Original Article

Surgical repair of unilateral absence of pulmonary artery in children with pulmonary hypertension: a single-center retrospective study

Xinjian Yan[1,2#], Jianzheng Cen[1,2#], Xiaokang Luo[1,2], Jimei Chen[1,2], Shusheng Wen[1,2], Jinlin Wu[1,2], Jian Zhuang[1,2]

[1]Department of Cardiovascular Surgery, Guangdong Cardiovascular Institute, Guangdong Provincial People's Hospital, Guangdong Academy of Medical Sciences, Guangzhou, China; [2]Guangdong Provincial Key Laboratory of South China Structural Heart Disease, Guangdong Provincial People's Hospital, Guangdong Academy of Medical Sciences, Guangzhou, China

Contributions: (I) Conception and design: X Yan, J Cen; (II) Administrative support: J Zhuang, J Chen, S Wen; (III) Provision of study materials or patients: J Cen, S Wen; (IV) Collection and assembly of data: X Yan, X Luo; (V) Data analysis and interpretation: X Luo, J Wu, X Yan; (VI) Manuscript writing: All authors; (VII) Final approval of manuscript: All authors.

[#]These authors contributed equally to this work and should be considered as co-first authors.

Correspondence to: Jian Zhuang, MD, PhD. No. 106 Zhongshan 2nd Road, Yuexiu District, Guangzhou 510080, China. Email: zhuangjian5413@tom.com.

Background: The early diagnosis of unilateral absence of pulmonary artery (UAPA) in children offers an opportunity for effective intervention. Due to the lack of clinical evidence, a consensus regarding surgical treatment has yet to be reported. The aim of this study is to evaluate the effectiveness and safety of pulmonary artery (PA) reconstruction with a "two-segment" technique to repair UAPA in patients with pulmonary hypertension.

Methods: Intraoperatively, the ligamentum arteriosum connecting the innominate artery and distal PA was dissected and occluded. A conduit created by fresh autologous pericardium formed the first "segment" of the neo-PA. The second "segment" was a Gore vascular graft with integrated rings anastomosed between the proximal end of the pericardial conduit and the main pulmonary artery (MPA).

Results: A total of five consecutive patients were included, and the absent PA was successfully reconstructed using the "two-segment" technique in all patients. Following revascularization, the direct measurement of the pressure in MPA during the operation showed that the average mean pulmonary artery pressure (mPAP) decreased from 31.3±16.0 to 16.8±4.2 mmHg (P=0.047). The average mPAP/radial mean arterial pressure (rMAP) ratio decreased from 0.59±0.27 preoperatively to 0.30±0.10 postoperatively (P=0.028). The mean follow-up period was 18.85±4.67 months. The median diameter of the reconstructed PA (pericardial segment) measured by transthoracic echocardiography (TTE) was 6.1 mm. One patient safely underwent a redo operation to repair relative stenosis in the neo-PA.

Conclusions: Early PA reconstruction may effectively alleviate pulmonary hypertension in children with UAPA. The "two-segment" technique is safe and can facilitate potential redo pulmonary arterioplasty. Anticoagulation and antiplatelet therapy, as well as frequent follow-up, is required after the operation.

Keywords: Congenital heart disease; unilateral absence of pulmonary artery (UAPA); pulmonary hypertension (PHT); surgical repair; two-segment technique

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Introduction

Unilateral absence of pulmonary artery (UAPA) is a rare congenital heart disease. It can occur as an isolated anomaly or in association with other congenital heart diseases, such as tetralogy of Fallot, aortic coarctation, or truncus arteriosus (1,2). To date, over 400 cases have been described in the literatures (2,3). Over 60% of the isolated UAPA were right side affected [absence of right pulmonary artery (RPA)] (2,4). The absence of a native pulmonary artery (PA) is caused by the malformation of the proximal sixth aortic arch during embryogenesis. A ductus arteriosus connecting to the hilar PA is usually found in most patients (5). Many of these patients develop pulmonary hypertension (PHT) after birth (6). Children presenting with PHT represent a difficult-to-treat subset of patients, and most of them die of PHT-induced right heart failure at an early age (7).

