Review Article

Management of partial anomalous pulmonary venous connections in patients requiring pulmonary resection: a case report and systematic review

Kieran Singhal[1,2], Andrew D. Newton[1,2], Christopher Corbett[1,2], Jarrod D. Predina[3]

[1]Center for Precision Surgery, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA; [2]Division of Thoracic Surgery, Department of Surgery, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, PA, USA; [3]Department of Surgery, Massachusetts General Hospital, Boston, MA, USA

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Correspondence to: Jarrod D. Predina, MD, MTR. 55 Fruit Street; Boston, MA 02108, USA. Email: jpredina@partners.org.

Abstract: Partial anomalous pulmonary venous connections (PAPVCs) are rare congenital anomalies that are frequently asymptomatic in adults. When PAPVCs are encountered in the patient requiring pulmonary resection, improper management can result in fulminant right-heart failure and death. In this report, we note our management of a 70-year-old male who presented with a right upper lobe ground-glass opacity (GGO) and a PAPVC. We also provide a systematic review of all contemporary reports and provide an algorithm for PAPVC management in the adult patient requiring pulmonary resection.

Keywords: Partial anomalous pulmonary venous connection (PAPVC); lung cancer; pulmonary resection

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Background

Partial anomalous pulmonary venous connections (PAPVCs) are a heterogeneous group of rare vascular malformations that describe incomplete connections from the pulmonary venous system to the systemic venous system or right heart. PAPVCs are encountered in approximately 0.5% of the pediatric population, where they are commonly associated with atrial septal defects (ASDs) (1). Within this cohort, treatment often requires operative repair (1). In the adult population, PAPVCs are exceedingly rare, thus disease-specific epidemiology and treatment protocols are undefined. Management of PAPVCs becomes a therapeutic dilemma when circulatory physiology may be rapidly altered, for example, during pulmonary resection. In this report, we highlight physiologic considerations of PAPVCs in the adult population by documenting our experiences involving a patient who presented with a suspicious pulmonary nodule and PAPVC. In addition to these comments, we provide a systematic review consisting of all contemporary reports describing management of PAPVCs in the patient undergoing resection for suspicious pulmonary lesions.

Case report

A 70-year-old male former smoker presented to our clinic for evaluation of an incidentally discovered 1.9 cm ground-glass opacity (GGO) of the right upper lobe (*Figure 1A*). Upon interview, the patient denied associated symptomatology. A preoperative PET-CT was obtained and revealed a standardized uptake value (SUV) of 4.9 (*Figure 1B*), and no suspicious lymph node involvement. The lesion was not amenable to transthoracic or endobronchial biopsy, thus the patient was consented for a video-assisted thoracic surgery (VATS) wedge biopsy. During initial dissection, an anomalous vein arising from the right superior pulmonary

