1.0050, 95% CI 1.0032–1.0070, p<0.0001) was associated with significantly increased risk of subsequent flare. In contrast, C3 and C4 levels were not statistically significantly associated with flare risk (p>0.05). In the multivariable model, both anti-dsDNA and ESR remained independently associated with risk of subsequent flare in the cCR (anti-dsDNA: HR 1.0005, 95% CI 1.0001–1.0008, *p*=0.0078; ESR: HR 1.0041, 95% CI 1.0020–1.0063, *p*=0.0002) and cCR-0 (anti-dsDNA: HR 1.0004, 95% CI 1.0000–1.0008, *p*=0.0188; ESR: HR 1.0045, 95% CI 1.0023–1.0067, p<0.0001) models. In the univariable model of patients in cLLDAS, ESR was also significantly associated with subsequent risk of flare (HR 1.0047, 95% CI 1.0029–1.0065, p<0.0001).
Conclusion: Despite attainment of T2T goals, elevated anti-dsDNA and ESR levels independently predicted flare risk in cSLE. These findings support continued biomarker monitoring even in clinically stable patients to guide long-term disease management. Further studies with larger international cohorts are needed to confirm these associations.
Disclosure
None declared

PT098

Predicting the recurrence risk of immunoglobulin a vasculitis due to clinical and laboratory features in children

D. Gezgin Yildirim[1], E. N. Sunar Yayla[2], C. G. Yildiz[1], N. Karacayir[1], B. Kucukali[1], N. Belder[1], M. Kutlar Tanıdır[1], P. Esmeray Senol[3], K. Ahmadova[1], B. Acun Sayili[1], O. Ozenli Yagci[1], S. A. Bakkaloglu[1]
[1]Pediatric Rheumatology, Gazi University, [2]Pediatric Rheumatology, Etlik City Hospital, Ankara, [3]Pediatric Rheumatology, Mersin City Hospital, Mersin, Türkiye
Correspondence: D. Gezgin Yildirim
Pediatric Rheumatology, **23(2):** PT098

Introduction: Immunoglobulin A vasculitis (IgAV) is the most common vasculitis in childhood, characterized by affecting the small vessels of the skin, gastrointestinal tract, and kidneys. Although IgAV is a benign and self-limited disorder, relapses may occur during follow-up.
Objectives: The aim of this study is to evaluate the frequency and predictors of IgAV relapses according to clinical and laboratory features in children.
Methods: We retrospectively reviewed the medical records of 546 pediatric IgAV patients between 2005 and 2025. IgAV-related recurrence was accepted as a recurrence of skin lesions and/or any systemic organ involvement after an asymptomatic period of at least 6 months. Patients were divided into two subgroups: Group 1, which consisted of IgAV patients who had at least one recurrence related to IgAV, and Group 2, which consisted of IgAV patients who had no recurrence.
Results: There were 546 IgAV patients with a male dominance. A total of 40 (7.3%) patients had a relapse related to IgAV after an asymptomatic period of at least 6 months (Group 1). Patients' characteristics and laboratory features were presented in Table 1. Male predominance, diarrhoea, bloody stool, bloody vomiting, gastrointestinal involvement, testicular involvement, WBC, creatinine, and pediatric vasculitis assessment scale (PVAS) at first visit were significantly higher, while mean age at diagnosis of IgAV was significantly lower in Group 1 than Group 2 (*p*=0.033, *p*=0.015, *p*=0.013, *p*=0.045, *p*=0.047, *p*=0.003, *p*=0.012, *p*=0.012, *p*=0.000, and *p*=0.015, respectively). The most recurrent clinical finding was purpuric skin eruption (65%) (Table 2). Recurrence free survival of Group 1 was illustrated in Fig1. Logistic regression analysis for the prediction of recurrence of IgAV was shown in Table 3. Male sex, diarrhoea, testicular involvement, Hb value, and PVAS were found as independent risk factors for the prediction of IgAV recurrence in the long-term follow-up (*p*=0.033, *p*=0.015, *p*=0.003, *p*=0.025, and *p*=0.005, respectively).
Conclusion: In this study, we have shown that male sex, diarrhoea, testicular involvement, higher Hb value, and higher PVAS at first visit were found as independent risk factors for the prediction of IgAV recurrence in the long-term follow-up. Close follow-up is important especially in the presence of male gender, testicular involvement in boys, presence of diarrhoea and high PVAS at first visit, considering that the disease may recur in the long term.
Disclosure
None declared

PT099

Coronary evolution and inflammatory response in ivig-resistant kawasaki disease treated with anakinra: a longitudinal study from a French tertiary center