An early diagnosis of UAPA in children offers an opportunity for surgical interventions (8). Surgical repair could be performed by staged operations (palliative procedure first) or single-stage correction to reconstruct the absent PA. Palliative procedures usually complicate the subsequent corrective interventions (9), while the long-gap discontinuity between the hilar PA and the main pulmonary artery (MPA) increases the difficulty of single-stage correction (10). Due to the lack of clinical evidence, a consensus regarding surgical treatment has yet to be reported.

In this study, we adapted the single-stage correction to reconstruct the absent PA with "two-segment" technique in order to accomplish the physiological correction as much as possible. The "two-segment" technique was designed aiming to facilitate potential redo pulmonary arterioplasty based on our experience from other operations related to PA reconstruction, i.e., PA repair during pulmonary atresia with ventricular septal defect (PA/VSD). The outcomes of this technique were also evaluated. We present the following article in accordance with the STROBE reporting checklist (available at https://tp.amegroups.com/article/view/10.21037/tp-22-491/rc).

Methods

Patients

The study was a single-center, retrospective study. The records of children who were diagnosed with UAPA and PHT and underwent surgical repair via the "two-segment" technique in Guangdong Provincial People's Hospital from March 2019 to April 2021 were retrospectively reviewed. The necessary medical data of each patient were extracted from the institutional database. The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). The study was approved by the Research Ethics Committee of Guangdong Provincial People's Hospital, Guangdong Academy of Medical Sciences (approval No. KY-Q-2021-267-01). Informed consents were obtained from the legal guardians of the children.

Pulmonary vein wedge angiography (PVWA) was the first choice for UAPA diagnosis; however, for children with high-risk factors for PVWA, a contrast-enhanced computed tomography (CT) was performed for diagnosis. Chest X-ray, transthoracic echocardiography (TTE), and contrast-enhanced CT were routinely performed on each patient to detect the accompanied anomalies before surgery. The reconstruction of the absent PA was assessed by TTE and contrast-enhanced CT scan postoperatively. All patients were followed up at our outpatient clinic after discharge from the hospital. Physical examination, chest radiography, and TTE were performed during the follow-up.

Surgical technique

Figure 1 illustrates the schematic of the "two-segment" technique. The operation was performed on a beating heart with cardiopulmonary bypass at normothermia. Following median sternotomy, the thymus anterior to the pericardium was removed. Cardiopulmonary bypass was established by cavoatrial and aortic cannulation. The aorta, superior vena cava, innominate artery, MPA, and ligamentum arteriosum connecting the innominate artery and distal RPA were sufficiently mobilized.

The RPA was fully exposed at the right hilum. The ligamentum arteriosum connecting the innominate artery and distal RPA was dissected and occluded. A tubular conduit was created using fresh autologous pericardium to form the first "segment" of the neo-RPA to enlarge the anastomotic stoma. The size of the RPA stump was measured using Hegar dilators. The diameter of the pericardial conduit was based on the development of the RPA at the hilum. The distal end of the pericardial conduit was anastomosed to the hilar RPA under the superior vena cava.

The second "segment" of the neo-RPA was a Gore vascular graft with integrated rings (GORE® INTERING® Vascular Graft, Gore & Associates Inc. Newark, DE, USA) placed at the posterior side of the aorta to prevent the distortion and stenosis caused by the compression of the

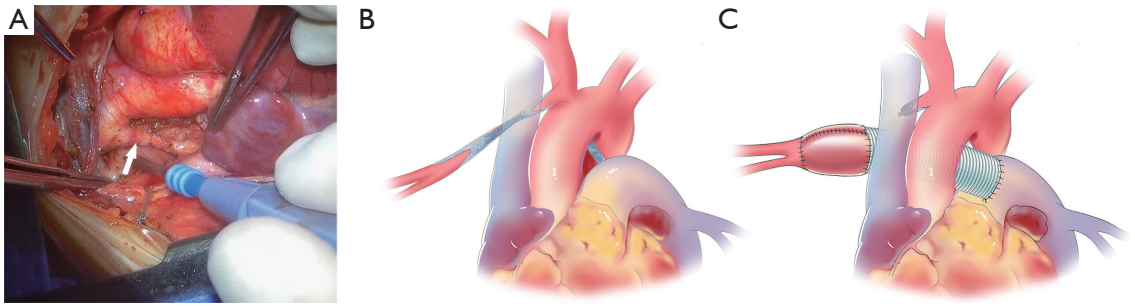
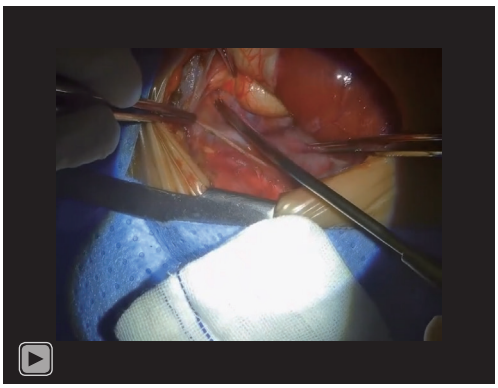


