Case Report

Transcatheter management of post-surgical superior vena cava obstruction

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ABSTRACT

Obstruction of the superior vena cava (SVC) is a rare complication after cardiac surgery in infants and children. We present the case of a 2-year-old male child who underwent surgery to repair a mixed total anomalous pulmonary venous connection. After 18 months of surgery, the child developed SVC syndrome. Transcatheter stenting was performed to relieve the SVC obstruction detected with transthoracic echocardiography. The patient was discharged after 5 days of hospitalization. The patient was doing well at follow-up appointments, with good laminar flow through the stent. In conclusion, transcatheter management of post-surgical complications of SVC obstruction was successful in this patient.

Key words: Mixed total anomalous pulmonary venous connection, Superior vena cava syndrome, Transcatheter stenting, Transthoracic echocardiography, Warden procedure

Superior vena cava (SVC) syndrome is an uncommon but serious complication that occurs after cardiac surgery in infants and children [1,2]. It is caused by the constriction or obstruction of the SVC, resulting in reduced venous drainage into the heart from the head, neck, and upper extremities [2]. This causes engorgement of blood in the upper portion of the body resulting in swelling and elevated pressures [1,3]. According to a systematic review, children with cardiac, malignant, or hematologic conditions may experience external compression and develop SVC syndrome [2]. The main underlying cause of SVC syndrome in children and adolescents has been identified as cardiac surgery for congenital heart defects [4]. Furthermore, the presence of surgical anastomosis and anatomical remodeling is regarded as plausible causes of SVC obstruction after cardiac surgery [5,6]. Pediatric SVC syndrome has been linked to substantial airway blockage and respiratory distress [7].

This case report describes our experience diagnosing and managing a case of SVC syndrome in a child after the repair of a rare congenital heart defect, which will help improve the existing clinical knowledge on pediatric SVC syndrome.

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CASE REPORT

A 2-year-old male child presented with facial swelling and mild respiratory distress for 1 month. He was diagnosed with mixed total anomalous pulmonary venous connection (TAPVC).

The upper right pulmonary vein was draining at a higher position into the SVC, whereas the lower right pulmonary vein opened into the junction between the SVC and right atrium. The left-sided pulmonary veins formed a common chamber, which was draining directly into the right atrium.

At 6 months, the Warden procedure for rerouting pulmonary veins was performed. In this procedure, the SVC was divided, and the end of the proximal SVC was closed. The right atrial appendage (RAA) was then resected, and the distal end of the resected SVC was anastomosed with the RAA. Subsequently, the atrial septal defect (ASD) was enlarged and closed using a Dacron patch. Consequently, the right-sided pulmonary venous drainage was redirected to the left atrium and the common chamber was anastomosed to the left atrium (Fig. 1).

The post-operative follow-up was uneventful. However, after 18 months, the patient developed SVC syndrome. His vitals were within normal limits. He had marked facial and neck swelling (Fig. 2a and b), engorged veins, and mild respiratory distress. The condition was functionally classified as the New York Heart

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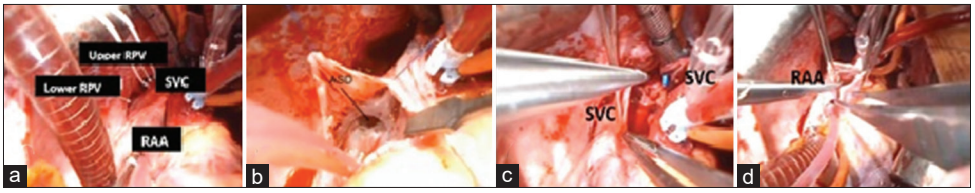
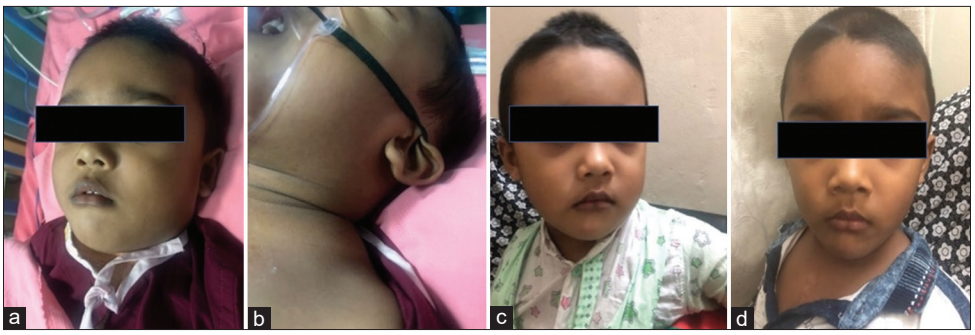


