Original Article

# Airway Anomalies in Cases of Anomalous Pulmonary Venous Connection – A Single-Center Experience


## Abstract

**Background:** Patients with congenital heart defects may present with concomitant defects involving other organ systems. Roughly 4 percent of this nature are airway anomalies. Presence of anomalous airways summon major challenge before the anesthesiologist, surgeon, and intensivist in the perioperative management of such patients. There is paucity of literature in the study of airway anomalies in the subset of congenital anomalous pulmonary venous connections. We present the analysis of three cases of airway anomalies in patients operated for anomalous venous drainage at our center. We hope to explicate the clinical implications and management of such rare presentations. **Methods:** The records of all patients who underwent surgical correction for anomalous venous return between January 2016 and January 2018 were reviewed retrospectively. The records were examined for presence of any airway issues, abnormal radiological findings, perioperative intubation or extubation issues and perioperative surgical findings. **Results:** Amidst the 410 cases operated for congenital heart defects in this period, 92 were operated cases for anomalous pulmonary venous return, of which 3 patients presented with airway issues. One patient had an aberrant right tracheal bronchus with normal carina and bilateral main bronchial stenosis, the second patient had a hypoplastic left lung and the third patient had congenital lobar emphysema of the left lung. **Conclusion:** Prudent perioperative management necessitates prior evaluation and preemptive planning for airway anomalies in patients with anomalous venous return, since they can belong to the "Malinosculation Syndrome" group, which involves anomalous communication by means of small openings between the different components of lung tissue, namely, the lung parenchyma, tracheobronchial tree, arteries, and veins.


**Keywords:** *Airway compression, anomalous pulmonary venous connection, congenital, intubation, tracheal bronchus*


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## Introduction

Patients with congenital heart disease (CHD) may have coexisting defects involving other organ systems too.[1] The incidence of airway anomalies in CHD has been found to be about 4%.[2] Patients with anomalous pulmonary venous connections (APVCs) constitute up to 10.6% of all CHDs reported in India.[3] At presentation for corrective surgery, presence of airway anomalies is a major encumbrance for the anesthesiologist, surgeon, and intensivist with regard to their perioperative management. However, there is limited literature pertaining to the study of airway anomalies in APVC alone. We present three cases of airway anomalies with different presentations, operated for APVC from our center. In addition, we elucidate the clinical implications and association of these concomitant lesions by means of a retrospective study.

## Materials and Methods

This is a retrospective analysis of all patients who underwent surgical repair of APVC at our institution between January 2016 and December 2017. The patient records were evaluated for any airway issues – abnormal radiological findings, perioperative intubation or extubation issues, and intraoperative surgical findings. Patients with external airway compression due to a vessel anomaly or chamber enlargement were excluded, as they were not considered cases of primary congenital airway disease.

## Results

*Patient characteristics*: Among the 410 congenital corrective procedures performed at our center in these 2 years, 92 were for APVCs [total APVC (TAPVC) in 21 and partial APVC in 71 patients]. There were more males (53) than females (39). The mean age and weight



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were 14.75 years (7 days–38 years) and 31.8 (2.5–48) kg, respectively.

Apart from the patients mentioned included in Table 1, one male patient had right upper pulmonary vein (RUPV) draining to the right atrium (RA) and left upper pulmonary vein to left superior caval vein (LSVC). Another female patient had RUPV draining to the azygous vein going to SVC [Table 2].

Herein, we discuss three interesting cases of associated airway abnormalities in APVC patients, which sprung a surprise during the postoperative period.

## Case Reports

### Case 1

A 2-year-old child presented with history of repeated lower respiratory tract infections treated with antibiotics and bronchodilators. She was diagnosed to have a supracardiac TAPVC by the cardiologist. She was taken up for surgery after the routine preoperative workup. An uncuffed endotracheal tube (ET) 4.5 mm ID was used during anesthesia and was fixed at 12.5 cm at the angle of lip. Intraoperative extensive dissection could not delineate any vertical vein and hence confirmed as cardiac TAPVC. After the patient met all criteria for extubation, she was extubated on the first postoperative day. Immediately after extubation, the child had paradoxical breathing with mild wheezing. Arterial blood gas revealed hypercapnia with normal oxygenation. She was reintubated with uncuffed ET size 4 (expecting stridor and airway edema), but since a large leak was encountered it was upsized to uncuffed ET size 4.5.

