EULAR Highlights: Systemische Sklerose

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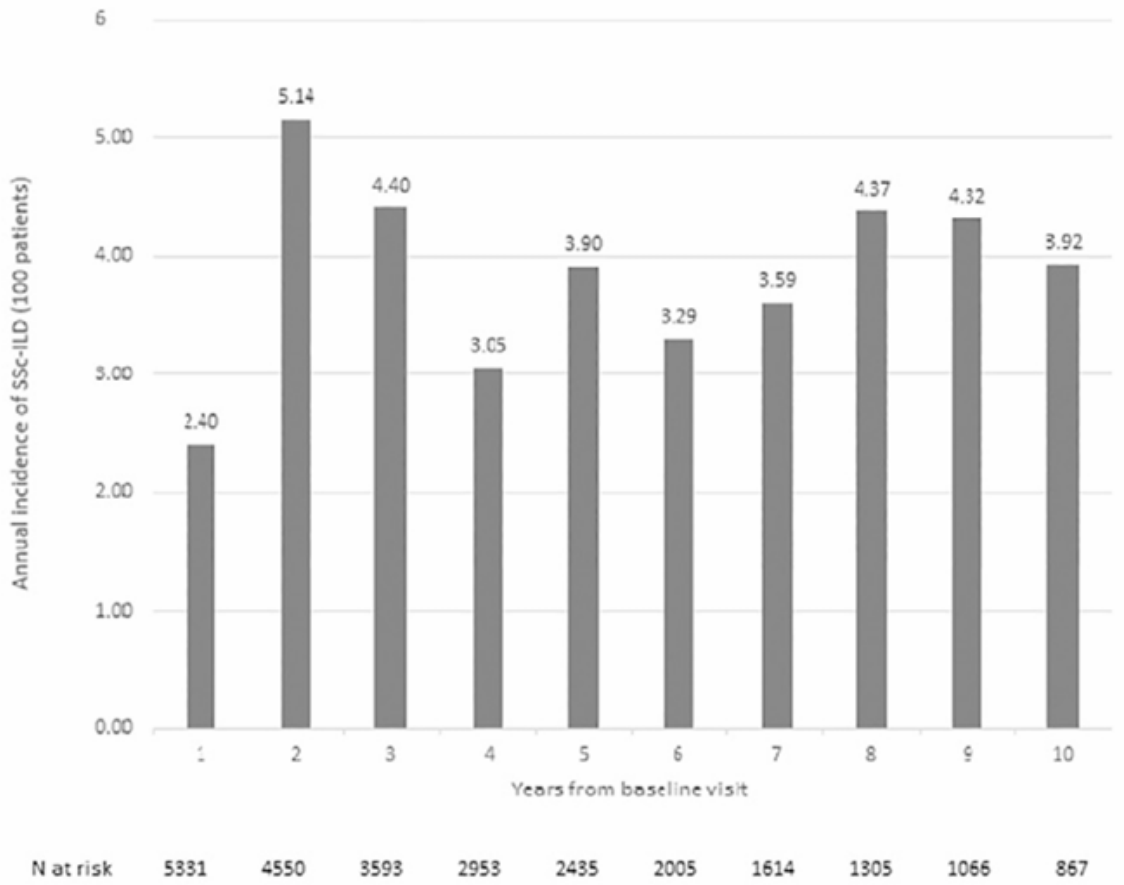
Disclosures

OD has/had consultancy relationship with and/or has received research funding from and/or has served as a speaker for the following companies in the last three calendar years:

4P-Pharma, Abbvie, Acepodia, Aera, Amgen, AnaMar, Anaveon, Argenx, AstraZeneca, Avalyn, Boehringer Ingelheim, BMS, Calluna, Cantargia, CSL Behring, EMD Serono, Galderma, Fimmcyte, Galapagos, Gossamer, Hemetron, Innovaderm, Kali, Lilly, Mediar, MSD Merck, Nkarta, Novartis, Oorja Bio, Orion, Pliant, Prometheus, Quell, Scleroderma Research Foundation, Skyhawk, Tandem, Topadur, UCB and Umlaut.bio.

Patent issued “mir-29 for the treatment of systemic sclerosis” (US8247389, EP2331143). Co-founder of CITUS AG.

Annual incidence rate of new onset of ILD in SSc per 100 patients from baseline EUSTAR visit



Petelytska L et al, Ann Rheum Dis. 2026 Apr;85(4):669-679.

JOURNAL ARTICLE

A screening tool to detect interstitial lung disease in systemic sclerosis: the ILD-RISC score

Cosimo Bruni ✉, Lorenzo Tofani, Håvard Fretheim, Sophie I E Liem, Arthiha Velauthapillai, Hilde Bjørkekjær, Imon Barua, Ilaria Galetti, Alexandru Garaiman, Mike O Becker, Anna-Maria Hoffmann-Vold, Jeska de Vries-Bouwstra, Madelon C Vonk, Jörg H W Distler, Marco Matucci-Cerinic, Oliver Distler

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ILD-RISC score calculator

Enter your data in the Input section, divided per each variable, according to the following instructions.
Maximum one input variable might be missing.

