Table 1. Autoantibody-defined subgroups using an unsupervised cluster analysis.

	Subgroups/ Medoids								
Variables	**1 Ro52**	**2 U1RNP**	**3 PMScl**	**4 Mi2**	**5 Jo1**	**6 Jo1/Ro52**	**7 TIF1**	**8 None***	**All**
n (%)	137 (10)	183 (14)	107 (8)	65 (5)	119 (9)	140 (10)	78 (6)	519 (39)	1348 (100)
Female (%)	93 (68)	116 (63)	79 (74)	45 (69)	76 (64)	96 (69)	64 (82)	313 (60)	882 (65)
Age at diagnosis, median (IQR)	56 (16)	51.5 (23)	51 (25)	57 (22.5)	47.5 (23.25)	52 (19.5)	53.5 (21.75)	58 (22)	55 (23)
Autoantibodies									
Anti-Jo1	0	6 (3)	0	1 (2)	119 (100)	140 (100)	0	0	266 (20)
Anti-PL7	7 (5)	13 (7)	0	0	0	0	0	0	20 (1.5)
Anti-PL12	5 (4)	3 (2)	1 (1)	0	1 (1)	0	0	0	10 (0.7)
Anti-EJ	2 (2)	0	0	0	0	0	0	0	2 (0.1)
Anti-OJ	0	7 (4)	0	0	0	0	0	0	7 (0.5)
Anti-TIF1	10 (7)	2 (1)	2 (2)	0	0	0	78 (100)	0	92 (7)
Anti-Mi2	1 (1)	1 (1)	1 (1)	65 (100)	0	2 (1)	0	0	70 (5)
Anti-SAE1	8 (6)	23 (13)	0	0	0	0	0	0	31 (2)
Anti-NXP2	1 (1)	23 (13)	1 (1)	0	0	0	0	0	25 (2)
Anti-MDA5	9 (7)	10 (6)	1 (1)	1 (2)	0	1 (1)	0	0	22 (2)
Anti-SRP	8 (6)	32 (18)	0	0	0	0	0	0	40 (3)
Anti-Ro52	137 (100)	16 (9)	0	0	0	140 (100)	0	0	293 (22)
Anti-PMScl	11 (8)	1 (1)	107 (100)	0	0	0	0	0	119 (9)
Anti-U1RNP	0	79 (43)	0	0	0	3 (2)	0	0	82 (6)

*IIM patients negative for the tested autoantibodies.

Results: Eight subgroups were defined by the autoantibody status (Table 1). Three of the subgroups (1, 2 and 6) have overlapping autoantibodies, while four are almost monospecific (3,4,5 and 7), and one (8) has patients negative for tested autoantibodies. Figure 1 represents the significant associations between *HLA-DRB1* alleles and the eight subgroups. Heliotrope rash and Gottron's sign were significantly more frequent in subgroups 3 (OR:2.2 95%CI:[1.1-4.8], OR:2.6 95%CI:[1.3-5.9], respectively), 4 (OR:12 95%CI:[3.6-75], OR:7.8 95%CI:[2.8-33], respectively) and 7 (OR:22 95%CI:[4.5-385], OR:10 95%CI:[3.1-65], respectively), and Raynaud's phenomenon was significantly more frequent in subgroup 6 (OR:3.3 95%CI:[1.2-11]).

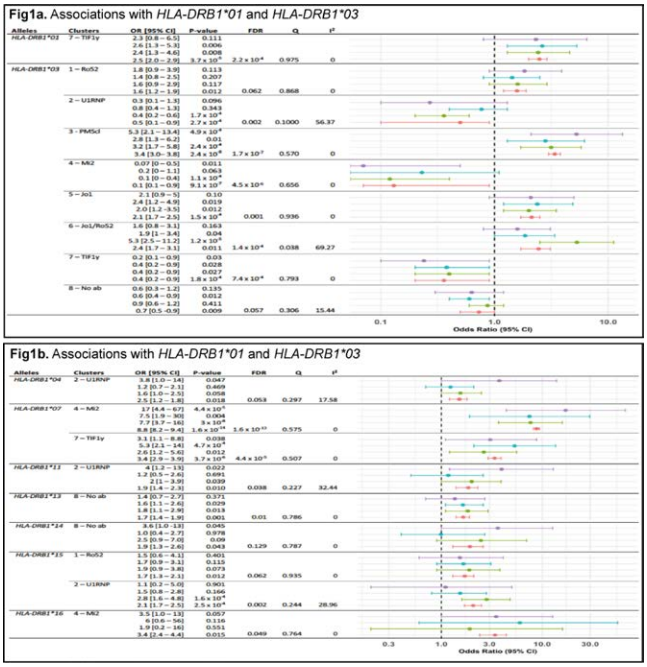


Figure 1. Forest plot of significant associations of *HLA. *DRB1* alleles with autoantibody-defined subgroups. Scandinavia includes patients from Denmark, Norway, and Sweden.

