of liver,abnormal fat metabolism and hemophagocytosis were common clinical features.Two cases were with ARDS and MOF in three and three died. The perforin A91V (NCBI:SNP rs35947132) gene was detected in seven systemic onset juvenile idiopathic arthritis compolicated with MAS cases, but no mutation were found. Glucocorticoid, intravenous immunoglobulin, immunoimpressive therapy were effective and HP (Plasmapheresis) used in one serious case was also effective.
Conclusions: MAS is a rare and potentially fatal complication of childhood rheumatoid diseases,especially systemic onset juvenile idiopathic arthritis. Most of our patients were male, and most cases were preceded by infection. Bone marrow studies support the diagnosis. MOF may be a poor prognostic sign.Aggressive early therapy is essential.
Disclosure of Interest: None declared
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AB0287 MUTUAL ASSENT TOWARDS COMPREHENSIVE DISEASE CONTROL: THE RELATIONSHIP BETWEEN US MEASURES AND PATIENT REPORTED OUTCOMES IN EARLY RHEUMATOID ARTHRITIS

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Objectives: Assessment of the relationship between US measures of joint inflammation/damage and patient reported outcomes (PROs): HAQ, pain and patient global assessment in early rheumatoid arthritis (early RA) patients over 5-years follow up period.
Methods: This longitudinal cohort of 261 patients with early RA was derived from the US monitoring study [1]. Adopting OMERACT definitions; correlations between total US scores (synovial hypertrophy, synovial fluid, Power Doppler, bone erosion and tenosynovitis) and PROs [2] namely functional disability (HAQ), pain and patient global scores were determined at 0, 1, and 5years. Radiological damage was assessed using modified Total Sharp score (mTSS). Univariate correlations as well as correlations between interval changes were assessed. Multivariable regression models were used to evaluate the associations over all time-points and their relationship to clinical disease activity measures.
Results: There were significant correlations ($p<0.01$) between total US score and HAQ (r=0.71), pain (r=0.69) and patient global scores (r=0.66) at all timepoints. The association tends to be stronger with increase disease duration (Spearman correlation 0.12 at baseline, 0.22 at 1-year and 0.41 at 5-years). Change in mTSS score at 5-years was not associated with changes in PROs. Improvements in US scores were also associated with improvements in PROs. Multivariate models revealed that synovial hypertrophy and Power Doppler scores were associated ($p<0.01$) with functional disability, pain and patient global assessment, controlling for clinical disease activity measures. Studying the pattern of joint involvement, it was associated significantly ($p<0.01$) with the US score of the affected joints. US total score at 1-year predicted subsequent 5-year HAQ score (R2=0.17). At 0, 1- and 5-years, total US scores were higher in patients whose HAQ score was >1 (9.26) compared to those below 1 (4.16, $p<0.01$).
Conclusions: the link between joint inflammation/structural damage and PROs is of critical importance to the care of patients with inflammatory arthritis. US measures of inflammation and structural damage correlated independently with physical function, pain and patient global assessments. A clear relationship between radiographic structure damage and the patient's perceived remission/flare provide the basis for comprehensive disease assessment and management.
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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.4485

AB0288 LABORATORY MARKERS OF INFLAMMATION AND SERUM NICOTINAMIDE PHOSPHORIBOSYLTRANSFERASE LEVEL IN RHEUMATOID ARTHRITIS PATIENTS

