A case report of a long-term survivor after inadvertent ligation of the left pulmonary artery during intended ductal ligation

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Background  Patent ductus arteriosus (PDA) can close on its own during childhood. Patent ductus arteriosus with left pulmonary artery (LPA) occlusion is rare. Here, we describe possible aetiologies of this condition and treatment strategies based on recent guidelines.

Case summary  A 35-year-old man experienced shortness of breath for 20 years. Physical examination revealed pitting oedema, digital clubbing, and bi-phasic murmur along the left sternal border at the 2nd and 3rd intercostal space. Congenital heart disease was suspected. Echocardiography revealed a severely dilated pulmonary trunk and PDA; however, the LPA was not visible. The patient has undergone PDA ligation surgery 30 years ago, which may have caused accidental LPA ligation; however, extreme elevation of pulmonary pressure led to increased difficulties in performing LPA reconstruction and PDA division. Therefore, pulmonary arterial hypertension (PAH) initial combination therapy with parenteral prostanoids was prescribed. The patient’s condition improved gradually.

Discussion  Inadvertent ligation of the LPA instead of PDA is a rare and usually fatal complication during ductal ligation. Patients who survive this catastrophic complication usually develop the progressive pulmonary vascular disease with increased pulmonary pressure and impaired lung parenchyma resulting in right heart and respiratory failure. Early and regular follow-up with cardiac imaging studies is important to diagnose this complication. Reconstruction of the ligated LPA and PDA late in the disease course is difficult due to the development of pulmonary arterial hypertension. Initial PAH combination therapy may be valuable for relieving the patients’ symptoms at that stage. Heart and lung transplantation can also be considered in suitable patients.

Keywords  Patent ductus arteriosus • Ligation • Left pulmonary artery occlusion • Case report

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Introduction

Failure of ductus arteriosus closure leads to patent ductus arteriosus (PDA). Patent ductus arteriosus with left pulmonary artery (LPA) occlusion is a rare condition in clinical practice. Careful identification of LPA occlusion is necessary. Aetiologies of LPA occlusion include thrombosis, vasculitis, congenital anomalies, and less frequently iatrogenic causes.

During PDA ligation surgery, LPA can be inadvertently ligated with high mortality and morbidity rates. Estimated incidence of LPA ligation during PDA ligation is approximately 1 in 1000 cases. However, non-routinely performed post-operative echocardiographic or other imaging examinations may have underestimated this rate. Herein, we describe the case of a long-term survivor after inadvertent ligation of LPA during ductal ligation surgery, which has not been recognized for more than 30 years.

Case presentation

A 35-year-old man has been experiencing shortness of breath for nearly 20 years. During the past 6 months, his symptoms worsened with the stiffening of his four limbs.

Physical examination revealed the following vital signs: blood pressure, 130/88 mmHg; heart rate, 100 beats per min; respiratory rate, 22 breaths per min; and oxygen desaturation rate, 60% on room air. Upon examination, the jugular vein and abdomen were distended. His heart sounds were regular, and bi-phasic murmur along the left sternal border was best heard at the 2nd and 3rd intercostal space, with mid-diastolic murmur at the 2nd aortic valve area. Pulmonary examination revealed bilateral rales, and decreased breath sounds in the left lung. Upon examination of the extremities, pitting oedema and digital clubbing were observed.

Electrocardiogram revealed right axis deviation and prominent elevated R wave in leads V1 through V2 (Supplementary material online, Figure S1). Laboratory test results showed serum N-terminal brain natriuretic peptide (NT-proBNP, normal range 0–150 pg/mL) level of 8768 pg/mL and hyperhaemoglobinemia (haemoglobin of 214 g/L, normal range 130–175 g/L). Markers for connective tissue diseases (antibodies), systemic vasculitis, and thrombophilia were negative. Echocardiography revealed enlarged right atrium (61 mm) and right ventricle (36 mm × 54 mm), flattening of the interventricular septum towards the left ventricle, and severe tricuspid regurgitation with pressure gradient of 90 mmHg. The tricuspid annular plane systolic excursion decreased to 13 mm, with preservation of left ventricular ejection fraction. Unexpectedly, occlusion of the LPA was observed with only a protruded root. A PDA with 20 mm in diameter was visible, and a mosaic bidirectional flow across this PDA was noted (Figure 1A and Supplementary material online, Video). Chest radiography showed right ventricular hypertrophy and bulging main pulmonary artery (Figure 2A). Chest computed tomography confirmed marked enlargement of the main pulmonary artery (74 mm) and large PDA (Figure 3A). The LPA root was visible. However, the distal branches were faintly filled (Figure 2B), subsequently, the left lung was dysplastic and collapsed (Figure 2B–D). The right pulmonary artery and its branches were dilated and relatively hyperaemic with disproportionate perfusion. No segmental or subsegmental mismatched perfusion was noted on dual-energy lung perfusion imaging. Based on these findings, a diagnosis of right heart failure was made. This failure was secondary to PDA with right-to-left shunting and complicated by LPA occlusion and pulmonary hypoplasia-induced hypoxaemia.

