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Complex Clinical Cases

TETRALOGY OF FALLOT WITH PULMONARY ATRESIA AND COMPLEX ANATOMY OF THE PULMONARY VASCULATURE - A NATURAL SURVIVOR

Poster Contributions
Hall B4-5
Saturday, April 6, 2024, 1:45 p.m.-2:30 p.m.

Session Title: 1280. Complex Clinical Cases: MD/PhD 05
Abstract Category: MD/PhD: Pediatric and Congenital Heart Disease
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Background: Tetralogy of Fallot (ToF) is the most common cyanotic congenital heart disease with broad spectrum of presentation, ranging from pink Fallot to pulmonary valve atresia (PVA). In this extreme anatomy, pulmonary blood flow depends entirely on bypass circulation.

Case: 22year-old man, diagnosed with PVA, pulmonary artery (PA) trunk agenesis and hypoplasia of the PA branches, with only left Blalock-Taussig shunt. Angiotomography showed aneurysmal dilation of the left PA confirmed by catheterization. Pulmonary pressure of 54/36 (45) mmHg and aorta 79/43 (53) mmHg. The patient was allegedly oligosymptomatic, but he had important cyanosis and during hospitalization he would assume squatting position frequently, classic in cyanotic ToF.

Decision-making: No possibility for unifocalization surgery of PA or cavopulmonary connection. Surgery or endovascular treatment of the aneurysm had high risk of obstructing important collateral vessels. It was opted to start sildenafil and cardiac rehabilitation. He progressed with clinical improvement, and ergoespirometry observed mild decrease in VO2 peak (23.7 to 20,11 ml.kg^{-1}.min^{-1}), with decrease of VE/VCO_2 slope (52.8 to 38.5), slight increase of METs (8.5 to 9.0), and saturation drop from 84% (at rest) to 71%.

Conclusion: Severe PA hypoplasia is a dramatic anomaly with few treatment resources to offer. The perception of symptoms is often influenced by the patient's adaptation to their clinical condition existing since birth, as in the case reported.