SUCCESSFUL PREGNANCY FOLLOWING MID LUNG EXCLUSION IN A CYANOTIC FONTAN PATIENT

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Category: Case Vignettes

Background: Many causes for cyanosis exist in Fontan palliated single ventricle patients (FpSVp). Diffuse extensive pulmonary AV malformations (AVMs) may make cyanosis resolution difficult. We present a patient with diffuse right lung AVMs (O2 sat 78%), poor exercise tolerance, and history of miscarriage, who successfully completed pregnancy following tarns-catheter partial right lung exclusion.

Case: A 26 yo FpSVp (D-Transposition of the great arteries, large ventricular septal defect, pulmonary stenosis and small left ventricle) was repaired abroad at 13 years of age. Initial interventions: BT shunt as a neonate; bidirectional Glenn at 2.5 years. Interval catheterizations for cyanosis included closure of Fontan fenestration, coiling of AVMs. She was left with substantial cyanosis related to extensive right lung AV malformations (AVMs). Patient moved to USA at 24 years and had exercise intolerance, good SV function, SaO2 in high 70% range despite hemoglobin normalization to the degree of cyanosis. Cardiac MRI: extensive bronchial and mediastinal collaterals directed to right lung. Patient had a strong desire to have a biological child, understanding all the associated risks to herself and the offspring.

Results: Cardiac catheterization: CI = normal, Fontan = 15 mmHg, LAP = 8 mmHg. Extensive diffuse AVMs in right middle lobe (RML). A large vascular plug placement in RML pulmonary artery excluded feeding AVMs. Additional small AVMs and small venous collaterals were also embolized. Patient's saturations improved to ~95%. At ~ 6-month follow-up, patient's SaO2 remained at 93-95% and exercise tolerance (peak VO2) had doubled. At 2 months follow-up patient continued to experience clinical improvement in exercise tolerance and her saturations remained elevated at 93% range. Subsequently she underwent a a successful pregnancy and delivery of a beautiful baby girl at 35 weeks gestation.

Conclusion: Diffuse AVMs may involve major lung segments of a lung with resultant cyanosis in FpSVp. Selective lung segment exclusion via embolization of the feeding branch of pulmonary artery may help improve clinical cyanosis. Tailored management approach may help with successful pregnancy outcomes in some complex CHD, cyanotic patients.

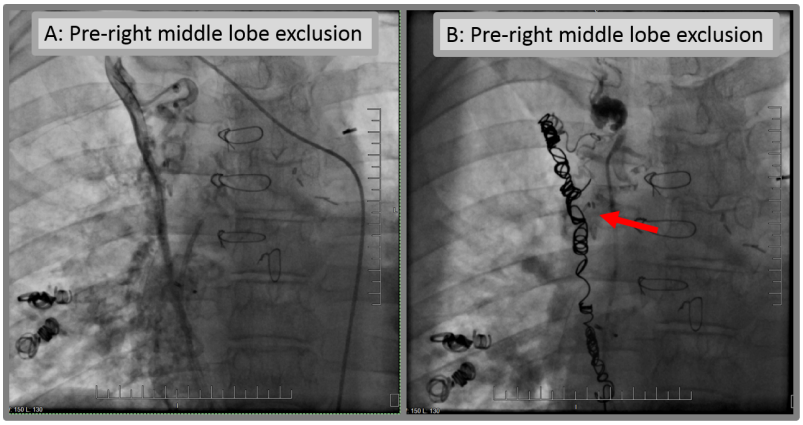

Image A: demonstrates pre coiling right middle lobe (RML) extensive AV malformations
Image B: image post coiling of collaterals and RML exclusion with vascular plug (red arrow)