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Hypogenetic right lung with partial anomalous pulmonary venous return and accessory diaphragm: a case of "scimitar lung"

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Partial anomalous pulmonary venous return (PAPVR) is a rare congenital cardiovascular condition in which some of the pulmonary veins drain into the systemic circulation. We report on the cadaveric dissection of a 71-year-old Caucasian male donor who died of chronic obstructive pulmonary disease with hypertension. We noted a faint incisional scar on the thorax extending from the parasternal region at the 4th intercostal level to the midaxillary line. Since the straight-line incision followed the ribs and the scar was quite faint, surgery probably occurred when the donor was young. We also observed numerous surgical interventions of the heart, lungs, and vasculature to correct various defects.

The morphology of the heart was normal, but was shifted more to the right side. An atrial septal defect (ASD) was closed with sutures. The right superior pulmonary vein that drained into the superior vena cava (SVC) was ligated close to the SVC and the right inferior, left superior, and inferior pulmonary veins all drained directly into the left atrium. We noticed a dilated coronary sinus entering the right atrium adjacent to the ASD; the ostium of the coronary sinus noticeably lacked the normal valve-like structure. We initially thought the right lung was a "horseshoe" lung, but realised that it was a "hypogenetic" lung with PAPVR and an accessory diaphragm. Compared to the left, the right secondary bronchi were much narrower and branched uncharacteristically, as seen in hypogenetic lung syndrome. The inferior lobe was highly disorganised, severely hypoplastic, and exhibited uncharacteristic morphology. The superior bronchopulmonary segment was markedly hypoplastic. The posterior and medial basal segments were not only hypoplastic and slender, but also extended like a tail to the left pulmonary cavity behind the heart/pericardium and in front of the oesophagus and aorta. The right lung, though hypoplastic, demonstrated patent bronchi and the lobes were inflatable. Based on the hypogenetic lung and PAPVR, we conclude that the donor exhibited 'scimitar' lung. (Folia Morphol 2023; 82, 4: 980–987)

Key words: scimitar lung, congenital cardiovascular anomaly, partial anomalous pulmonary venous return (PAPVR), partial anomalous pulmonary venous connection (PAPVC)

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INTRODUCTION

Partial anomalous pulmonary venous return (PAPVR), additionally recognised as partial anomalous pulmonary venous connection (PAPVC), is an inherited cardiac circulatory condition; it occurs infrequently and results in few, but not all of the pulmonary veins emptying into the general circulation, instead of the left atrium [19]. Present at birth, PAPVC is a multifaceted cardiac defect; it has no known cause, though there is a possibly multifactorial origin that includes genetic component [31]. Numerous variations of this vascular anomaly have appeared in the literature. The first description of scimitar syndrome, a variant of partial anomalous pulmonary venous drainage (PAPVD), appeared in 1836 [3, 4]. This rare, complex, congenital anomaly is characterised by abnormal pulmonic venous blood flow from the right lung shunted into the inferior vena cava (IVC) or right atrium via superior vena cava (SVC), thus a left-to-right shunt. Hypoplasia of the right lung, dextrocardia, malformations of the right pulmonary artery and bronchial tree, and abnormal arterial supply of the right lung (the so-called pulmonary sequestration) are associated anomalies that manifest frequently. Atrial septal defect happens in approximately 40% of cases with right-sided PAPVR [21]. Classic scimitar syndrome consists of hypoplasia of the right lung and right pulmonary artery, dextroposition of the heart, an abnormal pulmonary vein draining into the IVC, and a systemic collateral supply to the lung [3, 4].

In the complete form of the infrequently occurring, inherited abnormality called scimitar (hypogenetic) lung, systemic arterial blood flow supplies the right lung and its inferior lobe and empties partially or totally to the general veinous circulation. Hypoplasia of the pulmonary arteries and uncharacteristic bronchial branching accompany this disorder [23, 24]. The aberrant pulmonary venous return typically includes the right side [5]. A rare variation called horseshoe lung is often accompanied by vascular abnormalities of the hypogenetic lung [10, 17, 33].