An initial suspicion of pulmonary hypertensive crisis was made and echocardiography done.

The right ventricular systolic pressure (RVSP) was 25 mmHg (preoperative RVSP was 50 mmHg) which ruled out pulmonary hypertensive crisis. On the second trial of extubation, a similar sequence was noted. We ruled out phrenic nerve injury during dissection by observing bilateral normal diaphragmatic motion on ultrasonography during spontaneous breathing on T-piece. Appropriate antibiotics were started as per culture sensitivity pattern of ET secretions. A third trial of extubation after 3 days had similar results. Suspecting extrinsic vascular compression, a computed tomography (CT) thorax (with contrast and three-dimensional reconstruction) was done which revealed an aberrant right tracheal bronchus (TB) with normal carina and bilateral main bronchial stenosis [Figure 1]. A 2.8-mm intubation fiberscope (Karl Storz SE®, Germany) could not be inserted into the trachea beyond the division of TB making it critical tracheal stenosis (normal for age expected 8–9 mm). The opinion of a pediatric pulmonologist was taken for possible need for tracheal stenting. However, conservative management was continued as advised by pediatric pulmonologist. The patient was started on nebulized antibiotics and physiotherapy to clear her secretions.

Over a week, the patient was gradually weaned off invasive ventilation with introduction of noninvasive ventilation with high pressure control and pressure support. We accepted hypercapnea (upto 96 mmHg on venous blood gases) as the patient was neurologically and hemodynamically stable. She was gradually weaned off noninvasive ventilation over another week and was discharged on the 30th postoperative day with a clear chest.

### Case 2

A 20-day-old infant weighing 3 kg presented with difficulty in feeding and profuse sweating during the same. The child had a dysmorphic left upper limb. Auscultation revealed decreased breath sounds on the left side.

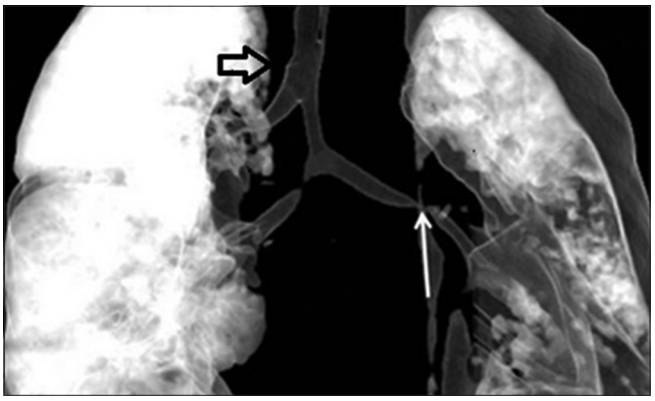

**Figure 1: Black arrow shows tracheal bronchus, white arrow shows bronchial stenosis**

**Table 1: Patient characteristics**

| | TAPVC | | | |
|---|---|---|---|---|
| | **Supracardiac** | **Cardiac** | **Infracardiac** | **Mixed** |
| Male | 4 | 0 | 1 | 4 |
| Female | 9 | 2 | 0 | 1 |

| | PAPVC | | | | | |
|---|---|---|---|---|---|---|
| | **RUPV Drainage to** | | | **RUPV, RMPV drainage to** | | |
| | **Right atrium** | **Right atriocaval junction** | **SVC** | | **Right atrium** | **Right atriocaval junction** | **SVC** |
| Male | 14 | 7 | 9 | Male | 3 | 2 | 4 |
| Female | 10 | 6 | 8 | Female | 1 | 2 | 3 |