Figures 1: (a-d) SVC was divided; the end of proximal SVC was closed. The RAA was resected; the distal end of the resected SVC was anastomosed with RAA. The ASD was enlarged, closed with a patch and finally the right-sided pulmonary venous drainage was redirected to the LA and the common chamber was anastomosed to the LA. ASD: Atrial septal defect, LA: Left atrium, RAA: Right atrial appendage, RPV: Right pulmonary vein, SVC: Superior vena cava

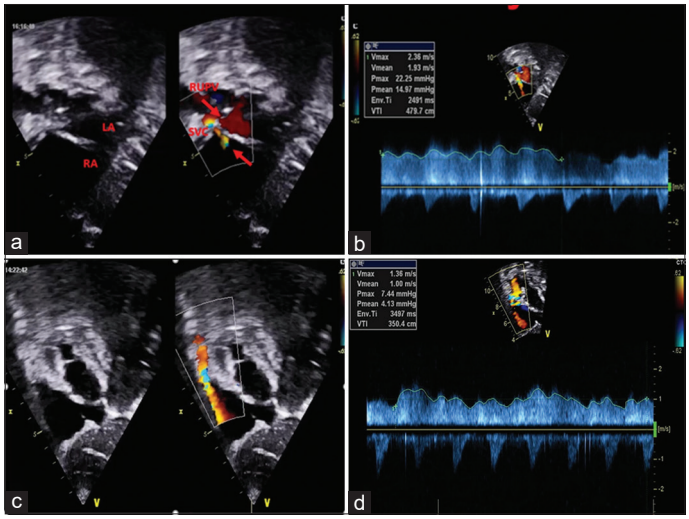


Figures 2: (a and b). SVC syndrome: Facial and neck swelling; (c). After a 1-month follow-up; (d). After 3-month follow-up. SVC: superior vena cava

Association Stage II category with no desaturation or pallor. The wound scar mark was healthy with no obvious visible pulsation. There were no signs of the right heart failure or a right heart failure murmur.

Electrocardiography and chest radiography findings were normal. Transthoracic echocardiography revealed an obstructed SVC flow with a peak gradient of 22 mmHg. Flow to other pulmonary veins was normal with no residual flow through the ASD. Mild pulmonary hypertension was observed. All chambers were of normal size with good biventricular functions (Fig. 3a and b). Transcatheter management of post-surgical SVC obstruction was planned thereafter**.**

The 5F Cordis vascular access system was used to obtain both femoral and jugular access. The patient was heparinized and a multipurpose catheter was introduced to check the hemodynamics. The right atrial, right ventricular, and pulmonary artery (PA) pressures were recorded. Pulmonary angiography revealed normal anatomy. Diluted contrast injection was given through the left forearm vein, which helped identify multiple venovenous collaterals. An angiogram from the internal jugular venous line (5F Cordis vascular access) revealed narrowing at the SVC site immediately distal to the connection with the innominate vein. A 014, 260 cm Run-through Floppy Coronary Guidewire (Terumo) was introduced, maneuvered, and parked into the right PA. A 4 mm × 15 mm non-compliant balloon was used to pre-dilate the stenosed segment and inflate it to 10 atm. A post-inflation angiography showed angiographic improvement, but no gradient reduction. A Nefro (Balton) peripheral stent 8 mm × 12 mm was positioned and inflated to 6 atm and later post-dilated up to 8 atm (Fig. 4). After completion of the procedure, an angiogram revealed good unobstructed flow through SVC to RA with a peak gradient of 7 mmHg (Fig. 3c and d). The patient was discharged after 5 days of hospitalization.

