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ORIGINAL ARTICLE

Utility and Importance of New Echocardiographic Screening Methods in Diagnosis of Anomalous Coronary Origins in the Pediatric Population: Assessment of Quality Improvement

Poonam P. Thankavel · Matthew S. Lemler · Claudio Ramaciotti

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Abstract Anomalous aortic origin of coronaries from the contralateral sinus (AAOCA) is rare but an important cause of cardiac death in the otherwise healthy young athlete. This necessitates prompt, accurate identification; transthoracic echocardiography (TTE) remains the primary screening tool. Assessment of accuracy is difficult since the true prevalence of the disease is unknown, with estimates at 0.3–1.07 %. The incidence by TTE remains much lower, between 0.09 and 0.17 % even with sophisticated equipment and a high index of suspicion. Our goal was to incorporate two new screening views to our standard TTE protocol and assess improvement in diagnosis of AAOCA by TTE in our laboratory. Recently (2011), we incorporated two new screening methods to standard protocol. The parasternal short axis sweep is extended to visualize the anomalous segment arising superiorly from the ascending aorta before exiting the root at a site close to a 'normal' origin. Secondly, the anomalous, interarterial coronary demonstrates an anterior, steep-angled course visualized in the parasternal long axis between the aorta and pulmonary artery. The echocardiogram database was searched for patients newly diagnosed with AAOCA in 2010 (prior to incorporation of new methods) and 2012. AAOCA incidence in our patient population improved from 0.02 % (2010) to 0.22 % (2012), age range from 4 days to 17 years. Teenagers and symptomatic patients with anomalous right coronary origin (6) underwent additional confirmatory imaging, and three underwent surgery. One patient with anomalous left coronary origin underwent surgical repair. Addition of the screening views can significantly increase the sensitivity of TTE in diagnosing AAOCA in the asymptomatic patients. We propose that these views be incorporated into the standard TTE evaluation of coronary arteries.

Keyword Anomalous coronary · Quality · Echocardiogram

Introduction

Anomalous aortic origin of a coronary artery from the contralateral sinus (AAOCA) is the second leading cause of sudden cardiac death in young athletes, and often exists in isolation [9]. The true prevalence of the anomaly is unknown, with a prospective angiographic study [1] estimating it at 1.07 % in the general population; however, the importance of these malformations is underscored by its high mortality rate in the otherwise healthy young adult [9]. While no current consensus exists on the management or natural history of asymptomatic patients, a new multicenter registry of these patients has been established to obtain the needed data [11]. The first step to accurately identify these patients, particularly in the asymptomatic setting, requires the development of reliable diagnostic tools. Transthoracic echocardiography (TTE) is a crucial part of initial cardiac evaluation, but its sensitivity, specificity, and accuracy remain unknown [8]. Even with a high index of suspicion, the TTE incidence in a large prospective pediatric study [5] remains at 0.17 %, which is significantly lower than angiographic estimates. This discrepancy is most likely due to poor diagnostic TTE accuracy. In an attempt to improve the accuracy and

P. P. Thankavel (✉) · M. S. Lemler · C. Ramaciotti
Division of Pediatric Cardiology, UT Southwestern Medical Center, Children's Medical Center of Dallas, 1935 Medical District Dr, Dallas, TX 75235, USA
e-mail: poonam.punjwani@childrens.com

reliability of TTE, we recently described two new screening methods [12, 13] to evaluate AAOCA. These screening methods were incorporated into the routine TTE evaluation in our laboratory in 2011. The goal of this study was to retrospectively assess the impact of these new policies in the TTE diagnosis of AAOCA in our laboratory. We compared the incidence of AAOCA before (2010) and after (2012) implementation of new screening techniques as an indication that routine TTE can be a more useful tool to identify this rare pathology.

Methods

Patients

This retrospective chart review was approved by the Institutional Review Board. The echocardiography database was reviewed for the diagnosis of anomalous coronary artery from January 1 to December 31, 2010 and from January 1 to December 31, 2012. The review included all transthoracic echocardiograms performed at Children's Medical Center, Dallas, in patients between 0 and 21 years of age. The anomalous coronary cohort was defined as anomalous coronary origin from the contralateral sinus; patients with co-existing congenital heart disease (with the exception of patent foramen ovale, small ventricular septal defect and/or a small patent ductus arteriosus), anomalous left coronary artery origin from the pulmonary artery, or single coronary artery were excluded. Patients referred to our institution specifically for evaluation of AAOCA were not included, as our intention was to assess the ability to diagnose AAOCA during routine exams. The total number of patients undergoing transthoracic echocardiograms was obtained, including those with congenital heart disease. The chart of AAOCA patients was reviewed for demographics, the indication for the TTE, and whether any additional confirmatory test was performed.

