


CASE REPORT

Open Access



Lung transplantation with concomitant cardiac repair for congenital hypoplasia of bilateral pulmonary arteries and patent ductus arteriosus

Che-Chih Cheng^{1,2}, Ming-Tai Lin^{3,4}, Shu-Chien Huang^{2,4} and Hsao-Hsun Hsu^{2,4*} 

Abstract

Background: Profound pulmonary arterial hypertension with end-stage right heart failure is considered to be the main cause of death in children with un-repaired congenital heart disease, and the traditional surgical treatment is heart–lung transplantation. We performed bilateral lung transplantation (LTx) with concomitant cardiac repair, and the patient has uplifting outcome.

Case presentation: We have reported the case of a patient with congenital hypoplasia of the bilateral pulmonary arteries and patent ductus arteriosus. The patient's clinical condition was gradually worsening and severely limited his ability to perform the activities of daily life. Bilateral LTx with concomitant patent ductus arteriosus repair was performed at the age of 11 years. The postoperative course was smooth and cardiopulmonary function nearly returned to normal according to radiological and laboratory examinations.

Conclusions: Bilateral LTx with concomitant cardiac repair may be superior to heart–lung transplantation in the case of the specific congenital heart disease.

Keywords: Congenital heart disease, Lung transplantation, Patent ductus arteriosus

Background

Pulmonary hypoplasia, a rare congenital heart disease (CHD), is characterized by the underdevelopment of the pulmonary structure, including the lung parenchyma, bronchus, and blood vessels [1, 2]. Although the true pathogenesis is unclear, it may be associated with an accident in utero or an embryologic defect of the lung or vascular tissue, which resulted in unilateral or bilateral absence of the pulmonary artery. For patients with congenital hypoplasia of the bilateral pulmonary arteries, the

pulmonary circulation depends on support from the systemic collateral circulation. The lack of normal pulmonary vessels leads to erythrocytosis, chronic hypoxemia, and cyanotic multisystemic complications, which result in an increase in pulmonary vascular resistance and progressive life-threatening pulmonary arterial hypertension (PAH) [3].

We herein report the case of an 11-year-old child with congenital hypoplasia of the bilateral pulmonary arteries and patent ductus arteriosus (PDA). The patient presented with central cyanosis and consequent PAH, and received a bilateral lung transplantation (LTx) with intraoperative PDA repair for his end-stage pulmonary hypoplasia accompanied with severe PAH and right heart decompensation.

