



Extracorporeal Membrane Oxygenation for Life-Threatening Cardiopulmonary Manifestations of Systemic Autoimmune and Vasculitic Disorders: A Patient-Level Review

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Abstract

Objective: Systemic autoimmune and vasculitic disorders may cause life-threatening respiratory or cardiac failure, including diffuse alveolar hemorrhage (DAH), acute respiratory distress syndrome (ARDS), fulminant myocarditis, and cardiogenic shock. Extracorporeal membrane oxygenation (ECMO) may provide temporary cardiopulmonary support while disease-directed therapy is initiated. This patient-level clinical synthesis characterized the clinical circumstances prompting ECMO, the underlying cardiopulmonary manifestations, VV-versus VA-ECMO use, disease-directed treatment administered during ECMO support, anticoagulation strategies, ECMO-related complications, and reported outcomes.

Methods: PubMed, Embase, Web of Science, and Scopus were searched from inception through 1 August 2026 using combinations of ECMO, systemic autoimmune and vasculitic disorders, and relevant cardiopulmonary manifestations. Reference lists were also screened. Eligible publications were case reports or case series providing sufficient individual-level clinical information.

Results: Twenty-seven publications describing 33 patients were included. Mean age was 36.0 ± 16.0 years; 17 (51.5%) were male. DAH with respiratory failure was the predominant manifestation, occurring in 25 patients (75.8%). VV-ECMO was used in 22 patients (66.7%) and VA-ECMO in 11 (33.3%). Median ECMO duration was 192 h (IQR, 120–312 h). ECMO-related complications were reported in 8 patients (24.2%); circuit or oxygenator thrombosis was most frequent (5 [15.2%]). Overall, 31 patients (93.9%) survived to hospital discharge and 2 (6.1%) died.

Conclusion: ECMO may provide temporary cardiopulmonary support in carefully selected patients with life-threatening respiratory and/or cardiac failure while disease-directed therapy is administered. However, the reported outcomes should be interpreted cautiously because of publication and reporting bias, clinical heterogeneity, incomplete patient-level information, and the absence of comparative controls.

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Introduction

Although most patients with systemic autoimmune and vasculitic disorders exhibit a subacute or chronic course, some develop life-threatening respiratory

and/or cardiac manifestations, including DAH, ARDS, fulminant myocarditis, and cardiogenic shock, that may require escalation to ECMO when conventional management is insufficient. ECMO can provide temporary support to stabilize gas exchange and/or maintain hemodynamic stability while disease-directed immunosuppressive therapy takes effect [1,2]. Such life-threatening manifestations arise from dysregulated immune responses that result in widespread inflammation, endothelial dysfunction, and immune-mediated vascular and tissue injury affecting vital organs [3,4]. The choice of ECMO configuration depends primarily on the predominant pattern of organ failure. VV-ECMO is used for severe respiratory failure when cardiac function is sufficient to maintain systemic circulation, providing extracorporeal gas exchange while allowing lung-protective ventilation, whereas VA-ECMO provides circulatory support, with or without respiratory support, for patients with cardiogenic shock or combined cardiopulmonary failure [5,6]. However, the use of ECMO in systemic autoimmune and vasculitic disorders is complicated by competing risks of bleeding and thrombosis, challenges in anticoagulation management during extracorporeal support, concurrent immunosuppressive therapy, susceptibility to secondary infections, and other management complexities related to both the underlying disease and the extracorporeal circuit [7,8]. Plasma exchange (PLEX) may also be incorporated into the management of selected severe autoimmune and vasculitic presentations, although its indications and potential contribution to outcomes remain uncertain and vary according to the underlying disease and clinical phenotype [9]. This patient-level clinical synthesis characterizes the clinical manifestations prompting ECMO, ECMO configuration and duration, disease-directed therapy and its timing relative to ECMO, PLEX, anticoagulation strategies, ECMO-related complications, and reported clinical outcomes.

Methods

We conducted a patient-level clinical synthesis of published case reports and case series describing the use of ECMO in patients with systemic autoimmune and vasculitic disorders who developed acute cardiopulmonary decompensation requiring extracorporeal support. For the purpose of this review, acute cardiopulmonary decompensation was defined as acute respiratory failure, circulatory failure, or combined cardiorespiratory failure of sufficient severity to require ECMO support after inadequate response to conventional management.

Study identification and search strategy

Study identification, selection, and reporting were performed in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) 2020 Statement [10], where applicable to the design of this patient-level clinical synthesis. Relevant publications

were identified through searches of PubMed, Embase, Web of Science, and Scopus from database inception through 1 August 2026. Although the databases were searched from inception, eligibility was restricted to publications from 2008 onward. The search strategy combined terms related to ECMO with terms describing systemic autoimmune and vasculitic disorders and/or relevant acute cardiopulmonary manifestations, including diffuse alveolar hemorrhage (DAH), acute respiratory distress syndrome (ARDS), respiratory failure, myocarditis, cardiogenic shock, and circulatory failure. Search terms were combined using Boolean operators, and the search strategies were adapted to the indexing structure and search syntax of each database. The detailed PubMed search strategy, including search terms, Boolean operators, and field tags, is provided in Supplementary Table S1. Google Scholar was used during the initial exploratory phase of study identification but was not included as a formal source in the final database search or PRISMA accounting. Publications identified during this exploratory process were subsequently cross-checked against the four bibliographic databases and the reference lists of relevant publications to avoid double counting of records. Thirty-one records entered the study-selection process. Bibliographic information was checked for duplicate publications by comparison of publication title, authors, journal, publication year, and digital object identifier or other available unique identifiers. No duplicate records were identified. Reference lists of relevant and eligible publications were also screened manually as a supplementary method of study identification. The study selection process is summarized in the PRISMA flow diagram.