The blending of posterior basilar segments of the right and left inferior lobes involving an incomplete parietal pleural deficiency constitutes horseshoe lung, which has a frequency that varies from 0.4% to 0.7% [11]. Most horseshoe lung cases are associated with right lung hypoplasia; approximately 80% are associated with PAPVR from the right lung to the IVC or right atrium (scimitar syndrome) [20, 37]. In PAPVR, one of the abnormal veins from the lung drains into general venous circulation, creating a left-to-right diversion, rather than normally occurring emptying of venous blood from the lung into the left atrium. In right-sided PAPVR, an anomalous pulmonary vein drains into either the SVC [18], right atrium [13], coronary sinus [22], or IVC [9]. Right upper lobe flow most commonly drains into the SVC and the condition is often associated with a high sinus venosus atrial septal defect near the orifice of the SVC [18]. An accessory diaphragm seen in the case reported here is a rare congenital anomaly associated with an atrial septal defect described in paediatric patients [28, 34] that occurs mostly on the right side [5] and is composed of fibromuscular tissue with a serosal lining [8]. In all the described cases, aplasia or certain extent of pulmonary hypoplasia was obvious on the involved side [5].

Embryology of lung development

As normal embryonic development occurs in the first 2 months, venous blood from the lungs empties into the systemic veins [9] and the common initial pulmonary vein develops from a bulge in the dorsal wall of the early stage left atrium [9, 11]. As development continues, the early-stage lungs attach to the common pulmonary vein and the connections that allow pulmonary venous return to the systemic veins disappear [9, 11]. Thus, the shared pulmonary vein integrates into the left atrium and four well-separated pulmonary veins arise, two from each lung [9, 11]. Abnormal resorption of the developing structures can bring about (1) changes in the size or the number of pulmonary veins or (2) atypical emptying into the general veins or right atrium.

MATERIALS AND METHODS

The cadaveric specimen was obtained from the willed body programme intended for the purpose of dissection by medical students. This study is based on the cadaveric dissection of a 71-year-old Caucasian male donor who died of chronic obstructive pulmonary disease/hypertension. We observed a number of what appeared to be surgical interventions of the heart, lung, and vasculatures, possibly due to malformations and anomalous venous drainage. In the dissection procedure, the anterior thoracic wall was removed, as guided by the faculty, and the thoracic viscera were dissected and studied. The students' dissection protocol called for opening the thoracic cage and studying in detail first the heart, followed by the respiratory system. Since the major

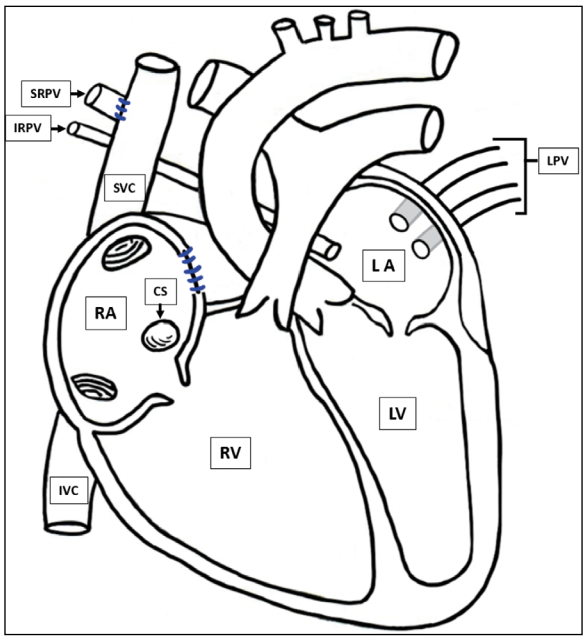

Figure 1. Schematic representation of the heart the students dissected and studied. The defects are corrected by surgical sutures as indicated by zig-zag suture lines, including right superior pulmonary vein that drained into the superior vena cava (SVC), which was ligated close to the SVC and atrial septal defect; RA — right atrium; LA — left atrium; LV — left ventricle; RV — right ventricle; CS — coronary sinus; IVC — inferior vena cava; SRPV — superior right pulmonary vein; IRPV — inferior right pulmonary vein; LPV — left pulmonary veins.

