Correction of Anomalous Systemic Venous Drainage in Heterotaxy Syndrome
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A 3-month-old patient with heterotaxy syndrome, complex intracardiac malformations, and severe heart failure underwent surgical correction. Anatomy included an interrupted inferior vena cava with hemiazygous continuation to a persistent left superior vena cava draining to the left atrium. The presence of partial anomalous pulmonary venous drainage precluded an intracardiac baffle. Systemic venous reconstruction was achieved using the left atrial appendage along with an intracardiac repair of the other defects.


Heterotaxy syndrome is frequently associated with complex cardiac malformations that may pose a significant surgical challenge [1, 2]. Surgical techniques for correction have been reported only sporadically [3–5]. We report the surgical management of an infant with polysplenia and cardiac malformations including left atrial isomerism, atrial septal defect, partial anomalous pulmonary venous return, and interrupted inferior vena cava with hemiazygous continuation to a persistent left superior vena cava draining to the left atrium.

The patient’s diagnosis included caudal regression syndrome, bradycardia, heterotaxy, and cardiac malformations including left atrial isomerism, interrupted inferior vena cava with hemiazygous continuation to a persistent left superior vena cava draining to the left atrium, levo-cardia, partial atrioventricular septal defect, mild tunnel subaortic stenosis, supravalvar pulmonary stenosis, partial anomalous pulmonary venous drainage to the right atrium near the superior vena cava, and coarctation of the aorta. Neonatal heart failure prompted extended end-to-end coarctation repair with no complications. Because of refractory heart failure, the patient underwent open surgical repair. Operative findings included massive cardiomegaly with predominantly right ventricular enlargement, small right superior vena cava, persistent left superior vena cava with hemiazygous continuation draining into the left atrium just at the left atrium, left atrial isomerism, left cava entering the left atrium at the base of the left atrial appendage with no coronary sinus communication, right pulmonary veins entering the rightward atrium near the orifice of the right superior vena cava, left-sided pulmonary veins entering near the orifice of the left superior vena cava, incomplete atrioventricular canal with a large primum defect, absent coronary sinus, dysmorphic inlet valves, supravalvar narrowing of the pulmonary artery, and mild tunnel subaortic stenosis (Fig 1).

Due to the anomalous systemic venous return, the intracardiac repair required hypothermia and circulatory arrest at 18°C. A pericardial patch was used to septate the atria, baffling the anomalous right pulmonary veins into the leftward atrium with closure of the atrial septal defect. The location of the pulmonary veins precluded an intracardiac baffle to route the anomalous systemic venous return to the right atrium. To reconstruct the systemic venous drainage, the left superior vena cava was transected, including the very long left atrial appendage, oversewing the cardiac side of the left atrium

Fig 1. External appearance including left atrial isomerism and hemiazygous continuation of interrupted inferior vena cava to persistent left vena cava draining into left atrium.
(Figs 2, 3). Bypass was reestablished and the heart resuscitated. The cut end of the left atrial appendage was closed, making a long tube to act as a neoinnominate vein. This was then brought anterior to the aorta and anastomosed in an end-to-side fashion to the right superior vena cava, diverting all the systemic venous blood to the right atrium. The pulmonary outflow tract was enlarged using a pericardial patch (Fig 4). The postoperative course of the patient was uneventful, with resolution of the heart failure and early extubation. Echocardiograms done 2 and 8 months after the operation showed a widely patent left superior vena cava.

