Partial anomalous pulmonary venous drainage

A novel approach to repair

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■ Isolated partial anomalous pulmonary venous drainage with an intact atrial septum is a rare finding. The authors describe their experience with three patients (ages 9, 37, and 54 years), with partial anomalous pulmonary venous connection to the superior vena cava, right atrium, and inferior vena cava, who underwent extracardiac conduit repair of this anomaly. In all three patients, a synthetic Gortex graft was used for reconstruction of the venous pathways to the left atrium. The follow-up period ranged from 10 to 82 months (mean, 42 months). All three patients were evaluated with intravenous digital angiography, transesophageal echocardiography, or both at 10, 33, and 82 months postoperatively. Patency of the grafts with no evidence of obstruction and excellent pulmonary venous flow was shown. This surgical technique is an excellent option for correction of this anomaly, and intravenous digital subtraction angiography is a useful diagnostic tool during the postoperative period to evaluate patency of the repair.

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ISOLATED PARTIAL anomalous pulmonary venous drainage with an intact atrial septum is a rare finding.[1,2] We briefly discuss this anomaly and describe our experience with three patients, with partial anomalous pulmonary venous connection to the superior vena cava, right atrium, and inferior vena cava, who underwent extra-cardiac conduit repair. A Gortex graft was used for reconstruction of venous pathways to the left atrium. To our knowledge, this is the first time this anomaly has been repaired in this manner and the long-term follow-up reported.

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CASE REPORTS

Case 1

A 36-year-old woman presented with recent onset of hemoptysis and dyspnea on moderate exertion. She had been well her entire life and had had two uncomplicated pregnancies and deliveries.

On physical examination, the second heart sound was widely split and fixed. A grade II/VI systolic murmur was noted along the left sternal border. The chest radiograph demonstrated a classic scimitar sign (*Figure 1*). Intravenous digital angiography (IV DSA) demonstrated anomalous pulmonary venous drainage of the right middle and lower pulmonary veins into the inferior vena cava (*Figure 2*). This finding was confirmed by cardiac catheterization. (For catheterization data, see *Table 1*.) The atrial septum was intact.

The patient underwent surgical repair with ligation of the anomalous pulmonary vein and the interposition of

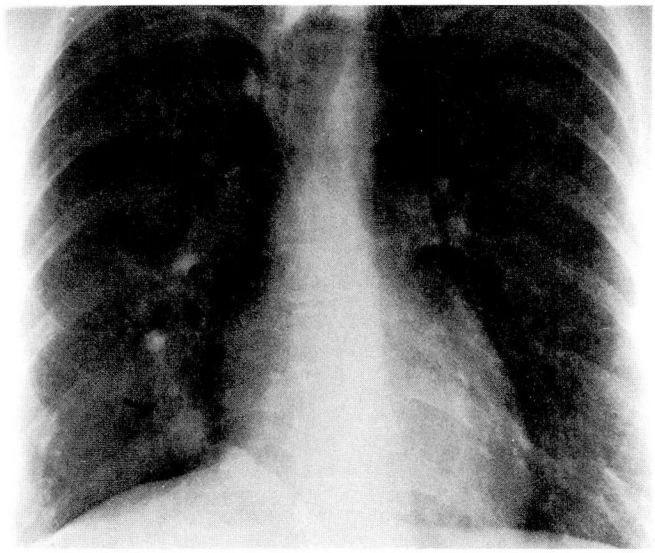

FIGURE 1. Case 1. Preoperative chest radiograph demonstrates the scimitar sign.

TABLE 1
CATHETERIZATION DATA

	Case 1	Case 2	Case 3
Cardiac output	7.0 L/min	3.2 L/min	2.4 L/min
Cardiac index	4.4 L/m/m^2	1.8 L/m/m^2	2.3 L/m/m^2
Pulmonary flow	–	5.4 L/m	5.2 L/m
Pulmonary index	–	3.0 L/m/m^2	5.0 L/m/m^2
QP:QS ratio	1.5:1	1.7:1	2.2:1
Systemic vascular resistance	14 U	43.9 U	39 U
Pulmonary vascular resistance	0.56 U	5.3 U	6 U
Left-to-right shunt	33%	41%	53%

an 18-mm Gortex graft between the anomalous vein and left atrium. Her postoperative course was unremarkable. IV DSA performed on the first postoperative day demonstrated patency of the graft (*Figure 3*). Attempts to repeat the study 16 months later failed due to poor venous access. The patient remained well 33 months postoperatively. Transesophageal echocardiography done at that time again demonstrated patency of the graft (*Figure 4*).

