

TITLE: Echocardiographic diagnosis and treatment of anomalous left coronary artery from the pulmonary artery; a 25 year experience

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INTRODUCTION: Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare form of congenital heart disease resulting in post-natal myocardial hypoperfusion and systolic dysfunction. Echocardiography has now replaced cardiac catheterization as the preferred method for diagnosis. The purpose of this study is to review a multi-center experience using echocardiographic markers as the primary diagnostic modality for ALCAPA, and then describe the short and long term surgical outcomes.

METHODS: Charts and echocardiograms from all patients undergoing ALCAPA repair from 1990-2015 at the Children's Hospital of Wisconsin and the Children's Hospital and Medical Center Omaha were identified and reviewed. Patients with additional intracardiac or structural coronary artery abnormalities were excluded.

RESULTS: A total of 36 patients (64% female) were identified. The median age at diagnosis was 9mo (1.3 – 219mo). Most, 67% of patients presented at <24 mo. of age. The majority, 55%, presented prior to 12mo of age. 90% of infants presented with congestive heart failure symptoms while older patients were more likely to be found incidentally or present with a murmur (63%).

During the study period, 70% of patients were diagnosed by echocardiography alone. No patient required cardiac cath in the final 10 yrs. of the experience. Coronary arteries origins were easily visualized in only 52% of diagnostic studies. However, indicators of ALCAPA were present and predicted the diagnosis in all cases. Specific markers included flow reversal within the left coronary artery (30/33; 90%), abnormal Doppler flow signals within the pulmonary artery (27/34; 79%), intramyocardial flow signals indicative of collateral coronary vessels (28/33; 85%), and right coronary artery dilation ≥ 2.0 mm (25/31; 81%). More generic markers included mitral valve insufficiency (26/35; 74%), left ventricular systolic dysfunction ($EF \leq 45\%$) (23/35; 66%) and left ventricular endocardial fibroelastosis (EFE) (20/35; 57%). 76% (n=25) of patients had ≥ 5 indicators present at the time of diagnosis. Infants were much more likely to present with left ventricular systolic dysfunction (90% vs. 37%, $p=0.0056$) and mitral insufficiency (85% vs. 62%, $p=0.38$) as compared to older children. Older children were more likely to present with collaterals (100% vs 75%, $p=0.038$) as compared to infants.

All patients underwent surgical repair within 30d of diagnosis. 80% (n=29) had direct coronary artery re-implantation and 20% (n=7) underwent the Takeuchi procedure (last in 1998). Only one patient required immediate post-operative ECMO support. There were no early (<30d) deaths or reinterventions. The median length of post-operative stay was 8 days (3-36 days). The median follow up was 10.47 yrs. (range 0.1 to 21 years). There was 1 late death and 1 patient was lost to follow up. At last follow up, 88% had normal systolic function ($EF > 50\%$), 36% had \leq mild mitral insufficiency, and only one patient (2.7%) had moderate mitral insufficiency. 11% (n=4) had residual coronary artery issues, 2 patients with stenosis at the site of repair (both having undergone with Takeuchi procedure), one with distal left main coronary artery stenosis and one with coronary artery dilation. By the last follow-up, 17% (n=6) of patients had required reintervention. 2 patients required coronary artery revisions, 2 patients required mitral valve revisions, 1 required implantable defibrillator placement, and 1 required pulmonary artery stenosis repair.

CONCLUSION: ALCAPA is a rare, but potentially fatal form of congenital heart disease that often requires an accurate and timely diagnosis. The majority of patients will present in infancy, but >30% will present in later childhood or beyond. Infants are more likely to present with overt heart failure with diminished LVEF and MR while older patients are often found serendipitously with echocardiographic evidence of collateralization. Echocardiography is the preferred method of diagnosis and can predict ALCAPA even if the CA origins are not well seen. In the modern era surgical outcomes are excellent and most patients will recover LV function and have improvement in MR. 20% of patients will require reintervention, with residual CA or MV issues being the most common indication.