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Case Report / Olgu Sunumu

Transcatheter management of obstructed pulmonary venous return and pulmonary atresia in a newborn with heterotaxy syndrome

Heterotaksi sendromlu bir yenidoğanda tıkalı pulmoner venöz dönüş ve pulmoner atrezinin transkateter tedavisi

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ABSTRACT

Heterotaxy syndrome refers to complex cardiac pathologies accompanied by several intracardiac development abnormalities. One of the main problems in these patients is total abnormal pulmonary venous connection. Although stenting the patent ductus arteriosus in newborns can be performed in several centers, stenting of the vertical vein in obstructed total abnormal pulmonary venous connection is not a well-established procedure. Herein, we report a newborn case with obstructive supracardiac total abnormal pulmonary venous connection and pulmonary atresia who was successfully treated with transcatheter palliation.

Keywords: Heterotaxy syndrome; newborn; obstructed total abnormal pulmonary venous connection; transcatheter management.

ÖZ

Heterotaksi sendromu, birçok intrakardiyak gelişim anomalisinin eşlik ettiği kompleks kardiyak patolojilerdir. Bu hastalardaki başlıca sorunlardan biri, total anormal pulmoner venöz bağlantıdır. Yenidoğanlarda patent duktus arteriyozus stenti birçok merkezde uygulanabilmesine rağmen, tıkalı total anormal pulmoner venöz bağlantıda vertikal ven stenti iyi tanımlanmış bir işlem değildir. Bu yazıda tıkalı suprakardiyak total anormal pulmoner venöz bağlantı ve pulmoner atrezisi olan, transkateter palyasyon ile başarılı bir şekilde tedavi edilen bir yenidoğan olgu sunuldu.

Anahtar sözcükler: Heterotaksi sendromu; yenidoğan; tıkalı total anormal pulmoner venöz bağlantı; transkateter tedavi.

Heterotaxy syndrome refers to complex cardiac pathologies accompanied by several intracardiac development abnormalities.[1] One of the main problems in these patients is cyanosis, which is a result of reduced pulmonary blood flow either pulmonary stenosis (PS) or pulmonary atresia.[2] Another problem is total abnormal pulmonary venous connection (TAPVC).[3] Management is adopted according to the obstruction in TAPVC and the severity of the PS.

Although stenting the patent ductus arteriosus (PDA) in newborns can be performed in several centers, stenting of the vertical vein in obstructed TAPVC is not a well-established procedure.[4,5] Herein, we report a newborn case with obstructive supracardiac TAPVC and pulmonary atresia who was successfully treated with transcatheter palliation.

CASE REPORT

A two-day-old male patient weighing 3.2 kg was referred to our clinic with pulmonary atresia. Upon physical examination, his overall condition was poor with significant dyspnea and cyanosis. The patient was initially intubated and admitted to the pediatric cardiac intensive care unit. His oxygen saturation level was 60% during oxygen supplementation and a prostaglandin-E_1 (PGE_1) infusion. Echocardiography showed right atrial isomerism, unbalanced complete atrioventricular septal defect, large PDA, obstructive supracardiac TAPVC in which the pulmonary veins were draining into a collector sac behind the atrium and also into the superior vena cava by means of a right-sided vertical vein, and pulmonary venous hypertension. Color Doppler ultrasound showed a

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turbulent flow in the mid-section of the vertical vein with a maximum 40 mmHg gradient. Chest X-ray revealed a bilateral multifocal ground glass appearance (Figure 1a). He had pulmonary venous hypertension and pulmonary edema due to the obstructed TAPVC.

An urgent cardiac catheterization was performed for stenting the obstructed TAPVC. A 5F sheath was inserted into the right jugular vein and a 4F sheath was placed into the right femoral vein using a percutaneous technique. Angiography revealed a large vertical PDA supplying blood to the pulmonary arteries, and venous return was delayed due to the narrowing in the vertical vein (VV) (Figure 2a). Intervention through the internal jugular vein was chosen, due to the angle of the VV into the superior vena cava (SVC). Simultaneous pressure recordings showed a 37 mmHg gradient between the pulmonary venous sac and the superior vena cava within the VV (Figure 2b). Following contrast injections, two stenotic sites were detected in the VV. Initially, a balloon was inflated inside the vertical vein and the stenotic sites were able to be localized to calculate the minimum length of the stent. In addition, the airway pressure was monitored during the balloon inflation to ensure that the stent would not compress the bronchus. Meanwhile, a 6×28 mm Omnilink® peripheral stent (Guidant Corp., Santa Clara, CA, US) was placed to cover both stenotic sites inside the VV within the 5F sheath (Figures 1b, 2c). Following the procedure, the ground glass appearance vanished (Figure 1b), and the patient was extubated on the second day of hospitalization. Following extubation, PGE_1 was discontinued during follow-up and PDA became narrower. Therefore, he was re-admitted to the catheterization laboratory for stenting of the PDA (Figures 1c, 2d). A 5F 4×20 mm coronary stent was advanced in the antegrade direction via a Judkins-R4 guiding catheter through the right femoral vein and implanted at the site of the ductus. The patient was followed in the intensive care unit for three days following the second procedure and discharged on the ninth day of his hospitalization. The patient was asymptomatic during follow-up and underwent Glenn operation + TAPVC repair, when he was six months old. He is now one year old and followed under scheduled follow-up visits.

