Case report - Cardiac general
An unusual manifestation of left partial anomalous pulmonary venous connection

Kalyana Javangula*, James Cole, Michael Cross, Philip H. Kay
Department of Cardiac Surgery, Leeds General Infirmary, E Floor, Jubilee Wing, Great George Street, Leeds LS1 3EX, UK

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Abstract
A 68-year-old male patient had aortic valve replacement for aortic valve endocarditis. The central line (left) position looked abnormal on chest X-ray. Contrast studies confirmed left sided partial anomalous pulmonary venous connection.

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Keywords: Anomalous pulmonary vein; Aortic valve

1. Introduction

A 68-year-old patient was operated for aortic valve endocarditis. The aortic valve replacement was done using standard cardiopulmonary bypass with aortic and right atrial cannulation. Myocardial protection strategy included antegrade cold blood cardioplegia into the aortic root and a second dose by direct coronary ostial route. The aortic valve was excised and a 21 mm mitroflow pericardial bioprosthetic valve was implanted. The only surprise finding was constant air in the venous line causing an air lock. All the usual sites for air entry like the atrial cannulation site, venous line connections and patent foramen ovale (PFO) were checked and ruled out. The causative organism was Streptococcus viridans and intravenous vancomycin was commenced preoperatively and continued postoperatively. The central venous line which was initially on the right side, was changed to left on the 10th postoperative day as per units protocol. The X-ray done after the new line insertion showed the central line going towards the aortic arch rather than towards left innominate [superior vena cava (SVC)] direction. The first suspicion was whether the line was in the carotid artery. Gas analysis of an arterial blood sample confirmed PO₂ of 15 kPa, PCO₂ of 4 kPa. The central line is connected to a transducer to monitor the trace which was surprisingly a venous trace. A bed side ultrasound examination confirmed the central line in internal jugular vein, but it could not be traced beyond the root of neck. During this time the patient was clinically stable apart from developing atrial fibrillation. He subsequently regained sinus rhythm following administration of amiodarone.

The patient was taken for a computed tomography (CT)-scan (Fig. 1) which confirmed the catheter in the internal jugular vein in the neck, but instead of going into the left innominate vein it was going towards the pulmonary hilum. There was no contact with either the carotid artery or the aortic arch. Contrast was given through the catheter which revealed a partial anomalous venous connection (Fig. 2). This anomalous vein drains the lingular segment into the left innominate vein. Following this, the catheter was repositioned in the left innominate vein. The patient made an uneventful recovery and was discharged home after the full course of antibiotics. He attended the follow-up clinic and was completely asymptomatic. Clinical examination

*Corresponding author. Tel.: +44 7747128401; fax: +44 1133928092. E-mail address: kalyanachakravarthi@hotmail.com (K. Javangula).
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Fig. 1. CT-reconstruction showing the left side central venous line going into left anomalous pulmonary vein communicating with left innominate vein.
never revealed any signs or symptoms of the volume overload or heart failure. Follow-up echocardiogram showed good prosthetic valve function and normal right and left ventricular function and normal estimated pulmonary artery pressures. As the partial anomalous pulmonary venous connection (PAPVC) was not impairing the patients haemodynamics, we decided not to intervene further. The patient has been kept under regular echocardiographic follow-up.

2. Discussion

PAPVC is a congenital anomaly present in 0.4–0.7% in postmortem examinations [1]. This is more common on the right side with only 10% involving left side veins. While most patient with PAPVC are asymptomatic, the natural history dictates that if significant left to right shunt exists, patients may develop irreversible pulmonary hypertension, irreversible pulmonary obstructive disease or right heart failure.

Our case is unique for several reasons. First, the left PAPVC was never recognized during preoperative investigations. Second, the on-table observation of constant air in the venous line of the bypass circuit (two stage venous cannula in the right atrium). Despite checking for all possible causes such as atrial tear, loose connections, septal defects or PFO [with trans-oesophageal echo (TOE)] we could not explain the source of the air. In retrospect, the left PAPVC might explain the air in the venous line. Third, the left side PAPVC came to light after positioning the central line which went down the anomalous connection. The initial blood gas analysis showed arterial PO₂ and PCO₂ but the pressure monitoring showed a venous trace which was also very interesting in this case. The imaging studies with contrast down the catheter confirmed the PAPVC. In our patient, it was only the lingual segment which drained into the left innominate vein. There are only two series of left side PAPVC reported in the literature, Van Meter et al. [2] and Dearani et al. [3].

