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Case Reports

Supracardiac Total Anomalous Pulmonary Venous Return Repair in a 7-Month-Old Infant

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Total anomalous pulmonary venous return is a congenital heart malformation characterized by anomalous pulmonary venous inflow to the right atrium. Surgical repair typically occurs during the first month of life, and survival beyond that age in untreated patients is unlikely. We report an extreme case of supracardiac total anomalous pulmonary venous return in an infant who survived 7 months despite atypical anomalous inflow without atrial-level communication and with right-to-left shunting only through a patent ductus arteriosus. We stabilized the patient's left-sided heart function before surgically repairing the anomalous venous return 2 months later. Three years postoperatively, the patient was well. ***(Tex Heart Inst J 2022;49(3):e207393)***

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Total anomalous pulmonary venous return (TAPVR), a congenital cardiac malformation characterized by anomalous pulmonary venous inflow to the right atrium (RA), accounts for 1% to 3% of all birth defects.[1] Of the 4 types of TAPVR, the supracardiac type occurs most often (45%–55% of cases), followed by the cardiac or coronary sinus (20%–25%), infracardiac (25%), and mixed types (5%).[2,3] Surgical refinements and advances in hospital care during the last 50 years have improved the survival rate of patients with TAPVR; early mortality rates of less than 10% have been reported after surgical correction.[4,5] We report a severe case of supracardiac TAPVR in an infant with atypical anomalous inflow who survived for months without atrial-level communication, and with right-to-left shunting only through a patent ductus arteriosus (PDA).

Case Report

A male infant was delivered at term by means of emergency cesarean section because of deceleration in his cardiotocogram. The first-time mother's pregnancy had been normal. The infant's birth weight was 5 lb 12 oz (2,600 g), and his Apgar score was 9 at one minute and 10 at 5 minutes. On postnatal day 3, his oxygen saturation was 80%, and a transthoracic echocardiogram (TTE) revealed possible TAPVR. During subsequent outpatient monitoring in the patient's native country, further medical follow-up was recommended because it was thought to be too early to correct the condition surgically. The infant became cyanotic during feeding and failed to thrive. At 7 months, after evaluation at a specialized international congenital heart center, he was considered to be in the terminal phase of TAPVR and thus inoperable.

The infant was admitted to our institution in dismal clinical condition with severe heart failure. After initial resuscitation, a TTE revealed the spontaneous closure of the foramen ovale (Fig. 1A) and extremely enlarged right-sided heart chambers (Fig. 1B). Apart from right-to-left shunting through a PDA, the infant's entire circulation was draining into the right side of the heart, which was functioning well. Conversely, the left side of the heart was completely collapsed with a closed aortic valve (Fig. 1C), similar to the circulation in patients who have hypoplastic left heart syndrome with retrograde flow in the aortic arch. Computed tomographic findings confirmed the diagnosis of atypical supracardiac TAPVR (Fig. 2A). Findings of right-sided heart

catheterization revealed supracardiac TAPVR with an atypical anomalous confluence of the pulmonary veins (PVs) to the superior vena cava (SVC), enlarged right-sided heart cavities and pulmonary artery (PA), a PDA, and PA hypertension with systemic pressure (Fig. 2B).

