Negative results - Congenital

Hybrid palliation for right atrial isomerism associated with obstructive total anomalous pulmonary venous drainage

Won-Kyoung Jhang\textsuperscript{a}, Yong-Jin Chang\textsuperscript{a}, Chun-Soo Park\textsuperscript{a}, Yeon-Mi Oh\textsuperscript{b}, Young-Hwue Kim\textsuperscript{b}, Tae-Jin Yun\textsuperscript{a,}\textsuperscript{*}

\textsuperscript{a}Division of Pediatric Cardiac Surgery, Asan Medical Center, College of Medicine, University of Ulsan, 388-1 Poongnap-dong, Songpa-gu, Seoul, Republic of Korea, 138-736

\textsuperscript{b}Pediatric Cardiology, Asan Medical Center, College of Medicine, University of Ulsan, Republic of Korea

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Abstract

A twenty-four-day-old girl, who was prematurely born at 36 weeks of gestation, and weighed 2.2 kg, and diagnosed with right atrial isomerism, functionally single ventricle, bilateral superior vena cava (SVC) and obstructive supracardiac total anomalous pulmonary venous drainage (TAPVD) draining to the junction between the right SVC and the right atrium, underwent a hybrid procedure in the operating room, which consisted of pulmonary artery banding, ductus ligation and stenting of the draining vein of TAPVD. Obstruction at the drainage site of TAPVD was initially relieved after stenting, but, one month after the procedure, the distal end of the stent became stenotic and she received bilateral sutureless repair of TAPVD. At postoperative seven months, she underwent bidirectional cavopulmonary shunt uneventfully, and she has been followed-up for two months in a stable state without any problem in the pulmonary venous pathway.

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Keywords: TAPVD; Right atrial isomerism; Hybrid procedure

1. Introduction

Although the prognosis of right atrial isomerism (RAI) associated with obstructive total anomalous pulmonary venous drainage (TAPVD) is very poor even in contemporary series \cite{1, 2}, aggressive TAPVD repair upon initial palliation was reported to improve the long-term outcome \cite{2}. Surgical intervention for TAPVD using cardiopulmonary bypass, however, may be too dangerous in small babies with RAI having multiple risk factors. We present a case of hybrid palliation for a baby with RAI and obstructive TAPVD without cardiopulmonary bypass.

2. Case

A female baby was born at 36 weeks of gestation with a birth weight of 2.2 kg. On fetal echocardiography, she was found to have RAI, unbalanced atrioventricular septal defect, double outlet right ventricle, large patent ductus arteriosus (PDA), bilateral superior vena cava (SVC), and supra-cardiac total anomalous pulmonary venous drainage (TAPVD) draining to the junction between the right SVC and the right atrium without obstruction. Initial oxygen saturation (SaO\textsubscript{2}) was 94% at room air. The postnatal echocardiography and cardiac computerized tomography (CT) (Fig. 1) confirmed the diagnosis. At the 10th day of life, she showed abdominal distension and elevation of C-reactive protein. On suspicion of neonatal sepsis and necrotizing enterocolitis, antimicrobial treatment was initiated. From the 15th day of life, she began to show desaturation (70%), tachypnea, and pulmonary venous congestion on chest X-ray. Follow-up echocardiography revealed that the draining site of TAPVD had become obstructive with a peak Doppler velocity of 2.2 m/s. Given her low body weight and suspected necrotizing enterocolitis, we elected to perform hybrid palliation. On the 24th day of life, she was brought to the operating room and underwent a hybrid procedure, consisting of (1) the ligation of PDA, (2) pulmonary artery banding (PAB), and (3) stent insertion into the draining vein of TAPVD (Fig. 2). For the intraoperative stenting, we used a balloon expandable drug eluting coronary stent (Endeaver\textsuperscript{m}, Medtronic Inc., Minneapolis), which was balloon dilated at 10 atmospheres with a final length and diameter of 12 mm and 4 mm, respectively. After the patient arrived in the intensive care unit (ICU), she showed fluctuations in SaO\textsubscript{2} and pulmonary artery pressure, which was deemed to be caused by increased pulmonary vascular reactivity after an abrupt relief of pulmonary venous obstruction. Thus, nitric oxide (NO) inhalation therapy was applied from then on. On postoperative day 4, she began to show a high SaO\textsubscript{2} (>95% at room air) and signs of pulmonary over-circulation, so NO therapy was discontinued and PAB was tightened in the ICU. She was extubated and transferred to the general ward at postoperative day 10 and 13, respectively. Postoperative echocardiography showed mild obstruction at the distal end of the stent with a peak velocity of 1.5 m/s in a continuous flow pattern at the SVC end of the stent, and

\*Corresponding author. Tel.: +82-2-3010-3589; fax: +82-2-3010-6966.
E-mail address: tjyun@amc.seoul.kr (T.-J. Yun).

