Original Article

Comparison of the use of venovenous extracorporeal membrane oxygenation in anti-melanoma differentiation-associated protein 5 positive dermatomyositis and other systemic rheumatic diseases associated with acute respiratory failure based on a single-center retrospective study

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Background: Systemic rheumatic diseases (SRDs), particularly anti-melanoma differentiation-associated protein 5 positive dermatomyositis (MDA5$^+$ DM), often affect the respiratory system and have a predisposition for developing into acute respiratory failure (ARF). Venovenous extracorporeal membrane oxygenation (VV-ECMO) can provide full respiratory support and can be used as a life-saving intervention. The present study describes the clinical profiles and prognoses of patients with MDA5$^+$ DM and other SRDs receiving VV-ECMO for ARF.

Methods: A single-center retrospective study of patients with SRD who received VV-ECMO between June 2017 and February 2022 was conducted. Demographic and laboratory data, treatments, extracorporeal membrane oxygenation (ECMO) parameters, and clinical outcomes were extracted from electronic medical records and compared between patients with MDA5$^+$ DM and other SRDs.

Results: Seven patients with MDA5$^+$ DM and four patients with other SRDs were included in the study. Treatment by ECMO was provided for 152 days. Only one patient experienced ECMO-related complications. Three patients in the other SRD group survived to ECMO decannulation, and two of these patients survived to discharge. However, no patients in the MDA5$^+$ DM group survived to decannulation or discharge.

Conclusions: Treatment by VV-ECMO could be safely applied to patients with SRDs to maintain normal respiration and oxygenation. However, patients with MDA5$^+$ DM associated with ARF who underwent VV-ECMO had worse outcomes.

Keywords: Venovenous extracorporeal membrane oxygenation (VV-ECMO); anti-melanoma differentiation-associated protein 5 positive dermatomyositis (MDA5$^+$ DM); systemic rheumatic diseases (SRDs); acute respiratory failure (ARF)

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Introduction

Systemic rheumatic diseases (SRDs) are a set of infrequent and heterogeneous disorders, including systemic lupus erythematosus, systemic sclerosis, primary Sjögren's syndrome, dermatomyositis, and systemic vasculitides, which commonly involve multiple organs and require immunosuppressant therapy and intensive care unit (ICU) management (1). The respiratory system, which covers respiratory compartments such as lung parenchyma, alveolar capillaries, and lung interstitium, has been identified as the system most vulnerable to SRDs (2). There are reports of up to 25% of patients with SRDs requiring hospitalization during the disease, and one third of them are likely to be admitted to the ICU, mainly due to respiratory failure. Respiratory failure carries a substantial risk of morbidity and mortality amongst SRDs, secondary to lung infections, rapidly progressive interstitial lung disease (RPILD), and acute respiratory failure (ARF) (1).

Anti-melanoma differentiation-associated protein 5 positive (MDA5$^+$) dermatomyositis (MDA5$^+$ DM) is characterized by a high frequency of RPILD and is recalcitrant to immunosuppressive therapies. It is also associated with mortality rates above 30% (3), with the majority of the patients succumbing to refractory respiratory failure (4). However, recent advances in the management of respiratory failure indicate that, when standard therapy fails, venovenous extracorporeal membrane oxygenation (VV-ECMO) remains a life-saving therapy in patients with MDA5$^+$ DM experiencing life-threatening hypoxemia or respiratory acidosis. Treatment with VV-ECMO, a well-known rescue technique, can temporarily replace lung function in critically ill patients with multiple lung diseases (5). It can also serve as a bridge to recovery or to organ transplantation, as previously reported in MDA5$^+$ DM patients with Deitchman (6) and by Bay *et al.* (7). To date, comparative studies of clinical characteristics, VV-*ECMO* parameters, adverse events, and outcomes of patients with MDA5$^+$ DM and those with other SRDs associated with ARF remain scarce.

