person specific and influenced by many factors (e.g. psychosocial factors, illness perceptions, sleep, comorbidity etc.). A thorough history alone usually suggests the single most likely cause for the patient's problem(s). The history should then guide the subsequent physical examination – an efficient targeted "rapier" approach is recommended in which the practitioner selects the appropriate skills from a range of competencies according to specific elements in the history. This contrasts with a more lengthy hypothesis-free "general screen" in which the same set of uniform procedures is undertaken in each patient.
This presentation will cover key principles and considerations of assessment and illustrate how the history guides the subsequent "rapier" examination (2). Examples include:
(1) in the *history:* determination of pain localisation and features that associate with radiated pain; important pain and stiffness characteristics that differentiate mechanical usage-related pain, inflammatory pain, acute crystal synovitis pain, destructive bone pain and neurogenic pain; non-specific symptoms of inflammation (2) in the *examination:* usual order of inspection at rest, inspection during movement, then palpation at rest and during movement of symptomatic regions; contrasting clinical findings that quickly differentiate joint and peri-articular problems; initial selection of the movement(s) that is affected first and most severely by arthropathy - the tight pack position(s); detection of "stress pain" (pain worse in tight-pack positions but reduced/absent in loose-pack positions - the most sensitive sign of inflammation); examination for effusion, soft-tissue and firm swelling; use of resisted active movements and stress tests for peri-articular lesions; a targeted screen for asymptomatic disease prompted by main diagnosis.
EULAR learning resources available at http://www.eular.org/edu_training_dvd.cfm include: (1) The "GALS" screen and (2) Principles of the musculoskeletal history and examination.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.2947

SP0103 **HOW TO ASSESS US COMPETENCY SKILLS**

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To optimize and standardize musculoskeletal ultrasonography education for rheumatologists, there is a need for competency assessments addressing the required training and practical and theoretical skills. Because of the increasing use of MSUS in rheumatology, there has been a focus over the past years on training.
A minimum training requirements are described by The European Federation of Societies for Ultrasound in Medicine and Biology (EFSUMB) where a 3 levels competency assessment (COMPASS) has been developed for rheumatologists, including an in-detail description of what theoretical and practical competencies to be acquired at each level with a related log book (1). The rheumatology-COMPASS levels are closely related to the levels of the EULAR MSUS courses, thereby ensuring that the content is supported by already provided courses such as the EULAR and EULAR-endorsed MSUS courses to facilitate the implementation of the rheumatology-COMPASS. In COMPASS level 1 the course contents resemble the EULAR MSUS basic and intermediate courses, level 2 resembles the EULAR MSUS advanced course whereas level 3 requires attendance in a "teach-the-teachers course" or experience as a teacher in at least 2 international MSUS courses. Level 3 also includes an academic level requiring research activity and acceptance of level 1 and 2 sonographers for training.
The EULAR MSUS courses have been organized since 1998 and the interest in these courses has been increasing. In 2007, the first 3 level EULAR MSUS course was conducted with great success and the 3 level courses (basic, intermediate and advanced) have been running ever since in relation to the EULAR congress, focusing mainly on the relevant content on the individual levels and the distribution between practical and theoretical skills.
EULAR has developed the following competence levels; level 1 and 2. The EULAR level 1 competency includes the performance of EULAR Online MSUS course and attendance to basic, intermediate and advanced MSUS courses, where attending the intermediate and advanced courses require a certain number of US examinations (however, if already reached COMPASS level 1, 2 or 3, there is no need of images for the EULAR courses), and the advanced course requires in addition to pass a practical examination. The EULAR level 2 competency is organized to ensure a minimum level of US knowledge for teachers in MSUS courses (2). This level includes the EULAR Teach the Teachers course as well as passing a theoretical and practical examination. Since there is a growing number of EULAR endorsed MSUS courses, it is of highly importance that the teachers in these courses have equal qualifications thereby providing comparable training and competencies beneficial for the clinical use of US. Information about the competence requirements is found at the EULAR website (http://www.EULAR.org).
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.7147