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Objectives: To study relationship between serum levels of nicotinamide phosphoribosyltransferase and laboratory markers of inflammation in patients with rheumatoid arthritis (RA).
Methods: We determined nicotinamide phosphoribosyltransferase level in sera of 140 patients with RA (96 women and 44 men) by indirect enzyme-linked immunosorbent assay (RaiBiotech, cat No. EIA-VIS-1). The control group consisted of 20 women and 10 men aged 22 to 55 years without complaints of pain in the joints throughout life. The mean duration of disease was 5.94±0.37 years.
Results: We divided all RA patients into 2 groups: one group (118 patients) with elevated levels of nicotinamide phosphoribosyltransferase serum (more than 3.9 ng/ml) and second group (22 patients) - with normal range.
In each of the two groups, the levels of CRP and ESR were determined.
Patients with elevated levels of Nampt had the following laboratory parameters (M±m): ESR – 37.83±1,57, CRP – 56.09±3,73 (rate - less than 5.0 mg/l). The second group had following data: ESR 22.46±0,56, CRP 21,65±1.38. Thus, patients with elevated levels of nicotinamide phosphoribosyltransferase had significantly higher concentrations of ESR and CRP ($p<0,001$).
Conclusions: There is the relationship between the level of nicotinamide phosphoribosyltransferase serum and laboratory markers of inflammation in RA (CRP and ESR). The data indirectly confirm the hypothesis that increased levels of nicotinamide phosphoribosyltransferase in RA patients is associated with disease activity.
Disclosure of Interest: None declared
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AB0289 PREDICTOR OF THE SIMPLIFIED DISEASE ACTIVITY INDEX 50 (SDAI 50) AT MONTH 6 DURING BDMARDS TREATMENT IN PATIENTS WITH LONG-ESTABLISHED RHEUMATOID ARTHRITIS: A SINGLE-CENTER, RETROSPECTIVE STUDY

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Background: The simplified disease activity index (SDAI) 50 has good agreement with the EULAR response measures for early rheumatoid arthritis (RA). Although there are reports on early RA, there have been no reports on long-established RA.
Objectives: In this study, we analyzed the relationships between various baseline factors and SDAI 50 after six months of biological disease-modifying antirheumatic drugs (bDMARDs) treatment to determine the prognostic factors for long-established RA.
Methods: The subjects were 332 RA patients who had been treated with bDMARDs for 6 months. The following characteristics were investigated: age, gender, disease duration, smoking history, body mass index, steroid and methotrexate dosage, previous bDMARDs use, combined csDMARDs use, ESR, CRP, serum matrix metalloproteinase-3 levels, SDAI score, health assessment questionnaire disability index score (for daily living activities) and short form-36 score (for quality of life). As a primary outcome index, SDAI response was defined as a 50% reduction in SDAI score between baseline and 6 months (SDAI 50).
Results: The group of RA patients who achieved SDAI 50 (Group A: 204 patients) had a higher tender joint count (p=0.041), swollen joint count (p=0.001), evaluator's global assessment (p=0.027) and SDAI (p=0.006) than did those who did not achieve SDAI 50 (Group B: 152 patients). Before the start of the treatment, steroid dosage (p=0.0187, odds ratio: 1.119, 95% CI: 1.029–1.229) and SDAI (p=0.0003, odds ratio: 0.953, 95% CI: 0.928–0.978) were determined based on logistic regression analysis. Comparisons were performed between Groups A and B and between before treatment and after 6 months of SDAI. Group A showed a significant improvement compared to Group B by repeated measure analysis of variance (ANOVA) (Interaction: p=0.000, Group A vs. Group B: p=0.000, before vs. after treatment: p=0.000).
Conclusions: Our study demonstrated that RA patients with a lower steroid dosage and higher SDAI baseline are more likely to achieve SDAI 50 with bDMARD treatment.
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Disclosure of Interest: Y. Miwa Grant/research support from: Astellas Pharma Inc., Mitsubishi Tanabe Pharma Corporation, AbbVie CK, Pfizer Japan Inc., Chugai Pharmaceutical Co., Ltd., Eizai Co., Ltd, Asahi Kasei Pharm Co., Ltd, YL biologics Ltd., Japan Blood Products Organization,. Ono Pharmaceutical Co., Ltd., Nippon Kayaku Co., Ltd., and Teijin Pharma Ltd., A. Nishimi: None declared, S. Nishimi: None declared, T. Tokunaga: None declared, S. Ishii: None declared, T. Kasama Grant/research support from: Mitsubishi Tanabe Pharma Corporation and AbbVie CK.
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Rheumatoid arthritis - comorbidity and clinical aspects

