was suspected and markers revealed serum ferritin 30.000 ng/ml, fibrinogen 250 mg/dl, Triglycerides 350 mg/dl and soluble CD25 12.000U/ml. Bone marrow aspiration confirmed the diagnosis. Cytomegalovirus (CMV) IgM and PCR were significantly high. With regard to all the previous clinical and laboratory features, a CMV-induced secondary MAS in JSLE was confirmed. Patient was managed according to HLH-2004 treatment protocol (pulsed methylprednisolone 30mg/kg/day, cyclosporine A 6mg/kg/day, IV etoposide 150mg/m²) and IVIG. Ganciclovir was also added (5mg/kg IV q12h x7 days). Patient showed marked clinical improvement together with significant diminution of the MAS laboratory markers to normal. Two weeks post discharge on oral dexamethasone and cyclosporine A, the patient showed a severe clinical and laboratory MAS relapse (serum ferritin 50.000 ng/ml). The previous treatment protocol was restarted together with adding Rituximab 375mg/m² x 3 doses two weeks apart. Patient showed marked clinical and laboratory improvement and was maintained on full dose oral prednisone and oral Cyclosporine A. Tight gradual withdrawal of oral steroids was done with close clinical and laboratory follow up.

Conclusion: JSLE is a major diagnostic conundrum in pediatrics owing to its extremely variable clinical manifestations which can mimic many common pediatric conditions (e.g. malignant disease, other auto-inflammatory conditions, malignancy and any specific organ associated disease). Compared to adults JSLE has a more severe disease presentation. MAS is a life threatening condition that requires high index of suspicion and prompt management. In our case, controlling JSLE activity using Rituximab was significantly beneficial in arresting HLH evolution.

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AB0922

UPDATES ON THE EVIDENCE BASED INTERDISCIPLINARY GUIDELINES FOR HENOCH-SCHÖNLEIN PURPURA

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Background: Henoch-Schönlein purpura (HSP) is the most common childhood vasculitis, it is characterized by inflammation of small vessels leading to non-thrombocytopenic purpura, arthritis/arthralgia, GI hemorrhage and glomerulonephritis¹. There is a very poor evidence base to guide the management of HSP, particularly for those with the severe forms of HSP nephritis. Up till now there are no EULAR or ACR recommendations for management of HSP.

Objectives: To set an updated interdisciplinary recommendations for the management of HSP

Methods: Interdisciplinary guidelines were developed with representative from PRINTO Egypt for pediatric rheumatology as well as patients' group. A systemic literature analysis in MEDLINE was performed, evidence and recommendations were graded, an algorithm for treatment and final statements were discussed in a consensus meeting (Nominal Group Technique). A preliminary draft was fine tuned and discussed thereafter by all participants (Delphi Procedure).

Results: Consensus was reached on recommendations, including a standardized treatment strategy according to the HSP severity state in the individual patient. In this updated interdisciplinary guideline for HSP; treatment was tailored to the patient's pathology and its severity (figure 1).

Criteria for admission were identified as: severe arthritis or arthralgia limiting ability to weight bearing and mobilization, severe abdominal pain, GI hemorrhage, evidence of nephritis/nephritic syndrome or renal impairment, evidence of neurological symptoms. Criteria for early referral to pediatric nephrologist were also identified including: hypertension, abnormal renal function, macroscopic hematuria for 5-days, nephrotic syndrome, acute nephritic syndrome, as well as persistent proteinuria.

Conclusion: A total of seven evidence based interdisciplinary recommendations for management of HSP have been formulated, that provide an up-to-date guidance of HSP management.

Recommendations for management of HSP

- **Joint symptoms:** bed rest and NSAIDs (avoid NSAIDs if renal disease present). Steroids have also been used in severe cases
- **Abdominal pain** usually settles within 72hrs and simple analgesia is adequate as (Ibuprofen at 5-10mg/Kg/dose tds +/-Paracetamol at 10-15mg/Kg/dose qds)
- **If abdominal pain is severe**, consider using steroids (2mg/Kg/day).
- **If abdominal pain is acute, severe and/or associated with blood in the stool**, then consider Intussusception seek **surgical opinion**. seek surgical opinion also in case of painful scrotal swelling
- **Skin manifestations** are generally benign and self-limited. Severe cases have been treated with Colchicine or dapsone.
- **Treatment of HSP nephritis in children:**
 - Children with HSP nephritis and persistent proteinuria 0.5-1 g/d are treated with ACE-I or ARBs.
 - Children with HSP nephritis and persistent proteinuria > 1 g/d after a trial of ACE-I or ARBs, and GFR > 50 ml/min per 1.73m² be treated with a 6-month course of corticosteroid therapy (either pulse followed by oral steroid 0.5 mg/kg prednisone on alternate days for 6 months or oral prednisone 0.8-1 mg/kg/d for 2 months and then reduced by 0.2 mg/kg/d per month for the next 4 months)
- **Treatment of crescentic HSP nephritis in children** (crescents in more than 50% of glomeruli in the renal biopsy with rapidly progressive renal deterioration):
 - use of pulse steroids and cyclophosphamide and azathioprine
 - Cyclosporine A is effective therapy in children resisted to first line therapy.
 - mycophenolate mofetil could be used in steroid-resistant cases
 - plasmapheresis and IVIG could be used in resisted cases
 - Rituximab could be used in adults with refractory or relapsing HSP but up till now its safety not established in children

Figure 1: recommendations for management of HSP

Figure 1

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AB0923

EFFECTS OF EXON 10 MUTATIONS VS NON-EXON 10 MUTATIONS ON FMF PHENOTYPE AND RESPONSE TO TREATMENT

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Background: Familial Mediterranean fever (FMF) is the most common monogenic periodic fever syndrome. The MEFV gene mutation encoding the pyrin protein results in an uncontrolled increase in interleukin-1. Today, more than 333 MEFV mutations have been identified; however, exon 10 mutations are still seem to be best correlated with clinical findings.

Objectives: In this study, we aim to investigate the role of exon 10 mutations vs non-exon 10 mutations on clinical features and response to treatment in patients with FMF.

Methods: Data charts of children (n=935) with FMF from Dokuz Eylul University childrens' hospital and Dr.B.Uz childrens' hospital were reviewed. Patients were divided into two groups with regard to having