A 3-month-old boy was admitted to Hainan Women and Children’s Medical Center on November 17, 2019, because of feeding difficulty, without a history of cyanosis, anemia, edema, oliguria, or chronic cardiac diseases, as well as a family history of cardiac diseases or other disorders. Physical examination revealed a systolic murmur of grade II at the left ventricular apex. Laboratory findings showed no abnormalities in either the serum creatine kinase or the serum creatine kinase isoenzyme and anemia, with arterial oxygen partial pressure (PaO₂) of 71 mmHg and arterial oxygen saturation (SaO₂) of 94%. Chest X-ray examination revealed a slightly thinner right lung texture than that of the left side (Fig. 1A). The electrocardiogram revealed no abnormalities. Color Doppler echocardiography (Fig. 1B) and electrocardiographic-gated computed tomography angiography (CTA) showed the absence of the right pulmonary artery and a starting segment of the collateral vessel that originated from the thoracic aorta (Fig. 1C and D). The subsequent cardiac catheterization and angiography indicated mild pulmonary hypertension (44/15 [24] mmHg), increased pulmonary vascular resistance (5.32 Wood U/m²), slightly elevated pulmonary capillary wedge pressure (20.5/10.5 [13.8] mmHg) (Fig. 2A), normal descending aorta pressure (72.7/49.8 [57.4] mmHg) (Fig. 2B), no image of the right pulmonary artery (Fig. 2C and D), and a collateral vessel originating from the thoracic aorta and zigzagged into the right lung (Fig. 2E and F). Pulmonary vein wedge angiography (the catheter was controlled to pass through the foramen ovale into the right upper pulmonary vein, and then a contrast agent was injected via the catheter retrograde to the “isolated” pulmonary artery) revealed an isolated right pulmonary artery with a starting segment diameter of approximately 3.75 mm (Fig. 2G and H). The patient was clinically stable for more than 2 years after direct anastomosis surgery between the “isolated” right pulmonary artery and the main pulmonary trunk.

Unilateral absence of pulmonary artery (UAPA) is a rare malformation with an estimated prevalence of 1 in 200,000 young adults that can present as an isolated lesion or may be associated with other congenital heart defects, such as truncus arteriosus, aortic coarctation, atrial septal defect, tetralogy of Fallot, right aortic arch, and pulmonary atresia. UAPA diagnosis is very difficult and is based on a complete medical history, physical examination, and imaging examinations. Recurrent respiratory infection was present in 35.4% of patients, hemoptysis in 41.5%, and exertional dyspnea in 41.5%. Early surgical repair of UAPA is necessary because it leads to contralateral pulmonary artery hypertension and ipsilateral pulmonary hypoplasia without treatment. Surgical strategies include direct anastomosis or interposition graft creation using flap techniques or prosthetic material. Preoperative pulmonary vascular anatomic and physiological data are important conditions to determine the surgical strategy. There is no blood flow entering into the “isolated” pulmonary artery...
Figure 1  (A) Chest X-ray examination showed a slightly thinner right lung texture than that of the left side (B) Color Doppler echocardiography showed an absence of the right pulmonary artery (C) and (D) CTA showed an absence of the right pulmonary artery, and a starting segment of collateral vessel originated from the thoracic aorta (white arrow).
Figure 2  (A) Plot waveform of the left pulmonary capillary wedge pressure (20.5/10.5 [13.8] mmHg) (B) Plot waveform of the descending aorta pressure (72.7/49.8 [57.4] mmHg) (C) and (D) Pulmonary artery trunk angiography showed no image of the right pulmonary artery (E) and (F) Descending aorta and selective vessel angiography showed a collateral vessel originated from the thoracic aorta and zigzagged into the right lung (G) and (H) Pulmonary vein wedge angiography revealed an isolated right pulmonary artery with a starting segment diameter of approximately 3.75 mm.
from the pulmonary artery trunk (no direct vessel access between them) in patients with UAPA, thereby making echocardiography, CTA, or conventional left and right cardiac angiography difficult. Pulmonary vein wedge angiography can reveal the “isolated” pulmonary artery, thereby facilitating the determination of surgical strategies. Therefore, pulmonary vein wedge angiography is important before choosing surgical strategies for UAPA.

**Author contributions**

Interpretation of data: D-F. Z., J-Z. Conception and design of the work: D-F. Z. Manuscript drafting: J-Z. Critical evaluation and revision of the manuscript: D-F. Z. All the authors have given their final approval of the version to be published.

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**Ethics approval and consent to participate**

This study was performed after permission from the Ethics Committee of Hainan Women and Children’s Medical Center (Approval number 2021—107), and informed written consent was got from the participating infants’ parents.

**Competing Interest**

The authors declare no conflict of interest.

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**References**