Age: age of the patient expressed in years

DU: Digital ulcers
0 = never
1 = current or previous

SSc antibody positivity (SSc-ATB)
2 = Anti-Centromere (ACA) positive
3 = Anti-Topoisomerase I (ATA) positive
4 = Anti-RNA polymerase III (ARA) positive
5 = isolated PM-Scl positive
0 = none of the above

DLCO/SB%: as continuous value of percent predicted

FVC%: as continuous value of percent predicted

0 < ILD-RISC score < 1

Maria Iacovantuono

Validation of the ILD-RISC score in special populations from the EUSTAR registry: tailored cut-off values for non-Caucasian ethnicities, patients with pulmonary hypertension and very early pre-disease

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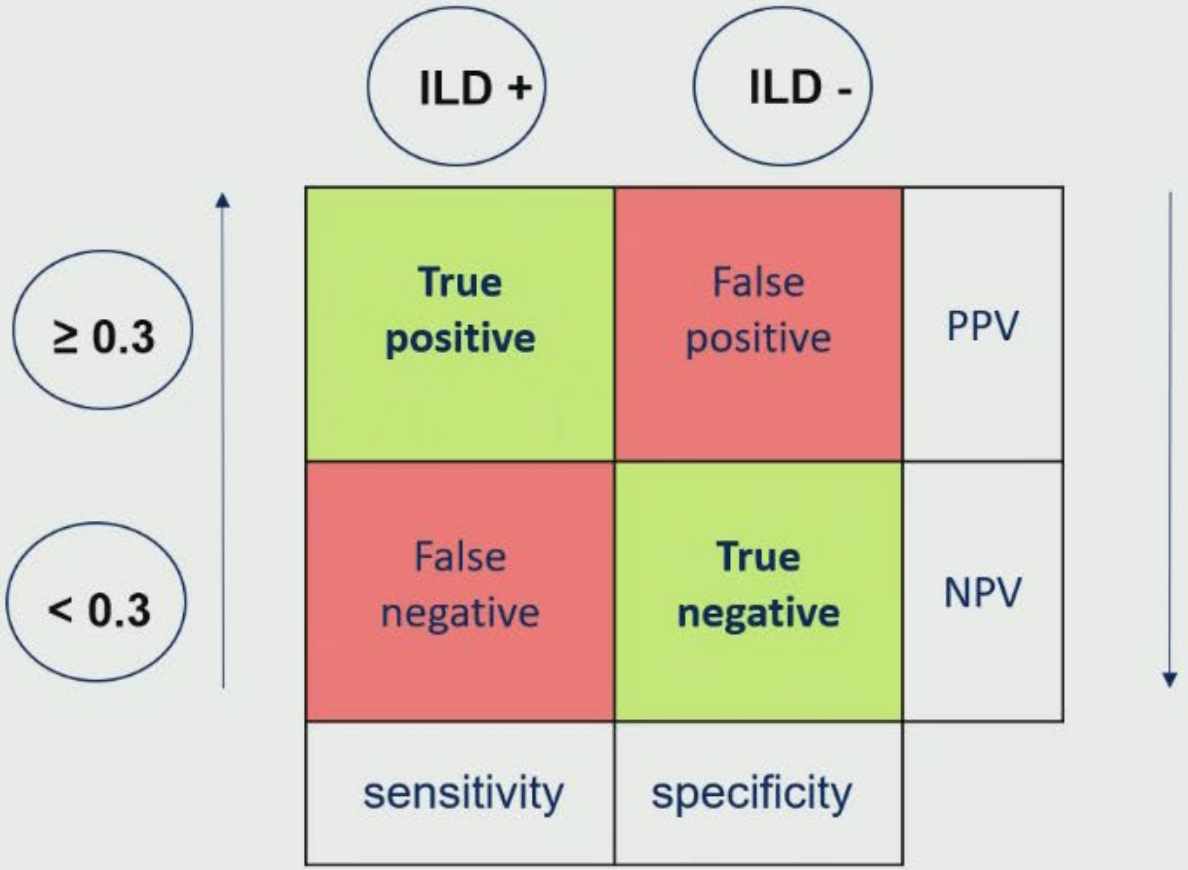
eular 26 EUROPEAN CONGRESS OF RHEUMATOLOGY

