Modern Rheumatology, **00**, 2024, 1–6
DOI: https://doi.org/10.1093/mr/roae066
Advance Access Publication Date: 8 August 2024
Original Article

Assessing the efficacy of haemoperfusion for dermatomyositis-associated acute exacerbation of interstitial lung disease: A multicentre retrospective study

Shuqin Sun[a], Yanfei Yu[b], Wenjing Feng[a], Luyan Sun[c] and Shuo Yang[b,*]

[a]Department of Geriatric Medicine, Affiliated Hospital of Qingdao University, Qingdao, China
[b]Department of Critical Care Medicine, Affiliated Hospital of Qingdao University, Qingdao, China
[c]Department of Oral Medicine, Affiliated Hospital of Qingdao University, Qingdao, China

*Correspondence: Shuo Yang; cliffsman@126.com; Department of Critical Care Medicine, Affiliated Hospital of Qingdao University, Qingdao, No.16 Jiangsu Road, Shinan District, Qingdao, Shandong Province 266000, China

ABSTRACT

Objectives: Haemoperfusion (HP) is used to treat various diseases, including sepsis and acute respiratory distress syndrome. However, few studies have explored the efficiency of HP in dermatomyositis-associated acute exacerbation of interstitial lung disease.

Methods: We conducted a retrospective study. Two hundred and sixteen patients with dermatomyositis-associated acute exacerbation of interstitial lung disease were included. Patients were divided into the HP group (treatment group) and the control group. Changes in oxygenation, haemodynamic parameters, lung ultrasound scores, and inflammatory cytokine levels were evaluated before and after HP in the treatment group. The length of intensive care unit (ICU) stays, duration of ventilator therapy, mortality rate, and incidence of complications were compared between the treatment and control groups.

Results: Haemodynamic and oxygenation variables in the treatment group significantly improved after treatment. However, the levels of the inflammatory factors significantly decreased after treatment. The length of ICU stay and the duration of ventilator therapy were significantly shorter in the treatment group than in the control group. The mortality rate of the treatment group was significantly lower than that of the control group.

Conclusions: This study demonstrated that HP could improve treatment efficacy in patients with dermatomyositis-associated acute exacerbation of interstitial lung disease.

KEYWORDS: Dermatomyositis; interstitial lung disease; haemoperfusion

Introduction

Dermatomyositis is a nonpurulent inflammatory disease that mainly affects the striated muscle and is characterized by lymphocyte infiltration [1, 2]. Its clinical symptoms manifest as skin damage, myopathy, and organ dysfunction [3]. In severe cases of dermatomyositis, damage to multiple organs, including the heart, lungs, and kidneys, may occur [3]. Interstitial lung disease is the most common and serious complication of dermatomyositis [4]. Moreover, infections, stress, and other causes can lead to acute exacerbation of interstitial lung disease [4]. Acute exacerbation of interstitial lung disease refers to a significant acute deterioration of respiratory function in patients with interstitial lung disease in a short time (usually within 1 month), mainly characterized by diffuse ground glass opacities and/or consolidation in both lungs newly appearing on chest High Resolution CT against the original background of interstitial lung disease [5]. Its clinical manifestations included fever, cough, and shortness of breath, followed by respiratory failure [4]. A previous study indicated that the cause of death in most patients with dermatomyositis was respiratory failure induced by acute exacerbation of interstitial lung disease [6]. A previous study also demonstrated multiple inflammatory mediators involved in acute exacerbation of dermatomyositis-associated interstitial lung disease, including tumour necrosis factor-alpha (TNF-α), interleukin (IL)-6, and IL-1 [7].

Haemoperfusion (HP) can effectively adsorb inflammatory cytokines and autoimmune antibodies [8]. Furthermore, HP is widely used to treat poisoning, inflammation, trauma, sepsis, and autoimmune diseases [9]. However, very few studies have evaluated the efficacy of HP for the acute exacerbation of dermatomyositis-associated interstitial lung disease.

This study investigated the efficacy of HP for dermatomyositis-associated acute exacerbation of interstitial lung disease. Outcomes included oxygenation and haemodynamic parameters after treatment. Secondary outcomes, including length of intensive care unit (ICU) stay, duration of ventilator therapy, and mortality rate, were also assessed.