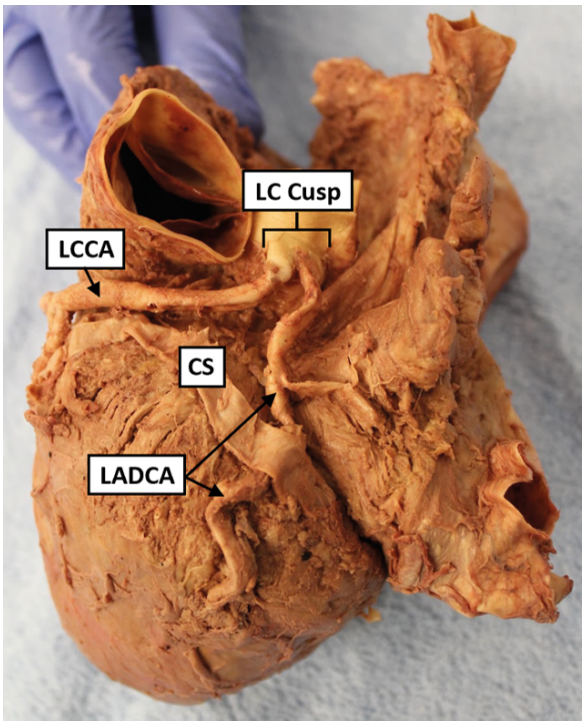

Figure 2. Actual cadaveric heart specimen with independent origins of the left anterior descending and circumflex coronary arteries from the left coronary cusps thus showing the lack of normal left coronary artery. Also shown is the dilated coronary sinus entering the right atrium; LC Cusp — left coronary cusp; LADCA — left anterior descending coronary artery; C-S — coronary sinus; LCCA — left circumflex coronary artery.

part of the dissection was performed by the students, some of the vascular and other structures were not optimally preserved. Additionally, the surgical corrective procedures disturbed the natural architecture of the gross morphology.

OBSERVATIONS AND RESULTS

On the right side of the thoracic wall, we observed an incision extending from the parasternal region at the 4th intercostal level to the midaxillary line, apparently to surgically address anomalies. Considering the straight line (not curved) incision following the ribs on the thorax, we believed that the surgery was performed during the donor's childhood, since the ribs are more horizontal in young age. An experienced in-house pathologist who examined the cadaver described the incision scar as faint and very old, suggestive of surgical intervention in childhood to correct problems in the thoracic viscera.

Heart, vasculatures, and surgical interventions

The morphology of the heart was normal in both size and shape. However, as reported in an earlier study, the heart and mediastinum were shifted more toward the right side [37]. We observed evidence of surgical procedures performed on the heart, likely to correct congenital defects. We found that the atrial septal defect (ASD) was corrected by closure with sutures (Fig. 1). The right superior pulmonary vein that drained into the SVC was ligated close to the SVC (Fig. 1). While the right coronary artery originated normally from the right coronary cusp, the left coronary circumflex and the left anterior descending arteries arose independently from the left coronary cusp, thus lacking the typical left coronary artery (Fig. 2). Additionally, we recognised a dilated coronary sinus entering the right atrium adjacent to the ASD (Fig. 2) and the ostium of coronary sinus noticeably lacked the normal valve-like structure.

Respiratory system: right lung

We initially thought the right lung was a typical 'horseshoe' lung, as described in the literature [21, 37]. Upon closer examination of all the thoracic viscera, vasculatures, and surgical interventions, we

realised that what we encountered was a hypogenetic right lung with PAPVR and accessory diaphragm (Fig. 3). The trachea was normal in size and location, bifurcated at the level of 4th thoracic vertebra; the carina was situated normally at the bifurcation. Upon division from the trachea, the right and left primary bronchi were of normal size. However, the right secondary bronchi were much narrower compared to the left and branched uncharacteristically, as seen in hypogenetic lung syndrome (Fig. 3) [24].

In the hypoplastic right lung, the oblique fissure separated the superior lobe from the inferior lobe and a horizontal fissure defining the small middle lobe was also observed. The inferior lobe was highly disorganised and severely hypoplastic and exhibited uncharacteristic morphology. The superior bronchopulmonary segment was markedly hypoplastic; the posterior basilar and medial basal segments were not only hypoplastic and fused, but also formed a slender "tail" that extended to the left pulmonary cavity behind the heart/pericardium and in front of oesophagus and aorta. However, the fused, tail-like right segments did not fuse with the similar segments of the left lung, as seen in "scimitar" lung. Notably, we also observed an accessory diaphragm separating the fused anterior and lateral basal segments (Fig. 3). Furthermore, the fused anterior and lateral basal segments that were wedged between the true diaphragm and the accessory diaphragm were inflatable. Though hypoplastic, the bronchi were patent; using an air pump, we were able to inflate the superior and middle lung segments. Importantly, we were also able to inflate all the lower lobe bronchopulmonary segments and pass a soft probe (pipe cleaner) into the pulmonary arterial branches of the lower lobe bronchopulmonary segments.

