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EULAR Highlights 2025
Inflammatory Myopathies

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COI

Research: AbbVie, Protagen, Novartis Biomedical; patent mir-29 for the treatment of systemic sclerosis issued (US8247389, EP2331143)

Lecturing: Boehringer-Ingelheim, GSK, Novartis, Otsuka

Consulting: Novartis, Boehringer Ingelheim, Janssen-Cilag, GSK

Congress support: Medtalk, Pfizer, Roche, Actelion, Mepha, MSD

Advisory Boards: Boehringer-Ingelheim, Janssen-Cilag

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Content

- (Co-)morbidity
- Treatment
  - IVIG
  - Novel monoclonal antibody
  - CAR T cells
  - Bispecific T Cell Engagers

Inflammatory Myopathies
Dermatomyositis
(Polymyositis)
Antisynthetase Syndrome
Immune-mediated necrotizing Myopathy
Cancer-associated Myositis
Overlap-Myositis (e.g. with systemic sclerosis, rheumatoid arthritis)
Secondary myositis (connective tissue disease, vasculitis, sarcoidosis)

Sporadic inclusion body myositis

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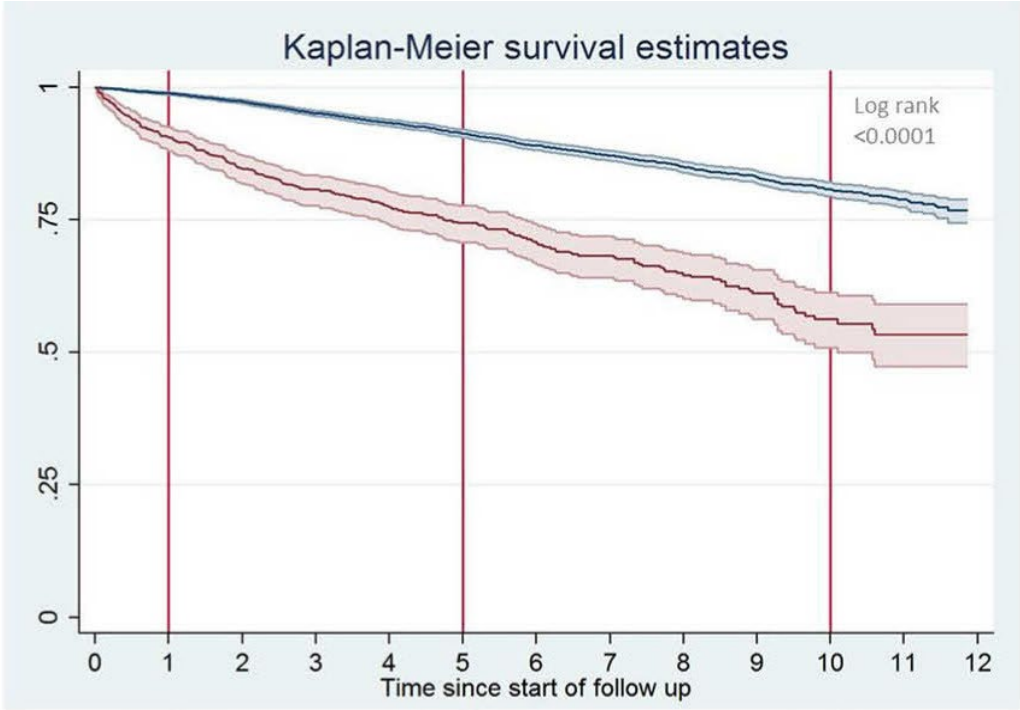
(Co-)Morbidity

Morbidity and Mortality of IIM



At risk, n	716/7100	648/7013	327/4037
Dead at end of interval, n	66/77	100/448	53/318
Cumulative mortality, %	9/1	23/7	31/12

- 80% long-term disability
- HR for death 5x increased
- Respiratory diseases, malignancies, cardiovascular diseases primary causes of death
- Survival not substantially changed during the last 20 years

Dobloug GC, et al. ARD;77: 40 - 47

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MYOSITIS PATIENTS IN THE ICU – MORBIDITY BEYOND MUSCLE INVOLVEMENT

Background

- To date, there are only limited data on the outcomes and treatments of IIM patients in the ICU.

