37.20 ± 7.36 U/L for severe patients ($P = 0.03$). Correlation analysis showed that there was a positive association between serum ADA levels and disease activity ($r = 0.43$, $P = 0.02$). Receiver operating characteristic analysis showed that 38.5 U/L was the optimum cut-off point of ADA level for severe disease activity (sensitivity 60%, specificity 92%, area under the curve: 0.81).

Conclusion: The evaluation of serum adenosine deaminase level in patients with autoimmune hepatitis should be considered a useful biomarker in the monitoring of their disease activity.

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AB1112

DIAGNOSTIC DELAY AND DAMAGE OF POLISH ADULT PATIENTS AFFECTED BY HEREDITARY AUTOINFLAMMATORY SYNDROMES

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Background: Autoinflammatory diseases (AIDs) cover a spectrum of diseases, which lead to chronic or recurrent inflammation caused by activation of the innate immune system, typically in the absence of high titre autoantibodies.¹ The most common monogenic AIDs are cryopyrin-associated periodic syndromes (CAPS), familial Mediterranean fever (FMF) and tumour necrosis factor receptor-associated periodic fever syndrome (TRAPS). In hereditary AIDs, chronic and recurrent inflammation can lead to both acute disease and irreversible damage.²

Objectives: To describe irreversible damage and diagnostic delay of AID in Polish adult patients (age ≥ 18).

Methods: We reviewed the records of 11 Caucasian (2 women, 9 men) and one female patient with Middle Eastern ancestry followed in our center from Aug 2012 to Jan 2019. We analysed demographic features, diagnostic delay, previous diagnosis and organ damage.³

Results: Eleven patients had documented variant mutation and AID diagnosis (6-TRAPS, 4-CAPS, 1-FMF). One patient with AA amyloidosis had negative genetic results. The mean age of the patients at the time of disease onset was 9.1 years (0.1- 37) and 33 years (16-62) at diagnosis. In all Caucasian patients diagnosis was established after the age of 18. Previous diagnosis was: JIA (2), AOSD (2), UCTD (4), vasculitis (1), seronegative RA (1), FMF (3 finally confirmed TRAPS) and allergy (3). Prior treatment included GCS (12/12), cDMARDs (7), antimalarials (6), colchicine (4), bDMARDs: tocilizumab-1, etanercept-2, adalimumab-1, CYC (2). After diagnosis 11 patients received IL-1 inhibitor, anakinra. Only one, 62 year old woman with TRAPS, did not need biological treatment. Only 3 TRAPS patients were free from damage accrual. We documented following damage categories: renal/amyloidosis (5 male cases), developmental (3 growth retardation), ear (severe hearing loss -3 CAPS cases). CINCA male patient at age 29 had multiple organ damage: neurological (epilepsy), severe hearing loss, severe ocular involvement (including optic nerve atrophy), joint restriction and deformity leading to complete physical disability. One FMF woman was advised elective abortion, one MWS had miscarriage. Two of 3 women had history of malignancy (one: breast cancer and ovarian cancer, and another thyroid cancer). No any mental

retardation occurred. During follow up 2 male patients with extensive amyloidosis died despite anakinra treatment.

Conclusion: AID are misdiagnosed and have high risk of damage. We advocate the need for efforts in order to increase awareness of AID to reduce the diagnostic delay. The frequency of malignancy in AID should be addressed in future international collaborative studies.

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Diagnostics and imaging procedures

AB1113

RELATION BETWEEN RESTRICTIVE PULMONARY FUNCTION TESTS AND ULTRASONOGRAPHIC CHANGES OF ASYMPTOMATIC ANTERIOR CHEST WALL JOINTS IN RHEUMATOID ARTHRITIS PATIENTS

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Background: Ultrasonography can detect different changes in anterior chest wall (ACW) joints in patients with Rheumatoid Arthritis (RA) even before being clinically manifested.⁽¹⁾ Pulmonary functions may be affected during the course of RA due to interstitial lung problem or chest wall problem.⁽²⁾

All studies usually demonstrate the interstitial pulmonary affection in RA but the chest wall joints is usually underestimated by the rheumatology community. Up to the best of our knowledge, there are no previous studies about the relationship between ultrasound detected subclinical changes in ACW and the pulmonary functions in RA patients.

Objectives: To detect the relation between ultrasonographic changes of asymptomatic ACW joints and pulmonary function tests (PFTs) in patients with RA.

Methods: The study included 44 subjects (22 RA and 22 control) in whom 88 sternoclavicular joints (SCJ) and 44 manubriosternal joints (MSJ) were studied. None of the participants had a history of respiratory complaints such as dyspnea, chronic cough, or chest pain. High resolution Computed Tomography (HRCT) was done on the chest to exclude interstitial lung problem that may affect chest expansion and PFTs. Ultrasound (US) assessments were performed to detect synovitis, erosions, ankylosis, osteophytes, or Doppler signals. Chest expansion was measured. PFTs was done and included measurement of the forced expiratory volume in 1 s (FEV1), forced vital capacity (FVC), and the ratio of forced expiratory volume in 1 s to the forced vital capacity (FEV1/FVC). In RA group, DAS28 and Health Assessment Questionnaire Disability Index (HAQDI) were recorded.

Results: US detected subclinical changes of ACW joints in (74.2%) of RA patient with significant difference between total US changes in RA (74.2%) and control (21.2%) ($p < 0.001$). MSJ ankylosing and erosions were highly associated with limited chest expansion in RA group ($P < 0.001$). PFTs were found to be restrictive in 13 RA patient (59.1%) with mean of FVC (65.5±5.5)%, FEV1 (70.4±9.1)%, FEV1/FVC (80.1±2.6) with significant difference to control with mean of FVC (93.6±7.2)%, FEV1 (94.3±4.5)%, FEV1/FVC (95.1±2.4). These restrictive PFTs were associated with SCJ synovitis ($p = 0.04$), SCJ PD activity ($p = 0.04$), SCJ erosions (0.02) and highly associated with MSJ ankylosing and erosions ($p < 0.001$). All RA patients (100%) with MS ankylosing and erosions and SCJ PD activity by US had limited chest expansion and restrictive PFTs. Restrictive PFTs were associated also with limited chest expansion with mean of (2.3±0.5) with significant difference ($p < 0.001$) to non-restrictive PFTs in RA with mean of (5.2±1.4).

In RA group, ultrasonographic changes and restrictive PFTs were found to be higher with smoking, longer disease duration and high DAS28.

Conclusion: Our study demonstrated that ultrasonographic subclinical changes in ACW joints is associated with restrictive pattern of PFTs and limited chest expansion in RA patients.