Transthoracic Echocardiograms

All echocardiographic examinations were performed using Siemens Sequoia C512 ultrasound equipment (Siemens Medical Solutions USA, Inc, Mountain View, CA, USA) or Philips iE33 (Philips Medical Systems, Bothell, WA, USA) with 10-, and 8-MHz transducers. All images were reviewed on a Syngo Dynamics workstation (Siemens Medical Solutions, Ann Arbor, MI, USA). The standard coronary artery evaluation in our laboratory includes two-dimensional (2D) and color Doppler imaging of the coronary arteries in the parasternal short axis view at the level of the aortic root. The new screening methods implemented in 2011 added extending the parasternal short axis superiorly into the ascending aorta in 2D with and without color Doppler, and the visualization of the proximal left main coronary course in the parasternal long axis sweep between the aorta and pulmonary artery, with careful attention to the angle of the proximal course [13].

Results

Prior to the implementation of new screening techniques (2010), 5,669 patients underwent TTE, with only 1 diagnosed with anomalous coronary origin (intramural left coronary from the right sinus), thus estimating the incidence at 0.02 %. In 2012, AAOCA was diagnosed in 14 out of 6,428 echocardiograms in children and young adults, improving the detection rate to 0.22 % (Yates' Chi squared test, $p = 0.004$; Table 1). Two patients (not included in the 2012 cohort) were referred specifically for evaluation of AAOCA and were found to have a right coronary arising from the left sinus.

The demographics of patients with AAOCA are summarized in Table 2. Of the 14 patients (age range 4 days to 17 years) diagnosed in 2012, 1 patient presenting with exertional syncope was diagnosed with an anomalous intramural left coronary artery from the right sinus. The anomaly was confirmed by cardiac magnetic resonance imaging (MRI) and by inspection during surgical unroofing. Of the remaining 13, all were diagnosed with anomalous right coronary from the left sinus. Four symptomatic young adolescents underwent additional confirmatory imaging with either cardiac MRI, CT (computed tomography), or catheterization. Two of those had surgical unroofing of the intramural component. The attending cardiologists of the nine remaining patients did not request confirmatory studies at the time of this publication. These patients were all asymptomatic and their decisions were primarily based on patients' young age and diagnosis of anomalous right coronary, the management of which is still controversial. In those patients, confirmatory imaging is planned at an older age. Nuclear exercise stress tests ($n = 4$) were unremarkable.

Table 1 Incidence of AAOCA at our institution in 2010 compared to 2012

Year	Number of AAOCA	Total number of patients	Incidence
2010	1	5,669	0.02 %
2012	14	6,428	0.22 %
			$p = 0.0004$

Table 2 Patient demographics

Patient no.	Age (years)	Sex	Indication for TTE	Diagnosis	Confirmation
1	9 years	Male	Exertional syncope	ALCO	CT scan, autopsy
2	15 years	Male	Hypertension	ARCO	MRI
3	11 years	Female	Palpitations, non exertional	ARCO	
4	16 years	Male	Function evaluation on chemotherapy	ARCO	
5	10 years	Female	Nonexertional chest pain	ARCO	
6	17 years	Male	Family history of early cardiac death	ARCO	MRI, OR
7	16 years	Male	Exertional chest pain	ARCO	MRI, OR
8	4 days	Male	Murmur, small VSD	ARCO	
9	6 years	Male	Murmur	ARCO	
10	4 years	Male	Sickle cell disease	ARCO	
11	1 month	Male	Murmur, small VSD	ARCO	
12	4 years	Male	Murmur	ARCO	
13	17 years	Male	Rule out hypertrophic cardiomyopathy	ARCO	Cath
14	16 years	Male	Fever, rule out endocarditis	ARCO	
15	14 years	Male	Exertional chest pain and syncope	ALCO	MRI, OR