Figure 1 Schematic of the "two-segment" technique. (A) Intraoperative picture showing the repair of UAPA. The white arrow shows the aberrant diverticulum at the base of innominate artery. (B,C) Before and after the reconstruction of pulmonary artery using the "two-segment" technique. UAPA, unilateral absence of pulmonary artery.



Video 1 The video shows the steps of "two-segment" technique. Firstly, mobilize the ligamentum arteriosum connecting the innominate artery and the distal RPA. Then, dissect and occlude the ligamentum arteriosum connecting the innominate artery and the distal RPA. Next, use fresh autologous pericardium to create the first "segment" of neo-RPA to enlarge the anastomotic stoma under the superior vena cava. Finally, use a Gore vascular graft with integrated rings as the second segment and place it at the posterior side of the aorta. Anastomose the graft to the proximal end of the pericardial conduit and the MPA. RPA, right pulmonary artery; MPA, main pulmonary artery.

adjacent vessels. The blood flow of the MPA was partly blocked using a Cooley clamp. Next, the vascular graft was anastomosed to the proximal end of the pericardial conduit and the MPA. All anastomoses were conducted using 7-0 or 8-0 Prolene sutures. The patency of the neo-RPA was confirmed by intraoperative transesophageal echocardiography (TEE). *Video 1* displays the steps of the technique.

Postoperatively, the patients were routinely transferred to the intensive care unit (ICU) for observation. Heparin was used for bridging to warfarin during the ICU stay. Patients were asked to take warfarin (0.1–0.2 mg/kg/d) for 6 months and maintain an international normalized ratio (INR) of 1.8–2.5. After the initial 6 months, aspirin 3–5 mg/kg a day was prescribed instead of warfarin.

Outcome indexes and follow-up

Postoperative follow-up was performed periodically and the patency of the neo-RPA was evaluated by TTE and CT scan.

Clinical outcomes: (I) the Z-score for the MPA; (II) the direct measurement of the pressure in MPA before and after the PA reconstruction; (III) the radial artery pressure before and after the PA reconstruction; (IV) the mean pulmonary artery pressure (mPAP)/radial mean arterial pressure (rMAP) ratio; (V) diameters of conduits for reconstructing the PA based on the diameters of the PA measured at the hilum position; (VI) complications which were recorded during the follow-up.

Statistical analysis

Because of the small sample size, it is not necessary to test the normality of the variables and there is no need to do comparative tests. All the analyses are descriptive. Continuous data were analyzed using the Shapiro-Wilk test and Q-Q plot for normality. Normal continuous data were expressed as the mean ± standard deviation (SD). Categorical data were expressed as the frequency or percentage. The Student's *t*-test was applied for normal data. The Wilcoxon and Kruskal-Wallis tests were applied for non-normal data. All tests were two-sided and P values of ≤0.05 were considered statistically significant.

Table 1 Characteristics of the included patients

No.	Gender	Age	Body weight (kg)	Diagnosis	3D CT	Z-score for the main PA	Diameters of conduits (mm)	Preoperative artery pressure (mmHg)	Postoperative artery pressure (mmHg)	Duration of follow-up (months)	Postoperative complications
1	Male	3.3 m	5.0	UAPA, PFO, TI, PHT	Yes, diverticulum	3.76	Pericardial: 7; vascular graft: 6	RAP: 71/37; PAP: 71/41	RAP: 73/35; PAP: 30/16	23.8	Underwent a second operation to repair relative stenosis at 9 m postoperatively
2	Male	1.8 y	10.0	UAPA, TI, PHT, PDA	Yes, diverticulum	3.96	Pericardial: 6; vascular graft: 6	RAP: 58/39; PAP: 34/18	RAP: 87/53; PAP: 20/10	16.7	In-hospital thrombosis disappeared before discharge from the hospital
3	Male	1.4 m	4.8	UAPA, PFO, PHT	Yes, diverticulum	1.24	Pericardial: 6; vascular graft: 6	RAP: 63/39; PAP: 31/16	RAP: 68/46; PAP: 21/11	21.8	None
4	Male	2.7 y	14.5	UAPA, PHT, cardiac situs inversus	Yes, diverticulum	1.63	Pericardial: 5; vascular graft: 5	RAP: 83/50; PAP: 29/14	RAP: 82/48; PAP: 22/10	13.4	None
5	Female	13 d	2.78	UAPA, PFO, PHT	Yes, diverticulum	2.22	Pericardial: 6; vascular graft: 6	RAP: 57/31; PAP: 40/10	RAP: 69/41; PAP: 29/10	18.6	None

