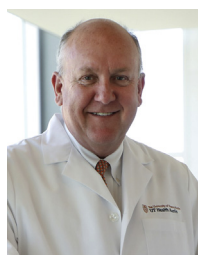


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Discussion

Presenter: Dr Anusha Jegatheeswaran



Dr Charles D. Fraser, Jr (Austin, Tex).

It's a great privilege to discuss yet another outstanding presentation by Dr Anusha Jegatheeswaran on the subject of anomalous aortic origin of a coronary or AAOCA. Anusha, just 1 year ago at that AATS plenary session, you brought us important information from the

CHSS AAOCA Registry. Important points from last year include ischemia or sudden death is more frequent in anomalous left coronaries, but it also occurs in anomalous right coronary arteries; patients with clinically significant anomalous right coronary arteries are more likely to have a long intramural segment; and patients undergoing surgery for AAOCA were thought to have at least a 10% risk of needing reoperation. Last year, we talked about you bringing us more information about the operations, what people are doing, and what are our patient outcomes.

So it is great to hear from you again on this very vexing subject. Unfortunately, this year you've brought us some really sobering information, in my estimation. Of the subgroup of 395 primary surgical patients of the original 682 patient cohort, I'll revisit some of what I heard of your important findings. Clinical observation of the patients and expectant management is not always successful. Of the 287 patients who did not undergo surgery, 9 died, including 6 with anomalous right coronary arteries. As you just related, surgery has risks. Despite the fact that study centers were exclusively congenital programs with operations being performed by experienced, largely board certified, congenital heart surgeons, there is significant morbidity and mortality associated with surgery for AAOCA.

In your study, 26 patients had postoperative ischemia and 3% of patients had new postoperative, ischemia. A total of 34 patients developed new, iatrogenic AI, and this appears to be related to commissural manipulation. Two percent of patients who had normal function going in to the operating room had a decreased EF coming out. Of the patients who are operated on without symptoms (eg, primarily for morphology), 51% did not undergo provocative testing before surgery. There were 42 reoperations in thirty patients. Fifteen patients required reoperation for a coronary artery misadventure. Freedom from any reoperation at 7 years was only 90%, and 4 patients died, 3 of whom had elective operations. So, we still have a long way to go with this challenging subject. In deciding on whether to operate on a given patient, it's easy if the patient had symptoms or sudden cardiac death. The tough ones are the asymptomatic patients. Among many important findings, you've documented that congenital heart surgeons are operating on the majority of patients solely based on morphologic determinants. Should we be more tempered in our decision-making after your data?



Dr Anusha Jegatheeswaran (Toronto, Ontario, Canada). You are correct.

From these data, we can see that the many surgeons operate on patients solely for morphology. Part of the reason for this occurring is that the guidelines suggest that patients with interarterial left AAOCA should undergo

surgery, without the need to demonstrate ischemia via testing, even in asymptomatic patients. From the data we presented last year, we know that a high proportion of patients have an interarterial course, perhaps rendering this a less useful discriminating feature.

What I would advocate for is the need for a collaborative team including surgeons, cardiologists, radiologists, nursing, and social work, all with a specific interest in AAOCA, to manage patients at each institution. We should be making decisions together regarding whether or not a patient needs surgery and should be maintaining a critical eye on whether the patient has been adequately evaluated. Especially because we increasingly coming to understand the risks involved with surgery. One approach would be to serially evaluate asymptomatic patients without proceeding to surgery immediately. However, we still don't know is the natural history of various lesions without surgery. This would allow us to determine how to balance the risk of surgery with the risk of ischemia.

Dr Fraser. I might also suggest that we need to involve, in an objective way, the patients or the parents in this decision-making. As per the above, it seems that our congenital heart surgeon colleagues have been slow to adopt the guidelines that we've developed. Specifically, we are not

adhering to the recommendation of preoperative provocative testing. Why is this, and should the guidelines be revisited?