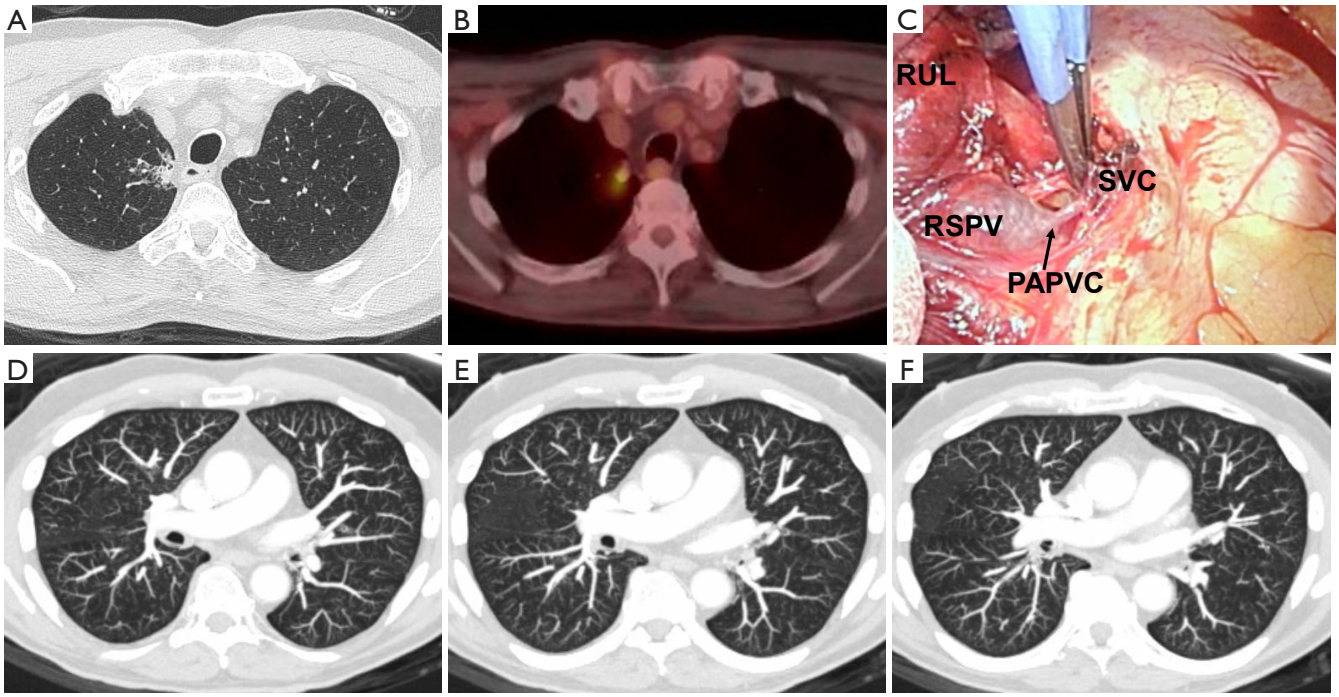

Figure 1 Preoperative and intraoperative imaging of subject presenting with a PAPVC originating in the right upper lobe pulmonary vein and draining into the superior vena cava. (A) The described subject presented with a 1.9 cm GGO of the right upper lobe; (B) preoperative PET demonstrated an SUV of 4.9; (C) during resection, a PAPVC from the right superior pulmonary vein to the vena cava was noted; (D-F) upon review of previous axial imaging, the PAPVC was identifiable. PAPVC, partial anomalous pulmonary venous connection; RUL, right upper lobe; RSPV, right superior pulmonary vein; SVC, superior vena cava.

vein and draining into the superior vena cava was identified (*Figure 1C*). Intraoperative review of a previous contrast enhanced chest CT confirmed partial anomalous pulmonary vein (*Figure 1D,E,F*), and revealed no other cardiopulmonary abnormalities.

Because the anomalous vein was associated with the lobe harboring the preoperatively identified GGO, the decision was made to proceed with resection without additional cardiopulmonary evaluation. A right upper lobectomy was performed, with ligation of the anomalous vein near the vena cava. After sampling lymph nodes for staging, the case was concluded in standard fashion. A postoperative echocardiogram was obtained, and ruled out septal defects. The patient is now 6 months from his procedure without evidence of cardiopulmonary compromise or disease recurrence.

Discussion

PAPVCs are rare congenital anomalies found in 0.4–0.7% of the population at autopsy (2). These anomalies are most frequently identified in children, where they are typically right sided and are associated with ASDs in 80–90% of cases (1,2). In the pediatric population, repair is indicated when pulmonary-to-systemic flow ratio (Qp/Qs) approaches 1.5 (2,3).

In contrast, PAPVCs in adults, particularly when associated with pulmonary nodules, are far less common with only 24 cases being reported to date (data summarized in *Table 1*). Given the rare presentation of PAPVCs in adults requiring pulmonary resection, treatment approaches are not established. Despite a small sample size, epidemiologic and therapeutic trends may be ascertained from systematic analysis of adult reports.

Upon review, the mean age of presentation of patients with PAPVCs and pulmonary nodules was found to be 59 years (SD 14 years). There appears to be no gender predominance (44% female, 56% male) nor laterality predisposition (46% are left-sided, 54% are right-sided). Approximately half of PAPVCs are found preoperatively