SENSITIVITY AND SPECIFICITY



AUC = 0.799

ILD on HRCT -	ILD on HRCT +

ILD-RISC score ≥ 0.3

Sensitivity 85%
Specificity 50%

Validation and longitudinal application

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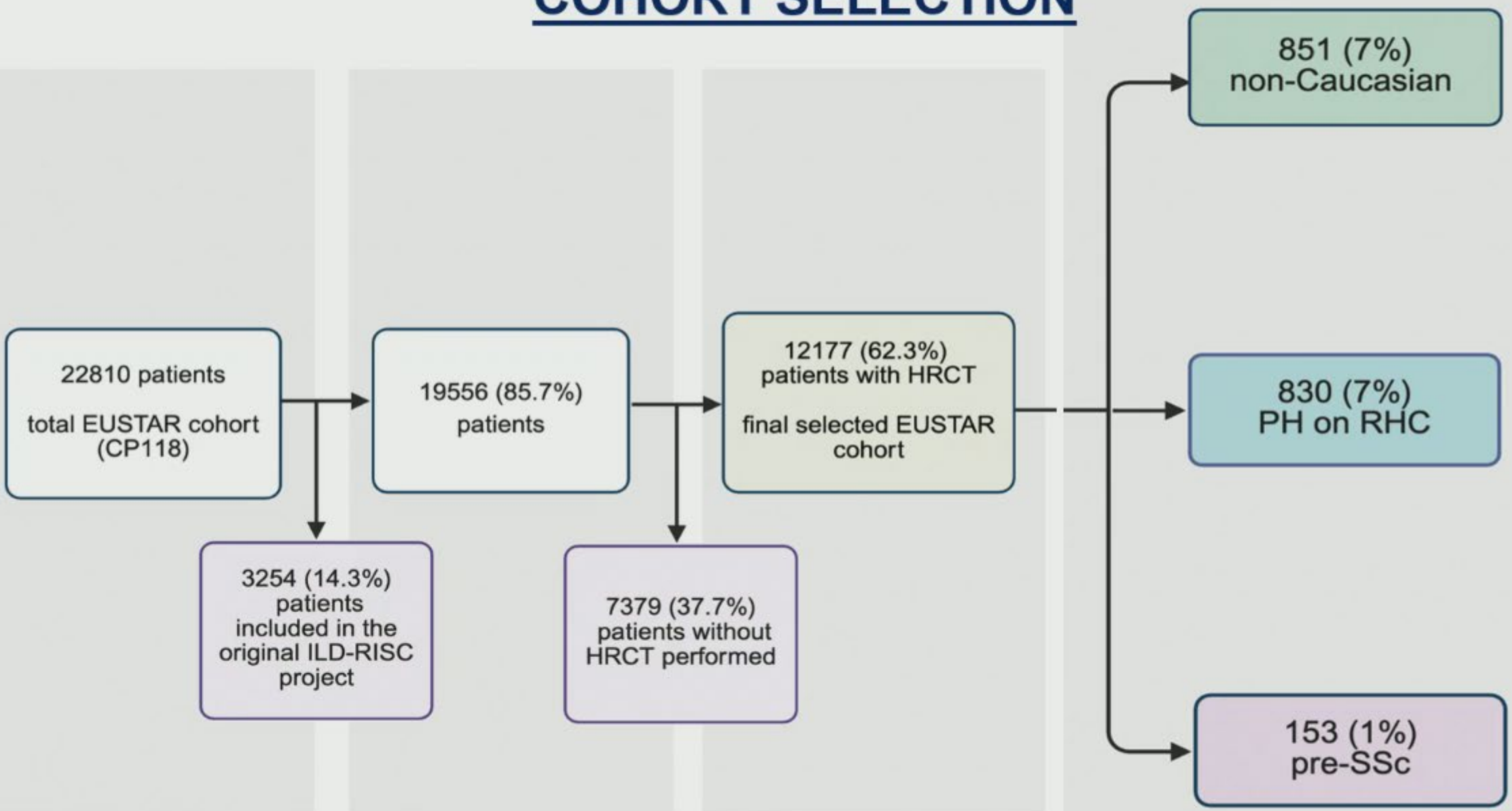
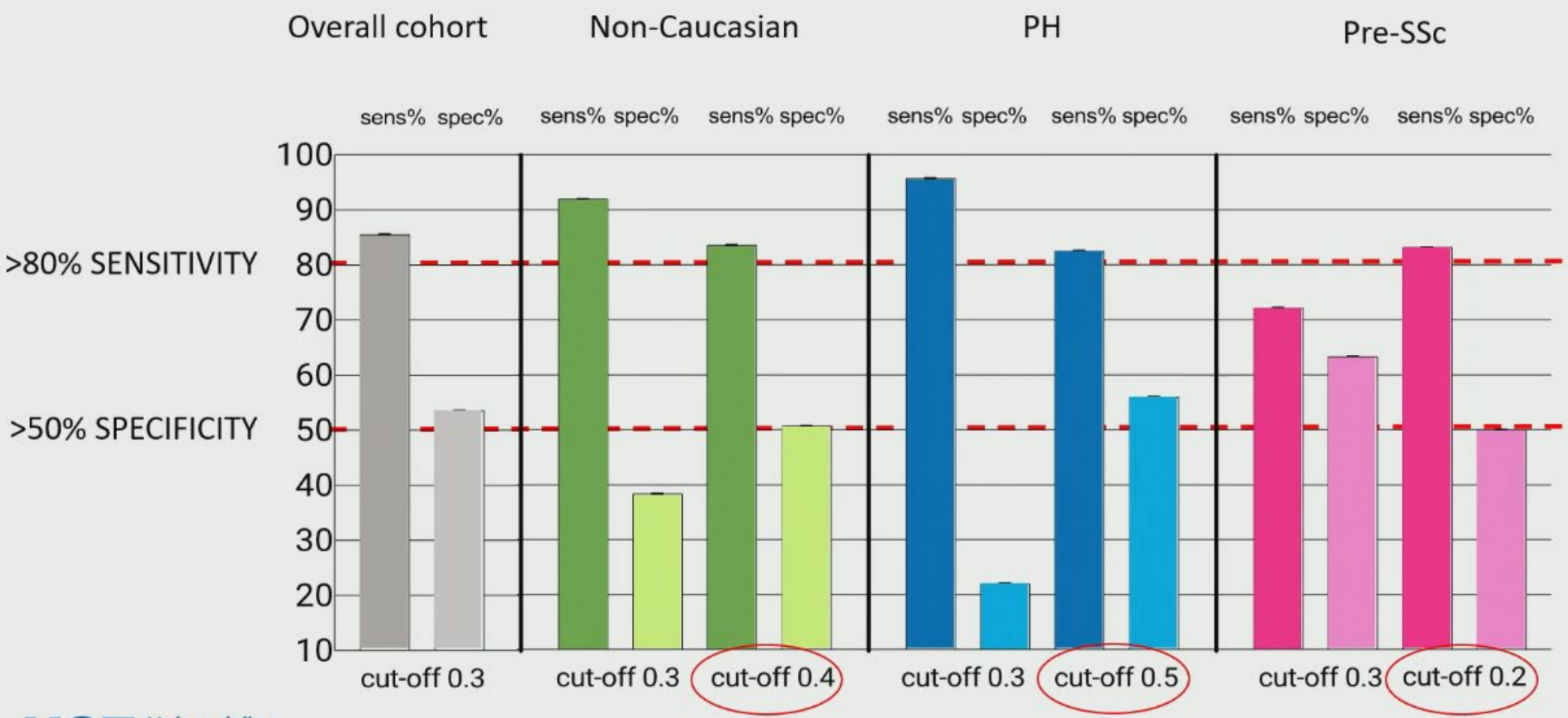
COHORT SELECTION



