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CASE REPORT

Artificial Lung / ECMO

# Efficacy of "Awake ECMO" for critical respiratory failure after pediatric open-heart surgery

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**Abstract** A 4-year-old boy with atrioventricular discordance, double-outlet right ventricle, pulmonary stenosis, and mitral regurgitation, was undergoing anatomical repair consisting of Senning, Rastelli, Damus–Kaye–Stansel procedures, and a mitral valve repair, complained of post-operative excessive airway tract secretion, which ultimately developed into acute respiratory distress syndrome (ARDS) 28 days after the operation. The cause of the ARDS was thought to be frequent manual positive pressure recruitment and prolonged inhalation of pure oxygen. At 45 days after the operation, hypercapnia and respiratory acidosis turned out to be irreversible, and therefore, veno-arterial extracorporeal membrane oxygenation (ECMO) was established utilizing the Endumo®4000 system. Pulmonic interstitial inflammation gradually improved while resting the lung under ECMO support; however, effective ventilation volume decreased critically because a massive pulmonary hemorrhage occurred at 2 and 9 days after the initiation of ECMO. To maximize the effectiveness of respiratory physical therapy, "Awake ECMO" was started and tidal volume dramatically increased with a regained cough reflex. Five days later, he was successfully weaned off from ECMO, and discharged 7 months after the operation without any neurological and physiological sequelae.

**Keywords** Extracorporeal membrane oxygenation · Pediatric · Endumo

The Awake ECMO procedure had been introduced for patients awaiting lung transplantation under veno-arterial ECMO support to prevent respiratory tract infection or pneumothorax and also to strengthen respiratory muscles [1, 2]. Recent reports have shown that it is a successful bridge to recovery or destination from cardiogenic shock in pediatric populations [3].

Herein, we reported a successful recovery from a post-cardiotomy severe respiratory failure with the Awake ECMO procedure.

## Case

A 4-year-old boy with atrioventricular discordance, double-outlet right ventricle, pulmonary stenosis, and mitral regurgitation underwent anatomical repair consisting of Senning, Rastelli, Damus–Kaye–Stansel (DKS), and mitral valve repair (MVP). The acute post-operative course was good, and the patient was extubated at 1 day after the operation. However, excessive airway tract secretion continued, and he was thus re-intubated 2 times. After that, still frequent manual positive pressure recruitment and prolonged inhalation of pure oxygen with noninvasive positive pressure ventilation (NPPV) could not be weaned off. Mild-to-moderate residual mitral regurgitation and dynamic obstruction of pulmonary venous pathway just after Senning procedure were thought to be a contributing

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cause of the congestive respiratory failure. The patient finally developed acute respiratory distress syndrome (ARDS) 28 days after the initial operation (Fig. 1a), and then, at 45 days after the operation, hypercapnia and respiratory acidosis became irreversible, and therefore, veno-arterial extracorporeal membrane oxygenation (ECMO) was established with an Endumo®4000 system (Fig. 1b).

Endumo 4000 system consisted of BIOCUBE 4000® (NIPRO, Osaka, Japan) as artificial lung with membranous surface area of 0.8 $m^2$, Rotaflow® (Maquet, Germany) as centrifugal pump, and Circuit was coated by heparin using T-NCVC coating® (TOYOBO, Osaka, Japan). The priming volume was 380 mL and maximum bypass flow is 3.0 L/min. A 12-Fr heparin-coated cannula for femoral arterial cannulation (Edwards Lifescience Duraflo II Femoral catheter FEM II type®, Edwards Lifesciences, Irvine, California) was cannulated from right internal carotid artery, and a 14-Fr heparin-coated cannula for femoral venous cannulation (Edwards Lifescience Duraflo II Femoral catheter FEM II type®, Edwards Lifesciences, Irvine, California) was cannulated from right internal jugular vein. These cannulae were connected to the ECMO circuit with non-heparin-coated connectors. A non-heparin-coated shunt tube was connected between the arterial and venous tubes, with heparin-coated t-shaped stopcocks. Heparin was continuously administered to control activated coagulation time of more than 180 s.