Eligibility criteria and study selection

Publications were eligible for inclusion if they were case reports or case series describing the use of ECMO in patients with systemic autoimmune or vasculitic disorders. Case reports and case series were deliberately selected because the objective of this patient-level clinical synthesis required sufficiently detailed individual patient-level information to characterize the underlying diagnosis, life-threatening clinical manifestation prompting ECMO support, ECMO configuration, management during extracorporeal support, and clinical outcome. The study population included patients with systemic autoimmune and vasculitic disorders, including ANCA-associated vasculitis (AAV), encompassing granulomatosis with polyangiitis, microscopic polyangiitis, myeloperoxidase-ANCA-associated vasculitis, and eosinophilic granulomatosis with polyangiitis, as well as other systemic autoimmune or inflammatory disorders associated with severe cardiopulmonary involvement requiring ECMO support. No restrictions were applied regarding patient age or sex. Publications were required to provide sufficient individual patient-level clinical information to permit meaningful extraction of the clinical and management variables relevant to the objectives of this review. Publications were excluded

if they were observational cohort studies, registry analyses, or other studies reporting only aggregated patient data; did not report original patient data; described ECMO use for conditions outside the scope of systemic autoimmune or vasculitic disorders or for cardiopulmonary failure not directly attributable to the underlying disease process; lacked sufficient individual-level clinical information for meaningful data extraction; represented duplicate publications; or did not meet the predefined temporal, reporting, or methodological criteria for inclusion. To focus on contemporary ECMO practice, only publications published from 2008 onward were eligible. This cutoff was selected because the period around 2008 marked the transition to a modern era of ECMO characterized by the increasing adoption of centrifugal pumps, low resistance polymethylpentene oxygenators, biocompatible circuit technologies, and more integrated ECMO systems [11]. Earlier reports were excluded because substantial differences in extracorporeal technology and clinical practice could limit comparability with contemporary ECMO management. Titles and abstracts of the identified records were independently screened by two reviewers. Records considered potentially relevant proceeded to full-text assessment. Given the highly specific clinical focus of the search and the nature of the identified records, all 31 records underwent full-text assessment. Disagreements regarding study selection were resolved through discussion and consensus. The methodological quality of the included reports was independently evaluated by two reviewers using the Joanna Briggs Institute (JBI) Critical Appraisal Checklists for Case Reports and Case Series, as appropriate [12] (Table 1). Following full-text assessment, four reports were excluded: two publications were excluded because they were published before the predefined 2008 cutoff for contemporary ECMO practice, one because the indication for ECMO was outside the predefined clinical scope of the review, and one because it did not meet the predefined reporting or publication criteria for inclusion. Twenty-seven publications were ultimately included.

Data extraction and outcomes

Data were extracted from each eligible publication at the individual patient level whenever sufficient information was available. Extracted variables included patient demographics; underlying autoimmune or vasculitic diagnosis and relevant underlying conditions; life-threatening clinical manifestation leading to ECMO support; ECMO-related complications; clinical outcome; ECMO configuration and duration of support; immunosuppressive treatment and its timing relative to ECMO; use of plasma exchange; and anticoagulation strategy where reported (Tables 2 and 3). When a publication reported more than one patient, data were extracted separately for each patient whenever sufficient information was available. The primary objective of this patient-level clinical synthesis was to characterize the published

clinical experience of ECMO use in systemic autoimmune and vasculitic disorders complicated by life-threatening cardiopulmonary manifestations, with particular emphasis on the clinical circumstances prompting ECMO, ECMO configuration, disease-directed treatment during ECMO support, and associated management strategies and clinical outcomes. Survival was recorded as a descriptive clinical outcome rather than as a measure of ECMO effectiveness. ECMO-related complications were recorded as described in the original publications and were not presumed to be directly attributable to ECMO unless such attribution was explicitly stated or supported by the clinical information provided in the original report.

Data synthesis and analysis

Owing to the descriptive nature of the available evidence, the small number of included publications and patients, and the substantial clinical and methodological heterogeneity among the case reports and case series, a formal meta-analysis was not performed. Data were collated in Microsoft Excel and analyzed descriptively at the patient level. Continuous variables were summarized using mean $\pm$ standard deviation, median, and range, as appropriate, whereas categorical variables were reported as frequencies and percentages. Missing or incompletely reported variables were not imputed, and analyses were based on the information available from the original publications. The final analysis included 27 publications reporting individual-level data for 33 patients.

Results

The literature search and study selection process are summarized in the PRISMA flow diagram (Fig. 1). A total of 31 records were identified through the database searches. No duplicate records were identified, and all records underwent full-text assessment. Following full-text assessment, four reports were excluded: two publications were excluded because they were published before the predefined 2008 cutoff for contemporary ECMO practice, one because full-text assessment showed that the indication for ECMO was not attributable to cardiopulmonary failure directly related to the underlying systemic autoimmune or vasculitic disease, as required by the predefined eligibility criteria, and one because it did not meet the predefined reporting and publication criteria for inclusion. Ultimately, 27 publications reporting individual-level data for 33 patients were included in the final analysis. The demographic characteristics, underlying disease, past medical history, life-threatening manifestations requiring ECMO support, ECMO mode and duration of support, and ECMO-related complications are presented in Table 2. Immunosuppressive therapy and its timing relative to ECMO initiation, plasma exchange, anticoagulation strategies, and clinical outcomes are presented in Table 3. The mean age was 36.0 ± 16.0 years (range, 13-70 years), with a median age of 33 years (interquartile range [IQR],