RESULTS (IV)

TAILORED CUT-OFF VALUES



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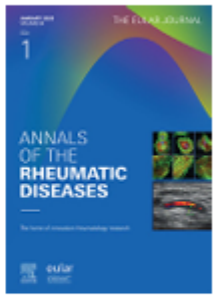

Treatment

Efficacy and safety of nerandomilast in patients with autoimmune disease-related progressive pulmonary fibrosis in the FIBRONEER-ILD trial

Anna-Maria Hoffmann-Vold[1,2,*], Shervin Assassi[3], Vincent Cottin[4], Michael Kreuter[5], Claudia Valenzuela[6], Marlies S Wijsenbeek[7], Hui Gu[8], Madhu Kanakapura[9], Ivana Ritter[9], Susanne Stowasser[9], Gerrit Weimann[9], Toby M Maher[10,11]

[1] *Department of Rheumatology, Oslo University Hospital, Oslo, Norway*
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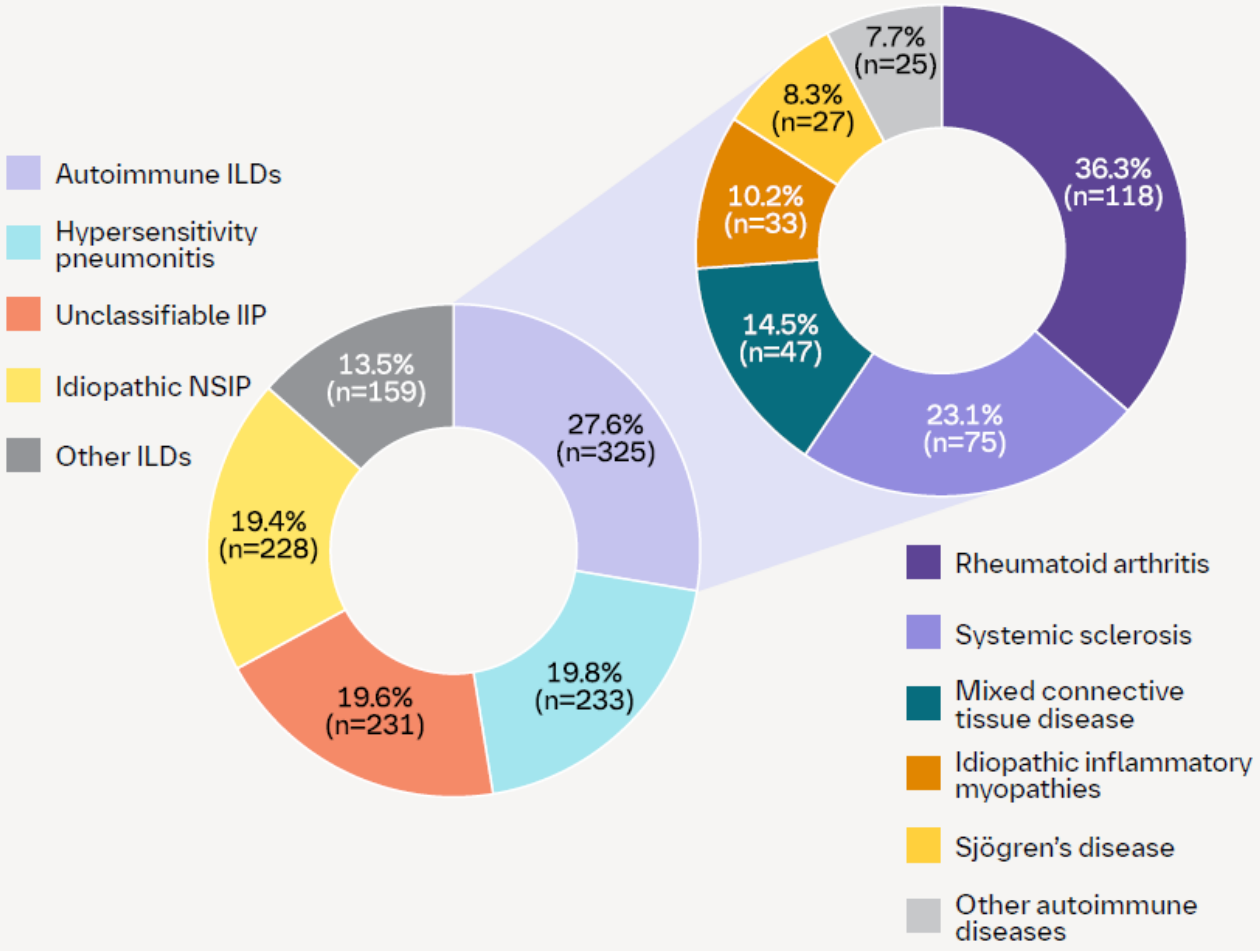
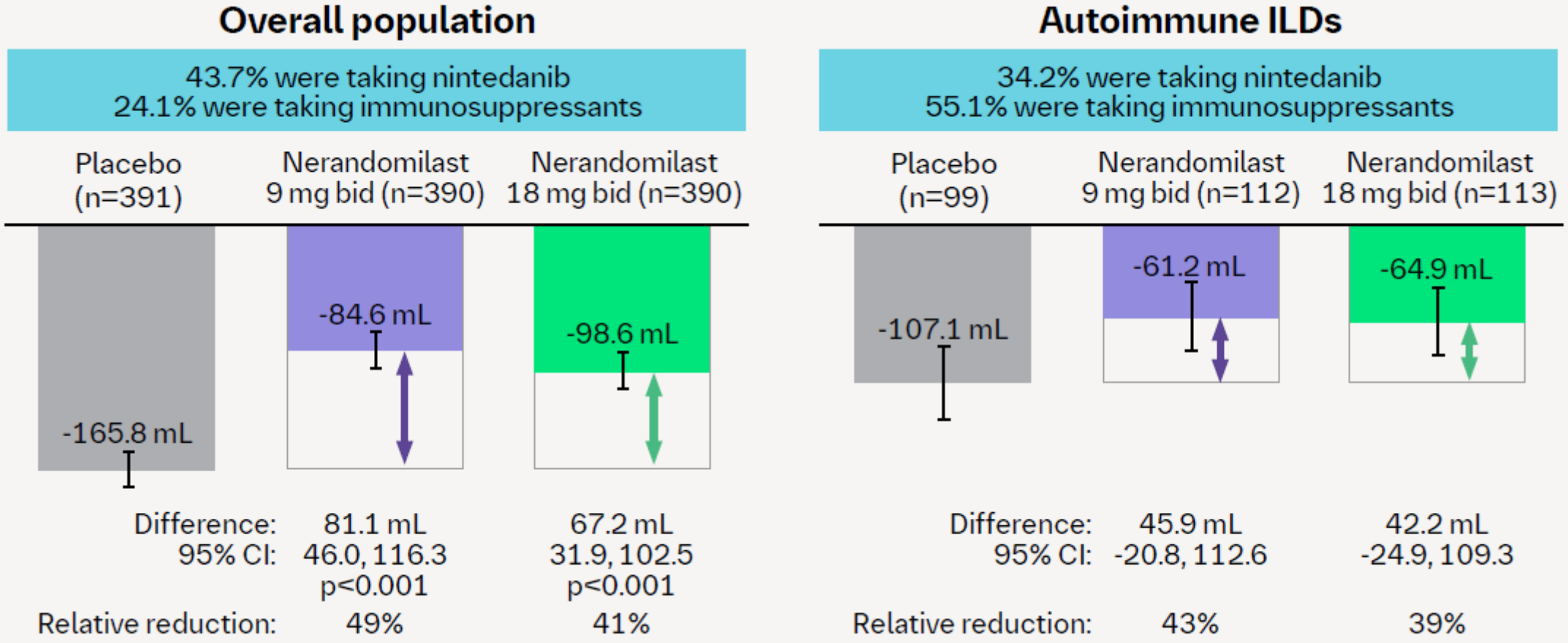
ILD diagnoses