Therefore, we undertook this study to depict the clinical profiles and VV-ECMO practice in SRD patients with respiratory failure and compare the clinical outcomes of patients with MDA5$^+$DM and those with other SRDs who received VV-ECMO. We present this article in accordance with the STROBE reporting checklist (available at https://jtd.amegroups.com/article/view/10.21037/jtd-24-650/rc).

Highlight box

Key findings

- Treatment by venovenous extracorporeal membrane oxygenation (VV-ECMO) could be safely applied to patients with systemic rheumatic diseases (SRDs) to maintain normal respiration and oxygenation.

What is known and what is new?

- Comparative studies of clinical characteristics, VV-ECMO parameters, adverse events, and outcomes of patients with anti-melanoma differentiation-associated protein 5 positive dermatomyositis (MDA5$^+$ DM) and those with other SRDs associated with acute respiratory failure (ARF) remain scarce.
- We undertook this study to depict the clinical profiles and VV-ECMO practice in SRD patients with respiratory failure and compare the clinical outcomes of patients with MDA5$^+$ DM and those with other SRDs who received VV-ECMO.

What is the implication, and what should change now?

- VV-ECMO may be considered an effective and safe supportive therapy and should be initiated early for patients with SRDs who develop ARF.

Methods

Study design and patient population

The present study retrospectively reviewed the records of adult patients with MDA5$^+$ DM or other SRDs who received VV-ECMO for ARF at the Shanghai Jiaotong University School of Medicine, Renji Hospital, between June 2017 and February 2022. The diagnosis of MDA5$^+$ DM was based on clinical and radiologic evidence of DM and interstitial lung disease (ILD) and the presence of anti-MDA5 antibodies.

The diagnosis of ILD was made according to the respiratory symptoms and the presence of bibasilar infiltrates on high-resolution computed tomography (HRCT) as follows: (I) subjective worsening of dyspnea within the past month; (II) appearance of new ground-glass opacities or consolidation on chest radiograph or HRCT; (III) evidence of hypoxemia, as defined by worsened or severely impaired gas exchange; (IV) no evidence of infection, as indicated by negative respiratory culture and serological test results for respiratory pathogens; and (V) no clinical evidence of pulmonary embolism congestive heart failure or pneumothorax as a cause of acute worsening.

The definition of ARF was progressive respiratory failure with new and worsening infiltrates visible on HRCT

imaging over the preceding three months, not readily explained by an infectious process or pulmonary edema. The diagnostic criteria were: mild (200 mmHg < PaO_2/FIO_2 ≤300 mmHg), moderate (100 mmHg < PaO_2/FIO_2 ≤200 mmHg), and severe (PaO_2/FIO_2 ≤100 mmHg) and 4 ancillary variables for severe ARDS: radiographic severity, respiratory system compliance (≤40 mL/cmH_2O), positive end-expiratory pressure (≥10 cmH_2O), and corrected expired volume per minute (≥10 L/min).

We excluded ARF caused by pulmonary infection identified through laboratory tests, imaging investigations, and the test of alveolar lavage fluid. Cardiogenic respiratory failure was excluded by examination of patients with echocardiography or brain natriuretic peptide. This study was conducted in accordance with the declaration of Helsinki (as revised in 2013). The study was approved by the ethics committee of Renji Hospital Affiliated to the Shanghai Jiaotong University School of Medicine and individual consent for this retrospective analysis was waived.

The primary endpoint was patients successfully weaned off extracorporeal membrane oxygenation (ECMO) defined as survival to 24 h after ECMO decannulation (8). The secondary endpoint was in-hospital mortality. VV-ECMO protocol was used with flow rate of 4–4.5 L/min, speed of 3,000 rpm, FIO_2 of 100%, heparin content of 2.5–2.7 U/kg/h. Withdrawal strategy was as the following: after confirming that the primary disease had been controlled and significant improvement had been confirmed by computed tomography (CT), ventilator was adjusted to the optimal support level, model lung ventilation was closed, and the shutdown test was conducted. If the patient had stable hemodynamics, sufficient gas exchange (dynamic review of arterial blood gas), and ventilator parameters were stable, withdrawal could be considered.