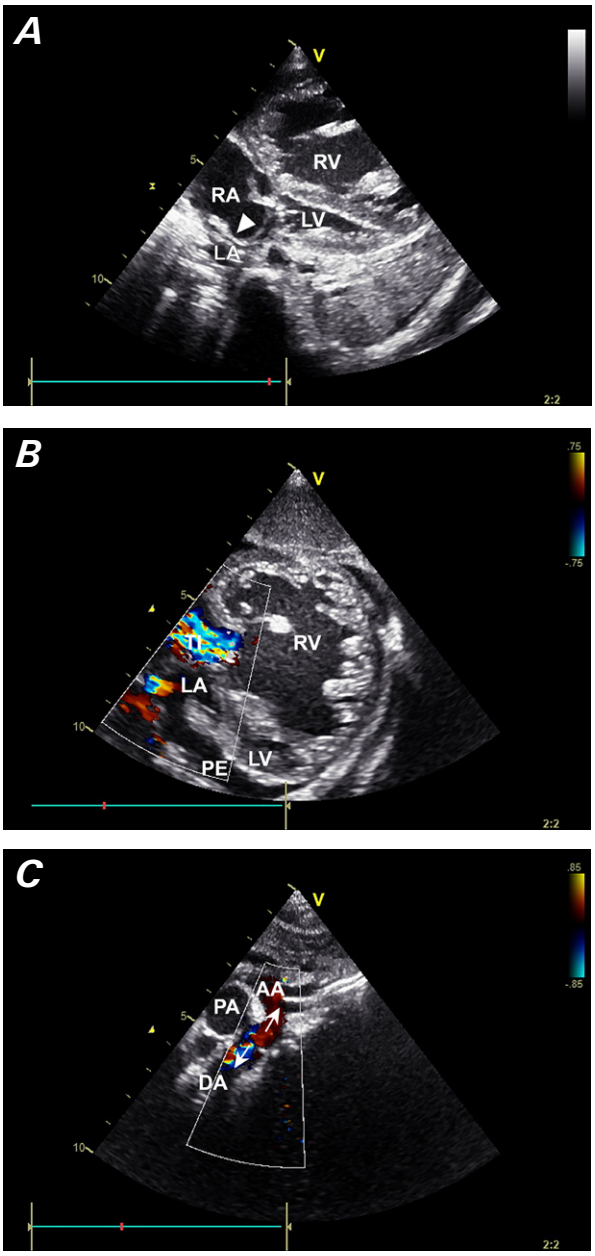

Fig. 1 *Transthoracic echocardiograms at the time of index admission.* ***A****) Apical 4-chamber view shows a closed foramen ovale (arrowhead), enlarged right-sided heart chambers, and residual cavities in the left side of the heart.* ***B****) Apical 4-chamber view shows severe tricuspid insufficiency (TI), an enlarged and hypertrophic right ventricle (RV), and a pericardial effusion (PE).* ***C****) Subxiphoid short-axis view shows a slender aorta filled retrograde through a patent ductus arteriosus (arrows indicate flow direction).*

AA = aortic arch; DA = descending aorta; LA = left atrium; LV = left ventricle; PA = pulmonary artery; RA = right atrium

Despite several attempts, percutaneous balloon atrial septostomy was not successful. We decided to perform an open RA septectomy on the 4th day of the patient's hospital stay. Aortic and bicaval cannulation and atriotomy of the grossly enlarged RA were performed through a standard median sternotomy. The RA septectomy resulted in a 15-mm-wide right-to-left shunt. The surgery and the patient's postoperative course were uneventful. A TTE revealed persistent PA hypertension and a bidirectional shunt at the atrial level (Fig. 3A).

Two months postoperatively, during the same hospitalization, results of right-sided catheterization and a TTE revealed that the left side of the patient's heart was well conditioned for systemic circulation, and the pediatric heart team decided to perform surgical TAPVR repair. Through a standard median sternotomy, aortic and bicaval cannulation was performed in the usual

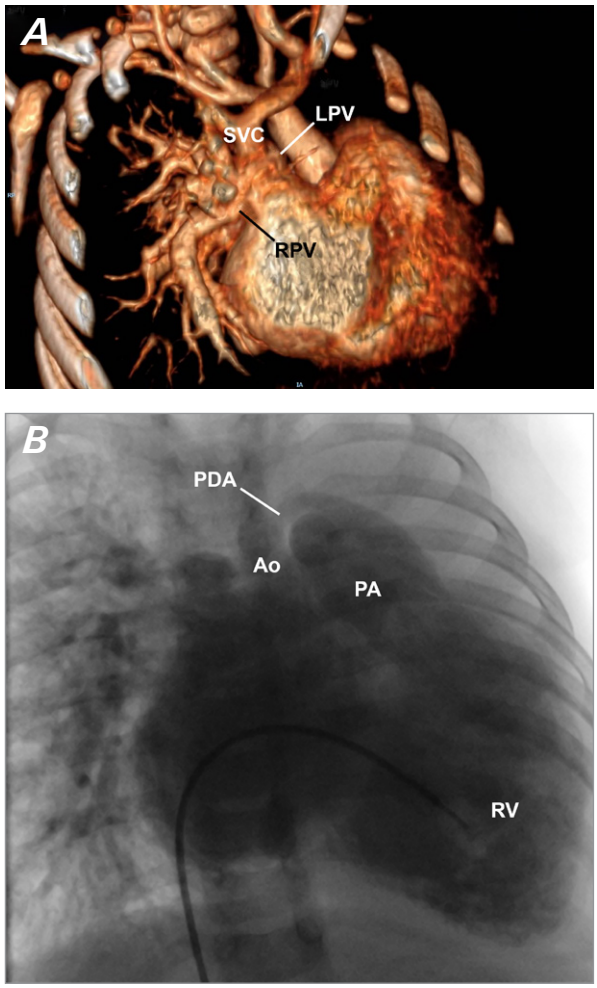

Fig. 2 *Before surgical repair at 7 months,* ***A****) a multislice computed tomogram (3-dimensional reconstruction) shows atypical total anomalous pulmonary venous return without vertical vein.* ***B****) Fluoroscopic image shows an enlarged right ventricle (RV) and patent ductus arteriosus through which the aorta (Ao) is filled.*