THURSDAY, 15 JUNE 2017

Macrophage M2 polarization: implications in fibrotizing diseases

SP0104 **REPROGRAMMING OF MYELOID CELLS IN CANCER: MECHANISMS AND SIGNIFICANCE**

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Chronic inflammation mediates tumor development by promoting a constant influx of inflammatory cells capable of modulating genes involved in cancerogenesis and creating micro and macroenvironments that support cancer growth. A major side effects of cancer inflammation is the pathological expansion and recruitment of myeloid cells endowed with immunosuppressive activity, to control the unresolved inflammation. Tumors reprogram myeloid cell differentiation and functions through various mechanisms, including altered metabolism, cancer-related inflammation andalteration of the hematopoietic output. These events govern the expansion of myeloid suppressor populations, mainly myeloid-derived suppressor cells (MDSCs) and tumor associated macrophages (TAMs). MDSCs and TAMs orchestrate tumor immunosuppression in concert with regulatory T cells, inhibitory cytokines and immune check points receptors, and act to subvert anti-tumor immunity, hence causing that eventually support immune evasion establishing a bottleneck for cancer immunotherapy. I will discuss inflammatory circuits and epigenetic events sustaining the expansion and the tumor-promoting reprogramming of myeloid cells in cancer bearers.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.7285

SP0105 **MACROPHAGES, METABOLISM AND INFLAMMATION**

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Macrophages are a heterogeneity population implicated in several diseases and correlated with distinct tissue outcomes after injury. M2 macrophages have been associated with tissue repair whereas M1 macrophages participate in early phase of tissue damage. Recent works have also suggested that upon activation, macrophages can use distinguished nutrients as source of energy and these metabolic pathways lead to their activation and differentiation. Nutrient sensors are intimae associated with innate receptors and therefore connected with inflammatory response. Uric acid is a damage-associated molecular pattern (DAMP), released from ischemic tissues and dying cells which, when crystalized, is able to activate the NLRP3 inflammasome through frustrated phagocytosis. Soluble uric acid (sUA) is found in high concentrations in the serum of great apes, and even higher levels in some diseases, before the appearance of crystals. sUA can be released in a hypoxic environment and triggers NLRP3 through the production of mitochondrial ROS, with increased maximum and reserve oxygen consumption ratio (OCR) and higher VDAC protein levels. This process is followed by ASC speck formation, caspase-1 activation and IL-1β release. These findings may have profound implications for inflammatory-related diseases.
Support: FAPESP and CNPq.
Disclosure of Interest: N. Camara Grant/research support from: FAPESP, CNPq
DOI: 10.1136/annrheumdis-2017-eular.7283

FRIDAY, 16 JUNE 2017

Mucosal B cells: gatekeepers of immune function

SP0106 **THE LUNG AS A DRIVER OF RA-ASSOCIATED AUTOIMMUNITY**

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Rheumatoid arthritis (RA) is a chronic inflammatory disease resulting from the complex interaction between genes and environment. In a large majority of patients this interaction leads to formation of disease-specific anti-citrullinated proteins antibodies (ACPA). Systemic autoimmunity may be triggered at mucosal sites (such as the lungs) long before the first signs of inflammation are starting in the joints. According to this model, smoking (and others environmental triggers) induces subclinical inflammation in the lungs, leading to increased local citrullination and formation of ACPA in genetically susceptible individuals. Lung changes on high-resolution computer tomography are present in both early-untreated ACPA positive RA and ACPA positive individuals at risk for but not yet having disease. Further, signs of subclinical inflammation and immune activation with germinal centers formation and ACPA enrichment is present in early untreated RA. Shared citrullinated targets have been described in the lungs and joints of patients with RA and more recent data unravels novel mechanisms showing how this extra-articular triggered autoimmunity progresses to joint-specific inflammation. Beside an initiating role, the lung might also be a secondary target for antibodies, especially in longstanding seropositive RA.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.7293

FRIDAY, 16 JUNE 2017

WIN & HOT session

SP0107 HOT SESSION: SCLERODERMA TREATMENT

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This presentation will provide an up to date summary of current management of SSc that can be applied across the disease spectrum. Systemic sclerosis (SSc) remains a challenging multifaceted rheumatic disease with high mortality and morbidity. However, treatments are emerging for some aspects of the disease and long term survival has improved significantly over the past decades. This session will review the clinical challenge and current therapeutic landscape of SSc focusing on practical aspects of management such as identifying and treating significant organ based complications in the lung, heart, kidney and gastrointestinal tract. Current approaches to overall disease management will be summarized including the use of haemopoietic stem cell transplant in selected poor prognosis cases. Recently updated EULAR/EUSTAR treatment recommendations will be reviewed and other evidence based management guidelines will be considered within a practical patient-focused framework.

