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CASE REPORT

Cardiopulmonary Bypass

Extracorporeal membrane oxygenation in spina bifida and (H1N1)-induced acute respiratory distress syndrome

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Abstract Acute respiratory distress syndrome (ARDS) is characterized as an acute hypoxemic and/or hypercapnic respiratory failure seen in critically ill patients and is still, although decreased over the past few years, associated with high mortality. Furthermore, ARDS may be a life-threatening complication of H1N1 pneumonia. We report on a 45-year-old spina bifida patient with confirmed H1N1 influenza virus infection causing acute respiratory failure, who was successfully weaned from 42-day veno-venous extracorporeal membrane oxygenation (vv-ECMO) treatment with an excellent outcome. Due to the physical constitution of spina bifida patients, we experienced challenges concerning cannula positioning and mechanical ventilation settings during weaning.

Keywords H1N1 · Acute respiratory distress syndrome · Extracorporeal membrane oxygenation · Spina bifida · Spinal cord injury

Abbreviations

ARDS	Acute respiratory distress syndrome
SCI	Spinal cord injury
ECMO	Extracorporeal membrane oxygenation
ICU	Intensive care unit
MV	Mechanical ventilation
MRI	Magnetic resonance imaging
CT	Computed tomography

Introduction

Acute respiratory distress syndrome (ARDS) is a severe respiratory condition characterized by diffuse inflammation of alveolar and vascular (capillary) lung structures leading to progressive hypoxemia and/or hypercapnia.

The American–European consensus conference definition has been applied since its publication in 1994 and has helped to improve knowledge about ARDS. However, in 2011, the European intensive medicine society agreed on the Berlin definition, which is now widely applied and accepted in diagnosis of ARDS [1]. In the management of patients with ARDS, extracorporeal membrane oxygenation (ECMO) has been successfully used as salvage therapy. ARDS severe enough to require ECMO therapy is estimated to occur in nearly 5–10 cases per million population per year [2]. The effectiveness of ECMO in ARDS patients with pneumonia, influenza A (H1N1), and/or trauma has recently been described and considered promising [3, 4], although is still associated with higher mortality [5].

The addition of prone positioning therapy to ECMO may improve alveolar recruitment and, therefore, reduce ventilator-induced lung injury [6, 7].

Adding prone positioning therapy to ECMO patients is recommended by the guidelines for adult respiratory failure from the extracorporeal life support organization if radiological imaging shows posterior consolidation of the lung fields [8].

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Case presentation

A 45-year-old (height 170 cm and 75 kg bodyweight) chronic paraplegic patient due to spina bifida presenting an extraordinary kyphoscoliosis (see Fig. 1) was admitted to the intensive care unit (ICU) after onset of an acute respiratory failure due to H1N1 influenza.

Prior to ARDS onset, the patient underwent urologic treatment for pyelonephritis that had led to sepsis and required intensive medical care. As respiratory failure proceeded and pulmonary function could not be maintained despite extensive mechanical ventilation (MV), the patient met the criteria for extracorporeal membrane oxygenation (ECMO) and has, therefore, been transferred to our hospital (ECMO center).

According to the patients' medical history, a magnetic resonance imaging (MRI) conducted years ago showed no signs of Chiari malformation.

Microbial testing revealed H1N1 virus infection as the probable cause for the ongoing ARDS.

His chest X-ray and computed tomography (CT) showed diffuse bilateral infiltration, and blood gas test showed a severe hypoxemia and hypercapnia refractory to the conventional MV (Table 1; Fig. 2).

On the day of admission and after completion of diagnosis, we percutaneously (right jugular vein 17 French size + right femoral vein 21 French size) administered a veno-venous extracorporeal membrane oxygenation. Cannula (Maquet HLS, Rastatt, Germany) positioning has been verified immediately using ultrasound, but was challenging due to a monstrous thoracic scoliosis.

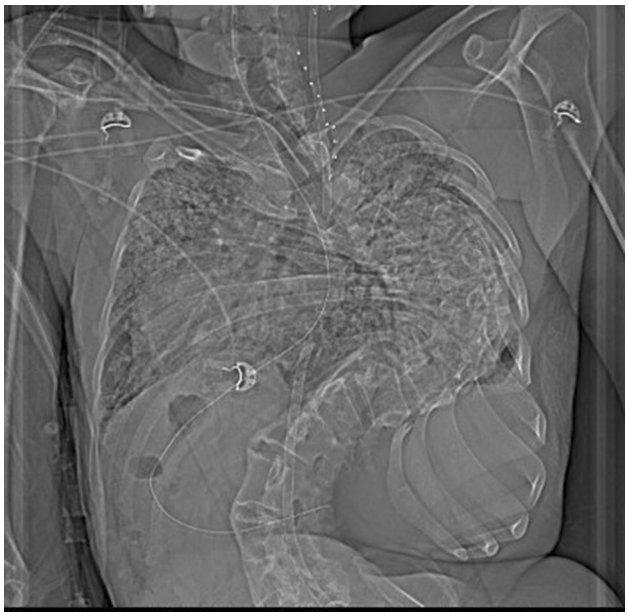

Fig. 1 Chest and abdomen X-ray overview in anteroposterior view. Significantly deforming scoliosis

