Gözde Kübra Yardımcı: None declared, reyhan bilici salman: None declared, Levent Kılıç: None declared, mehmet akif ozturk: None declared, Berna Goker: None declared, seminur haznedaroglu: None declared, Umut Kalyoncu Grant/research support from: MSD, Roche, UCB, Novartis and Pfizer, Consultant for: MSD, Abbvie, Roche, UCB, Novartis, Pfizer and Abdi Ibrahim, Speakers bureau: MSD, Abbvie, Roche, UCB, Novartis, Pfizer and Abdi Ibrahim, abdurrahman tufan: None declared

DOI: 10.1136/annrheumdis-2019-eular.4766

THU0563

COMPARISON BETWEEN PATIENTS WITH IDIOPATHIC PULMONARY FIBROSIS AND INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES: A PROSPECTIVE COHORT

Gianluca Sambataro^{1,2}, Domenico Sambataro^{2,3}, Sebastiano Emanuele Torrisi¹, Vancheri Ada¹, Mauro Pavone¹, Francesca Pignataro⁴, Nicoletta Del Papa⁴, Stefano Palmucci⁵, Carlo Vanchieri¹, Lorenzo Malatino³, Michele Colaci¹.
¹Teaching Hospital "Policlinico – Vittorio Emanuele", Clinical and Experimental Medicine, Catania, Italy; ²Artroreuma srl, Outpatient of Rheumatology accredited with National Health System, Mascalucia (CT), Italy; ³Hospital "Cannizzaro", Clinical and Experimental Medicine, Catania, Italy; ⁴Day Hospital of Rheumatology, Hospital "G. Pini – CTO", Department of Rheumatology, Milan, Italy; ⁵Teaching Hospital "Policlinico – Vittorio Emanuele", Department of Medical Surgical Sciences and Advanced Technologies – Radiology I Unit, Catania, Italy

Background: The term "Interstitial Pneumonia with Autoimmune Features (IPAF)" is used to describe patients with Interstitial Lung Disease in combination to clinical, serological and/or pulmonary features that are suggestive, but insufficient to satisfy classification criteria of a specific Connective Tissue Disease (CTD). Some retrospective studies available in literature described patient cohorts with IPAF heterogeneous in terms of clinical, serological, and radiographic manifestations

Objectives: To prospectively recruit a cohort of consecutive ILD patients classified as IPAF or as affected by Idiopathic Pulmonary Fibrosis (IPF); to describe their clinical, serological, and radiological features by a multi-disciplinary team composed by Pulmonologists, Radiologists, and Rheumatologists, comparing IPAF and IPF patients.

Methods: In the last 2 years, they were enrolled 45 patients with IPAF and 143 with IPF among a total of 506 patients with Interstitial Lung Disease (ILD). All patients were evaluated clinically by both rheumatologists and pulmonologists, also by means of chest high resolution computed tomography (hrCT), pulmonary function tests (PFT), and nailfold videocapillaroscopy.

Results: In IPAF cohort the most common characteristics from the clinical, serological and morphological domain were Raynaud's phenomenon (RP) (31.1%) antinuclear antibodies positivity with titre $\geq 1/320$ or any titre for centromeric or nucleolar pattern (17.7%) and nonspecific interstitial pneumonia (68.8%) respectively. The majority of patient (88.9%) had the minimum of 2 criteria at the recruitment. Female gender was more common in IPAF compared to IPF (62.2% vs 23%, $p < 0.0001$) and the mean age of onset was lower in patient with IPAF (66 vs 73, $p < 0.0001$). NVC resulted positive in 2 patients, both without RP. Patients with IPAF showed better performances in PFT than patients with IPF than those with IPF (mean forced vital capacity 83% vs 74%, $p = 0.01$; mean diffusing lung capacity for carbon monoxide 64% vs. 52%, $p = 0.01$). Patients with IPAF less commonly than patients with IPF needed oxygen therapy (44.4% vs 77.6%, $p < 0.0001$) and more commonly showed at hrCT NSIP (68.8% vs 6.9%, $p < 0.0001$) than UIP pattern (0% vs 86%, $p < 0.0001$).

