

Neonatal Repair of Total Anomalous Pulmonary Venous Connection with Goldenhar Syndrome and Unilateral Lung Agenesis—A Case Report

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Research Article

Keywords: total anomalous pulmonary venous connection, Goldenhar syndrome, unilateral lung agenesis

Posted Date: May 27th, 2021

DOI: <https://doi.org/10.21203/rs.3.rs-419649/v1>

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Abstract

Background: Total anomalous pulmonary venous connection with unilateral lung agenesis and Goldenhar syndrome is extremely rare and high mortality.

Case presentation: We present a case of total anomalous pulmonary venous connection with unilateral lung agenesis and Goldenhar syndrome. It was diagnosed from Transthoracic echocardiography and enhanced Computed Tomography. Total absence of the lung, the bronchial tree, and vascular structures were detected on the right side, and the left pulmonary veins returned abnormally to the innominate vein. There was apparent indication of pulmonary venous obstruction, the operation was performed at 3 days after birth. The common pulmonary venous chamber with vertical vein and the left atrium was anastomosed using 7-0 PDS running sutures through a median sternotomy. Postoperative echocardiography and Computed Tomography 1 year after the surgery, between the common pulmonary venous chamber and the left atrium was no stenosis.

Conclusion: A extremely rare case of total anomalous pulmonary venous connection with unilateral lung agenesis and Goldenhar syndrome successfully repaired at 3 day after birth was reported. Anastomosis between the common pulmonary venous chamber and the left atrium using the vertical vein is a reasonable choice in patient with small common pulmonary venous chamber.

Background

Goldenhar syndrome is a rare congenital disease characterized by craniofacial abnormalities included the incomplete development of the eye, ear, jaw[1]. The average incidence rate of this syndrome is estimated to be between 1:3000 and 1:5000 live births. Cardiovascular malformations have been reported in 5–58% of patients [1]. The prevalence of lung agenesis is reported to 34 per 1,000,000 live births[2]. Cases of repair for total anomalous pulmonary venous connection with lung agenesis have previously been reported, operative mortality was high. In addition, total anomalous pulmonary venous connection with unilateral lung agenesis and Goldenhar syndrome were reported only 2 cases. We report a rare case of total anomalous pulmonary venous connection with unilateral lung agenesis and Goldenhar syndrome successfully repaired at 3 day after birth. To the best of our knowledge, this is the first report survived more than 1 year after surgery.

Case Presentation

A 25-year-old pregnant woman at 25 weeks of gestation was diagnosed with right lung agenesis and right hypoplastic eye, but heart malformation was not diagnosed.

A female neonate was delivered at 37 weeks with caesarean section. The birth weight of the neonate was 2101g, postnatally, she was noticed craniofacial abnormalities and suspected Goldenhar syndrome. She deteriorated requiring intratracheal ventilation. Chest radiograph, Transthoracic echocardiography and enhanced Computed Tomography confirmed the diagnosis. Total absence of the lung, the bronchial tree,

and vascular structures were detected on the right side, and the left pulmonary veins returned abnormally to the innominate vein(Fig.1 A,B,C). In addition, she had a large arterial septal defect, a small ventricle septal defect and a patent ductus arteriosus. There was apparent indication of pulmonary venous obstruction, the operation was performed at 3 days after birth.

The heart was exposed through a median sternotomy. Cardiopulmonary bypass was instituted with ascending aortic perfusion, direct bicaval venous drainage. The patent ductus arteriosus was ligated. The pulmonary artery venting via the left pulmonary artery, cardiac arrest was achieved using antegrade cardioplegia infusion. We approached from superior of the left pulmonary artery without inversion. The vertical vein was divided and incised to the common pulmonary venous chamber, the posterior wall of left atrium was incised towards the left atrial appendage longitudinally. The common pulmonary venous chamber with vertical vein and the left atrium was anastomosed using 7-0 PDS running sutures. The atrial septal defect was closed directly. The aortic clamp times was 48 minutes. The left and right ventricular function were good with catecholamine supports and nitric oxide, weaning from cardiopulmonary bypass was smooth and the sternum was closed. Postoperative echocardiography and Computed Tomography 1 year after the surgery, between the common pulmonary venous chamber and the left atrium was no stenosis(Fig.2).

