, PA):

Jonathan Chen from Philadelphia. The overall arc of these two presentations is really fantastic and it puts into light for us data versus religion, right, that we've carried for so long. This is just-- it's not really a question but more of a plea which is that the title of your abstract uses the word 'major', and in all of your limitations and the conclusion's part, you say 'selected'. And the request or urgent, hopeful request is that the ultimate version of this is 'selected'. Because the truth is that these are a very selected group of genetic abnormalities that we know of, and parent groups, that's the overlap here, the third rail of trisomy 18 and 13 and genetic syndromes is that the parent groups become vocal advocates, and we become, especially in the transplant forum as Bob is saying, we are stewards of these donor organs. 20 years ago, the question for Stanford
was can you transplant a kid with Down’s? And they got sued. And so now all we-- and we all do. And the question now will be outside of the 10 genetic syndromes you're talking about, we'll have families who will come to us waving the paper and saying, "Major genetic abnormalities do not impact on mortality. You should take my child who has X. That's not one of those 10." And so, the plea would be emphasizing the word 'selected' and not 'major'.

Dr. Alsoufi:

Well, that's very easy Jonathan, because as is my tradition, I'm always late writing the paper, so that's an easy task.

Dr. Chen:

Great. Thank you very much.

Dr. Alsoufi:

Thank you very much.