**Table 2: Bronchopulmonary vascular malinosculation classification**

| Color | Type | Defects Included |
|---|---|---|
| I | Isolated bronchial PM | Tracheal bronchus, tracheal stenosis, Tracheal cyst, Congenital Bronchial stenosis, bronchial atresia, brochogenic cyst, congenital lobar emphysema, CPAM |
| II | Isolated Arterial PM | Interrupted Pulmonary Artery, Isolated Systemic Arterial supply to normal lung, Dual arterial supply to normal lung |
| III | Isolated venous PM | APVC, TAPVC, Isolated Scimitar vein, Meandering pulmonary vein, Scimitar variant, Anomalous unilateral single pulmonary vein |
| IV | Mixed Bronchoarterial PM | Typical intralobar sequestration |
| V | Mixed bronchovenous PM | Combination of Bronchial (I) and Pulmonary venous anomalies (III) |
| VI | Mixed Arterio- Venous PM | Fistula between the following: 1.Systemic/pulmonary artery and vein 2.Scimitar vein and systemic artery supply |
| VII | Mixed Broncho-Arterial-Venous anomalies | 1.Classical Scimitar syndrome 2.Typical extralobar sequestration 3.Variations in bronchial, arterial and venous anomalies |

Chest X-ray [Figure 2a] showed absence of normal air bronchogram on the left side. Echocardiography revealed TAPVC to coronary sinus with large ostium secundum atrial septal defect (ASD), bearing right to left shunt. Intraoperative findings confirmed TAPVC draining to coronary sinus, bilateral SVC with larger left SVC, and hypoplastic left lung with small left pulmonary artery. Under total circulatory arrest, coronary sinus was deroofed, rerouting all pulmonary veins to left atrium (LA) through ASD using untreated autologous pericardial patch and LSVC was routed to LA appendage. The sternum was not closed due to significant edema and the child was shifted to the intensive care unit. The child underwent a delayed sternal closure the next day with stable hemodynamics. The child was extubated on the second postoperative day.

While weaning, the $PCO_2$ was noted to be as high as 55 mmHg with associated parameters being normal. Postoperative echocardiography was normal with RVSP 30 mmHg and normal biventricular function. On postoperative day 5, the child had to be reintubated in view of aspiration and was weaned off ventilator over the next 3 days. The child was discharged and advised pneumococcal and influenza vaccination. Follow-up at 8 months showed normal growth with normal biventricular function. X-ray of the upper limb revealed absent ulna on the left side for which she was advised conservative management by an orthopedician [Figure 2b].

### Case 3

A 1-year-old child presented with repeated respiratory tract infection and was diagnosed as sinus venosus ASD with APVC. Preoperative chest X-ray showed hyperlucency in the left upper zone without any mediastinal shift. There was slight decreased breath sounds on the left upper zone although no specific percussion findings could be elucidated. CT cardiac done for mapping the pulmonary venous connection confirmed congenital lobar emphysema (CLE) of the left lobe of the lung [Figure 3]. During surgery, i.v. access was established the previous night while blood sample was sent for cross matching. The child was premedicated with i.v. ketamine 0.25 mg/kg with midazolam 0.05 mg/kg intubated with i.v. vecuronium 0.1 mg/kg.

Anesthesia was maintained with air:$O_2$, isoflurane with positive pressure ventilation. Perioperatively, we used pressure regulated volume control mode for ventilation setting tidal volume around 8 mL/kg keeping peak airway pressures less than 25 $cmH_2O$. Intraoperative findings showed right superior pulmonary vein draining into azygous vein, draining into SVC. Warden's procedure was performed. Postoperative extubation and clinical course were, however, uneventful.

## Discussion

In our institute, we had three patients who were operated for APVC and had coexisting airway anomalies. This initiated us to conduct a retrospective analysis of our hospital data regarding the association between the two issues – CHD and airway abnormalities. Our institutional incidence for this occurrence was 3.2%. A review of the available literature was made with an effort to analyze the possible associations between APVC and airway anomalies.

