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REVIEW

Unilateral absence of pulmonary artery: Pathophysiology, symptoms, diagnosis and current treatment

Atrésie unilatérale de l'artère pulmonaire : physiopathologie, symptômes, diagnostic et traitement

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Summary Unilateral absence of pulmonary artery (UAPA) is a rare malformation that can present as an isolated lesion or may be associated with other congenital heart defects. UAPA is often associated with other congenital cardiovascular anomalies, such as tetralogy of Fallot, atrial septal defect, coarctation of aorta, right aortic arch, truncus arteriosus and pulmonary atresia. Diagnosis of UAPA is very difficult and is based on taking a complete medical history, physical examination and imaging examinations. Clinical symptoms include exercise intolerance, haemoptysis and recurrent respiratory infections. Adult patients with UAPA are often asymptomatic. There is no consensus regarding the treatment for UAPA. The therapeutic approach should be based on symptoms of the patient, pulmonary artery anatomy and associated

Abbreviations: CT, Computed tomography; MRA, Magnetic resonance angiography; MRI, Magnetic resonance imaging; PA, Pulmonary artery; PHT, Pulmonary hypertension; UAPA, Unilateral absence of pulmonary artery.

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aortopulmonary collaterals. Treatment options for these patients include partial or total pneumonectomy, closure of selected collateral arteries not solely responsible for pulmonary blood flow or a primary versus staged pulmonary artery anastomosis. This review summarizes pathophysiology, symptomatology and current diagnosis and treatment of this disease.

MOTS CLÉS
Cardiopathie congénitale ;
Absence unilatérale de l'artère pulmonaire ;
Pathophysiologie ;
Symptômes

Résumé L'absence unilatérale de l'artère pulmonaire est une malformation rare dont le mode de présentation peut être unique, isolée ou associée à d'autres cardiopathies congénitales. Elle est souvent associée en effet à la tétralogie de Fallot, la communication interauriculaire, la coarctation aortique, l'arche aortique droit, la persistance du canal artériel ou l'atrésie pulmonaire. Le diagnostic de l'absence unilatérale de l'artère pulmonaire est difficile, fondé un interrogatoire complet, un examen physique et clinique exhaustif. La symptomatologie inclut la dyspnée à l'effort, l'hémoptysie et les infections respiratoires récurrentes. Les patients adultes porteurs de cette pathologie sont souvent asymptomatiques. Il n'y a pas de consensus concernant le traitement de l'absence unilatérale de l'artère pulmonaire. L'approche thérapeutique doit être orientée par la symptomatologie décrite par le patient, l'anatomie artérielle pulmonaire et associée aux collatérales aorto-pulmonaires mises en évidence. Les options thérapeutiques incluent une pneumonectomie partielle ou totale sur l'occlusion sélective des artères collatérales et pas seulement responsables du flux artériel pulmonaire ou encore une anastomose réglée de l'artère pulmonaire. Cette revue résume la physiopathologie, la symptomatologie et les modalités diagnostiques et thérapeutiques de cette affection rare.

Background

Unilateral absence of pulmonary artery (UAPA) is a very rare congenital cardiovascular malformation. It was first described by Frentzel in 1868; in his initial paper, Frentzel reported that 30% of patients with this finding could remain asymptomatic until adult life [1,2].

Pool et al. [3] reviewed 32 cases before 1962, whereas Shakibi et al. [4] studied 47 cases from 1962 to 1976. Ten Harkel et al. [5] added a review of 107 cases from 1978 to 2000. The prevalence of isolated UAPA without associated cardiac anomalies ranged from 1 in 200,000 to 1 in 300,000 adults [6,7]. In 2011, Bockeria et al. found 352 cases of UAPA in the world literature. In 237 of these cases, UAPA was associated with congenital heart defects [8].