PA, pulmonary artery; UAPA, unilateral absence of pulmonary artery; m, months; y, years; d, days; PFO, patent foramen ovale; TI, tricuspid insufficiency; PHT, pulmonary hypertension; RAP, radial artery pressure; PAP, pulmonary artery pressure; PDA, patent ductus arteriosus; 3D CT, three-dimensional reconstruction of computed tomography.

All statistical analyses were performed using IBM SPSS Statistics 22.0 software (Armonk, New York, USA).

Results

Patient characteristics

A total of five consecutive patients were included (four males and one female). The ages of the patients at the time of operation ranged from 13 days to 2.75 years (median: 3 months 8 days), and their body weights ranged from 2.78 to 14.5 kg (median: 5.0 kg). Transcutaneous oxygen saturation was above 95% on admission in all patients, and the main symptom was cough. Cardiac malformations were detected by prenatal echocardiography in three patients. All patients were preoperatively diagnosed with PHT by TTE (three severe, one moderate, and one mild). An intraoperative assessment of pulmonary pressure was conducted by penetrating a catheter connected to a pressure sensor into the MPA. The mPAP was 31.3±16.0 mmHg (range, 20–55 mmHg), and the average rMAP was 49.6±8.0 mmHg (range, 40–60 mmHg) at the same time.

Three patients had accompanying patent foramen ovale, of whom one was a right to left shunt and two were bidirectional shunts. Two patients were associated with tricuspid regurgitation (*Table 1*). One patient had a history of surgical closure of patent ductus arteriosus (PDA).

The preoperative TTE showed that the unilateral PA was absent. Several small systemic to affected lung collaterals were also detected in each patient. Four patients were diagnosed with unilateral absence of the RPA, while the remaining patient was diagnosed with cardiac situs inversus accompanied by a unilateral absence of the left PA. The mean diameter of the MPA was 12.44±3.22 mm (range, 8.20–14.00 mm), with a mean Z score of 2.56 (range, 1.24–3.96). The mean preoperative left ventricular ejection fraction (LVEF) was 73.2%±6.53%, and the mean left ventricle fractional shortening (FS) was 40.2%±4.87%.

The X-rays showed a hyperlucent lung in the ipsilateral lung fields. The discontinuity and absence of intrapericardial PA were confirmed by PVWA or three-dimensional (3D) CT reconstruction. Two patients underwent PVWA. The angiogram clearly showed the right pulmonary vein and blind end of the native RPA connecting other right lung vessels. Notably, after 3D CT reconstruction, an aberrant diverticulum was found at the base of the innominate artery in every patient (*Figure 2*). The normally developed pulmonary vasculature and the stump of the absent PA were clearly demonstrated. The discontinuity of the RPA was consistent with the PVWA findings.

