Coarctation of the aorta: Future risk despite adequate repair

Melinda Cory, MD[1], Thomas M. Zellers, MD, MSc[2], William Scott, MD, MS[2], Claudio Ramaciotti, MD[2] and V. Vivian Dimas, MD[2]

[1]Pediatrics, Children's Medical Center, Dallas, TX; [2] Pediatrics, Division of Cardiology, University of Texas Southwestern Medical Center, Dallas, TX

Background

- Patients with repaired coarctation of the aorta (CoA) have a shorter life expectancy than their peers
- Myocardial infarction and cerebrovascular accident are the two most common etiologies of late death
- It is thought that these patients have abnormalities of arterial endothelium, despite adequate repair, placing them at increased risk for a future cardiovascular incident
- Risk factors for cardiovascular incidents include ambulatory hypertension, abnormalities of arterial endothelial function, early atherosclerosis and exercise induced hypertension
- Carotid intima media thickness (cIMT) is a validated surrogate end point for atherosclerosis
- Flow mediated vasodilation (FMV) is a validated test to assess coronary artery endothelial dysfunction
- There is limited data for pediatric patients with repaired coarctation

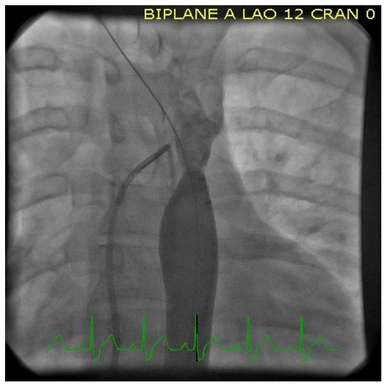

Figure 1: CoA pre-stent

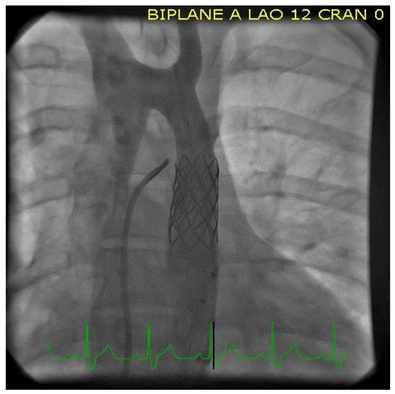

Figure 2: CoA post-stent

Objectives

- Study Objective: To evaluate patients with repaired CoA for evidence of arterial dysfunction
- Hypothesis: Patients with CoA would show endothelial dysfunction and abnormalities of carotid intima media thickness despite successful repair
- Additional known risk factors for cardiovascular events including ambulatory hypertension and exercise induced hypertension were also evaluated

Materials & Methods

- Patients > 5 yrs of age status post repair of CoA without associated significant congenital heart disease were included.
- Successful repair defined as residual gradient < 20 mmHg by echo or upper and lower extremity BP readings
- 24 patients were identified from August 1, 2011 until March 31, 2014, with successful enrollment of 10 pts.
  - One patient was later excluded due to previously unidentified associated congenital heart defects.
- Evaluation included:
  - Echocardiogram
  - cIMT measurement
  - FMV testing
  - Exercise stress testing (EST)
  - 24 hour ambulatory blood pressure monitoring (ABPM)
- Results were compared to normative data from pediatric patients if available, otherwise adult normative data was used to determine abnormal values
- Not all enrolled patients were able to complete all components of the evaluation and some had unsatisfactory data for interpretation

Results

- Patient characteristics
  - The average age was 15.5 ± 3.7 yrs
  - The average corrected echo gradient post-repair was 11.3 ± 8.5 mmHg
  - One pt whose echo gradient exceeded 20 mmHg underwent catheterization where the peak to peak gradient was 3 mmHg
  - 5 patients (55%) had recurrent coarctation which was repaired
  - 4 patients (45%) had associated bicuspid valve
  - On initial repair, 5 patients (55%) were corrected surgically while the remaining 4 (45%) had stent placement (figures 1 and 2)
  - All 4 patients with recurrent CoA had a stent placed at second correction
- EST completed by 7 patients
  - Three (43%) patients met criteria for exercise induced hypertension (peak systolic BP > 95th percentile)
  - One patient had systolic BP in the 90-95th percentile
- cIMT measurement completed by 6 patients
  - Four (67%) were abnormal (> 95th percentile)
- FMV completed by 8 patients (7 with interpretable data)
  - Two (28%) were abnormal.
- ABPM completed by 8 patients (3 adequate for interpretation)
  - All 3 patients met criteria for ambulatory hypertension
  - Two of the 3 patients met criteria for severe ambulatory hypertension.
  - All 3 patients with abnormal ABPM also had abnormal exercise stress tests.
  - Two of the 3 patients also completed cIMT measurements and both were abnormal.
- There were no significant associations among CoA type, or repair type with abnormal EST or cIMT.
- Results summary in Table 1

Table 1: Patient Characteristics and Summary of Results

Patient	Age (yrs)	Echo Gradient (mmHg)	cIMT	FMV	EST	ABPM
001	23.14	5	NC	A	A	A
002	14.44	13	A	N	A	A
003	13.95	30	NC	ID	A	NC
004	17.00	12.5	N	NC	N	ID
005	13.59	14	A	N	N	ID
006	10.11	4	NC	NC	NC	NC
008	18.59	11	A	N	N	NC
009	15.49	12	N	A	N	NC
010	13.65	0	A	N	A	A

A (Abnormal), N (Normal), NC (Not Completed), ID (Insufficient Data)

Summary

- CIMT is a validated predictor of future cardiovascular events in adults and was abnormal in 67% of patients who underwent cIMT measurement.
- All patients who were able to complete ABPM had abnormal findings
- Prospective enrollment remains a challenge in clinical research studies

Conclusions

Patients with CoA demonstrate persistent arterial abnormalities despite adequate repair. This is consistent with our hypothesis that these patients have abnormalities in their arterial endothelium which will place them at increased risk for future cardiac events at an early age. Further study is warranted in a larger population to better define this risk profile and reduce late morbidity and mortality.