Aim

- To analyze the clinical characteristics and outcomes of patients with IIM in the ICU to provide better insight into the management of critically ill patients

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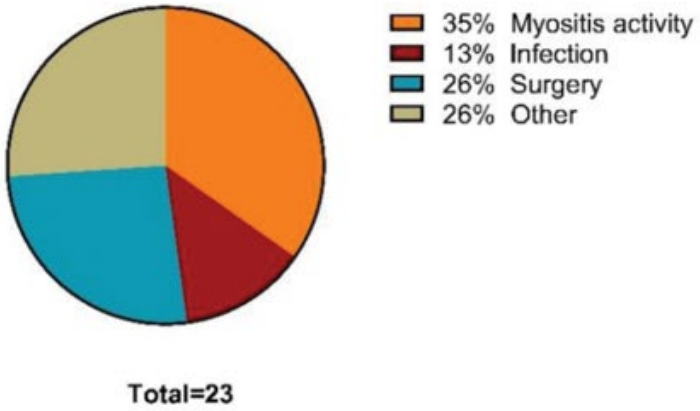
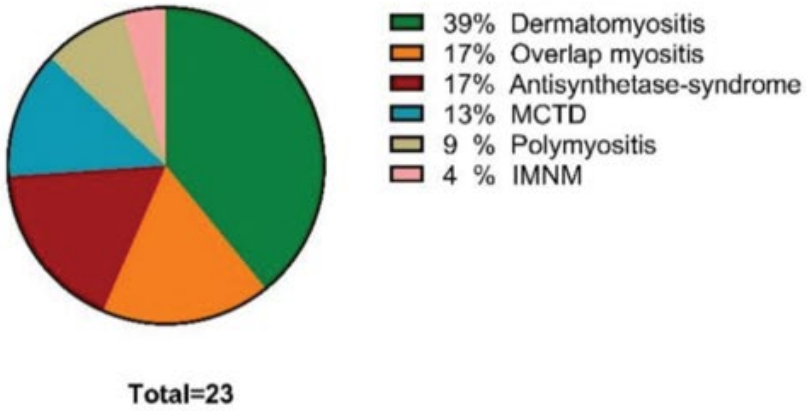
Results I

- Mean age at ICU admission 58 years (range 19-83 years), mean duration of ICU treatment 32 days (range 1-503 days), 5 patients died in the ICU,14 patients were intubated, 21 were under immunosuppression

Reason of ICU admission



Myositis subtype



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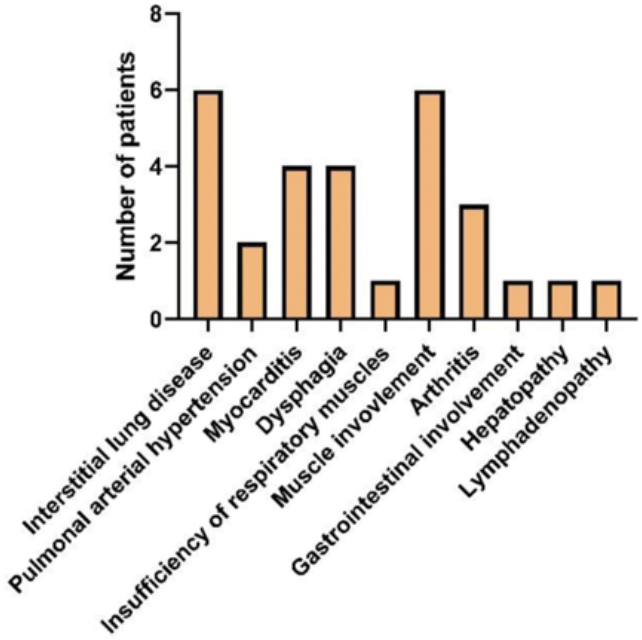
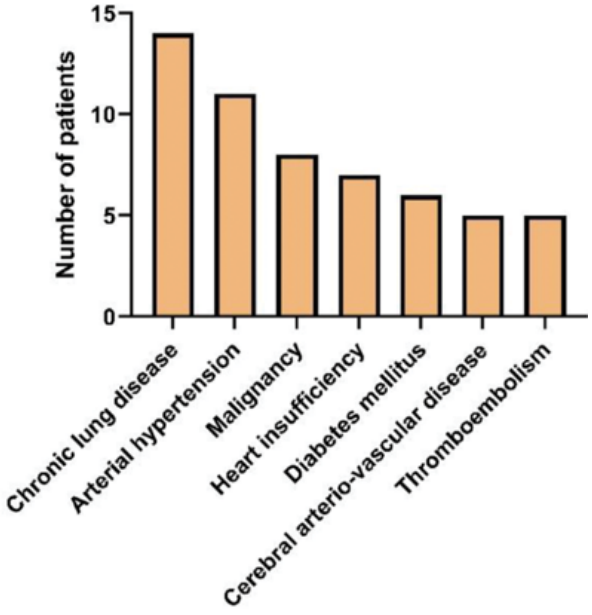
Results II