DISCUSSION

Surgical or transcatheter intervention should be performed during the neonatal period to ensure the pulmonary blood flow in patients with pulmonary atresia. A poor overall preoperative condition, neonatal period, and comorbidities which require an urgent surgical intervention significantly increase the mortality of the intervention.[6] Early age, low body weight, and obstruction of the pulmonary venous return all adversely affect the results of a surgical repair.[7,8] Furthermore, several studies have shown that surgical intervention has unfavorable outcomes, when performed on obstructed TAPVC patients with a single functional ventricle in heterotaxy syndrome.[9] The mortality rate of simultaneous obstructive TAPVC repair with shunt palliation procedure in the neonatal period is extremely high in most centers.[9]

There is a high risk for mortality in patients with heterotaxy syndrome accompanied by complex cardiac pathologies, particularly in cases undergoing surgical palliation during the neonatal period.[9-11] In a study published in 1996, Sadiq et al.[10] reported death

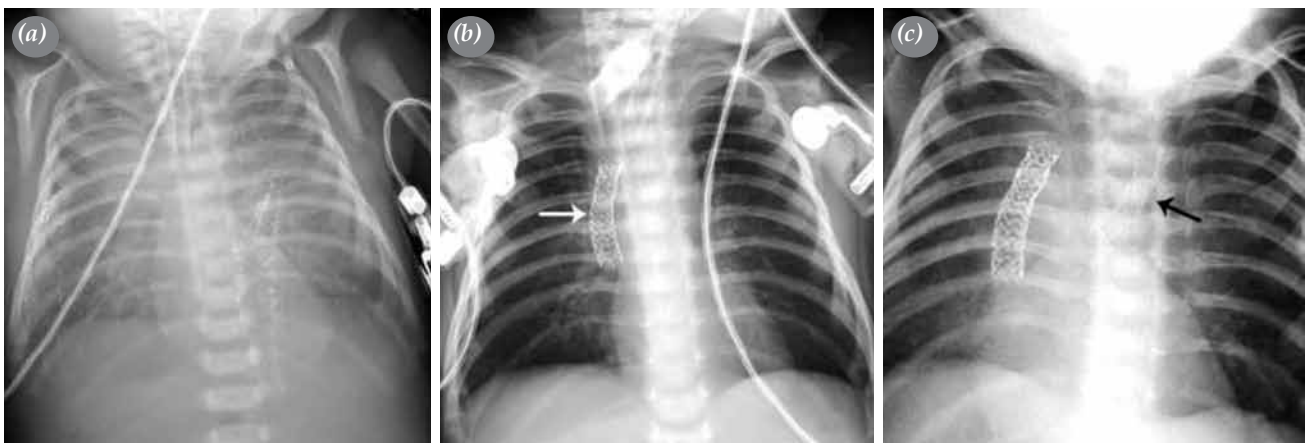

Figure 1. (a) A preoperative chest X-ray images showing a bilateral multifocal ground glassy appearance. **(b)** Following stenting, the vertical vein (white arrow shows the stent) ground glassy appearance was diminished. **(c)** A postoperative chest X-ray image following stenting the patent ductus arteriosus (black arrow shows the stent).