Disclosure of Interest: C. Denton Grant/research support from: Inventiva, CSL Behring, GSK, Bayer, Consultant for: GSK, Actelion, Inventiva, Roche

DOI: 10.1136/annrheumdis-2017-eular.2534

FRIDAY, 16 JUNE 2017

Comorbidities in rheumatoid arthritis

SP0108 HERPES ZOSTER: HOW TO PREVENT, TO DIAGNOSE AND TO TREAT

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Herpes Zoster is a major public health problem and is an infection that results from re-activation of latent varicella infection acquired most commonly naturally or more recently through immunization. The incidence of HZ is approximately twice that of the general population in patients with immune mediated inflammatory diseases (IMIDs). Underlying mechanisms are largely those which compromise cell mediated immunity and epidemiologic risks largely follow immunosenescent patterns (i.e. aging). Rheumatologists use a large variety of immunosuppressive drugs which further increase the risk of HZ and are obliged to recognize the clinical syndrome, its complications, apply effective therapy and be actively engaged in strategies to maximize immunization and prevention. This discussion will focus on recent advances in each of these areas highlighting newly described complications of HZ such as stroke and advances in vaccine development.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.7214

FRIDAY, 16 JUNE 2017

Life-threatening presentation of rheumatic diseases

SP0109 ACUTE RESPIRATORY FAILURE, MACULO-PAPULAR RASH, INDURATIVE EDEMA OF THE EXTREMITIES AND CERVICAL LYMPHADENOPATHY IN A 6-WEEK-OLD INFANT

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Case report: A previously healthy 40-days-old male infant, from non-consanguineous parents from Morocco, presented in a peripheral hospital with fever for 48 hours, associated with rhinorrhea, mild diarrhea and progressive irritability during the last 24 hours. Initial laboratory studies revealed elevated acute phase reactants (CRP 116 mg/L), mild neutrophilia, elevated liver enzymes (AST 315, ALT 174 U/L), direct hyperbilirubinemia, discoagulopathy. Cerebral spinal fluid analysis and microbiology cultural workup resulted negative. A wide-spectrum antibiotic and antiviral therapy was initiated. On the 6th day of illness he developed diffuse maculo-papular rash, indurative edema of the extremities, right cervical lymphadenopathy and bilateral conjunctival injection. On the basis of a certain clinical diagnosis of Kawasaki disease and the infant was treated with IGIV 2 g/kg. An echocardiography performed prior to the IGIV infusion showed homogeneously dilated coronary arteries (left coronary artery 3.2 mm, right coronary artery 2.2 mm). Twelve hours after the end of IVIG infusion, the child presented a rapidly progressive, severe respiratory failure requiring endotracheal intubation and was transferred to our ICU. On admission (day 7th), physical examination revealed a feverish, critically ill-infant with hepatomegaly (5–6 cm below the right costal margin), diffuse maculo-papular rash, “sock-like” erythema and swelling of the feet, cheilitis, bilateral conjunctival injection and right cervical adenopathy. The urine output was markedly decreased; he rapidly developed hemodynamic instability with hypotension and tachycardia. Complete blood count showed anemia (6.8 g/dl), thrombocytopenia (16.000/mmc), elevated CRP (240 mg/L), hypoalbuminemia (18 g/dl) and hypofibrinogenemia (0.83 g/L); liver enzymes were normal. Intensive ventilatory and hemodynamic support therapy were started, in addition to a massive transfusional regimen.

Given the clinical and hematological picture, the diagnosis of MAS was considered and subsequently confirmed by high ferritin level (2197 mcg/L), AST above the normal value (61 U/L) and hypertriglyceridemia (181 mg/dl) [2]. The clinical suspicion was supported by persistent cytopenia despite daily transfusions, low erythrocyte sedimentation rate (3 mm/h) with concomitant rising CRP, elevated IL2-R level (28.320 KU/L) and decreased NK function. The patient was treated with high dose methylprednisolone pulse therapy (25 mg/kg) for 3 consecutive days 12–14), followed by a maintenance of 1 mg/kg/daily. By day 15th, a progressive decrease in inflammatory markers and a concomitant improvement of general conditions was observed, with the possibility to discontinue inotropic support on day 12th and invasive ventilation on day 25th. Since day 17th, a diffuse cutaneous desquamation was noted. The fever settled on day 35th.

