Anomalous Left Coronary Artery from Pulmonary Artery (ALPACA) - Is It Really so Rare?

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Background: Abnormal left coronary artery from pulmonary artery (ALCAPA) is described in the literature as a rare congenital cardiovascular defect that occurs in approximately 1/300000 live births. The mortality of untreated ALPACA has been estimated to range from 35 % -85% in the first year of life. Our data and experience shows a much higher incidence of this entity.
Methods: Hospital data bases were retrospectively searched for cases of ALPACA diagnosed at our center over the past 10 years.
Results: During a period of 10 years, 2001-2011, we diagnosed and treated 9 patients with ALPACA. Seven patients came to medical attention because of respiratory distress and asthma like symptoms that developed during the first month of life. Two patients were referred for cardiac evaluation because of failure to thrive and systolic murmur. Age at clinical presentation ranged from 1-7 months. Diagnosis was made at 2-48 months, average 15 months. The main initial diagnostic tool was echocardiography and diagnosis was confirmed by cardiac catheterization. During the last period of study, CT and MRI were used in order to confirm the diagnosis. All 9 patients had successful surgical repair by re implantation of the left coronary artery to the aorta.
Conclusion: Relying on the published incidence and based on the annual birth rate in our hospital we should have diagnosed and treated only one tenth of the actual diagnosed patients over a period of 10 years. Our rate is tenfold higher than expected. We speculate that ALCAPA is either more frequent in certain geographic areas or simply under diagnosed worldwide. A high index of suspicion and new diagnostic modalities like MRI and CT angiography may show that the current reported incidence is actually too low.