Most patients who have no associated cardiac anomalies have only minor or absent symptoms and survive into adulthood. Isolated UAPA without other cardiac anomalies is rare. Adult patients with UAPA are often asymptomatic and therefore undiagnosed. They present with exercise intolerance, haemoptysis or are incidentally detected during chest radiography.

There is no consensus on treatment of UAPA. The choice of treatment is based on symptoms of the patient, pulmonary artery (PA) anatomy and associated aortopulmonary collaterals, associated cardiovascular anomalies and pulmonary hypertension (PHT).

In this review, we summarize the pathophysiology, symptomatology, diagnosis and current treatment of UAPA.

Pathophysiology and embryogenesis of unilateral absence of pulmonary artery

The embryologic explanation for the origin of the absent PA is believed to be as follows. The intrapulmonary pulmonary arteries arise from the lung buds and the extrapulmonary pulmonary arteries arise from the proximal portion of the sixth aortic arch. The main PA is derived from the truncoaortic sac. The ductus arteriosus, which forms from the distal portion of the sixth arches, connects to the primitive dorsal aorta, which becomes the underside of the aortic arch ipsilateral to the arch or the base of the innominate artery contralateral to the arch. An absent PA is caused by the involution of the proximal sixth aortic arch and persistence of the connection of the intrapulmonary PA to the distal sixth aortic arch. It has been pointed out that all reported cases of absent PA with satisfactory angiographical, surgical or autopsy documentation had a ductus arteriosus or ligamentum ipsilateral to the absent PA [2,9]. The preferred terms for this condition have thus been suggested to be ''PA proximal interruption'', ''non-confluent PA'' or ''ductal origin of the distal PA'' [10].

If the ductus arteriosus closes after birth, the ipsilateral intrapulmonary PA will lose its source of blood supply and diminish in size and thus will not be visible with imaging [2].

In early embryonic development, transient systemic-to-pulmonary collateral arteries may arise during two extended periods. During maldevelopment of the pulmonary outflow tract, these transient connections may persist as systemic-to-pulmonary collateral arteries.

The timing and extent of pulmonary outflow tract maldevelopment may determine the origin and distribution of the collateral arteries. When pulmonary obstruction occurs at a very late stage of foetal development or after birth, bronchial arteries can develop into systemic-to-pulmonary collateral arteries [11]. Fadel et al. [12] showed that PA occlusion stimulates angiogenesis in the systemic circulation of the ipsilateral lung and increases systemic-to-pulmonary blood flow.

After a period of pulmonary occlusion, revascularization normalizes the systemic blood flow to the lung and induces

a partial loss of collateral vessels. It has been shown that, in congenital heart disease, certain aortopulmonary collateral arteries have a marked histological similarity to the ductus arteriosus [13].

Collaterals to the affected lung usually arise from bronchial arteries [7], but have also been documented to arise from intercostal, subdiaphragmatic, subclavian [5] and even coronary arteries [14]. It is argued that these connections can persist in human beings as aortopulmonary collateral vessels, in certain cases with abnormalities in the pulmonary part of the cardiac outflow tract [13].