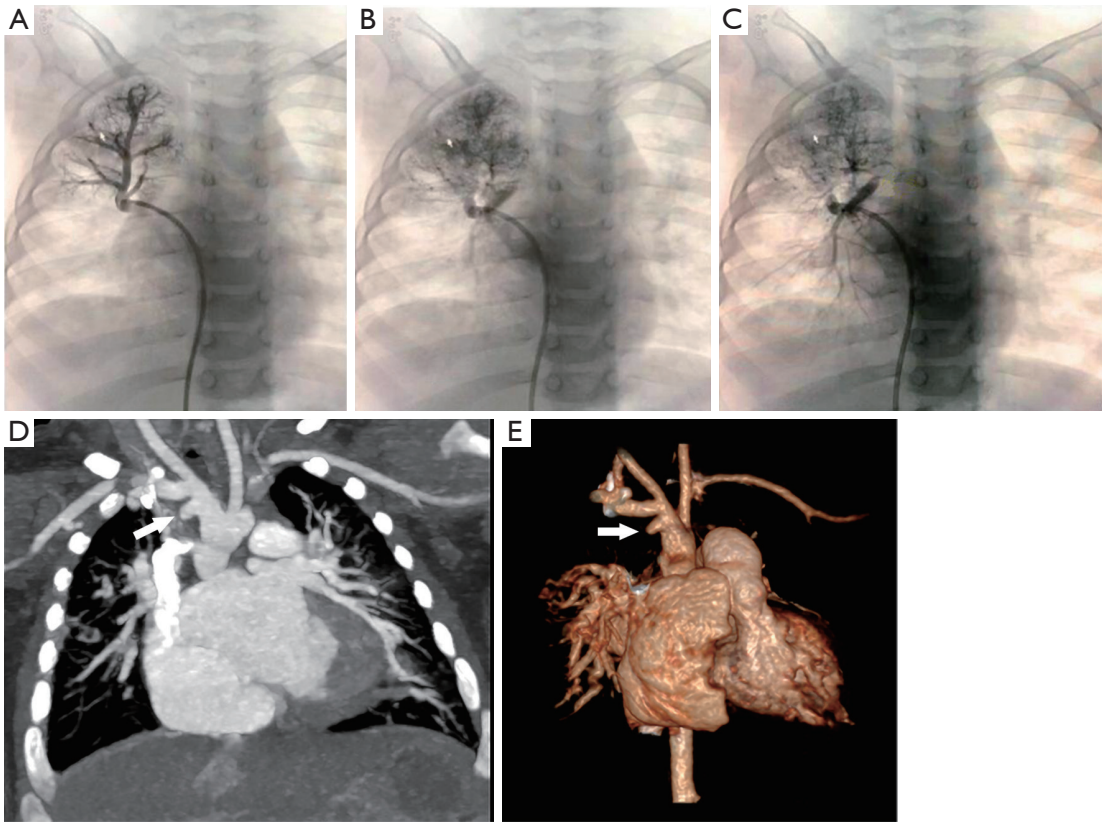


Figure 2 Preoperative imaging of the patients. (A-C) The three phases of preoperative PVWA shows the normally developed pulmonary vasculature (white arrow) and the stump of the absent PA. (D,E) Computed tomography 3D reconstruction shows the aberrant diverticulum at the base of innominate artery (white arrow). 3D, three-dimensional; PVWA, pulmonary vein wedge angiography; PA, pulmonary artery.

Postoperative outcomes

After a careful exploration during surgery, the absent PA was successfully reconstructed using the "two-segment" technique described above in all patients. Three of the patients underwent the procedure with cardiopulmonary bypass, while other procedures were performed off-pump. Following revascularization, the direct measurement of the pressure in MPA during the operation showed that the average mPAP decreased from 31.3±16.0 to 16.8±4.2 mmHg (P=0.047, *Figure 3A*). The average mPAP/rMAP ratio decreased from 0.59±0.27 preoperatively to 0.30±0.10 postoperatively (P=0.028, *Figure 3B*). The average mechanical ventilation time was 45.0±27.6 hours. The mean postoperative hospital stay was 11.8±3.3 days (range, 7–15 days), and the mean total hospital stay was 25.8±6.3 days (range, 17–34 days).

The perioperative patency of the neo-RPA was demonstrated by postoperative TTE. The postoperative LVEF was 65.0%±2.4%, and the postoperative FS was 33.2%±2.5%. The median diameter of the reconstructed PA (pericardial segment) was 5.3 mm (range, 3.0–6.9 mm). The mean blood velocity of the neo-RPA was 2.1±2.1 m/s. The three patients accompanied by patent foramen ovale all changed to a left to right shunt. Three patients were still found with PHT, but the severity had decreased (one moderate, two mild). The postoperative CT scan showed that the neo-PA was patent without compression or fold (*Figure 4*). A thrombosis was observed in the reconstructed PA in one of the patients during postoperative hospitalization, and heparin 10–20 U/kg/h was immediately infused via an infusion pump. The thrombus disappeared before discharge from the hospital. There were no in-hospital deaths.