Table 1 MV settings and blood gas test

	At time of admission	12 h of ECMO	After ECMO removal
Mode	BIPAP	BIPAP	CPAP/ASB
FiO_2	1.0	0.4	0.4
RR	26/min	16/min	22/min
PEEP	17 mbar	15 mbar	9 mbar
Pinsp	32 mbar	27 mbar	14 mbar
pH	7.29	7.43	7.38
pCO_2	70 mmHg	48 mmHg	51 mmHg
pO_2	110 mmHg	81 mmHg	85 mmHg
SO_2%	98	96	97
BE	+4	+8	+4

MV mechanical ventilation, *BIPAP* biphasic positive airway pressure, *CPAP* continuous positive airway pressure, *ASB* assisted spontaneous breathing, *FiO_2* fraction of inspired oxygen, *RR* respiratory rate, *PEEP* positive end-expiratory pressure, *Pinsp* inspiratory pressure, *pH* potential of hydrogen, *pCO_2* partial pressure of carbon dioxide, *pO_2* partial pressure of oxygen, *So_2%* oxygen saturation, *BE* base excess

The cannulation is always performed in cooperation with the cardiotechnology following a standardized protocol.

Subsequently, MV settings were adjusted to ensure protective ventilation.

Because of the patients distinct physical constitution, the pump flow strongly depended on the positioning of the patient. Very small positional changes (head tilt and rotation, hip flexion, or rotation) caused severe, recurrent, and prolonged episodes of decreased blood flow rate and increased negative pressure of the venous inflow, leading to an inadequate systemic oxygenation and to an increased risk of intra-oxygenator blood clotting. Therefore, patient and cannula positioning had to be adapted constantly.

Once the negative pressure exceeded a limit value and subsequently ECMO blood flow decreased, we immediately tried to ensure optimal suction of the ECMO cannula by small changes in the position of the body. In the event of persistent insufficient blood flow, we performed whole body position changes (e.g., head-low, legs-up, 60/90/120°). Due to the enormous physical deformation, especially, whole body changes proved to be challenging and required partly improvised support aids.

Due to that there is no standardized protocol/regimen available, concerning ECMO therapy and positioning therapy in patients suffering challenging deformity, we adapted to incident by trial and error but agreed on the following scheme to solve the recurrent inflow pressure problems:

1st step checking for kinking of the cannules;

2nd step minimal body changes, e.g., head tilt and rotation, hip flexion, or rotation;

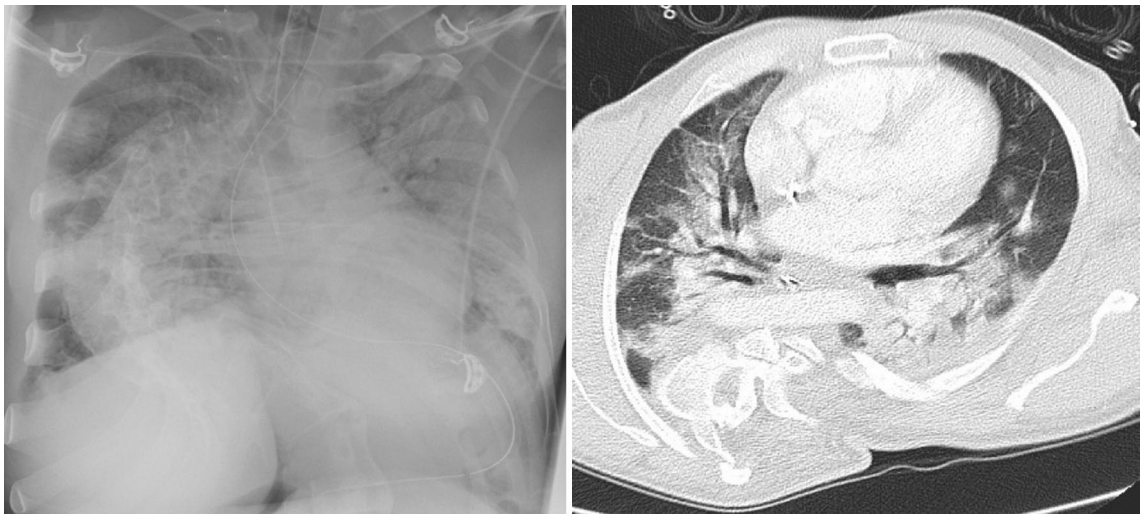

Fig. 2 Chest X-ray in anteroposterior view and CT scan in supine position at the time of admission. Bilateral opacities, posterior consolidation of lung fields

3rd step whole body position changes, e.g., head-low, legs-up, 60/90/120°.

In the initial stage, moving the patients' body/body positioning therapy was conducted only to maintain or improve ECMO blood flow (see scheme above). On the 5th day of ECMO therapy as episodes of decreased blood flow rates persisted and one oxygenator already had to be replaced, the blood flow direction was reversed. Although the pump flow remained strongly dependent on the patients position, the incidence and duration of decreased blood flow periods decreased. However, until termination of ECMO therapy and due to clotting, a total of three oxygenators had to be replaced as the patient highly depended on the extracorporeal oxygenation.