Data collection

The following information was collected in standardized forms: demographic data, clinical parameters of patients with MDA5$^+$ DM and other SRDs, rheumatism-specific antibodies, pre-ECMO data and laboratory findings, indications, and complication(s). Respiratory ECMO Survival Prediction and Predicting Death for Severe Acute Respiratory Distress Syndrome on VV-ECMO scores (survival predictors in VV-ECMO patients), ICU therapeutics, prehospital and in-hospital agents (particularly immunosuppressive medications), ECMO-weaning status, complications of primary disease, and ICU and hospital stays. HRCT status was evaluated through the pulmonary HRCT score referred to the previous scoring standard (9,10).

Statistical analysis

Categorical variables were expressed as percentages, while continuous or non-normally distributed variables are expressed as medians {25–75th percentile interquartile range [IQR]}. Statistical analyses were performed using the GraphPad Prism version 8.0 software (GraphPad Software, La Jolla, CA, USA).

Results

General patient characteristics and extracorporeal membrane oxygenation-related treatment parameters

Seven patients with anti-MDA5$^+$ DM and RPILD {3:4 female to male ratio, median age: 58 [52–64] years}, and four patients with ARF and other SRDs with three cases of myositis and one case of lupus {2:2 female to male ratio, median age: 40 [28–63] years} requiring ICU admission, received VV-ECMO. None of the patients was a smoker. Comorbid illnesses which might affect the lungs include lung infection disease, acute pulmonary edema and drug-related lung injury. Patient demographics, characteristics, ECMO-related treatment parameters, and comorbid illnesses which might affect the lungs are shown in *Table 1*.

Treatments and outcomes of patients with MDA5$^+$ DM or other SRDs

Treatments and outcomes of patients in the two groups are shown in *Table 2*.

Of the seven patients in the MDA5$^+$ DM group receiving VV-ECMO, six were given mechanical ventilation and vasopressors, and two patients needed renal replacement therapy. Of the four patients in the other SRD group, one needed biphasic intermittent positive airway pressure ventilation, while three patients needed mechanical ventilation, vasopressors, and renal replacement therapy.

During admission, the seven patients in the MDA5$^+$ DM group received immunosuppressive therapy and medium doses of methylprednisolone, intravenous immunoglobulins (IVIG), and Janus kinase inhibitors (JAKi, tofacitinib 5 mg twice daily) treatment. No obvious adverse reactions (such as thrombosis or infection) were observed in any patients treated with JAKi during the hospitalization.

Additionally, in the MDA5$^+$ DM group, one patient

Table 1 Baseline characteristics of the patients with MDA5$^+$ DM and other SRDs

Variables	MDA5$^+$ DM (n=7)	Other SRDs (n=4)
Age, years	58 [52–64]	40 [28–63]
Female	3 [43]	2 [50]
BMI, kg/m^2	23.5 [20.6–23.8]	24 [21–27]
Comorbidity		
HBsAg positive	1 [14.3]	–
Diabetes	1 [14.3]	–
Clinical course		
Duration of ILD presence before admission, days	60 [45–60]	110 [93–165]
Days from symptom onset to admission	7 [7–20]	14 [12–15]
Days from admission to ECMO	12 [3–42]	15 [4–26]
Rheumatism specific antibody		
Anti-MDA5	7 [100]	0
Anti-Ro52	5 [67]	3 [75]
Anti-SSA	1 [14]	1 [25]
Anti-ribosomal P	NA	1 [25]
Anti-Smith	NA	1 [25]
dsDNA	NA	1 [25]
Laboratory data		
ESR, mm/h	40 [21–51]	15 [2–57]
PCT, μg/L	0.13 [0.07–1.09]	0.57 [0.16–3.41]
CRP, mg/L	12 [3–35]	67 [8–131]
WBC, ×10^9/L	9 [8–11]	21 [5–26]
Lymphocyte count, ×10^9/L	0.42 [0.22–0.65]	1.09 [0.15–2.13]
CK, U/L	63 [35–268]	174 [30–730]
Creatinine, μmol/L	47 [34–65]	112 [61–153]
LDH, U/L	736 [601–1,049]	727 [411–1,082]
Ferritin, ng/mL	1,690 [1,500–3,216]	597 [323–909]
CD4 count, cells/μL	68 [42–218]	49 [9–341]
CD8 count, cells/μL	31 [17–80]	63 [16–488]
BNP, pg/mL	105 [27–210]	202 [116–262]
Illness severity		
APACHE II	27 [16–29]	27 [20–39]
SOFA score	10 [9–14]	13 [7–14]
VV-ECMO score		
RESP score	–3 [–4 to 1]	3 [–5 to 4]
PRESERVE score	9 [6–9]	7 [6–9]