25–45 years). Seventeen patients (51.5%) were male and 16 (48.5%) were female. The most frequently represented diagnoses were granulomatosis with polyangiitis (GPA) in 11 patients (33.3%), systemic lupus erythematosus (SLE) in 7 (21.2%), microscopic polyangiitis (MPA) in 4 (12.1%), and myeloperoxidase-ANCA-associated vasculitis (MPO-AAV) in 3 (9.1%). Eosinophilic granulomatosis with polyangiitis and mixed connective tissue disease were each reported in 2 patients (6.1%), whereas macrophage activation syndrome, polymyositis, IgG4-related disease, and Sjögren's syndrome were each represented by 1 patient (3.0%). Overall, AAV accounted for 20 patients (60.6%), comprising granulomatosis with polyangiitis (GPA; n = 11), microscopic polyangiitis (MPA; n = 4), MPO-AAV (n = 3), and eosinophilic granulomatosis with polyangiitis (EGPA; n = 2). Systemic lupus erythematosus (SLE) accounted for 7 patients (21.2%), while the remaining 6 patients (18.2%) had mixed connective tissue disease (MCTD; n = 2), macrophage activation syndrome (MAS; n = 1), polymyositis (PM; n = 1), IgG4-related disease (IgG4-RD; n = 1), or Sjögren's syndrome (SjS; n = 1). Twenty-one patients (63.6%) had no significant past medical history, while 12 (36.4%) had specified underlying conditions. DAH with respiratory failure was the predominant life-threatening manifestation, occurring in 25 patients (75.8%). Severe cardiac manifestations, including cardiogenic shock, fulminant myocarditis, or left ventricular failure, were reported in 7 patients (21.2%). One patient (3.0%) presented with decreased level of consciousness, fever, and hypotension requiring intubation. VV-ECMO was used in 22 patients (66.7%), whereas VA-ECMO was used in 11 (33.3%). VV-ECMO was predominantly used in patients presenting with severe respiratory failure, particularly DAH, whereas VA-ECMO was used primarily in patients with severe cardiac or combined cardiorespiratory failure. The mean duration of ECMO support was 234.2 ± 137.5 h, with a median of 192 h (IQR, 120–312 h; range, 48–600 h). No ECMO-related complications were reported in 25 patients (75.8%), whereas complications were reported in 8 (24.2%). Circuit or oxygenator thrombosis was the most frequently reported complication, occurring in 5 patients (15.2%). Minor extrapulmonary bleeding with thrombocytopenia or suspected heparin-induced thrombocytopenia, abdominal hematoma, and increased hemoptysis following heparin escalation were each reported in 1 patient (3.0%). Corticosteroids were used in all 33 patients (100%), followed by cyclophosphamide in 18 (54.5%), rituximab in 7 (21.2%), and intravenous immunoglobulin in 7 (21.2%). Azathioprine was used in 3 patients (9.1%), while mycophenolate mofetil, anakinra, and avacopan were each reported in 1 patient (3.0%). Immunosuppressive treatment was administered before ECMO in 16 patients (48.5%), during ECMO in 19 (57.6%), and after ECMO in 11 (33.3%); these categories were overlapping because several patients received treatment across more than one phase of ECMO support. Plasma

exchange was performed in 17 patients (51.5%), while its use was not reported in 16 patients (48.5%). Anticoagulation strategies varied considerably across the cohort. Systemic anticoagulation was initiated at ECMO initiation in 6 patients (18.2%), whereas anticoagulation was initially withheld or minimized and subsequently initiated in 7 (21.2%). Five patients (15.2%) received no systemic anticoagulation during ECMO, including one patient who received a single heparin bolus at cannulation followed by no documented subsequent systemic anticoagulation. Citrate anticoagulation was used in 1 patient (3.0%), while the anticoagulation strategy was not reported in 14 patients (42.4%). Among the 33 patients included in this review, 31 (93.9%) survived to hospital discharge, while 2 (6.1%) died.

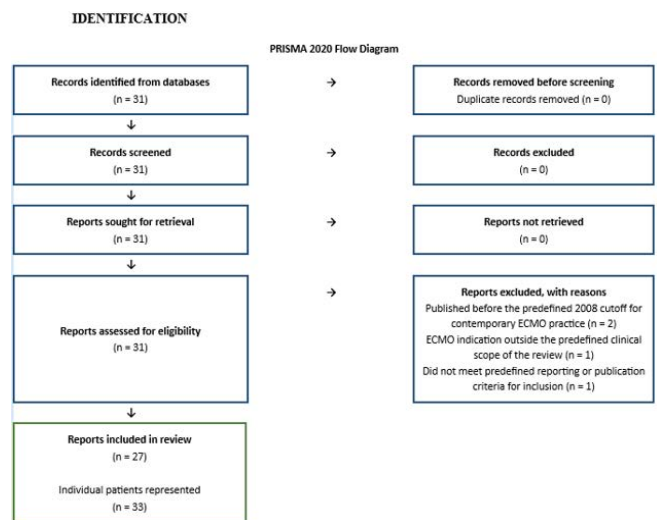


Figure 1: PRISMA 2020 flow diagram illustrating the study selection process. Twenty-seven reports providing individual-level data for 33 patients met the eligibility criteria and were included in the review.

Table 1: Joanna Briggs Institute (JBI) Critical Appraisal Checklist criteria used for the included case reports and case series. The appropriate checklist was applied according to study design; each criterion was assessed as Yes, No, Unclear, or Not applicable. Checklist wording follows the JBI 2020 critical appraisal tools [12].

Panel A. JBI Critical Appraisal Checklist for Case Reports					
No.	Appraisal criterion	Yes	No	Unclear	Not applicable
1	Were the patient's demographic characteristics clearly described?	—	—	—	—
2	Was the patient's history clearly described and presented as a timeline?	—	—	—	—
3	Was the current clinical condition of the patient on presentation clearly described?	—	—	—	—

4	Were diagnostic tests or assessment methods and the results clearly described?	—	—	—	—
5	Was the intervention(s) or treatment procedure(s) clearly described?	—	—	—	—
6	Was the post-intervention clinical condition clearly described?	—	—	—	—
7	Were adverse events (harms) or unanticipated events identified and described?	—	—	—	—
8	Does the case report provide takeaway lessons?	—	—	—	—
Panel B. JBI Critical Appraisal Checklist for Case Series					
No.	Appraisal criterion	Yes	No	Unclear	Not applicable
1	Were there clear criteria for inclusion in the case series?	—	—	—	—
2	Was the condition measured in a standard, reliable way for all participants included in the case series?	—	—	—	—

3	Were valid methods used for identification of the condition for all participants included in the case series?	—	—	—	—
4	Did the case series have consecutive inclusion of participants?	—	—	—	—
5	Did the case series have complete inclusion of participants?	—	—	—	—
6	Was there clear reporting of the demographics of the participants in the study?	—	—	—	—
7	Was there clear reporting of clinical information of the participants?	—	—	—	—
8	Were the outcomes or follow-up results of cases clearly reported?	—	—	—	—
9	Was there clear reporting of the presenting site(s)/ clinic(s) demographic information?	—	—	—	—
10	Was statistical analysis appropriate?	—	—	—	—

Table 2: Patient characteristics, underlying conditions, clinical presentation, ECMO configuration and duration, and ECMO-related complications.

