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Clinical science

Extracorporeal membrane oxygenation for acute lung injury in idiopathic inflammatory myopathies—a potential lifesaving intervention

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Abstract

Objectives: Idiopathic inflammatory myopathies (IIM) can present with acute IIM-related lung injury and respiratory failure, leading to a high mortality risk in intensive care units (ICU). Extracorporeal membrane oxygenation (ECMO) in acute respiratory distress syndrome can be lifesaving. We aimed to report a case series of IIM patients that received ECMO.

Methods: Patients with IIM from tertiary care centres in Belgium, Canada, Denmark, USA and Sweden who underwent ECMO were reviewed to describe clinical characteristics, disease outcomes and hospitalization course. Clinical characteristics at admission and during ICU stay including ECMO complications and mortality causes were summarized.

Results: The study included 22 patients (50% female, mean ± SD age at admission 47 ± 12 years) with anti-MDA5 positive dermatomyositis (68%), anti-synthetase syndrome (14%), polymyositis (9%), overlap myositis (5%) and non-MDA5 dermatomyositis (5%). Patients had low comorbidity scores and 46% had received immunosuppression before their ICU admission. Eight (36%) patients died in the ICU, six (27%) were bridged to recovery and eight (36%) were bridged to transplant. When comparing patients bridged to recovery and those who died in the ICU, those who died were older ($P = 0.03$) and had higher median Charlson comorbidity index scores ($P = 0.05$). Both groups had similar frequencies of ECMO-related complications (33% vs 50%, $P = 0.94$).

Conclusion: In the patients exposed to ECMO in this case series, 14 were successfully bridged to recovery or transplant, while 8 died in the ICU. Large studies are needed to collect data on clinical outcomes in patients with IIM-ILD exposed to ECMO to identify the best candidates for the intervention.

Keywords: dermatomyositis, extracorporeal membrane oxygenation, lung transplantation, rapid progressive interstitial lung disease, anti-MDA5 antibody.

Rheumatology key messages

- Patients with idiopathic inflammatory myopathy and rapidly progressive interstitial lung disease have poor outcomes.
- In certain cases, ECMO can be helpful to bridge those patients to recovery or transplant.

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Introduction

Certain subsets of patients with Idiopathic inflammatory myopathies (IIM) are at high risk of rapidly progressive interstitial lung disease (RPILD) and can present with acute lung injury indistinguishable from ARDS. Despite aggressive immunosuppression and supportive care in the intensive care unit (ICU), outcomes in that context are poor [1]. Extracorporeal membrane oxygenation (ECMO) can serve as a bridge to recovery or lung transplantation when mechanical ventilation becomes insufficient. ECMO is associated with lower mortality in severe ARDS [2], however, its benefit in IIM remains uncertain. We report a case series of individuals with IIM receiving ECMO and their outcomes to inform clinical decisions.

Methods

Individuals exposed to ECMO for IIM-related respiratory failure were reviewed from six tertiary care institutions (University Hospitals Leuven, Belgium; McGill University and University of British Columbia hospitals, Canada; Copenhagen University Hospital, Rigshospitalet, Denmark; Karolinska Institutet, Sweden; Johns Hopkins Hospital, USA). Records were reviewed from 2010 to 2020 for all centres except for Johns Hopkins Hospital that covered the period from 2000 to 2020. Patients were included if cannulated to ECMO and diagnosed with: (1) IIM per expert rheumatologist opinion, or (2) ARDS with a positive myositis-specific autoantibody in the absence of another cause. Individuals with previous lung transplant were excluded. Clinical characteristics at hospital admission and during ICU stay were described. Disease duration was defined from first IIM symptom to hospital admission. Clinical features were considered present or absent based on physician judgement. Chest computed tomography and pathological findings were reviewed. Comorbidity burden was measured using the age-adjusted Charlson Comorbidity Index (range 0–37) [3]. Time to death, to ECMO, or to lung transplant was calculated from day of ICU admission. Duration of hospitalization was calculated from first admission to hospital until discharge or death. Follow-up duration and vital status were based on last documented follow-up.

