



Case Report

Effects of preoperative veno-venous extracorporeal membrane oxygenation management and left atrial pressure reduction via balloon atrial septostomy on respiratory system compliance in a neonate with transposition of the great arteries with intact ventricular septum

Keisuke Nishida^{*}, Yusuke Seino, Muneyuki Takeuchi

Department of Critical Care Medicine, National Cerebral and Cardiovascular Center, 6-1, Kishibe-Shimmachi, Suita, Osaka, 564-8565, Japan

ARTICLE INFO

Handling Editor: DR AC Amit Chopra

Keywords:

Respiratory system compliance
Left atrial pressure
Veno-venous extracorporeal membrane oxygenation
Balloon atrial septostomy
Congenital heart disease
Transposition of the great arteries

ABSTRACT

Background: Transposition of the great arteries with intact ventricular septum causes severe hypoxia immediately after birth. Balloon atrial septostomy is often required for oxygenation improvement, and severe cases may require veno-venous extracorporeal membrane oxygenation (VV-ECMO). This case report highlights the critical role of balloon atrial septostomy in improving respiratory system compliance and facilitating early ECMO discontinuation in a neonate with transposition of the great arteries.

Case summary: A neonate with transposition of the great arteries experienced persistent hypoxia, hypotension, and lactic acidosis, prompting VV-ECMO on the first day of life. Echocardiography revealed a restrictive atrial septal defect, and balloon atrial septostomy was performed. This resulted in increased oxygen saturation, improved respiratory system compliance (from 0.19 to 0.43 mL/cmH₂O/kg), and reduced left atrial pressure. ECMO was successfully withdrawn on day 2, which was later followed by a successful Jatene procedure.

Conclusion: Balloon atrial septostomy can significantly improve oxygenation and respiratory system compliance in patients with transposition of the great arteries, enabling rapid VV-ECMO discontinuation and informing future congenital heart disease management.

1. Introduction

Transposition of the great arteries (TGA) with intact ventricular septum is a congenital heart disease characterized by anomalous positioning of the aorta and pulmonary arteries; it leads to cyanosis and severe hypoxia immediately after birth. Timely intervention for TGA is crucial to ensure an adequate oxygen supply and prevent long-term complications. When an atrial septal defect (ASD) is restrictive, balloon atrial septostomy (BAS) becomes a vital intervention that can address immediate postnatal hypoxia with TGA because it facilitates the mixing of oxygenated and deoxygenated blood by creating or enlarging the ASD. For severe cases or those complicated by neonatal persistent pulmonary hypertension (PPHN), veno-venous (VV) extracorporeal membrane oxygenation (ECMO) can provide temporary cardiopulmonary support [1]. Successful weaning from VV-ECMO requires improved oxygenation and

^{*} Corresponding author.

E-mail address: nishida.keisuke@ncvc.go.jp (K. Nishida).

increased respiratory system compliance (CRS). This report describes the case of a neonate with TGA who required VV-ECMO for severe hypoxia management immediately after birth and subsequently underwent BAS.

2. Case description

A male neonate who was prenatally diagnosed with TGA was born at 40 weeks of gestation with a weight of 3400 g. Severe hypoxia developed immediately after birth; therefore, the patient was intubated and admitted to our pediatric intensive care unit. Despite the initiation of inhaled nitric oxide therapy, persistent hypoxia (SpO₂ of approximately 50 %), hypotension (mean arterial pressure of approximately 30 mmHg), and lactic acidosis (20 mmol/L) were observed, indicating PPHN. Because of ongoing shock, VV-ECMO was initiated on the day of birth. After ECMO initiation, the SpO₂ level improved to 80 % with a VV-ECMO flow rate of 120 mL/kg/min, and inotropic support comprising adrenaline and dobutamine was administered to maintain hemodynamic stability. The lactate level rapidly decreased after ECMO initiation.

On postnatal day 1, CRS decreased from 0.26 mL/cmH₂O/kg to 0.19 mL/cmH₂O/kg after ECMO initiation (Fig. 1). The mode of mechanical ventilation was pressure-controlled ventilation (peak inspiratory pressure, 22 cmH₂O; positive end-expiratory pressure, 8 cmH₂O), and the tidal volume decreased to 2.7 mL/kg. Chest radiography showed significant pulmonary congestion, and a repeat echocardiogram revealed a restrictive ASD with left atrial enlargement (Figs. 2 and 3). The decrease in CRS was attributed to pulmonary congestion caused by increased left atrial pressure, likely caused by fluid overload and improved oxygen saturation with VV-ECMO. This reduced pulmonary vascular resistance and increased pulmonary blood flow through the patent ductus arteriosus, further increasing left atrial pressure.

