PERSISTENT ALCAPA PHYSIOLOGY AFTER ALCAPA REPAIR

Background: Left ventricular (LV) perfusion in anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) relies on right coronary artery (RCA) collaterals prior to repair. The Takeuchi repair for ALCAPA entails the creation of an aortopulmonary baffle (APB) which incorporates the left coronary. We present an adult case of Takeuchi repair failure.

Case: 44 year old male with ALCAPA s/p Takeuchi repair at age 19 presented with dyspnea. Cardiac imaging showed a massively dilated LA (272ml), severe MR and several jet flows from the APB into the pulmonary artery (PA) with normal LV function. Cardiac cath revealed the LV is primarily perfused by RCA collaterals (Fig IB) and the proximal APB is partially obstructed with poor antegrade flow (Fig 1A) as it feeds the APB-PA fistula. Elevated filling pressures and pulmonary artery pressure (PAP) were also confirmed by cath.

Decision-making: The APB failure has reinstated the LV dependent RCA collateral circulation and added a new left to right shunt (APB-PA), resulting in elevated PAP. The ischemic watershed area between the right and left coronary circulation has led to chronic MR and a massive LA. Surgical repair included APB-PA patch repair with removal of the proximal baffle fibrosis, mitral valve repair and LA plication. Repeat cath showed APB anterograde flow (Fig 1C), no shunting and improved PAP.

Conclusion: Complications of the Takeuchi repair require a multidisciplinary approach recognizing all aspects of the disease for a successful outcome.