

MYOCARDIAL ISCHEMIA IN ANOMALOUS AORTIC ORIGIN OF A RIGHT CORONARY ARTERY (AAORCA): MEDIUM-TERM FOLLOW-UP IN A LARGE PROSPECTIVE COHORT

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Background: AAORCA may lead to myocardial ischemia (MI) and sudden death in the young. Asymptomatic patients are deemed low-risk if normal exercise stress test (EST). We aimed to determine the occurrence of MI in AAORCA following a standardized approach in a prospective cohort.

Materials/Methods: All patients with AAORCA from 12/2012 to 10/2019 were enrolled. CTA and EST (>8 years) and stress perfusion imaging (sPI) (>10 years) were performed, and both EST and sPI in younger patients if concern for MI.

Results: A total of 222 patients (male 135, 61%) with AAORCA were enrolled, median age of 11.6 [IQR 6.2-14.7] years, follow up 1.8 [IQR 0.7-4.2] years. At presentation, 169 patients (76%) had no exertional symptoms (chest pain/syncope). EST was performed in 113/169 (67%), only one positive (<1%). sPI was performed in 98 (58%) with 10 (10%) positive for abnormal perfusion. Of those with negative EST, 9 (10%) had positive sPI. In symptomatic patients (53, 24%), EST was performed in 49 (94%), only one positive (2%). sPI was performed in 42 (79%) and 7 (17%) were positive. Of those with negative EST, 7 (18%) had positive sPI. Comparing patients with positive and negative sPI, no difference in intramural length was seen (5.5 ± 2.3 vs 6 ± 2.3 mm, $p=0.4$).

Conclusions: Asymptomatic patients with AAORCA may present with inducible MI on sPI. EST is a poor predictor of MI, regardless of symptoms. Intramural length of the RCA does not appear to correlate with MI in this cohort. At follow up, all patients are alive and most exercising with no restriction. Long-term follow-up is necessary to determine the natural history of those with and without MI on sPI.