Patient 1 was diagnosed in 2010, subsequent patients diagnosed in 2012

ARCO anomalous right coronary origin, *ALCO* anomalous left coronary origin, *Cath* cardiac catheterization, *CT* computerized tomography, *MRI* magnetic resonance imaging, *OR* operating room, *VSD* ventricular septal defect

Discussion

The goal of our study was to assess the impact of new screening techniques in the diagnosis of AAOCA by routine TTE. We noted an improvement in accuracy of TTE diagnosis by an increase in incidence from 0.02 % (2010) to 0.22 % (2012). While we recognize that not all patients had additional confirmatory tests, in our experience, false positives are uncommon, whereas the pitfall of TTE in this diagnosis is usually false negatives [12]. The incidence in 2010 at our institution is similar to prior years, and the continued use of our new protocol diagnosed 18 new patients in 2013. Since the true prevalence of the disease in the general population is unknown due to its rarity (estimates vary from 0.1 to 1.07 %), evaluation of the sensitivity and negative predictive value of this diagnostic test become problematic [8]. However, TTE remains the primary, and sometimes only, diagnostic modality used to image patients at risk. Therefore, an improvement in TTE sensitivity significantly enhances our ability to understand this rare, but potentially fatal anomaly. Our study supports the notion that the addition of the proposed screening images to routine TTE can achieve that goal. We believe that these techniques can be easily added to every routine TTE without significant increase in the time or difficulty in image acquisition or interpretation.

There are case reports [2, 5] and anecdotal evidence that this anomaly can be missed in patients that undergo TTE

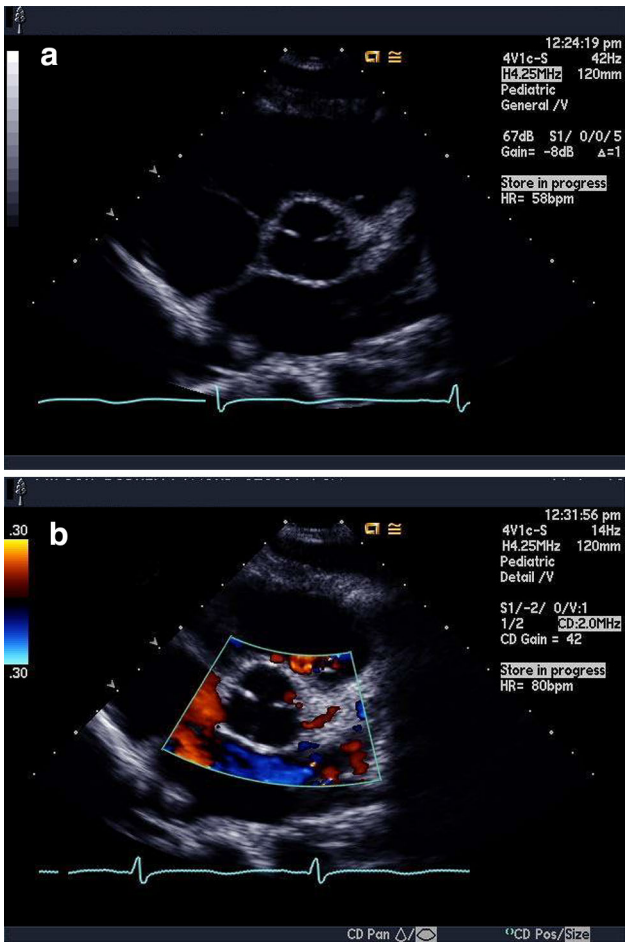

Fig. 1 False negative: normal appearing left main coronary artery in **a** 2D and **b** color Doppler (*red*)

