Background: Deficiency of adenosine deaminase 2 (DADA2) is a monogenic autoinflammatory disease whose pathogenesis has not been clearly elucidated. **Objectives:** To investigate the role of vascular inflammatory factors in the pathogenesis of DADA2, to compare the vascular inflammation profiles of DADA2 patients with different phenotypes, and to compare DADA2 patients with classic polyarteritis nodosa (PAN).

Methods: The study included eighteen DADA2 patients, ten PAN patients, and eight healthy controls. Plasma levels of sST2, sRAGE, Tie-2, sCD40L, Tie-1, sFlt-1, LIGHT, TNF- α , PIGF, IL-6, IL-18, IL-10, MCP-1 were studied by cytometric bead-based multiplex assay panel.

Results: Among the DADA2 patients, five had hematological manifestations, 13 had vasculitic findings, and accompanying immunological findings were present in seven patients. Nine patients had neurological findings, five of whom had neuropathy. Hematological findings were Diamond-Blackfan anemia-like phenotype in four patients and bicytopenia (anemia and thrombocytopenia) in one patient. Disease characteristics of DADA2 and PAN patients revealed that neurological involvement and livedo reticularis were more frequent in DADA2 patients ($p=0.034$ and $p=0.009$, respectively), while myalgia was more common in PAN patients ($p=0.001$).

Plasma levels of Tie-1 and sFlt-1 were higher in the overall DADA2 patients compared to healthy controls and PAN patients ($p<0.001$ and $p=0.004$, respectively). DADA2 patients with PAN-like features had higher sRAGE, Tie-2, and TNF- α levels compared to PAN patients ($p=0.013$, $p=0.003$, and $p=0.001$, respectively). There was no significant difference in the levels of vascular inflammation markers between DADA2 patients with vasculitis and hematological involvement except IL-18. The plasma IL-18 levels were higher in the DADA2 patients with hematological findings compared to vasculitic phenotype ($p=0.001$). Finally DADA2 patients with neuropathy had higher sRAGE concentrations than patients without neuropathy and healthy controls ($p=0.03$ and $p=0.008$, respectively).

Conclusion: We suggest that the high plasma IL-18 levels may be associated with an activated IFN pathway, the pathogenesis of hematologic manifestations, and unresponsive to anti-TNF treatment. Higher concentrations of Tie-1, Tie-2, sFlt-1, sRAGE, and TNF- α distinguished DADA2 patients with PAN-like features from PAN patients. We identified sRAGE as a potential biomarker of neuropathy in DADA2 patients.

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Challenges for working people with RMDs in pandemic times

OP0169-PARE

DEVELOPMENT OF POINTS TO CONSIDER WHEN SUPPORTING PERSONS WITH RHEUMATIC AND MUSCULOSKELETAL DISEASES TO PARTICIPATE IN HEALTHY AND SUSTAINABLE PAID WORK

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Background: Despite earlier diagnosis and improved management of rheumatic and musculoskeletal diseases (RMDs), a work participation gap remains when compared to the general population. To bridge this work participation gap, EULAR's current strategy states that 'by 2023, EULAR's activities and related advocacy will have increased participation in work by people with RMDs'. To achieve this goal, guidance is needed how to support people with RMDs to remain in paid work or (re)-enter the labour force.

Objectives: To develop Points to Consider (PtC) when supporting people with RMD to participate in healthy and sustainable paid work.

Methods: An international expert group, established by Eular in 2019, convened twice to agree on a protocol for the development of PtC. EULAR's standard

operating procedures were followed. The group (a) defined the target audience, (b) identified areas from which knowledge should be derived to enable formulation of the PtC, (c) agreed on a strategy to collect evidence, (d) established an international taskforce to formulate and agree on the PtC, and (e) proposed an implementation plan.

Results: The target audience are professionals involved in clinical care for patients with RMDs as well as their organisations, persons with RMDs and their organisations, and administrators responsible for healthcare and work policies. Six knowledge areas are identified (Table). Depending on the specific knowledge area, strategies for collecting evidence comprise synthesis of published and grey literature, surveys among various organisations and collection of case studies from employers. Whenever evidence in RMDs is limited, evidence from other chronic diseases will be sought. The international taskforce includes experts from different disciplines in rheumatology (one in other chronic diseases), EU policy makers, and representatives of PARE, HCP, EULAR Public Affairs and EMEUNET. As part of the implementation, close cooperation with national professional and patients societies is planned. Additionally, results will be included in EULAR's activities at the EU policy level.

Table 1. Knowledge areas and strategy to retrieve evidence

	In persons with RMDs:	Sources of evidence
1	Is work relevant for the clinical outcome of disease	LR; Review of management recommendations and Care Standards
2	What are barriers and facilitators to enter or stay in the labor force	SLR
3	What is the effectiveness of interventions to enter or stay in the labor force	SLR; Survey among professional and patient organisations
4	Which social security systems are more effective when entering or staying in the labor force	LR
5	How does disease influence the cycle of work	SLR; Grey literature
6	What (not) to do by employers to let patients enter or stay in the work force	SLR; Survey among large companies; case studies

LR: Literature review; SLR: Systematic literature review

Conclusion: The proposed initiative to develop PtC should ultimately result in improvement of healthy and sustainable labor force participation of people with RMDs.

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Progress in myositis and scleroderma research - I

OP0170

DECLINE IN FORCED VITAL CAPACITY (FVC) IN PATIENTS WITH SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE (SSC-ILD) WITH AND WITHOUT DYSPNOEA: DATA FROM THE SENSICIS TRIAL

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Background: Some patients with SSC-ILD develop dyspnoea secondary to parenchymal lung disease, while others do not report dyspnoea even when their