Table 1 (*continued*)

Table 1 (*continued*)

Variables	MDA5$^+$ DM (n=7)	Other SRDs (n=4)
Pre-ECMO ABG and ventilator parameters[†]		
pH	7.45 [7.38–7.50]	7.18 [7.03–7.42]
PaO_2, mmHg	45 [40–60]	52 [44–74]
$PaCO_2$, mmHg	37 [30–43]	62 [29–132]
Lactate, mmol/L	3.3 [2.6–5.4]	6.1 [4.4–12.9]
FiO_2	0.9 [0.9–0.9]	0.9 [0.7–0.9]
PIP, cmH_2O	24 [20–25]	23 [15–24]
PEEP, cmH_2O	9 [8–9]	9 [4–10]
P/F ratio	60 [55–80]	74 [56–85]
ECMO details		
ECMO flow, L	4.2 [3.9–4.5]	4.1 [4.0–4.4]
ECMO FiO_2	0.8 [0.7–0.8]	0.8 [0.8–0.8]
Duration of ECMO run, days	11 [6–12]	20 [14–24]
Pneumothorax and mediastinal emphysema	7 [100]	2 [50]
ICU-acquired lung infection	5 [71]	3 [75]
ECMO-related complications	0	1 [25][‡]

Data are expressed as n [%] or median [interquartile range]. "NA" represents patients not having this clinical characteristic. [†], one each in the MDA5$^+$ DM and other SRDs group did not receive invasive mechanical ventilation because of pneumothorax and being considered inevitable progression to refractory respiratory failure needing VV-ECMO, and the desire to maintain an awake state, respectively. [‡], one patient occurred ECMO-related gastrointestinal hemorrhage. MDA5$^+$ DM, anti-melanoma differentiation-associated protein 5 positive dermatomyositis; SRDs, systemic rheumatic diseases; BMI, body mass index; HBsAg, hepatitis B surface antigen; ILD, interstitial lung disease; ESR, erythrocyte sedimentation rate; PCT, procalcitonin; CRP, C-reactive protein; WBC, white blood cell; CK, creatine phosphokinase; LDH, lactate dehydrogenase; BNP, brain natriuretic peptide; APACHE II, Acute Physiology, Age, and Chronic Health Evaluation II; SOFA, sepsis-related organ failure assessment; VV-ECMO, venovenous extracorporeal membrane oxygenation; RESP, Respiratory Extracorporeal Membrane Oxygenation Survival Prediction; PRESERV, PRedicting dEath for SEvere ARDS on VV-ECMO; ECMO, extracorporeal membrane oxygenation; ABG, arterial blood gas; PIP, peak inspiratory pressure; PEEP, positive end expiratory pressure; P/F ratio, the ratio of arterial oxygen concentration to the fraction of inspired oxygen; ICU, intensive care unit.

received cyclophosphamide, one received antifibrotics, and three received calcineurin antagonists before admission. All four patients in the other SRD group received immunosuppressive therapy and pulse-dosed methylprednisolone and IVIG in the hospital, and two received cyclophosphamide, and one received calcineurin antagonists before admission.