Received 29 February 2024; Accepted 26 June 2024

Materials and methods

Patient selection

In this multicentre retrospective study, 216 patients with dermatomyositis-associated acute exacerbation of interstitial lung disease admitted to the Department of Critical Care Medicine with 160 beds and 4 branch centres in different regions at the Affiliated Hospital of Qingdao University between June 2020 and June 2023 were enrolled (shown in Figure 1). All patients were ≥18 years of age, and all of them received ventilator therapy (invasive mechanical ventilation or high-flow nasal cannula). All patients underwent Corona Virus Disease -19 nasopharyngeal swab and tested negative. The dermatomyositis extrapulmonary manifestations of patients were described as follows: cutaneous manifestations, myopathy, vasculitis, and gastrointestinal manifestation. Among the 216 patients in the study, 184 cases were anti-MDA-5 antibody-positive subtype and the other 32 cases were anti-ARS antibody-positive subtype. The patients who underwent HP were included in the treatment group ($n = 98$), whereas those who refused HP therapy (economic reasons, etc.) were included in the control group ($n = 118$). According to the guidelines for acute exacerbation of interstitial lung disease, each patient was given high-dose methylprednisolone (1000 mg/d) intravenously for initial 3 consecutive days. After high-dose treatment, the hormone maintenance dose was 40 mg/d. The patient was treated with a maintenance dose of 40 mg/d for 4 weeks. All patients were not treated with immunosuppressive agents during ICU stay. Patient demographic data were obtained from medical and nursing electronic medical records, laboratory results, and imaging systems.

The haemodynamic and oxygenation parameters before and after all rounds of HP treatment were compared between the treatment groups. Clinical data of before and after treatment were collected at the time of admission and 1 h after the last HP procedure, respectively. Indicators including the length of ICU stay, duration of ventilator therapy, mortality rate, and complications were compared between the treatment and control groups.

Implementation of HP

A single-needle double-lumen tube was inserted into the patient's femoral or internal jugular vein for HP. A Fresenius blood filtration system machine (Fresenius, Germany) and an HA280 cartridge (Jafron Biomedical, Zhuhai, China) were used for HP, with blood flow controlled within the range of 180–200 ml/min. The patient was treated with first HP session for 4 h, which was repeated once more within 24-h intervals. All patients were given HP treatment 4 times, if possible. Blood coagulation, blood pressure, heart rate, respiratory rate, and oxygen saturation were monitored during HP. For patients treated with mechanical ventilation, they underwent intubation immediately after being admitted to the Department of Critical Care Medicine. All patients with mechanical ventilation treatment were in an intubation state during HP therapy.

Pulse indicator continuous cardiac output study

Patients were required to complete the pulse indicator continuous cardiac output (PICCO) monitoring within 3 h of ICU admission. Ductus arteriosus was placed in the femoral artery to measure the continuous cardiac output (CO), arterial blood pressure, and other parameters. A venous catheter was placed in the superior vena cava, and the temperature response curve was calculated.

Lung ultrasound study

Lung ultrasound assessment was performed using a Sonosite X-Porte and a 2–5 MHz curved array probe (FUJIFILM SonoSite, USA). The ultrasonic examination was performed by one of three experienced ICU physicians. Twelve points in the body longitudinal axis were checked during lung ultrasound assessment for each patient according to the lung ultrasound systematic protocol. Moreover, three experienced ICU physicians scored the ultrasound images. Lung ultrasound patterns and lung aeration were described as four categories: (1) normal patterns (N): normal aeration corresponding to the presence of lung sliding with A lines or fewer than two isolated B lines; (2) B1: moderate loss of lung aeration corresponding to multiple well-defined B lines or spaced ultrasound lung called 'comet-tail artefact'; (3) B2: severe loss of lung aeration corresponding to multiple coalescent B lines or multiple abutting ultrasound lung comet tails issued from the pleural line; and (4) C: lung consolidation corresponding to the presence of a tissue pattern containing hyperechoic punctiform images representative of air bronchograms, presence or absence of regional pulmonary blood flow, and/or dynamic bronchograms. The lung ultrasound scores (LUSsc) of each region were calculated based on the worst image value. The scores for each pattern are as follows: N = 0, B1 = 1, B2 = 2, and B3 = 3 [10, 11].

Statistical analysis

Continuous variables were presented as mean ± standard deviation or median (interquartile range). Categorical variables were presented as frequencies (percentages). The chi-square test was used to compare categorical variables. Student's *t*-test, Mann–Whitney U test, or Wilcoxon signed rank test was performed to evaluate the differences between the groups. Paired *t*-test was applied for comparing variables before and after HP treatment. Kaplan–Meier survival curves were plotted, and the log-rank test was used to compare 30-day mortality rates. The Cox proportional-hazards model was used to analyse risk factors for 30-day mortality. Sequential Organ Failure Assessment (SOFA) cardiac, SOFA respire, and age were considered as variables of multivariate analysis. Statistical significance was set at $P < .05$. Statistical analyses were performed using SPSS (version 26.0; IBM Corp., USA).