During ECMO support, the mechanical ventilator was set lower: at 5 $mH_2O$ positive end-expiratory pressure (PEEP) and at 15 $cmH_2O$ peak inspiratory pressure (PIP) to rest of the lung (Table 1). Pulmonic interstitial inflammation gradually improved; however, the effective ventilation volume critically decreased because massive pulmonary hemorrhages had occurred at 2 and 9 days after the initiation of ECMO. Under deep sedation without the coughing reflex, atelectasis did not improve, so "Awake ECMO" was started at 16 days after the initiation of ECMO support, to maximize the effectiveness of respiratory physical therapy (Table 1). Activated coagulation time was controlled at between 160 and 180 s. By maintaining Ramsay sedation scale 4 with continuous intravenous infusion of midazolam hydrochloride (0.2–0.4 mg/kg/h) and morphine hydrochloride (1–2 μg/kg/min), the coughing reflex was recovered and effective tidal volume improved with aggressive respiratory physical therapy using biphasic cuirass ventilation (RTX®) (Fig. 2a, b). Four days after the initiation of Awake ECMO, he was successfully weaned off from ECMO. It took a long time for the patient to complete physiotherapy and respiratory rehabilitation, but he was finally discharged at 7 months after the operation without any neurological and physiological sequelae. A post-operative catheter examination 1 year after the operation showed less than mild mitral regurgitation and a pulmonary capillary wedge pressure of 10 mmHg.

## Comment

Safe long-term use of the Endumo system without circuit exchange and the possibility of "Awake ECMO" under such stable ECMO support had been already reported [4]; however, this research reports the first successful weaning off from ECMO support after severe respiratory failure following a complex congenital heart surgery using the "Awake ECMO" procedure. Although the recovery of respiratory function was mainly achieved through the improvement of ARDS by resting lung therapy under veno-arterial ECMO support, there is no doubt that the recovery from critically reduced tidal volume incurred due to a subsequently occurring massive pulmonary hemorrhage was truly obtained by the effective respiratory physical therapy with the "Awake ECMO" procedure.

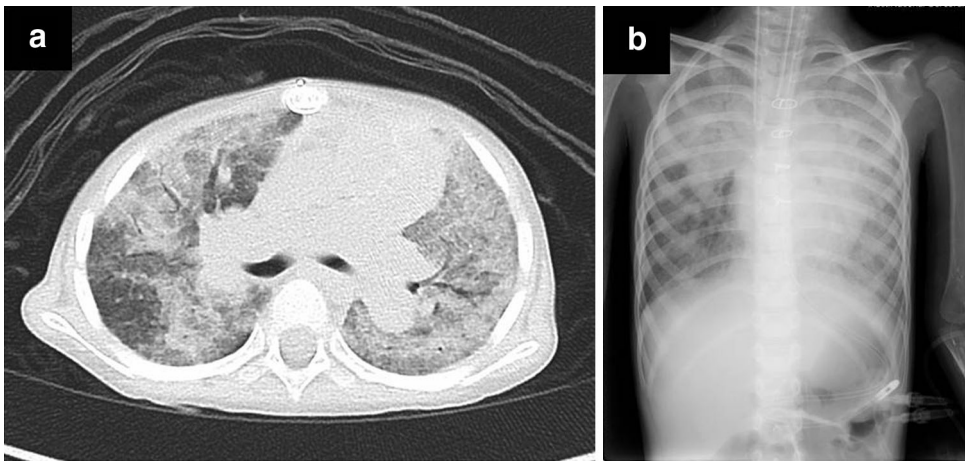


**Fig. 1** Chest computed tomography just before ECMO cannulation representing a massive interstitial fibrotic change (**a**). Chest X-ray just after the initiation of ECMO (**b**)