Case 2

A 54-year-old woman presented with palpitations, atypical chest pain, and dyspnea of 18 months duration. She had a five-year history of asthma. She also had undergone left partial pneumonectomy at the age of 16 for tuberculosis.

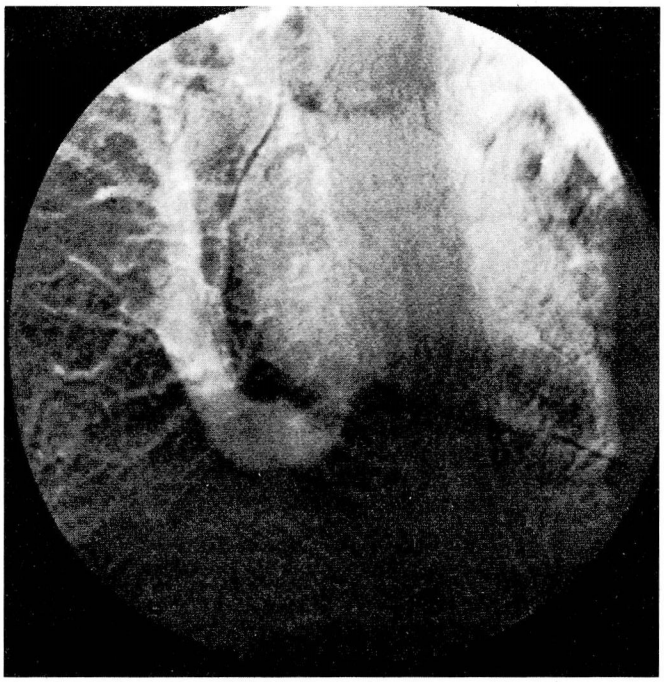

FIGURE 2. Case 1. Preoperative IV DSA shows partial anomalous venous return to the inferior vena cava.

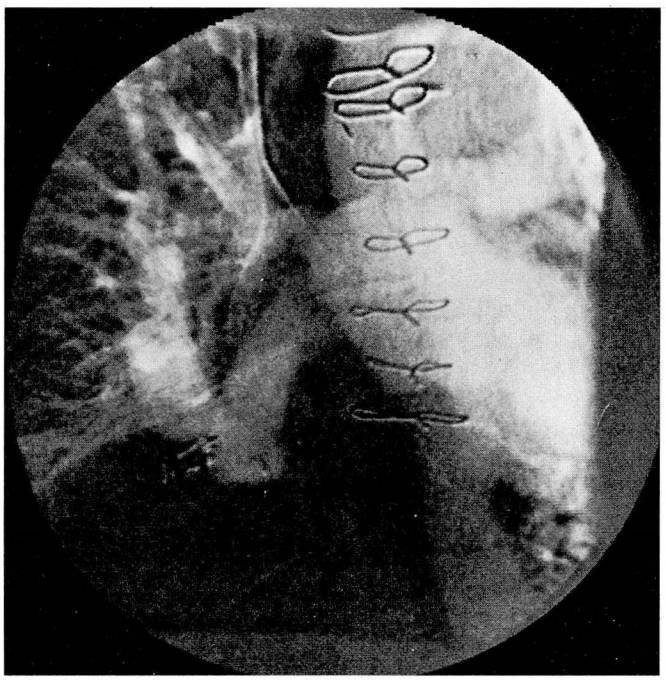

FIGURE 3. Case 1. Postoperative IV DSA demonstrates surgical correction of the anomalous pulmonary vein and the widely patent Gortex graft.

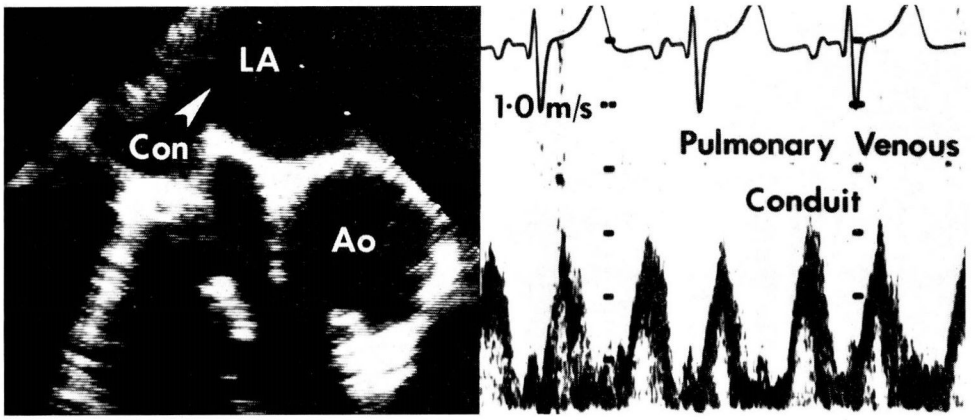

FIGURE 4. Case 1. Postoperative transesophageal echocardiography shows patency of the conduit repair with a normal pulmonary venous flow.