Descriptive statistics were used to describe the study cohort. Continuous variables were expressed as means with standard deviation (SD) or medians with interquartile range [IQR] depending on distribution, and categorical variables as frequencies with percentages. Comparisons between patients who were bridged to recovery and those who died in the ICU were done using Student's *t test*, Chi squared test or Kruskal–Wallis test. No comparison was done with the patients that were transplanted as their outcomes were expected to follow different trajectories than non-transplanted patients. The study received ethical approval from the Medical/Biomedical Research Ethics Committee of the West-Central-Montreal-Health (MP-2021–2402) in agreement with the Helsinki declaration for clinical studies and all patients gave informed consent to participate in this study.

Results

We included 22 individuals with IIM (Belgium $n=5$, Canada $n=7$, Denmark $n=2$, Sweden $n=4$ and USA $n=4$) that received veno-venous ($n=20$) or veno-arterial ($n=2$) ECMO for respiratory failure (Tables 1 and 2). Most subjects were newly diagnosed with IIM (68%) and had positive anti-MDA5 autoantibodies (68%). At hospital admission, about half of the patients had heliotrope and/or Gottron's rashes, muscle weakness and/or creatine kinase elevation. Isolated ILD was found in only two (9%) patients. On imaging, ground glass opacities with or without consolidation were found in all subjects. At ICU admission, all subjects were on corticosteroids, and 21 (95%) patients on at least one more immunosuppressor. Respiratory co-infections were suspected at ICU admission in nine (41%) patients. All patients had met severe ARDS criteria with a ratio of partial pressure of oxygen in arterial blood (PaO2) to the fraction of inspiratory oxygen concentration (FiO2) ≤100mmHg. Median time-to-ECMO after ICU admission was five days [IQR 1, 12].

Of the 22 patients included, six (27%) were bridged to recovery, eight (36%) were bridged to transplant and eight (36%) died in the ICU (Table 1, 2 and Supplementary Table 1, available at *Rheumatology* online). Of the patients that were bridged to recovery, two (33%) had anti-MDA5+ dermatomyositis, two (33%) anti-EJ+ anti-synthetase syndrome, one (17%) polymyositis and one (17%) overlap myositis (anti-Ku+). Of those, three (50%) were new IIM diagnoses, and two (33%) had isolated ILD. At ICU admission, one (17%) patient had a respiratory co-infection. The median duration of ECMO in that group was 11 days [IQR 7, 27] and two (33%) patients experienced ECMO-related major bleeding requiring transfusion. Of the six patients bridged to recovery, four had >6 months post-hospital discharge follow-up and one was lost to follow-up after being transferred to another institution (Supplementary Table 1, available at *Rheumatology* online).

Of the eight patients that died in the ICU, seven (88%) had anti-MDA5+ dermatomyositis, and one (13%) an anti-Jo1 anti-synthetase syndrome. Of those, six (75%) were new IIM diagnoses. At ICU admission, three (38%) had respiratory co-infections. The median duration of ECMO in that group was 24 days [IQR 18, 37]. Four (50%) patients experienced ECMO-related complications including two patients with neurological complications, one with major bleeding and one with ischaemia, major bleeding, neurological and cannulation-related complications. Four (50%) patients had withdrawal of care, three (38%) died from respiratory failure and one (13%) from ECMO-related major haemorrhagic stroke.

Of the eight transplanted patients, six (75%) had anti-MDA5+ dermatomyositis, one (13%) a non-MDA5 dermatomyositis (anti-Ro60+) and one (13%) a polymyositis. Of those, six (75%) were new IIM diagnoses. At ICU admission, five (63%) had respiratory co-infections. The median duration of ECMO in that group was 30 days [IQR 15, 41] and the median time to transplant from ICU admission was 43 days [IQR 38, 84]. Three (38%) patients had ECMO-related complications including a patient with a cannulation-related DVT, another with major bleeding, and one with ischaemia, major bleeding and neurological complications (Supplementary Table 1, available at *Rheumatology* online). Of the eight transplanted patients, five had >6 months post-hospital discharge follow-up including one patient who died 290 days post-transplant due to respiratory failure and two who were lost to follow-up after being transferred to

