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Hugely Dilated Coronary Arteries in Congenitally Corrected Transposition of Great Vessels with Restricted Pulmonary Flow with Dextrocardia in an Adult: Case Report

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Abstract

The number of adults surviving with complex congenital heart diseases has increased dramatically over the past few years because of significant advances in diagnosis and medical and surgical care. Dilatation of the extramural coronary artery in cyanotic congenital heart disease is found in response to vascular remodeling induced by endothelial vasodilator substances like nitric oxide and vascular endothelial growth factor. Here, we describe a case of a 20 years young man who was diagnosed with congenitally corrected transposition of great vessels and underwent a successful bidirectional Glenn procedure. Pre-operative workup revealed hugely dilated, non-aneurysmal coronary arteries which were confirmed intra-operatively.

Keywords: Coronary dilatation; Extramural coronary artery; Dextrocardia

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Introduction

Patients with Cyanotic Congenital Heart Disease (CCHD) bear a complex multisystem disorder with its distinctive pathophysiology, requiring tertiary care. Recent advancements in medical and surgical techniques have now enabled the survival of these patients well into adulthood [1,2]. Bjork described ectasia of coronary arteries in a cyanotic patient in 1966 and after two years, his observation was confirmed by angiography and at necropsy [3]. At high altitude and in cyanotic patients, hypoxia-induced viscous erythrocytotic perfusate with its increased shear stress initiates coronary artery dilatation, but this dilatation subsequently reaches beyond expectations, as predicted on vasodilator therapy alone [4,5]. Due to increased levels of hypoxia-induced vascular endothelium growth factor, there is pronounced peripheral ramification of arterioles [6].

Case Summary

A 20 years of age young man presented with a history of shortness of breath on exertion and cyanosis since 5 years of age. On physical examination, he had severe cyanosis and grade IV clubbing with SPO2 of 62%. Cardiovascular examination revealed normal peripheral pulses and Grade 3/6 ejection systolic murmur in the pulmonary area. Chest X-ray showed dextrocardia with poor bronchovascular markings. 2D-echocardiography revealed dextrocardia, congenitally corrected Transposition of Great Arteries (ccTGA) with large inlet Ventricular Septal Defect (VSD) and severe pulmonary stenosis as shown in **Figure 1**.

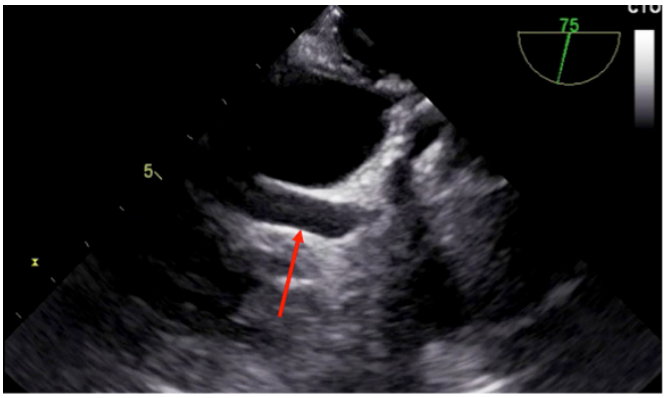

Figure 1: Echocardiography showing enlarged right coronary artery (red arrow).

Cardiac catheterization confirmed the diagnosis of dextrocardia with ccTGA, VSD with restricted pulmonary flow. Aortic root angiography showed dilated coronary arteries with left coronary from the right sinus and right coronary from left sinus in **Figure 2**.

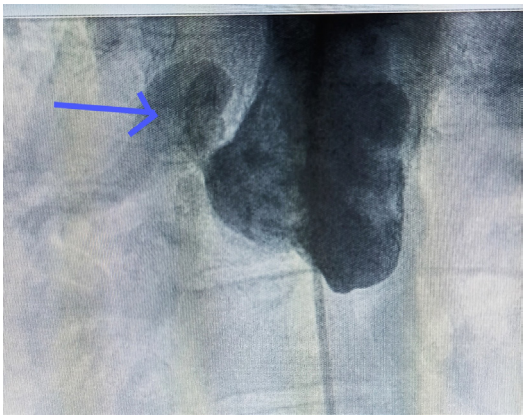

Figure 2: Angiography showing enlarged right coronary artery (blue arrow).