Hoffmann-Vold AM et al. EULAR 2026 POS0382

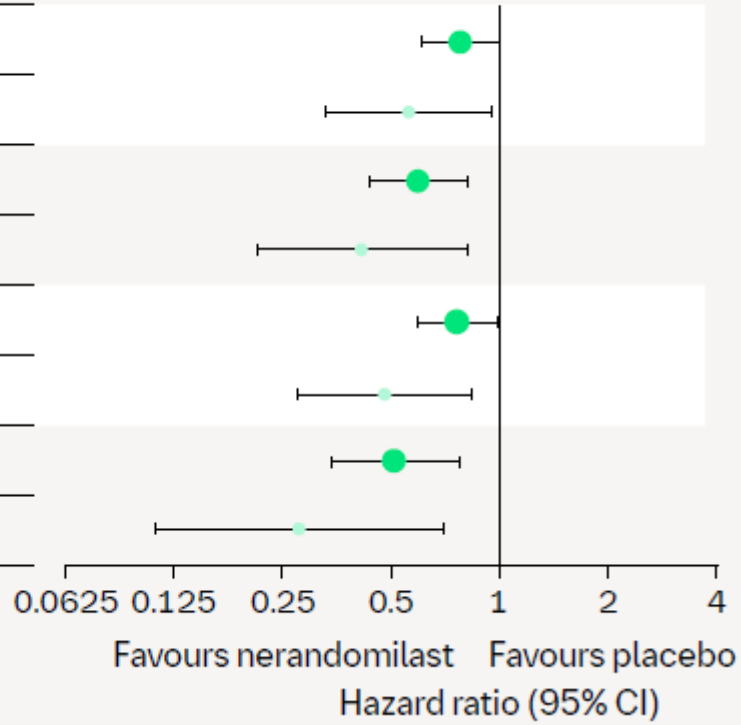
Change in FVC (mL) at week 52



Hoffmann-Vold AM et al. EULAR 2026 POS0382

Time-to-event endpoints with nerandomilast 18 mg bid vs placebo

		n (%) with event		
		Placebo	Nerandomilast	Hazard ratio (95% CI)
Acute exacerbation of ILD, hospitalisation for respiratory cause, or death	Overall	143 (36.5)	113 (28.9)	0.77 (0.60, 0.99)
	Autoimmune	33 (33.0)	28 (24.8)	0.56 (0.33, 0.94)
Acute exacerbation of ILD or death	Overall	97 (24.7)	59 (15.1)	0.59 (0.43, 0.82)
	Autoimmune	23 (23.0)	13 (11.5)	0.41 (0.21, 0.82)
Hospitalisation for respiratory cause or death	Overall	131 (33.4)	103 (26.3)	0.76 (0.59, 0.99)
	Autoimmune	31 (31.0)	23 (20.4)	0.48 (0.27, 0.84)
Death	Overall	64 (16.3)	34 (8.7)	0.51 (0.34, 0.78)
	Autoimmune	16 (16.0)	7 (6.2)	0.28 (0.11, 0.69)



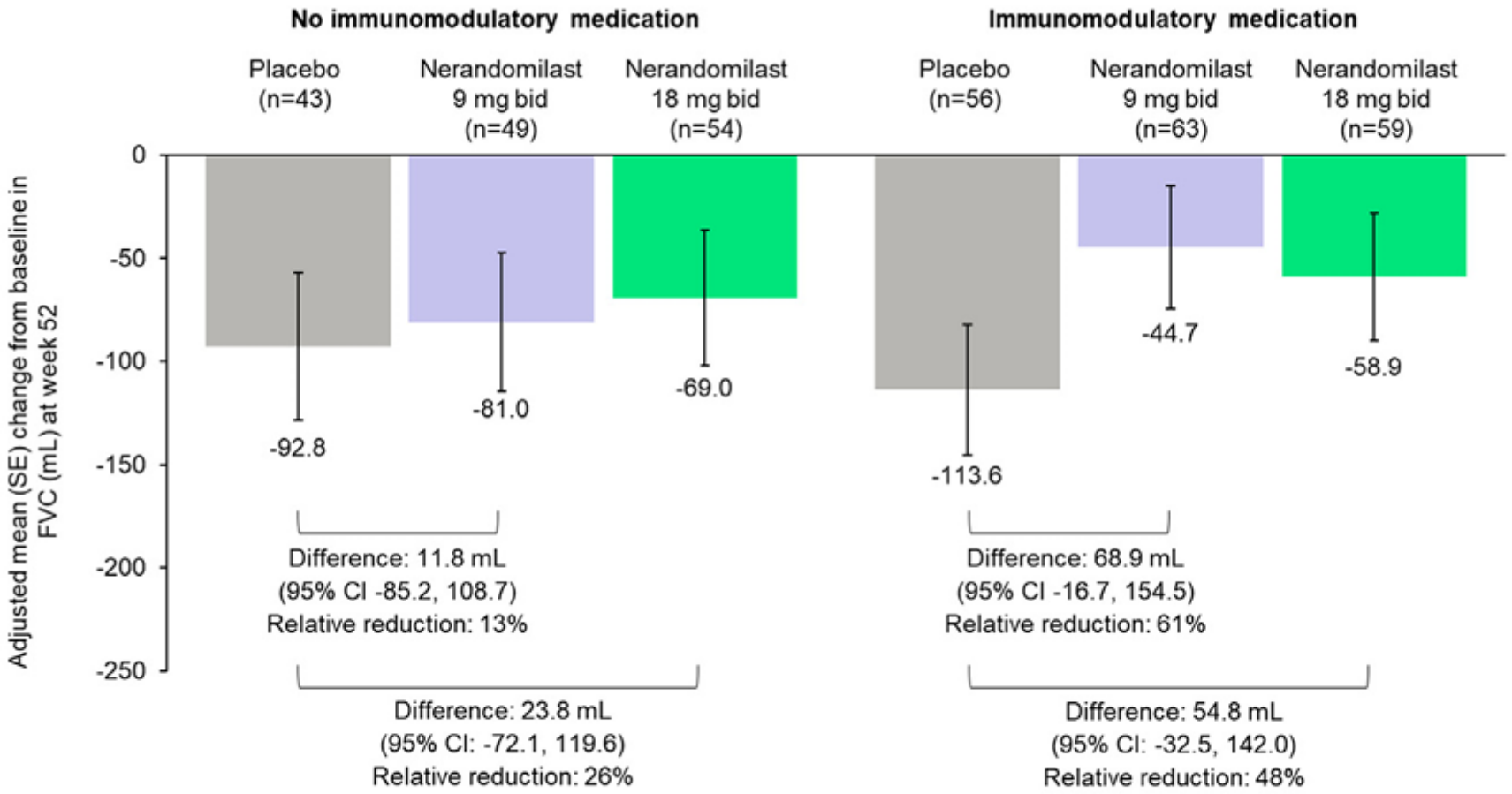
Hoffmann-Vold AM et al. EULAR 2026 POS0382