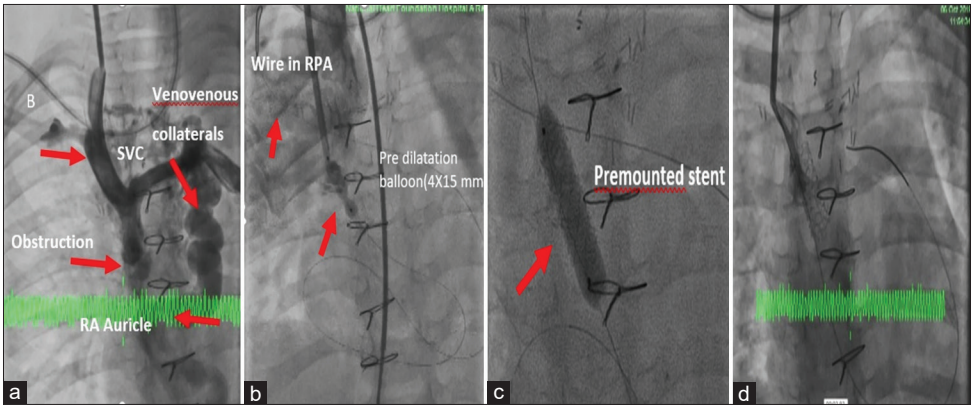


Figures 3: (a and b). A subcostal bicaval view of the pre-procedure transthoracic echocardiography (TTE) showed an obstructed SVC flow with a mean gradient of 15 mmHg and peak gradient of 22 mmHg. Good flow to other pulmonary veins was observed; (c and d). The post-procedure TTE showed decline of pressure gradient from 22/15 mm Hg to 7/4 mm Hg. A good unobstructed flow was seen through SVC to RA. SVC: Superior vena cava, RA: Right atrium

The patient was called for an initial follow-up at 1 month and for subsequent follow-ups at 3, 6, and 12 months. There was a marked improvement in the patient's facial and neck swelling at 1-month and 3-month follow-ups (Fig. 2c and d). The patient was doing well 6 months after surgery, with normal activity and weight gain. Laminar flow was noted through the SVC stent with a peak gradient of 5 mmHg.

DISCUSSION

The patient, in this case, underwent surgery to repair mixed TAPVC, a rare congenital heart defect; however, 18 months after



Figures 4: (a) Diluted contrast through venous access showing venovenous collaterals; (b) Run through floppy wire parked at RPA across the stenosis and pre-dilatation of the stenosed segment; (c) Pre-mounted Nefro stent (12 mm × 8 mm) dilated at 6 atm; (d). Post-stenting angiogram showed unobstructed flow. RPA: Right pulmonary artery, SVC: Superior vena cava, RA: Right atrium

the procedure, SVC syndrome manifested as a post-operative complication. This could be attributed to a relatively slow occlusion with negligible signs during the early course, wherein the patient is asymptomatic followed by a sudden development of SVC syndrome with complete venous obstruction and a variety of symptoms such as swelling of the face, neck, and arms; coughing; shortness of breath; difficulty swallowing; and engorged veins in the chest or neck [4]. Consistent with the findings about the association of pediatric SVC syndrome with significant airway obstruction and respiratory symptoms, the patient displayed marked facial and neck edema with engorged veins and mild respiratory distress, indicating the onset of SVC syndrome [7].

Transcatheter stenting and open surgical bypass are two treatment options for SVC syndrome patients with malignant and benign causes as they provide immediate symptom relief and allow the patient to continue treatment for the underlying condition [8]. The use of transcatheter stents is the preferred primary treatment modality over open reconstruction surgery because of low post-operation morbidity and fast recovery time [2,9]. Compared to balloon angioplasty, endovascular stenting relieves SVC obstruction more effectively with a lower recurrence rate [10]. A study conducted by Tzifa *et al.* demonstrated that endovascular therapy is the first-line treatment that alleviates SVC obstruction and relieves accompanying symptoms while providing long-term benefits [5].

CONCLUSION

Mixed TAPVC is a rare condition noted in children; the occurrence of SVC syndrome after cardiac surgery is less common in infants and children. Transcatheter management of this complication was successful in the current case.

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