On physical examination, the second heart sound was normal with normal splitting. A grade II/VI systolic murmur was auscultated at the second right intercostal space with radiation to the neck and upper left chest. The electrocardiogram showed right bundle branch block. The chest radiograph showed changes related to the left pneumonectomy. The echocardiogram revealed right atrial and right ventricular enlargement. Cardiac catheterization demonstrated drainage of an anomalous right upper pulmonary vein to the superior vena cava. The atrial septum was intact.

At surgery, a Gortex graft was inserted between the right superior pulmonary veins and the left atrium, and the anomalous connection was ligated. Postoperative recovery was unremarkable.

Ten months later, IV DSA demonstrated patency of the graft. The patient continues to be categorized as New York Heart Association Functional Class I.

Case 3

A 9-year-old girl was admitted for evaluation of recent onset of dyspnea and fatigue with exercise. She had been hospitalized at five months, eight months, and 13 months of age for pneumonia and bronchitis. A heart murmur was noted at that time, and cardiac catheterization revealed right partial anomalous pulmonary venous drainage.

On physical examination, she was acyanotic. The second heart sound was split normally. There was a II/VI systolic murmur at the right upper sternal border and a II/VI longer-blowing systolic murmur at the left lower sternal border. The chest radiograph showed that the heart was shifted to the right. Right atrial enlargement with increased pulmonary vascular markings was suggested. There were several rib and hemivertebral anomalies. The echocardiogram revealed right ventricular hypertrophy, right atrial enlargement, and paradoxical septal motion. Cardiac catheterization demonstrated anomalous pulmonary venous drainage from the right lung to the right atrium. The right ventricle was dilated and moderately dysfunctional. The atrial septum was intact.

The patient underwent ligation of the anomalous vein and insertion of a Gortex graft from the pulmonary vein to the left atrium. The postoperative course was unremarkable. Cardiac catheterization in the immediate postoperative period demonstrated patency of the graft and no residual left-to-right shunt.

The patient resumed a normal lifestyle. The most recent follow-up was at 82 months postoperatively. Graft patency was again demonstrated at that time by IV DSA (*Figure 5*).

DISCUSSION

The exact incidence of partial anomalous pulmonary venous drainage has not been well established in clinical studies. It has been estimated by Hughes and Rumore[3] that partial pulmonary venous drainage occurs in about 0.7% of routine autopsies.[3,4] In the complicated form of the anomaly, a variety of associated intracardiac defects have been described most commonly as an atrial septal defect.[1]

The isolated form of the anomaly with an intact atrial septum is less common.[1,2] The pulmonary veins affected are mainly those from the right lung, and the site of termination varies but is most commonly the superior vena cava.[5]

Clinical symptoms in patients with isolated partial anomalous pulmonary venous drainage are governed by the degree of left-to-right shunt and the resultant volume overload of the right ventricle. Physiologically, the anomaly resembles a left-to-right shunt at the atrial level, and the clinical picture usually resembles an atrial septal defect. When symptoms of fatigue, dyspnea, and finally, congestive heart failure develop, surgical correction is the definitive therapy.

Various surgical techniques have been described. Most often, the anomalous pulmonary veins are redirected through a surgically created atrial septal defect to the left atrium via an intra-atrial baffle.[5,6] In our patients, however, we interposed a Gortex graft between the anomalous pulmonary vein and the left atrium. The procedure was carried out on cardiopulmonary bypass. The anomalous venous connection to the system venous circulation was interrupted by simple ligation. In all three patients, long-term patency of the grafts was confirmed by IV DSA or transesophageal echocardiography or both. All three patients resumed a normal lifestyle postoperatively and are currently classified as New York Heart Association Functional Class I.

The concept of using synthetic conduits in venous pathways has probably occurred to most thoracic surgeons. The main concern, however, is thrombosis and venous obstruction. None of our patients was given maintenance anticoagulant treatment. In spite of our concern that there might be a problem with the Gortex grafts, all grafts appear to be functioning well in the short and intermediate term with no evidence of obstruction. The long-term fate of these grafts is still to be determined. Until further studies are carried out, we believe that this surgical technique should not necessarily be the standard or preferred approach in the routine case. However, it certainly should be part of the cardiac surgeon's armamentarium when treating this disorder, particularly in the occasional case where the anatomical relationships make it technically difficult and sometimes impossible to use the standard methods.