Follow-up

The follow-up rate was 100%, and the mean follow-up period was 18.85±4.67 months (range, 13.42–23.75 months).

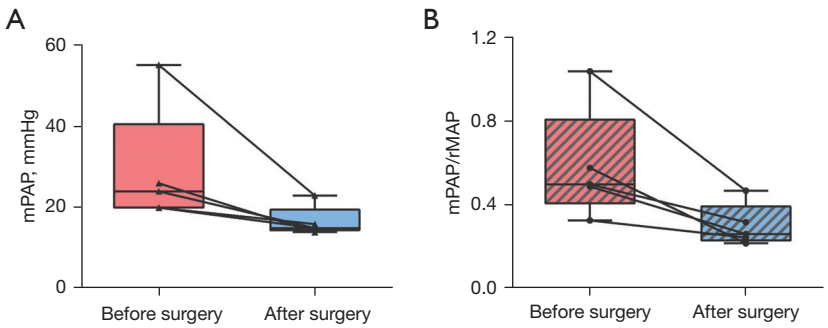


Figure 3 mPAP (A) and the mPAP/rMAP ratio (B) decreased after the operation. The central line within the box denotes the median, while the box contains the 25th to 75th percentiles of the dataset. The whiskers mark the maximum and minimum values of the dataset. mPAP, mean pulmonary artery pressure; rMAP, radial mean arterial pressure.

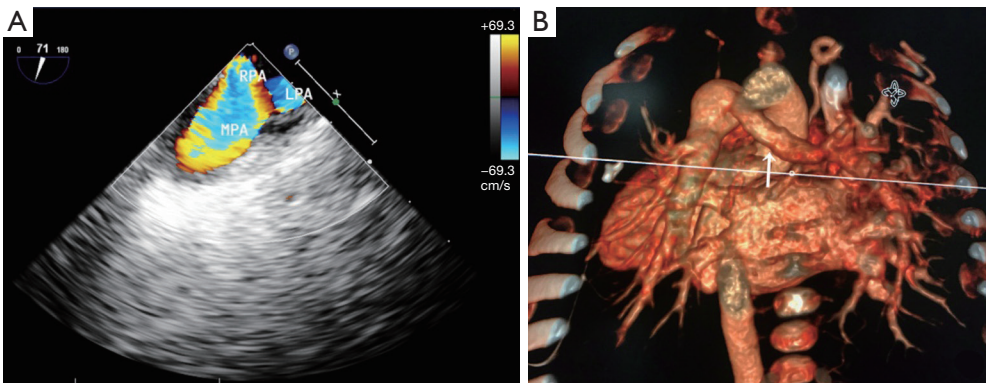


Figure 4 Postoperative evaluation of the reconstructed PA shows the patency of the neo-PA by echo (A) and CT (B). The white arrow shows the reconstructed PA. The white line was the auxiliary line for 3D CT reconstruction. CT, computed tomography; LPA, left pulmonary artery; RPA, right pulmonary artery; MPA, main pulmonary artery; PA, pulmonary artery.

TTE follow-up was available for all patients. The patency of the neo-RPA was evaluated by TTE and CT scan. The 3D CT reconstruction demonstrated that the neo-PA was patent. The LVEF and FS were 64.3%±5.7% and 36.0%±5.7%, respectively. No pulmonary arterial hypertension was found by TTE. The median diameter of the reconstructed PA (pericardial segment) measured by TTE was 6.1 mm (range, 2.7–8.1 mm).

Relative stenosis of the neo-PA was detected by TTE in one patient, with an increased blood velocity and a pressure gradient of 25 mmHg. This patient underwent a redo pulmonary arterioplasty to repair the stenosis of the neo-RPA 9 months after the primary operation. During the redo pulmonary arterioplasty, the reconstructed PA was easily recognized and mobilized due to the guidance of the Gore vascular graft with integrated rings. In addition, after the removal of the Gore vascular graft, we noticed that the pericardial segment was encircled by autologous tissue, connecting with the native hilar PA without any boundaries. Under these circumstances, it was not necessary to remove the pericardial conduit or mobilize the native PA in the hilum. We simply dissected the pericardial conduit along the long axis to expose the stenotic lumen of the neo-artery. We found that the stenosis was caused by a mural thrombus within the pericardial conduit. After inspection, the thrombus was removed and the pericardial conduit incision was enlarged with a pericardial patch. Then, a new Gore vascular graft with a larger diameter was implanted to reconstruct the PA. The patient was discharged soon after surgery, and the reconstructed RPA remained patent during the subsequent follow-up.

