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CASE REPORT

Functionally univentricular heart with systemic venous anomalies: surgical palliation and pulmonary arterial reconstruction with a roll of left atrial appendage

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Abstract

In this report, we describe a 3-year-old patient with a functionally univentricular heart (UVH), who had a combination of double outlet right ventricle (DORV) along with an unrouteable interventricular communication (VSD), severe infundibular and pulmonary valvar stenosis, and severe left pulmonary artery (LPA) ostial stenosis. This patient also had an interrupted inferior caval vein (IVC) with bilateral superior caval veins (SVC). We were able to undertake a successful Kawashima procedure with interruption of the antegrade pulmonary blood flow, reconstructing the LPA using a pedicled roll of the left atrial appendage (LAA).

Keywords Kawashima · Pulmonary artery reconstruction · Left atrial appendage

Introduction

In patients with an interrupted IVC, all the venous return from lower half of body (other than the hepatic venous return) drains via the azygous venous system into a SVC [1]. The hepatic veins typically form a confluence, cross the diaphragm, and most often drain directly into the atrium receiving the remainder of the systemic venous return. On occasion, however, IVC interruption can be found with usual atrial arrangement. Without other intracardiac malformations, this drainage of the systemic veins into the right-sided atrium (RA) is asymptomatic. It assumes importance if associated intracardiac malformations requiring surgical correction are present. Association of IVC interruption with bilateral SVC may be seen in some patients [2]. Should patients with an interrupted IVC also have a functionally UVH, and restricted pulmonary blood flow, the surgical procedure of choice is a bidirectional superior cavopulmonary anastomosis (BDG), known as the Kawashima procedure. When this combination is encountered with bilateral SVC, then the BDG must be constructed bilaterally. These patients may also have varying degrees of right ventricular outflow tract obstruction with narrowing of the pulmonary arteries requiring reconstruction. A flap of the LAA has been used in the past for a variety of right ventricular outflow tract obstructions [3, 4]. Use of the LAA as a roll to bridge the defect between pulmonary artery segments, however, at least to the best of our knowledge, has not been described. In this report, we discuss the unique morphological aspects and intra-operative management of one patient with these unusual combinations of cardiac malformations.

Case report

A 3-year old male child, weighing 9 kg, presented with cyanosis on feeding. He had been failing to thrive since the age of 3 months. On physical examination, there was central cyanosis and mild tachypnea. Resting saturation on room air was

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75%. An ejection systolic murmur (grade 3/4) was audible in the left 2nd intercostal space and was conducted all over the precordium. Chest radiograph revealed usual arrangement of the abdominal organs, an enlarged heart occupying two-thirds of the chest width, an apex of "right ventricular type," and evidence of reduced flow of blood to the lungs.

Transthoracic echocardiography showed DORV with a large VSD, but unrouteable to the aorta. There was infundibular and pulmonary valvar stenosis, with a peak gradient of 80 mmHg across the right ventricular outflow tract. There were bilateral SVC, with the left one draining through a dilated coronary sinus (CS) into the left atrium (LA). The coronary sinus appeared unroofed. Injection of agitated saline into the left antecubital vein showed air bubbles entering the LA, thus confirming this finding. There was suspicion of interruption of the IVC.

As is the policy at our institution, a computed tomography (CT) angiogram (Fig. 1) was performed to confirm the diagnosis and it confirmed the normal arrangement of the abdominal and thoracic organs, with normal venous connections apart from the bilateral SVC and an interrupted IVC without any communicating channel between the SVC. The right-sided SVC drained into the morphologically right atrium (RA) as expected. The images also showed that the left SVC drained into a dilated CS, but that the sinus itself opened into the morphologically LA. The infrahepatic portion of the IVC was absent, with the hepatic veins draining directly into the morphologically RA. The interrupted IVC continued as a left-sided hemizygous vein, which then drained into the left-sided SVC. The pulmonary venous drainage was normal. There was a large atrial septal defect in the oval fossa, but the images did not show whether there was another communication through the mouth of the CS. The atrioventricular connections were concordant, with the hypoplastic left ventricle emptying to a dominant and hypertrophied morphologically right ventricle through a 2-cm VSD that was remote from the aorta and extended into the inlet septum. There was also grade 2 straddling of the tricuspid valve. The ascending aorta was large and was situated anterior to the hypoplastic pulmonary trunk. The right pulmonary artery (RPA) was 7 mm in diameter, but the LPA was severely stenosed at the site of attachment of a large persistently patent arterial duct (PDA). The narrowest diameter was no more than 2 mm, although the artery expanded distal to the stenosis, with a diameter of 6.6 mm. The descending aorta measured 13.4 mm at the level of diaphragm. The aortic arch was left-sided, with the large PDA occupying its expected position, but with a focal narrowing at the site of its insertion into the LPA. The diagnosis was that of a patient having a functionally UVH, with bilateral SVC and an interrupted IVC draining via the hemizygous vein and CS into the LA. Bilateral BDG without interrupting the left-sided hemizygous vein was planned. We reasoned that this modification of the Kawashima procedure would direct the systemic venous return into the pulmonary circulation, thus improving the arterial saturation. Ligation and division of the PDA, with reconstruction of the LPA was also planned.