Results

The demographic characteristics of the patients are presented in Table 1. No significant differences were observed in characteristics between the treatment and control groups.

Haemodynamic variables

Haemodynamic variables before and after HP were compared in the treatment group (Table 2). The results indicated no significant changes in heart rate (75.99 ± 23.27 vs 78.64 ± 23.73, $P > .05$), systolic blood pressure (121.66 ± 24.91 vs 125.68 ± 23.45, $P > .05$), and diastolic blood pressure (71.55 ± 11.80 vs 68.65 ± 11.00, $P > .05$) after

Table 1. Characteristics of demographics.

Characteristics ($n = 216$)	Treatment ($n = 98$)	Control ($n = 118$)	*P* value
Sex (male)	53 (54.08)	50 (42.37)	.089
Age, years	50.18 ± 17.25	49.89 ± 16.64	.899
Comorbidities			
Chronic heart failure	23 (23.47)	25 (21.19)	.688
Chronic liver disease	19 (19.39)	15 (12.71)	.181
Chronic renal disease	10 (10.20)	15 (12.71)	.284
Chronic obstructive pulmonary disease	33 (33.67)	38 (32.20)	.819
Asthma	6 (6.12)	5 (4.23)	.485
Hypertension	32 (32.65)	27 (22.88)	.109
Diabetes	27 (27.55)	36 (30.51)	.634
Cerebrovascular disease	8 (8.16)	11 (9.32)	.765
Laboratory features			
White blood cells ($\times 10^9$/l)	10.41 (10.01–11.04)	10.71 (9.94–11.24)	.276
Neutrophils ($\times 10^9$/l)	8.27 (8.03–8.59)	8.35 (8.10–8.57)	.445
Lymphocyte ($\times 10^9$/l)	1.83 (1.74–1.90)	1.85 (1.78–1.89)	.289
Platelet ($\times 10^9$/l)	218.50 (206.50–234.00)	224.00 (210.75–236.00)	.172
CRP (mg/l)	128.30 (104.45–167.90)	140.74 (131.90–150.48)	.105
ALT (U/l)	20.00 (18.00–23.00)	21.20 (17.70–23.70)	.335
AST(U/l)	25.50 (22.00–29.00)	26.00 (23.00–29.25)	.211
TBil (μmol/l)	7.14 (6.45–7.85)	7.37 (6.86–7.84)	.088
BUN (mmol/l)	10.68 (9.55–11.79)	10.94 (9.28–11.87)	.863
Creatinine (μmol/l)	100.68 ± 8.57	102.46 ± 9.12	.143
Admission SOFA score	5 (4–7)	5 (3–7)	.487
Admission APACHE II score	14.12 ± 5.59	14.03 ± 5.50	.898
MV	63 (64.28)	72 (61.02)	.621
HFNC	35 (35.71)	46 (38.98)	.621
Initial P/F ratio	123.70 ± 19.08	127.74 ± 21.54	.150

Abbreviations: ALT, alanine aminotransferase; AST, aspartate aminotransferase; BUN, blood urea nitrogen; CRP, C-reactive protein; HFNC, high-flow nasal cannula oxygen therapy; MV, mechanical ventilation; TBil, total bilirubin.

HP. The results of the PICCO variable demonstrated that CO (2.99 ± 1.07 vs 3.65 ± 1.02, $P < .01$) and global ejection fraction (13.89 ± 3.23 vs 17.19 ± 3.43, $P < .01$) significantly improved after treatment. Both cardiac function index (CFI) and cardiac power index (CPI) were parameters used in PICCO technology to assess cardiac function. CFI was defined as the ratio of CO to the global end-diastolic volume [12]. It reflected the relationship between cardiac pumping function and myocardial cell stretch length [13]. CFI could evaluate total cardiac contractility and be applied for early diagnosis of heart failure. CPI was calculated as the product of mean arterial pressure and cardiac index divided by 451. CPI was related to blood flow velocity and vascular resistance [14]. It was an indicator for evaluating the overall heart condition [15]. The results of CPI and CFI also showed improvement (CPI, 0.49 ± 0.03 vs 0.50 ± 0.04, $P < .01$; CFI, 4.04 ± 0.28 vs 4.16 ± 0.37, $P < .05$). Extravascular lung water index (ELWI) (14.08 ± 1.83 vs 11.69 ± 2.02, $P < .01$) significantly reduced after treatment.

