

What Can Cause Pulmonary Vascular Disease in Functionally Single Ventricle?

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Editorial

According to a classical concept, there are multiple pathophysiological factors that may cause pulmonary vascular disease in congenital heart defects [1]:

Increased blood flow and pressure in small pulmonary arteries (damages endothelium and starts cascade of biochemical reactions);

Increased pulmonary vein pressure (causes reflex spasm of pulmonary arteries);

High oxygen tension in pulmonary arteries (damages endothelium);

Systemic hypoxemia and acidosis as well as polycythemia with increased blood viscosity (causes micro embolism of pulmonary vessels).

Definition of “functionally single ventricle” includes a wide spectrum of anatomical variants such as double inlet left or right ventricle, mitral or tricuspid atresia, unbalanced atrioventricular canal, heterotaxy syndrome with one normally developed ventricle or other congenital heart defects with severe hypoplasia of one of the ventricles [2]. Anatomical and hemodynamic abnormalities in these congenital heart lesions are characterized by presence of a common ventricular chamber, where blood from the left and right atrium mixes and then enters the aorta and pulmonary artery. This can cause pulmonary hypervolemia and hypertension (in patients without pulmonary stenosis), difficulties with blood outflow (in patients with mitral atresia/stenosis combined with restrictive atrial septal defect), intake of highly oxygenated blood into the pulmonary artery (in patients with transposition of the great arteries) as well as systemic hypoxemia and polycythemia.

During the second half of the XX century, when the lung biopsy was believed to have a high diagnostic potential, multiple morphological studies were conducted to document the presence of pulmonary vascular disease in patients with functionally single ventricle [3-6]. However up to now, there are no fundamental works devoted to causes of pulmonary vessel morphological changes in this complex anomaly.

Pulmonary blood flow obstruction has a major impact on a single ventricular hemodynamics. In 1985, Juaneda and Haworth reported almost normal structure of pulmonary vessels in patients with single ventricle, pulmonary stenosis and decreased pulmonary blood flow [4]. However, patients with increased pulmonary blood flow but with “normal” pulmonary arterial pressure/resistance demonstrated hypertrophy of media in pulmonary arterioles [5]. Increased pulmonary arterial pressure almost always is associated with

hypertrophy of media as well as often with proliferation of intima [4,5]. Direct unrestricted blood flow from the single ventricle to pulmonary circulation in patients without pulmonary stenosis is associated with severe pulmonary hypervolemia, pulmonary hypertension and early morphological changes in pulmonary vessels. In 1976, Macartney et al. suggested that the genesis of pulmonary vascular disease is identical to a large ventricular septal defect [7]. However, Juaneda and Haworth demonstrated more rapid progression of morphological changes in pulmonary vessels in patients with functionally single ventricle and increased pulmonary blood flow, compared to patients with the ventricular septal defect [4]. They showed that patients with double inlet left ventricle and stenosis of systemic atrioventricular valve have more profound morphological changes of pulmonary vessels. Importantly, that the severity of media hypertrophy as well as intima fibrosis was the same in pulmonary arteries and pulmonary veins.

Full mixing of arterial and venous blood occurs in 16-36% of patients with functionally single ventricle [7,8]. In the rest of cases, blood flow is “selective”: in other words, blood mixing is incomplete. There might be a “favorable” hemodynamic variant, when pulmonary venous blood is directed predominantly to the aorta and blood from caval veins – to the pulmonary artery. In case of an “unfavorable” hemodynamic variant, arterial blood is directed predominantly to pulmonary circulation and venous blood – to systemic circulation. It is quite possible that an “unfavorable” type of intracardiac blood mixing in functionally single ventricle (similar to transposition of the great arteries) may serve as an additional cause for the development of pulmonary vascular disease. The course of pulmonary vascular disease can be worsened by micro embolisms to pulmonary vessels because of polycythemia and increased blood viscosity.

Thus, all aforementioned pathophysiological mechanisms can contribute to the development of pulmonary vascular disease in functionally single ventricle. Investigation of these mechanisms are challenging because of a big variety of anatomical and hemodynamic variants of single ventricle as well as because relatively small number of patients with this pathology.

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