Active organ involvement



Most common co-morbidities



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MYOSITIS PATIENTS IN THE ICU – MORBIDITY BEYOND MUSCLE INVOLVEMENT

Conclusions

- Active disease (e.g. ILD or cardiac involvement) and infectious complications can lead to severe morbidity with a potentially life-threatening course.
- There is a need for interdisciplinary treatment in the ICU setting.

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BONE HEALTH DETERMINANTS IN INFLAMMATORY MYOPATHIES

Background

- Use of high doses of glucocorticoid (GC) for prolonged periods, muscle weakness and reduced mobility compromise bone health in IIM.

Aim

- To assess the prevalence and risk factors for fragility fractures (FFx) in a monocentric cohort of IIMs patients in a third level rheumatology center

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BONE HEALTH DETERMINANTS IN INFLAMMATORY MYOPATHIES

Results

- 191 patients (125 female, 65.4%), mean age of 67.3 ± 13.5 years, mean disease duration of 10.4 ± 7.4 years
- Osteoporosis (OP) occurred in 70 patients (36.6%; 64 females, mean age 72.3 years)
- FFx occurred in 29/70 patients (15.2% of the total cohort; 25 females, mean age 72.5 years)
- Age, female sex and disease duration were risk factors for both OP and FFx ($p<0.01$); no association found for GC dose
- OP associated with higher HAQ and lower SF-36 values in both physical and emotional domains ($p\leq0.04$)

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BONE HEALTH DETERMINANTS IN INFLAMMATORY MYOPATHIES

Conclusions

- OP occurred in 30% of the enrolled IIM patients and > 40% of them developed at least one FFx
- OP substantially adds to the overall disease burden – physically and emotionally
- IIM patients should be carefully screened for individualized management strategies

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REDUCING CARDIOVASCULAR RISK IN MYOSITIS: FOCUS ON LIPID-LOWERING THERAPY

Background

- Statin therapy reduces CVD risk, but its use in IIM is limited due to potential muscular adverse events.
- International recommendations for CV risk reduction are lacking.

Aims

- To assess CVD risk in a myositis cohort using the SCORE-2 prediction system, carotid artery Doppler ultrasound measurement and biomarkers
- To recommend individual lipid-lowering treatment with 6 months follow-up regarding efficacy and adverse events

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REDUCING CARDIOVASCULAR RISK IN MYOSITIS: FOCUS ON LIPID-LOWERING THERAPY

Results

- 80 patients, mean age of 56.2 ±13.4 years, disease duration of 9 (5-15) years
- SCORE-2: 78.8% medium/high CVD risk; 73.13% asymptomatic carotid plaque; risk factors: hypertension (71.3%), diabetes (25%)
- After 6 months
  - Cholesterol levels significantly lower
  - No progression of carotid plaques
  - 37.5% of patients shifted into a lower SCORE-2 risk category
  - No negative side effects

Conclusions

- Not to neglect assessment and management of CVRF in IIM

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Treatment

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PHASE 2 RESULTS FROM THE ALKIVIA STUDY

Background

- Efgartigimod = IgG1 antibody Fc fragment blocking the neonatal Fc receptor (FcRn) without impacting antibody production, albumin levels, or other parts of the immune system

Aim

- To evaluate the safety and efficacy of subcutaneous (SC) efgartigimod (coformulated with recombinant human hyaluronidase PH20) compared with placebo in patients with IIM receiving standard of care (SOC) medications