Supplementary Table S2. Use of immunomodulatory medications at baseline in diagnostic subsets (RA-ILD, SSc-ILD, MCTD-ILD) in the FIBRONEER-ILD trial.

	RA-ILD			SSc-ILD			MCTD-ILD		
	Placebo (n=32)	**Nerandomilast 9 mg bid (n=45)**	**Nerandomilast 18 mg bid (n=41)**	**Placebo (n=23)**	**Nerandomilast 9 mg bid (n=25)**	**Nerandomilast 18 mg bid (n=27)**	**Placebo (n=12)**	**Nerandomilast 9 mg bid (n=16)**	**Nerandomilast 18 mg bid (n=19)**
Any immunomodulatory medication (except prednisone)	21 (65.6)	29 (64.4)	30 (73.2)	13 (56.5)	12 (48.0)	10 (37.0)	6 (50.0)	9 (56.3)	12 (63.2)
Methotrexate	7 (21.9)	10 (22.2)	11 (26.8)	3 (13.0)	1 (4.0)	1 (3.7)	0	0	4 (21.1)
Azathioprine	0	3 (6.7)	2 (4.9)	6 (26.1)	4 (16.0)	2 (7.4)	1 (8.3)	3 (18.8)	2 (10.5)
Hydroxychloroquine sulphate	5 (15.6)	7 (15.6)	4 (9.8)	2 (8.7)	0	2 (7.4)	4 (33.3)	3 (18.8)	1 (5.3)
Hydroxychloroquine	2 (6.3)	6 (13.3)	2 (4.9)	1 (4.3)	2 (8.0)	1 (3.7)	0	2 (12.5)	4 (21.1)
Tacrolimus	1 (3.1)	0	0	2 (8.7)	2 (8.0)	3 (11.1)	1 (8.3)	1 (6.3)	2 (10.5)
Leflunomide	5 (15.6)	5 (11.1)	5 (12.2)	0	0	0	0	0	1 (5.3)
Ciclosporin	1 (3.1)	1 (2.2)	0	0	4 (16.0)	0	0	2 (12.5)	1 (5.3)
Abatacept	1 (3.1)	4 (8.9)	7 (17.1)	0	0	0	0	0	0
Methotrexate sodium	0	1 (2.2)	3 (7.3)	1 (4.3)	0	1 (3.7)	0	0	0
Iguratimod	0	3 (6.7)	1 (2.4)	0	0	0	0	0	1 (5.3)
Adalimumab	1 (3.1)	1 (2.2)	2 (4.9)	0	0	0	0	0	0
Colchicine	1 (3.1)	0	0	1 (4.3)	0	0	1 (8.3)	0	0

Data are n (%). Medications (other than prednisone) used by >1% of all patients with autoimmune ILDs at baseline are shown.

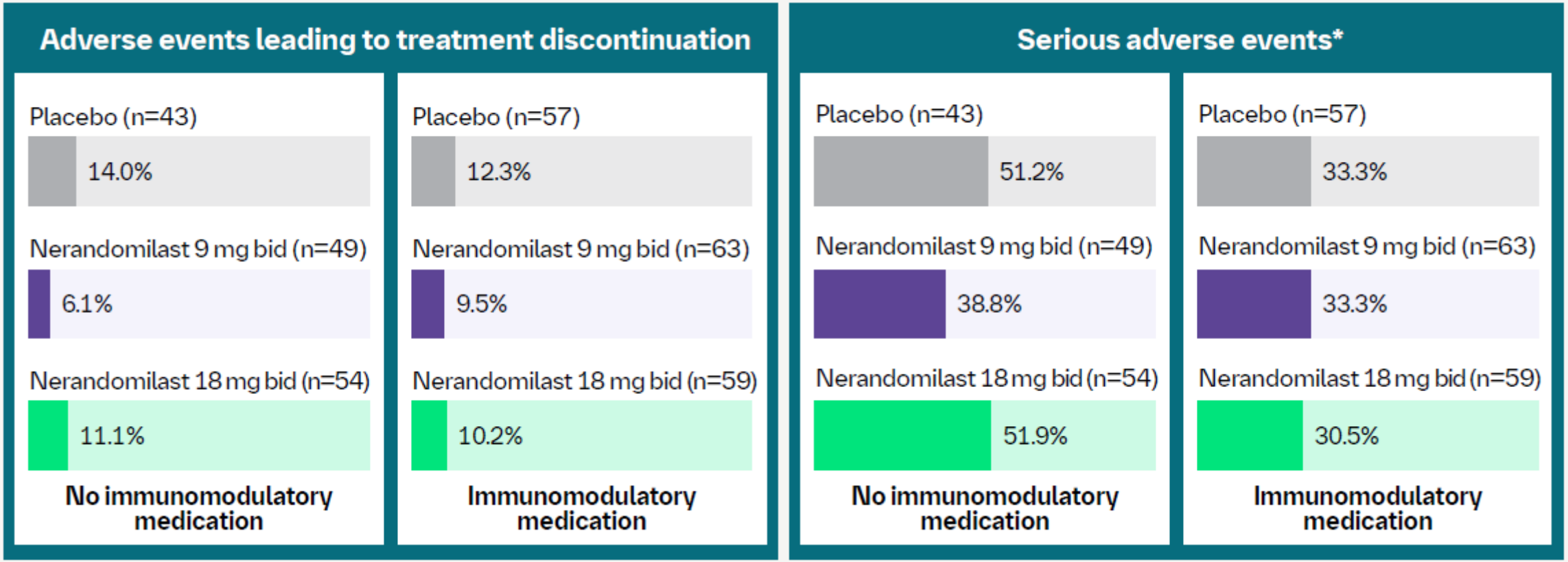
Effect of non-ILD background immunosuppressive medications