Comment

Heterotaxy syndromes are characterized by the presence of situs ambiguous (with associated polysplenia or asplenia) and frequent cardiac malformations, including atrial isomerism, cardiac malposition, systemic and pulmonary venous anomalies, transposition of great vessels, pulmonary stenosis or atresia, single coronary artery, single ventricle, single atrophicventricular valve, and absent coronary sinus [2]. A persistent left superior vena cava occurs in as many as 30% to 50% of patients with the heterotaxy syndrome [2, 6]. The left superior vena cava may drain to the right atrium via the coronary sinus or, less frequently, to the left atrium. In the latter situation surgical correction is necessary to separate the systemic and pulmonary venous blood. There have been few reports addressing the technical aspects of this surgical repair, which may be done by anastomosing the left superior vena cava to the right atrial appendage or to the left pulmonary artery, or by an intraatrial baffle [7]. In this case, the combination of anomalous right pulmonary veins, atrial septal defect, interrupted inferior vena cava with hemiazygous continuation to a persistent left superior vena cava, and absent coronary sinus presented a challenging anatomic situation. Because of the interrupted inferior vena cava with hemiazygous continuation, unobstructed repair of systemic venous return was critical. An intraatrial baffle was not possible due to partial anomalous return of the right pulmonary veins, which could not technically be routed at the same time to the left side. A left superior cavopulmonary anastomosis was not created because of concerns of potential pulmonary hypertension in the setting of
subaortic narrowing and possible left atrioventricular valve regurgitation. Atrial septation and pulmonary venous reconstruction was accomplished using a pericardial patch. As the length of the left superior vena cava was insufficient for direct right atrial anastomosis, the appendage was used to create a conduit. The left atrial tube was then anastomosed to the superior vena cava. This appeared to offer the most tension-free repair. Due to the marked right ventricular enlargement, direct anastomosis to the right atrial appendage appeared less favorable. Conduit patency was confirmed by echocardiography 2 weeks, 2 months, and 8 months after the operation. Although this condition is rare, this technique may be of use in similar cases requiring complex venous reconstruction.

References

INVITED COMMENTARY
This patient described by Drs Palacios-Macedo and Fraser presented with a complex cardiac malformation including a persistent left superior vena cava (LSVC) with hemiazygous continuation of an interrupted inferior vena cava. The transfer of the LSVC in continuity with a long left atrial appendage as a tube to the right superior vena cava has not been reported previously. A postoperative echocardiogram at 8 months shows the anastomosis to be patent. The question posed to Drs Palacio-Macedo and Fraser is whether there was need for an extracardiac conduit or whether an intracardiac tunnel could be performed in this case. They suggested that the configuration of the pulmonary veins precluded the use of an intraatrial baffle. Shumacker and associates [1] in 1967 reported a similar technique using part of the left atrium as a flap. This tubular extension was implanted into the right atrium. Sherafat and colleagues [2] in 1970 and Kabbani and co-workers [3] in 1973 both reported that these grafts were thrombosed. They both cited the risk of thrombosis and compression between the aorta and the sternum in an extended extracardiac conduit. Gontijo and associates [4] reported the use of a polytetrafluoroethylene tubular graft that extended across to the right atrium in a 14-year-old patient. The use of nonviable material in an infant or growing child would not be advisable. Furthermore, one would seek to use a ringed conduit in the older patient to prevent compression.

An intracardiac repair would be my method of choice for rerouting the anomalous LSVC. This can be performed with the use of pericardium, Dacron, polytetrafluoroethylene, or the child’s own tissue. Because pericardium can degenerate, become fibrotic, or calcify, I have used polytetrafluoroethylene since 1984. The intracardial reroutings could be difficult if one used the methods described by Rastelli and associates [5] and Kirklin and Barratt-Boyes [6] in which the tunnel is created on the posterior wall of the left atrium or in which the pulmonary veins drain the tricuspid valve. Doctors Palacios-Macedo and Fraser stated that the intraatrial baffle could not be used because of the configuration of the pulmonary veins. Two previously reported methods may have been possible. Komai and colleagues [7] reported inverting the left appendage and swinging the edge of the appendage flap to the posterior wall of the left atrium around a cannula, forming a tunnel between the LSVC and the right atrium. The remaining atrial septal defect was closed with pericardium. Another intraatrial method described by Sand and associates [8] uses a Dacron patch to construct a baffle on the roof of the left atrium. The baffle begins at the base of the LSVC and left atrial appendage junction, extending across the right atrium superior and anterior to the pulmonary veins. An atrial septal defect is created if not present, and the atrial septum is reconstructed at the lip of the intraatrial tunnel. The major part of the tunnel is the child’s own tissue. These methods would avoid the potential obstruction of the pulmonary veins and the mitral valve. They also avoid the area of the conduction system.

The extended extracardiac tube proposed by Palacios-Macedo and Fraser presents a novel and interesting approach to the rerouting of the LSVC to the right side. I am concerned about the potential for thrombosis or compression of this channel and await a longer follow-up report on this child.

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