No	Included publication	Age (years)	Sex	Disease	Past medical history	Life-threatening manifestation	ECMO Mode	ECMO duration (h)	ECMO related complications
1	Di Maria et al., 2008 ⁽¹³⁾	13	Male	MPA	No significant past medical history	DAH with respiratory failure and shock	VA	120	No complications reported
2	Joseph et al., 2011 ⁽¹⁴⁾	13	Male	GPA	No significant past medical history	DAH with respiratory failure	VV	120	No complications reported
3	Hohenforst-Schmidt et al., 2013 ⁽¹⁵⁾	65	Female	GPA	Chronic sinusitis	DAH with ESRD and respiratory failure	VV	240	Minor extrapulmonary bleeding; thrombocytopenia/ suspected HIT
4.1	Pacheco Claudio et al., 2014 ⁽¹⁶⁾	33	Female	SLE	No significant past medical history	DAH with respiratory failure and sepsis	VV	48	No complications reported
4.2		36	Male	SLE	No significant past medical history	DAH with respiratory failure and sepsis	VV	144	ECMO circuit thrombosis
5	Patel & Lipchik et al., 2014 ⁽¹⁷⁾	28	Female	SLE	SLE with nephritis; cardiomyopathy, HTN, and DM	DAH with respiratory failure, cardiac tamponade, and cardiac arrest	VV	96	No complications reported

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6.1	Yusuff et al., 2015 ⁽¹⁸⁾	23	Female	GPA	No significant past medical history	DAH with respiratory failure	VV	240	No complications reported
6.2		27	Male	GPA	No significant past medical history	DAH with respiratory failure	VV	240	ECMO circuit/ oxygenator thrombosis
7	Choi & Kang et al., 2016 ⁽¹⁹⁾	43	Female	GPA	No significant past medical history	DAH with respiratory failure	VV	192	No complications reported
8	Rawal et al., 2016 ⁽²⁰⁾	28	Male	GPA	No significant past medical history	DAH with respiratory failure	VV	504	No complications reported
9	Ammirati et al., 2016 ⁽²¹⁾	25	Male	EGPA	No significant past medical history	Cardiogenic shock	VA	312	No complications reported
10	Abdelaty et al., 2017 ⁽²²⁾	18	Female	SLE	History of lupus nephritis	Diffuse alveolar hemorrhage and respiratory failure	VV	600	No complications reported
11	Vanoli et al., 2017 ⁽²³⁾	33	Male	GPA	Recurring sinusitis	Diffuse alveolar hemorrhage and respiratory failure	VV	216	No complications reported
12	Lee CF et al., 2018 ⁽²⁴⁾	14	Male	MAS	No significant past medical history	DAH with respiratory failure	VA	120	No complications reported
13.1	Delvino et al., 2019 ⁽²⁵⁾ (two cases)	45	Female	GPA	No past medical history	DAH with respiratory failure	VV	144	No complications reported
13.2		45	Male	GPA	No significant past medical history	DAH with respiratory failure	VV	336	Abdominal hematoma
14	Mabe et al., 2019 ⁽²⁶⁾	54	Male	MCTD	No significant past medical history	DAH with respiratory failure and shock	VA	216	No complications reported
15	Tong et al., 2019 ⁽²⁷⁾	60	Female	MPA	No significant past medical history	DAH with respiratory failure	VV	168	Increased hemoptysis with heparin escalation; improved after heparin dose reduction
16	Liu et al., 2020 ⁽²⁸⁾	47	Female	SLE	No significant past medical history	DAH with respiratory failure	VA	168	No complications reported
17	Lopes et al., 2020 ⁽²⁹⁾	22	Male	EGPA	Mild persistent asthma, allergic rhinitis	Myocarditis with Cardiogenic shock	VA	168	No complications reported
18	Wang et al., 2021 ⁽³⁰⁾	32	Male	GPA	Chronic occupational exposure to metallic dust and chemical aerosols	DAH with respiratory failure	VV	192	No complications reported

19	Finkel et al., 2022 ⁽³¹⁾	13	Male	GPA	No significant past medical history	DAH with respiratory failure	VV	384	No complications reported
20	Smesseim I et al., 2022 ⁽³²⁾	47	Female	MPO-AAV	No significant past medical history	DAH with respiratory failure	VV	360	Oxygenator membrane thrombosis
21.1	Ohta et al., 2022 ⁽³³⁾	70	Female	MPO-AAV	HTN and dyslipidemia	DAH with respiratory failure	VV	120	Oxygenator membrane thrombosis
21.2		63	Male	MPO-AAV	HTN	DAH with respiratory failure	VV	288	Oxygenator membrane thrombosis
22	Phoophiboon et al., 2023 ⁽³⁴⁾	31	Female	SLE	No significant past medical history	↓ LOC, fever, and hypotension requiring intubation	VV	192	No complications reported
23	Hamana et al., 2023 ⁽³⁵⁾	22	Female	MCTD	Photosensitivity, autoimmune arthritis, and skin rash	Fulminant myocarditis	VA	96	No complications reported
24.1	Depta et al., 2024 ⁽³⁶⁾ (Three cases)	44	Male	PM	No significant past medical history	LV failure with VT and hemodynamic collapse	VA	552	No complications reported
24.2		34	Male	IgG4-RD	Type 2 myotonic dystrophy and IgG4-RD	LV failure with VT and hemodynamic collapse	VA	384	No complications reported
24.3		36	Female	SLE	Raynaud phenomenon; prior COVID-19 vaccination	LV failure	VA	336	No complications reported
25	Tran TH et al., 2025 ⁽³⁷⁾	30	Female	SjS	No significant past medical history	LV failure with sustained polymorphic VT and hemodynamic collapse	VA	96	No complications reported
26	Endo et al., 2026 ⁽³⁸⁾	31	Male	MPA	Atopic dermatitis; no other significant past medical history	DAH with severe respiratory failure	VV	72	No complications reported
27	Jung et al., 2026 ⁽³⁹⁾	62	Female	MPA	No significant past medical history	DAH with severe respiratory failure and renal dysfunction	VV	264	No complications reported

