Experience in the management of abnormal origin of coronary artery from the opposite sinus at childhood age

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The objective of this study was to assess the clinical presentation, management and outcomes of children diagnosed with abnormal course of coronary artery from the opposite aortic sinus (ACAAO).

Material and methods: All patients with ACAAO and less than 18y of age at diagnosis were included in this single-center retrospective study. Demographics, clinical presentation and symptoms, echocardiographic data treatment and outcomes were assessed.

Results: From 1993 to 2016, 34 patients aged 10.5±5.8y, had ACAAO (23 males=67.6%): 19 right coronary artery (RCA) from left sinus (56%), 9 left coronary artery (LCA) from right sinus (26%) and 6 others (single coronary artery and circumflex from right coronary artery). An Inter-arto-pulmonary course was present in 79.4% and intramural segment in 67.6%. Mean age at diagnosis was 8y in females and 11.7y in males (p=0.08) and did not differ with anatomy. Three patients had cardiac arrest (9%), 7 chest pain at exercise (21%), 8 syncope (23%), 3 dyspnea (9%), 2 acute pulmonary edema (6%), 2 arrhythmias (6%) and 9 had no symptom (27%). All patients with severe symptoms (i.e. 59% of the cases) had an intramural segment, 57% of RCA and 89% of LCA had severe symptoms. Time from onset of symptoms to diagnosis was 1.9±2.6y (median 9months), not different with anatomy and shorter in females (0.9y) than males (2.5y, p= 0.27). Surgical reimplantation was performed in 20 cases (59%): 63% of RCA and 89% of LCA from the opposite sinus, none of the single coronaries and abnormal circumflex. Surgery was more frequent in case of intramural segment (80% versus 28%, p = 0.0027) and severe symptoms (86% versus 27% in mild or no symptoms, p= 0.01). Age at surgery was 12.9±4.5y (median 13.6y, min 5.3y). Time from diagnosis to surgery was 190±470 days (median 50days) : shorter in LCA (50days) than RCA (253days,p= 0.44) and in severe symptoms (67 versus 723days, p = 0.02). Follow-up was 4.4±3.7y. No death occurred.

Conclusion: ACAAO in children can impair vital prognosis, even in RCA from opposite sinus. Intramural trajet is associated with worse outcome. Surgical option depends on clinical status and anatomy, and has been uneventful.