Clinical symptoms and complications of unilateral absence of pulmonary artery

UAPA is twice as common on the right side. According to a study by Bockeria et al. [6] among 182 patients with isolated UAPA, the right PA was absent in 60% of cases (109/182) [8]. It is, however, noteworthy that about 80% of the reported cases involving the left PA have been associated with other congenital cardiovascular anomalies, such as tetralogy of Fallot, atrial septal defect, coarctation of aorta, right aortic arch, truncus arteriosus, patent ductus arteriosus and pulmonary atresia [5,15]. Two types of presentations are described. The first presentation is the one seen in infants, where they usually present with congestive cardiac failure and PHT [4,16]. The other presentation is in older patients, who are often asymptomatic; they present with exercise intolerance (18—40%), haemoptysis (20%) or are incidentally detected during chest radiography [4,16]. A literature review by Ten Harkel et al. [4] found that PHT was present in 44%, haemoptysis in 20%, recurrent pulmonary infections in 37% and limited exercise tolerance in 40% of patients with isolated UAPA. The prognosis depends on associated cardiovascular anomalies and the degree of PHT. Overall mortality in this series was 7% [5]. Recurrent pulmonary infections, decreased exercise tolerance and mild dyspnoea during exertion are the most common symptoms [5]. The aetiology of recurrent infections observed in patients with UAPA is likely to be multifactorial. Lack of arterial blood flow to the affected lung may result in poor delivery of inflammatory cells to sites of inflammation and impaired ciliary function. In addition, poor blood flow to the affected lung may result in alveolar hypocapnia, leading to secondary bronchoconstriction and mucous trapping. Chronic infection can lead to bronchiectasis in some patients [7,17]. Haemoptysis in patients with UAPA is caused by excessive collateral circulation [18]. Haemoptysis may be self-limiting for many years [19], but can also lead to massive pulmonary haemorrhage and death [20].

The incidence of PHT among the patients with unilateral absence of PA has been reported to be between 19% and 44% in different case series [3—5].

PHT may result from blood flow directed away from the absent PA to the remaining PA. Increased blood flow in the contralateral PA leads to shear stress within the endothelium, with subsequent release of vasoconstrictive compounds, such as endothelin. Chronic vasoconstriction of the pulmonary arterioles may lead to remodelling, resulting in increased resistance of the pulmonary vasculature and PHT.

Infants with isolated absence of the one of the PAs frequently present with PHT [4,16]. Pool et al. [3] documented medial hypertrophy of the PA of the normal lung (opposite to the affected side) in 53% of 17 patients. The affected lung (lacking a PA) was free of these changes in 83% of cases. Haworth et al. [21,22] described the changes in the right and left PA after ligation of the left PA and ductus arteriosus in an animal model (14 newborn pigs). In the right lung, pulmonary arterial pressure and resistance fell to normal after birth; however, structurally, muscularity remained high in arteries less than 75 μm in diameter. The left PA and branches were small, with a disorganized elastin wall structure, although the muscularity of the arteries in the left side was reduced. The mean PA pressure was elevated (20—35 mmHg), along with features of hypertrophied right ventricle [21,22]. Other reasons postulated for PHT were insufficient elasticity of the pulmonary vascular bed of the normal side receiving full cardiac output and abnormal response to vasoconstrictor [23,24]. Pool et al. [3] opined that those patients with isolated unilateral absence of PA who develop PHT generally do so at an early age and die from right heart failure. However, if PHT does not develop at an early age, it is unlikely that it will develop later.

Diagnosis of unilateral absence of pulmonary artery

Diagnosis of UAPA is very difficult and is based on taking a complete medical history, physical examination and imaging examinations. A high index of suspicion is needed to make the diagnosis. In infancy, the signs can be subtle and can be easily missed [6,9,16]. Examination may reveal an asymmetrical chest with abnormal breath sounds in the affected side. There may be a systolic ejection murmur across the pulmonary outflow tract [25].

The electrocardiogram is usually normal in patients with uncomplicated isolated absence of PA (without PHT), whereas it shows right ventricular dominance in cases associated with PHT [5]. The chest radiograph of patients with UAPA typically shows asymmetric lung fields (Fig. 1), with an ipsilateral small hemithorax holding a hyperlucent lung. The mediastinum will be shifted towards the affected side and the hilar vasculature on that side will be absent or greatly diminished. The ipsilateral hemidiaphragm may be elevated [6,17]. When suspicious findings are noted on a chest radiograph, the diagnosis of UAPA can be made definitively by computed tomography (CT), magnetic resonance imaging (MRI) or transthoracic echocardiography. On cross-sectional imaging, the absent PA will typically terminate within 1 cm of its expected origin from the main PA (Fig. 2). Other findings that may be noted on CT or MRI include intact peripheral branches of the PA, variable collateral circulation, mosaic parenchymal changes and bronchiectasis secondary to recurrent infections. In addition to depicting static morphological structures, magnetic resonance angiography (MRA) with contrast also provides real-time assessment of the haemodynamic status [6,7,17]. Transthoracic echocardiography is a good tool for establishing the diagnosis, excluding any other cardiac or major vessel abnormalities and evaluating the presence of

