

MACROPHAGE ACTIVATION SYNDROME, RARE CAUSE OF ACUTE LIVER FAILURE WITH POOR PROGNOSIS

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Poster

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A retrospective case series of eight patients with macrophage activation syndrome presenting as acute liver failure found that all four patients meeting King's College criteria for transplantation died despite intensive support, while four patients responding to corticosteroids achieved long-term remission with etoposide.

KEY POINTS

- Eight patients with MAS-related acute liver failure were identified between January 2005 and November 2022, with underlying causes including EBV infection in five cases and adult Still disease in three cases
- All eight patients had strongly elevated circulating ferritin (average 19321 µg/l), leukocytosis, and thrombocytopenia, and all but one had at least one negative bone marrow or lymph node examination prior to definitive histological diagnosis
- Four patients met King's College criteria for urgent liver transplantation; two were transplanted but all four died on average 34 days after symptom onset despite mechanical ventilation, renal replacement therapy, and circulation support
- Four patients who responded to high-dose corticosteroid therapy achieved long-term remission after specialized haematological treatment with etoposide
- The authors suggest that liver transplantation should be carefully considered in MAS patients meeting King's College criteria given the observed mortality in this series

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