### Developmental anatomy of the lungs and bronchus

Normal tracheobronchial development is initiated at 24–26 days as a median bulge of the ventral wall of the pharynx that develops at the caudal end of the laryngotracheal groove. At 28–30 days, the lung buds elongate into primary bronchi; at 30–32 days, five lobar bronchi appear as a monopodial outgrowth of the primary bronchi. All segmental bronchi are formed by 36 days. Three theories, namely, reduction, migration, and selection, postulate the abnormal bronchi. Supernumerary occurs early in the development, at about 29–30 days of the embryonic life, as the lobar bronchi begin to differentiate. Displaced bronchi are more likely to occur after 32 days of embryonic life, as the bronchi elongate and branch further.[4]

Pulmonary veins develop from a rich capillary plexus that initially drains the developing lung buds. This plexus

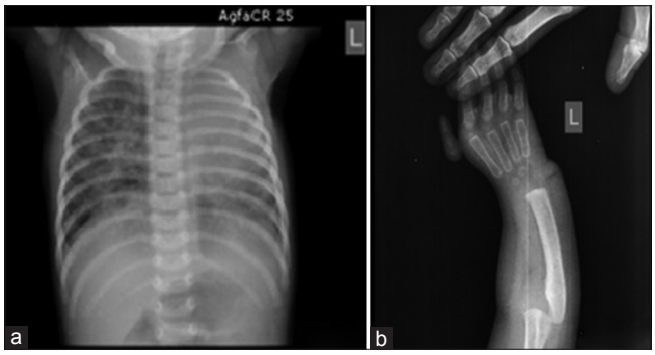


**Figure 2: (a) Preoperative X-ray of patient. (b) X-ray of forearm bones**

initially drains into portions of anterior and posterior cardinal system and right vitelline venous systems, but these primitive connections regress. With later development, the pulmonary veins form a confluence or a common chamber that drains into an outpouching of the left side of the developing atrium. Failure of these connections results in anomalous pulmonary venous drainage.[5]

It is enthralling to note these anomalies being part of the same spectrum of "malinosculation syndromes;" a patient with APVC could also have coexisting airway anomalies.

**Tracheal bronchus**

The term tracheal bronchus was first described by Sandisfort in 1785.[6] It was defined as an aberrant, accessory, or ectopic bronchial branching originating from the right lateral wall of the trachea with diameter ranging from 0.5 to 1.0 cm and its length ranging from 0.6 to 2.0 cm. It has a higher male preponderance. Anthropologically, "swine bronchus" is a normal finding in some animals like sheep, swine, and cattle, but it is a rare finding in humans. Most of the cases of the TB are asymptomatic, and they are detected only incidentally during bronchoscopy or radiologic examinations.

Depending on the number of segmental bronchi of the anatomical right upper lobe bronchus, they can be categorized as follows:

1. Displaced: right upper lobe bronchus bifurcates. One segmental branch of the anatomically normal upper lobe bronchus is simultaneously absent and "replaced" by an aberrant bronchus originating from the trachea
2. Supernumerary: right upper lobe bronchus trifurcates. TB exists in addition to an anatomically normal branching upper lobe bronchus.[6]

The term "pig bronchus" or "bronchus suis" is used when there is tracheal origin of the entire right upper lobe bronchial system.

Kubik and Muntener and Boyden termed tracheal bronchus in relation to pulmonary artery. On the right side, TB arising proximal and distal to the origin of the upper lobe bronchus was called preeparterial and posteparterial, respectively. Similarly, on the left side, TB

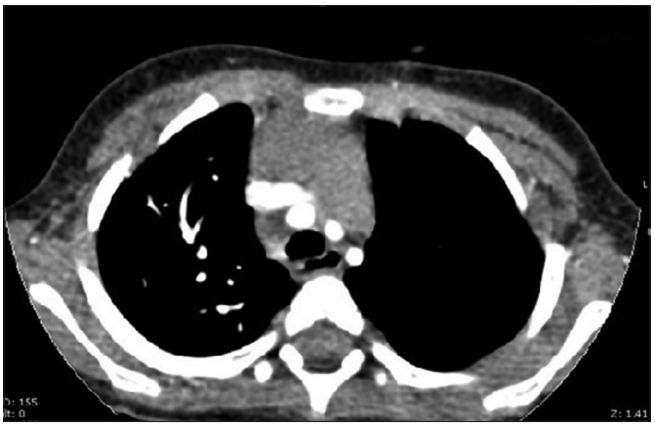

**Figure 3: Congenital lobar emphysema of left lung**

was termed eparterial or prehyparterial and posthyparterial depending on its origin proximal or distal to the normal bronchus.[4] The supernumerary TB aerates the normal lung parenchyma, a cyst, or an accessory segment of the right upper lobe. The lung tissue can be intra- or extralobar, depending on whether it shares the pleura of the upper lobe. In addition, it may have independent blood supply, from either the systemic or the pulmonary arterial system.