Table 2. Haemodynamic variables.

Haemodynamic variables (treatment group)	Before treatment	After treatment	*P* value
Heart rate	75.99 ± 23.27	78.64 ± 23.73	.431
SBP (mmHg)	121.66 ± 24.91	125.68 ± 23.45	.250
DBP (mmHg)	71.55 ± 11.80	68.65 ± 11.00	.097
CO (l/min)	2.99 ± 1.07	3.65 ± 1.02	<.001*
GEF (%)	13.89 ± 3.23	17.19 ± 3.43	<.001*
ELWI (ml/kg)	14.081.83	11.69 ± 2.02	<.001*
CPI (W/m^2)	0.49 ± 0.03	0.50 ± 0.04	.006*
CFI (l/min)	4.04 ± 0.28	4.16 ± 0.37	.022*

*P <.05. Abbreviations: CPI, cardiac work index; DBP, diastolic blood pressure; GEF, global infective fraction; SBP, systolic blood pressure.

Table 3. Oxygenation parameters.

Oxygenation parameters (treatment group)	Before treatment	After treatment	*P* value
pH value	7.42 ± 0.045	7.41 ± 0.049	.372
PO_2	61.09 ± 7.42	67.19 ± 6.87	<.001*
PCO_2	39.67 ± 2.84	39.80 ± 2.87	.776
Lactic acid	1.43 ± 0.53	1.45 ± 0.60	.745
P/F ratio	141.50 (115.86–167.86)	155.77 (132.67–176.82)	<.001*
LUSsc	25.67 ± 6.24	18.54 ± 5.56	<.001*

*P <.05. Lung ultrasonography has been validated for the diagnosis and evaluation of various changes in lung morphology and oxygenation. The results demonstrated the LUSsc significantly decreased after treatment (25.67 ± 6.24 vs 18.54 ± 5.56.

Oxygenation parameters

Blood gas analysis indicators and lung ultrasound were used to evaluate changes in oxygenation before and after HP treatment (Table 3). The blood gas analysis results indicated that partial pressure of oxygen (PO_2) (61.09 ± 7.42 vs 67.19 ± 6.87, $P < .01$) and P/F ratio [141.50 (115.86–167.86) vs 155.77 (132.67–176.82), $P < .01$] significantly increased after HP. Additionally, no statistically significant changes were observed in pH value (7.42 ± 0.045 vs 7.41 ± 0.049, $P > .05$), partial pressure of carbon dioxide (PCO_2) (39.67 ± 2.84 vs 39.80 ± 2.87, $P > .05$), and lactic acid (1.43 ± 0.53 vs 1.45 ± 0.60, $P > .05$).

Changes in inflammatory cytokines

We also explored the changes in inflammatory cytokine levels (Table 4). C-Reactive protein levels significantly decreased after treatment (136.63 ± 37.44 vs 110.19 ± 25.43, $P < .01$). The result indicated that in the treatment group, levels of

Table 4. Inflammatory cytokines.

inflammatory cytokines (treatment group)	Before treatment	After treatment	*P* value
CRP (mg/l)	136.63 ± 37.44	110.19 ± 25.43	<.001*
IL-1β (pg/ml)	120.67 ± 11.48	69.12 ± 5.80	<.001*
IL-6 (pg/ml)	50.66 ± 5.50	25.13 ± 2.78	<.001*
IL-8 (pg/ml)	26.07 ± 1.14	25.79 ± 0.06	.016*
TNF-α (pg/ml)	59.76 ± 5.99	43.06 ± 4.16	<.001*
IL-4 (pg/ml)	0.89 ± 0.26	0.96 ± 0.23	.042*
IL-10 (pg/ml)	0.45 ± 0.05	0.47 ± 0.05	<.001*

*P <.05. Abbreviation: CRP, C-reaction protein.

Table 5. Prognosis and complication.