Contrast injection into descending thoracic aorta showed multiple collaterals supplying the lung. He underwent a successful bidirectional Glenn shunt under cardiopulmonary bypass. Severely dilated but non-aneurysmal coronary arteries were noted with a maximum diameter of 20 mm at the origin in **Figure 3**. Also, the branch coronaries were hypertrophied and tortuous. The postoperative course was uneventful with room air saturation of 84%. The patient is under close follow up and is planned for Fontan's completion surgery soon.

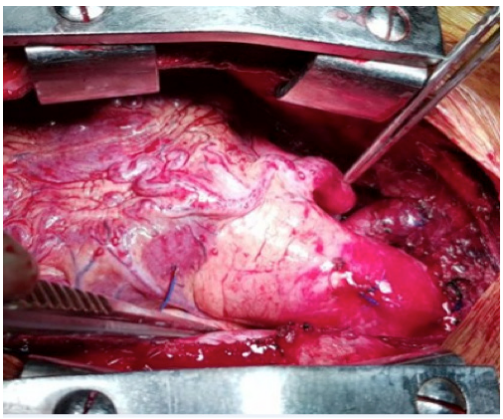

Figure 3: Intraoperative photograph showing large coronary arteries (forceps).

Discussion

Extramural coronaries are reported to be mildly dilated, moderately dilated or ectatic and tortuous in 83% (49/59) of the coronary arteriograms in cyanotic CHD [5]. In patients with CCHD, basal myocardial blood flow is considerably higher in both ventricular free walls and septum, potentially trespassing on flow reserve. As the extramural coronaries are already in a state of chronic maximal dilatation, there is little room for further dilatation. Aneurysmal dilatation of coronaries is a consequence of the associated abnormalities of media leading to mural attenuation. Hypoxia is considered to induce VEGF release from myocardial smooth muscle cells, with up-regulation of VEGF receptor-1 in myocardial endothelial cells. A recent study on CCHD patients revealed decreased bioavailability of nitric oxide and impaired response of acetylcholine on endothelium-dependent vasodilation. This impaired nitric oxide-mediated angiogenesis might lead to the limited growth of terminal arterioles in CCHD. The enhanced vasodilatory capacity of this resistance might be a contributing factor [6].

Aneurysm and ectasia of coronary arteries due to media attenuation had been described in various studies, in our case; these left and right coronaries were significantly dilated rather than aneurysmal, up to 20 mm in maximum diameter. While the abnormality exists in children and may become more pronounced with increasing age, a correct preoperative angiographic diagnosis of this rare anomaly is important. For clinical implications, it is important to recognize them perioperatively to avoid damage or compression at the time of surgery. During cardiopulmonary bypass, there are chances of easy distal run-off of cardioplegia with the risk of air-embolism and arrhythmia in the perioperative period.

It has been noticed that cyanotic patients have lower lipoprotein levels than the general population, which might be correlated with hypoxemia, secondary erythrocytosis with up-regulation of nitric oxide, hyperbilirubinemia and low platelet counts. Still, they have a higher ratio of LDL/HDL cholesterol with an increased risk of atherosclerosis [7]. As a growing population beyond adult age in this group, it is important to consider for atherosclerotic preventive measures during follow up.

Conclusion

Recent advancement in medical and surgical techniques has now enabled the survival of patients with complex cyanotic heart disease. Associated hypoxia leads to vascular remodeling of extramural coronary arteries by vascular endothelial growth factor (vasculogenesis) and nitric oxide (angiogenesis). Careful pre-operative angiographic diagnosis and proper intraoperative handling of these coronary arteries can avoid any unwanted complications during surgery.

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Conflicts of Interest

None.

Ethical Standards

The authors assert that all procedures contributing to this work comply with institute guides on the care.

Informed written consent was taken from the child's parents.

No prior abstract or poster presentation for this case.

The institutional ethics committee has approved this manuscript.

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