Cardiology in the Young

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Surgical treatment of anomalous right upper lobe pulmonary vein obstruction caused by compression between pulmonary artery and trachea: a case report

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Brief Report

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Abstract

The normal anatomical course of right upper lobe pulmonary vein involves drainage anteriorly to the pulmonary artery, ultimately reaching the left atrium. However, anomalies can occur with the most common variation involving the convergence of the right upper lobe pulmonary vein with the superior vena cava. In a rare pulmonary vascular malformation, the anomalous right upper lobe pulmonary vein takes a path between the right pulmonary artery and right main bronchus [1]. During a clinical consultation, a patient presented in our hospital with this specific anomalous right upper lobe pulmonary vein, along with an atrial septal defect and a patent ductus arteriosus. As a consequence of this aberrant positioning, the right upper lobe pulmonary vein was compressed between the pulmonary artery and trachea, leading to pulmonary vein obstruction. Thus, a successful pulmonary vein replantation was performed to correct the congenital malformation.

Pulmonary vein obstruction can arise from diverse aetiological factors encompassing both congenital influences, such as genetics and embryology, as well as acquired factors, such as traction, compression, and surgical interventions. pulmonary vein obstruction can manifest in various forms, including single and multiple pulmonary vein obstructions. An exceptionally rare pulmonary vein obstruction variant involves the compression of the right upper lobe pulmonary vein by the pulmonary artery and trachea.[1] In the current study, we present the diagnostic approach and surgical management of such an anomalous right upper lobe pulmonary vein, complicated by the presence of an atrial septal defect and a patent ductus arteriosus.

Case presentation

A 24-year-old female was admitted to our hospital with a heart murmur. Upon physical examination (oxygen saturation -95%), a systolic murmur was detected in the second intercostal space at the left edge of the sternum. Pre-operative echocardiography revealed a secundum atrial septal defect (32 mm in size), along with mild tricuspid regurgitation, mild pulmonary valve regurgitation, pulmonary hypertension, and dilation of the main, left, and right pulmonary arteries. A chest computed tomographic angiogram confirmed the presence of a secundum atrial septal defect, patent ductus arteriosus (2 mm in size), pulmonary hypertension, and obviously compressed right upper lobe pulmonary vein by the right pulmonary artery and right main bronchus. Prior to oxygen inhalation, the pre-operative cardiac catheterisation determined a shunt fraction (Qp/Qs) of 1.17, mean pulmonary artery pressure of 35 mmHg, and pulmonary vascular resistance of 4.94 wood units. Subsequently, after oxygen inhalation, mean pulmonary artery pressure reduced to 32 mmHg, Qp/Qs increased to 3.32, and pulmonary vascular resistance decreased to 1.58 wood units.

A standard median sternotomy was performed to access the surgical site. The pericardial cavity was opened to expose the heart. Enlargement of the right atrium and main, left, and right pulmonary arteries was noted during the surgical procedure. Firstly, the patent ductus arteriosus was identified, separated, and ligated using 5-0 polypropylene sutures. Cardiopulmonary bypass was established with the ascending aorta and bicaval drainage. During cardiopulmonary bypass, cut the right pleura, collapse the right lung, right upper lobe pulmonary vein and right pulmonary artery were fully dissected from the right chest hilum. To facilitate the passage of the right upper lobe pulmonary vein, the pericardium was incised at the right pulmonary hilum. Next, the ascending aorta was cross-clamped to induce cardiac arrest and an incision was made in the right atrium. The anomalous right upper lobe pulmonary vein was dissected near its junction with the left atrium within the pericardial, then carefully excised and positioned in