In our study, our first patient had an aberrant right TB with normal carina and bilateral main bronchial stenosis in addition to cardiac type of TAPVC. The second patient had TAPVC draining into coronary sinus, bilateral SVC with larger left-sided SVC, and hypoplastic left lung with small left pulmonary artery. The third patient had left lobe CLE with right superior pulmonary vein draining into azygous vein draining into SVC.

We further noted that Pryce used the term "sequestration" to describe the abnormal lung that was "disconnected" or "secluded" from the normal bronchial tree and had anomalous systemic arterial supply (classically described as intralobar sequestration). However, he found variations and termed the variants as Pryce types I, II, and III.[7]

Sade *et al.* proposed the term "sequestration spectrum" in 1974, to include all the various combinations of lung and vascular anomalies encountered.[7] Later in 1987, Clements and Warner proposed a new term "malinosculation." This term was used to include the entire gamut of abnormalities where there is anomalous communication by means of small openings between the different components of lung tissue, namely, the lung parenchyma, tracheobronchial tree, arteries, and veins.[7] This classification is a systematic approach for the evaluation of bronchopulmonary vascular malinosculation, taking into account isolated and concurrent abnormalities of airway, arteries, and veins.

By this classification [Figure 4], patients 1 and 3 would be in group V and patient 2 in group VII of BVPM.[8-10]

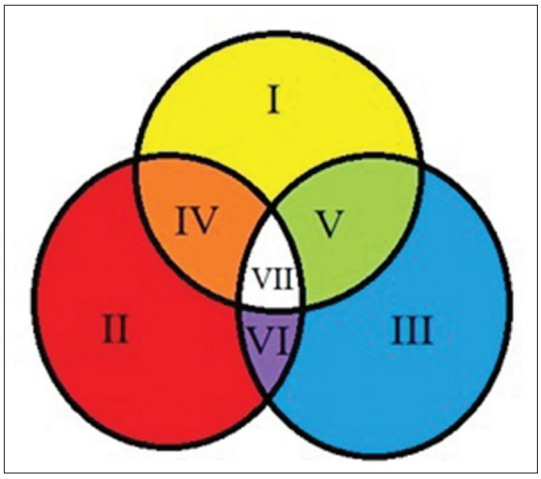


**Figure 4: Graphical representation of various types of bronchopulmonary vascular malinosculation with color coding indicating an overlap of the lesions[8-10]**

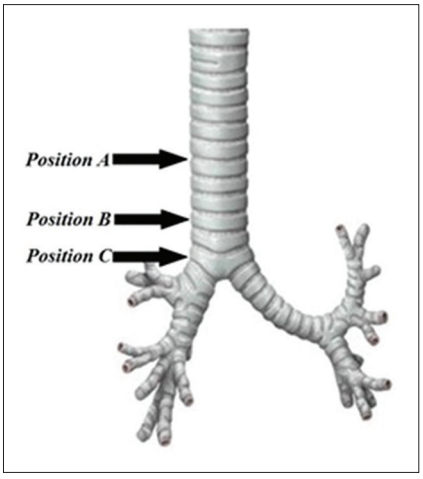


**Figure 5: Tracheal bronchus arising at different positions**

## Clinical implications

*TB*: This condition is usually asymptomatic; diagnosis is often accidental and achieved by bronchography, bronchoscopy, or chest CT. Although TB are usually reported to be asymptomatic, they may cause stridor in children. They may also be related to inflammatory conditions affecting the lung on that side, including recurrent pneumonia nonobstructive bronchiectasis and postobstructive pneumonia distal to an adenoma arising in the TB. Diagnosis of this collapsed area is difficult in children because of the brief distance between atelectasis areas and normally ventilated areas which may mislead auscultation that may be apparently normal due to transmission of sounds from adjacent areas.