Conclusion: Our study reveals that certain subgroups of IIM patients are characterized by overlap of myositis -specific and -associated autoantibodies, which in turn are associated with different *HLA-DRB1* alleles including potential novel associations. These results point to different disease mechanisms in the subgroups, as well as suggest that IIM classification could be improved by integrating broader serological and genetic data.

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OP0161

TREATMENT PATTERNS OF IDIOPATHIC INFLAMMATORY MYOPATHIES: RESULTS FROM AN INTERNATIONAL COHORT OF OVER 1,400 PATIENTS

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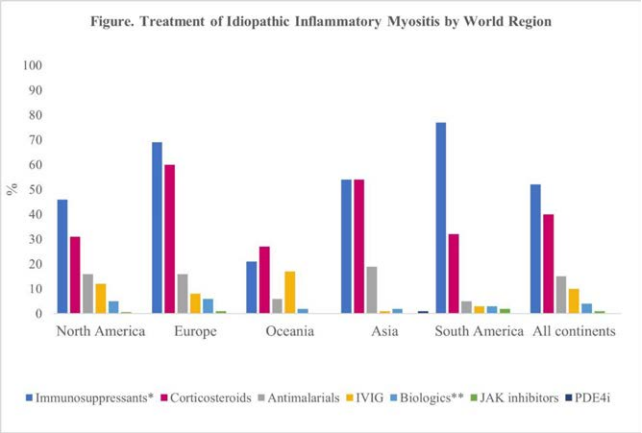
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Background: Idiopathic inflammatory myopathies (IIM) are a group of heterogeneous autoimmune disorders with limited standardization of treatment protocols.
Objectives: To evaluate frequency and patterns of various treatments used for IIM based on disease subtype, world region, and organ involvement.
Methods: Cross-sectional data from the international CoVAD self-report e-survey[1] was extracted on Sep 14th, 2021. Patient details included demographics, IIM subtypes (dermatomyositis (DM), polymyositis (PM), inclusion body myositis (IBM), antisynthetase syndrome (ASSD), necrotizing myositis (NM) and overlap myositis (OM)), clinical symptoms, disease duration and activity, and current treatments. Treatments were categorized in corticosteroids (CS), antimalarials, immunosuppressants (IS), intravenous immunoglobulins (IVIG), biologics, and others. Typical clinical symptoms (dyspnea, dysphagia) were used as surrogate for organ involvement. Factors associated with IS were analyzed using multivariable logistic regression, adjusting for IIM subtype, demographics, world region, disease activity, and prevalent clinical symptoms (>10%).
Results: In 1418 patients with IIM, median age was 61 years [IQR 49-70], 62.5% were females, median disease duration was 6 years [IQR 3-11], most common subset was DM (32.4%).
The most used treatments were IS (49.4%, including Methotrexate 19.6%, Mycophenolate Mofetil 18.2%, Azathioprine 8.8%, Cyclosporine 2.7%, Tacrolimus 2%, Leflunomide 1.6%, Sulfasalazine 1%, and Cyclophosphamide 0.6%), followed by CS (40.8%), antimalarials (13.8%) and IVIG (9.4%). Biologics were used in 4.3% of patients.
Treatment patterns differed significantly by IIM subtypes with a higher frequency of IS (77.7%) and CS (63.4%) use in ASSD; antimalarials (28.6%) and biologics (9.8%) use in OM and IVIG use in NM (24.6%) (Table 1). Also, treatment patterns were different in regions of the world (Figure 1), with a higher frequency of CS use in Europe (60.5%) and IS use in South America (77.2%). Antimalarials were most used in Asia (19.4%), while IVIG use was most common in Oceania (16.9%). Dyspnea was associated with higher use of IS (69.9%) and CS (65.8%) (p<0.001), whereas dysphagia was negatively associated with IS (39.7%) and CS (32.7%) likely due to a higher proportion in IBM patients reporting dysphagia.
Multivariable logistic regression analysis showed an association of IS with the IIM subtype (least used in IBM (OR 0.07 [95%CI 0.04-0.13] compared to DM), world region (most used in South America (OR 2.35 [1.12-4.91] compared to North America), active and worsening disease activity (OR 3.49 [1.76-6.91] compared to remission), and some clinical features (dyspnea, fatigue, and muscle weakness).
Conclusion: IIM treatment patterns differ significantly by disease subtypes, world regions and organ involvement, highlighting the need for unified international consensus-driven guidelines.