	Treatment	Control	*P* value
Length of ICU stay (days)	11.61 ± 4.13	16.11 ± 5.94	<.001*
Ventilator duration (days)	8.40 ± 2.84	11.55 ± 3.95	<.001*
Mortality rate (ICU stay)	10 (10.20)	24 (20.03)	<.001*
Complications			
Acute heart failure	6 (6.12)	18 (15.25)	.020*
Acute renal failure	5 (5.10)	17 (14.41)	.024*
Acute liver failure	5 (5.10)	14 (12.71)	.081

*P <.05

IL-6, TNF-α, IL-8, and IL-1 beta (β) decreased significantly after treatment (IL-6, 50.66 ± 5.50 vs 25.13 ± 2.78, P < .01; TNF-α, 59.76 ± 5.99 vs 43.06 ± 4.16, P < .01; IL-8, 26.07 ± 1.14 vs 25.79 ± 0.06, P < .05; IL-1β, 120.67 ± 11.48 vs 69.12 ± 5.80, P < .01). On the other hand, the levels of anti-inflammatory cytokines IL-4 and IL-10 were elevated (IL-4, 0.89 ± 0.26 vs 0.96 ± 0.23, P < .05; IL-10, 0.45 ± 0.05 vs 0.47 ± 0.05, P < .01).

Prognosis and complication assessment

The effect of treatment on patient prognosis and complications was worth noting as well. The length of ICU stays, duration of ventilator therapy, mortality rate, and incidence of complications were compared between the two groups (Table 5). The length of ICU stay was significantly shorter in the treatment group than in the control group (11.61 ± 4.13 vs 16.11 ± 5.94, P < .01). The ventilator therapy duration in the treatment group was significantly reduced compared to the control group (8.40 ± 2.84 vs 11.55 ± 3.95, P < .01). The mortality rate during the ICU stay in the treatment group was significantly lower than that in the control group [10 (10.20) vs. 24 (20.03), P < .01]. The incidence of acute organ failure was significantly lower in the treatment group than in the control group [acute heart failure: 6 (6.12) vs. 18 (15.25), P < .05; acute renal failure: 5 (5.10) vs. 17 (14.41), P < .05]. Survival curve analysis revealed that the 30-day mortality rate was significantly lower in the treatment group than in the control group [18 (18.37) vs 36 (30.51), P < .05] (shown in Figure 2). The Cox proportional-hazards model (Table 6) indicated that older age and high SOFA cardiac score were associated with 30-day mortality of whole patient.

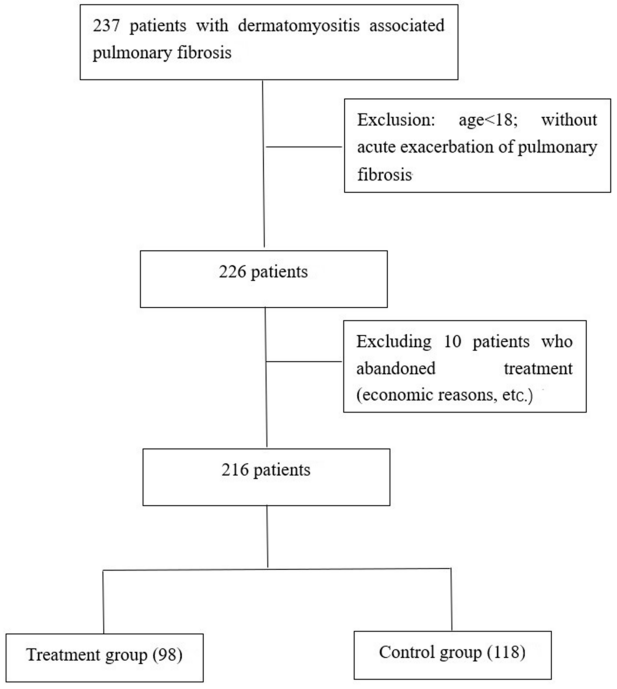

Figure 1. Patient selection scheme.

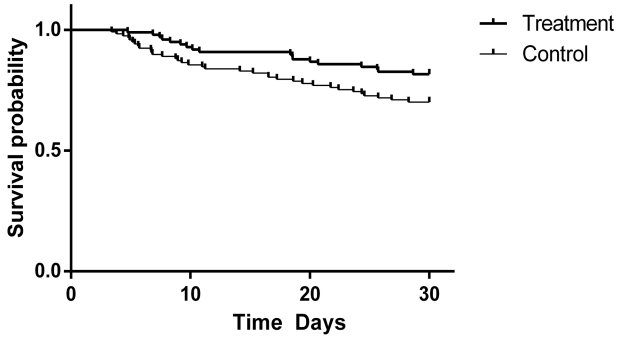

Figure 2. A Kaplan–Meier survival curve analysis.

Table 6. Risk factors for 30-day mortality in whole patients (Cox regression).

