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Resolution of Pulmonary Arterial Hypertension after Successful Repair of a Large Aortopulmonary Window (APW) in an Adult Patient

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Introduction: Aortopulmonary window (APW) , if unrepaired early in life, leads to fixed pulmonary hypertension and Eisenmenger's Syndrome. We report the case of a 23 year old man with a large type I APW who was judged to remarkably still have reversible pulmonary hypertension, which indeed resolved after successful surgical repair.

Methods: Auscultation of a cardiac murmur in an asymptomatic 23 year old 70 kg army recruit led to the diagnosis of a large, 4 cm (by ECHO), aorto-pulmonary window. Cardiac catheterization at another institution revealed pulmonary artery (PA) pressure at systemic level, pulmonary to systemic flow ratio (Qp/Qs) greater than 4:1, and pulmonary vascular resistance (PVR) approximately 1/10 of systemic. Oxygen challenge resulted in further increase of Qp/Qs and PVR decreased. Lung biopsy via left thoracotomy showed no evidence of irreversible pulmonary hypertension (PAH), and the patient was referred to our center for further management. Surgical repair was performed through a median sternotomy, cardiopulmonary bypass with aortic and bicaval cannulation, and cardioplegic arrest. The aorta and pulmonary artery were separated at the level of the window, and the resulting defects in the great vessels were closed with a Dacron Hemashield patch to the aortic side, and a separate autologous pericardial patch to the pulmonic side.

Results: Immediately postoperatively, under anesthesia, PAP pressure was reduced to approximately one quarter of systemic. The postoperative period was uneventful, and the patient was discharged in 7 days on prophylactic sildenafil therapy, at which time estimated PA pressure was half systemic. At follow-up one year later, the patient remains clinically well, at NYHA class I, and with echocardiographically estimated PA pressure remaining at the same level.

Conclusion: Although previously unrepaired APW in adults is typically inoperable due to the establishment, early in life, of irreversible pulmonary arterial vascular disease and hypertension, nonetheless, if careful evaluation demonstrates reversibility of PA hypertension, surgical repair should be offered, as it can achieve restoration of normal anatomy, abolishment of shunting, and resolution of pulmonary hypertension. Long term close follow-up is recommended to monitor PA pressure, in the hope of eventual complete discontinuation of sildenafil therapy.