Dr Jegatheeswaran. As I mentioned, for a patient with an interarterial left AAOCA, the guidelines seem to suggest that you don't need testing and can proceed to surgery based solely on morphology. However, more concerning was the fact that there a large proportion of patients within our cohort with right AAOCA who did not have preoperative testing. One thing to note is that our cohort starts in 1999 because of the retrospective aspect component, and the various guidelines were not available at that time. As such, physicians seem to have been doing what they felt like at that time.

However, we are doing this repair to get rid of ischemia. So, first, it would be nice to know if there is ischemia in patients who are asymptomatic, and second, it would be nice to follow up and see whether we really corrected ischemia through surgical repair.

With respect to the guidelines, I think the information from this study can add some useful information. I think we still have a lot of ways to go in terms of having high-level evidence that will allow for Class A recommendations. What we really need is a protocol for assessment and management of patients before we can provide definitive answers.

Dr Fraser. As I did last year, I would like you to offer another management opinion based on a theoretical, but a realistic scenario: A 12-year-old girl is incidentally found to have anomalous right coronary artery from the left sinus and an 8-mm intramural course behind the left-right commissure. All provocative testing is normal. Would you recommend surgery, and, if so, what operation?

Dr Jegatheeswaran. We don't have the answer to a very important question yet, because we still don't know the risk of ischemia and sudden cardiac death, in comparison with the risk of surgery. As such, I don't know if I would be able to provide a definitive answer because this study was not intended to answer that question. However, using our current understanding, this patient does have a high-risk feature for ischemia and sudden cardiac death that we elucidated last year, that of a long intramural course. We know that in that our cohort there were patients who had a long course, in the setting of right AAOCA, who had sudden events. So, although patients with right AAO-CAs were previously thought to have a benign lesion, this is no longer the case. However, other than an appreciation that a long intramural course is a risk factor, we do not yet have a cutoff, and most centers would not operate on patients with right AAOCA without some kind of positive testing. In the setting of negative testing, centers doing a large number of these cases often perform serial evaluation.

What we really need before we can answer this question is more research, more protocolized studies, and international collaboration, which will allow the capture of thousands of patients. This is required because the sudden cardiac event rate is so low.

In the interim, the pros and cons of surgery need to be balanced and discussed with the family. This patient would require an unroofing with careful resuspension of the commissural post or creation of a neo-ostial window, which may decrease the risk of new aortic regurgitation.



Dr David M. Kalfa (*New York, NY*). Congratulations on your work. Your work shows that a surgery doesn't always treat or prevent coronary ischemia and can even sometimes lead to ischemia. You also stated that coronary unroofing was by far the most frequent technique applied to these patients.

I strongly believe that, from a technical standpoint, a coronary unroofing procedure needs to be done in an adequate and proper way to be effective. You need to be aggressive in terms of unroofing the coronary artery to really "open" the coronary ostium, open the commissure-related or fibrosis-related obstruction of the coronary ostium. So my question for you is do you have any granular data about how these coronary unroofing procedures that you included in your study were performed? Do you have granular data coming from the operative notes? And, if yes, did you analyze this data in your study?

Dr Jegatheeswaran. For this study, I personally reviewed all the operative notes, which totaled just under 400 in number. The problem that we have with all studies of this nature, where we are using retrospective data is that we do not always have the granular data we need. Sometimes surgeons will just write "We did an unroofing." They do not always clearly write whether they simply resected a small flap at the ostium or whether they have tacked the intima after takedown. As such, although we reviewed the data we collected carefully, what we really require is, as you mentioned, granular data collection with all those details, which likely needs to be prospectively collected.



Dr Gosta Pettersson (*Cleveland, Ohio*). Thank you for presenting this sobering data. I do a fair number of reoperations, and even in this category of patients, there is a number of patients who had previous operations for their anomaly—in the last one we did the entire root had been destroyed by a surgeon who had tried a minimally invasive approach to unroofing of an anomalous right coronary only to recognize that the artery did not after all did not have an intramural course, and ending up having to reconstruct the root and bypass both coronary arteries.

