

MYOCARDIAL ISCHEMIA IN CHILDREN WITH ANOMALOUS AORTIC ORIGIN OF A CORONARY ARTERY WITH INTRASEPTAL COURSE

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Background: Intraseptal anomalous aortic origin of a coronary artery (AAOCA) is considered a benign condition. However, there have been case reports of patients with myocardial ischemia, arrhythmia, and sudden cardiac death. The purpose of this study was to determine the clinical presentation, myocardial perfusion upon provocative stress testing, and management of children with AAOCA with an intraseptal course in a prospective cohort.

Materials/Methods: Patients with AAOCA and intraseptal course were prospectively enrolled from 12/2012-05/2019, evaluated, and managed following a standardized algorithm. Myocardial perfusion was assessed using stress imaging. Fractional flow reserve (FFR) was performed in patients with myocardial hypoperfusion on non-invasive testing. Exercise restriction, beta-blockers, and surgical intervention were discussed with the families.

Results: Eighteen patients (female 6, 33.3%), who presented with no symptoms (10, 55.6%), non-exertional (4, 22.2%) and exertional symptoms (4, 22.2%), were enrolled at a median age of 12.4 years (0.3-15.9). Perfusion imaging was performed in 14/18 (77.8%) and was abnormal in 7/14 (50%); FFR was positive in 5/8 (62.5%). All 4 patients with exertional symptoms and 3/10 (30%) with no or non-exertional symptoms had myocardial hypoperfusion. Coronary artery bypass grafting was performed in a 4-year-old patient; beta-blocker and exercise restriction were recommended in 4 patients not suitable for surgery. One patient had non-exertional chest pain and 17 were symptom-free at median follow-up of 2.5 years (0.2-7.1).

Conclusions: Up to 50% of patients with intraseptal AAOCA had inducible myocardial hypoperfusion during non-invasive provocative testing. Long-term follow-up is necessary to understand the natural history of this rare anomaly.