LPV = left pulmonary vein; PA = pulmonary artery; RPV = right pulmonary vein; SVC = superior vena cava

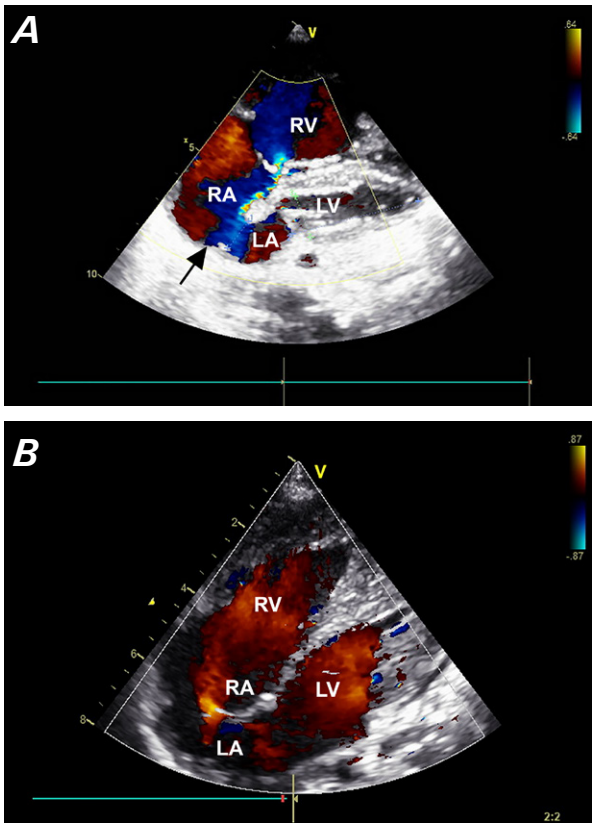

Fig. 3 *Transthoracic echocardiograms (left parasternal long-axis views).* ***A****) Persistent pulmonary artery hypertension and a bidirectional shunt (arrow) at the atrial level are seen immediately after open atrial septectomy.* ***B****) Two months later, the left ventricle (LV) is developed and dominant.*

LA = left atrium; RA = right atrium; RV = right ventricle

fashion. Intraoperatively, the following conditions were observed: inflow of the right PVs to the RA–SVC connection, flow from the left PVs without vertical vein to the same RA–SVC connection, and a greatly enlarged RA and right ventricle. The PDA was directly closed. After aortic cross-clamping and root administration of cardioplegic solution, the RA was opened. With use of 6-0 polypropylene sutures, a polytetrafluoroethylene Gore-Tex baffle patch (W.L. Gore & Associates, Inc.) with a 5-mm fenestration was attached to the atrial septal defect that had been surgically created previously, so that the PVs were connected to the left atrium through the defect. The SVC was detached from the RA, and the stump was closed with an equine pericardial Matrix Patch (Autotissue). The Warden procedure was then performed, with use of a Matrix Patch, to modify the SVC inflow in the RA. Sinus rhythm was observed after aortic unclamping.

On postoperative day 1, the patient was weaned from mechanical ventilation; on day 7, enteral feeding was introduced. He was discharged from the hospital 14 days after the second surgery, in a good clinical state.

Two months after the second operation, a TTE showed a developed, dominant left ventricle; mild tricuspid insufficiency with normal PA pressure; and spontaneous subtotal occlusion of the patched fenestration at the level of the atrial septum (Fig. 3B). Three years postoperatively, the patient's TTE showed normal results, he was taking no medications, and he was developing adequately.

Discussion

Our patient had an anatomic variant of supracardiac TAPVR in which a vertical vein did not connect to the brachiocephalic vein; instead, a confluent formation directly connected all PVs to the SVC-to-RA inflow. Of note, our patient survived for 7 months without atrial-level communication and only because of right-to-left shunting through a PDA. According to Harada and colleagues,[6] the median age for TAPVR repair is 1 month, with survival prospects worsening at later ages. In our patient, percutaneous atrial septostomy was attempted; however, it was not successful because of the small left atrium and the fibrous tissue of the closed foramen ovale. Careful preoperative planning enabled us to perform a surgical atrial septectomy to promote the growth of the left side of the heart. Although a Blalock-Hanlon operation without cardiopulmonary bypass was an option, we thought that partial atrial clamping would cause more hemodynamic compromise than would short cross-clamping of the aorta. Thus, the prudent first step was to enable left-sided volume growth and conditioning of the left ventricle so that TAPVR repair could later be performed.

Conclusion

After accomplishing left-sided cavity enlargement and stabilizing our patient, we successfully repaired his rare variant of supracardiac TAPVR. Although patients with TAPVR who are in a terminal state of disease rarely survive beyond newborn age without emergency surgery, we consider it prudent to tailor approaches to individual cases.

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