Prenatal diagnosed single ventricle associated with TAPVC, TGA, right isomerism, pulmonary stenosis

Introduction

Complex heart diseases, particularly the combination of single ventricle with right isomerism, TGA and PS associated with abnormal venous return is very rare. Such cases have severe postnatal clinical and consequently poor outcome.

Case report

A cardiac malformation was first detected by sonologist. Then patient was referred to our clinic for detailed fetal cardiac assessment. We presented the case of 28 years old woman (G2P1) at 28 gestational weeks of pregnancy. Single ventricle, TAPVR, TGA, PA stenosis, juxtaposition of the IVC and PLSVC were our findings in such a complex CHD. Moreover, the stomach was observed on the left side of the fetal abdomen, IVC anterior or juxtaposed to the aorta. No morphologically left atrium was detected accompanying by infracardiac type TAPVR and absent coronary sinus. Putting all these sings together our definitive diagnosis was right isomerism. The prognosis for the neonate would be unfavorable. Patients decided to terminate the pregnancy.

Discussion

Presented case right isomerism with absent interatrial septum forms single atrium anatomy. In case of TAPVR with single ventricle and single atrium it loses the essence of where the drainage of pulmonary venous return will occur. The important point is whether the TAPVR is obstructive and the amount of blood that has high saturation returns from the lungs to the heart chambers. Even if the pulmonary venous obstruction is not revealed during the pregnancy, it occurs in 50% of cases within the first 48 hours of postnatal life. Furthermore, because of amount of blood flow to the lungs as a result of pulmonary stenosis decreases, it is thought that cyanosis will be more severe with the need for early surgical intervention, and mortality increase.

Conclusion

This type of complex cardiac malformation with abnormal situs (right isomerism) is not compatible with life. Detailed examination during the pregnancy helps to reach the correct diagnosis to manage it properly. However, patients decided not to continue the pregnancy.