Variables	Beta coefficient	Exp(*B*) Odd Ratio	95% Confidence interval for exp(*B*) Low	High	*P* value
Age	0.022	1.023	1.005	1.041	.014*
SOFA cardio	0.898	2.453	1.961	3.069	<.001*

*P <.05

Discussion

Dermatomyositis is an autoimmune connective tissue disease that primarily affects the skin and muscles [16]. Various diseases can complicate dermatomyositis among which

pulmonary interstitial fibrosis is a common and serious complication [17]. The mortality rate increases significantly when dermatomyositis is complicated by pulmonary interstitial fibrosis [18]. The main lesion site of dermatomyositis complicated by pulmonary fibrosis is the pulmonary interstitium, which can manifest as a cough and dyspnoea after exercise [19]. Infection is one of the main causes of acute exacerbation of pulmonary fibrosis in dermatomyositis, which can quickly lead to respiratory failure in patients [20]. During acute exacerbation of interstitial lung disease, the oxygenation ability of the patient is significantly impaired and may progress to multiple organ dysfunction [21].

Previous studies demonstrated that inflammatory cytokines play an important role in the acute exacerbation of pulmonary interstitial fibrosis in patients with dermatomyositis [22]. Gono et al. suggested that the activity of dermatomyositis and interstitial lung disease is associated with elevated levels of TNF-α and IL-6 [23]. Sun et al. indicated that IL-1 involved dermatomyositis-associated acute exacerbation of interstitial lung disease [24]. The increase in vascular permeability caused by inflammatory factors can lead to changes in haemodynamics and oxygenation and even progress to shock [16]. Therefore, effective therapy of early clearance of inflammatory factors is crucial for the efficacy and prognosis of patients. However, there are still limited studies on the role of HP in the dermatomyositis-associated acute exacerbation of pulmonary interstitial fibrosis.

Inflammatory cytokines and toxins are effectively removed by HP [25]. He et al. reported that HP significantly reduced the levels of IL-6, IL-8, and IL-10 in patients undergoing cardiopulmonary bypass [26]. Zhu et al. suggested after HP, the levels of IL-6 and TNF-α in children with Henoch-Schönlein purpura decreased significantly [27]. The present study indicated that the levels of inflammatory cytokines including IL-1, IL-6, IL-8, and TNF-α significantly decreased after HP.

Acute exacerbation of pulmonary fibrosis can lead to organ oedema and rapid deterioration of oxygenation and haemodynamic indicators in patients [28]. We evaluate patient oxygenation and haemodynamic variables through PICCO and blood gas analysis. The present study found that the oxygenation and haemodynamic parameters, including PO_2, P/F ratio, CO, and ELWI, also significantly improved after HP.

Lung ultrasound has been validated for the diagnosis and evaluation of various lung diseases including pulmonary oedema, pneumonia, and acute respiratory distress syndrome (ARDS) [29, 30]. The aforementioned procedure is noninvasive, simple, and easy to perform. Previous studies indicated that LUSsc can be used to assess lung recruitment in patients with ARDS [31, 32]. Bouhemad et al. indicated that LUSsc is an ideal decision-making tool for antimicrobial therapy in ventilator-associated pneumonia [33]. Baldi et al. demonstrated that LUSsc can be used to monitor pulmonary oedema after lung ventilation [34]. In the present study, lung ultrasound manifestations before and after HP treatment were analysed, and the results demonstrated a significant improvement in the LUSscs of patients.

Dermatomyositis-associated acute exacerbation of pulmonary interstitial fibrosis seriously affects patient prognosis [35, 36]. Previous studies showed that it had a high mortality rate [37]. Accordingly, the study further focused on the overall prognosis and incidence of complications in both groups. It is worth mentioning that compared to the control group, the ICU stay length and duration of ventilator therapy were significantly shortened in the treatment group. The mortality during ICU stay and 30-day mortality rate were significantly decreased in the treatment group. This study also demonstrated that the incidence of complications decreased significantly in the treatment groups. It is probably attributed to the reduction in inflammatory response–alleviated organ damage.

This study has some limitations. First, the sample size was insufficiently large. Further study with larger sample sizes should be encouraged. Second, formal protection against HP is still lacking. Third, the study was a retrospective study. Randomized controlled studies in this field are worth exploring in the future.

In conclusion, this study identified that HP effectively cleared inflammatory cytokines and improved lung morphology and oxygenation in patients with dermatomyositis-associated acute exacerbation of interstitial lung disease. Moreover, HP improved patient prognosis and reduced mortality rates. As a promising treatment, the clinical application of HP for dermatomyositis-associated acute exacerbation of interstitial lung disease is worthy of further exploration.