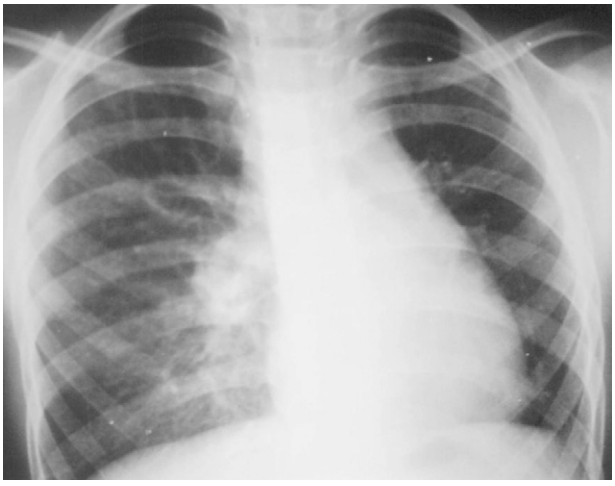

Figure 1. Chest X-ray showing a small left hemithorax, hyperinflation of the right lung, deviation of the trachea and mediastinum to the left and an engorged vascular shadow in the right hilum. The volume and vascularity of the left lung appear to be diminished. From Kruzliak et al. [45].

associated PHT. Cases without PHT can be further examined by echocardiography to detected early signs of this condition.

Angiography remains the gold standard for diagnosis of PA agenesis. Currently, with the development of CT, MRI and MRA techniques, it is rarely performed unless embolization is indicated for massive haemoptysis [5,17,26]. Pulmonary venous wedge angiography is particularly useful in delineating the presence of an ipsilateral hilar PA and intrapulmonary vessels, which is required before revascularization [1,9]. Cases without PHT can be followed with echocardiography to detect early signs of the same. Ventilation perfusion scanning is not necessary for the diagnosis of UAPA; if done, it shows normal or diffusely diminished Xenon-127 uptake during the wash-in equilibrium phase, coupled with absent or greatly diminished perfusion in the affected lung [1].

Unilateral absence of pulmonary artery treatment

There is no consensus regarding the treatment for isolated absence of PA. The therapeutic approach for isolated absence of PA should be based on symptoms of the patient, PA anatomy, associated cardiovascular anomalies and aortopulmonary collaterals and PHT. No treatment is required in patients without any evidence of cardiopulmonary dysfunction (as seen in adults with incidental detection); they should be followed up on a regular basis [4].

Most agree that treatment should be reserved for the small number of patients with haemoptysis, recurrent lower respiratory tract infection or PHT. Treatment options for these patients include partial or total pneumonectomy, closure of selected collateral arteries not solely responsible for pulmonary blood flow or a primary versus staged PA anastomosis [27—30]. Others advocate an early search for the occult PA in all patients, regardless of symptoms, followed by a staged repair to promote distal PA growth and lung development. Early re-establishment of pulmonary blood flow, either primarily or after a shunt procedure, may allow