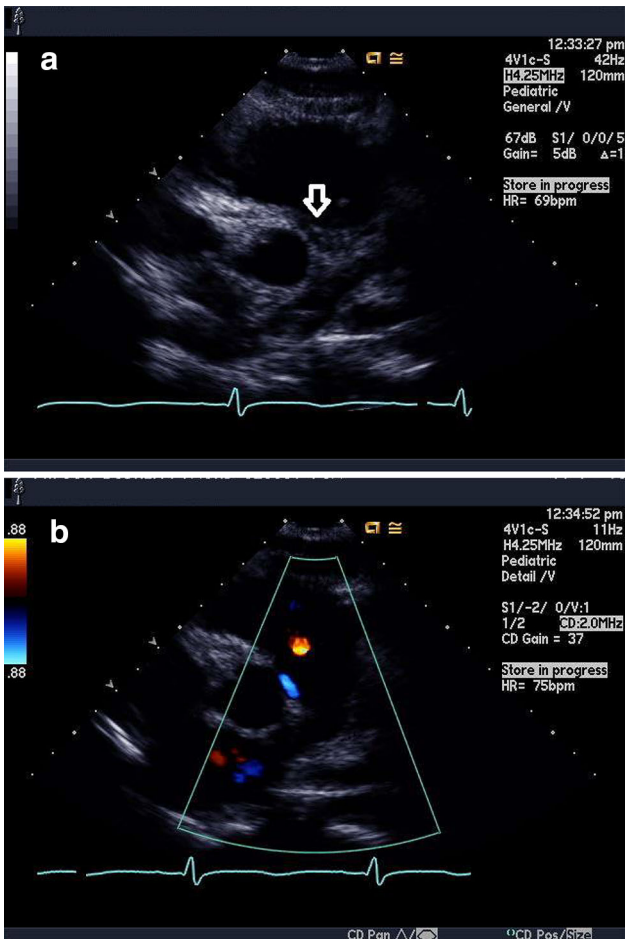

Fig. 2 Superior parasternal short axis (in same patient as Fig. 1) demonstrating anomalous left coronary in same patient in **a** 2D (*arrow*) and **b** color Doppler (*blue*)

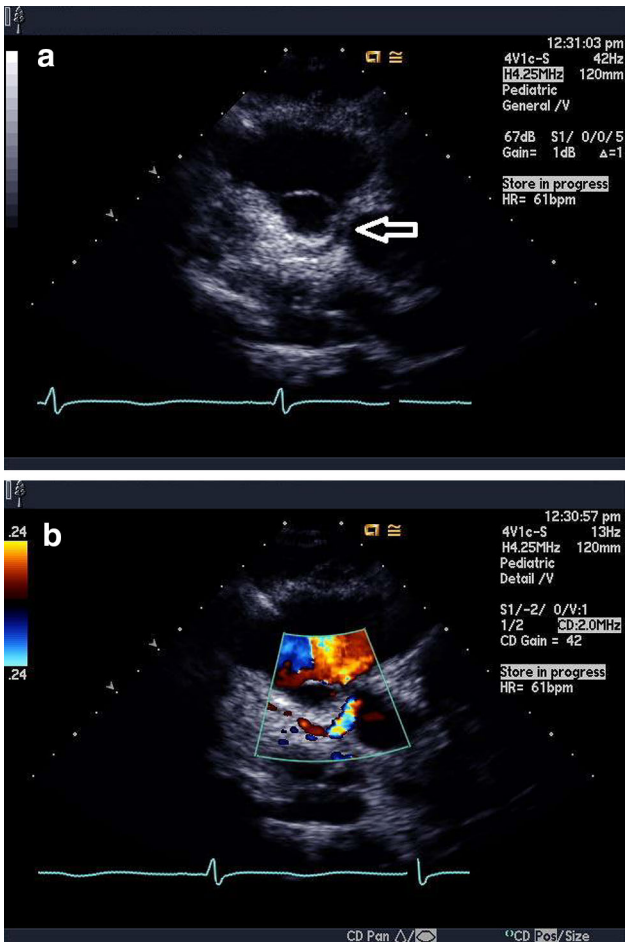

Fig. 3 Parasternal long axis evaluation of anomalous left coronary artery in the same patient with the anomalous course identified in **a** 2D (*arrow*) and **b** color Doppler (*blue*)

evaluation using the routine views. In fact, in most of our patients with AAOCA, we were able to produce images with normal appearing coronary arteries, both in 2D and color Doppler (Figs. 1, 2, 3, 4). Our index case in 2010 had a normal appearing left main coronary artery on initial presentation. He was subsequently diagnosed with an anomalous, intramural left coronary artery from the right sinus on the fourth day of hospitalization while on extracorporeal membrane oxygenation (ECMO) support, and unfortunately, died secondary to severe left ventricular dysfunction and neurological injury. Evaluation of his heart on autopsy revealed an anatomic characteristic that has been mentioned in prior reports [7] the coronary origin was high on the ascending aorta with an intramural, steep leftward course as it traveled toward the left sinus, exiting the aorta such that on external evaluation it appeared almost normal (Fig. 5). The recognition of this anatomic feature sheds insight into false negative TTE findings when the coronary arteries are evaluated in the routine parasternal short axis view [12]. Color Doppler flow mapping has rightfully been recognized as an important tool in coronary evaluation [6], but the parasternal short axis sweep must be extended superiorly into the ascending aorta to unmask the anomalous course.