Respiratory system: left lung

Both lobes of the left lung were normal in size and exhibited typical gross morphology. An oblique fissure separated the upper and lower lobes and surface visceral impressions appeared normal (Fig. 3). The left lung shifted more towards the right, causing the observed mediastinal shift, and no pathology was noted. The left lung was inflatable by air pump, showing a normal patency of the bronchi and their functional ability. We were able to pass a soft probe (pipe cleaner) into the pulmonary arterial branches of all bronchopulmonary segments.

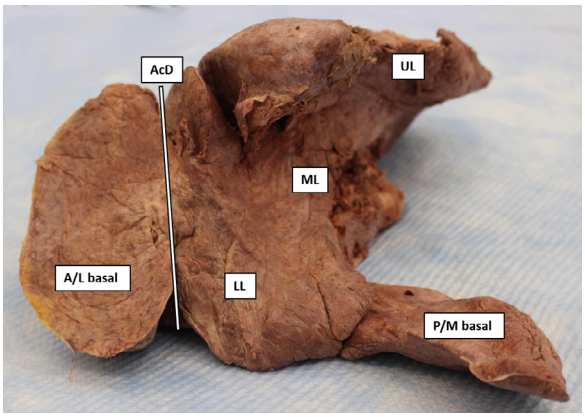

Figure 3. The cadaveric lungs showing normal left lung and the hypogenetic right lung with aberrant morphology. The inferior lobe was highly disorganized and severely hypoplastic and exhibited uncharacteristic morphology. The right superior bronchopulmonary segment was markedly hypoplastic; the posterior basilar and medial basal segments were not only hypoplastic and fused, but also formed a slender "tail" like morphology. We also observed an accessory diaphragm (indicated by a line) separating the fused anterior and lateral basal segments. The fused anterior and lateral basal segments that were wedged between the true diaphragm and the accessory diaphragm were inflatable; UL — upper lobe; LL — lower lobe; ML — middle lobe; AcD — accessory diaphragm; A/L basal — anterior and lateral basal fused segments; P/M basal: — posterior basilar and medial basal fused segments.

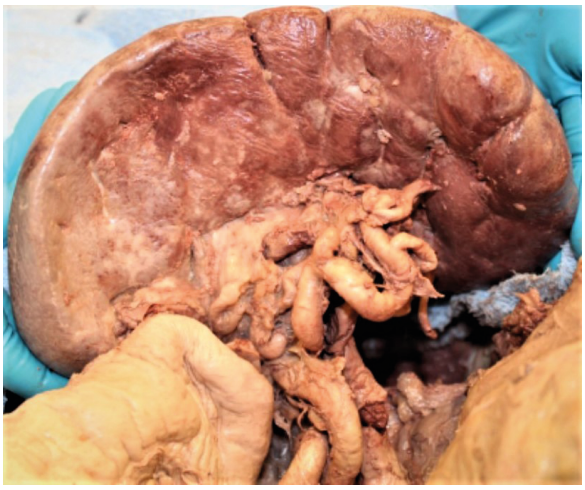

Figure 4. Splenomegaly with greatly enlarged and coiled vasculatures.

Pulmonary veins

The right superior pulmonary vein drained directly into the SVC; the inferior right pulmonary vein from the lower lobe drained into the left atrium. The left superior and inferior pulmonary veins drained directly into the left atrium. In this case, the anomalous pulmonary vein that drained from the right lung into the SVC was corrected by ligation close to the SVC (Fig. 1).

Apart from the cardiopulmonary changes, we also observed hitherto unreported splenomegaly (Fig. 4) and greatly enlarged and coiled splenic vasculatures. The spleen was 28 cm long, 16 cm wide, 6 cm in thickness, and weighed 1,856 g. The liver, pancreas, and kidneys were normal in size without any pathology such as liver cirrhosis, pancreatic cancer, or others.

DISCUSSION

In the following paragraphs, we compare the findings of this case, highlighting relevance with similar cases described in the literature. We have included pertinent embryology related to the case. Oftentimes, in childhood, infants with scimitar syndrome or PAPVR are asymptomatic and this condition goes unnoticed; in adults, it is incidentally observed during imaging studies. However, the person (donor) in this report had surgical interventions at a young age to correct anomalous venous return and cardiac anomaly such as ASD. In addition, we also observed other cardiovascular anomalies, such as a widened coronary sinus and the left anterior descending and left circumflex arteries arising independently from the left coronary cusp.