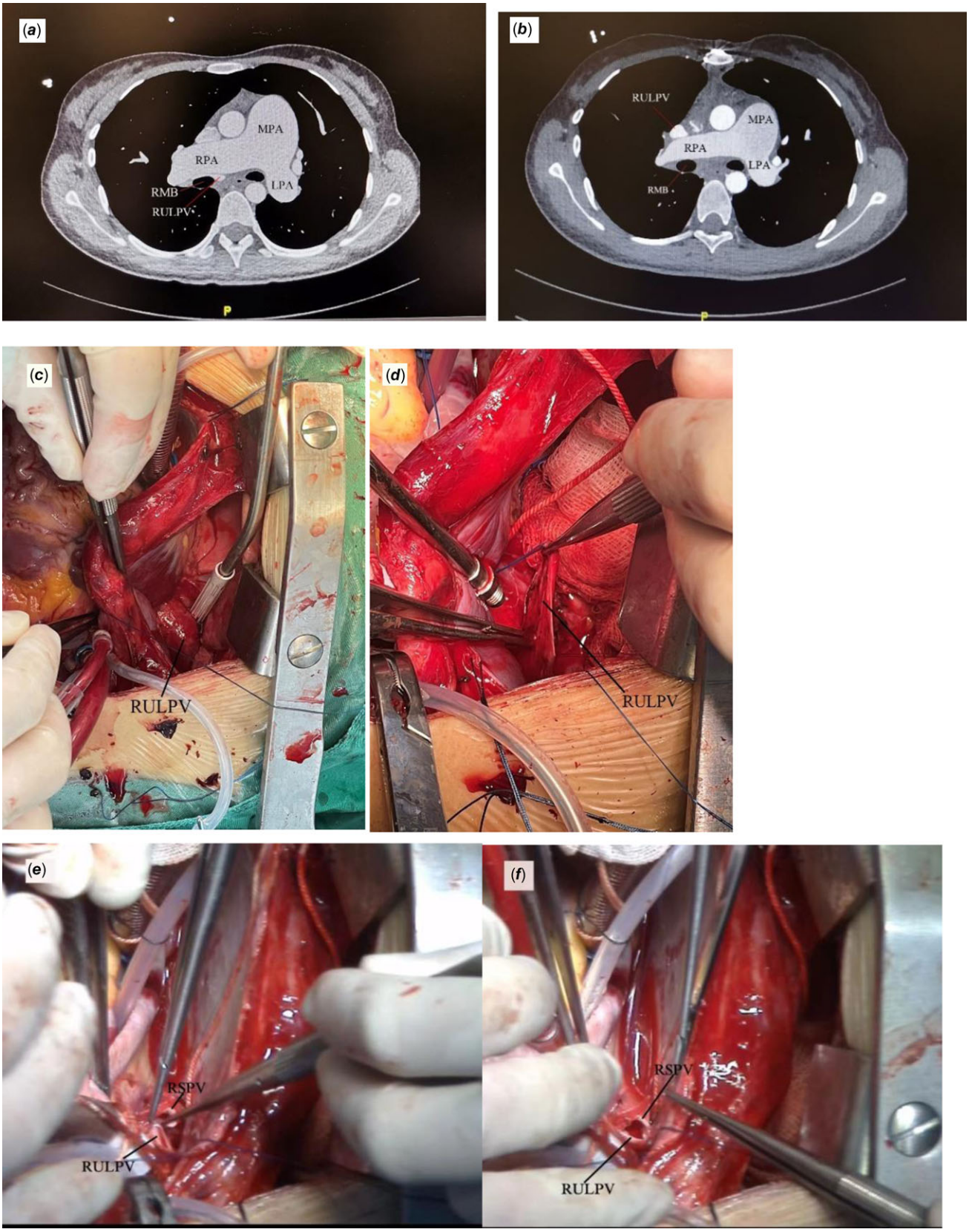

Figure 1. (***a***) Right upper lobe pulmonary vein before the operation was compressed by the pulmonary artery and right main bronchus, with signs of pulmonary hypertension and enlargement of left, right, and main pulmonary arteries. (***b***) Normal right upper lobe pulmonary vein after operation, with significantly smaller left, right, and main pulmonary arterial diameters compared to pre-operative measurements. (***c***) Right upper lobe pulmonary vein was fully separated from the pulmonary hilum. (*d*) rRight upper lobe pulmonary vein was incised and positioned in front of the right pulmonary artery. (***e*** and ***f***) Right upper lobe pulmonary vein was anastomosed with right superior pulmonary vein. Abbreviations: MPA, main pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery; RULPV, right upper lobe pulmonary vein; RSPV, right superior pulmonary vein; RMB, right main bronchus.