AB0290 PULMONARY AMYLOIDOSIS IN RHEUMATOID ARTHRITIS – A POSTMORTEM CLINICOPATHOLOGIC STUDY OF 161 PATIENTS

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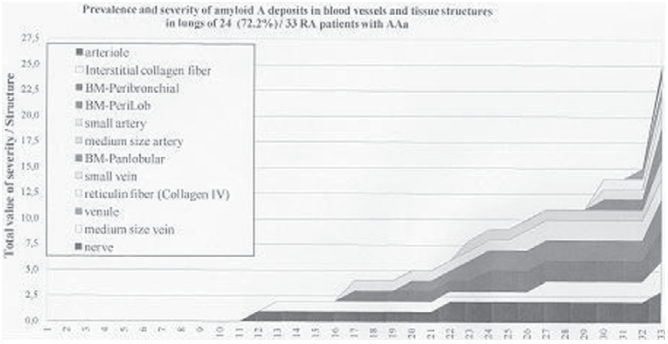
Background: AA amyloidosis (AAa) is one of the most insidious systemic complications of rheumatoid arthritis (RA), which furtively may lead to death [1].
Objectives: The aim of this study was to determine the prevalence and location of amyloid A deposition in the lungs of RA patients at the time of death.
Methods: A randomized autopsy population of 161 in-patients with RA was studied. AAa complicated RA in 34 (21.1%) cases [1].

At least four tissue samples of lungs (from apical and basal regions of both lungs) were available for histologic evaluation in 33 of these 34 patients.
RA was confirmed clinically according to the criteria of the ACR.
The presence of amyloid A deposits in various structures of the lungs was determined histologically by amyloid specific Congo red staining, according to Romhányi [2].
The extent of amyloid A deposition was evaluated by semi-quantitative, visual estimation on a 0 to 3 plus scale, based on the number of involved tissue structures per light microscopic field [1]. ("0": no amyloid deposits, "1": Sporadic, minimal amyloid deposits on different tissue structures, "2": less than five, "3": five or more involved tissue structures per microscopic field at objective magnification of x20)
Results: Amyloid A deposition in the lungs was detected in 24 of 33 (72.2%) patients.
Amyloid deposition in various structures does not begin at the same time.
In the early stage of systemic amyloidosis there were histologically detectable amyloid deposits only in a few structures (arterioles, interstitial collagen fibers, peribronchial and perilobular basement membranes). In other structures (small and medium size arteries, panlobular basement membranes, small veins, collagen IV reticulin fibres, venules, medium size veins and nerves) deposits were seen only in late stages of amyloidosis (with massive involvement of the mentioned structures).



Conclusions: Amyloidosis is a progressive, cumulative process, involving in its early stage only a few structures in some organs, and increasingly more in the later stages of the disease [1]. Amyloid A deposition starts in the most frequently involved structures of the most frequently involved organ [1].
In the lungs amyloid A deposition starts in the wall of arterioles and in interstitial collagen fibers. As time progresses, basement membranes of peribronchial and peripheral regions of lobules, small and medium sizes arteries become involved. Still later panlobular deposition of basement membranes, small veins, reticulin fibers (collagen IV) of subpleural fat tissue, venules and medium size veins become involved. The involvement of nerves indicates advanced stages of amyloid deposition in the lung.
This chronology of amyloid A deposition allows an indirect assessment of the stage of amyloidosis. Based on the involvement of structures in lung biopsy specimens the pathologist may be able to estimate involvement of the other structures, even if not present in the sections. Involvement of arterioles alone (without involvement of small arteries) indicates an early stage of amyloidosis, whereas amyloid A deposits in veins or peripheral nerves suggests an advanced stage with massive involvement of other pulmonary structures.
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Disclosure of Interest: None declared
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AB0291 EPIDEMIOLOGY AND COMORBIDITY OF RHEUMATOID ARTHRITIS IN UPPER EGYPT, A HOSPITAL BASED STUDY