Consequently, BAS was performed on the same day, resulting in adequate interatrial mixing, which was confirmed by follow-up echocardiography. After BAS, the SpO₂ levels increased to 90 %, left atrial size was reduced (Fig. 3), and CRS improved markedly (from 0.19 mL/cmH₂O/kg to 0.43 mL/cmH₂O/kg) within 12 h of the procedure (Fig. 1).

Remarkably, ECMO was successfully withdrawn on postnatal day 2 without the anticipated need for aggressive fluid balance management (Fig. 1) or improvement of edema of the thorax and abdomen. Fluid balance during ECMO was not significantly negative, and early withdrawal was facilitated primarily by the improvement in oxygenation and reduction in left atrial pressure via BAS. Subsequently, the patient underwent the Jatene procedure on postnatal day 15. He was discharged from the pediatric intensive care unit on day 25. On day 46, he was discharged home.

3. Discussion

This case highlights the critical aspects of managing TGA in a neonate by focusing on the role of BAS in improving CRS and oxygenation during VV-ECMO. For typical TGA cases, BAS is performed to improve hypoxia; however, this case suggested that BAS also plays a crucial role in dramatically improving CRS by reducing left atrial pressure. To enable successful weaning from VV-ECMO,

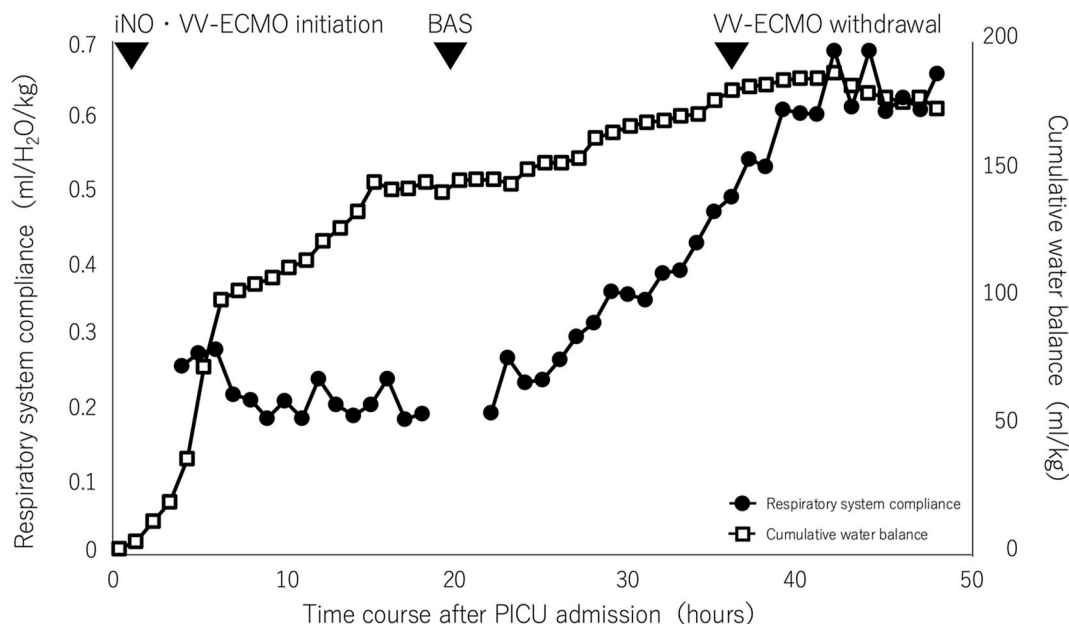


Fig. 1. Changes in respiratory system compliance and cumulative water balance after pediatric intensive care unit admission. Respiratory system compliance was calculated as measured tidal volume/set pressure control level, with no spontaneous breath during this course. BAS, balloon atrial septostomy; iNO, inhaled nitric oxide; PICU, pediatric intensive care unit; VV-ECMO, veno-venous extracorporeal membrane oxygenation.