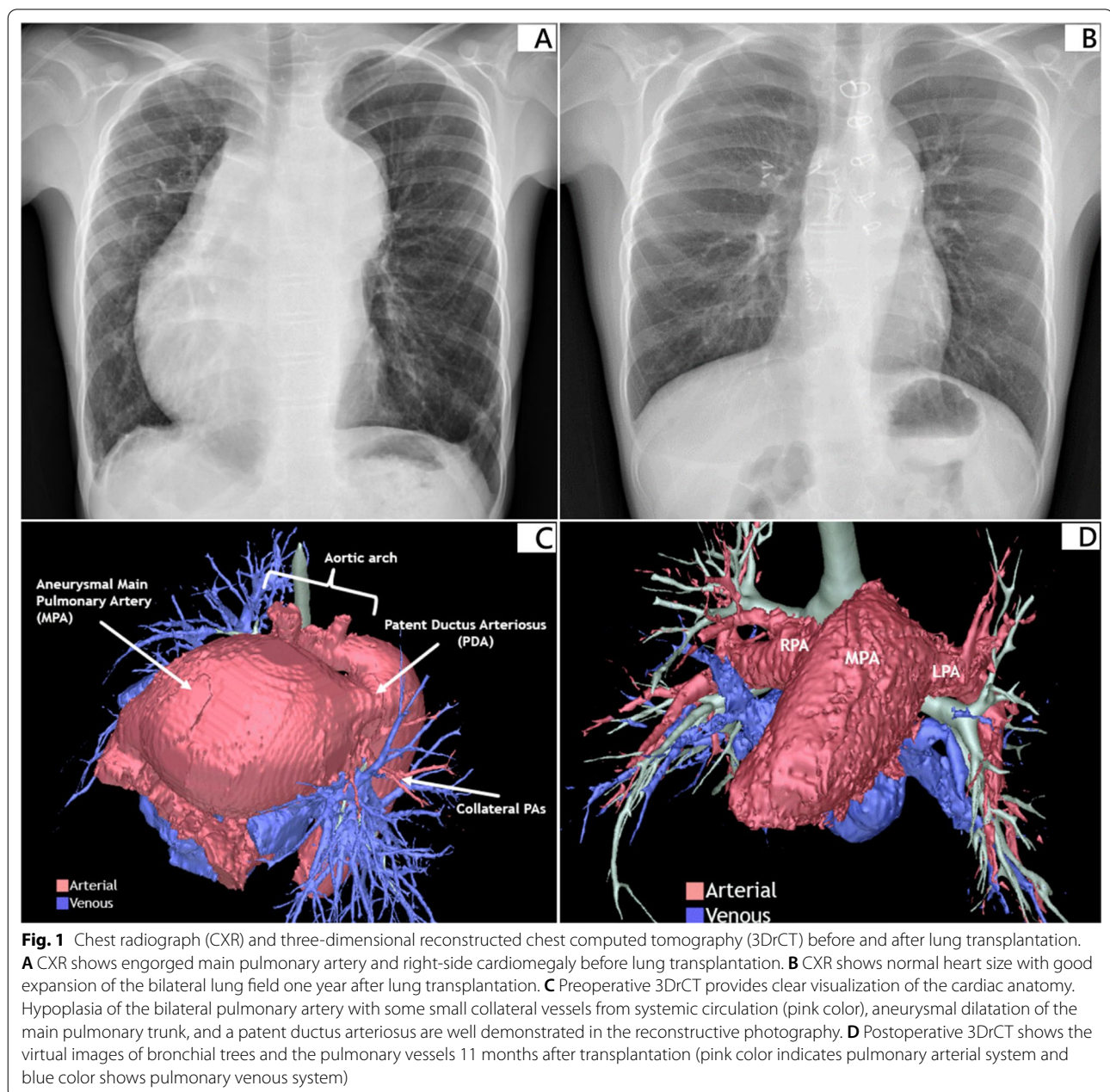
*Correspondence: ntuhsu@gmail.com

² Division of Thoracic Surgery, Department of Surgery, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan

Full list of author information is available at the end of the article



© The Author(s) 2022. **Open Access** This article is licensed under a Creative Commons Attribution 4.0 International License, which permits use, sharing, adaptation, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if changes were made. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit <http://creativecommons.org/licenses/by/4.0/>. The Creative Commons Public Domain Dedication waiver (<http://creativecommons.org/publicdomain/zero/1.0/>) applies to the data made available in this article, unless otherwise stated in a credit line to the data.



Case presentation

A 10-year-old boy with CHD presenting with central cyanosis and severe PAH was referred to us for LTx evaluation. The diagnosis of congenital hypoplasia of the bilateral pulmonary arteries and PDA was confirmed by right heart catheterization at birth. The PAH-targeted therapy, Sildenafil, was initially prescribed at the age of 8 weeks. In order to relieve his symptoms, the PAH combination therapy of Sildenafil and Bosentan was started at 17 weeks. The dose was modified according to his body weight. However, his clinical condition deteriorated

resulting in an increasing number of hospital admissions due to end-stage CHD presenting as severe hypoxia and shortness of breath. The repeated right heart catheterization before referral showed a mean pulmonary arterial pressure of 97 mmHg (the same level as that in the ascending aorta) and the 6-min-walk test distance was only 80 m. Pre-operative chest radiography and multidetector thoracic computed tomography (CT) illustrated aneurysmal dilatation of the main pulmonary trunk and right-side cardiomegaly, accompanied with severe hypoplasia of the bilateral pulmonary arteries and a large PDA