Abbreviations: DM, diabetes mellitus; EGPA, eosinophilic granulomatosis with polyangiitis; ESRD, end-stage renal disease; HIT, heparin-induced thrombocytopenia; HTN, hypertension; IgG4-RD, immunoglobulin G4-related disease; LOC, level of consciousness; LV, left ventricular; MAS, macrophage activation syndrome; MCTD, mixed connective tissue disease; MPO-AAV, myeloperoxidase-ANCA-associated vasculitis; PM, polymyositis; SjS, Sjögren's syndrome; VT, ventricular tachycardia.

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Table 3: ECMO management and disease-directed treatment.

No.	Included publication	Immunosuppressive treatment	Timing relative to ECMO initiation	PLEX	Anticoagulation strategy	Outcome
1	Di Maria et al., 2008 ⁽¹³⁾	IV methylprednisolone, cyclophosphamide, rituximab	Methylprednisolone before ECMO; cyclophosphamide during ECMO; rituximab after decannulation	Yes—5 daily sessions, initiated during ECMO	Citrate anticoagulation; ACT 160–180 s; no significant bleeding during ECMO	Survived
2	Joseph et al., 2011 ⁽¹⁴⁾	High-dose IV methylprednisolone	Before and during ECMO	Yes—7 daily sessions, initiated during ECMO	Reduced-intensity anticoagulation; ACT 180–220 s.	Survived
3	Hohenforst-Schmidt et al., 2013 ⁽¹⁵⁾	IV prednisolone, cyclophosphamide → azathioprine	Prednisolone before ECMO; cyclophosphamide on ECMO day 2	Yes daily ×15 days, then every other day ×21 days; initiated before ECMO	No systemic anticoagulation; ACT 160–200 s	Survived
4.1	Pacheco Claudio et al., 2014 ⁽¹⁶⁾	IV methylprednisolone + cyclophosphamide	Initiated after ECMO	No, not reported	No systemic anticoagulation	Died
4.2		IV methylprednisolone + cyclophosphamide	At ECMO initiation	No, not reported	Initially no systemic anticoagulation; temporary heparin after circuit thrombosis	Survived
5	Patel & Lipchik et al., 2014 ⁽¹⁷⁾	IV methylprednisolone	Initiated after ECMO	No, not reported	Unfractionated heparin; ACT >200 s	Survived
6.1	Yusuff et al., 2015 ⁽¹⁸⁾	IV methylprednisolone + cyclophosphamide	Initiated before ECMO	No, not reported	Minimal anticoagulation; single 5,000 IU IV heparin bolus at cannulation, followed by no systemic anticoagulation for 48 hours.	Survived
6.2		IV methylprednisolone + rituximab + IV immunoglobulin, followed by mycophenolate mofetil	Initiated after ECMO	Yes-5 daily sessions, initiated during ECMO	Initially no systemic anticoagulation after a 2,500 IU IV heparin bolus at cannulation; systemic heparin was introduced on ECMO day 2 because of circuit/oxygenator clot formation, targeting an aPTT ratio of 1.5–1.8	Survived
7	Choi & Kang et al., 2016 ⁽¹⁹⁾	IV methylprednisolone + IV immunoglobulin	initiated before ECMO	Yes, initiated after ECMO; number of sessions not reported	Low-dose UFH (ACT 140–160 s initially, then 180–200 s)	Survived
8	Rawal et al., 2016 ⁽²⁰⁾	IV methylprednisolone + cyclophosphamide	Initiated before ECMO	Yes, initiated after ECMO; number of sessions not reported	Systemic heparin targeting ACT 140–160 seconds	Survived
9	Ammirati et al., 2016 ⁽²¹⁾	IV methylprednisolone 10 mg/kg/day for 3 days, followed by prednisolone 1 mg/kg/day	Initiated after VA-ECMO and diagnostic endomyocardial biopsy	No, not reported	Not reported	Survived
10	Abdelaty et al., 2017 ⁽²²⁾	IV methylprednisolone	Specific timing relative to ECMO not reported	No, not reported	No systemic anticoagulation	Survived

