Objectives: We compared serum levels of brain injury markers and IFNs between children with cSLE and controls and investigated the relationship of markers to cSLE disease features and serum-IFN levels.
Methods: We utilized prospectively-collected cross-sectional data from cSLE participants (ages 12-17) recruited from the Lupus Clinic at a Canadian tertiary children's hospital from January 2020–December 2023 and age-, sex-matched healthy controls. Serum brain injury markers (serum neurofilament light (sNFL), glial fibrillary acidic protein (GFAP), Tau) were quantified using Simoa Human Neurology 4–Plex B assay; IFN-α and IFN-γ were also quantified by their respective Simoa assays (Quanterix, Billerca, MA, USA). Disease features included disease activity (SLEDAI-2K), damage (SLICC damage index, SDI >0), glucocorticoid (GC) dose at study visit, and cumulative GC exposure (prednisone-equivalent). Wilcoxon rank sum test compared markers and IFNs; Spearman correlation tested associations.
Results: 56 cSLE participants (mean age=15.1±1.8 years, 86% female) and 43 controls (mean age=15.1±1.7 years, 81% female) were included. For cSLE, median disease duration was 22.6 months (IQR 12.5-43.9), median SLEDAI-2K was 2.5 (IQR, 2-5.5), 13% had disease damage, 41% were using glucocorticoids at study visit, and median cumulative GC exposure was 1.9 grams (IQR 0.6-6.9). One patient had a NPSLE diagnosis. GFAP (114.0 vs 74.3 pg/mL) and Tau (3.57 vs 2.58 pg/mL) levels were significantly higher in cSLE compared to controls ($p<0.05$), as were serum IFN-α (0.278 vs 0.018 pg/mL) and IFN-γ (0.100 vs 0.068 pg/mL). All brain injury markers had significant positive correlations with SLEDAI-2K and GC dose; sNFL and Tau associated with disease damage. Higher levels of sNFL and GFAP correlated with IFN-α, while GFAP also associated with IFN-γ. No correlations were found between Tau and IFNs.
Conclusion: Serum brain injury markers and IFNs were elevated in cSLE, with brain injury markers correlating with disease features, IFN-α, and IFN-γ. This suggests a link between IFN-mediated inflammation and neuronal/glial injury, and potential utility of sNFL, GFAP and Tau as diagnostic and monitoring biomarkers in cSLE. Future studies will explore relationships between brain injury markers and IFNs in larger cSLE cohorts over time.
Disclosure
None declared

P003

Comparative evaluation of chatgpt and llama for reliability, quality and accuracy in familial mediterranean fever

A. Uzun Bektaş, S. E. Ünsal, B. Bora
Pediatric Rheumatology, Dokuz Eylul University, İzmir, Türkiye
Correspondence: A. Uzun Bektaş
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Introduction: In the era of advanced technology, large language models (LLMs) such as OpenAI's ChatGPT and Meta's LLaMA offer rapid access to medical information for both patients and healthcare professionals. Unlike traditional search engines, LLMs empower clinicians to retrieve clinical data rapidly, facilitating decision-making processes. Despite their broad potential and widespread accessibility, studies have highlighted their accuracy and reliability concerns. The utilization of LLMs in rheumatology, particularly in the context of Familial Mediterranean Fever (FMF) remains underexplored.
Objectives: This study aims to evaluate and compare the accuracy, quality, and reliability of responses generated by ChatGPT-4o and LLaMA-3.1 405B models to medical questions related to FMF.
Methods: A total of 31 questions related to FMF were prepared from a clinician's perspective, based on the SHARE and EULAR guidelines. Each question was asked to ChatGPT-4o and LLaMa-3.1 405B only once in separate chat sessions. The randomized responses were independently and blindly evaluated by two pediatric rheumatologists, each with over 20 years of experience, in terms of reliability, quality, and accuracy. Reliability was assessed using the Modified DISCERN score, and quality using the Global Quality Scale (1). The accuracy of the responses was evaluated according to the evidence-based guideline on the genetic diagnosis of FMF published by SHARE and the guideline on FMF management published by EULAR (2,3). The Shapiro-Wilk test, the Intraclass Correlation Coefficient, and the Wilcoxon Signed Rank test were used to analyze the score distribution, inter-rater agreement, and the models' comparative performance, respectively. Readability was assessed using the Flesch Reading Ease, Flesch-Kincaid Grade Level, Gunning Fog Index, Coleman-Liau Index, and SMOG Index (4).
Results: Both models demonstrated moderate reliability according to the modified DISCERN score and high response quality on the Global Quality Score. Although both models aligned with guidelines in terms of accuracy on average, LLaMA exhibited notable inconsistencies. Despite achieving a high average score, some of its responses either directly contradicted guideline-based protocols or contained partially aligned guidelines but erroneous information. In contrast to LLaMA, ChatGPT's responses adhered to guidelines without contradictions; however, certain responses demonstrated informational gaps. Statistical analyses showed that ChatGPT's responses were superior to LLaMA's, with significantly higher scores in accuracy, quality, and reliability. Readability assessments showed that understanding the answers from both LLMs required at least a college-level education.
Conclusion: It is crucial for the clinicians to consider that LLMs may provide incomplete or misleading information.
Disclosure
None declared