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front of the right pulmonary artery. Using 6-0 polypropylene stitches, anastomosis of the severed right upper lobe pulmonary vein was performed in situ. The anastomosis allowed for smooth passage of an outflow tract bougie (13#) from the atrial septal defect through the right upper lobe pulmonary vein. The atrial septal defect was subsequently closed in the standard manner using an autologous pericardial patch. Concurrently, tricuspid valve valvuloplasty was performed due to the enlargement of the tricuspid valve ring. Once the heart spontaneously resumed normal function, the incision in the right atrium was closed and the patient was successfully weaned off cardiopulmonary bypass.

The patient was discharged on post-operative day 6. Subsequent post-operative computed tomographic angiogram revealed unobstructed drainage of the right upper lobe pulmonary vein from the pulmonary artery to the left atrium. Additionally, there was a significant reduction in the diameter of the left and right pulmonary arteries compared to pre-operative measurements. Post-operative echocardiography demonstrated normal flow velocity in the right upper lobe pulmonary vein, with no obstruction, no residual atrial septum leakage, no tricuspid regurgitation, and successful patent ductus arteriosus closure. Pulmonary arterial pressure also showed a marked decrease, and the electrocardiogram was normal. Follow-up evaluation was performed three months after surgery in the outpatient department. The patient exhibited good health, and echocardiography demonstrated normal flow velocity in the right upper lobe pulmonary vein, indicating continued positive progress (Fig. 1).

Discussion

The anatomical structure of the right pulmonary veins, particularly the right superior pulmonary vein, is known to display significant variability. One frequently observed anomaly involves the right upper lobe pulmonary vein draining directly into either the right atrium or the superior vena cava.[2] The right upper lobe pulmonary vein may also drain directly into the left atrium, but with pulmonary vein branches crossing behind the intermediate bronchus, right upper lobe vein posterior to the bronchus intermedius, and right isolated superior posterior branch.[3] To the best of our knowledge, only four cases have been reported where the right upper lobe pulmonary vein anomalously resides between the right pulmonary artery and right main bronchus,[4] with none reporting simultaneous treatment of this type of anomalous right upper lobe pulmonary vein during cardiac surgery.

The embryological aetiology of this type of anomalous right upper lobe pulmonary vein may stem from the atypical positioning of the pulmonary artery and pulmonary vein during the formation of the pulmonary bud. The positioning of the pulmonary vein behind the pulmonary artery can occur alone or with other CHDs, although often without obvious clinical symptoms. Consequently, many patients may be misdiagnosed with primary pulmonary hypertension without further examination. Therefore, a pulmonary vessel computed tomographic angiogram should be performed when pulmonary hypertension caused by pulmonary vein obstruction is suspected[5] In the present case, the patient exhibited pulmonary hypertension inconsistent with an atrial septal defect. In patients with CHD and pulmonary hypertension, it can be difficult to identify the cause of pulmonary hypertension, even for highly experienced echocardiologists. Therefore, computed tomographic angiogram examination should be performed routinely to exclude pulmonary vein disease.

In accordance with the 2020 ESC guidelines for adult CHD,[6] surgical intervention is recommended for large atrial septal defects demonstrating signs of right ventricular volume overload and pulmonary vascular resistance below 3 wood units after oxygen inhalation. In this instance, informed by the type, cause, and location of the pulmonary vein obstruction, simultaneous replantation of the anomalous right upper lobe pulmonary vein was undertaken in order to alleviate the pulmonary vein obstruction.

The surgical approach involved the dissection of the right upper lobe pulmonary vein, followed by its reorientation anterior to the right pulmonary artery and in situ anastomosis of the transected right upper lobe pulmonary vein. This operative technique has two primary advantages. Firstly, it optimally utilises the pleural and pericardial cavities, ensuring a clear surgical field and ease of operation without increasing procedural complexity. Secondly, the in situ anastomotic position is subject to minimal tension, contributing to a favourable prognostic outcome.

In summary, this case report provides a surgical approach tailored to this specific type of anomalous right upper lobe pulmonary vein. While short-term post-operative outcomes were satisfactory, the long-term effects require further monitoring and evaluation.

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Competing interests. The authors declare no competing interests.

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