11	Vanoli et al., 2017 ⁽²³⁾	IV methylprednisolone + cyclophosphamide	Initiated during ECMO	Yes-initiated during ECMO; number of sessions not reported	Not reported	Survived
12	Lee CF et al., 2018 ⁽²⁴⁾	Cyclophosphamide + rituximab + methylprednisolone	Initiated during ECMO	Yes, initiated during ECMO; number of sessions not reported	No systemic anticoagulation	Survived
13.1	Delvino et al., 2019 ⁽²⁵⁾ (two cases)	IV methylprednisolone + cyclophosphamide	Initiated during ECMO	Yes-initiated during ECMO; number of sessions not reported	Not reported	Survived
13.2		IV methylprednisolone + cyclophosphamide	Initiated during ECMO	Yes-initiated during ECMO; number of sessions not reported	Not reported	Survived
14	Mabe et al., 2019 ⁽²⁶⁾	IV methylprednisolone	Initiated during ECMO	No	Not reported	Survived
15	Tong et al., 2019 ⁽²⁷⁾	IV methylprednisolone + IVIG + cyclophosphamide	Initiated before ECMO	Yes-initiated before ECMO; number of sessions not reported	Systemic heparin from ECMO initiation; target aPTT 45–50 s, adjusted using thromboelastography	Survived
16	Liu et al., 2020 ⁽²⁸⁾	IV methylprednisolone + IVIG; cyclophosphamide	Initiated after ECMO	No, not reported	Not reported	Survived
17	Lopes et al., 2020 ⁽²⁹⁾	IV methylprednisolone; cyclophosphamide	Methylprednisolone after ECMO initiation; cyclophosphamide later (timing not specified)	No, not reported	Not reported	Survived
18	Wang et al., 2021 ⁽³⁰⁾	Methylprednisolone + IVIG	Initiated before ECMO	Yes-4 sessions. timing relative to ECMO not specified	Unfractionated heparin; ACT approximately 200 s; aPTT approximately 50 s	Survived
19	Finkel et al., 2022 ⁽³¹⁾	IV methylprednisolone + rituximab + IVIG + cyclophosphamide	Methylprednisolone + rituximab + plasma exchange before ECMO; IVIG + cyclophosphamide after ECMO initiation	Yes-5 daily sessions, initiated before ECMO	No anticoagulation for 48 h; low-dose heparin thereafter	Survived
20	Smesseim I et al., 2022 ⁽³²⁾	IV methylprednisolone + cyclophosphamide	Methylprednisolone before ECMO; cyclophosphamide during ECMO	Yes-initiated before ECMO; number of sessions not reported	Anticoagulation initially withheld due to hemoptysis; UFH started after oxygenator thrombosis	Survived
21.1	Ohta et al., 2022 ⁽³³⁾	IV methylprednisolone + cyclophosphamide + rituximab	Methylprednisolone during ECMO; cyclophosphamide and rituximab during ECMO	Yes-2 sessions during ECMO	Initially no systemic anticoagulation; heparin initiated after PLEX stabilized anemia, targeting an aPTT twice the pre-heparin value	Survived
21.2		IV methylprednisolone + rituximab	Methylprednisolone + rituximab during ECMO	Yes-1 session during ECMO	Initially no systemic anticoagulation; heparin initiated on ECMO day 7 after circuit exchange, targeting an aPTT twice the pre-heparin value.	Survived
22	Phoophiboon et al., 2023 ⁽³⁴⁾	IV methylprednisolone + anakinra	Methylprednisolone before and during ECMO; anakinra after ECMO	No, not reported	Not reported	Died
23	Hamana et al., 2023 ⁽³⁵⁾	IV methylprednisolone + prednisolone + IV cyclophosphamide	Methylprednisolone before ECMO; prednisolone + cyclophosphamide during ECMO	No, not reported	Not reported	Survived

24.1	Depta et al., 2024 ⁽³⁶⁾ (Three cases)	Prednisone + azathioprine + IV methylprednisolone + IVIG	Prednisone + azathioprine before ECMO; methylprednisolone during ECMO; IVIG during ECMO	No, not reported	Not reported	Survived
24.2		Prednisone	Prednisone before ECMO	No, not reported	Not reported	Survived
24.3		IV methylprednisolone + prednisone + azathioprine	Methylprednisolone during ECMO; prednisone + azathioprine after ECMO	No, not reported	Not reported	Survived
25	Tran TH et al., 2025 ⁽³⁷⁾	IV methylprednisolone	Methylprednisolone during ECMO	No, not reported	Not reported	Survived
26	Endo et al., 2026 ⁽³⁸⁾	IV methylprednisolone + prednisone + rituximab + avacopan	Methylprednisolone before ECMO; rituximab during ECMO; avacopan after ECMO	Yes-3 daily sessions, initiated before ECMO and continued during ECMO	Not reported	Survived
27	Jung et al., 2026 ⁽³⁹⁾	IV methylprednisolone + IV cyclophosphamide	Methylprednisolone before ECMO; cyclophosphamide during ECMO	No, not reported	Single 1,000 IU IV heparin bolus at cannulation; no systemic anticoagulation for 12 h, followed by low-intensity heparin targeting aPTT 40–50 s	Survived

Abbreviations: ACT, activated clotting time; aPTT, activated partial thromboplastin time; IU, international units; IV, intravenous; IVIG, intravenous immunoglobulin; UFH, unfractionated heparin.

Discussion

The principal finding of this patient-level clinical synthesis is that, in severe systemic autoimmune and vasculitic crises, the acute organ-failure phenotype appeared to be more closely related to the required ECMO configuration than the underlying rheumatologic diagnosis. DAH with respiratory failure was the predominant indication for extracorporeal support and was associated with predominant use of VV-ECMO, whereas VA-ECMO was used primarily in patients with severe cardiac or combined cardiorespiratory failure. Despite substantial diagnostic heterogeneity, the clinical circumstances leading to extracorporeal support were therefore largely characterized by the pattern and severity of acute cardiopulmonary failure. By reconstructing individual clinical trajectories rather than considering rheumatologic diseases as a single group, this clinical synthesis characterizes the relationships among presenting manifestations, ECMO configuration, disease-directed therapy, PLEX, anticoagulation strategies, and reported clinical outcomes. Although vessel-size classification and serological categorization provide crucial diagnostic frameworks, the clinical phenotype during life-threatening decompensation appeared to be more closely related to the specific territory and intensity of inflammatory organ injury [40]. In small-vessel disorders such as GPA, MPA, and MPO-AAV, necrotizing inflammation of the pulmonary microvasculature produces pulmonary capillaritis and fulminant DAH [40, 41]. Similarly, EGPA and SLE can trigger profound alveolar capillary or myocardial damage, manifesting as life-threatening DAH or cardiogenic shock [42-45]. MCTD may

present with life-threatening DAH or acute myocarditis, while MAS generates a hyperinflammatory cytokine storm that may result in severe ARDS, DAH, or circulatory failure [46-48]. Furthermore, cardiac involvement affecting the myocardium or conduction system in PM, IgG4-RD, and SjS can lead to acute LV failure, malignant VT, or hemodynamic collapse [49-52]. Thus, while the underlying immunopathology varied widely among the included patients, these diverse conditions ultimately converged on a shared clinical endpoint: severe pulmonary, cardiac, or combined cardiopulmonary collapse necessitating VV-ECMO or VA-ECMO support. The broad demographic and clinical background of the included patients further illustrates that no uniform baseline clinical profile was apparent among patients who ultimately required ECMO. Patients represented a wide range of ages, and most had no significant prior medical history. Among those with reported pre-existing conditions, the clinical background varied from lupus nephritis and chronic or recurrent sinusitis to cardiomyopathy, hypertension, diabetes mellitus, asthma, allergic disease, Raynaud phenomenon, and environmental inhalational exposure. These conditions were inconsistently represented across the reported cases and did not reveal a recognizable baseline pattern preceding the need for extracorporeal support. Life-threatening cardiopulmonary deterioration may therefore occur either as the presenting manifestation of previously unrecognized systemic autoimmune disease or during an acute exacerbation of established disease. The selection of ECMO configuration closely reflected the predominant pattern of physiological failure. VV-ECMO was predominantly used in the setting of severe respiratory failure, particularly DAH, whereas VA-