Tsuchida et al[7] have recently described a case of scimitar syndrome, which was successfully repaired by incorporating a synthetic conduit. However, in their case, an intra-atrial approach was used. A 14-mm woven Dacron graft was used for the construction of a pathway to the left atrium through an enlarged atrial septal defect.

The use of IV DSA to evaluate pulmonary venous anomalies has been previously described.[8] We have already detailed its use in the evaluation of systemic and pulmonary venous anomalies, including the scimitar syndrome.[9] This paper demonstrates its application in the postoperative follow-up of patients with isolated partial anomalous pulmonary venous drainage who underwent extracardiac conduit repair. All three patients described have been studied postoperatively using IV DSA, which clearly demonstrated patency of the grafts (*Figures 3* and 5).

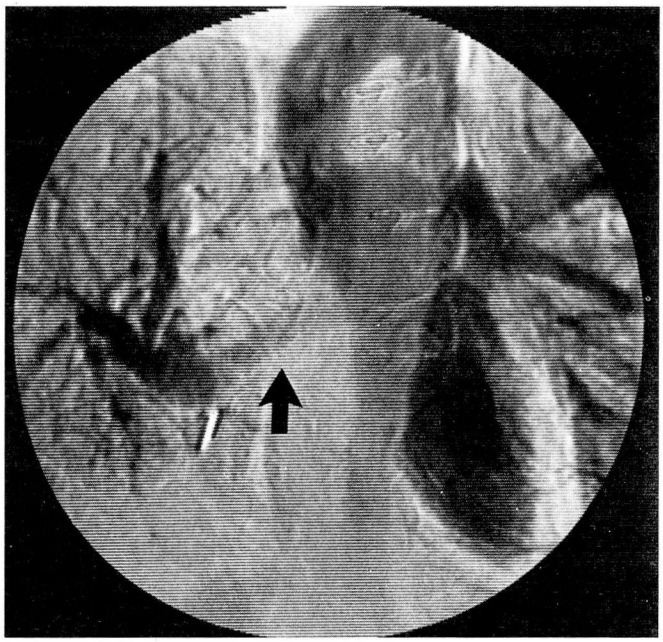

FIGURE 5. Case 3. IV DSA (82 months after operation) demonstrates a patent conduit draining to the left atrium (arrow).

CONCLUSION

Based on the excellent results seen in our patients who have undergone conduit repair of partial anomalous pulmonary venous return, we believe that this technique is an excellent option for the correction of this anomaly. Also, IV DSA is a useful diagnostic tool in the postoperative period to evaluate patency of the repair.

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Commentary

ELIOT Rosenkranz, MD, Department of Thoracic and Cardiovascular Surgery, The Cleveland Clinic Foundation, comments: The method of repair employed for isolated partial anomalous pulmonary venous return is dictated by the anatomic type of anomalous return encountered. The most common is isolated drainage of a single pulmonary vein, usually the right upper pulmonary vein, to the superior vena cava. This is typically repaired by constructing a pericardial baffle between the orifice of the anomalous vein and the usually coexisting sinus venous atrial septal defect. Rarely, the atrial septal defect is small or nonexistent. In that situation, the atrial septal defect is enlarged or created by excising the secundum septum, and the baffle is then constructed as described.

The right pulmonary vein can also drain directly to the body of the right atrium. Repair of this defect is identical to that employed for anomalous drainage to the superior vena cava.

A third type of partial anomalous drainage, to the inferior vena cava, is frequently called scimitar syndrome. A pericardial baffle is constructed within the inferior vena cava and right atrium, directing the anomalous venous drainage across the atrial septum to the left atrium.

A right- or left-sided anomalously draining pulmonary vein may enter the coronary sinus. This is repaired by incising the atrial septum between the os of the coronary sinus and the secundum septum defect and constructing a pericardial baffle directing the coronary sinus drainage to the left atrium.

In the case of the rare type of partial anomalous drainage to the left superior vena cava or the innominate vein, the anomalous pulmonary vein is detached from the systemic vein and reimplanted in the left atrial appendage or body of the left atrium itself.

The method of repair described by Hanhan et al is a novel technique that uses an extracardiac conduit to drain pulmonary venous blood to the left atrium. As the authors point out, this method may be especially useful when access to the draining vein is difficult. Their concerns about the risks of conduit thrombosis or stenosis secondary to pseudointimal hyperplasia must be taken into consideration when a more standard method of repair might be safely employed.