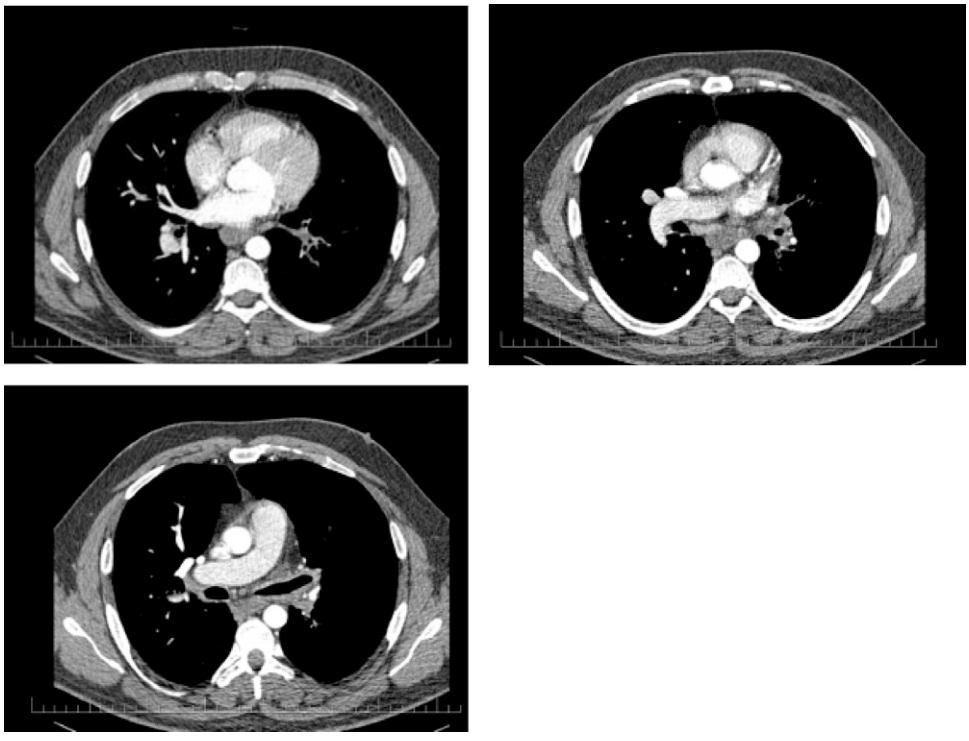

Figure 2. Computed tomography findings of pulmonary artery trunk enlargement and the absence of the left pulmonary artery with lower vascularity in the left lobe of the lung and light displacement of the heart and mediastinum to the left. From Kruzliak et al. [45].

the affected lung to develop more normally and improve PHT [31].

The surgical approaches have been either the creation of an aortopulmonary shunt [32] or connection of the affected PA to the main PA [2,4]. The provision of adequate antegrade blood flow to the unilateral absent PA before hypoplasia is established may increase the probability of long-term patency [9].

If the intrapulmonary arteries are well developed, a primary anastomosis with the central PA can be made. If the intrapulmonary branches are small, a modified Blalock-Taussig shunt can be used; this allows better growth of the affected intrapulmonary arteries before the segments are connected with the main PA during subsequent surgery [2,33]. Welch et al. [2] reported two cases of direct anastomosis of right PA to the main PA under the ascending aorta in a neonate and a 3-month-old infant. Direct anastomosis could be possible in the neonatal period or early infancy and should be recommended if possible [2]. Other surgical options are an interposition tube graft with autologous pericardium or prosthetic material (Dacron/GORE-TEX) and mobilization with end-to-side anastomosis of the affected artery to the main PA [2]. Because a size mismatch between the graft and native PA develops as the child grows, follow-up surgery for revision and/or catheter intervention can be anticipated. It has been reported that reconstruction using the autologous tissue or autologous pericardium may have potential to grow or could at least dilate. The major concern with these techniques is that the pedicled part of the pericardium is sutured and it is caused by ischemic shrinkage or growth disturbance [34]. Although reconstruction with pedicled autologous pericardium is reported to be useful to prevent late stenosis, long-term results are unclear [35]. Moreno-Cabral et al. [27] reported a reconstruction case using an autologous pericardial tube in front of the superior vena cava and ascending aorta. A postoperative pulmonary angiogram 4 years later showed a patent new right PA that appeared to have grown with the child; the anastomotic site to the main PA appeared stenotic, probably due to compression of the ascending aorta. Krammoh et al. [36] described their two-stage therapeutic approach in three patients. The first stage was PDA stenting, followed by surgical anastomosis in the second stage. The patients required anticoagulation therapy after the ductal stenting. The authors reported good outcome in three cases during midterm follow-up. Another report described a 21-month-old child with PHT and congestive heart failure in whom a hilar artery was reconstructed with a conduit (GORE-TEX), leading to the normalization of pulmonary pressure and improvement of congestive heart failure. This condition remained stable during a follow-up period of 2 years [37].