Discussion

Goldenhar syndrome is a rare congenital disease and has also been reported cases with cardiac and lung malformations. Most frequent is tetralogy of Fallot, followed by septal defects and situs inversus. Total anomalous pulmonary venous connection with lung agenesis have been reported only 6 cases[2, 3, 4], The combination of total anomalous pulmonary venous connection, unilateral lung agenesis, and Goldenhar syndrome were only two cases, however surgical outcomes were unknown[3, 4]. To the best of our knowledge, this is the only case survived more than 1 year after surgery. Hasegawa and colleagues[4] reported cardiac procedures with lung agenesis were performed through a median sternotomy or a posterolateral thoracotomy, however the posterolateral thoracotomy approach were high mortality. Therefore, this case was performed through a median sternotomy, the total anomalous pulmonary venous connection and the posterior wall of left atrium were well in sight through a superior approach without inversion. The common pulmonary venous chamber and the left atrium was anastomosed using the vertical vein because the common pulmonary venous chamber was small. Following up more than 1 year after surgery, pulmonary venous obstruction due to use of the vertical vein was none.

Conclusion

A rare case of total anomalous pulmonary venous connection with unilateral lung agenesis and Goldenhar syndrome successfully repaired at 3 day after birth was reported. Anastomosis between the common pulmonary venous chamber and the left atrium using the vertical vein is a reasonable choice in patient with small pulmonary chamber.

Abbreviations

LPV: left pulmonary vein

LPA: left pulmonary artery

VV: vertical vein

InnV: innominate vein

SVC: supra vena cava

LUPV: left upper pulmonary vein

LLPV: left lower pulmonary vein

LA: left atrium

LAA: left atrial appendage

PVC: pulmonary venous chamber

Declarations

Ethics approval and consent to participate

Not applicable. Institutional review board was not required was waived for the purpose of this study.

Consent for publication

Informed consent was obtained from the patient.

Availability of data and materials

The data that support the findings of this report are available from Chiba Children's Hospital. The author can make it available upon reasonable request.

Competing interests

There are no competing interests for all authors.

Finding

None

Authors' contributions

Takahiro Ito:ito19880530@yahoo.co.jp: wrote the manuscript and reviewed the literature.

Ikuo Hagino: ihagino@e23.jp : reviewed and edited the paper.

Mitsuru Aoki: m.aoki@outlook.com: reviewed and edited the paper.

Acknowledgments

None

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References

1. Minati Choudhury, Poonam Malhotra Kapoor. Goldenhar Syndrome: Cardiac Anesthesiologist's Perspective. *Ann Card Anaesth.* 2017; 20: S61–S66.
2. Yuji Kaku· Mitsugi Nagashima · Goki Matsumura et al. Neonatal Repair of Total Anomalous Pulmonary Venous Connection and Lung Agenesis. *Asian Cardiovasc Thorac Ann.* 2015;23(6):716-8.
3. Markus Fischer, Matthias Gorenflo, Joachim Eichhorn et al. First description of unilateral lung agenesis and anomalous pulmonary venous connection in a newborn with Goldenhar syndrome. *Cardiology in the Young* (2012), 22, 344–345.
4. Tomomi Hasegawa, MD, Yoshihiro Oshima, MD, Ayako Maruo, MD et al. Pediatric Cardiothoracic Surgery in Patients With Unilateral Pulmonary Agenesis or Aplasia. *Ann Thorac Surg.* 2014 May;97(5):1652-8.

