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Commentary: I guess I'm just confused…isn't this information sobering?

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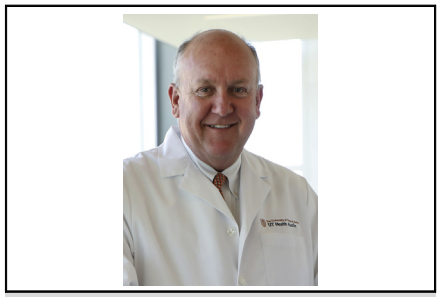

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CENTRAL MESSAGE

AAOCA is being diagnosed in many people; some are symptomatic, but most are asymptomatic. Many of these patients end up undergoing cardiac surgery at a time where indications for operation are not completely clear. Are these operations safe? What are the current complication rates? We really *have* to know, and the data suggest reason for more attention.

I have been anxiously waiting for the article by Jegatheeswaran and colleagues[1] concerning the outcomes after surgery for anomalous aortic origin of a coronary artery (AAOCA) to be published. I was an invited discussant for the oral presentation of these data at the annual meeting of the American Association for Thoracic Surgery almost 1 year ago. At the time, I thought these data needed to be published as soon as possible. In fact, I commented to several colleagues that the accompanying article merited an expedited review. I found the information concerning and critically important to what have become very frequent (and lengthy) conversations with patients and families about AAOCA surgery. Maybe I was confused, but I thought we really need to pay close attention to what is going on with this subject.

The study is not perfect, but what study is? It is a retrospective look at a voluntary registry from 45 centers contributing to the Congenital Heart Surgeons Society. Thus, there must be some selection bias, but I argue that this may influence the data toward a more optimistic than practically realistic viewpoint. More to come on that. One criticism of the article was that it does not compare outcomes between surgical and expectant (nonoperative) management. That was not the intention of the study, and, of course, to make such a comparison, one would have to agree to nonoperative treatment of patients with the

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substrate for ischemia that will never happen (nor should it). The follow-up is short (~2 years), perhaps the conclusions could have been reworded, and so forth.

So, was I confused about how important these data are? I don't think so. The patients in this study were enrolled from congenital heart surgery centers, and the participants should have been familiar with guideline recommendations for AAOCA care, but many of them did not follow guidelines for assessing patients for provocative evidence of ischemia.[2] Only 165 of the 395 operated patients had provocative preoperative ischemia testing. This is concerning. How are we to know that we are doing the right thing for our patients if we don't agree on how to assess candidacy? As a comparator, consider the evolution of indications for coronary artery bypass grafting and the mass of information available to surgeons and cardiologists to assess risk and predict outcomes. Even if AAOCA guidelines are imperfect and incomplete, they are based on best available evidence. So why have congenital heart surgeons and their colleagues in cardiology largely disregarded the guideline recommendations? Should a surgeon ever take an asymptomatic patient with AAOCA to the operating room without provocative ischemia testing?

The operations for AAOCA have real risk, maybe more than we think. As previously noted, the study cohort is biased. The registry is voluntary, and the patients should have been operated on by congenital heart surgeons. These are the same surgeons who perform arterial switch operations, perform neonatal Ross–Konno procedures, reimplant anomalous left coronary arteries from the pulmonary artery, and on and on. These surgeons should be intimately familiar with the risks and nuances of manipulating coronary ostia. We know that in the real world, AAOCA surgery is also being performed by cardiothoracic surgeons without specific congenital heart surgery training. Maybe the data in the current study underestimate the risk profile of AAOCA operations in general practice. In the hands of what should be the most qualified surgeons, members of the Congenital Heart Surgeons Society, asymptomatic patients undergoing elective surgery without preoperative evidence of ischemia died. Patients who did not have evidence for ischemia before surgery developed new ischemia. Approximately 10% of patients undergoing an operation developed new aortic insufficiency. This was importantly related to commissural manipulation. Patients going into surgery with normal heart function developed new myocardial dysfunction.

Consider a theoretical, but very realistic (and frequent) scenario. A 12-year-old child undergoes a screening physical examination to be cleared for school sports. The examination results are normal. The hypervigilant parents have expressed concerns about lay-press articles on sudden cardiac death in athletes and request an echocardiogram. The echocardiogram reveals anomalous aortic origin of the right coronary artery from the left aortic sinus of Valsalva with a retro-commissural, short intramural course. A consultant pediatric cardiologist will not clear the child for sports. The parents are very anxious, and the child is essentially crippled by the diagnosis of AAOCA, so surgery is considered. Shouldn't these parents know that their child has a chance of dying, having a coronary ostial misadventure, or developing new aortic valve insufficiency after an operation that we don't even know is indicated? Maybe I'm confused, but this is serious stuff: prophylactic operations where guidelines are disregarded and patients die. We had better pay close attention to these "imperfect" studies.

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