Congenital pulmonary venolobar syndrome

Felson [12] coined the term congenital pulmonary venolobar syndrome (CPVS) in 1973; it incorporates a mixed group of unusual anomalies that may occur alone or in combination; abnormal unions of the lung parenchyma, the lung and general vasculature, and, infrequently, the digestive system may be seen [12]. Components of CPVS are hypogenetic lung, such as lobar agenesis, aplasia or hypoplasia, PAPVR, lack of pulmonic artery and/or IVC, pulmonic impounding, general pulmonic arterialization, and secondary diaphragm [25].

In scimitar syndrome, a type of PAPVR, venous blood from the hypoplastic lung flows through an abnormal pulmonic vein and drains into the general venous system. This is one of the rare inherited variations of developmental pulmonic venolobar syndrome that almost always occur on the right side [5]. The two most constant features of this syndrome are anomalous pulmonary venous return into systemic circulation and lung hypoplasia [1, 15, 16].

In the case of PAPVR, inconsequential right upper lobe venous drainage into SVC is frequently observed [18]. Most patients live symptom free; some with coexisting cardiac anomalies or dangerous forms of lung atresia or hypoplasia may have medical signs early in life that require surgical intervention. In the case reported (Fig. 1), the hypoplasia of the right lung and cardiovascular anomalies, as well as evidence of surgical interventions, confirm that the donor was symptomatic as an infant. The CPVS is displayed when one or additional, but not all, of the pulmonic veins empty into a general vein, causing a left-to-right bypass. The wide range of clinical presentations of this syndrome comprises hypogenetic lung (including lobar agenesis, aplasia, or hypoplasia), PAPVR, lack of pulmonic artery and/or IVC, pulmonic impounding, general pulmonic arterialisation, and extra diaphragm [12].

A small component of CPVS includes horseshoe lung, wherein a basal segment herniation of the right pulmonary lobe may cause a bridge of lung parenchyma to extend from the right lung bottom through the midline backside of the pericardium/heart to join the posterobasal segments of the left lung: a true horseshoe lung [14]. The herniated portion may be attached or may be separated by pleural membranes. Pleural separation of pulmonary lobes distinguishes pseudo-horseshoe appearance from a true horseshoe lung [35]. In the case reported here, the fusion of the right and left posterobasal segments were not observed, thus establishing the presence of a pseudo-horseshoe lung (Fig. 3). The herniated right posterobasal segment was a functional unit, as it could be aerated using an air pump.

Development of pulmonary vein and related anomaly

The pulmonary vein develops early in embryonic life and the blood from the lung buds empties into the visceral plexus, which connects with paired central veins and umbilicovitelline veins. The right central vein develops into the SVC, while the left cardinal vein mostly disappears. The umbilicovitelline veins develop into the IVC, portal venous system, and ductus venosus. During the 4th fetal week, a shared pulmonic vein arises from the wall behind the left atrium and joins the general pulmonic vein that empties blood flow from the lung buds. Thus, the common pulmonary vein becomes incorporated into the dorsal wall of the left atrium, eventually giving rise to four separate pulmonary veins. If the common pulmonic vein fails to join the visceral plexus and a visceral plexus connection with a principal or umbilicovitelline vein perseveres, some type of full abnormal pulmonic venous link (TAPVC) or limited pulmonic venous link/return PAPVR will occur [11]. An earlier report on

TAPVC described four types of cardiovascular anomalies: supracardiac, infracardiac, cardiac, and mixed types [6]. A recent case of PAPVR describes multiple congenital anomalies, such as annular pancreas and patent umbilical vein with connection to left portal vein, in addition to cardiovascular anomalies [32].

Hypogenetic lung

Though infrequent, inherited underdevelopment of one or more pulmonic lobes is found in patients with CPVS [12]. Reports describe pulmonic agenesis, pulmonary aplasia, and pulmonary hypoplasia, wherein the air sac and bronchial tube are present, but the involved section is undersized [12]. Collectively, these three abnormalities are mentioned as hypogenetic pulmonic disorder [12, 26]. In the case reported here, we observed that the right lung was present, but the three lobes were extremely hypoplastic; as in most cases of hypogenetic lung, the trachea was normal. The case presented here is the form with pulmonary hypoplasia: air sac and bronchial tube are present, but the involved part is small, as described. Hypogenetic lung is much more common on the right side [5] and involvement of multiple lobes frequently occurs [26]. The large percentage of patients with lone hypogenetic lung exhibit no signs or symptoms [17, 19]; nevertheless, when other cardiac and vascular structures are affected, as in the case reported here, surgical intervention is warranted.