Table 1. Clinical characteristics at hospital admission of IIM patients exposed to ECMO

Clinical characteristics, *n* (%)	All (*n* = 22)	No transplant			Bridged to transplant (*n* = 8)
		Survived ICU (*n* = 6)	Died in ICU (*n* = 8)	*P*-value	
Age at admission, years, mean±SD	47 ±12	41 ±8	53 ±10	0.03	44 ±14
Female, *n* (%)	11 (50)	3 (50)	3 (38)	1.00	5 (63)
Ethnicity, *n* (%)					
Asian	3 (14)	0 0	1 (13)	0.35	2 (25)
Black	1 (5)	1 (17)	0		0
Caucasian	17 (77)	5 (83)	7 (88)		5 (63)
Other	1 (5)	0	0		1 (13)
New IIM diagnosis, *n* (%)	15 (68)	3 (50)	6 (75)	0.69	6 (75)
Disease duration, months, median [IQR]	3 [2, 7]	4 [1, 7]	3 [2, 10]	0.52	3 [2, 4]
IIM subsets, *n* (%)					
DM (anti-MDA5+)	15 (68)	2 (33)	7 (88)	0.18	6 (75)
DM	1 (5)	0	0		1 (13)
Antisynthetase syndrome	3 (14)	2 (33)	1 (13)		0
Polymyositis	2 (9)	1 (17)	0		1 (13)
Overlap myositis	1 (5)	1 (17)	0		0
Skin involvement, *n* (%)	14 (64)	1 (17)	6 (75)	0.11	7 (88)
Heliotrope and/or Gottron's	12 (55)	1 (17)	4 (50)	0.47	7 (88)
Erythroderma	2 (9)	0 (0)	1 (13)	0.80	1 (13)
Mechanics hands	4 (18)	1 (17)	2 (25)	1.00	1 (13)
Ulceration	6 (27)	0 (0)	4 (50)	0.15	2 (25)
Periungual erythema, *n* (%)	14 (64)	2 (33)	6 (75)	0.31	6 (75)
Raynaud phenomenon, *n* (%)	4 (18)	1 (17)	2 (25)	1.00	1 (13)
Arthritis, *n* (%)	9 (41)	1 (17)	5 (63)	0.24	3 (38)
Dysphagia, *n* (%)	3 (14)	0	3 (38)	0.30	0
Muscle weakness, *n* (%)	12 (55)	3 (50)	4 (50)	1.00	5 (63)
Creatine kinase elevation, *n* (%)	13 (59)	3 (50)	5 (63)	0.64	5 (63)
Isolated ILD, *n* (%)	2 (9)	2 (33)	0	0.32	0
Radiological findings					
Consolidation	18 (82)	6 (100)	6 (75)	0.42	6 (75)
Ground-glass	22 (100)	6 (100)	8 (100)	1.00	8 (100)
Fibrosis	8 (36)	1 (17)	3 (38)	0.39	4 (50)
Pathological findings[a]					
Available lung pathology	10 (46)	3 (50)	2 (25)		5 (63)
UIP	1 (10)	1 (17)	0	0.23	1 (13)
NSIP	2 (20)	1 (17)	0	0.29	1 (13)
Organizing pneumonia	2 (20)	2 (33)	0	0.32	0
Diffuse alveolar damage	8 (80)	2 (33)	1 (13)	0.38	5 (63)
Discrete inflammation	1 (10)	0	1 (13)	0.18	0
Charlson comorbidity index, median [IQR]	2 [2, 2]	1.5 [1, 2]	2.5 [2, 3]	0.05	2 [2, 2]
Immunosuppression prior to admission, *n* (%)	10 (46)	2 (33)	3 (38)	1.00	5 (63)

IIM, idiopathic inflammatory myopathy; ECMO, extracorporeal membrane oxygenation; ICU, intensive care unit; DM, dermatomyositis; ILD, interstitial lung disease; ARDS, acute respiratory distress syndrome; UIP, usual interstitial pneumonia; NSIP, non-specific interstitial pneumonia.
[a] Eight had open or video-assisted thoracoscopic lung biopsy, one had a transbronchial biopsy showing discrete inflammation, and one had explanted lung pathology.

another institution (Supplementary Table 1, available at *Rheumatology* online).