After median sternotomy, standard cardiopulmonary bypass (CPB) was established after cannulating the aorta and the RA, with the heart kept beating under normothermia. Both SVC were dissected and widely mobilized. The azygous vein was not visualized on the right side, even after complete dissection of the right SVC. A large hemizygous vein was present draining into the left SVC, but was not divided since it carried all the venous return from the IVC. PDA was divided. The LPA was divided at its site of union with the PDA, and ductal tissue was excised completely. Even after complete mobilization of both the pulmonary arteries up to the hilum of both lungs, it was not possible to reconnect the ends of the divided LPA. Hence, we created a roll from the LAA, which was interposed between the ends of the LPA, thus restoring its continuity (Fig. 2). Posterior suturing was onto pericardium and both ends of the LPA like in a suture less repair. Anteriorly, it was augmented using in situ LAA as a roll to restore the luminal continuity of the LPA. As the LAA appendage was used as a pedicled roll without violating its lumen, its ligation was not performed.

Following LPA reconstruction, the right SVC was divided and its cardiac end was sutured closed. Using the open technique of BDG as previously described by us [5], we created an end-to-side anastomosis between the right SVC and the RPA. The left SVC was similarly divided at its cardiac end, well below the insertion of the hemizygous vein, with two cardiotomy suckers placed to drain the SVC and IVC return. We then created a standard anastomosis between the left SVC to LPA, having placed a clamp on the origin of the RPA to avoid flooding of the operative field by the blood coming from the right BDG.

Following completion of the bilateral BDG, we weaned the patient from CPB, noting the favorable rise of systemic saturations to 90%. At this the time, the FiO2 was set at 60%, the tidal volume was set at 8 ml/kg without any positive end expiratory pressure (PEEP), and he was receiving dobutamine (5 mcg/kg/min), dopamine (5 mcg/kg/min), and sodium nitroprusside (0.5 mcg/kg/min). The pressure in the right SVC, however, as measured by a small intravenous cannula that had been placed into the right external jugular vein percutaneously before the operation, was 25 mmHg, with a pulsatile pressure waveform. Intra-operative measurement of pressures between the SVCs and their respective pulmonary arteries revealed no gradient across the anastomoses. On temporary occlusion of the pulmonary trunk, the measured pressure fell to 18 mmHg, with the pressure waveform no longer being pulsatile. In view of these findings which indicated significant antegrade flow, we re-established CPB and divided the main pulmonary trunk and sutured both the ends separately with exclusion of pulmonary valve, thus interrupting the antegrade