The patient’s history was carefully reviewed to confirm the presence of congenital PDA with unilateral agenesis of the LPA. After birth, the presence of a harsh heart murmur without cyanosis raised
a suspicion of congenital heart disease. Thereafter, PDA was confirmed. Subsequently, PDA ligation surgery was performed at the age of 4. According to his parents, surgery was uneventful except for the development of hoarseness, which has since persisted. He lost to regular follow-up. Three years later, echocardiography revealed the persistence of PDA with left-to-right shunt. However, since no symptoms were noticed, his parents refused a second surgery. At the age of 31, the patient presented with more severe symptoms including haemoptysis and exercise intolerance. Right heart catheterization revealed a pulmonary artery pressure of 140/87 (m104) mmHg and pulmonary vascular resistance of 46.5 Wood Unit QP/QS 0.22.

Discussion

Unilateral pulmonary artery occlusion can either be congenital or secondary in aetiology. Congenital unilateral agenesis of the pulmonary artery is a rare anomaly. Our patient demonstrated an LPA proximal root, ruling out congenital agenesis of the LPA. Moreover, since no segmental or subsegmental perfusion defects were observed, LPA occlusion caused by thromboembolic disease was unlikely. Additionally, other possible causes, including Takayasu's arteritis and fibroinuous mediastinitis, were excluded because their diagnostic criteria and specific imaging features were not fulfilled.

Accidental division of LPA instead of the PDA is a rare and fatal complication of ductal closure. We searched PubMed using topics of 'left pulmonary artery ligation,' 'ductus arteriosus,' and 'surgery' to identify literatures discussing unintentional ligation of the LPA during ductal ligation. Among 22 cases, 9 were reviewed (Table 1). To the best of our knowledge, the present case is the only long-term survivor after LPA ligation with PDA patency for more than 30 years.

Pulmonary arterial hypertension is common in the setting of shunt lesions and might be associated with corrective surgery. Large PDA with PAH is more misleading, as the ductus is often as large as the aortic arch and may partially obscure the arch causing obstruction. Short-term complications of LPA ligation include respiratory failure, recurrent infection, and increased perioperative mortality (Table 1). Of note, our patient showed no significant deterioration immediately after LPA ligation, which was consistent with previous reports (Table 1). The patient showed severe pulmonary vascular remodelling, which may be secondary to left lung dysplasia, right lung overinflation, and pulmonary vascular disease due to patent intracardiac shunting. To allow for early detection of this complication, echocardiography in the early post-operative period and regular follow-up should be performed. Our patient was found to have patent PDA on echocardiography 4 years after the primary surgery. However, since the patient did not exhibit symptoms, his parents refused a second surgery. Moreover, the patients' clinical condition worsened at 17 years of age, indicating that reconstruction was overdue. Echocardiography revealed isolated right-sided chamber enlargement, a finding not consistent with isolated persistent PDA. Regarding this situation, detailed echocardiographic evaluation during strict and regular follow-ups is needed, which allow for timely referral to an expert pulmonary hypertension centre when suspicious findings are present.

Prompt diagnosis and reoperation of ligated LPA at a specialized cardiovascular centre is important to minimize risks of reconstructive surgery. Otherwise, division of the LPA with continued patency of

Figure 1 Echocardiography showing patent ductus arteriosus and left pulmonary artery. (A) Mosaic bidirectional flow across the patent ductus arteriosus is shown. (B) Severe dilatation of the pulmonary trunk and right pulmonary artery is shown. The left pulmonary artery was missing after protruding its root (red arrowhead).
Figure 2. Images of the patient’s lung. Bedside chest X-ray showed an aneurysm of the pulmonary trunk, dilated right heart, and reduced left lung volume (A). Cross-sectional chest computed tomography revealed left lung parenchymal dysplasia and right lung vascular hyperaemia (B). A 3D reconstruction of the pulmonary airway demonstrated reduced volume and disorganized parenchyma of the left side from the anterior (C) and posterior (D) views.