Discussion

This study summarized our experience of surgical repair of UAPA in children with PHT. We used 3D CT reconstruction combined with PVWA for the diagnosis of UAPA and evaluated the pulmonary vasculature development. We also presented an effective and safe "two-segment" technique to reconstruct the absent PA. The postoperative follow-up illustrated the patency of the neo-PA, and the risks of postoperative complications were acceptable.

UAPA was first reported by Frentzel in 1868 (11). The prevalence of isolated UAPA is estimated to be 1/200,000–1/300,000 (12,13), and the main symptoms in infant patients are congestive heart failure and PHT. PHT is a severe complication of UAPA and is not known to regress spontaneously, leading to a high mortality rate (reported to be 7%) (13). However, some children with an isolated UAPA are asymptomatic and therefore remain undiagnosed until adulthood. Such patients gradually develop recurrent hemoptysis, lower respiratory tract infection, or PHT, which eventually affects their quality of life and natural lifetime. So, we can assume that early diagnosis and physiological correction in childhood are reasonable interventions to improve the prognosis.

Previously, diagnosis mainly depended on invasive angiography. PVWA is believed to be a "gold standard" for UAPA diagnosis. However, there might be an increased risk for newborn infants undergoing such an examination. The difficulty of performing PVWA also limits the widespread use of this invasive technique. With the development of diagnostic techniques, such as the 3D reconstruction of CT scans, PVWA is tending not to be essential (14).

Most pulmonary vessels can be clearly presented through a 3D reconstruction after contrast-enhanced CT, which facilitates the detection of asymptomatic UAPA. Pfefferkorn *et al.* reported their results of cardiac catheterization and found a diverticulum of the innominate artery in patients in whom the aortic arch and absent PA were on opposite sides (15). These findings were subsequently supported by other reports (5,16). By using 3D reconstruction of CT scans, we found a similar diverticulum in all of our patients. An implication of this finding may be that the diverticulum indicating fetal blood supply to the affected lung might be an important clue for the diagnosis of UAPA. In addition, it is also worth noting that the anomalies of three patients were observed in prenatal screening by fetal ultrasound examination, which suggests fetal echocardiography could be an effective method for early screening for UAPA.

Ten Harkel *et al.* reported that 44% of patients had accompanying PHT (4). Children with UAPA and PHT tend to have a poor prognosis, and infants with UAPA may have an increased risk of persistent PHT (17). PHT is caused by the imbalance between the decreased pulmonary vascular bed and the relatively increased blood flow. It may eventually develop in untreated patients and lead to further deterioration and higher mortality (13). Despite the lack of consensus regarding PHT prevention, previous studies have shown that early revascularization of the absent PA improved the condition of patients (1,18). It has been reported that patients who receive delayed surgical interventions may develop regression of the affected PA and have a less favorable outcome (7).

All patients in this study were diagnosed with UAPA accompanied by PHT. Following reconstruction of the absent PA, the PA pressure and mPAP/rMAP ratio decreased significantly with the blood flow to the neo-PA. No fatal complications occurred perioperatively. Our experience suggested that early reconstruction of an absent PA can markedly alleviate PHT, and this physiological correction might be more favorable for the long-term prognosis.

The surgical strategy for revascularization is another topic of debate. Some researchers have reported their experience with the two-stage strategy. Firstly, a systemic-pulmonary shunt is created by a modified Blalock-Taussig shunt (19,20) or by interventional catheterization (21), which tends to promote the growth of pulmonary vasculature and the development of ipsilateral lung. Subsequent corrective surgery is then conducted to reconstruct the absent PA. However, the majority of patients with UAPA have the ductus arteriosus supplying the affected pulmonary vessel during embryogenesis. So, the intrapulmonary vasculature and the distal portion of the absent PA may develop normally. Moreover, a systemic-pulmonary shunt would inevitably increase the preload of the left ventricle, which increases the risk of congestive heart failure, especially in patients with poor right ventricle function. Thus, we believe that single-stage correction is feasible for most patients diagnosed with UAPA. Our perioperative results showed that the PA pressure decreased immediately after the operation and there were no patient deaths, suggesting that this strategy was appropriate.

