Anomalous right-sided pulmonary venous connection to the superior vena cava

Connessione anomala di vena polmonare destra alla cava superiore

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Over the years, different techniques have been introduced for the repair of sinus venous atrial septal defect (ASD) with anomalous right-sided pulmonary venous connection to the superior vena cava. We report the case of a 9-year-old girl, who presented with dyspnea and peripheral cyanosis. Preoperative echocardiography and angiography findings suggested a partial anomalous pulmonary venous connection. On cardipulmonary bypass, the ASD was dilated, and the anomalous pulmonary vein was anastomosed to the right atrium and redirected to the left atrium using an intraatrial baffle and a tube graft. The intraoperative and postoperative periods were uneventful, and the patient is currently in good health at 4.5 years’ follow-up.

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patch with 5-0 polypropylene so that it could be drained into the left atrial cavity (Figure 2). The suture line of the intraatrial patch was placed away from the sinus node and the pulmonary veins. The SVC defect was repaired with a pericardial patch. The aortic clamp was released, and the patient was weaned off bypass. The sternotomy was closed, and patient’s sinus rhythm was recovered.

The postoperative course was uncomplicated, and the patient was discharged on the seventh postoperative day. At discharge, an echocardiography investigation showed an LVEF of 55% and mild TR with a PAP of 22 mmHg. She has been followed in a Cardiology Clinic every year ever since. She had an echocardiography revealing mild TR and mild RV dilatation with PAP of 43 mmHg 9 months ago. Currently at 4.5 years’ follow-up, she is on no medications and remains asymptomatic.

### Discussion

Surgical repair for PAPVC to the SVC ideally includes a complete closure of the septal defect and redirection of the anomalous pulmonary veins into the left atrium without pulmonary venous or SVC obstruction or injury of the sinus node or its blood supply. However, when the veins enter further up the SVC, surgical treatment can be more complex [1]. Most commonly, surgical correction of these lesions is performed using the internal patch technique, or a modification thereof, to redirect the anomalous pulmonary venous return through the sinus venosus defect by baffling these structures with a pericardial or synthetic patch with or without performing a patch cavoplasty as needed [3, 4]. If the insertion of the pulmonary veins is more than 2 cm above the atrio caval junction, the surgeon may face a challenging...
In 1984, Warden and colleagues [8] reported a new technique for high PAPVC, in which caval continuity was established by direct cavoatrial anastomosis. The SVC was divided above the orifices of the anomalous pulmonary veins, and the cephalic end of the divided SVC was then anastomosed directly to the right atrial appendage after amputation of the tip. A patch within the right atrium diverted the pulmonary blood flow from the orifice of the SVC through the associated sinus venosus ASD. The cavaal end of the divided SVC was closed by sutures. Further clinical experiences with this technique or a modified one have been reported by other authors [1, 3, 9, 10]. De Leon and coworkers [11] have described an incision crossing the atrio caval junction. Having recognized that the incision was carried across the region of the sinoatral node, they subsequently modified their approach by creating two separate incisions, one in the lateral atrial wall and one on the distal SVC, to avoid injury to the sinus node or its artery.

In our patient, we applied an innovation for the reconstruction of the high PAPVC. The connection of the right anomalous pulmonary veins was separated from the SVC. Then the proximal end of the extended pulmonary vein was anastomosed directly to the RA. In this technique, two autologous pericardial patches were needed to repair the pulmonary orifices on the SVC and the dilated ASD. The procedure had some advantages. Firstly, it avoided long baffles and extensive SVC patching. In contrast to the Warden procedure, the SVC was not divided into the cephalic and caudal ends; therefore, the caval continuity was intact. In the SVC translocation with the highest anomalous pulmonary vein, it can be assumed that the cephalic end of the divided SVC becomes more distant from the right atrial appendage and the cavoatrial anastomosis has more tension and likelihood of stenosis [1]. Another advantage of this technique was the avoidance of the creation of conduits inside the SVC. In our technique, however, the foreign material as a tube graft was settled to enhance the right pulmonary vein and redirect anomalous pulmonary venous flow into the LA.

The repair of the high insertion of the anomalous pulmonary veins by redirecting the venous return through the sinus venosus ASD by using intraatrial baffling can be a good choice with a favorable long-term prognosis. The reanastomosis to the RA and extension of the right anomalous pulmonary vein is a safe and effective correction for PAPVC with high insertion into the SVC, and avoids long baffles, extensive SVC patching, and caval translocation.

**References**