We need to separate the different anomalous carefully, take them one by one and analyze the approach risk, indication for intervention and choice of intervention for each one. We are very critical when it comes to preoperative evaluation of these patients and require not only symptoms but functional testing and use all available technology, including intravascular ultrasound and fractional flow reserve to prove significance. We also try to convince patients preoperatively to submit themselves to postoperative examinations as well. I'd like to congratulate you on this very important study, again demonstrating that we still have a long way to go.

Dr Jegatheeswaran. We have a long way to go.



Dr Sabine Hellevi Daebritz (*Zürich, Switzerland*). I've done a lot of those cases, and I used to do a thoracic artery bypass in addition for security for the postoperative period being aware that this will occlude after 6 months if it's not needed. There is obviously another peak in the fourth decade in those patients;

they never present with myocardial infarction and they often have symptoms but you cannot reproduce ischemia. These patients are often reluctant to undergo surgery. Can you comment on implanting a defibrillator because, obviously, they are not prone to infarction but to sudden cardiac death because of rhythm problems due to ischemia.

Dr Jegatheeswaran. With respect to our cohort, all patients were aged less than 30 years of at enrollment, with mostly medium-term follow-up, a median of 2.8 years, and we only had 3 patients in our cohort who had an implantable cardioverter defibrillator. Two implantable cardioverter defibrillators were for prevention based on scar, and one was for secondary prevention after an event that occurred after the repair. As such, unfortunately this Registry does not contain adequate data to provide you with an answer regarding whether or not patients should receive an implantable cardioverter defibrillator.



Dr Pravana Sinha (*Washington, DC*).

I have a question specifically about the intramural anomalous coronaries. In your data you show that commissural manipulation leads to more AI. Do you think that in itself is a surrogate for the anatomic variation where the intramural course runs below the commissure? Rather than the message that you should avoid commissural manipulation, should the message be that

that's a different anatomy that requires commissural manipulation?

Dr Jegatheeswaran. I don't believe that should be the message. When you have a coronary that is running behind the commissure, you have to decide what surgical strategy you are going to use. The most common option is unroofing, with commissural take-down and hopefully with resuspension. Some surgeons don't always resuspend, especially if the coronary is just at the top of the pillar. After reading almost 400 operative notes, it is clear that different surgeons use different strategies with respect to resuspension.

Our study simply found that there is an association between commissural takedown and new AI. At the present time, we do not know what the right repair is. We do not know the long-term outcomes for the various repair strategies that could be used in this setting, such as neo-ostial windows and reimplantation after transection from outside the aorta without the use of a button. As a result, we still do not know which one has the best outcomes in this setting.

Dr Sinha. I mean, the surgeon should not have to balance between doing a complete unroofing and commissural resuspension because one will lead to more AI, the other will lead to more ischemia. So we have adopted a technique that we presented last year. It's a small series of 26 patients because I believe that no matter what the anatomy is, when you unroof the coronary, there is a decrease in the commissure that is equal to the diameter of the intramural segment. So we prophylactically suspend the commissure in 100% of our cases and at the same time achieve complete unroofing of the coronary.



Dr Antonio F. Corno (*Leicester, United Kingdom*). Don't you think that instead of only sharing the outcomes of surgery, you should also collect all possible data from all the patients with myocardial ischemia in the presence of anomalous coronary artery? These are the data we're missing,

the common denominator, and there are alarming data reported from the pathology registries.

Dr Jegatheeswaran. I would definitely agree with that. Our Registry does collect data from all patients with AAOCA, whether or not they're medically treated or surgically treated. The focus of this study was surgical risks, but we do also have that data. However, I will make this plea: We need everyone's data, especially medically treated patients who may not be seen by surgeons, for us to figure out what is happening.