Partial anomalous pulmonary venous return can roughly be typed into two forms: one that occurs in association with ASD and the other in association with hypogenetic lung [7]. The second form of PAPVR is a part of CPVS. In PAPVR, an atypical vein empties a portion or the whole hypogenetic lung [5, 12, 26]. The configuration of the anomalous vein (right pulmonary vein draining into the IVC) on anterior-posterior thoracic radiograms has been equated to a Turkish sword or scimitar [2] and so "scimitar syndrome" describes hypogenetic lung and PAPVR presenting together [2]. While hypogenetic lung might occur on the left, scimitar syndrome occurs nearly exclusively on the right-side [2, 5, 30]. In virtually all occurrences, the anomalous vein is a pulmonic vein [2, 5, 12].

In scimitar syndrome, the abnormal pulmonic vein most often empties into the IVC inferior to the right hemidiaphragm [2, 12]; rarely, PAPVR empties into the suprahepatic segment of the IVC [5, 13, 29]. When PAPVR empties into a general vein or to the right atrium, there is a left-to-right bypass; in general, patients experience no symptoms unless the shunt is 2:1 or larger [13]. In the case presented here, the right superior pulmonary vein drained into the SVC. Because the donor had both hypogenetic lung and PAPVR, we concluded that the person experienced a "scimitar" lung.

Accessory diaphragm

An accessory diaphragm is a thin fibromuscular membrane perhaps associated with partial descent of the septum transversum [8]. This anomaly is predominantly a right-sided condition. Why this occurs is unclear, but it was acceptably explained that the timing difference in growth of the two lung buds is probably involved [8]. In other reports, aplasia or a certain amount of pulmonic hypoplasia was apparent on the involved side; this also occurred in the case presented here (Fig. 3). The accessory diaphragm thus separates the pleural cavity on the right side into a part that traps part or all of the right middle or a part that traps the lower lobes beneath it [5]; this is also seen in the case we present (Fig. 3). If the lung trapped beneath the accessory diaphragm is aerated, as seen in the case presented here, it will move with respiration [12]. Most interestingly, we observed an accessory diaphragm separating the fused anterior and lateral basal segments that could be aerated using a pump, thus providing evidence that it was functional. Furthermore, the 'sequestered' segments were supplied by a branch of the pulmonary artery.

In the case reported here, we identified a dilated coronary sinus entering the right atrium adjacent to the ASD and the ostium of coronary sinus noticeably lacked the typical valve-like structure. Widening of the coronary sinus resulted from an increased volume of flow of blood into the right atrium through anomalous communication of the right pulmonary vein with SVC (PAPVR) and the presence of left-to-right shunt through the ASD. We speculate that the excessive volume of blood shunted into the right atrium and an increase in intra-atrial pressure associated altered haemodynamics caused the observed widened coronary sinus. In 2017, the coexistence of ASD and 'unroofed coronary sinus' with TAPVC in a 16-year-old female was reported [27]. It seems that dilated coronary sinus is seen in association with ASD [27].

Aetiology

Partial anomalous pulmonary venous return is a congenital cardiac defect with no known cause; it

may have a multifactorial origin, including a genetic component. No evidence has implicated common teratogens (e.g., drugs, infections) in the genesis of PAPVC. No evidence for a genetic predisposition has been reported [31], though it is reportedly seen with Turner syndrome [36].

In addition to the cardiopulmonary changes, we also observed splenomegaly (the spleen was 28 cm long, 16 cm wide, 6 cm in thickness, and weighed 1,856 g) and greatly enlarged and coiled splenic vasculatures, probably due to vascular or macrophage sequestration (Fig. 4). The size of a normal, healthy spleen can vary considerably from person to person. Women tend to have smaller spleens than men and taller people tend to have larger spleens than shorter people. In general, an adult spleen is about 12.7 cm long, 7.6 cm wide, 3.8 cm thick, and weighs about 6 ounces. It is hard to even speculate a cause for the splenomegaly, since the portal system, liver and pancreas were normal.

CONCLUSIONS

In conclusion, we present a case of hypogenetic right lung with other unusual features such as partial anomalous pulmonary venous return, pseudo horseshoe lung, atrial septal defect, and accessory diaphragm. The right superior pulmonary vein returned blood to the SVC instead of the left atrium. Considering the fact that the donor has both hypogenetic lung and PAPVR, we conclude that this represents a "scimitar" lung. To sustain life, some of these defects were corrected in childhood, which allowed the donor to live for 71 years.

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Conflict of interest: None declared

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