Conclusion: IPAF criteria recruit a rare cohort of patients with a disease probably less severe than IPF, even though clinically relevant. Further prospective studies may better define the long-term prognosis of ILD in IPAF. The follow-up of these patients by a multi-disciplinary team may be useful in order to early recognize and treat the new cases of CTDs. Some IPAF criteria probably are useless due to their high specificity for definite CTD and the classification need a revision in the light of the prospective experiences. Nevertheless, IPAF classification can recruit incomplete form or early onset of CTD that can allow a timely treatment in patients without a structured damage.

REFERENCES:

- [1] Sambataro G, Sambataro D, Torrisi SE, Vancheri A, Pavone M, Rosso R, Schisano M, Crimi C, Pignataro F, Fischer A, Del Papa N, Vancheri C. State of the art in interstitial pneumonia with autoimmune features: a systematic review on retrospective studies and suggestions for further advances. Eur Respir Rev 2018;27(148)

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.6735

THU0564

COULD A PROBABILISTIC REASONING AI ACCELERATE RARE DISEASE DIAGNOSIS? EVALUATING THE POTENTIAL IMPACT OF A DIAGNOSTIC DECISION SUPPORT SYSTEM IN A RETROSPECTIVE STUDY

Simon Ronicke^{1,2}, Martin C. Hirsch², Ewelina Türk², Katharina Larionov¹, Daphne Tientcheu¹, Annette D. Wagner¹. ¹Outpatient clinic for rare inflammatory systemic diseases, Department of Nephrology, Hannover Medical School, Hannover, Germany; ²Ada Health GmbH, Berlin, Germany

Background: The diagnosis of rare diseases is often delayed by years [1]. The main factor for this delay is believed to be the lack of knowledge and awareness regarding rare diseases [2]. Probabilistic diagnostic decision support systems (DDSS) have the potential to accelerate rare disease diagnosis by highlighting differential diagnoses for physicians [3, 4]. DDSS's are based on case input and incorporated medical knowledge.

Objectives: We examine a probabilistic DDSS prototype and assess its potential to provide accurate rare disease suggestions early in the course of rare disease cases.

Methods: Retrospectively, information from the medical records of 93 patients was transferred to the DDSS. Each of these patients had a confirmed rare inflammatory systemic disease. The accuracy of the DDSS disease suggestions was assessed for all documented visits over time. Time to correct top fit (TF) and top five fit (T5F) disease suggestion was assessed, as was the original time to clinical diagnosis (TD). TF/TD as well as T5F/TD were calculated to allow for comparison of TF respective T5F normalized to TD. Wilcoxon signed-rank test was conducted for TD-TF and TD-T5F.

Results: The DDSS suggested the correct disease at a time *earlier* than the time of clinical diagnosis among the top five fit disease suggestions in 53.8% of cases (50 of 93), and as the top fit disease suggestion in 37.6% of cases (35 of 93). Median advantage of correct disease suggestions compared to the time point of clinical diagnosis was 3 months or 50% for top five fit respective 1 month or 21% for top fit. The correct diagnosis was suggested at the *first* documented patient visit among the top five fit disease suggestions in 33.3% (top five fit), respective 16.1% of cases (top fit). Wilcoxon signed-rank test shows a significant difference between the time to clinical diagnosis and the time to correct disease suggestion for both top five fit and top fit (z-score -6.68, respective -5.71, $\alpha = 0.05$, p-value < 0.001). The DDSS suggested the correct rare disease at the time of diagnosis in 89% of cases (83 of 93)

Figure 1. Distribution of TF/TD by TD. Visualisation of the time to correct top fit suggestion relative to the time to diagnosis, grouped by time to diagnosis. Number of cases per group: 0m: 5; 1-12m: 33; 1-5y: 30; >5y: 25