Meanwhile, the patients underwent percutaneous dilatational tracheostomy and as septic multi organ failure proceeded received continuous renal replacement. As ECMO blood flow could be reduced and, therefore, resulted in less inflow pressure problems and less dependency on the patients' position, we began to establish a regimen of intermittent prone positioning therapy to improve alveolar recruitment of the posterior consolidated lung fields and, therefore, pulmonary capacity (Fig. 3).

As the respiratory function improved, ECMO support was gradually decreased and terminated after 42 days. Prior to ECMO termination, blood flow has been lowered to two l/min and oxygen flow down to 0 l/min (withdrawal trial).

2 days after ECMO removal, we transferred the patient at his own wish closer to his home to a clinic specialized on prolonged weaning from MV.

At the time of discharge from our hospital, he was able to breath spontaneously for short periods of time depending

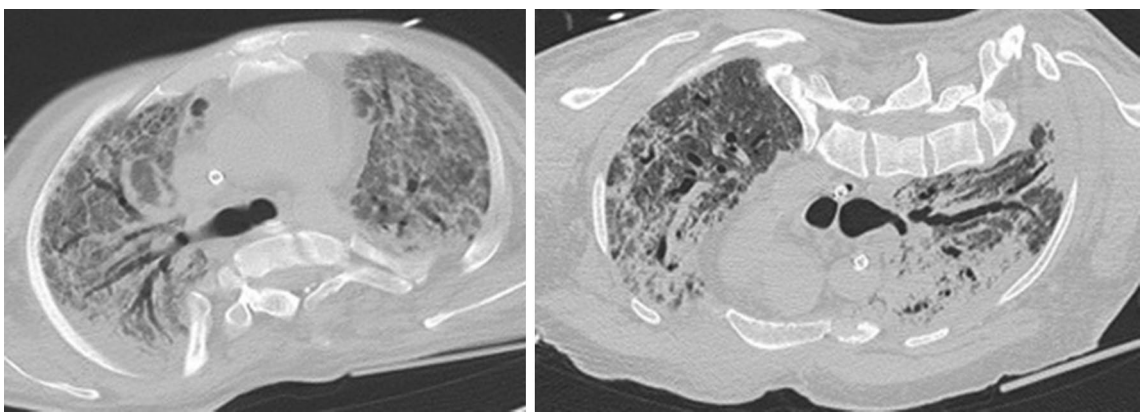

Fig. 3 CT scan in supine (left) and prone position (right). Alveolar recruitment of dorsal lung fields in prone position, starting consolidation of anterior lung fields

on oxygen insufflation, showed no cognitive impairment and could already sit in his wheelchair.

After 4 additional weeks, the patient was discharged to a rehabilitation center, breathing spontaneously and being able to mobilize himself in his wheelchair.

Discussion

Severe kyphoscoliosis causing restrictive respiratory impairment may result or predispose acute respiratory failure [9]. In addition, spinal cord injury (SCI), due to several factors, e.g., reduction in respiratory muscle strength, fatigue (motor impairment), retention of secretions (ineffective coughing) [10] aggravate the risk of respiratory failure.

Although pulmonary complications in SCI patients are common, yet, there is insufficient evidence about their management; current practice is mainly based on clinical experience and expert opinion [11].

ECMO is increasingly emerging into the ICUs and ARDS therapy. Depending on the physical constitution of the patient, ECMO therapy and/or cannula positioning may be more challenging in patients suffering physical deformities. Technical complications of ECMO therapy responsible for acute pump or circuit dysfunction are fortunately not common, but if so may cause devastating ECMO therapy failure [12].

Literature review on ECMO in SCI, spina bifida, or severe kyphoscoliosis reveals a lack of evidence. To our knowledge, only one case series ($n=7$) of ECMO in SCI addresses this specific issue [13].

The intention was to provide the experiences of long-term ECMO therapy in a spina bifida patient. Although the patient of this present case suffered severe technical complications due to spina bifida deformity, ECMO is feasible and the outcome was excellent.

While experiencing recurrent episodes of increased negative pressure of the venous inflow and decreased blood flow rates in the initial stage, we applied positioning therapy (e.g., 60/90 and 120° positioning) to resolve the inflow complications. Alternatively, the addition of a second venous inflow cannula may significantly increase blood flow rate and decrease negative pressure (suction) of the venous inflow line [14].

However, at the end of the ECMO treatment and once the patients' pulmonary function had been improved/partially restored and the patient showed less dependency on ECMO, we were able established a positioning therapy regimen including the prone position as an adjunct to ECMO without experiencing any technical problems. Similarly, a systematic review by Culbreth et al. also reported limited complications and highlighted the clinical efficacy, although evidence is still insufficient [15].

However, both prone positioning and ECMO are well established in treating refractory hypoxemia in ARDS. Combining these therapies may increase the potential risk of cannula dislodgement. This case report suggests that these therapy options can be used in tandem despite considerable physical deformities.

Compliance with ethical standards

Conflict of interest All the authors of this article report that they have no conflict of interest.

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