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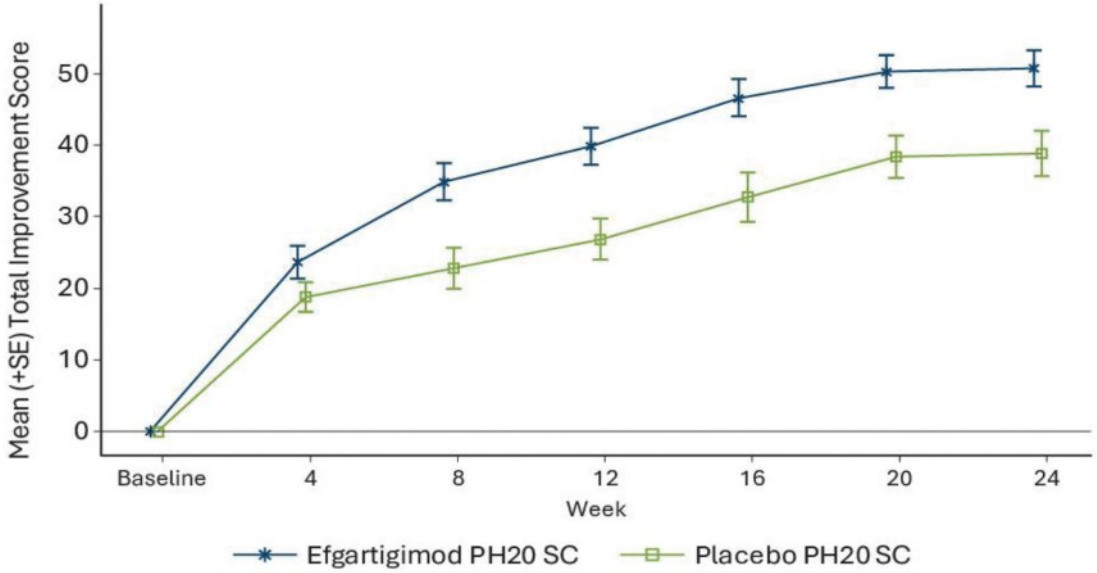
Results

Figure 1A. Mean (±SE) TIS over time



n (mean):						
Efgartigimod PH20 SC: 47	44 (23.64)	45 (35.00)	44 (39.83)	45 (46.72)	42 (50.24)	41 (50.67)
Placebo PH20 SC: 42	42 (18.81)	40 (22.81)	37 (26.89)	36 (32.71)	36 (38.33)	36 (38.96)

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PHASE 2 RESULTS FROM THE ALKIVIA STUDY

Conclusions

- Efgartigimod PH20 SC led to significant improvement over placebo in TIS and key secondary endpoints with good safety and tolerability.
- The results demonstrate the mechanistic relevance of FcRn inhibition in IIM, indicating the potential pathogenicity of autoantibodies in IIM.
- These findings support further evaluation of efgartigimod PH20 SC in IIM in the ongoing phase 3 part of the study.

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EFFICACY OF IVIG ON PULMONARY MANIFESTATIONS IN DERMATOMYOSITIS

Background

- ProDERM study was a randomized Phase 3 study designed to evaluate the efficacy and safety of IVIG in adult DM patients

Aim

- Post-hoc analysis of the ProDERM study data to assess the effect of IVIg on pulmonary symptoms of DM