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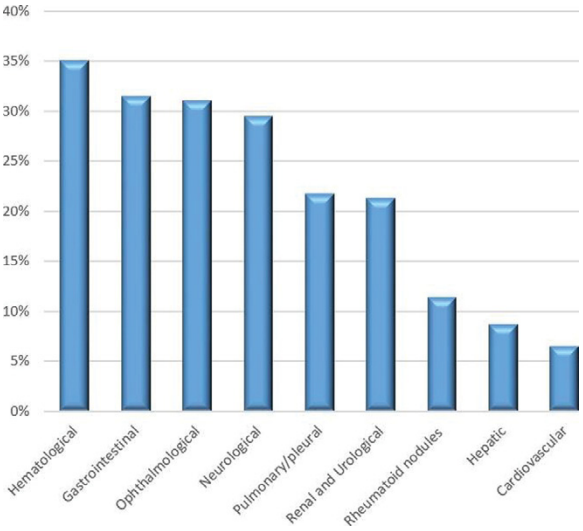
Background: Rheumatoid arthritis (RA) is one of the commonest autoimmune diseases. It affects about 1% of the population worldwide (1). The prevalence of RA varies widely between different countries (2). Not only the prevalence of the disease which differs among different continents, races, ages and socioeconomic levels but also the disease pattern. Studies explaining the epidemiology of RA in Egypt in general and in upper-Egypt, in particular, are very limited (3).
Objectives: To estimate the comorbidity of rheumatoid arthritis and its relation to disease activity, duration, disease pattern and demographic features of RA patients in upper Egypt.
Methods: This study was carried out on 923 patients who fulfilled ACR/EULAR criteria 2010. All of them live in Sohag governorate and aged 18 years or older DAS28-ESR score, first involved joint, joint distribution, disease pattern, extra-articular comorbidities including gastrointestinal, urinary, cardiac, haematological and neurological were estimated. The activity of daily living was valued by Erlangen score (E-ADL).
Results: The mean age of the participants was 45±10.9 years, with a range (19–70). The median of the disease duration was 5 years, with a range (0.5–40 years). Most of the participants were female (691, 74.9%). Disease onset was gradual or insidious in 94.3% of cases and acute in 5.7% of them. First joint group affected were the small joints of the hands (MCPs and PIPs), recorded in 48.9% of cases, Followed by wrist joints (29.3% of cases), then knees (9%), ankles and small joints of the foot (6%) and lastly other joints collectively recorded in only 6.8%. The commonest extra-articular comorbidities were haematological, seen in 323 cases; 35%, followed by gastrointestinal in 290 cases (31.4%), then ophthalmological in 31%, entrapment syndromes in 29.4%, pulmonary in 21.7%, urological in 12.4%, rheumatoid nodules in 11.4%, liver cirrhosis in 8.7%, renal impairment in 8.5% and Cardiovascular diseases in 6.5%. The activity of daily living (E-ADL) showed that most of the cases fell in score 4 (58.2%). Regarding DMARDs treatment of the study population, Methotrexate (MTX) was used regularly by 78.3% of cases, hydroxychloroquine (HCQ) by 78.1%, followed by Leflunomide (LEF) by 26.4% and sulfasalazine (SSZ) by 13.1%. The majority of cases used combination therapy of either MTX+HCQ, MTX+SSZ, MTX+HCQ+SSZ or MTX+LEF. Regarding other drugs, 99% of cases used NSAIDs (regularly in 30.2% and on demand in 68.8%). Steroids were regularly used by 28.8% of cases.



Conclusions: The commonest comorbidities were haematological, gastrointestinal, ophthalmological and neurological ones; respectively. Erosion, deformity and Das28-ESR score have a great impact on E-ADL score.
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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.1440

AB0292 CLINICAL FEATURES OF RHEUMATOID ARTHRITIS AT 75 YEARS OF AGE AND OLDER IN JAPAN – COMPARISON WITH POLYMYALGIA RHEUMATICA IN THE SAME AGE GROUP

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Background: As Japan is a super-aged society, we have many chances to care for elderly patients in our hospital. Elderly-onset rheumatoid arthritis (RA) (onset age >60 years) may present similar symptoms to those of polymyalgia rheumatica (PMR). We consider that differential diagnosis of RA and PMR is more difficult in patients over 75 than those under 74 in clinical practice.
Anti-cyclic citrullinated peptide antibody (ACPA) was reported to be a helpful tool in the differential diagnosis of EORA from PMR. However, when elderly patients with negative ACPA complained of bilateral shoulder and/or girdle pain, it was difficult to differentiate PMR from RA.
Objectives: The study aimed to explore clinical features of RA and PMR at onset age 75 years. For the present investigation, we used a novel diagnostic