Acknowledgements

We are grateful to Yili Wu and Sumei Li (Statistics Teaching and Research Office of Qingdao University) for their assistance in data analysis.

Conflict of interest

None declared.

Funding

This study was supported by the Youth Fund of Qingdao University Affiliated Hospital (No. QDFYQN202102035).

Data availability

All data generated or analysed during this study are included in this article. Further inquiries can be directed to the corresponding author.

Ethical approval

This study was approved by the Ethics Committee of the Affiliated Hospital of Qingdao University (No. QYFY WZLL 28061). Informed consent was waived for this study.

Author contributions

S.Y., W.F., and L.S. collected and analysed the data. S.Y. drafted the manuscript. S.Y. and Y.Y. conceived and designed the study. S.S. reviewed and edited the manuscript. All authors have read and approved the final manuscript. S.Y. and S.S. confirmed the authenticity of all the raw data.

References

[1] DeWane ME, Waldman R, Lu J. Dermatomyositis: clinical features and pathogenesis. *J Am Acad Dermatol* 2020;**82**:267–81.

[2] Palmucci S, Di Mari A, Cancemi G *et al*. Clinical and radiological features of interstitial lung diseases associated with polymyositis and dermatomyositis. *Medicina* 2022;**59**:58.

[3] Li H, Cai HR. The relationship between immunosuppressive therapy and clinical isolates for patients diagnosed with dermatomyositis-related interstitial lung diseases. *Int Immunopharmacol* 2022;**113**:109351.

[4] Wang H, Lv J, He J *et al*. The prevalence and effects of treatments of rapidly progressive interstitial lung disease of dermatomyositis/polymyositis adults: a systematic review and meta-analysis. *Autoimmun Rev* 2023;**22**:103335.

[5] Collard HR, Ryerson CJ, Corte TJ *et al*. Acute exacerbation of idiopathic pulmonary fibrosis. An international working group report. *Am J Resp Crit Care* 2016;**194**:265–75.

[6] Waseda Y. Myositis-related interstitial lung disease: a respiratory physician's point of view. *Medicina* 2021;**57**:599.

[7] Lou Y, Zheng Y, Fan B *et al*. Serum levels of interleukins and S100A8/A9 correlate with clinical severity in patients with dermatomyositis-associated interstitial lung disease. *BMC Pulm Med* 2020;**20**:196.

[8] Kielstein JT. Hemoperfusion and plasmapheresis in the intensive care unit. *Wien Klin Mag* 2023;**1**:1–6.

[9] Grafe C, Weidhase L, Liebchen U *et al*. Hemoperfusion in anesthesia and intensive care medicine: benefits, risks, and evidence for different systems. *Anaesthesiologie* 2023;**72**:843–51.

[10] Prat G, Guinard S, Bizien N *et al*. Can lung ultrasonography predict prone positioning response in acute respiratory distress syndrome patients? *J Crit Care* 2016;**32**:36–41.

[11] Smit MR, Hagens LA, Heijnen N *et al*. Lung ultrasound prediction model for acute respiratory distress syndrome: a multicenter prospective observational study. *Am J Respir Crit Care Med* 2023;**207**:1591–601.

[12] Su WL, Shui HA, Lan CC *et al*. Cardiovascular parameters associated with troponin i as indicators for 14-day mortality in patients with septic shock. *Am J Med Sci* 2018;**356**:244–53.

[13] Meybohm P, Gruenewald M, Renner J *et al*. Assessment of left ventricular systolic function during acute myocardial ischemia: a comparison of transpulmonary thermodilution and transesophageal echocardiography. *Minerva Anestesiol* 2011;**77**:132–41.

[14] Chen WY, Zhang ZH, Tao LL *et al*. Afterload-related cardiac performance identifies cardiac impairment and associates with outcome in patients with septic shock: a retrospective cohort study. *J Intensive Care* 2021;**9**:33.

[15] Huber W, Fuchs S, Minning A *et al*. Transpulmonary thermodilution (tptd) before, during and after sustained low efficiency dialysis (sled). A prospective study on feasibility of tptd and prediction of successful fluid removal. *PLoS One* 2016;**11**:e153430.

[16] Zhao L, Shen C, Xie S *et al*. The role and mechanism of myeloperoxidase in dermatomyositis. *Int Immunopharmacol* 2023;**124**:110803.