Review of two patients at our institution that had normal initial echocardiograms and were subsequently diagnosed with left AAOCA demonstrated the anomalous angle in the parasternal long axis view [13]. When studied retrospectively, this was a finding present on AAOCA patients but not in controls. This TTE sweep is an important view to evaluate the left coronary because it is routinely done at most centers during evaluation of the right ventricular outflow tract, and provides an easy screening tool for AAOCA when special care is taken to identify the proximal left main coronary artery. Furthermore, in our experience, this screening can also be used to identify the anomalous right coronary artery.

Although the diagnosis of AAOCA is commonly incidental and its management remains controversial [10], when symptoms are present they tend to be exertional in

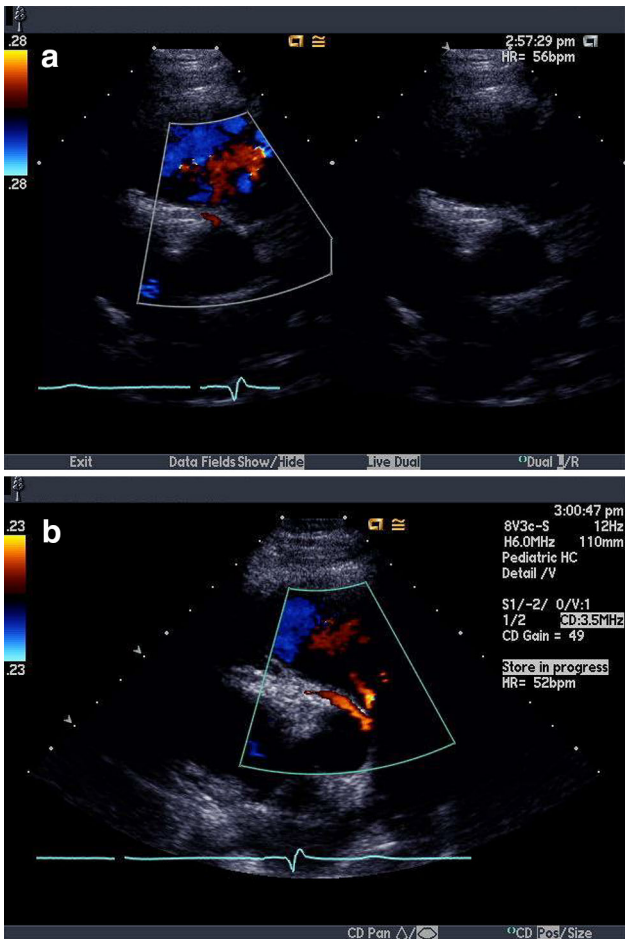

Fig. 4 False negative in a different patient with anomalous right coronary artery when imaged in the **a** standard parasternal short axis view; the anomalous coronary is visualized more superiorly (**b**)

nature [2]. With an increased effort toward cost-effective cardiac screening in young athletes, it is imperative to provide a better assessment of coronary arteries by TTE as ECG and exercise tests are often unremarkable in this subgroup [4], and no further imaging tests are pursued if the coronaries are deemed normal by TTE. Based on our results, we speculate that the addition of the proposed screening techniques the specificity of even targeted TTE's for young athletes can be significantly improved.

While a significant proportion of our patients (9 of 14) with a diagnosis of AAOCA did not undergo confirmatory tests due to young age and/or lack of exertional symptoms, this management strategy is reflective of current nation-wide practice. A recent survey [3] of practitioners around the country revealed that 92 % relied on echocardiography for the initial diagnosis, with only approximately 25 % obtaining confirmatory imaging (CT, MRI, or cardiac