**Table 1** Setting of ECMO, hemodynamic and respiratory indices

| Event | Days after ECMO initiation | | | | | |
|---|---|---|---|---|---|---|
| | 0 (before initiation) | 0 (after initiation) | 15 | 16 (awake day 0) | 19 (awake day 4) | 20 (decannulation) |
| ECMO driving condition | | | | | | |
| Rpm | – | 2830 | 2660 | 2660 | 2660 | – |
| Supported blood flow without shunt (L/min) | – | 1.0 | 1.3 | 1.3 | 1.3 | – |
| $O_2$ flow (L/min) | – | 1.0 | 2.5 | 2.5 | 3.0 | – |
| $FiO_2$ | – | 0.8 | 0.8 | 0.8 | 0.8 | – |
| Settings of mechanical ventilation | | | | | | |
| $FiO_2$ | 0.9 | 0.21 | 0.5 | 0.5 | 0.5 | 0.40 |
| PEEP (mmHg) | 5 | 5 | 5 | 5 | 5 | 5 |
| Pressure support ($cmH_2O$) | 25 | 15 | 15 | 15 | 15 | 14 |
| Respiratory frequency (/min) | 26 | 15 | 15 | 15 | 15 | 28 |
| Tidal volume (mL) | – | 52 | 71 | 102 | 121 | 216 |
| Peak inspiratory pressure ($cmH_2O$) | – | 21 | 21 | 21 | 19 | 21 |
| Hemodynamics | | | | | | |
| Arterial blood pressure (mmHg) | 90/50 (65) | 90/70 (75) | 110/70 (80) | 95/65 (70) | 100/65(75) | 95/65 (70) |
| CVP (mmHg) | – | 5 | 10 | 9 | 7 | 9 |
| Heart rate (/min) | 155 | 80 | 90 | 80 | 85 | 85 |
| The blood gas data | | | | | | |
| pH | 7.27 | 7.476 | 7.45 | 7.46 | 7.45 | 7.40 |
| $PO_2$ (mmHg) | 43.5 | 174 | 168 | 240 | 236 | 111 |
| $PCO_2$ (mmHg) | 62.1 | 42.2 | 39.6 | 37.9 | 39.6 | 40.3 |
| P/F ratio | 43.5 | 217 | 210 | 300 | 295 | 277 |
| Base excess (mmol/L) | 1.8 | 7.0 | 3.3 | 3.2 | 3.3 | 0.5 |

*ECMO* extracorporeal membranous oxygenation, *Rpm* revolutions per minute, *PEEP* positive end-expiratory pressure, *CVP* central venous pressure

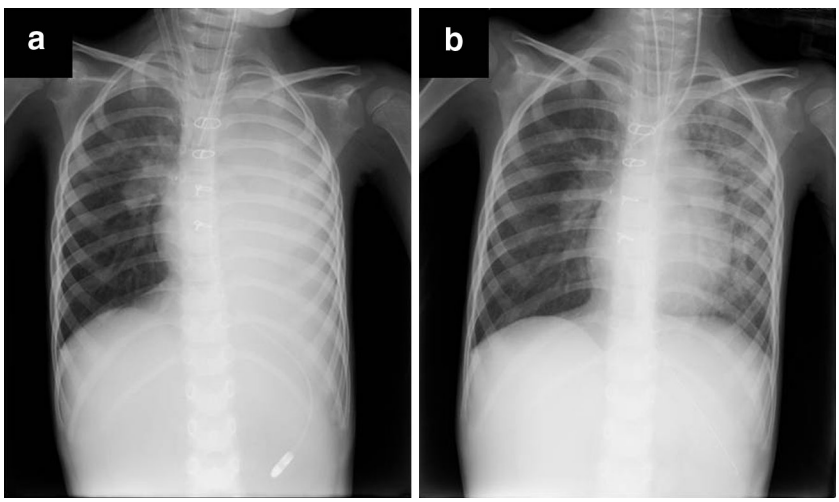


**Fig. 2** Chest X-ray before (**a**) starting "Awake ECMO" and just after (**b**) successful weaning off from ECMO

Even though surgical outcomes of congenital heart surgery have been gradually improving over the past two decades, post-operative respiratory problems are still the second main cause of death [5]. ARDS is one of the most serious respiratory complications; however, its etiology is often unclear, and conventional treatments such as steroid pulse therapy are sometimes ineffective. Whereas the initiation of veno-venous ECMO was reported to be an

effective therapy for ARDS [6], veno-arterial ECMO was selected for the present case because ventricular volume unloading and the volume unloading of pulmonary circulation had been expected to attenuate mitral regurgitation and also dynamic pulmonary venous obstruction, which were thought to be the causes of the ARDS.

Because blood–gas exchange should be fully supported by ECMO during Awake ECMO procedure, it is essential to maintain adequate sedation level to obtain comfortable patient's condition and prevent accidental kinking, dislocation, or discontinuation of ECMO circuit. As described above, administration of morphine hydrochloride as an analgesic agent in addition to midazolam hydrochloride is our preferred regimen for sedation, then intensive care unit nurse specialists assess and control patient's sedation level to Ramsey sedation scale 4, for recovery of the cough reflex.

At the time of writing this report, ventricular assist devices for pediatric patients are being introduced in Japan, but the institutions allowed to use them are limited, and there is a serious shortage of donors for heart transplants in pediatric patients. In order to recover from post-operative cardiopulmonary dysfunction after congenital heart surgery, the development of a durable and safe ECMO system and the effective use of ECLS like the "Awake ECMO" procedure may be used to rescue more patients in the future.

**Compliance with ethical standards**

**Conflicts of interest** None.

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