Conclusion: The DDSS was capable of providing accurate rare disease suggestions in most of the rare disease cases. In many cases it provided correct rare disease suggestions early in the course of the disease, sometimes in the very beginning of a patient's journey. The interpretation of these results suggests that DDSS's have the potential to highlight the possibility of a rare disease to physicians early in the course of a case. Limitations of this study derive from its retrospective and unblinded design, data input by a single user, and the optimization of the knowledge base during the course of the study. Whether the use of this DDSS leads to a reduced time to rare disease diagnosis in a clinical setting should be validated in prospective studies.

REFERENCES:

- [1] Blöb S, et al. Diagnostic needs for rare diseases and shared prediagnostic phenomena: Results of a German-wide expert Delphi survey. PLOS ONE. 2017 feb;12(2):e0172532. Available from: <http://dx.plos.org/10.1371/journal.pone.0172532>.
- [2] Nationales Aktionsbündnis für Menschen mit Seltenen Erkrankungen (NAMSE). Nationaler Aktionsplan für Menschen mit Seltenen Erkrankungen. 2013. Available from: http://www.namse.de/images/stories/Dokumente/nationaler_aktionsplan.pdf.
- [3] Kostopoulou O, et al. Early diagnostic suggestions improve accuracy of family physicians: a randomized controlled trial in Greece. Family Practice. 2015 jun;32(3):323–328.
- [4] Riches N, et al. The effectiveness of electronic differential diagnoses (DDX) generators: A systematic review and meta-analysis. PLoS ONE. 2016 mar;11(3):e0148991. Available from: <http://dx.plos.org/10.1371/journal.pone.0148991>.

Disclosure of Interests: Simon Ronicke Employee of: Ada Health GmbH, Berlin, Martin C. Hirsch Shareholder of: Ada health GmbH, Ewelina Türk Employee of: Ada Health GmbH, Berlin, Katharina Larionov: None declared, Daphne Tientcheu: None declared, Annette D. Wagner: None declared

DOI: 10.1136/annrheumdis-2019-eular.6866

THU0565

COMPARISON OF SURVIVAL RATES AMONG SUBGROUPS OF PATIENTS EVALUATED FOR FEVER OF UNKNOWN ORIGIN

Emre Bilgin, Berkan Armagan, Alper Sarı, Ertuğrul Çağrı Bölek, Bayram Farisoğulları, Gözde Kübra Yardımcı, Levent Kılıç, Ali Akdoğan, Omer Karadag, Şule Apraş Bilgen, Ali İhsan Ertenli, Sedat Kiraz, Umut Kalyoncu. Hacettepe University Medical School Internal Medicine, Rheumatology, Ankara, Turkey

Background: Fever of unknown origin (FUO) is one of the most challenging clinical situations. Although several studies showed a relatively benign course of patients remained undiagnosed, long-term outcome of patients with a certain diagnosis remain non-established (1).

Objectives: To describe the follow-up results of patients investigated for FUO and had a certain diagnosis.

Methods: Data from patients who admitted to Hacettepe University Hospitals, inpatients sections of department of the internal medicine with the complaint of FUO collected prospectively from January 2015 to October 2017. Patients with an uncertain diagnosis after all diagnostic procedures excluded. Patients were divided into 3 main subgroups: rheumatologic, infectious and malignant groups. We compared Kaplan-Meier curves for all diagnosis-to-death time frames with the standart log-rank test. $p < 0.05$ was considered as statistically significant.