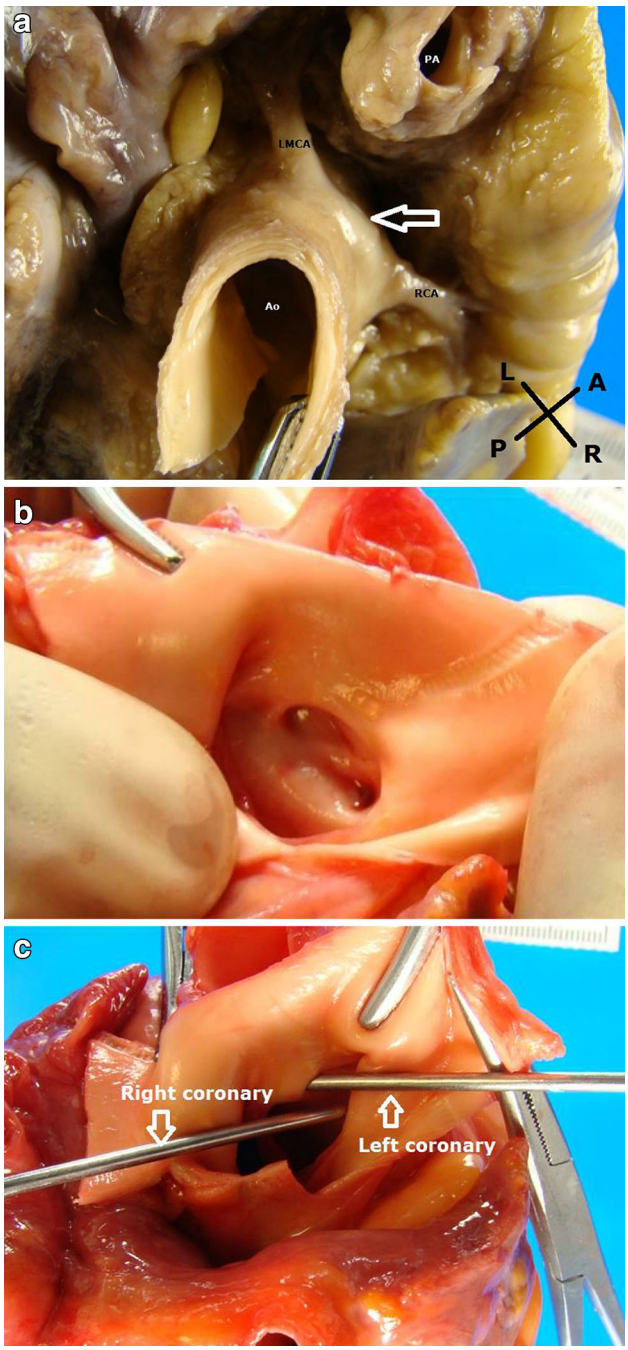

Fig. 5 **a** External appearance of anomalous left coronary from the right sinus. The *arrow* denotes the anomalous proximal course which is subtle on external inspection, with an almost normal appearance to the 'exit' site from the ascending aorta (*Ao* aorta, *LMCA* left main coronary artery, *PA* pulmonary artery, *RCA* right coronary artery). **b** Internal inspection of the aorta reveals both coronaries arise from the same (right) sinus. **c** Probe placement demonstrates that the anomalous proximal left coronary artery has a more superior take-off and proximal course compared to the right coronary (Photo credit for Dr Sarah Johnson-Welch)

catheterization). In addition, approximately 20 % do not refer for surgery if the patient is asymptomatic and have no evidence of ischemia. Of those that refer asymptomatic patients for surgical repair, there is a wide range of age threshold.

The main limitations of our study are inherent to its retrospective nature; due to the vast number of studies, we were unable to review every negative TTE study retrospectively to evaluate if all coronary views were performed or if acoustic windows were adequate for diagnosis. Therefore, the false negatives (from inadequate visualization of coronaries) in our population are unknown.

The study included all patients with TTEs performed in our laboratory as our 'population'. An evaluation limited to young adults undergoing TTE for workup of chest pain or syncope might have yielded a higher incidence, but was not technically feasible for a retrospective chart review. In addition, we cannot gauge the effect of increased awareness and interest in AAOCA in the increase in their diagnosis.

In conclusion, our data show that the addition of the screening views adopted recently in our institution can significantly increase the sensitivity of TTE in diagnosing AAOCA in the asymptomatic general population. We propose that these views be incorporated into the standard TTE evaluation of coronary arteries, specifically in the young athlete, thereby increasing the sensitivity of even limited TTE for sports clearance in this population.

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