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Muserref Kasap-Cuceoglu and Melis Pehlivanturk-Kizilkan contributed equally.
Patient Consent Received
No

Disclosure of Interest
None declared

Table 1 (abstract P310). Measurements of capileroscopy findings according to patient groups

	AN+RF+ n=4	AN+RF- n=13	BN n= 2	Primer raynoud kontrol n=6	p
Microangiopathy score	0.84 (IQR 0.98)	0.92 (IQR 0.96)	0.71	0.65 (IQR0.23)	0.62
- **Capillary loss score**	0.22 (IQR 0.62)	0.06 (IQR 0.41)	0.15	0.0 (IQR 0.26)	0.58
- **Capillary ramification score**	0.18 (IQR 0.25)	0.13 (IQR 0.25)	0.09	0.09 (IQR 0.18)	0.88
- **Disorganized capillaries score**	0.38 (IQR 0.48)	0.56 (IQR 0.68)	0.46	0.43 (0.32)	0.98
Enlarged capillaries score (20-50)	0.56 (IQR 0.78)	0.31 (IQR 0.72)	0.73	0.58 (IQR 0.42)	0.60
Giant capillaries score (>50)	0.06 (IQR 0.22)	0.00 (IQR 0.03)	0.18	0.03 (0.14)	0.14
Microhaemorrhage score	0.00	0.00	0.00	0.01	0.67

P311

Maculopapular rash and fever: a diagnostic challenge to the paediatrician in the COVID-19 pandemic

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Introduction: Several clinical conditions can manifest with fever and a maculopapular rash in paediatric age. Although some presentations are benign, others may be medical emergencies, which demand a prompt diagnosis and treatment. Some of the more common causes of fever and maculopapular rash include infectious diseases (Sars-CoV-2, Parvovirus B19; Coxsackie; Epstein-Barr virus infection, Mycoplasma Pneumoniae, etc), hypersensitivity reactions, Autoinflammatory syndromes, vasculitis, Kawasaki disease (KD), autoimmune diseases.
Objectives: In the COVID-19 pandemic era these symptoms need a well-organized hospital strategy to rapidly exclude Sars-CoV-2 infection and to distinguish severe and rapidly developing patients.
Methods: We evaluated the medical records of children admitted to a paediatric tertiary centre in the years 2020-202, excluding children with suspected or documented COVID-19 infection.
Results: We retrospectively identified 21 patients (13M; 9F), age: 0.7-12 years, admitted with the diagnosis of fever and rash and with a definite diagnosis. 10 children had a documented infection (2 Mycoplasma; 2 Parvovirus; 5 EBV; 1 Adenovirus); 3 patients had a KD; 4 had an autoimmune disease; 3 had an Autoinflammatory syndrome, 1 a vasculitis; 1 had a Macrophage Activation Syndrome (MAS). Distribution of the rash, a persistent/vanishing rash, the associated lymphadenopathy did not contribute to the differential diagnosis. Haemoglobin levels were significantly lower in KD (8.3-11.2). CRP was significantly higher in KD (3.23-34) vs autoimmune diseases and Autoinflammatory syndromes. The other laboratory parameters did not contribute to the differential diagnosis, otherwise reached by specific IgM and PCR. In children with clinical signs of suspicion of Autoinflammatory syndromes, the genetic approach permitted to reach the treat-to-target.
Conclusion: The numerous viral skin diseases that affect children present a diagnostic challenge to the clinicianIn some situations, viral rash may be difficult to clinically differentiate from nonviral diseases; extensive laboratory evaluation isolates the virus. Otherwise, autoimmune diseases must be excluded and, in this suspicion, the alert must be high to promptly diagnose a MAS. A most severe presentation can hide the first attack of an Autoinflammatory syndrome: hence, the genetic study of these condition is a milestone in the differential diagnosis and avoid a diagnostic delay.
Patient Consent Received
Yes

Disclosure of Interest
None declared

P312

Primary Sjögren's syndrome: description of a monocentric pediatric cohort with focus on the interferon pathway activation

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Introduction: Primary Sjögren's Syndrome (pSS) is a chronic autoimmune disease that can involves both exocrine glandular and extra-glandular systems. This condition is rare in children in whom diagnosis may be less straightforward due to the absence of well-established diagnostic criteria. Compelling evidence suggests a significant role for type I interferon (IFN) in the pathogenesis of SS [1].
Objectives: 1) To describe clinical and diagnostic features of a cohort of children with pSS;
2) To examine IFN pathway activation in pSS.
Methods: We included retrospectively 16 pSS children with disease onset until age 16 years that referred to our Unit. For each patient we collected: clinical data including the disease activity indexes (*ESSDAI* - EULAR Sjögren's Syndrome Disease Activity Index, *ESSPRI* - EULAR Sjögren's Syndrome Patient Reported Index) [2], laboratory tests and US examination of salivary glands. In addition, expression of 6 type I IFN-related genes (IFI27, IFI44, IFIT1, ISG15, RSAD2, SIGLEC1) was tested in blood samples by standard Real-Time PCR techniques.
Results: Almost all patients (15/16) were females. The mean age at onset was 10.08 years (range 5.1-15.7); the mean time interval between onset of symptoms and diagnosis was 2.03 ± 1.6 years; the mean follow up time was 3.36 years. *Table 1* shows the clinical features at disease onset. Rheumatoid factor (RF), antinuclear antibodies (ANA), anti-SSA, anti-SSB were positive in the majority of cases (68.7%, 68.7%, 75%, 62.5% respectively); hypergammaglobulinemia and high