ECMO was mainly used when severe cardiac dysfunction, cardiogenic shock, malignant ventricular arrhythmias, or combined cardiopulmonary failure predominated. VV-ECMO provides respiratory support and permits reduction of ventilator intensity in patients with severe pulmonary injury, whereas VA-ECMO provides circulatory support when myocardial function and systemic perfusion are critically compromised [5, 6]. This distinction is clinically important because the required ECMO configuration reflects the predominant physiological consequences of the disease rather than the autoimmune diagnosis itself. The same underlying disease may therefore require different forms of extracorporeal support depending on whether pulmonary, cardiac, or combined organ failure dominates the presentation. Beyond maintaining physiological stability, extracorporeal support may provide a window for diagnostic and therapeutic interventions that would otherwise be difficult to undertake during profound respiratory or circulatory failure. The published cases included diagnostic procedures such as bronchoscopy [15], endomyocardial biopsy [21], and lung biopsy [25], as well as therapeutic interventions including bronchoscopic administration of recombinant factor VIIa and plasma exchange [32]. These experiences suggest that the clinical role of ECMO may extend beyond immediate rescue by providing a period of physiological support during which diagnostic clarification and disease-directed interventions can be undertaken in otherwise critically unstable patients. The duration of extracorporeal support varied considerably across the reported cases, reflecting the heterogeneous clinical trajectories of patients with severe autoimmune and vasculitic disease. This variability may reflect differences in the extent and reversibility of organ injury, disease activity, response to treatment, and the occurrence of intercurrent complications. Because ECMO provides physiological support without directly modifying the underlying autoimmune pathomechanism, liberation from extracorporeal support ultimately requires sufficient recovery or stabilization of native organ function. In life-threatening autoimmune and vasculitic disease, the duration of support may therefore reflect the time required for disease-directed therapy to control the inflammatory process, for injured organs to recover sufficient function, and for associated critical-care complications to be managed. Disease-directed immunosuppression constituted the therapeutic backbone across the reported cases, although the specific regimens and their timing relative to ECMO varied considerably. This therapeutic heterogeneity reflects differences in the underlying disease, pattern and severity of organ involvement, previous treatment exposure, and the clinical circumstances surrounding ECMO initiation rather than evidence favoring one regimen over another. In AAV, glucocorticoids combined with rituximab or cyclophosphamide remain established remission-induction approaches, although the optimal treatment strategy in severe, mechanically ventilated DAH

remains uncertain [40, 41]. Recent literature likewise emphasizes that treatment decisions in severe DAH should be individualized according to disease severity, organ involvement, and the balance between immunosuppressive benefit and treatment-related complications [41]. In the present cohort, the frequent use of corticosteroids and the variable addition of other immunosuppressive therapies are consistent with this individualized approach, while the case-based nature of the evidence does not permit comparison of the relative efficacy of different treatment regimens. The timing of immunosuppressive therapy relative to ECMO was heterogeneous, with treatment initiated before, during, or after extracorporeal support, and several patients receiving therapy across more than one phase. This pattern is clinically relevant because ECMO and immunosuppression address different components of the disease process: extracorporeal support provides temporary physiological support for failing organs, whereas immunosuppressive therapy targets the underlying inflammatory process responsible for organ injury. In rapidly progressive DAH or fulminant myocarditis, clinical deterioration may continue despite initiation of disease-directed therapy, making concurrent organ support and treatment of the underlying inflammatory disease necessary in selected patients. Recent case-based experience in severe AAV-associated DAH similarly illustrates the use of VV-ECMO to maintain adequate gas exchange while induction immunosuppression is administered (39). Importantly, the available evidence cannot determine whether earlier initiation of immunosuppression, a particular treatment regimen, or a specific sequence relative to ECMO is associated with better outcomes. Plasma exchange (PLEX) was incorporated selectively into the management of severe autoimmune and vasculitic presentations, with considerable variation in its indications and timing relative to ECMO and immunosuppressive therapy. This heterogeneity, together with the case-based nature of the available evidence, precludes determining the independent contribution of PLEX to clinical outcomes. This caution is particularly relevant in AAV, where contemporary evidence has not established a consistent clinical benefit sufficient to support routine use of PLEX across all patients, despite its continued consideration in selected severe presentations [41]. Accordingly, the present case-based experience should be interpreted as illustrating how PLEX has been incorporated into individualized management strategies rather than as evidence supporting its routine use in patients receiving ECMO. Anticoagulation represented one of the most challenging aspects of ECMO management, particularly in patients with active DAH, in whom the need to maintain circuit patency must be balanced against the risk of exacerbating pulmonary or systemic bleeding. Anticoagulation strategies varied substantially across the reported cases, ranging from initiation at ECMO commencement to delayed, reduced-intensity, or absent systemic anticoagulation. Circuit or oxygenator thrombosis