Table 1 Reports of PAPVCs encountered during pulmonary resection

Case	Age (years)	Gender	Tumor location	Pathology	PAPVC source	Resection	PAPVC drain	ASD present?	PAPVC corrected?**	Author and year
1	72	Female	RML & RLL	NSCLC	LUL	Pneumonectomy	Left brachiocephalic vein	No	No	Black *et al*., 1992 (4)
2	68	Male	LUL	NSCLC	LUL	Lobectomy	Left brachiocephalic vein	No	No	Takamori *et al*., 1995 (5)
3	58	Female	LLL	NSCLC	LUL	Lobectomy	Left brachiocephalic vein	No	Yes	Takei *et al*., 2002 (6)
4	79	Female	LUL	NSCLC	LUL	Lobectomy	Left brachiocephalic vein	No	No	Miwa *et al*., 2004 (7)
5	48	Male	LUL	NSCLC	RUL	Pneumonectomy	Superior vena cava	No	Yes	Sakurai *et al*., 2005 (8)
6	45	Female	LLL	NSCLC	LUL	Lobectomy	Left brachiocephalic vein	No	No	Yamada *et al*., 2005 (9)
7	62	Male	RUL	NSCLC	RUL	Lobectomy	Azygous vein and superior vena cava	No	No	Galetta *et al*., 2006 (10)
8	59	Male	RUL	NSCLC & TB	RUL	Lobectomy	Superior vena cava	No	No	Sasaki *et al*., 2006 (11)
9	45	Female	LLL	Carcinoid	LUL	Lobectomy	Left brachiocephalic vein	No	Yes	Smith *et al*., 2006 (12)
10	81	Male	RUL	NSCLC	RUL	Lobectomy	Superior vena cava	No	No	Tanaka *et al*., 2008 (13)
11	65	Female	RML	NSCLC	RML	Lobectomy	Right inferior pulmonary vein	U	No	Akiba *et al*., 2008 (14)
12	44	Female	LLL	Granuloma	LLL	Lobectomy	Azygous vein	U	No	Sumida *et al*., 2009 (15)
13	80	Male	RUL	NSCLC	RUL	Lobectomy	Superior vena cava	No	No	Pryshchepau *et al*., 2009 (16)
14	64	Male	RUL & RLL	NSCLC	RUL	Lobectomy	Superior vena cava	No	No	Mikubo *et al*., 2013 (2)
15	U	U	LUL	NSCLC	LUL	Lobectomy	Left brachiocephalic vein	No	No	Szkorupa *et al*., 2013 (17)
16	45	Male	RLL	NSCLC	RUL	Bilobectomy (RML and RLL)	Superior vena cava	U	No	Tourmousoglou *et al*., 2014 (18)
17	64	Male	LUL	NSCLC	LUL	Lobectomy	Left brachiocephalic vein	No	No	Asakura *et al*., 2014 (19)
18	26	Female	LUL	NSCLC	LUL	Lobectomy	Left brachiocephalic vein	U	No	Shapiro *et al*., 2014 (20)
19	48	Male	RLL	NSCLC	RLL	Pneumonectomy	Inferior vena cava	No	No	Liu *et al*., 2015 (21)
20	61	Female	RUL	NSCLC	RUL	Lobectomy	Azygous vein	No	No	Heineman *et al*., 2015 (22)
21	59	Male	RUL	NSCLC	RUL	Lobectomy	Superior vena cava	No	No	Inafuku *et al*., 2016 (23)
22	74	Female	LLL	NSCLC	LUL	Lobectomy	Left brachiocephalic vein	No	Yes	Kawagoe *et al*., 2017 (3)
23	45	Male	RUL	NSCLC	RUL	Lobectomy	Azygous vein	U	No	Kawasaki *et al*., 2017 (24)
24	69	Male	RUL	NSCLC	RUL	Lobectomy	Superior vena cava	No	No	Current case, 2017

**, correction describes reimplantation. PAPVC, partial anomalous pulmonary venous connection; NSCLC, non-small cell lung cancer; ASD, atrial septal defect; RUL, right upper lobe; RML, right middle lobe; RLL, right lower lobe; LUL, left upper lobe; LLL, left lower lobe; TB, tuberculosis; U, unknown.