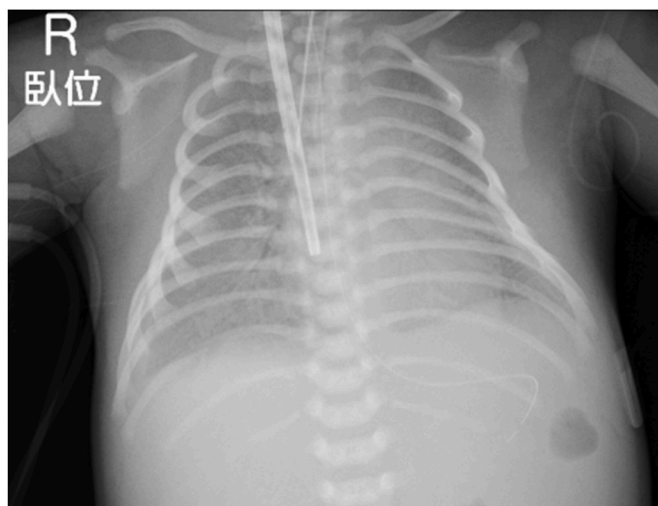


Fig. 2. Echocardiography images of the left atrial size before (left) and after (right) balloon atrial septostomy.

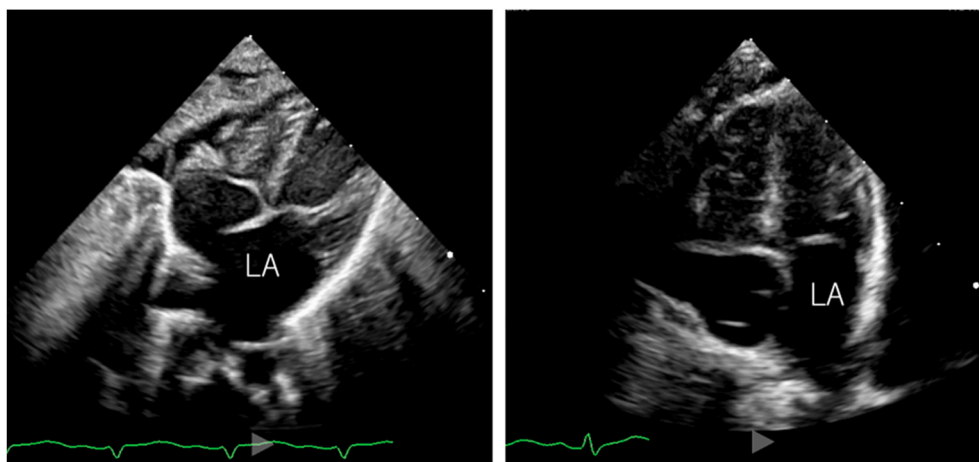


Fig. 3. Radiography image of the chest after initiation of veno-venous extracorporeal membrane oxygenation before balloon atrial septostomy. LA, left atrium.

improvements in both CRS and oxygenation are important [2]. For this patient, BAS provided significant improvements in CRS and oxygenation, via a mechanism distinct from those in typical VV-ECMO cases.

Severe hypoxia in neonates with TGA immediately after birth is often caused by a restrictive ASD, intact atrial septum, or complications of PPHN [1]. Nicholson et al. reported a similar case of a neonate with TGA and PPHN who required VV-ECMO despite an initial improvement in oxygenation after BAS [2]. Although VV-ECMO is occasionally used for neonates with PPHN, several days of weaning are typically required. For instance, one study showed that neonates with PPHN treated with VV-ECMO required an average of 7 days of support [3]. In contrast, our case demonstrated that although PPHN was initially suspected as the primary cause of hypoxia, further evaluation revealed that a restrictive ASD was the main issue. Thus, performing BAS during VV-ECMO improved oxygenation immediately and stabilized the condition of the patient, thereby enabling earlier VV-ECMO discontinuation.