C. Matucci Cerinic[1], M. Gattorno[1], I. Kone-Paut[2], P. Dusser-Benesty[2]
[1]IRCCS Istituto Giannina Gaslini, Rheumatology and autoinflammatory diseases, Genoa, Italy, [2]Department of Paediatric Rheumatology and CEREMAIA, Bicêtre University Hospital, APHP, university of Paris Saclay, Paris, France
Correspondence: C. Matucci Cerinic
Pediatric Rheumatology, **23(2):** PT099

Introduction: Treatment of intravenous immunoglobulin (IVIG)-resistant Kawasaki disease (KD) remains non-standardized. IL-1 blockade with anakinra (ANK) has shown promising results in case series and a phase II open-label trial.
Objectives: To describe the longitudinal clinical and coronary evolution of IVIG-resistant KD patients treated with anakinra, from treatment initiation to long-term follow-up.
Methods: We retrospectively analyzed IVIG-resistant KD patients treated with ANK at a French tertiary referral center and enrolled in the JIR cohort. Clinical, laboratory, treatment, and echocardiographic data were collected at predefined timepoints: baseline (T0), day 7, day 14, day 30, day 365, and last follow-up. Coronary artery aneurysms (CAA) were defined according to American Heart Association (AHA) criteria. Z-score normalization was defined as <2.
Results: Thirty patients were included (56.7% male, mean age 2.17 ± 2.31 years). Mean follow-up duration was 16.95 ± 17.26 months. All received aspirin and IVIG (63.3% one dose, 30% two doses, 6.7% three doses). Before ANK, 40% received corticosteroids and 6.6% infliximab. At ANK initiation, all had persistent inflammation, and 11 (36.6%) had CAA (2 at diagnosis). Two had myocarditis. Three additional patients developed new CAA under ANK, all affecting the proximal right coronary artery (RCA). CRP normalized in 77% of patients at day 14, and in 82.6% (19/23) during follow-up (median time 33 days, IQR 33.5). The median duration of ANK therapy was 14 days (range 1–324); 26.6% received corticosteroids after ANK. Seven patients were switched to another biologic.
Among patients with CAA, the proximal RCA showed the highest initial Z-score (median 3.48 [IQR 2.6–7.6]). Despite a frequent transient increase in coronary Z-scores around day 7 to 14 after ANK initiation, the median time to normalization across all coronary segments was 5 days (IQR 0–181; n=13). At last follow-up, only 3 patients had persistent CAA.
Conclusion: In this French tertiary cohort, anakinra was associated with favorable 1-year outcomes regarding systemic inflammation and coronary artery evolution in IVIG-resistant KD. Prospective studies are needed to confirm these findings.
Disclosure
None declared

PT100

Elevated serum brain injury markers correlate with disease features and interferons in children with systemic lupus erythematosus

G. Ramanathan[1], O. Mwizerwa[1], J. Ledochowski[1], T. El Tal[2], L. Ng[1], A. Jeyanathan[1], A. Davis[1], L. Hiraki[1], D. Levy[1], J. Wither[3], Z. Touma[3], A. Yeh[1], A. Knight[1]
[1]Hospital for Sick Children, Toronto, [2]Children's Hospital of Eastern Ontario, Ottawa, [3]Schroeder Arthritis Institute, Krembil Research Institute, Toronto Western Hospital, Toronto, Canada
Correspondence: A. Knight
Pediatric Rheumatology, **23(2):** PT100

Introduction: Childhood-onset systemic lupus erythematosus (cSLE) involves interferon (IFN)-mediated inflammation emerging during the critical period of adolescent brain development. Neuropsychiatric lupus (NPSLE) manifests as syndromes like cognitive dysfunction, seizures, and psychiatric Clinicians face challenges diagnosing/treating brain inflammation in cSLE, due to suboptimal diagnostic tools. Neuronal/glial structural proteins may be useful biomarkers of brain injury in cSLE.

Pediatric Rheumatology (2025) 23:93
https://doi.org/10.1186/s12969-025-01138-8

Pediatric Rheumatology

MEETING ABSTRACTS

Open Access

Proceedings of the 32nd European Paediatric Rheumatology Congress

Helsinki, Finland. 17-20 September 2025
Published online: 16 September 2025

O01

Reconstitution of the B cell compartment following anti-CD19 CAR-T cell therapy in refractory patients with Juvenile Dermatomyositis (JDM)

E. Marasco[1], R. Nicolai[1], A. Ariolli[1], M. Becilli[2], C. Bracaglia[1], P. Merli[3], M. I. Petrone[1], F. Del Bufalo[3], M. Algeri[3], M. G. Cefalo[3], F. Locatelli[3], F. De Benedetti[1]
[1]Rheumatology Unit, [2]Department of Hematology/Oncology, Cell and Gene Therapy, [3]Department of Hematology/Oncology, Cell and Gene Therapy, Ospedale Pediatrico Bambino Gesù, Rome, Italy
Correspondence: E. Marasco
Pediatric Rheumatology, **23(2):** O01