When comparing patients with IIM on ECMO who were bridged to recovery and those who died in the ICU, those who died were older ($P = 0.03$) and had higher median Charlson comorbidity index scores ($P = 0.05$) (Table 1). No other significant differences were found between those groups in terms of clinical presentation at ICU admission, immunosuppressive therapy exposure or ECMO-related complications (Tables 1 and 2).

Discussion

This large case series summarizes our experience with the use of ECMO in severe IIM-related lung disease where six patients were successfully bridged to recovery, eight bridged to transplant, and eight died. Patients with IIM and RP-ILD have very poor short-term prognosis, especially those with anti-MDA5 positivity. In a retrospective multicentre French study on patients with IIM admitted to the ICU with acute respiratory failure, in-hospital mortality was 84% for those with anti-MDA5 and 29% for those with anti-synthetase [4]. In our series, all patients met criteria for severe ARDS prior to ECMO initiation [5], which has been associated with an in-hospital mortality of 46% outside of the IIM context [6]. Because mortality generally occurs within the first 3 months in IIM-RPILD, these patients are treated aggressively [7]. Thus ECMO, a potentially lifesaving intervention in other types of ARDS [8, 9], is worth consideration as part of the arsenal of management options for severe lung disease in IIM. Neither the diagnosis of an underlying IIM nor use of immunosuppression should be factors discouraging ECMO as rescue therapy in this population, but other factors such as age, and comorbidities should be carefully assessed.

Table 2. Hospital course and outcomes of IIM patients exposed to ECMO

Clinical characteristics	All ($n = 22$)	No transplant			Bridged to transplant ($n = 8$)
		Survived ICU ($n = 6$)	Died in ICU ($n = 8$)	*P*-value	
Duration of hospitalization, days, median [IQR]	89 [50, 116]	100 [51, 118]	47 [30, 71]	0.14	47 [30, 71]
Treatment received during hospitalization *n* (%)					
Corticosteroids (IV and/or PO)	22 (100)	6 (100)	8 (100)	1.00	8 (100)
Cyclophosphamide	16 (73)	4 (67)	6 (75)	1.00	6 (75)
Intravenous immunoglobulins	10 (46)	1 (17)	4 (50)	0.47	5 (63)
Mycophenolate mofetil	8 (36)	2 (33)	2 (25)	1.00	4 (50)
Rituximab	8 (36)	0	4 (50)	0.15	4 (50)
Calcineurin inhibitor	7 (32)	0	2 (25)	0.58	5 (63)
Azathioprine	1 (5)	0	0	–	1 (13)
Three agents	7 (32)	0	4 (50)	0.15	3 (38)
>Three agents	2 (9)	0	1 (13)	1.00	1 (13)
Plasmapheresis	5 (23)	2 (33)	2 (25)	1.00	1 (13)
Duration ICU stay, days, median [IQR]	61 [27, 97]	45 [27, 82]	32 [25, 48]	0.47	107 [87, 156]
Respiratory co-infection at ICU admission *n* (%)	9 (41)	1 (17)	3 (38)	0.80	5 (63)
Respiratory viral infection	1 (5)	0	1 (13)	1.00	0
Pneumocystis jirovecii	3 (14)	0	0	–	3 (38)
Other respiratory infection	7 (32)	1 (17)	2 (25)	0.59	4 (50)
CMV infection at ICU admission *n* (%)	2 (9)	2 (33)	0	0.32	0
Dialysis during ICU *n* (%)	9 (41)	3 (50)	3 (38)	1.00	3 (38)
Sepsis during ICU *n* (%)	14 (64)	5 (83)	4 (50)	0.47	5 (63)
Time to ECMO after ICU admission, days, median [IQR]	5 [1, 12]	2 [1, 6]	4 [1, 7]	0.85	31 [3, 45]
Duration of ECMO, days, median [IQR]	22 [8, 38]	11 [7, 27]	24 [18, 37]	0.27	30 [15, 41]
ECMO-related complications *n* (%)	9 (41)	2 (33)	4 (50)	0.94	3 (38)
Peripheral vascular ischaemia	2 (9)	0	1 (13)	1.00	1 (13)
Neurologic complications	4 (18)	0	3 (38)	0.30	1 (13)
Major bleeding requiring transfusion	6 (27)	2 (33)	2 (25)	1.00	2 (25)
Local cannulation related	2 (9)	0	1 (13)	1.00	1 (13)
Oxygen dependence at hospital discharge *n* (%)	6 (27)	3 (50)	2 (25)	0.23	1 (13)