Figures

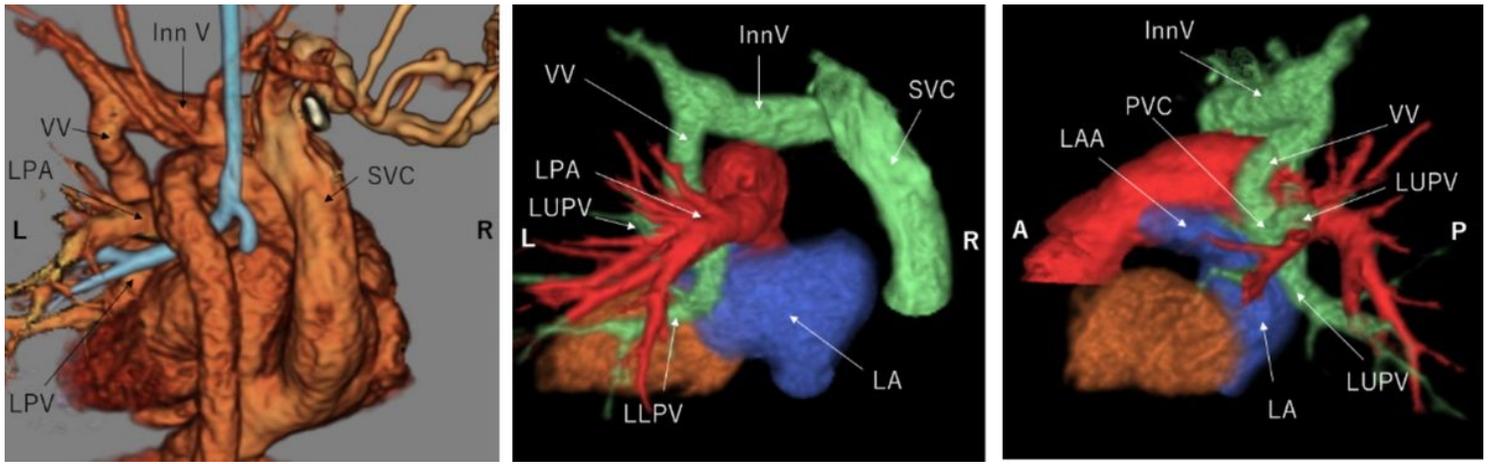


Figure 1

Preoperative 3-dimensional tomography (A) total absence of the right bronchial tree and right vascular structures. (posterior to anterior view) (B),(C) the left pulmonary veins returned abnormally to the innominate vein. (posterior to anterior view and left side to right side view) LPV: left pulmonary vein; LPA: left pulmonary artery; VV: vertical vein; InnV: innominate vein; SVC: supra vena cava; LUPV: left upper pulmonary vein; LLPV: left lower pulmonary vein; LA: left atrium; LAA: left atrial appendage; PVC: pulmonary venous chamber

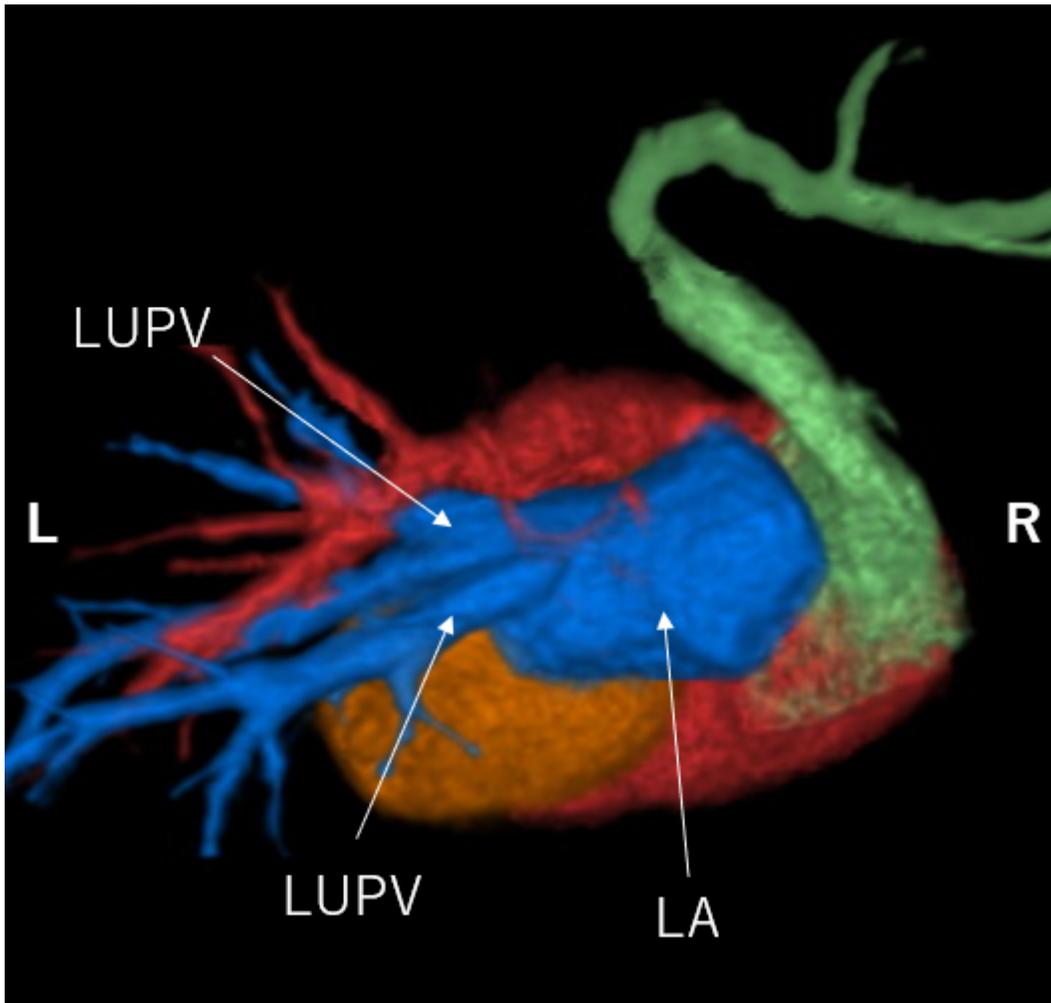


Figure 2

Postoperative 3-dimensional tomography demonstrating the left pulmonary veins connected to left atrium. LUPV: left upper pulmonary vein; LLPV: left lower pulmonary vein; LA: left atrium

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