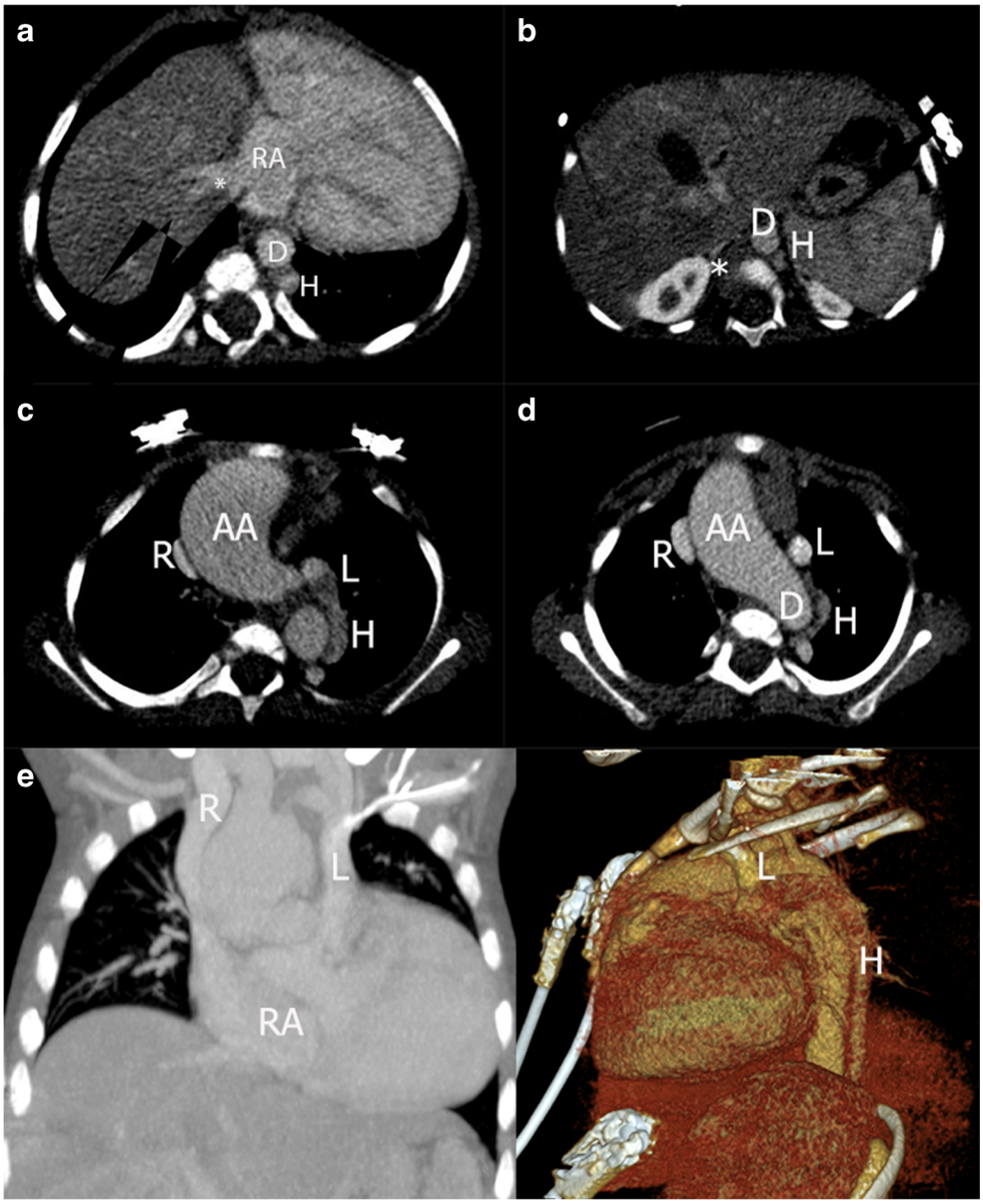

Fig. 1 **a** The axial section shows the morphologically right atrium (RA), the descending thoracic aorta (D), the hemizygous vein (H), with the asterisk showing drainage of the hepatic veins draining to the right atrium. **b** The inferior caval vein is interrupted (asterisk). **c**, **d** The large hemizygous vein (H) drains to the left superior caval vein (L). The right-sided superior caval vein (R) is also seen. (E) Coronal section showing the right superior caval vein (R) draining into the right atrium (RA) and the left superior caval (L) draining to coronary sinus that drains into the left atrium. (E) VRT image showing hemizygous vein (H) joining with the left superior caval (L) and draining to coronary sinus. There is no intercommunicating vein between left and right superior caval veins

flow of blood to the lungs. The patient was then uneventfully weaned off CPB. The total CPB time was 112 min.

After weaning from mechanical ventilation, the systemic saturation was in excess of 90% on room air. Subsequent postoperative recovery was uneventful. At 12 months of follow-up, the saturation is above 90% on room air and an echocardiogram shows normal ventricular function with good SVC flow into both the pulmonary arteries. We plan to perform another CT angiogram to assess the reconstructed LPA. A cardiac catheterization is planned for detailed assessment prior to a future redirection of the hepatic venous return into the pulmonary arteries, that we may be able to achieve transabdominally by a side to side anastomosis of the IVC and the hemizygous vein along with ligation of the suprahepatic IVC.

Discussion

Anomalies of individual systemic venous connection are not uncommon. Abnormal connections often produce abnormal venous drainage, but this is not always the case and therefore, “connection” and “drainage” are not synonymous [1]. The obvious example of this paradox is the diagnosis in our patient. He had persistence of the left SVC, which received the venous return from the abdomen through hemizygous continuation of an interrupted IVC. The left SVC, in turn, connected to the CS, but the sinus then opened into the morphologically LA. The hepatic veins drained into the morphologically RA, along with the right SVC. Despite the plethora of anomalous venous drainage, therefore, the combination cannot be labeled as representing a totally anomalous systemic venous