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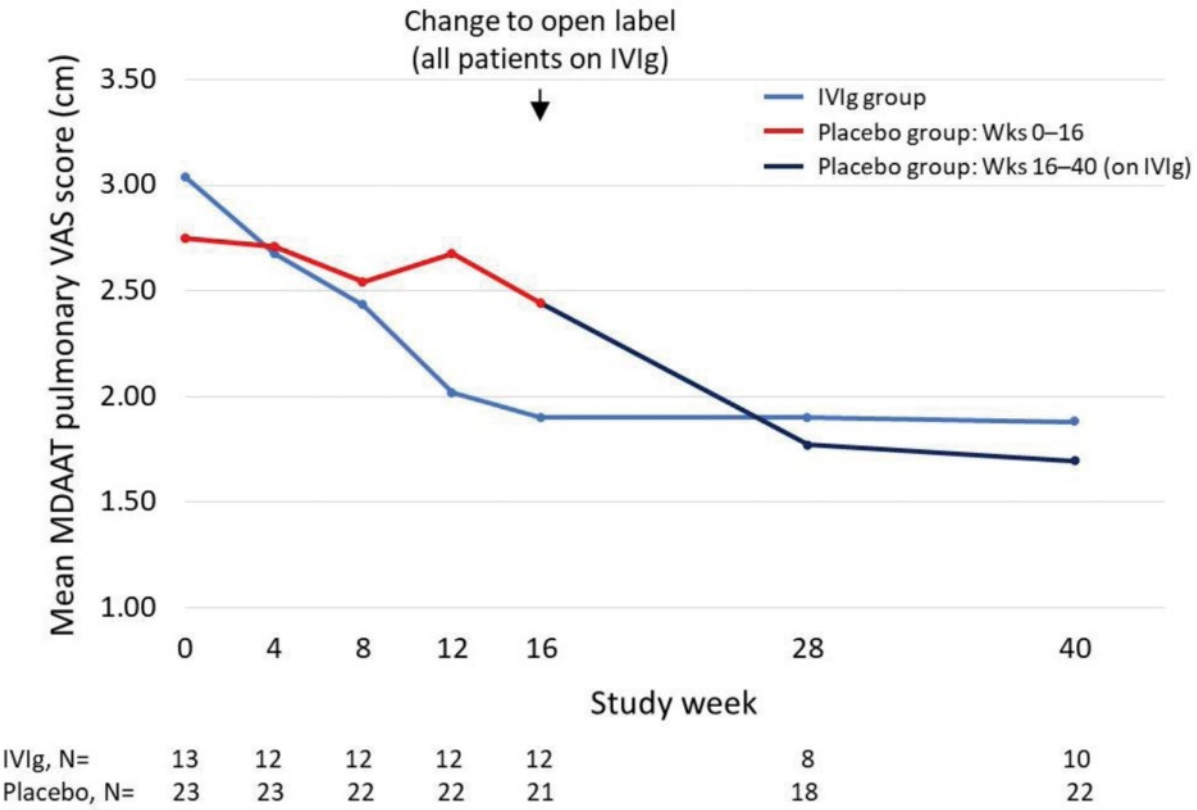
Results



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EFFICACY OF IVIG ON PULMONARY MANIFESTATIONS IN DERMATOMYOSITIS

Conclusions

- IVIg may have favourable treatment effects on active pulmonary manifestations of DM.
- Future studies should consider including pulmonary outcomes to assess the effect of treatment on these symptoms.

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Upcoming Therapies I

CAR T Cells

- RESECABTAGENE AUTOLEUCEL, A FULLY HUMAN, AUTOLOGOUS 4-1BB ANTI-CD19 CAR T CELL THERAPY; DOI: annrheumdis-2025-eular.B1119
- SAFETY AND PRELIMINARY EFFICACY OF CD19 CAR T-CELL TREATMENT IN RHEUMATIC DISEASE – DATA FROM THE PHASE I/II CASTLE BASKET STUDY; DOI: annrheumdis-2025-eular.B1038
- SUCCESSFUL BCMA-CAR T-CELL SALVAGE THERAPY IN A PATIENT WITH IDIOPATHIC INFLAMMATORY MYOSITIS RELAPSING AFTER CD19-CAR T-CELL THERAPY; DOI: annrheumdis-2025-eular.E504

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Upcoming Therapies II

Bispecific Antibodies

- SAFETY AND EFFICACY OF T CELL ENGAGER THERAPY IN PATIENTS WITH REFRACTORY AUTOIMMUNE DISEASE; DOI: annrheumdis-2025-eular.B1336
- INDUCTION OF GC-FREE REMISSION BY THE BISPECIFIC CD19xCD3 T CELL ENGAGER BLINATUMOMAB IN PATIENTS WITH SEVERE, THERAPY-REFRACTORY ANTI-SYNTHETASE SYNDROME; DOI: annrheumdis-2025-eular.E688

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Thank you for your attention!

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