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OP0162 EFFICACY AND SAFETY OF LENABASUM IN THE PHASE 3 DETERMINE TRIAL IN DERMATOMYOSITIS

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Background: Safe and effective treatments are of significant unmet need in DM. Lenabasum, a CB2 agonist that activates resolution of inflammation, improved skin disease, patient-reported outcomes, and biomarkers in a Phase 2 study of DM patients with active skin disease.
Objectives: To evaluate the efficacy and safety of lenabasum in a Phase 3 double-blind study in DM.
Methods: DM patients ≥ 18 years old with active skin with or without muscle involvement were enrolled in 55 sites in North America, Europe, and Asia-Pacific. Stable doses of background immunosuppressants were allowed. Subjects were randomized 2:1:2 to lenabasum 20 mg BID, lenabasum 5 mg BID, or placebo BID for 52 weeks, with visits ≤ 8 weeks apart. The study was stopped after all subjects completed Week 28. Some subjects had completed Week 52 by then. The primary efficacy endpoint was Total Improvement Score (TIS) at Week 28 and a secondary efficacy endpoint was TIS at Week 52, for lenabasum 20 mg BID vs placebo.
Results: 175 subjects (69 lenabasum 20 mg BID, 35 lenabasum 5 mg BID, 71 placebo BID) received study drug; 167 completed Week 28, and 103 completed Week 52. The most common reasons for study discontinuation were study stopped by Sponsor (34.3%), withdrawal of consent (4.5%), and adverse events (AEs, 3.9%), with similar rates among groups. Baseline demographics and disease measurements were similar among groups and in total subjects were (mean or %): age 52.0 years; 81.1% female, 75.4% White; MMT-8 133.3; CDASI activity score 23.4; HAQ-DI 0.84, MDGA 5.55, EMGA 5.23; and PtGA

Table 1. Current Treatments for IIM, Stratified by Disease Subtypes

	Dermatomyositis	Polymyositis	Inclusion Body Myositis	Anti-synthetase syndrome	Necrotizing myositis	Overlap syndrome	All IIM	p-value
Number of patients	**459**	**182**	**348**	**148**	**57**	**224**	**1418**	
Immunosuppressants*	269 (58.6)	107 (58.8)	39 (11.2)	115 (77.7)	40 (70.2)	130 (58.0)	700 (49.4)	<0.001
Corticosteroids	208 (48.0)	81 (46.8)	32 (9.7)	90 (63.4)	32 (59.3)	103 (50.0)	546 (40.8)	<0.001
Antimalarials	99 (21.6)	7 (3.8)	0 (0.0)	25 (16.9)	1 (1.8)	64 (28.6)	196 (13.8)	<0.001
Intravenous Immunoglobulins	54 (11.8)	16 (8.8)	19 (5.5)	10 (6.8)	14 (24.6)	20 (8.9)	133 (9.4)	<0.001
Biologics**	17 (3.7)	7 (3.8)	0 (0.0)	13 (8.8)	2 (3.5)	22 (9.8)	61 (4.3)	<0.001
Others***	6 (1.3)	0 (0.0)	0 (0.0)	1 (0.7)	0 (0.0)	5 (2,2)	12 (0.8)	0.098

**Methotrexate (278), Mycophenolate Mofetil (258), Azathioprine (125), Cyclosporine (38), Tacrolimus (28), Leflunomide (23), Sulfasalazine (14), Cyclophosphamide (9). **Rituximab (44), Abatacept (5), TNF inhibitors (4), Tocilizumab (3), Belimumab (3), Secukinumab (1). ***JAK(10) and PDE4 inhibitors (2)*

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