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follow-up cardiac CT showed that the distal end of the stent was partially occluded by the lateral wall of the right SVC, which had become smaller than it had been preoperatively. Because the patient’s clinical state was relatively stable with mild desaturation (75% at room air), she was discharged on postoperative day 24 with a prescription of home oxygen therapy. Six days after the discharge, however, she came back to the hospital in a moribund state with severe cyanosis and tachypnea, necessitating endotracheal intubation and mechanical ventilatory support. Her body weight was 2.7 kg at this time. Echocardiography revealed a severe obstruction at the distal end of the stent with a peak velocity of 3 m/s. She was immediately taken to the operating room and underwent an emergency operation consisting of (1) retrieval of the stent, and (2) stentless repair of TAPVD. Intra-operative findings showed obstruction of the distal end of stent by the shrinkage of the right SVC, which was presumably caused by the catheter intervention on the thin wall of the right SVC (Fig. 2b). The stent per se was patent without thrombosis or abnormal tissue in-growth. Postoperative course after the 2nd operation was uneventful, although she needed NO inhalation for six days and delayed sternal closure. Postoperative echocardiography showed normal biphasic flow pattern of the individual pulmonary veins without obstruction and adequately tight pulmonary artery banding with a flow velocity of 3.5 m/s. She was discharged home on postoperative day 38. At postoperative seven months, she underwent bidirectional cavopulmonary shunt and division of the main pulmonary artery uneventfully, and she has been followed-up for two months in a stable state without any problem in the pulmonary venous pathway.

3. Comments

Although RAI has still remained a clinical entity with a dismal prognosis [3, 4], contemporary surgical series have indicated that aggressive initial treatment of TAPVD, whether obstructive or not, may improve the long-term outcomes [2]. For the repair of TAPVD, primary stentless repair has become the procedure of choice because conventional repair for RAI-associated TAPVD may lead to post-repair pulmonary vein stenosis [1]. In this case, however, we elected to perform hybrid palliation for TAPVD, instead of primary stentless repair, because the risk of stentless technique, employing cardiopulmonary bypass and cardiac arrest, would have been too high, given her low body weight and suspected neonatal sepsis. Stenting of the obstructive vertical vein in isolated TAPVD has been reported as a rescue procedure for critically ill patients [5, 6], but stenting of the obstructive TAPVD in patients with RAI and functionally single ventricle as a part of a hybrid procedure has not been reported yet. In our experience, this novel technique was deemed partially successful in that TAPVD repair had to be performed on an emergency basis before the 2nd stage operation (i.e. bidirectional cavopulmonary shunt). Nevertheless, it is prudent to say that the risk of stentless repair of TAPVD would have been greater if the patient had undergone the procedure in the first place. Furthermore, if there had not been shrinkage of the right SVC, or if the right SVC had been larger than it really was, the stent would have remained patent for a longer term period.

To employ this novel technique, the draining vein of TAPVD should be surgically accessible. In this regard, cardiac/infra-cardiac type TAPVD with draining site obstruction, or mixed type TAPVD without supra-cardiac drainage, may not be amenable to this approach. Patients with long segment obstruction or individual pulmonary vein stenosis can also be regarded as poor candidates for this procedure.

In conclusion, for critically ill neonates with RAI and obstructive TAPVD, stenting of the draining vein of TAPVD, along with other procedures, can be a potent option for stabilization. Favorable morphology of TAPVD and draining vein, adequate size of the SVC or innominate vein where
stents are to be placed, minimal injury to the vessels upon catheter intervention, and meticulous post-stenting surveillance are caveats of this approach for long-term durability.

References