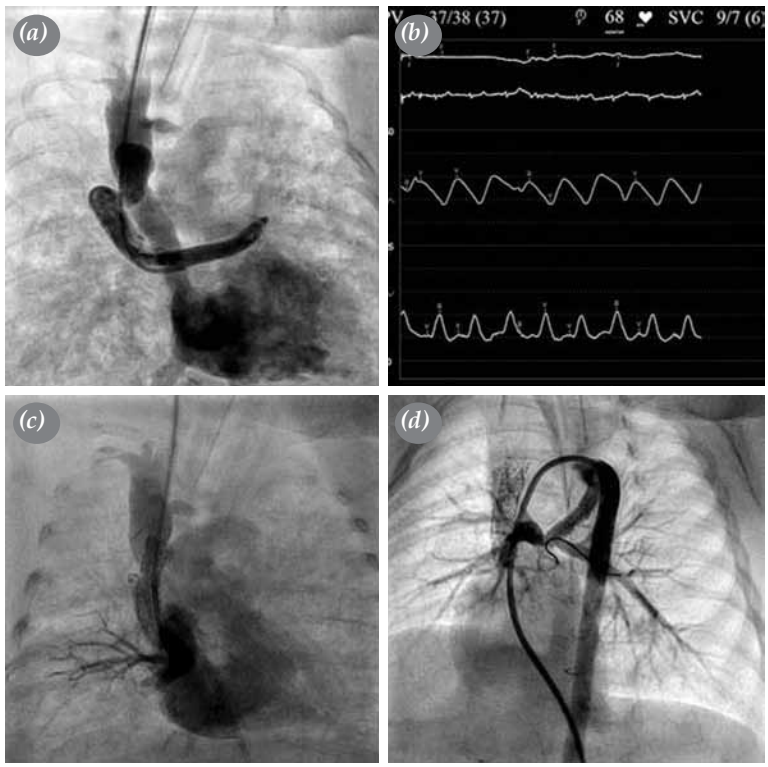

Figure 2. (a) A posteroanterior view of vertical vein injection showing the narrowing sites. **(b)** Simultaneous pressure recordings of the pulmonary vein and superior vena cava. **(c)** A posteroanterior view of the vertical vein stent. **(d)** An aortogram showing the pulmonary arteries filled by the duct stent. Also note the narrowing parts of the vertical stent.

PV: Pulmonary vein; SVC: Superior vena cava.

during the early period in seven out of 11 patients (64%) (eight patients with accompanying obstructive TAPVR) and in the late period in nine patients (82%).[10] Khan et al.[12] conducted a current, multi-center study and found that early period mortality continued to pose an important problem in heterotaxy syndrome patients who underwent TAPVR repair.[12] In another study, Nakayama et al.[9] reported that an age of <2 months and a body weight of <3.5 kg during surgery, as well as aortopulmonary shunt surgery constituted a significant risk factor for both early and late in-hospital mortality.[9] In several studies from the late 1990s, the mortality risk was observed to be significantly higher (25 to 60%) during the onset palliations in the neonatal period in patients with heterotaxy syndrome and single ventricle physiology, and this rate could increase up to 95% in the presence of accompanying obstructive TAPVR.[13,14] In another study reported by Ota et al.,[11] the patients with right atrial isomerism were divided into two groups as "pre-2003" and "2004 and later." The five-year survival rate was found to be 53.8% in pre-2003 patients, while it was 81.7% in the patient group of "2004 and later." The surgical results for right atrial isomerism patients had a tendency to improve and the palliative surgery practices in the neonatal period (particularly in those receiving shunt) constituted an important risk factor for early mortality.[11] Timing of the TAPVR is also of utmost importance in TAPVR patients with heterotaxy syndrome and single ventricle physiology. Sebastian et al.[15] showed that not performing a TAPVR repair during the neonatal period was a more appropriate approach.[15] However, these patients may face a high risk of surgical mortality, since the intervention is unavoidable in the neonatal period in the presence of obstructive TAPVR.[14] Thus, we believe that TAPVC repair may be postponed to a later period in isomeric pulmonary atresia patients with non-obstructed TAPVC. If the TAPVC is obstructive, an intervention should be performed during the newborn period. Transcatheter interventions available in selected cases, as in our case, may prevent a high surgical mortality in this patient population.

Currently, the mortality rates due to shunts placed in pulmonary atresia cases, where the only pulmonary blood source is a ductus remain at an undesired level. However, thanks to transcatheter PDA stenting performed at centers with experience in skilful hands, the patient can be safeguarded against the risk of neonatal shunt mortality. Our case, in whom we prevented the neonatal shunt mortality by implanting a transcatheter PDA stent, was also protected from the potential femoral artery complications thanks to the antegrade route for stenting through the femoral vein. This method also ensured a more comfortable stent advancement into the vertically extending PDA.

In conclusion, the delivery of a vertical vein stent and patent ductus arteriosus stent via a transcatheter route may be an alternative approach to surgical treatment in newborns with complex congenital cardiac diseases accompanied by obstructive total abnormal pulmonary venous connection and duct dependency.

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