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P004

Is it possible to predict the need for tonsillectomy in children with PFAPA?

B. Öksel[1], Z. Özaslan[1], A. Şen[2], S. Atamyıldız Uçar[3], M. Çakan[4], S. N. Tığrak[5], B. Menentoğlu[6], F. Kaya[7], D. Alkan[2], G. O. Yener[8], H. Adıgüzel Dündar[9], Ö. Altuğ Gücenmez[9], F. Demir[10], H. Ilgaz Tüzen[11], N. Belder[12], A. Tanatar[13], G. Vatansever[14], H. Kazanasmaz[15], K. Ulu[16], A. U. Bektaş[17], F. G. Demirkan[18], B. Bozkaya Yücel[19], T. Kasap[20], B. K. Demir[11], D. G. Yıldırım[12], S. S. Kilic[21], M. Kalyoncu[15], S. Yüksel[22], B. Bora[17], K. Öztürk[7], N. A. Ayaz[6], S. Türkuçar[5], S. Balcı[23], N. Şahin[1], S. Özdel[2], B. Sözeri[3], H. E. Sönmez[1]
[1]Department of Pediatric Rheumatology, Kocaeli University, Kocaeli, [2]Department of Pediatric Rheumatology, Ankara Etlik City Hospital, Ankara, [3]Department of Pediatric Rheumatology, Health Sciences University, Umraniye Training and Research Hospital, [4]Department of Pediatric Rheumatology, Zeynep Kamil Women and Children's Diseases Training and Research Hospital, İstanbul, [5]Department of Pediatric Rheumatology, Pamukkale University, Denizli, [6]Department of Pediatric Rheumatology, İstanbul University, [7]Department of Pediatric Rheumatology, İstanbul Medeniyet University, İstanbul, [8]Department of Pediatric Rheumatology, Eskişehir City Hospital, Eskişehir, [9]Department of Pediatric Rheumatology, Dr. Behcet Uz Pediatric Diseases and Surgery Training and Research Hospital, İzmir, [10]Department of Pediatric Rheumatology, Acıbadem Ataşehir Hospital, İstanbul, [11]Department of Pediatric Rheumatology and Nephrology, İzmir Katip Çelebi University, İzmir, [12]Department of Pediatric Rheumatology, Gazi University, Ankara, [13]Department of Pediatric Rheumatology, Gaziantep City Hospital, Gaziantep, [14]Department of Pediatric Rheumatology, Bursa Uludag University Faculty of Medicine, Bursa, [15]Department of Pediatric Rheumatology, Karadeniz Technical University, Trabzon, [16]Department of Pediatric Rheumatology, Sancaktepe Prof. Dr Ilhan Varank Training and Research Hospital, İstanbul, [17]Department of Pediatric Rheumatology, Dokuz Eylul University, İzmir,

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Pediatric Rheumatology

MEETING ABSTRACTS

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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
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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

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