

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

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## BACKGROUND

- Anomalous aortic origin of a coronary artery (AAOCA) with intraseptal course is rare and considered a benign condition.
- However, there have been case reports of myocardial ischemia, arrhythmia, and sudden cardiac death in this condition.

## HYPOTHESIS

- Myocardial ischemia is not uncommon in AAOCA with intraseptal course.
- We prospectively determined the clinical presentation, myocardial perfusion upon provocative stress testing, and proposed management in a prospective cohort of children with intraseptal AAOCA.

## METHODS

- Patients were prospectively evaluated and managed using a standardized algorithm from 12/2012-05/2019.
- Stress nuclear and/or magnetic resonance perfusion imaging in patients  $\geq 8.0$  years old and/or symptoms of ischemia.
- Diastolic fractional flow reserve during dobutamine (dFFR<sub>d</sub>) and/or instantaneous wave-free ratio (iFR) were performed in patients with perfusion deficits or high-risk features.
- The risks/benefits of exercise restriction, beta blockers, and surgery were discussed.

## Non-invasive and intracoronary hemodynamics assessment of the intraseptal coronary artery

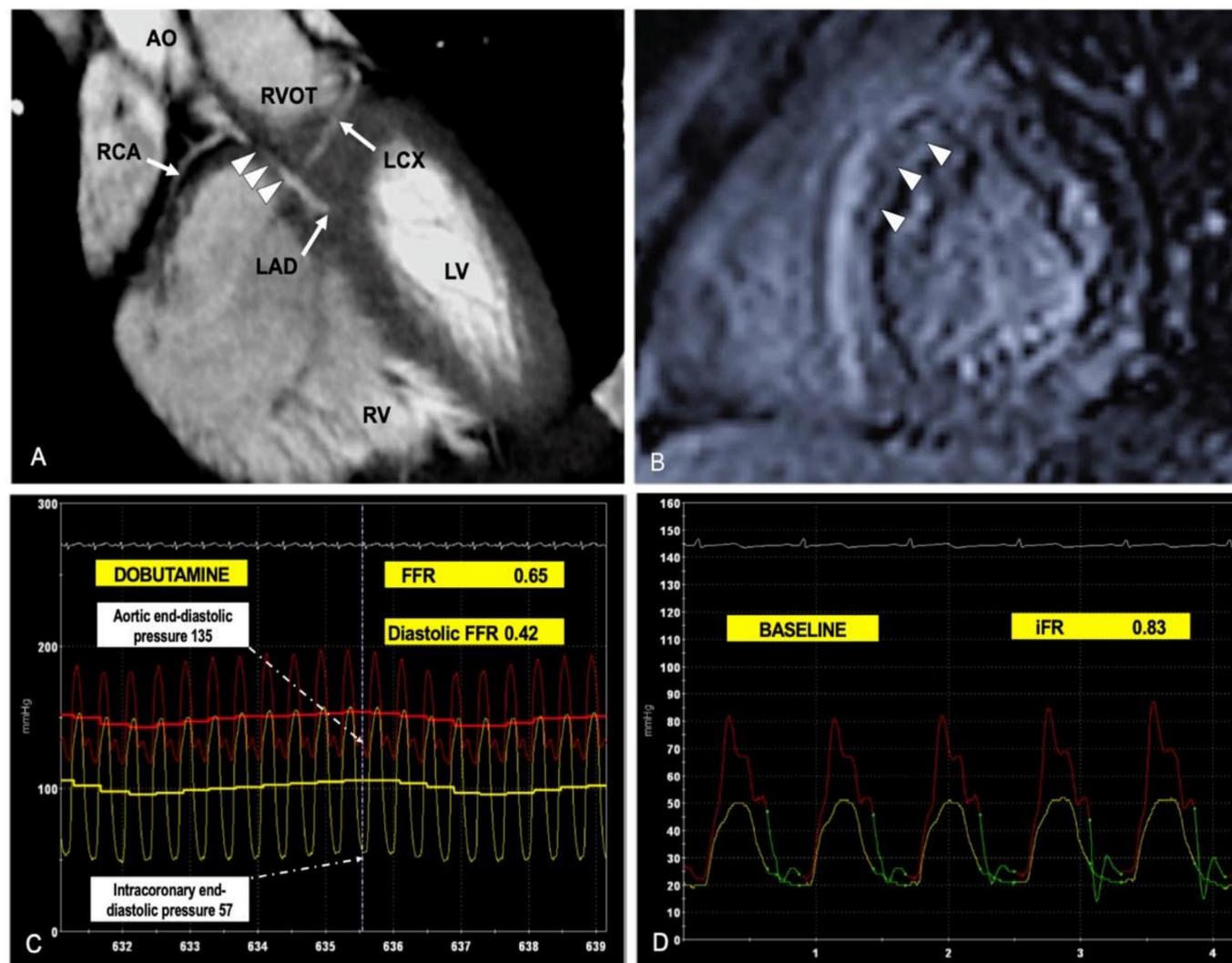


Figure 1. Non-invasive and invasive intracoronary hemodynamics assessment of the anomalous coronary artery. (A) CTA image demonstrating anomalous aortic origin of the LCA with an intraseptal course (arrow heads); (B) scMR showed perfusion defect in the anterior septum during peak dobutamine stress (arrowheads); FFR and diastolic FFR during dobutamine infusion (E), and tracings of iFR at baseline (F). LCA = left main coronary artery; AO = aorta; RVOT = right ventricular outflow tract; LV = left ventricle; RV = right ventricle; RCA = right coronary artery; LAD = left anterior descending coronary artery; LCX = left circumflex coronary artery; CTA = Computerized tomography angiography; FFR: Fractional flow reserve; iFR: Instantaneous wave-free ratio; scMR: Stress cardiac magnetic resonance imaging.

## RESULTS

- Prospective study (2012-2019)**
- 18 pts (6 female, 33.3%) with no symptoms (10, 55.6%), non-exertional (4, 22.2%), exertional symptoms (4, 22.2%)
  - Symptoms 8/18 (44%)
  - Stress perfusion imaging Inducible ischemia 7/14 (50%)
  - Diastolic FFR  $\leq 0.80$  5/8 (63%)
  - Perfusion imaging performed in 14/18 (77.8%)
  - All 4 pts with exertional symptoms and 3/10 (30%) with non-exertional symptoms had inducible hypoperfusion.
  - CABG performed in a 4-year-old pt;  $\beta$ -blocker & exercise restriction recommended in 4 pts deemed not suitable for surgery.
  - One patient had non-exertional chest pain and 17 were symptom-free at a 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 provocative non-invasive testing.
- Long-term follow-up is necessary to understand the natural history of this rare anomaly