Introduction: CD19 CAR T-cell therapy is an innovative strategy to induce profound B cell depletion in patients with refractory idiopathic inflammatory myopathies (IIM). However, its effects on B cell phenotypes in pediatric patients remain largely unknown.
Objectives: To characterize the reconstitution profile of B cell subsets in three patients with juvenile dermatomyositis (JDM) treated with anti-CD19 CAR T-cell therapy.
Methods: Three patients with refractory JDM received a single infusion of autologous, second-generation anti-CD19 CAR T cells (1×10^6 CAR T cells/kg; lentiviral vector), manufactured on the CliniMACS Prodigy system. Lymphodepletion was achieved using cyclophosphamide (1000 mg/m^2 over 2 days) and fludarabine (90 mg/m^2 over 3 days). B cell immunophenotyping was performed according to standard operating procedures (SOPs) before and after CAR T-cell infusion. A comparator group of five JDM patients treated with rituximab (RTX) was included.
Results: B cell reconstitution occurred at a median of 8 weeks (IQR: 8.0-12.0) post-CAR T therapy, with an immature transitional B cell phenotype. B cell subsets distribution was assessed at a median of 27.3 weeks (IQR: 22.5–40.2) after treatment, when all B cell subsets were present for each patient, and compared to pre-treatment profiles. In the RTX group, B cell phenotyping was performed at a median of 47 weeks (IQR: 33.4–50.7) post-treatment. The proportion of CD19$^+$ B cells among lymphocytes was similar before and after CAR T-cell treatment (pre: 32% [IQR: 23–40], post: 21% [IQR: 20–24]). B cell subset analysis showed comparable levels of transitional, naïve and plasmablast populations pre- and post-treatment. However, there was a notable reduction in total memory B cells (MBCs) post-treatment (pre: 6.4% [IQR: 6.3–9.1], post: 3.9% [IQR: 3.1–4.0]), particularly in the switched MBC subset. Interestingly, JDM patients treated with RTX showed higher frequencies of switched MBCs during B cell reconstitution compared to those treated with CAR T cells: CAR T treated-patients 16.0 % [IQR: 8.5-18.0], RTX group 52.0% [IQR: 28.0, 60.0].
Conclusion: These data show that following CAR T cell treatment, the memory B cell compartment presents a lower frequency of switched MBCs when compared to pre-treatment levels and to patients treated with RTX, suggesting a specific effect of CAR T cell treatment in regulating the memory B cell compartment in JDM patients.
Disclosure
None declared

O02

Anti-CD19 Car-T cell therapy in refractory juvenile dermatomyositis (JDM): a single-center case series

R. Nicolai[1], M. Becilli[2], E. Marasco[1], P. Merli[2], C. Bracaglia[1], F. Del Bufalo[2], M. I. Petrone[1], M. G. Cefalo[2], M. Algeri[2], F. De Benedetti[1], F. Locatelli[2,3]
[1]Division of Rheumatology, [2]Department of Hematology/Oncology, Cell and Gene Therapy, IRCCS Ospedale Pediatrico Bambino Gesù, [3]Catholic University of the Sacred Heart, Rome, Italy
Correspondence: R. Nicolai
Pediatric Rheumatology, **23(2):** O02

Introduction: CD19 CAR T-cell therapy represents an innovative approach emerging as a promising treatment option in patients with refractory idiopathic inflammatory myopathies (IIM), but the experience in children so far is limited.
Objectives: To report on the safety and efficacy of anti-CD19 CAR T-cell therapy in 3 JDM patients.
Methods: All 3 patients received a single infusion of fresh, autologous second-generation anti-CD19 CAR T product (lentiviral vector) manufactured on the Prodigy device ($1x10^6$ CAR T cells/kg), after lymphodepletion with cyclophosphamide (1000 mg/m^2 over 2 days) and fludarabine (90 mg/m^2 over 3 days).
Results: Maximum follow-up after CAR T-cell therapy was 20 months. Patients 1 and 2 had a history of severe chronically active disease with extensive calcinosis and ulcerations. Patient 3 presented interstitial lung disease (ILD) and persistently active muscle and skin disease with ulcerations. Short-term effects of CAR T-cell therapy on Patient 1 have been previously reported (Nicolai A&R 2024). All patients were refractory to multiple immunosuppressive treatment lines and presented significant side effects from chronic glucocorticoid treatment. After infusion, CAR T cells rapidly expanded, reaching peak concentrations after a median of 7 days (range 7-9). At peak, the median number of circulating CAR T cells was 32.69 per microliter (range 31.04-535.02). CD19+ B cells were undetectable in peripheral blood after a mean of

© The Author(s) 2025. **Open Access** This article is licensed under a Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License, which permits any non-commercial use, sharing, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if you modified the licensed material. You do not have permission under this licence to share adapted material derived from this article or parts of it. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit http://creativecommons.org/licenses/by-nc-nd/4.0/.