

AATS consensus statement helps manage treatment of coronary anomalies

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A diagram of the 'unroofing' technique, which is one of the recommended surgical interventions discussed in the guidelines. Credit: The Journal of Thoracic and Cardiovascular *Surgery*

The issues surrounding congenital coronary anomalies and their effect on sudden death are complex. Researchers are still trying to fully



understand anomalous aortic origin of a coronary artery (AAOCA) and its relationship to adverse health outcomes in humans, especially children. Using the most up-to-date literature, as well as the input of leading experts in the field, the American Association for Thoracic Surgery (AATS) has released practical guidelines for the identification and treatment of AAOCA, including an overview of the latest data surrounding population-based risk.

AAOCA is a heart abnormality that can increase an individual's risk for sudden cardiac death (SCD) due to ischemia and arrhythmias. These risks increase with vigorous exercise, particularly in a competitive environment in which participants are regularly driven to the upper limits of physical endurance. The consequences are perhaps most devastating in youth sports. Many children with these anomalies present no symptoms and sometimes the only indication of the defect is when sudden death occurs. According to data, as many as 600,000 children and young adults could have AAOCA, putting them at real risk for sudden cardiac death during or shortly after intense exercise.

AATS assembled a multidisciplinary group of experts, including congenital cardiac surgeons and adult and pediatric cardiologists, to formulate evidence-based guidelines. "The recommendations are based on expert consensus opinion as well as on the best available evidence," explained lead author James S. Tweddell, MD, Executive Co-Director of the Heart Institute and Professor of Surgery, Cincinnati Children's Hospital Medical Center, Cincinnati, OH. "Despite important knowledge gaps, we feel it is important and timely to review the available evidence and present expert opinion based on best practices."

The consensus statement begins by offering important background information on the prevalence of AAOCA among young people and the risk for SCD. Investigators point out that the risk from SCD in <u>young</u> <u>people</u> who do not participate in competitive sports is incredibly low;



however, among those with AAOCA, the cumulative risk of death over a 20-year period participating in sports is 6.3% for AAOCA originating from the left side (AAOCLA) and 0.2% for AAOCA originating on the right (AAOCRA).

Investigators also address various diagnostic methods. While some AAOCAs are caught during testing because a child is symptomatic, two reports analyzed by the group indicate that 50% of SCD due to AAOCA are first events without prior symptoms. The guidelines suggest that the best method of initial screening for AAOCA is a thorough transthoracic echocardiogram with Doppler color flow mapping. The experts recommend this method based on wide availability, cost-effectiveness, ease of performance, and absence of radiation exposure. If the echocardiogram is inconclusive or the anatomy cannot accurately be defined, coronary computed tomographic (CT) angiography or cardiac magnetic resonance imaging is recommended to get a better picture of the structure of the heart.

In terms of treatment, the guidelines discuss surgical interventions, as well as non-surgical medical management including exercise restrictions and declining to participate in competitive sports. "For children with AAOCA, the decision to participate in competitive or high intensity recreational sports should include a thorough family discussion and counseling regarding the risks and benefits of observation versus surgical management of this lesion," said Dr. Tweddell. "In addition, we recommend that automated external defibrillators (AEDs) should be available and accessible for sporting practices and competition."

With so many lives, especially young lives, at stake, it is important for practitioners to have easy access to the latest information surrounding AAOCA. "Although the risk for any single affected individual is small, the loss of an otherwise healthy person is particularly devastating," concluded Dr. Tweddell. "Surgical and interventional therapies have



been developed that appear to be protective but these therapies carry risks. The challenge is identifying those individuals at such risk that there is net benefit to the therapy. While we acknowledge that there is still much to be learned with this entity, the guidelines put forth in this document are based on best practice and knowledge regarding the risk of SCD that we have to date."

More information: Julie A. Brothers et al, Expert consensus guideline: Anomalous aortic origin of a coronary artery, *The Journal of Thoracic and Cardiovascular Surgery* (2017). DOI: 10.1016/j.jtcvs.2016.06.066

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