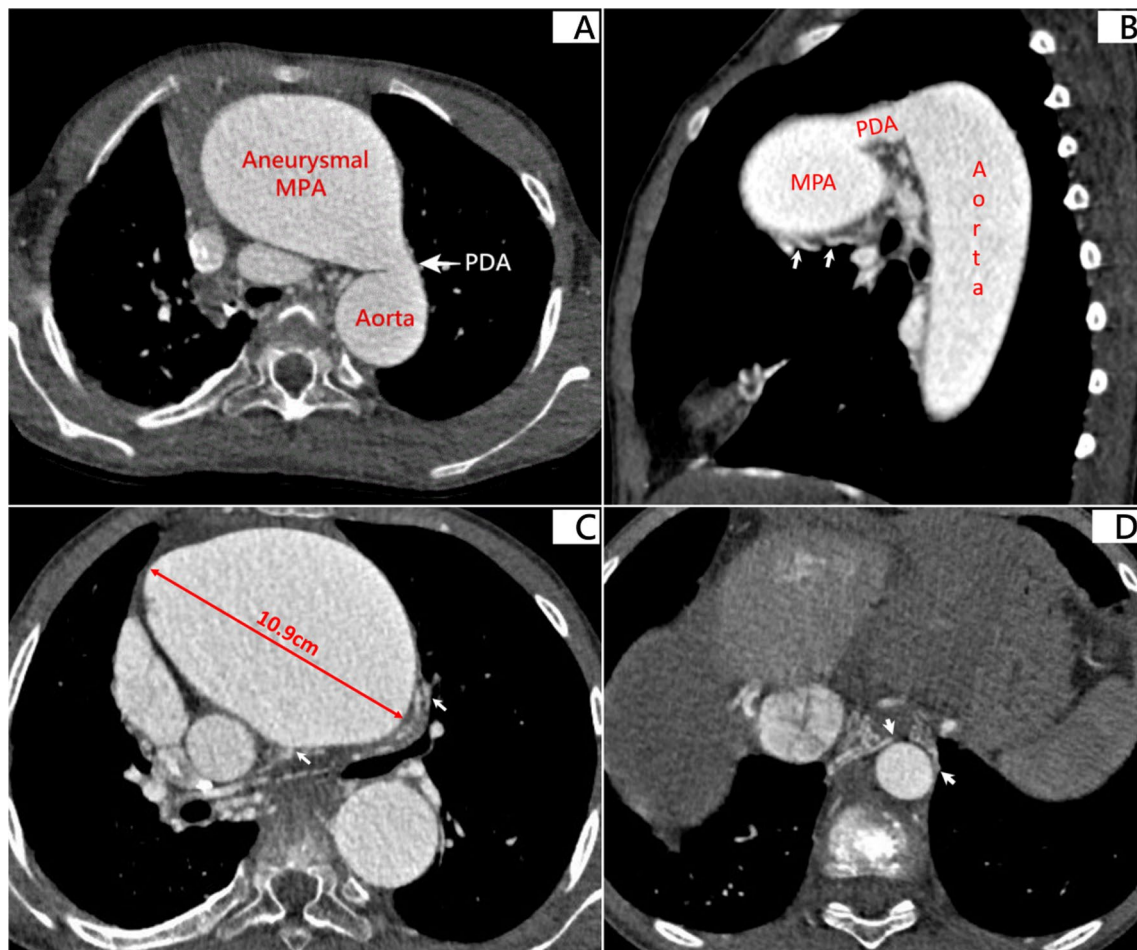


Fig. 2 Contrast thoracic computed tomography shows the patient's cardi thoracic anatomy before lung transplantation. **A** Patent ductus arteriosus (approximately 1.1 cm in diameter) identified between the aneurysmal main pulmonary artery and the descending aorta. **B** Axial view of thoracic computed tomography reveals patent ductus arteriosus between the main pulmonary artery and the descending aorta. Arrows indicate several collateral branches of bilateral pulmonary arteries (approximately 4–5 mm in diameter). **C** Maximum diameter of the aneurysmal main pulmonary trunk is 10.9 cm. The Arrows indicate several collateral branches of bilateral pulmonary arteries (approximately 4–5 mm in diameter). **D** Arrows indicate bilateral major aortopulmonary collateral arteries (MAPCAs) from the descending aorta (approximately 2–4 mm in diameter)

(Figs. 1A, C, and 2). After multidisciplinary evaluation, bilateral LTx under cardiopulmonary bypass support with concomitant intraoperative PDA repair was offered. The patient was then listed as status 1 exception in the LTx waiting list.