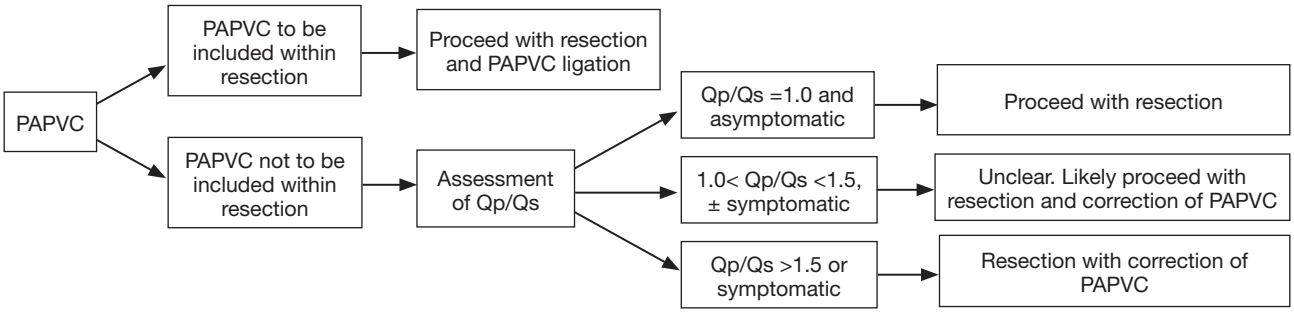

Figure 2 Proposed treatment algorithm for adult patients with PAPVC requiring pulmonary resection. If the PAPVC originates from the pulmonary segment undergoing resection, standard resection with PAPVC ligation is appropriate. If the PAPVC originates from a pulmonary segment that is not being included in the pulmonary resection, management depends on preoperative Qp/Qs and the extent of resection. For patients with normal Qp/Qs, pulmonary resection alone is sufficient. For patients with Qp/Qs >1.5, PAPVC correction should occur along with pulmonary resection. For patients with Qp/Qs ranging from 1.0 to 1.5, PAPVC correction should likely take place, particularly for patients requiring extensive pulmonary resection. PAPVC, partial anomalous pulmonary venous connection.

with the other half found intraoperatively (3). The most common drainage patterns in adults involve connections from the left superior pulmonary vein to the innominate vein (41.6%) and the right superior pulmonary vein to the superior vena cava (29.1%). Perhaps most interestingly, although PAPVCs are highly associated with ASDs in the pediatric population, there are no reports of adult patients with a PAPVC and lung nodules who have been found to have an ASD. These data may suggest that additional studies aimed at assessing septal abnormalities are of little added utility.

Management of PAPVCs in the setting of pulmonary resection requires careful consideration of several factors including PAPVC anatomy, preoperative cardiopulmonary physiology (as assessed by Qp/Qs), and the extent of planned pulmonary resection. For PAPVCs that arise from a pulmonary segment that will be included in the resection specimen, simple ligation is adequate. In fact, some reports suggest that resection may result in improvements of pulmonary hypertension (19).

When a PAPVC arises from an anatomic segment which will not be included in the resection, the management approach is more challenging and requires a pre-resection assessment of Qp/Qs and consideration of resection extent. These factors are important as significant pulmonary resections in patients with elevated Qp/Qs ratios can result in fulminant right-heart failure due to acute increases in shunt fraction through the PAPVC and elevated pulmonary arterial pressures (3,4). Although a clear Qp/Qs cut-off is unknown, patients with Qp/Qs greater than 1.5 or symptomatic PAPVCs are at high risk for post-operative complications, and thus should undergo PAPVC repair. Successful PAPVC repair has been described both prior to and during pulmonary resection with good success (3,6,8,12). For patients with asymptomatic PAPVCs and normal Qp/Qs, standard pulmonary resection is likely sufficient. Finally, in patients with lesser resections and borderline Qp/Qs (ranging from 1.0 to 1.5), it remains unclear if PAPVC correction is necessary with reports demonstrating success with both approaches (9,12,18). A summary of this treatment algorithm is provided in *Figure 2*.

Although these data provide a basic blueprint for both diagnostic and therapeutic approaches for the rare patient presenting with PAPVCs and resectable pulmonary nodules, some potential limitations should be noted. First, the compiled data is based almost exclusively on single case reports from a variety of centers across the world. This methodology introduces heterogeneity in the treatment approach and limits interpretation of long term follow-up data. Second, there is no randomization or control group which can complicate outcome interpretation. Finally, the prevalence of PAPVCs is likely underestimated as nearly half of these anomalies were found only intraoperatively during resection. Evaluation of the contralateral hemithorax is thus impossible during resection. Nevertheless, these results represent the first formal review characterizing trends and provides an applicable treatment algorithm for this challenging cohort.

In summary, management of PAPVCs during resection is an uncommon clinical scenario that requires careful consideration of venous anatomy, preoperative

cardiopulmonary physiology, and resection extent. For patients with PAPVCs arising from the pulmonary segment to be included in the resection specimen, simple vein ligation is appropriate. For those patients with PAPVCs arising from pulmonary segments which will not be included in the resection, consideration of preoperative Qp/Qs and resection extent is essential when determining the need for PAPVC correction.

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Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Informed Consent: Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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