None of the patients was a candidate for lung transplantation due to either the severity of their illness, a shortage of donors, an inability to consent, an inability to rehabilitate, or other economic or poor prognostic factors. No patients in the MDA5$^+$ DM group survived to ECMO decannulation or discharge. Three patients in the other SRD group survived to ECMO decannulation, and two of these survived to discharge.

High-resolution computed tomography scans of patients with MDA5$^+$ DM or other SRDs and ARF before and after VV-ECMO

Figure S1A,S1B illustrates that bilateral interstitial infiltrates and pneumothorax worsened on the HRCT scans in the MDA5$^+$ group but improved in the other SRD group after VV-ECMO (Figure S2A,S2B).

Table 2 Treatments and outcomes for patients between MDA5$^+$ DM group and other SRDs group

Items	MDA5$^+$ DM (n=7)	Other SRDs (n=4)
ICU therapeutics		
HFNO or BIPAP	1 [14]	1 [25]
Mechanical ventilation	6 [86]	3 [75]
Vasopressors	6 [86]	2 [50]
Renal replacement therapy	2 [29]	3 [75]
Agents before admission		
Median methylprednisolone dose†, mg/d	60 [40–60]	60 [60–75]
Methylprednisolone duration, days	60 [45–60]	110 [93–165]
Cyclophosphamide‡	1 [14]	2 [50]
Calcineurin antagonists§	3 [43]	1 [25]
Antifibrotic drug¶	1 [14]	1 [25]
New or added agent regimen after admission		
Median methylprednisolone dose, mg/d	80 [80–80]	70 [60–80]
Max methylprednisolone dose, mg/d	120 [120–160]	240 [240–435]
JAKi	7 [100]	0
IVIG use	7 [100]	4 [100]
Outcomes		
Time in ICU, days	18 [10–23]	21 [18–25]
Time in hospital, days	24 [18–51]	41 [28–43]
Survival to ECMO decannulation	0	3 [75]
Survival to hospital discharge	0	2 [50]

Data are expressed as n [%] or median [interquartile range]. †, estimated by total methylprednisolone dose divided by total days of use. ‡, one patient in MDA5$^+$ DM and two patients in other SRDs group both received four doses of cyclophosphamide. §, three patients in MDA5$^+$ DM group received 18, 20 and 22 days of calcineurin antagonists compared to 80 days for one patient in other SRDs group. ¶, one each patient in MDA5$^+$ DM and other SRDs group received 30 and 60 days of antifibrotic drug (nintedanib) respectively. MDA5$^+$ DM, anti-melanoma differentiation-associated protein 5 positive dermatomyositis; SRDs, systemic rheumatic diseases; ICU, intensive care unit; HFNO, high-flow nasal oxygen therapy; BIPAP, biphasic intermittent positive airway pressure ventilation; JAKi, Janus kinase inhibitors; IVIG, intravenous immunoglobulin; ECMO, extracorporeal membrane oxygenation.

Discussion

The condition MDA5$^+$ DM usually affects the lungs, and most patients develop RPILD, which quickly progresses into respiratory failure. This single-center case series describes seven patients with MDA5$^+$ DM who developed severe respiratory failure and four patients with other SRDs and ARF who were supported by VV-ECMO between June 2017 and February 2022.

All dermatomyositis patients had laboratory-confirmed MDA5$^+$ and chest scan CT imaging of ILD. Despite receiving corticosteroids, JAKi treatment (9), and/or antifibrotic medicine, the outcomes of patients with MDA5$^+$ DM supported by VV-ECMO were extremely poor.