was the most frequently reported ECMO-related complication in this cohort. Notably, all reported episodes of circuit or oxygenator thrombosis occurred in patients in whom systemic anticoagulation had initially been withheld or minimized and was subsequently initiated. Although this observation raises concern regarding the thrombotic risks associated with reduced anticoagulation, the small number of events and the descriptive nature of the case reports preclude attribution of thrombosis directly to the anticoagulation strategy. These findings underscore the need for individualized anticoagulation decisions based on the trajectory of bleeding, circuit function, and thrombotic risk. This approach is consistent with emerging evidence from the 2026 RATE randomized trial [53], in which low-dose unfractionated heparin and therapeutic low-molecular-weight heparin were non-inferior to standard-dose unfractionated heparin for a composite of severe bleeding, severe thromboembolic complications, and 6-month mortality in adults receiving ECMO, with fewer severe bleeding events observed in the lower-intensity groups. Accordingly, reduced-intensity anticoagulation may be a reasonable consideration in selected patients with high bleeding risk, including active DAH, whereas anticoagulation-free ECMO remains a more individualized strategy that requires particularly close surveillance for circuit dysfunction and patient-related thrombosis [8-53]. The relatively limited number of reported ECMO-related complications in this case-based cohort should be interpreted cautiously. Case reports and small case series frequently provide incomplete or uneven reporting of secondary complications, and attribution to ECMO, critical illness, the underlying disease, or immunosuppressive therapy is often difficult. Accordingly, the complication profile observed in this review should be regarded primarily as an illustration of the competing bleeding, thrombotic, and critical-care challenges encountered during extracorporeal support rather than as an estimate of their true incidence. The findings should also be considered alongside broader observational and registry-based evidence. In a French monocenter study of 90 critically ill patients with systemic rheumatic diseases requiring ECMO, Bay et al. reported an in-hospital mortality of 51.1%; among patients treated with VV-ECMO, vasculitis was independently associated with more favorable outcomes, whereas markers of greater physiological severity, including higher SAPS II scores and arterial lactate levels, were associated with mortality (54). Similarly, an ELSO registry analysis of 135 patients with AAV supported with ECMO reported pulmonary involvement in 93% and survival to hospital discharge of 73%. Lower survival was associated with older age, VA-ECMO, ECPR, and cardiac arrest before ECMO, whereas pulmonary hemorrhage itself was not associated with worse outcome [55]. Taken together, these larger observational datasets provide an important context for the present case-based findings and suggest that outcomes during ECMO may be influenced substantially by the severity and pattern of

acute physiological failure rather than by the presence of pulmonary hemorrhage alone. The higher survival observed in the present cohort, 93.9% (31/33), should therefore be interpreted cautiously when compared with these larger datasets. The apparent difference is likely influenced by study design, as case reports and small case series selectively capture individual clinical experiences and are particularly susceptible to publication and reporting bias, whereas registry and observational cohorts encompass a broader spectrum of disease severity and clinical outcomes. Differences in patient selection, timing of ECMO initiation, ECMO configuration, institutional expertise, and concomitant treatment further limit direct comparison between these populations. Nevertheless, the available evidence suggests that meaningful survival is possible in selected patients with life-threatening autoimmune or vasculitic disease requiring ECMO, while providing no basis for interpreting the high survival observed in published case reports as an estimate of ECMO effectiveness. The present review should also be distinguished from the broader systematic review by Falaas et al. [56], which synthesized ECMO use in rheumatologic disease across 144 studies comprising 734 patients. The broader review included a wide spectrum of rheumatologic diseases and incorporated both case-based reports and retrospective observational studies, whereas the present review deliberately focused on systemic autoimmune and vasculitic disorders with acute, life-threatening cardiopulmonary decompensation and restricted inclusion to reports providing sufficiently detailed individual-level information. This narrower design enabled reconstruction of individual clinical trajectories, including the relationship between life-threatening presentation, organ-failure phenotype, ECMO configuration, disease-directed therapy, anticoagulation, and clinical outcome. The two reviews therefore address complementary questions rather than providing competing estimates of ECMO effectiveness. Whereas the broader review characterizes the overall published landscape of ECMO use across rheumatologic disease, the present synthesis focuses more specifically on the clinical sequence and management of rare, life-threatening autoimmune and vasculitic presentations. Accordingly, the two reviews should be interpreted as complementary descriptions of the published evidence rather than comparative estimates of treatment effectiveness.

Conclusion

Published case reports and case series suggest that VV-ECMO and VA-ECMO can provide temporary cardiopulmonary support in selected patients with life-threatening systemic autoimmune and vasculitic disorders who develop refractory respiratory, circulatory, or combined cardiopulmonary failure despite conventional management. Across the reported cases, ECMO was integrated with disease-directed therapy, providing physiological support

while immunosuppressive and other disease-specific interventions were instituted and native organ function recovered or stabilized. However, the favorable outcomes observed among the included patients should be interpreted cautiously because of substantial publication and reporting bias, clinical heterogeneity, incomplete patient-level information, and the absence of comparative controls. The available evidence therefore supports the clinical feasibility of ECMO as a supportive component of multidisciplinary management in selected life-threatening presentations, rather than establishing its effectiveness. Prospective multicenter registries and collaborative studies are needed to better define patient selection, optimal timing of ECMO initiation, anticoagulation and concomitant disease-directed treatment strategies, and short- and long-term outcomes in this rare and critically ill population.

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Supplementary material: Supplementary Table S1. Detailed literature search strategy...; Supplementary Table S2. Excluded publications and reasons for exclusion.

Limitations

Several methodological limitations should be considered

when interpreting the findings of this review. The evidence base consisted exclusively of published case reports and small case series, making the synthesis inherently susceptible to publication and reporting bias, whereby successful recoveries and favorable outcomes may be more likely to be reported than unsuccessful or fatal cases. Accordingly, the observed survival proportion of 93.9% (31/33 patients) should not be interpreted as an estimate of expected survival among patients with life-threatening autoimmune or vasculitic disease requiring ECMO. Substantial clinical heterogeneity was also present across the cohort with respect to underlying diagnoses, patterns and severity of cardiopulmonary involvement, ECMO configuration and duration, timing of ECMO initiation, anticoagulation strategies, and concomitant immunosuppressive and PLEX regimens. In addition, the completeness and granularity of patient-level information varied considerably among reports, limiting consistent assessment of baseline disease severity and other clinical variables. Finally, the absence of a non-ECMO comparator group precludes determination of the absolute or incremental benefit of ECMO compared with conventional management alone. The findings should therefore be regarded as a descriptive synthesis of reported patient trajectories and management rather than evidence of comparative treatment effectiveness.

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