Due to extreme scarcity of cadaveric donors in Taiwan and the small size of our recipient (160 cm in height and 38 kg in weight), a suitable donor was available a year later (a brain-dead 32-year-old male with a good size match between the donor and recipient). Under precise donor-recipient team cooperation, a standard donor procurement technique with Perfadex® solution for antegrade and retro-grade organ perfusion and preservation was performed. After median sternotomy, the PDA was resected, and the aortic side of the PDA was repaired by

continued suture after cardiopulmonary bypass had been established. The oversized main pulmonary trunk was trimmed to approximately half of its original diameter using linear stapler and reinforced by 4–0 Prolene running suture. The recipient's right lung was removed and then the donor's right lung was implanted by sequential anastomosis of right main bronchus and left atrium first. The donor's right main pulmonary artery was anastomosed to the recipient's main pulmonary trunk using end-to-side anastomosis. And the left donor lung was implanted using a similar technique. The total allograft ischemia time was 513 min.

The hospitalization included an intensive care unit (ICU) stay of 67 days and a general ward stay of 15 days. There were two major events during the ICU stay.

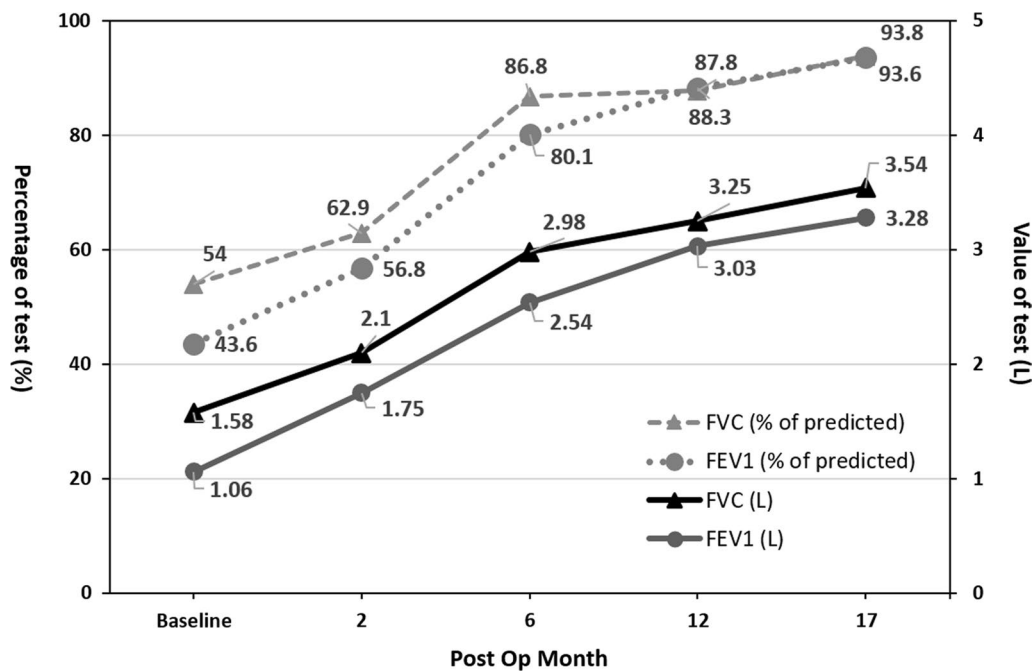


Fig. 3 Pulmonary functional test before and after bilateral lung transplantation

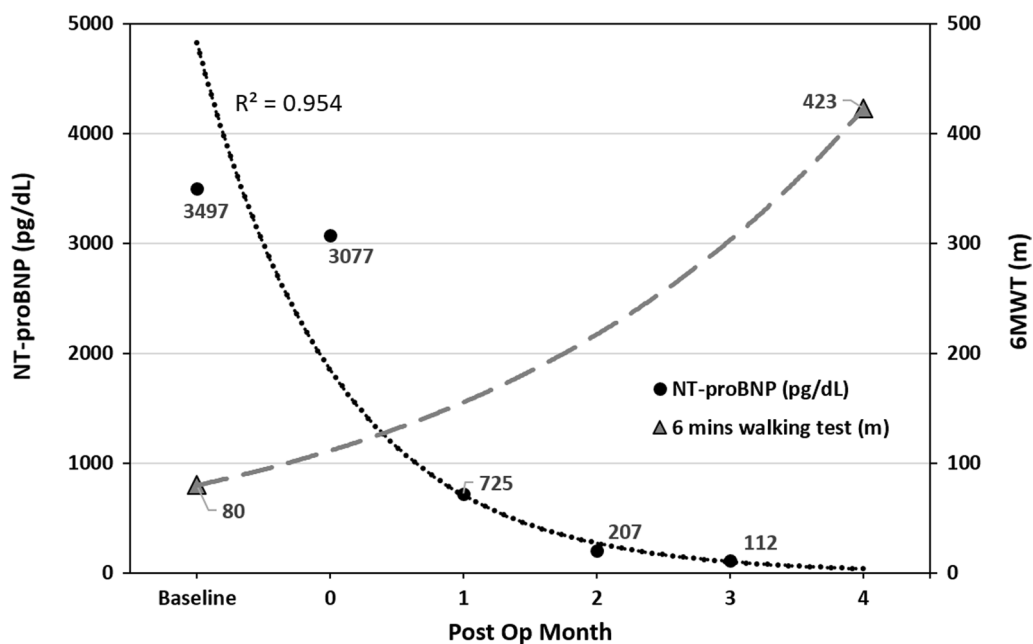


Fig. 4 Curvilinear relationship between time and blood level of NT-proBNP and distance of 6-Minute-Walk test

The first was pneumonia with sepsis attributed to the donor's nosocomial infection that delayed removal of the endotracheal tube; this lasted for 12 days. The second was pancytopenia with intermittent fever. After a series of examinations and drug modifications, a complex

web of causes was determined, including collapsed lung pneumonia, cytomegalovirus infection related splenomegaly complicated with thrombocytopenia, and drug reaction (Tazocin), that led to an ICU stay of approximately 38 days. The patient was discharged in a

relatively stable condition on postoperative day 82 after an improved clinical status and rehabilitation in the general ward. Thoracic CT revealed that the diameter of dilated pulmonary trunk significantly decreased from 10.9 cm preoperatively to 4.9 cm postoperatively. The postoperative chest radiograph demonstrated a nearly normal cardiac shadow, and a reconstructive chest CT illustrated that the patient's anatomy was similar to normal (Fig. 1B, D). The patients exercise capacity, such as pulmonary function and 6-min-walk-test distance, improved significantly and the level of NT-pro BNP returned to the normal range, compared with that of the preoperative condition (Figs. 3, 4). The patient had no physical limitations and enjoyed a good quality of life and returned to school 1 year after surgery.

