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Early View

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Extracorporeal life support as a bridge to lung transplantation strategy in anti-MDA5+ rapidly progressive interstitial lung disease is life-saving but with persistent difficulties at the bedside

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To the Editor:

In a recent issue of the journal, Bay *et al*. reported fifteen patients with anti–melanoma differentiation–associated protein 5 (anti-MDA5+) rapidly progressive interstitial lung disease (RP-ILD) requiring Bayextracorporeal life support (ECLS) [1]. In five patients, ECLS implementation allowing a bridge to emergency lung transplantation within a median delay of 8 days led to survival whereas in the ten others, fatal outcome occurred since no lung transplantation was possible, either in the absence of listing for transplantation by the medical team in charge or due to disqualification related to underlying morbidities.

We would like to congratulate the authors for their impressive findings demonstrating that emergency lung transplantation should be acknowledged as the only chance of survival in anti-MDA5+ RP-ILD patients referred with life-threatening respiratory failure requiring ECLS. However, we wish to comment on some practical aspects, which may help encouraging physicians in charge considering ECLS as bridge to lung transplantation, since this strategy is still not consensually valued as the unique rescue emerging therapy to propose to MDA5+ RP-ILD patients with refractory respiratory failure [2].

First, given the considerable survival benefits now established in this specific indication and since early diagnosis of such a rare disease in young patients with fulminant and irreversible phenotype is needed urgently, emergency screening of the highly specific anti-MDA5 antibodies may represent a limiting condition to register RP-ILD patients on the high-emergency waiting list for lung transplantation. The role of tertiary expert centres, to which patients should thus be referred, has to be acknowledged, knowing additionally that availability of anti-MDA5 antibody testing remains a limiting step.

Second, a high-rate diagnosis of MDA5+ dematomyositis at the time of ECLS initiation was reported in both survivors (80%) and non-survivors (40%) in Bay's work [1]. Interestingly, skin lesions including characteristic ulcerations of Gottron's papules, painful palmar papules, oral ulcerations and alopecia, mostly recognised as pathognomonic in MDA5+ dematomyositis in RP-ILD patients [3], were observed with an elevated prevalence (60%) similarly in survivors and non-survivors. Skin lesions frequently represent a key trigger for emergency anti-MDA5 antibody screening. We thus suggest that these cutaneous lesions should be considered as a surrogate diagnostic marker if MDA5 antibody testing is not available on time in order not to delay the implementation of ECLS support as a bridge-to-transplantation strategy in critically ill patients at high risk of complications.

Finally, given the limited information provided on post-lung transplantation follow-up in the five survivors described in this work [1], establishing future recommendations to manage anti-MDA5+ RP-ILD patients will need to determine the exact risks of infection, graft rejection and disease recurrence, the requirement of adjunctive immunomodulatory therapies and long-term life expectancy for these

very particular lung transplant recipients. Bay's interesting preliminary findings should thus be completed by long-term outcome data to definitively convince physicians in charge, since almost half of the patients on ECLS were not transplanted as just not listed for lung transplantation.

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