Echocardiography follow-up revealed an increasing, irregular dilation of left (max 5 mm) and right (max 3.5 mm) coronary arteries, with a progressive left ventricular apex hypocinesis, but a stable ejection fraction (55%). ECG showed persistent repolarization abnormalities.

Of note, Adenovirus-PCR was found positive in the bronchoalveolar washing performed on admission.

The differential diagnosis included: Kawasaki disease complicated by respiratory distress syndrome and MAS, familiar hemophagocytic lymphohistiocytosis, autoimmune lymphoproliferative syndrome, criopyrinopathies and immunodeficiency.

Comment: Kawasaki disease can be catastrophic in the early infancy, due to atypical presentation and resistance to the conventional therapy. Despite typical in its presentation, this case was complicated by two challenging conditions, respiratory distress syndrome and MAS, which hampered the diagnostic and therapeutic management during the course of the disease and required massive intensive support.These two complication are rarely associated to KD. In this particular case, respiratory failure can probably be explained by a combination of causes: fluid overload, systemic vasculitis and the concomitant and probably triggering Adenovirus infection.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.7176

SP0110 LIFE-THREATENING COMPLICATIONS IN SYSTEMIC VASCULITIS

M.C. Cid on behalf of G. Espigol-Frigolé, S. Prieto-González, J. Hernández-Rodríguez, A. García-Martínez. *Systemic Autoimmune Diseases, Hospital Clinic. University of Barcelona, Idibaps, Barcelona, Spain*

Systemic vasculitis may present with life-threatening complications that need to be promptly recognized and appropriately managed to ensure patient survival and minimize irreversible organ damage.

The most common life-threatening events differ between large and medium or small-vessel vasculitis. In large-vessel vasculitis, particularly giant-cell artertis (GCA), vascular remodelling in response to inflammation may lead to severe stenosis leading to ischemic stroke in 3–6% of patients and, rarely, myocardial infarction, mesenteric ischemia or critical extremity ischemia. For these complications, particularly when happening in treated patients, intensifying immunosuppressive therapy is not the best or only option and additional interventions may be required. Stroke occurs as a consequence of carotid or vertebral stenosis. Stenosis of the carotid siphon has been repeatedly reported. In necropsy studies, vasculitic involvement and thrombosis of proximal intracranial branches has been observed. Infarcts are usually multiple, usually happen shortly after the initiation of glucocorticoid therapy, convey a 30% mortality or lead to remarkable disability. If critical stenosis is suspected before irreversible infarction, percutaneous intraluminal angioplasty may be function and life saving.

Acute aortic syndrome (aortic dissection or intramural haematoma) is an increasingly recognized hurdle in patients with GCA. It is usually a delayed complication occurring months and frequently years after diagnosis. Its frequency has not been delimitated but in a recent prospective follow-up study it was demonstrated to affect at least 2% of patients. Emergency open surgery repair, when feasible, is the best option for ascending aorta involvement (type A) and endovascular repair for involvement of the descending aorta (type B).

In small-vessel vasculitis, life-threatening presentations include rapidly progressive glomerulonephritis leading to kidney failure, alveolar haemorrhage, alithiasic colecistitis with perforation and intestinal ischemia. Rapidly progressive glomerulonephritis and alveolar haemorrhage are more frequently seen in anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) and anti glomerular basement membrane disease. Alveolar haemorrhage can be occasionally seen in cryoglobulinemic or IgA vasculitis. In addition to supportive measures, these patients are usually treated with high-dose methyl-prednisolone, cyclophosphamide or rituximab and plasma exchange. Plasma exchange has been found superior to IV methyl prednisolone mega-doses in preserving or recovering renal function but this advantage do not seem to persist over long-term follow-up. This approach is usually applied also to alveolar haemorrhage although there is not strong evidence supporting it.

Gastrointestinal complications, particularly intestinal ischemia and intestinal or gallbladder perforation are life-threatening complications which may require emer-