Discussion and conclusions

Because of early diagnosis and the advances in pediatric cardiac surgery, the life expectancy of patients with CHD has improved significantly over recent decades. For CHD children with a complex anatomy who are not suitable candidates for surgical repair, unrepaired intracardiac or extracardiac shunts could lead to a progressive increase in pulmonary vascular resistance and life-threatening PAH [4, 5]. As these CHD patients grow up, a progressive decline in cardiopulmonary function due to irreversible PAH and cyanotic multisystemic complications increase the morbidities and affect their daily activities. As a consequence, most deaths in CHD patients with refractory PAH occur in adulthood. According to the "Guidelines for the Management of Grown up Congenital Heart Disease", cardiothoracic transplantation, such as heart–lung transplantation (HLT_x) or LT_x with simultaneous repair of the cardiac defect, is recommended to be considered when the short-term prognosis is poor or quality of life is unacceptable [6].

For end-stage CHD patients referred for thoracic transplantation, HLT_x theoretically could be accomplished with a shorter operation and bypass time because there is no need to deal with complex congenital defects and the number of anastomoses between donor and recipient is limited, compared to that of LT_x with additional reconstructive surgery. For CHD patients with Eisenmenger syndrome receiving thoracic transplantation, Waddell et al. reported that the 30-day (80.7%) and 1-year survival rates (70.1%) in the HLT_x group were better than those in the LT_x group (68% and 55.2%) [7]. However, a cohort study conducted in Nordic patients with Eisenmenger syndrome who underwent transplantation demonstrated that there was no difference in the median survival after HLT_x and LT_x with concomitant repair of cardiac defects [8]. This multicenter study also illustrated that there was no survival difference between complex and simple

anatomical defects, between pediatric and adult transplantations, or between centers. Compared to HLT_x, the additional benefit of LT_x with simultaneously cardiac repair for CHD patients is that the donor heart could be preserved and offered to another recipient on the heart transplant list. In the current era of organ scarcity, this therapeutic option would be considered favorable due to the large disparity between the patients on the waiting list and the number of cadaveric allografts available.