Most patients with MDA5$^+$ DM in the present study presented with respiratory symptoms similar to those presented by patients described in reports from France, which indicated the involvement of the lungs in MDA5$^+$ DM (10). Shortness of breath was the most common symptom, and the mean interval between symptom onset and admission was four weeks (11). Consistent with previous

reports (12), MDA5$^+$ DM typically presents with distinctive mucocutaneous features, including Gottron's papules and Mechanic's hand, and a low incidence of myositis. Lymphocytopenia and high ferritin are frequently seen upon hospital admission, which also has been implicated in the occurrence of MDA5$^+$ RPILD in a large Chinese dermatomyositis/clinically amyopathic dermatomyositis patient cohort (13) and other meta-analysis (14).

All 11 patients received methylprednisolone and immunosuppressive treatments. Recent data indicate that early aggressive combination therapies consisting of glucocorticoids, calcineurin antagonists, and cyclophosphamide are likely to improve the outcomes of patients with MDA5$^+$ DM RPILD (15,16). However, this improvement was not observed in the present study. This poor response may be due to the critically ill subgroup of patients with MDA5$^+$ DM and the relatively long duration of their ILD prior to admission. Although rare, RPILD can rapidly progress to severe respiratory failure and even death (17). Therefore, aggressive multi-agent immunosuppression in all patients with MDA5$^+$ DM was recommended, even in those with initially mild ILD.

In the present study, 11 patients underwent VV-ECMO, a life support method for blood oxygenation, to correct severe hypoxemic respiratory failure. Most patients with other SRDs (3/4) who were supported with VV-ECMO survived after weaning, and half (2/4) survived to discharge, indicating that VV-ECMO is an effective choice for managing other SRDs. This result is in agreement with the extracorporeal life support registry report, which showed that the survival rate of patients with adult pulmonary infection supported by VV-ECMO is 59–60% (8).

At the time of the present study, ECMO support in severe respiratory failure secondary to anti-MDA5$^+$ DM RPILD has been described in only ten studies. The median duration of VV-ECMO in the seven patients with MDA5$^+$ DM associated with RPILD in the present study was 11 days [6–12 days], and all the patients died. Similarly, a retrospective study of six patients with refractory respiratory failure who received VV-ECMO reported that all patients died (18). Four other studies reported poor outcomes and no survivors (19-22). In the largest case series reported to date, nine patients with myositis-associated RPILD who received VV-ECMO with no lung transplantation also had poor outcomes, with only one patient surviving (23,24).

Finally, the present study showed a higher proportion (5/7) of patients who developed pneumothorax and pneumomediastinum before ECMO initiation and that is consistent with the finding reported by Ma *et al.* (25) After implementing ECMO, platform pressure dropped from 18–26 to 10–18 cmH_2O in patients with mechanical ventilation. Unexpectedly, despite a significant reduction of platform pressure, pneumothorax newly occurred in the remaining two patients. Guidelines (8) suggest that ECMO therapy plays an effective role in reducing pneumothorax incidence in the presence of dermatomyositis-related RPILD, and should be considered early to avoid complications associated with mechanical ventilation (26).

However, this study was limited by small samples and the retrospective design, causality cannot be established; further research is necessary to examine the relationship between ECMO and pneumothorax or pneumomediastinum.

Conclusions

In conclusion, VV-ECMO may be considered an effective and safe supportive therapy and should be initiated early for patients with SRDs who develop ARF. Patients with other SRDs may be more likely to survive to ECMO decannulation or to discharge compared with patients with MDA5$^+$ DM.

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Footnote

Reporting Checklist: The authors have completed the STROBE reporting checklist. Available at https://jtd.amegroups.com/article/view/10.21037/jtd-24-650/rc

Data Sharing Statement: Available at https://jtd.amegroups.com/article/view/10.21037/jtd-24-650/dss

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Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://jtd.amegroups.com/article/view/10.21037/jtd-24-650/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. This study was

conducted in accordance with the declaration of Helsinki (as revised in 2013). The study was approved by the ethics committee of Renji Hospital Affiliated to the Shanghai Jiaotong University School of Medicine and individual consent for this retrospective analysis was waived.

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