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Brief Report

The impact, and surgical implications, of isolated anomalous connection of one pulmonary vein

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Abstract Partially anomalous pulmonary venous connection of a solitary pulmonary vein in the setting of an intact atrial septum is often subclinical, and the indications for surgical repair are controversial. Here we describe a patient who developed a significant shunt over a period of 10-years. Flow through the anomalously connected pulmonary segment depends on the difference of pressure between the right and left atrium, and may increase with age.

Keywords: Partially anomalous pulmonary venous connection; intact atrial septum; pulmonary hypertension

In the current era, the indications for surgical correction of partially anomalous pulmonary venous connection of a solitary vein, in the setting of an intact atrial septum, are far from clear. We describe our own recent experience with a solitary patient, which shows that the surgical indications may change with time, and are affected by several complicating factors.

Case report

A woman with partially anomalous pulmonary venous connection was referred to the National Cardiovascular Center at the age of 44 years with a one-month history of intermittent ankle oedema. The atrial septum was intact, and there was no other interatrial communication. The right upper pulmonary vein was connected to the proximal part of the superior caval vein, with the other pulmonary veins joining in normal fashion to the left atrium. She had long-standing systemic hypertension. The electrocardiogram at that time demonstrated atrial fibrillation. Elective catheterization showed mild pulmonary hypertension and a raised pulmonary arterial wedge pressure (Table 1). Because of the relatively small ratio of pulmonary to systemic flows, there was a consensus at our medico-surgical meeting that surgery should not be recommended. Her symptoms resolved with a small dose of diuretic agents.

At the age of 54 years, she developed congestive heart failure. The chest radiogram then showed moderate cardiomegaly with pulmonary congestion (Fig. 1). Cardiac catheterization this time showed an increased left-to-right shunt, together with moderate pulmonary hypertension (Table 1). The consensus was now for surgical repair of the anomalous pulmonary vein.

Under standard cardiopulmonary bypass and cardioplegia, the intact atrial septum was incised and a sufficiently large interatrial communication was created at the oval fossa, incorporating also the superior interatrial fold. The right upper pulmonary vein was redirected to the left atrium through the surgically created hole using a baffle of 0.1 millimetre-thick expanded polytetrafluoroethylene. As is often the case with this particular malformation, the dilated proximal superior caval vein accommodated the channel quite nicely, without producing any obstruction to the systemic venous pathway.

The patient had a smooth postoperative course throughout. Postoperative catheterization, carried

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Table 1. Summary of catheterization data.

	Preoperative		Postoperative
Age	44 y	54 y	54 y
RAP	(11)	(18)	(9)
PAP	47/16(31)	54/29(38)	38/16(23)
PCWP	(15)	(24)	(15)
Qp/Qs	1.19	1.90	–
CI	5.33	3.51	3.01
LVEDVI	96.1	–	90
EF	56	–	57

RAP = right atrial pressure [mmHg]; PAP = pulmonary arterial pressure[mmHg]; PCWP = pulmonary capillary wedge pressure[mmHg]; y = years; Qp/Qs = pulmonary/systemic flow ratio; CI = cardiac index [l/min/m^2]; LVEDVI = left ventricular end-diastolic volume index [ml/m^2]; EF = ejection fraction [%].

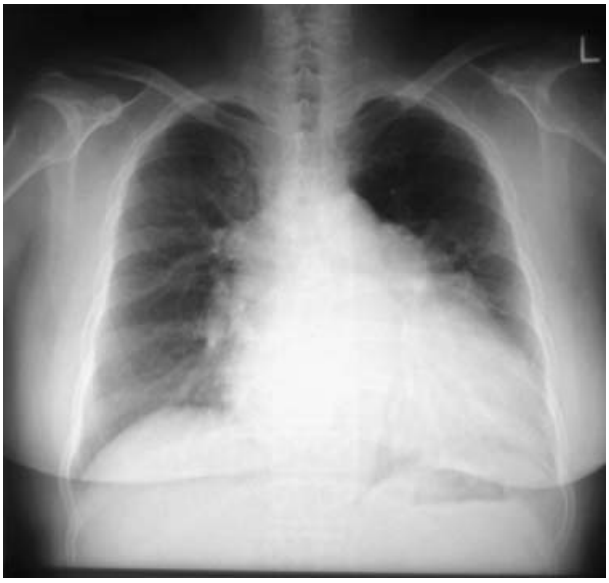

Figure 1.
Chest radiogram at the age of 54 years before surgery. There is moderate cardiomegaly and pulmonary hypercirculation particularly in the right lung.

out 2 weeks after the surgery, showed improvement in the pulmonary hypertension (Table 1). The heart rate was around 70 beats per minute, but still in atrial fibrillation. She is currently doing well 3 years after the surgery, receiving anticoagulation for the persistent atrial fibrillation.