IIM, idiopathic inflammatory myopathy; ECMO, extracorporeal membrane oxygenation; ICU, intensive care unit One patient from the transplanted group was lost to follow-up after being transferred to another institution.

A previous series of nine patients with IIM (67% with anti-MDA5) receiving ECMO reported very high early mortality, with only one survivor [10], but none of those patients had ECMO used as bridge to transplant. In our study, 6 out of 14 (43%) non-transplanted patients survived. Also contrary to this previous series, we did not exclude patients with concomitant respiratory infection which can be triggers of ILD flares and ARDS [11, 12]. While having an infection may result in more easily reversible lung disease, only one of the non-transplanted survivors in our series had an infection. After exclusion of all subjects with a concomitant infection, 5 out of 10 (50%) patients who received ECMO without lung transplant survived to ICU discharge.

The use of ECMO is a complex and personalized decision based on patient and institutional factors. Our series examined the use of ECMO based on clinician decision without any pre-specified criteria. While our study was not powered to identify risk factors for poor ECMO outcomes, we did identify patients who survived being younger and having a lower Charlson comorbidity index. The proportion of patients with anti-MDA5 was also numerically lower among those who survived without lung transplant, compared with those who died which could reflect the severity of anti-MDA5-related RPILD.

Lung transplant in IIM-related ILD has similar survival outcomes as in other indications such as idiopathic pulmonary fibrosis with a 5-year survival probability of 70% [13, 14]. All eight transplant recipients in our series were anti-MDA5 positive. One patient died at nine months after hospital discharge, and two patients had unknown status after ICU discharge. However, the decision and ability to use ECMO for bridging to transplant in eligible patients is highly dependent on institutional capabilities and entails a complex interdisciplinary discussion that need to be evaluated on a case-by-case basis.

Several limitations to our study need to be acknowledged. First, we identified cases retrospectively and might have missed cases of IIM-ILD patients cannulated to ECMO that were not identified by their treating team, notably those with no myositis autoantibodies. Recognizing IIM with RPILD in the acute care setting can be challenging especially if patients present with isolated ILD or no muscle involvement and might have not been captured. Moreover, it is possible that some ARDS cases included in our study were not experiencing an ILD flare, but a respiratory infection, and might have biased our results to better outcomes [15]. Of note, we also did not compare clinical outcomes of IIM-ILD patients in the ICU with similar features that were not cannulated to ECMO, potentially reporting on patients that were considered good candidate for the intervention with biased results towards better outcomes. Finally, a large proportion of patients included in this case series had anti-MDA5+ dermatomyositis limiting the generalizability of our findings to other IIM subsets.

Conclusion

Although ECMO is an invasive intervention with potential severe complications, it should be considered as a rescue option on a case-by-case basis in patients with IIM presenting

with acute respiratory failure depending on institutional capabilities. This intervention can serve as a bridge to allow immunosuppression to take effect or for lung transplant. Large international registries are needed to collect data on clinical outcomes in patients with IIM exposed to ECMO to identify the best candidates for the intervention.

Supplementary material

Supplementary material is available at *Rheumatology* online.

Data availability

The data underlying this article cannot be shared publicly for the privacy of individuals that participated in the study. The data will be shared on reasonable request to the corresponding author.

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