Successful Repair of Anomalous Single Coronary Artery from the Pulmonary Artery with an Intramural Right Coronary Artery

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Purpose: Anomalous single coronary artery from the pulmonary artery (ASCAPA) is a rare congenital anomaly with less than 30 cases described in the literature. Six of 10 (60%) patients are reported to have survived operative intervention. We present our experience with diagnosing and successfully managing ASCAPA in a 6-week-old, 3.7-kg female.

Methods: Echocardiography demonstrated an intramural right coronary artery arising from a single coronary artery with possible origin from the pulmonary artery. Cardiac catheterization confirmed a single coronary artery arising from the right pulmonary artery. At the time of surgical correction, a large ostial button was excised from the right and main pulmonary arteries. Mobilization of the intramural right coronary artery was accomplished by excising the aortic wall en block and performing an aortic valvuloplasty. Deep hypothermic circulatory arrest was required for direct re-implantation of the single coronary artery on the transverse aortic arch. The aorta and pulmonary artery were reconstructed with decellularized bovine pericardial patch and autologous pericardium.

Results: The patient was supported with extracorporeal membrane oxygenation for 4 days postoperatively. The patient's chest was closed on the seventh postoperative day. Postoperative computed tomographic angiogram showed a widely patent coronary artery.

Conclusion: ASCAPA is a rare anomaly which may require creative positioning of the coronary button for a successful outcome.