[17] Dsouza KG, Alexander AS, Watts JJ *et al*. Management of interstitial lung disease in patients with autoimmune disease-related interstitial lung disease. *Multidiscip Respir Med* 2023;**18**:890.

[18] Zhang H, Liang R, Yuan X *et al*. Serum IgA levels for predicting the development of rapidly progressive interstitial lung disease in dermatomyositis. *Respir Med* 2023;**216**:107322.

[19] Wang Q, Gao C, Zhang C *et al*. Tumor markers are associated with rapidly progressive interstitial lung disease in adult-dermatomyositis. *Clin Rheumatol* 2022;**41**:1731–9.

[20] Lou Y, Wei Q, Fan B *et al*. The composition of the lung microbiome differs between patients with dermatomyositis and rheumatoid arthritis associated with interstitial lung disease. *FEBS Open Bio* 2022;**12**:258–69.

[21] Li Y, Li Y, Wang Y *et al*. A clinical risk model to predict rapidly progressive interstitial lung disease incidence in dermatomyositis. *Front Med Lausanne* 2021;**8**:733599.

[22] Brunasso AM, Aberer W, Massone C. New onset of dermatomyositis/polymyositis during anti-TNF-alpha therapies: A systematic literature review. *ScientificWorldJournal* 2014;**2014**:179180.

[23] Gono T, Kaneko H, Kawaguchi Y *et al*. Cytokine profiles in polymyositis and dermatomyositis complicated by rapidly progressive or chronic interstitial lung disease. *Rheumatology* 2014;**53**:2196–203.

[24] Sun Y, He Y, Liu Y *et al*. Biologics in polymyositis and dermatomyositis-associated interstitial lung disease. *Curr Pharm Biotechnol* 2014;**15**:521–4.

[25] Ronco C, Samoni S, Bellomo R. Hemoperfusion and immunomodulation. *Contrib Nephrol* 2023;**200**:142–8.

[26] He Z, Lu H, Jian X *et al*. The efficacy of resin hemoperfusion cartridge on inflammatory responses during adult cardiopulmonary bypass. *Blood Purif* 2022;**51**:31–7.

[27] Zhu Y, Dong Y, Wu L *et al*. Changes of inflammatory mediators and oxidative stress indicators in children with Henoch-Schonlein purpura and clinical effects of hemoperfusion in the treatment of severe Henoch-Schonlein purpura with gastrointestinal involvement in children. *BMC Pediatr* 2019;**19**:409.

[28] Luppi F, Sebastiani M, Salvarani C *et al*. Acute exacerbation of interstitial lung disease associated with rheumatic disease. *Nat Rev Rheumatol* 2022;**18**:85–96.

[29] Campbell SJ, Bechara R, Islam S. Point-of-care ultrasound in the intensive care unit. *Clin Chest Med* 2018;**39**:79–97.

[30] Mongodi S, De Luca D, Colombo A *et al*. Quantitative lung ultrasound: technical aspects and clinical applications. *Anesthesiology* 2021;**134**:949–65.

[31] Chen WT, Wang HY, Ye QH *et al*. The diagnostic values of lung ultrasound for ARDS. *Eur Rev Med Pharmacol Sci* 2022;**26**:6642–6.

[32] Bitker L, Talmor D, Richard JC. Imaging the acute respiratory distress syndrome: past, present and future. *Intensive Care Med* 2022;**48**:995–1008.

[33] Bouhemad B, Liu ZH, Arbelot C *et al*. Ultrasound assessment of antibiotic-induced pulmonary reaeration in ventilator-associated pneumonia. *Crit Care Med* 2010;**38**:84–92.

[34] Baldi G, Gargani L, Abramo A *et al*. Lung water assessment by lung ultrasonography in intensive care: a pilot study. *Intensive Care Med* 2013;**39**:74–84.

[35] Mathai SC, Danoff SK. Management of interstitial lung disease associated with connective tissue disease. *BMJ* 2016;**352**:h6819.

[36] Toyoda Y, Hanibuchi M, Kishi J *et al*. Clinical features and outcome of acute exacerbation of interstitial lung disease associated with connective tissue disease. *J Med Invest* 2016;**63**:294–9.

[37] Solomon JJ, Fischer A. Connective tissue disease-associated interstitial lung disease: a focused review. *J Intensive Care Med* 2015;**30**:392–400.

Modern Rheumatology, 2024, 00, 1–6, DOI: https://doi.org/10.1093/mr/roae066, Advance Access Publication Date: 8 August 2024, Original Article