Results: Total 106 patients were included, 58(55%) of them were female. Median age was 48 (18-81) years. Patients were also divided into three subgroups: rheumatologic (RHE) (n=49, 46.2%), infectious (INF) (n=28, 26.4%) and malignant (MLG) (n=29, 27.4%) causes; adult-onset Still's disease (n=20; 41% of), tuberculosis (n=9; 32%) and lymphoma (n=19; 66%) were the most common diagnosis among groups, respectively. Mortality rates in decreasing order were RHE, INF and MLG groups, respectively. As an interaction was found between gender and survival, results of survival analysis were given as stratified according to gender. While there was no survival difference between RHE and INF groups among men, both of these groups had better survival than MLG group (ROM-INF $p=0.13$; ROM-MLG $p=0.001$; INF-MLG $p=0.022$). Among women, only there was a significant survival difference between RHE and MLG groups (ROM-INF $p=0.15$; ROM-MLG $p=0.007$; INF-MLG $p=0.42$).

Conclusion: Among patients evaluated for FUO, survival rate was higher in patients who had a rheumatological diagnosis. Further diagnostic algorithms are needed to identify these subgroups, because of the higher mortality among INF and MLG groups.

REFERENCES:

- [1] Knockaert DC, Dujardin KS, Bobbaers HJ. Long-term follow-up of patients with undiagnosed fever of unknown origin. Arch Intern Med. 1996;156(6):618.

Figure 1. Kaplan-Meier curves of male patients (RHE vs. INF $p=0.13$; RHE vs. MLG $p=0.001$; INF vs. MLG $p=0.022$)

Figure 2. Kaplan-Meier curves of female patients (RHE vs. INF $p=0.15$; ROM vs. MLG $p=0.007$; INF vs. MLG $p=0.42$)

Disclosure of Interests: Emre Bilgin: None declared, Berkan Armagan: None declared, Alper Sarı: None declared, Ertuğrul Çağrı Bölek: None declared, Bayram Farisoğulları: None declared, Gözde Kübra Yardımcı: None declared, Levent Kılıç: None declared, Ali Akdoğan: None declared, Omer Karadag: None declared, Şule Apraş Bilgen: None declared, Ali İhsan Ertenli: None declared, Sedat Kiraz: None declared, Umut Kalyoncu Grant/research support from: MSD, Roche, UCB, Novartis and Pfizer, Consultant for: MSD, Abbvie, Roche, UCB, Novartis, Pfizer and Abdi Ibrahim, Speakers bureau: MSD, Abbvie, Roche, UCB, Novartis, Pfizer and Abdi Ibrahim

DOI: 10.1136/annrheumdis-2019-eular.3374

THU0566

OCULAR FEATURES IN 381 PATIENTS WITH SYSTEMIC SARCOIDOSIS AND ITS CORRELATION WITH THE IWOS CRITERIA. STUDY IN A UNIVERSITY HOSPITAL

Belén Atienza-Mateo, José Luis Martín-Varillas, Rosalía Demetrio-Pablo, Vanesa Calvo-Río, Raúl Fernández Ramón, D. Prieto-Peña, Monica Calderón-Goercke, Lara Sánchez Bilbao, Iñigo González-Mazón, Miguel Á. González-Gay, Ricardo Blanco. Hospital Universitario Marqués de Valdecilla, Rheumatology and Ophthalmology, Santander, Spain

Background: Sarcoidosis is a multisystemic inflammatory disease characterized by non-caseating epithelioid granulomas that can affect any organ system. The three most frequency affected organs are lung, skin and eyes. Ocular involvement is the presenting symptom in approximately 20-30% and can involve any part of the eye and its anexal tissues. Sarcoidosis may cause uveitis, conjunctivitis, episcleritis/scleritis, optical nerve disease and orbital inflammation.

Objectives: To analyze the prevalence of ocular involvement is systemic sarcoidosis, the clinical patterns and their correlation with the International Workshop on Ocular Sarcoidosis (IWOS) criteria. These criteria classify ocular sarcoidosis as definite, presumed, probable and possible, according to some ophthalmological and analytical findings. They are especially useful if a biopsy is not obtained or it is negative.

Methods: Retrospective study of patients admitted to a single reference University Hospital between 1999 and 2019 with diagnosis of sarcoidosis. Clinical findings, demographics features, anatomic location and IWOS intraocular signs were recorded. We also collected serum angiotensin

converting enzyme (ACE), liver enzyme test, chest radiography, chest computed tomography scan, treatment and biopsy if performed.