The difficulty of directly connecting the hilar PA and the MPA is the substantial distance between the two stumps (22). A direct anastomosis would inevitably lead to excess

tension on the anastomosis. Several studies have reported interposition of the tube graft created with autologous pericardium (23) or prosthetic material (19). The disadvantage of prosthetic material is that the graft cannot grow naturally with age (8). However, the autologous pericardium tube is vulnerable to the compression of the adjacent vessels, with blood flow stasis forming a thrombus that eventually occludes (18). Moreover, a redo surgery for repeated pulmonary arterioplasty would be inevitable because of a mismatch between the diameter of the tube graft and the patient's growth. Based on our experience, reconstruction of the PA in the hilum is usually challenging during redo surgery. In most cases, the native PA in the hilum is deeper and poorly developed. The native PA in the hilum is always messy following removal of the tube graft, which increases the difficulty of anastomosis, with a high risk of anastomotic stenosis or uncontrollable hemorrhage. Therefore, a comprehensively preventive plan during primary surgery is highly necessary.

In our study, we modified the corrective procedure with a "two-segment" technique by reconstructing the absent PA with fresh autologous pericardium and a Gore vascular graft with integrated rings. By using autologous pericardium, we successfully augmented and prolonged the confluence of the affected PA at the hilum. The reconstruction was then completed by the second segment of a Gore vascular graft with integrated rings, connecting the proximal end of the pericardial conduit and the MPA. The pericardial conduit provided the growth potential for future development, and the vascular graft prevented deformation caused by the compression of the ascending aorta. Notably, in one case of redo pulmonary arterioplasty, it was not necessary to remove the pericardial conduit after removing the Gore vascular graft. Meanwhile, the pericardial segment was equivalent to the prolongation of the hilar artery. A redo pulmonary arterioplasty could be simply conducted by a dissection of the pericardial conduit along the long axis and enlargement of the pericardial incision with a pericardial patch, followed by implantation of a new Gore vascular graft. The "two-segment" technique might simplify the procedure and decrease the risks of a potential redo pulmonary arterioplasty. This preventive technique for redo pulmonary arterioplasty has been preliminarily verified in our limited practice.

Stenosis of the neo-PA is common and remains a problem after reconstruction of the absent PA (23,24), which is usually caused by thrombosis in the neo-PA. Considering the utilization of artificial material in the "two-segment" technique, sufficient anticoagulation is essential for maintaining the patency of the neo-PA and improving the long-term outcomes (8). Owing to the high risk of hemorrhage after anticoagulation therapy in children, we utilized anticoagulation therapy for a short period followed by antiplatelet therapy. Based on our experience heparin therapy was effective in cases of perioperative thrombosis. Additionally, considering that there was one patient who developed thrombosis and stenosis in the reconstructed PA during follow-up, frequent and close follow-up is indispensable.

Obviously, our results were still limited. Due to the small number of samples, the valuable and interesting finding of aberrant diverticulum for diagnosis of UAPA with 3D CT reconstruction cannot be applied broadly. Also, the sample size was not large enough to confirm the changes in pulmonary pressure following pulmonary reconstruction. Furthermore, the duration of follow-up was not long enough to evaluate the long-term patency of the reconstructed PA. However, we believe that the conclusions of this study are still reliable. More prospective and large-scale studies with longer follow-up periods are expected.

Conclusions

Early reconstruction of the PA may effectively alleviate PHT in children with UAPA. The "two-segment" technique is an effective and safe surgical technique to reconstruct the absent PA and might facilitate potential redo pulmonary arterioplasty. Anticoagulation or antiplatelet therapies are required postoperatively, and frequent and close follow-up is indispensable to prevent thrombosis or stenosis in the neo-PA.

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Footnote

Reporting Checklist: The authors have completed the STROBE reporting checklist. Available at https://

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Data Sharing Statement: Available at https://tp.amegroups.com/article/view/10.21037/tp-22-491/dss

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://tp.amegroups.com/article/view/10.21037/tp-22-491/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). The study was approved by the Research Ethics Committee of Guangdong Provincial People's Hospital, Guangdong Academy of Medical Sciences (approval No. KY-Q-2021-267-01). Informed consents were obtained from the legal guardians of the children.

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