Figure 3. Computed tomographic pulmonary artery angiography. (A) Sagittal view of the patent ductus arteriosus (red arrow) and (B) left pulmonary artery root (red arrowhead). (B) Dilation of the pulmonary trunk and the right pulmonary artery and its branches are shown. The left pulmonary artery was missing after protruding its root (red arrowhead).
**Table 1** Inadvertent ligation of left pulmonary artery in literature

| Ref       | Age       | Comorbidities       | Post-ligation symptoms                  | Diagnostic methods                | Treatments                  | Outcomes        |
|-----------|-----------|---------------------|----------------------------------------|-----------------------------------|-----------------------------|-----------------|
| Yucel et al. | 17 days   | Aortic coarctation  | No improvement in pulmonary function   | Echo, CTA confirmed               | Reconstructive surgery 3 days later | Survived        |
| Pontius et al. | 4 months | Down syndrome       | NA                                     | Autopsy                           | No                          | Died 8 h after surgery |
|           | 4 months  | VSD                 | Atelectasis                            | Oesophageal stethoscope during surgery | Reconstructive surgery immediately | Died < 1 year    |
|           | 24 years  | VSD                 | None                                   | Angiography 12 years later        | VSD closure                 | Died after surgery |
|           | 4 months  | Levo-transposition  | None                                   | Cineangiogram 2 years later        | Conservative                | Survived         |
|           | 5 days    | None                | NA                                     | Oesophageal stethoscope            | Reconstructive surgery next day | Survived         |
|           | 10 months | Down syndrome       | Fail to thrive                         | Cardiac catheterization            | Reconstructive surgery 5 months later | Survived        |
|           | 2 years   | Seckel’s bird-headed dwarf | None                                  | Cardiac catheterization 4 years later | Conservative               | Survived         |
|           | Infant    | None                | None                                   | NA                                | Reconstructive surgery 3 years later | Survived         |
|           | 7 months  | None                | None                                   | Cardiac catheterization 21 months later | Reconstructive surgery 21 months later but no flow to LPA | Survived well   |
|           | 3 weeks   | Coarctation          | None                                   | X-ray, Cardiac catheterization 2 weeks later |                    | Died            |
| Tefera et al. | 10 years | None                | None                                   | Echo                              | Reconstructive surgery 3 years later. | Survived         |
|           | 33 days   | None, 28 weeks preterm| Sepsis                                | Physical examination, Echo, Chest X-ray, CTA confirmed | Reconstructive surgery 46 days later | Extensive tissue damage. No reperfusion in the LPA. Died 6 months later |
| Terlemez et al. | 10 months | Down syndrome       | NA                                     | X-ray, Echo, Angiography confirmed | Reconstructive surgery 5 months later | Survived         |
|           | 11 days   | None                | NA                                     | X-ray, Echo, Angiography confirmed | Reconstructive surgery on the same day | Survived         |
|           | 20 months | None                | NA                                     | X-ray, CT confirmed               | Closure of PDA              | Survived         |
|           | 4 days    | NA                  | X-ray                                  |                                   |                             | Survived         |
PDA over a long period of time might lead to markedly increased difficulty in reconstruction with a high risk of morbidity and mortality. In some case reports, a combination of percutaneous stenting with surgical repair in specific situations has been tried, which resulted in good results. Addition al pulmonary stenting may be effective in alleviating patients’ pulmonary pressure and pulmonary vascular resistance, especially in patients who were not candidates for reconstruction surgery. After consulting with the surgeon and the patient’s family, LPA reconstruction and PDA division were not recommended for this present patient. In addition, LPA total occlusion, left lung parenchyma dysplasia, and severe pulmonary vessel remodelling have led to increased risk and difficulties in performing correction surgery or percutaneous stenting. Initial combination therapy including parenteral prostanoids was prescribed in the present high-risk patient according to the recent guidelines. Fortunately, our patient’s condition gradually improved after adding PAH therapy. Furthermore, PAH medication can help ameliorate symptoms. However, in the setting of Eisenmenger (as present in our patient) and end-stage heart failure, heart transplantation may also be considered. The present case might have been counselled regarding heart and lung transplantation for severe dysplasia of both lungs. Regular follow-up was then performed to monitor the patient’s potential to be a candidate for reconstruction or heart and lung transplantation.

Conclusion

Inadvertent ligation of the LPA is a rare and harmful complication of ductal ligation. We present a long-term survivor after mistakenly undergoing ligated LPA with patent PDA for more than 30 years. A combination of PAH therapy was helpful in ameliorating symptoms. In addition, regular follow-up is important while heart and lung transplantation might be considered in the future.

Lead author biography

Qi-xian Zeng, MD, PhD, graduated from the Chinese Academy of Medical Sciences and Peking Union Medical College in 2018, now is a cardiology attending at Fuwai Hospital in Beijing, China. Her line of research is focused on pulmonary vascular disease and pulmonary vessel intervention.

Supplementary material

Supplementary material is available at European Heart Journal—Case Reports online.

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.
Consent: The authors confirm that written consent for submission and publication has been obtained from the patients in line with COPE guidance.

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