The two schools of thought about managing left side PAPVC are, first early repair before the patient develops symptoms of right heart volume overload and pulmonary hypertension. Second, where surgical correction is indicated only if the patient is symptomatic because the correction of left PAPVC is not without its complications such as atrial fibrillation, complete heart block, cardiac arrest and pulmonary venous obstruction [4, 5]. The Mayo group (Elbardissi et al.) showed that intraoperative echocardiograms as well as the measurement of anastomotic gradients with monitoring lines (more than 4 mmHg is significant) makes these corrections effective with good outcomes. They also showed that the correction can be done both on-pump as well as off-pump.

In conclusion, the left side PAPVC is a significant anomaly with serious consequences depending on the amount of volume load on the right heart. It can be corrected safely provided proper imaging studies are done preoperatively and adequate precautions are taken during surgery.

References


eComment: Left-sided partial anomalous pulmonary venous connection – should diagnosis lead to surgery?

Author: Frank Edwin, Walter Sisulu Pediatric Cardiac Center, Sunninghill Hospital, Johannesburg, South Africa
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Left-sided partial anomalous pulmonary venous connection (PAPVC) occurs 10 times less frequently than right-sided PAPVC, in isolation both have similar hemodynamics. Left PAPVC affects the upper lobe drainage twice as commonly as it affects the entire drainage of the left lung. Commonly, the anomalous vein drains via a vertical vein into the innominate vein. Rare patterns of drainage include drainage into the left subclavian vein, persistent left superior vena cava, coronary sinus, or directly into the right atrium. Isolated PAPVC is often asymptomatic and tends to go unnoticed until adulthood as encountered in the current report [1], or during investigation of another illness. This author recently encountered a case of left-sided PAPVC in a two-year-old discovered on investigation of a suspected vascular ring.

Left untreated, long-standing PAPVC predisposes the patient to right-sided volume overload, tricuspid regurgitation (TR), arrhythmias, pulmonary hypertension, irreversible pulmonary vascular disease, right ventricular dysfunction and right ventricular failure.

The development of symptoms and complications from isolated PAPVC depends on the shunt fraction and thus on the number of pulmonary veins
anomalously draining to the right heart. A single anomalous vein is not usually hemodynamically significant in the short-term unless the hemodynamic effect is exaggerated by coexistent left-to-right shunts like atrial septal defects (ASDs). The hemodynamic effect of an ASD may be out of proportion to its size when PAPVC coexists. PAPVC occurs in approximately 10–15% of patients with secundum ASDs and in 85% of patients with sinus venosus ASDs [2]. It is imperative therefore to rule out a coexistent PAPVC prior to percutaneous ASD closure.

The occurrence of symptoms such as fatigue, dyspnea, effort intolerance, palpitations, and chest pain indicates significant right heart overload and chronicity. In earlier times, catheter-based angiography was the imaging modality of choice; this has been superseded by echocardiography, computerized tomographic angiography, and magnetic resonance imaging.

Javangula et al. [1] mention the lack of consensus regarding indications for surgery. Generally, indications for surgery have paralleled those for isolated ASDs. Sachweh et al. [3] found 27% and 17% of adult patients (44±17 years) with secundum ASDs had moderate (32–50 mmHg) and severe (>50 mmHg) systolic pulmonary hypertension, respectively; it was not possible to predict from the study which patients would develop hypertensive pulmonary vascular disease (HPVD). The hemodynamic similarity between ASD and PAPVC indicates the potential for HPVD in the latter also. The diagnosis of PAPVC may therefore be considered an indication for surgery on the basis of this potential for HPVD in adulthood [3, 4].

In the current era, symptomatic PAPVC and asymptomatic patients with Qp:Qs >1.5, right ventricular dilation, mild-to-moderate TR, or early stages of HPVD require surgical treatment in order to prevent the development and progression of right ventricular failure and irreversible pulmonary vascular disease [5]. Asymptomatic patients without evidence of right ventricular dilation or TR may not be subjected to surgery on the basis of these recommendations.

References