**Introduction**

Recently there has been a rise in the number of fetal heart disease cases concomitant with the increase of fetal heart screening, but fetal diagnosis of total anomalous pulmonary venous connection (TAPVC) is still difficult and rare. In this case, we were able to save the patient by diagnosing TAPVC with severe pulmonary vein obstruction (PVO) in the fetal period. We present the case to demonstrate the benefit of fetal TAPVC diagnosis.

**Case**

The mother, at 34 weeks of gestation, was referred to us with suspected fetal congenital heart disease, TAPVC being particularly likely because of the wide post-LA space which was evident in the fetal heart screening. Detailed fetal echocardiography was performed and we diagnosed the fetus as isolated TAPVC with severe PVO. The common chamber was small, with the vertical vein being narrow and meandering as it drained to the azygos vein. The flow pattern of both right and left pulmonary veins showed a continuous severe PVO pattern.

At 38 weeks, the baby was delivered. Postnatal echocardiography confirmed the fetal diagnosis. The baby showed severe cyanosis, needing urgent intubation and oxygen administration. Despite such medical treatment, his cyanosis and condition didn’t improve and an urgent TAPVC repair operation was performed on the day. The operation was successful and he was discharged from our hospital at 6 months old.

**Conclusion**

Without the correct diagnosis in the fetal period, the patient wouldn’t have survived. We must make an effort to identify the disease using effective screening methods such as post-LA space index.