In conclusion, although the operation time was longer and expertise in the surgical technique is required, our case demonstrated that LT_x with concomitant cardiac repair at centers with expertise in both cardiac surgery and thoracic transplantation could be an acceptable and favorable therapeutic option for some selective end-stage CHD patients with complex cardiac defects.

Abbreviations

CHD: Congenital heart disease; CT: Computed tomography; CXR: Chest radiography; HLT_x: Heart–lung transplantation; LT_x: Lung transplantation; MAPCA: Major aortopulmonary collateral artery; PAH: Pulmonary arterial hypertension; PDA: Patent ductus arteriosus; 3DrCT: Three-dimensional reconstructed chest computed tomography.

Acknowledgements

The authors sincerely thank donor and transplant coordinators at the National Taiwan University Hospital, and the Taiwan Organ Registry and Sharing Center for their continuous efforts in increasing the safety, availability, and quality of donors for organ transplantation. The following persons are acknowledged for their contribution with data acquisition and overall support of this report: Pei-Chen Hsu and Yu-Mei Lai (Department of Surgery at National Taiwan University Hospital, National Taiwan University College of Medicine, Taipei, Taiwan). This work was supported by grants from National Taiwan University Hospital (MS441).

Authors' contributions

CCC, MTL, SCH, and HHH enrolled and treated the patient. HHH and CCC searched the literature and collected clinical data. CCC and HHH drafted the article and submitted the paper for publication. All authors read and approved the final manuscript.

Funding

All authors declare that there were no financial conflicts of interest.

Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Not applicable

Consent for publication

Completed

Competing interests

The authors declare that they have no competing interests.

Author details

¹Department of Surgery, Kaohsiung Armed Forces General Hospital, Kaohsiung, Taiwan. ²Division of Thoracic Surgery, Department of Surgery, National Taiwan University Hospital and National Taiwan University College of Medicine,

Taipei, Taiwan. ³Department of Pediatrics, National Taiwan University Hospital, Taipei, Taiwan. ⁴College of Medicine, National Taiwan University, Taipei, Taiwan.

Received: 18 September 2021 Accepted: 12 March 2022

Published online: 23 March 2022

References

1. Lauria MR, Gonik B, Romero R. Pulmonary hypoplasia: pathogenesis, diagnosis, and antenatal prediction. *Obstet Gynecol.* 1995;86(3):466–75.
2. Fuloria M, Aschner JL. Persistent pulmonary hypertension of the newborn. *Semin Fetal Neonatal Med.* 2017;22(4):220–6.
3. Arvanitaki A, Giannakoulas G, Baumgartner H, Lammers AE. Eisenmenger syndrome: diagnosis, prognosis and clinical management. *Heart.* 2020;106(21):1638–45.
4. Engelfriet P, Boersma E, Oechslin E, et al. The spectrum of adult congenital heart disease in Europe: morbidity and mortality in a 5 year follow-up period. The Euro Heart Survey on adult congenital heart disease. *Eur Heart J.* 2005;26(21):2325–33.
5. Diller GP, Korten MA, Bauer UM, et al. Current therapy and outcome of Eisenmenger syndrome: data of the German National Register for congenital heart defects. *Eur Heart J.* 2016;37(18):1449–55.
6. Deanfield J, Thaulow E, Warnes C, et al. Management of grown up congenital heart disease. *Eur Heart J.* 2003;24(11):1035–84.
7. Waddell TK, Bennett L, Kennedy R, Todd TR, Keshavjee SH. Heart-lung or lung transplantation for Eisenmenger syndrome. *J Heart Lung Transplant.* 2002;21(7):731–7.
8. Hjortshøj CS, Gilljam T, Dellgren G, et al. Outcome after heart-lung or lung transplantation in patients with Eisenmenger syndrome. *Heart.* 2020;106(2):127–32.

Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Ready to submit your research? Choose BMC and benefit from:

- fast, convenient online submission
- thorough peer review by experienced researchers in your field
- rapid publication on acceptance
- support for research data, including large and complex data types
- gold Open Access which fosters wider collaboration and increased citations
- maximum visibility for your research: over 100M website views per year

At BMC, research is always in progress.

Learn more biomedcentral.com/submissions