Patients who have delayed diagnosis and delayed surgical interventions may develop irreversible hypoplasia and regression of the affected PA and have a less favourable outcome [38,39].

Binnetoglu et al. [40] described the case of a missing right PA associated with anomalous left pulmonary venous connection in a 4-month-old girl. In this clinical case, the ratio of pulmonary-to-systemic blood flow was calculated as 2.04 by Fick's method and the pulmonary vascular resistance was 5.14 Wood units. The pulmonary angiogram showed a dilated main PA, but the right PA was not visible. Although the pulmonary-to-systemic flow ratio was 2 to 1, a surgical procedure to correct the abnormal pulmonary venous drainage could not be performed due to the long distance between the vertical vein and the left atrium. Medical treatment with a dual endothelin receptor antagonist (bosentan) was considered, but the patient died of right heart failure within 2 months.

In older patients, revascularization is not encouraged or even feasible because the intrapulmonary arteries have been found to be severely narrowed or even completely obstructed by fibrosis [3,41].

Haemoptysis may be self-limiting over many years [19,42], but massive haemoptysis can be treated only by selective embolization of the systemic collaterals [43] or even pneumonectomy of the affected side [41,44]. Kruzliak et al. [45] reported the case of UAPA in an adult with self-limiting haemoptysis. Selective embolization is less invasive and may carry less procedural risk than a pneumonectomy; however, limitations include lack of expertise, technical difficulty for safe coil release and higher long-term recurrence rates (up to 25%) due to the extensive collateralization [46]. Furthermore embolization may take longer, potentially compromising patient safety in an acute scenario. The contribution of the affected lung to functional gas exchange is usually minimal, which negates a lung-preserving procedure. Embolization, however, should be considered in less fit patients or as an adjunct to surgery to reduce subsequent surgical blood loss by devascularizing the pleural space in preparation for a thoracotomy [47].

Bekoe et al. [48] reported the case of an adult with UAPA, who was treated with pneumonectomy followed by a good outcome. The authors recommended pneumonectomy for patients with recurrent haemoptysis caused by UAPA, because the lung affected by UAPA does not contribute to ventilation.

Severe infections may require lobectomy or pneumonectomy, and any pulmonary surgery in a patient with UAPA may be complicated by the presence of systemic collaterals [5].

For patients who are not considered suitable for revascularization or when PHT does not improve, therapeutic measures for PHT may be helpful. Pharmacotherapy for PHT is indicated and is based on endothelin receptor antagonists (e.g. bosentan) or parenteral prostacyclins. Long-term vasodilator therapy may improve survival. Sildenafil, calcium channel blockers and continuous intravenous infusion of prostacyclin have been tried with variable responses [5,49]. Infants who present with severe PHT represent the most difficult subset to treat. PHT in this group is not known to regress spontaneously [16]. The overall mortality across all the age groups is reported to be 7% and infants with severe PHT have poor outcome.

Conclusion

Isolated absence of right PA is a rare entity. In infancy, it presents with respiratory distress and severe PHT. Adult patients are often asymptomatic. PHT is a severe complication of UAPA and significant increases the mortality rate of this disease. Patients who have delayed diagnosis and delayed surgical interventions may develop irreversible hypoplasia and regression of the affected PA and have a

less favourable outcome. It is very important to establish an early diagnosis and provide adequate treatment for this disease.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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