After ECMO initiation, fluid overload and improved oxygenation increased the pulmonary blood flow, leading to increased left atrial pressure, which resulted in pulmonary congestion and decreased CRS. For cases involving a restrictive ASD, this increase in pulmonary blood flow can further increase left atrial pressure, potentially causing the ASD to be more restrictive or even causing it to close [4]. For our case, BAS facilitated interatrial mixing, rapid lowering of left atrial pressure, and significant improvement of CRS, as evidenced by a marked increase in the tidal volume within 12 h after the procedure. This improvement allowed for early ECMO withdrawal, primarily because of reduced left atrial pressure after BAS rather than the usual mechanisms of fluid balance correction and improved chest wall compliance. Typically, correcting the fluid balance with diuretics is necessary to reduce left atrial pressure and improve CRS; however, this process often requires several days. However, in our case, the rapid reduction in left atrial pressure achieved by BAS led to a dramatic improvement in CRS, thus enabling early ECMO withdrawal. Increased left atrial pressure is known

to negatively affect respiratory mechanics [5]. Although left atrial pressure in this case was not directly measured, echocardiography and chest radiography findings supported the hypothesis that BAS-induced reduction of left atrial pressure significantly improved CRS.

In conclusion, this case highlights the effectiveness of VV-ECMO for managing severe hypoxia in a neonate with TGA with intact ventricular septum. Furthermore, it underscores the critical role of BAS in improving oxygenation, reducing left atrial pressure, and improving CRS. The rapid and significant improvement in CRS after BAS, which facilitated early ECMO withdrawal, represents an important finding that may influence future management strategies for neonates with complex congenital heart disease.

CRediT authorship contribution statement

Keisuke Nishida: Conceptualization, Supervision, Writing – original draft, Writing – review & editing. **Yusuke Seino:** Conceptualization, Writing – original draft, Writing – review & editing. **Muneyuki Takeuchi:** Conceptualization, Writing – original draft, Writing – review & editing.

5. Patient consent

The patient's parents provided informed consent for the publication of this case report and accompanying images.

4. Conflicts of interest and source of funding

This work was supported by a grant from JSPS KAKENHI to Keisuke Nishida and Muneyuki Takeuchi (grant number 24K12190). Yusuke Seino declare no conflict of interest.

Declaration of competing interest

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests: Keisuke Nishida reports financial support was provided by JSPS KAKENHI. Keisuke Nishida reports a relationship with JSPS KAKENHI that includes: funding grants. Muneyuki Takeuchi reports a relationship with JSPS KAKENHI that includes: funding grants. The other author declares no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Acknowledgments

We would like to thank Editage (www.editage.jp) for English language editing.

References

- [1] N. Yam, R.H. Chen, B.A. Rocha, K.S. Lun, T.C. Yung, T.W. Au, Preoperative venovenous extracorporeal membrane oxygenation for transposition of great arteries with severe pulmonary hypertension in a newborn, *Ann. Thorac. Surg.* 109 (2020) e329–e330, <https://doi.org/10.1016/j.athoracsur.2019.08.077>.
- [2] C. Maratta, R.M. Potera, G. van Leeuwen, A. Castillo Moya, L. Raman, G.M. Annich, Extracorporeal Life support organization (ELSO): 2020 pediatric respiratory ELSO guideline, *ASAIO J.* 66 (2020) 975–979, <https://doi.org/10.1097/MAT.0000000000001223>.
- [3] D.A. Lazar, D.L. Cass, O.O. Olutoye, S.E. Welty, C.J. Fernandes, P.T. Rycus, T.C. Lee, The use of ECMO for persistent pulmonary hypertension of the newborn: a decade of experience, *J. Surg. Res.* 177 (2012) 263–267, <https://doi.org/10.1016/j.jss.2012.07.058>.
- [4] M.S. Cohen, B.W. Eidem, F. Cetta, M.A. Fogel, P.C. Frommelt, J. Ganame, B.K. Han, T.R. Kimball, R.K. Johnson, L. Mertens, S.M. Paridon, A.J. Powell, L. Lopez, Multimodality imaging guidelines of patients with transposition of the great arteries: a report from the American society of echocardiography developed in collaboration with the society for cardiovascular magnetic resonance and the society of cardiovascular computed tomography, *J. Am. Soc. Echocardiogr.* 29 (2016) 571–621, <https://doi.org/10.1016/j.echo.2016.04.002>.
- [5] R. Hughes, A.J. May, J.G. Widdicombe, The effect of pulmonary congestion and oedema on lung compliance, *J. Physiol.* 142 (1958) 306–313, <https://doi.org/10.1113/jphysiol.1958.sp006017>.