Discussion

It is well recognized that isolated partially anomalous pulmonary venous connection of one whole lung, or in the setting of the scimitar syndrome, should be corrected surgically.[1,2] The need for surgical correction is less clear when only a small part of one lung has anomalous venous return in the setting of an intact atrial septum, since such a situation tends to appear clinically unimportant. According to authoritative sources, life expectancy may be normal, and surgery is unlikely to be indicated where the ratio of pulmonary to systemic flows in this setting is less than 1.8.[1,2]

The distribution of flow to the right upper lobe is stated as approximately one-fifth in the normal lung.[3] Provided that the distribution remains comparable to the normal in the abnormally draining right upper lobe, the ratio of pulmonary to systemic flows is theoretically estimated to be 1.23 if the right upper lobe only drains anomalously. The initially measured ratio of flows in our patient was in keeping with this estimation. There are some reports, however, that call attention to the development of either significant shunting or pulmonary vascular obstructive disease in this setting.[4–7]

It is also known that, in patients with atrial septal defects, the ratio of pulmonary to systemic flows increases with age, because systemic hypertension progressively makes the left ventricle less compliant. In isolated partially anomalous pulmonary venous connection, nonetheless, this is not the case, since an interatrial communication is lacking. An alternative mechanism of change in the ratio of pulmonary to systemic flows is the different atrial pressures by which the individual pulmonary venous drainage is confronted. The pulmonary arterial pressure is identical in each lung segment. The normal lung segments drain into the left atrium, whereas the anomalously draining ones connect to the right atrium. Hence, the ratio of pulmonary to systemic flows depends on the difference of pressure between the two atrial chambers.[7,8] In reality, there is a report of isolated partially anomalous pulmonary venous connection of one pulmonary vein in combination with mitral valvar stenosis developing a significant left-to-right shunt.[9] In our current patient, the difference between the right atrial pressure and the pulmonary capillary wedge pressure became greater, from 4 to 6 millimetres of mercury, over a period of 10 years.

The pulmonary hypertension seen in our patient is less likely to be related to the mild increase in flow of blood to the lungs. It is generally accepted that pulmonary hypertension does not progress in patients with isolated partially anomalous pulmonary venous connection of only a small part of one lung in terms of the circumstance of the ratio of pulmonary to systemic flows.[1,2] Instead, it is likely that the long-standing systemic hypertension, and the raised end diastolic pressure of the left ventricle, will have been the major factor. Persistent atrial

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fibrillation had also adversely affected the atrial pressures.

When to intervene in this situation remains unclear. There are no "cast iron" indications for surgery. When a prophylactic aspect of repair is taken into account, then consideration should be given to operating at a much earlier time. In this respect, we do not disagree with conventional wisdom that isolated partially anomalous pulmonary venous connection should be left uncorrected during infancy and childhood. Because of the compliant ventricles, and usually insignificant differences in pressure between the right and the left atriums, the left-to-right shunt dose not produce a haemodynamically crucial impact. Also, repair of one pulmonary vein in small children is far from a simple procedure. It should be technically feasible, but no precise prognosis has yet been documented in the longer term after surgical repair during early childhood in this particular setting. The situation differs from repair of totally anomalous pulmonary venous connection, in which all pulmonary venous blood flows across the anastomosis. With partially anomalous connection of a solitary vein, the reconstructed channel accommodates a quarter amount of blood, and might be liable to occlusion. We do not, therefore, recommend aggressive early repair. Nonetheless, taking into account our experience with the current patient, we need to warn parents of children with this minor pulmonary venous abnormality that their situation should not be regarded as entirely innocent malformation.

In the past, some patients with interatrial communications of superior sinus venosus type with associated anomalous connection of the right upper pulmonary vein underwent simple closure of the defect, leaving the connections of the anomalous branches to the superior caval vein. Furthermore, in some patients with complicated mixed type of totally anomalous pulmonary venous connection, a single anomalous pulmonary vein may be left uncorrected.[10] The postoperative situation of these patients is the same as isolated partially anomalous pulmonary venous connection. Long after the initial procedure, such patients can suffer problems similar to those encountered in our patient, in other words an increasing left-to-right shunt, atrial arrhythmias, and pulmonary hypertension. Our current experience, therefore, provides us with a good opportunity to reconsider our surgical strategy for isolated partially anomalous pulmonary venous connection, as well as "palliatively" repaired partially anomalous pulmonary venous connection with atrial septal defect or mixed type of total anomalous pulmonary venous connection.

If a similar patient were referred today because of clinical symptoms, we would arrange repair of the anomalous pulmonary venous drainage soon, irrespective of the ratio of pulmonary to systemic flows, and employ anti-arrhythmic manoeuvres such as the maze procedure depending on the duration of atrial fibrillation. When neither systemic hypertension nor arrhythmia is present, the magnitude of shunting may not increase with time. Even so, we would maintain close observation to avoid unexpected deterioration of the overall situation.

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