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# A rare and catastrophic manifestation of hyperinflammation; haemophagocytic syndrome (HPS); analysis of 21 patients from Turkey

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## Objectives
Hemophagocytic syndrome (HPS) is a catastrophic and life-threatening clinical condition caused by exaggerated immune response along with the tissue destruction due to different etiological factors.. In this study, we aim to investigate the clinical features and outcome of patients of HPS patients in tertiary hospital institutions from Turkey.

## Methods
We contacted the university hospital centres ( department of haemotology,rheumatology and infectious diseases) at Turkey with e-mail. They were asked to fill in the clinical (symptoms and signs), laboratory and treatment data from HPS patients records on an excel file retrospectively. Clinical features and treatment outcomes of 21 patients were retrospectively analysed. Sex, age, duration of fever, duration of diagnosis time, and laboratory data were explored for relationship with mortality.

## Results
As etiological factors triggered the HPS in our cases revealed in 15 ( 71%) patients. Infectious diseases in six cases ( 2 *Crimean-Congo hemorrhagic* fever (CCHF), 1 Ebstein Barr Virus (EBV), 1 Cytomegalovirus (CMV), 1 Influenza virüs, 1 Toxoplasmosis ), Rheumotologic disease in six cases ( 4 AOSD , 1 Rheumatoid arthritis , 1 SLE), Haemotological malignancies ( Diffuse Large B-cell lymphoma in one patient) and ulcerative colitis (one patient) had related to HPS. Six of the 21 patients received only high dose steroids as initial management and only two of them clinically resolved. The mortality rate of all-cause HPS was 23,8% (5 of 21 patients). Eritrocyte sedimentation rate and lactate dehydrogenase levels of our patients were remarkable.

## Conclusions
We would like to draw attention a more complex clinical condition that share many similar clinical and laboratory findings with septicemia and FUO, most often we come across in our daily practice as infectious disease specialists. Although ESR and LDH levels are not spesific biochemical marker to diagnose HPS , can be predictor factors for mortality as in our study.