TABLE. Demographic and clinical features of the 50 patients with ocular sarcoidosis.

	Patients (n=50)
Age, mean (SD) years	45.5 (16.7)
Sex, men/women, n/n	23/27
Affected organs, n (%)	
Lung	30 (60)
Skin	14 (28)
Liver	4 (8)
Kidney	3 (6)
Musculoskeletal system	3 (6)
Central nervous system	3 (6)
Hearth	2 (4)
Ocular signs, n (%)	
Bilaterality	22 (44)
Snowballs/strings of pearls	19 (38)
Mutton-fat KPs	12 (24)
Multiple chorioretinal peripheral lesions	7 (14)
Periphelebitis	5 (10)
Anterior synechiae	1 (2)
Optic disc granulomas	1 (2)
IWOS criteria, n (%)	
Definite	22 (44)
Presumed	13 (26)
Possible	4 (8)
Treatment, %	
Prednisone (oral)	44 (88)
Methylprednisolone (bolus)	5 (10)
Conventional IS	
.....MTX	21 (42)
.....AZA	9 (18)
.....CsA	2 (4)
.....MMF	2 (4)
Biologic therapy	
.....IFX	5 (10)
.....ADA	4 (8)
.....GLM	2 (4)

Abbreviations: ADA, adalimumab; AZA, azathioprine; CsA, cyclosporine A; GLM, golimumab; IFX, infliximab; IS, immunosuppressor; KPs, keratic precipitates; MMF, mycophenolate mofetil; MTX, methotrexate; SD: standard deviation.

Results: We selected patients with ocular inflammation from a cohort of 381 patients with sarcoidosis (n=50, 13%). Most of the cases were women (54%) and median age was 45.5±16.7 years. In these 50 cases, the most affected organ was lung (60%), followed by skin (28%). Forty patients had uveitis, 32 of them with ocular symptoms. Thirty-nine out of 50 patients (78%) met one of the 4 IWOS diagnostic categories: 22 with definite (44%), 13 presumed (26%) and 4 with possible (8%) sarcoidosis. Eleven patients did not meet IWOS criteria. The most common ocular signs were bilaterality (44%), snowballs or strings of pearls (38%), mutton-fat KPs (24%), multiple chorioretinal peripheral lesions (14%) and periphelebitis (10%). The median value of ACE was 69 U/l. Forty-four patients (88%) received oral corticosteroids, 21 (42%) received methotrexate, 11 (21%) received another conventional immunosuppressor and 11 (21%) a biological treatment. TABLE shows demographic and clinical features.

Conclusion: In our population the IWOS Criteria had a sensitivity of 78%. Even though there is no gold standard for diagnosing ocular sarcoidosis yet, IWOS signs can help clinicians suspect it.

REFERENCES:

- [1] Herbert CP, Rao NA, Mochizuki M; members of Scientific Committee of First International Workshop on Ocular Sarcoidosis. International criteria for the diagnosis of ocular sarcoidosis: results of the first International Workshop On Ocular Sarcoidosis (IWOS). *Ocul Immunol Inflamm.* 2009 May-Jun;17(3):160-9.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.4248

THU0567

POLYMYALGIA RHEUMATICA-LIKE SYNDROME FROM CHECKPOINT INHIBITOR THERAPY: CASE SERIES AND SYSTEMATIC REVIEW OF THE LITERATURE

cassandra calabrese¹, Laura Cappelli², Marie Kostine³, Elizabeth Kirchner¹, Tawnie Braaten², Leonard Calabrese¹. ¹Cleveland Clinic Foundation, *Rheumatic and Immunologic Disease, Cleveland, United States of America;* ²Johns Hopkins University, *Rheumatology, Baltimore, United States of America;* ³Centre Hospitalier Universitaire de Bordeaux, *Rheumatology, Bordeaux, France*