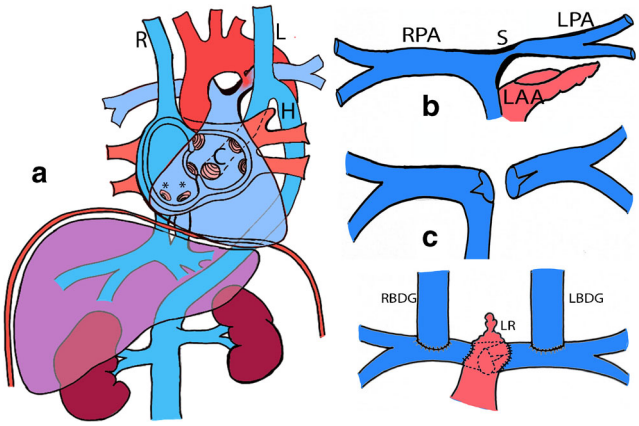

Fig. 2 **a** Drawing of the anatomy. **b** Pulmonary artery anatomy and its relation with left atrial appendage (LAA). The patent arterial duct is not shown. **c** The ductal tissue and the stenosed segment have been excised. (D) The final reconstruction. R: right superior caval vein, L: left superior caval vein, RPA: right pulmonary artery, LPA: left pulmonary artery, RBDG: right Glenn, LBDG: left Glenn, LR: roll of left atrial appendage

connection. The combination of interruption of IVC with bilateral non-communicating SVC, however, may be sometimes encountered in patients with a functionally UVH [2, 6–8]. A series of eight patients has been reported from Saudi Arabia, with all of these patients having hypoplasia of the morphologically right ventricle [2]. Some reports describe anomalies of systemic venous drainage co-existing with DORV (Table 1). Interruption of IVC as part of totally anomalous systemic venous connection has also been described [1].

Juxta ductal pulmonary arterial stenosis is a well-known feature of congenital heart defects that are associated with a decreased pulmonary blood flow. In such patients, reconstruction of the pulmonary arteries has been accomplished by excision of ductal tissue and subsequent direct anastomosis, or by using a patch of pericardium, pulmonary or aortic homograft, or other prosthetic material. In patients where there is a wide gap resulting from excision of the ductal tissue, an interposition roll created out of the autologous pericardium, autologous pulmonary vein conduit, or a flap of the pulmonary trunk [11] has been used to restore the continuity of the pulmonary artery. A patch of the LAA was first used by Barbero-Marcial and colleagues in 1990 [3], with the pedicled LAA subsequently used for reconstruction of the right ventricular outflow in five patients by Aeba and colleagues [4]. Gerelli et al. [12] have described a novel method of reconstruction using LAA called "autologous tissue reconstruction" in patients with VSD and pulmonary atresia. In this method, the LAA was sutured to the distal end of the right ventriculotomy and to the pulmonary confluence that was left in its anatomical position. The entire posterior wall of the right ventricular outflow tract was thus made of autologous vascularized tissue. The LAA, however, has not been used for reconstruction of the LPA in the fashion that we used.

In view of the proximity of the LAA to the LPA, we used a roll prepared from its walls to bridge the gap between the ends of the artery after excision of the ductal tissue. We have used LAA as anterior wall while pericardium formed the posterior wall as in suture less repair. We reasoned that the use of in situ living tissue would retain the potential for growth and hopefully avoid restenosis of the reconstructed LPA.

By using an open technique for the BDG, we were able to avoid the need for cumbersome bicaval cannulation. The free egress of the systemic venous return with the open technique prevents the cerebral congestion that may result after placement of a large-sized cannula, or after intermittent clamping of the SVC [5].

Creation of a fenestrated Kawashima procedure has been reported previously, especially in infants or a young child with reactive pulmonary vasculature [6, 7]. Interrupting the antegrade flow of blood to the lungs avoided the need for any fenestration in our patient. The exclusion of the blood from the hepatic veins is known to promote the development of pulmonary arteriovenous fistulas causing reduction in

Table 1 List of previous publications describing anomalies of systemic venous drainage co-existing with double outlet from the right ventricle

Author	*N*	Age	Morphology	Procedure	Follow-up
Hannan RL, 2003 [9]	1	8 months	DORV	Fenestrated bilateral bidirectional cavopulmonary anastomosis	Uneventful. Only immediate post-operative follow-up described
Picarelli D, 2005 [10]	1	5 months	DORV	Fenestrated bilateral bidirectional cavopulmonary anastomosis	12 months
Sersar SI, 2009 [2]	8	NA	Right heart hypoplasia	Bilateral bidirectional cavopulmonary anastomosis	1 death, rest uneventful. Only immediate post-operative follow-up described
Shahabuddin S, 2009 [6]	2	7 years	Tricuspid atresia, DORV	Bilateral bidirectional cavopulmonary anastomosis	5 years, uneventful in one, only immediate post-operative follow-up described immediate post-operative period

oxygen saturation and has been a subject of an ongoing debate. Nonetheless, in our patient, at his last follow-up visit, there is no reduction in oxygen saturation to suggest the development of pulmonary arteriovenous fistulas. The child is on regular follow-up with a plan of surgical hepatic venous redirection to the pulmonary artery. At that time, a follow-up CT angiogram is also planned to evaluate the efficacy of our method of LPA reconstruction.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Statement of human rights/ethical approval All procedures performed in this study were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. For this study, formal consent was obtained.

Informed consent Informed consent was obtained from all individual participants included in the study.

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