EP14.03 Taussig-Bing Heart  
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Introduction

Double outlet right ventricle (DORV) is a type of ventriculoarterial connection in which both great vessels arise either entirely or predominantly from the right ventricle. From the wide spectrum of DORV variants, Taussig and Bing reported in 1949 a heart with the aorta transposed, arising entirely from the right ventricle and the pulmonary trunk, to the left, overriding a ventricular septum defect Fig. 1. Depending on the relationship between the pulmonary valve and the interventricular septum, the so called “Taussing-Bing heart” can be considered either a DORV or a transposition of great arteries (TGA) Fig. 2.

Case report

A 20 years old primigravida with a 24 weeks pregnancy presented for a suspicion of cardiac malformation. The ultrasound examination revealed in the “4 chamber view” a perimembranous ventricular septum defect (VSD), but otherwise normal cavities Fig. 3 and in the “3 vessel view” a malalignment and discrepancy in size of the aorta (3.6mm) compared to the pulmonary trunk (6.9mm) Fig. 4. Also, the great arteries had a parallel course with the aorta arising from the right ventricle, but the outflow tract of the left ventricle was not clearly visible Fig 5. The diagnosis was TGA. An amniocentesis was performed and revealed a normal karyotype.

Upon reexamination the enlarged pulmonary trunk seemed to override the VSD, arising predominantly from the right ventricle and parallel to the aorta Fig. 7,8, thus the diagnosis of DORV was taken into discussion. The discrepancy in size between the two great arteries was explained either by an obstruction at the level of the aortic arch or because the outflow of both ventricles was directed mainly into the pulmonary trunk. At 29 weeks, we obtained a clear image of the pulmonary trunk arising predominantly from the right ventricle alongside the aorta and the VSD extending into the “outlet” portion of the septum Fig 8, 9. Also coarctation of the aorta was diagnosed with a marked narrowing distal of the left subclavian artery Fig. 10.

The woman gave birth at 35 weeks and the newborn showed good adaptation to extrauterine life. The cardiac ultrasound confirmed the antenatal diagnosis of DORV with subpulmonary VSD, coarctation of aorta, including it in the Taussig-Bing heart pathology spectrum. Despite prostaglandin therapy the ductus arteriosus began to close, resulting in inadequate systemic perfusion and increased pulmonary pressure. Shortly after, heart failure and neonatal death occurred. Necropsy was performed and it confirmed the diagnostic of Taussig-Bing heart and aortic coarctation Fig. 11-13 (the arrows indicate the aorta and pulmonary artery arising from the right ventricle).

Discussions

It is often difficult to differentiate DORV – Taussig-Bing variant from a TGA with a VSD, especially prenatally using ultrasound. The altered cardiac anatomy makes it impossible to obtain the classic outflow tract sections and thus, as in our case we obtained images that suggested both the diagnosis of DORV and TGA. Nonetheless a clear diagnostic should be made as the two pathologies are mutually exclusive, both representing different forms of abnormal ventriculoarterial connections.