FRIDAY, 14 JUNE 2019

15:30:00 – 17:00:00

Calming the cytokine storm in children and adults__**SP0153 CALMING THE STORM IN ADULT MAS/HLH IN RHEUMATIC DISORDERS**

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Background: Hemophagocytic lymphohistiocytosis (HLH) is a rare and often overlooked clinical manifestation of an aberrant hyper inflammatory immune response leading to a fatal cytokine storm(1). Diagnostic criteria according to The HLH Study Group of the Histiocyte Society include fever, splenomegaly, bicytopenia, hypertriglyceridemia and/or hypofibrinogenemia, hyperferritinemia, low/absent NK activity, increased soluble CD-25 levels and hemophagocytosis(2). Prompt recognition and differentiation from severe sepsis is essential to improve the outcome. HLH reflects a disbalanced immune system in response to infectious, malignancy, or autoinflammatory/autoimmune mediated triggers(1). The latter group of patients are regarded as having Macrophage Activation Syndrome (MAS-HLH)(3). In adults MAS-HLH comprises 12.5% of all HLH causing triggers(4). The most frequent immunological disorders associated with adult HLH-MAS are systemic lupus erythematosus and adult onset Still's disease, but every other immunological disorder can be involved(4, 5).

Treatment: Treatment of adults HLH patients requires swift recognition and an experienced team of specialists that are acquainted with the critical factors influencing the balance between co-morbidity, cytokine storm induced septic-like symptoms and toxicity of chemo-immunotherapy(6). In general, adult patients do not tolerate doses of etoposide that are given in (HLH-94 based) chemo-immunotherapy schedules that have been shown effective in children(7,8). Dose and frequency modifications of etoposide may avoid prolonged neutropenia, infectious complications or hepatic toxicity. The same precautions account for adults patients with MAS-HLH in which treatment differs from the HLH schedules. A step up approach depending on clinical features and severity is warranted(9, 10). High doses of corticosteroids are recognized as first-line treatment(9, 10). In patients with MAS-HLH and SLE steroids, cyclophosphamide or etoposide are given (10). Cyclosporine can be added in patients with insufficient immediate response(9). The cytokine blocking agent anakinra (anti-IL-1 β) given in high doses is emerging as an alternative or additional treatment of adult MAS-HLH(11, 12, 13). Another promising cytokine blocker is anti-IL-6 (tocilizumab). Etoposide at a reduced dose can be initiated in patients with severe non-responsive and active disease or CNS-involvement(10).

Conclusion: Awareness of the clinical emergency MAS-HLH in patients with autoinflammatory/autoimmune mediated disorders is essential for timely recognition and potential life-saving treatment. The management of MAS-HLH in adults requires a team of dedicated specialists with experience in the treatment of critically ill patients with immunological disorders and anticipation on its rapidly changing and deteriorating nature.

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Know your methods! Interactive discussion_____**SP0154 REASONS FOR QUALITATIVE RESEARCH FOR HPR**

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Qualitative research plays a pivotal role in clinical practice by providing insights from the patient's perspective. This becomes all the more relevant, as we move into the era of patient-centered care. In this presentation, we shed light on the different approaches to qualitative data collection and analysis, as well as systematic literature reviews of qualitative research. We will also elaborate on how qualitative research methods are used to better understand which outcomes are relevant to patients, self-management of symptoms, non-adherence to medication and non-pharmacological methods, the doctor-patient relationship, and to evaluate policy and interventions within the realm of RMDs. Finally, in an open panel, we will discuss how qualitative and quantitative methods triangulate, contributing to mixed methods approaches.

Disclosure of Interests: None declared

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SP0155 REASONS FOR QUANTITATIVE RESEARCH FOR HPR

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Health professionals in rheumatology are expected to provide care that is evidence-based. Evidence-based practice is the careful integration of best research evidence with clinical expertise, patient values and needs in the delivery of high-quality, cost-effective health care.¹ A sound understanding of quantitative methods is essential for correct evaluation of research and appropriate implementation into clinical practice.

Quantitative methods are used in a wide range of topics, from validating patient-reported outcome measures, sample size estimation, assessing associations between variables or predicting risk factors and evaluating the effectiveness of interventions. Furthermore, quantitative methods can be used to pool the results of several studies into a meta-analysis and thus provide better estimates of effectiveness than what can be achieved in individual studies.

This presentation will debate the case for quantitative research using examples of research conducted by health professionals in rheumatology. It is hoped that delegates will gain more confidence in using quantitative methods in their own research or evaluating evidence for clinical practice.

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