For an anesthesiologist, the site where TB arises in relation to the carina is important.[8] Accordingly, they can be three positions significant as shown in Figure 5. When the bronchus arises at Position A, that is, far away from carina, symptoms are most severe, with children, either with wheeze or stridor, often misdiagnosed and managed for asthma.[9] At Position B, the TB is about 1 cm above the carina, and at Position C, it arises at the carina. A TB has been reported as high as 6 cm above the carina. In such patients, a tracheal tube could obstruct an ectopic TB or an ectopic TB could be intubated, causing atelectasis, hypoxaemia, or both. The usage of short tubes to prevent ET entry into the TB and fiber optic confirmation of the position of the tip of the ET post intubation are preferable.[10] Using a standard length ET could obstruct the TB when it arises at Position B and causes a hypoxic shunt. In desaturation, the ET should be carefully withdrawn, monitoring for improvement in oxygen saturation while auscultating the apical zone of the lung.

In our first patient, the tube was fixed about 0.5 cm higher than for her age (expected 13 cm) during surgery, merely based on auscultatory findings. This probably avoided the oversight of missing ventilation of the right upper lobe. Hence, the problem was not diagnosed intraoperatively.

Preoperative assessment of the anatomy and adequate planning is essential for one-lung ventilation (OLV) when required in patients with TB to avoid serious hypoxaemia, atelectasis, and complications.[11] In TB with a narrowed distal trachea, a bronchial blocker (BB) may have advantages over the conventional DLT in achieving OLV. But there are several cases reported which cite the inability to isolate the right lung with BB.[12] Using a left-sided DLT made it possible to perform complete OLV in some cases after failing to collapse the right lung, using BB. Typically, patients with TB can be well-managed with a left DLT. If a left DLT is not adequate, other potential alternatives include the use of a Univent tube alone; use of a Fogarty catheter to block the TB; use of BB with Univent tube; use of 2 BB, one to block the TB and another to block the normal bronchus.[12-15]

Our patient had TB with distal tracheal and bronchial stenosis which was conservatively managed at this stage and was advised for yearly follow-up. However, depending on the severity of stenosis, several options exist for management[16] including slide tracheoplasty, segmental resection, and anastomosis for discreet, short-segment stenosis. Postoperative airway stenosis is managed with endoscopic balloon dilatation techniques. Tracheal stenting is used as an adjuvant therapy with its own additional morbidity and is currently only used as salvage procedure. Newer techniques incorporate bioabsorbable materials and three-dimensional printer devices. These design airway stents are to be anatomically specific and custom-made.

Regarding our second patient, another manifestation of malinosculation could be the occurrence of "hypoplastic" bronchus/lung or "spongy" lung. This child in the absence of the cardiac lesion could have directly presented for noncardiac surgery as upper limb reconstruction as in this case.

Our third patient had CLE which *per se* was asymptomatic and did not pose any perioperative problem. Literature suggests CLE as a rare entity with male predominance, usually unilateral affecting more often the left upper lobe (43%) followed by the right middle lobe (32%). Bilateral involvement has been reported in 20%. Associated CHD is present in approximately 15% of patients with CLE.[17] Classical presentations are life-threatening respiratory distress during infancy secondary to hyperinflated lung due to ball valve effect.[18] This results in compression atelectasis of ipsilateral or contralateral side with mediastinal shift, hypoxia, and associated hypotension. The disease is frequently demented with pneumonia and pneumothorax.[19] Insertion of an intercostal drainage may worsen the situation. Controversy still exists regarding the management of CLE. Although conservative management is suitable for milder cases, lobectomy is the mainstay of treatment. Our patient was managed conservatively.

## Conclusion

It is important to seek and recognize airway anomalies in APVC patients keeping in mind that they could be a part of the spectrum of "malinosculation syndromes." This could lead to better perioperative planning and management of these patients rather than being taken aback with unpleasant surprises on table.

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**Conflicts of interest**

There are no conflicts of interest.

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