Background: Checkpoint inhibitor therapy has caused a paradigm shift in the field of oncology, producing significant survival benefits in patients with an ever-growing list of malignancies, but their use is attended by a spectrum of immune related adverse events (irAEs). Many rheumatic irAEs have been described. A critical and presently unanswered question is what proportion of these rheumatic irAEs represent new appearances of classic rheumatic diseases or, alternatively, represent new clinical variants with potentially different pathogenesis, clinical course and treatment responsiveness. Little is known about the PMR-like entity that has been described in the setting of ICI therapy.

Objectives: To assess whether the polymyalgia rheumatica (PMR)-like syndrome reported as an irAE from checkpoint inhibitor therapy is consistent with the 2012 EULAR/ACR provisional criteria for PMR¹.

Methods: The cases were derived from two sources. Group 1 represents reported cases from three contributing centers. Group 2 was derived from a systematic review of the literature searching for all cases reported as PMR or PMR-like illness associated with checkpoint inhibitor therapy. Cases were assessed for the quality of reporting and then analyzed to determine whether they fulfilled the 2012 EULAR/ACR provisional criteria for PMR.

Results: A total of 49 patients were included for analysis, including 9 cases from Cleveland Clinic (Table 1), 4 cases from Johns Hopkins University, and 7 cases from University Hospital of Bordeaux, in addition to the 29 separate cases found by systematic review. Among the entire group, 37 (75%) were designated "complete" indicating that they had sufficient data to reliably apply the 2012 EULAR/ACR criteria of which 28 (75%) cases fulfilled complete criteria for PMR. A number of cases also demonstrated some clinical features unusual for idiopathic PMR.

Conclusion: This study suggests a high proportion of reported cases of checkpoint inhibitor-related PMR fulfill preliminary criteria for PMR yet in one quarter clinical details were incomplete making verification problematic. Furthermore, in the absence of a gold standard for the diagnosis of PMR, the relationship of checkpoint inhibitor-related PMR to the idiopathic form remains unclear.

REFERENCES:

- [1] Dasgupta B, Cimmino MA, Maradit-Kremers H, et al. 2012 provisional classification criteria for polymyalgia rheumatic: a European League Against Rheumatism/American College of Rheumatology collaborative initiative. *Ann Rheum Dis* 2012;71:484-492.

Disclosure of Interests: cassandra calabrese Speakers bureau: Regeneron/Sanofi, Laura Cappelli Grant/research support from: Bristol-Myers Squibb, Consultant for: Regeneron/Sanofi Genzyme, Marie Kostine: None declared, Elizabeth Kirchner Consultant for: Celgene, Horizon, Novartis, Regeneron, Speakers bureau: Merck and Sanofi, Tawnie Braaten Grant/research support from: supported by T32 grant and "research reported in this publication was supported by the National Institute of Arthritis and Musculoskeletal and Skin Diseases of the National Institutes of Health under Award Number T32AR048522. The content is solely the responsibility of the authors and does not necessarily represent the official views of the National Institutes of Health.", Leonard Calabrese Consultant for: Bristol-Myers-Squibb, Genentech and Astra-Zeneca.

DOI: 10.1136/annrheumdis-2019-eular.1389

THU0568

CLINICAL CHARACTERISTICS OF SEVEN RELAPSING POLYCHONDROITIS CASES MISDIAGNOSED AS ARTHRITIS

Xiaoyu Cao, Jiu-Liang Zhao, Dong Xu, Yong Hou, Xiaofeng Zeng. *Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Graduate School of Peking Union Medical College, Beijing, China*

Background: Relapsing polychondritis (RPC) is a rare autoimmune disease. It mainly affects the tissue abundant of cartilage, such as ear, nose, and tracheoles. The multiple systems including eye, joint, mucocutaneous, and kidney could be involved¹. As the presentation of arthritis was nonspecific and could appear in several connective tissue diseases,