Nominal treatment-by-subset interaction p=0.39 for nerandomilast 9 mg bid and p=0.64 for nerandomilast 18 mg bid.

Hoffmann-Vold A et al, Ann Rheum Dis. 2026 Apr 29:S0003-4967(26)00214-1

Adverse events leading to discontinuation and serious adverse events in patients with autoimmune ILDs by use of immunomodulatory medication at baseline



Hoffmann-Vold AM et al. EULAR 2026 POS1207

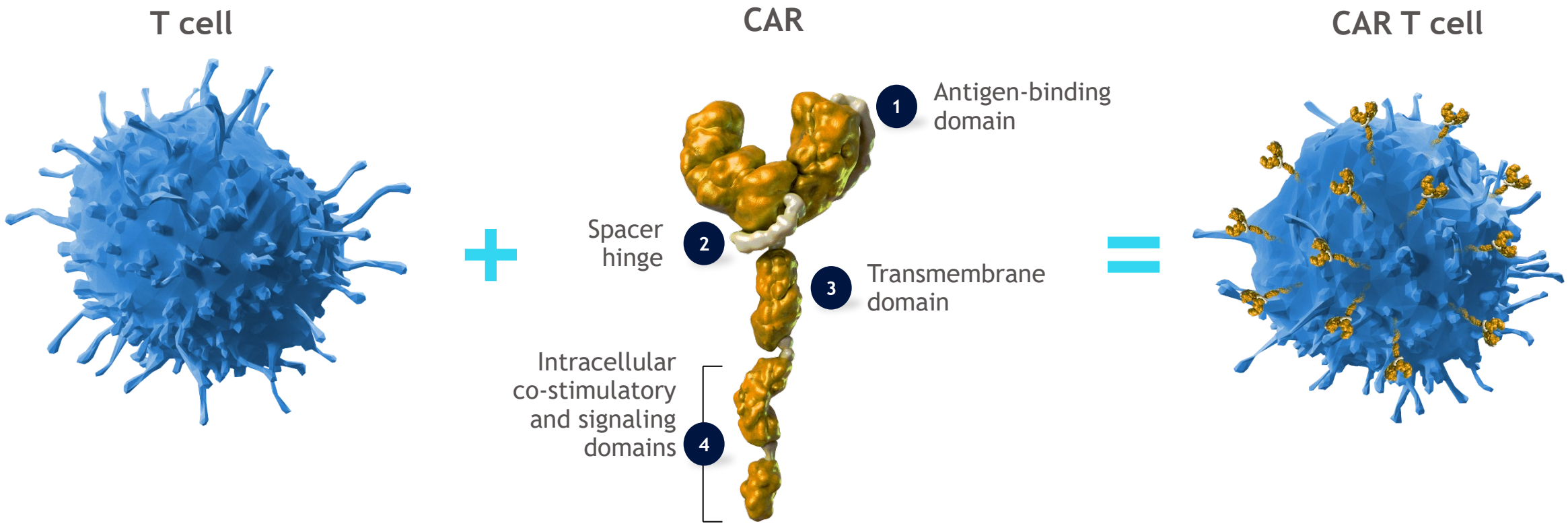
Most frequent adverse events in patients with autoimmune ILDs by use of immunomodulatory medication at baseline

	No immunomodulatory medication			Immunomodulatory medication		
	Placebo (n=43)	Nerando-milast 9 mg bid (n=49)	Nerando-milast 18 mg bid (n=54)	Placebo (n=57)	Nerando-milast 9 mg bid (n=63)	Nerando-milast 18 mg bid (n=59)
Diarrhoea	9 (20.9)	12 (24.5)	14 (25.9)	15 (26.3)	20 (31.7)	21 (35.6)
Cough	7 (16.3)	5 (10.2)	5 (9.3)	5 (8.8)	10 (15.9)	10 (16.9)
Condition aggravated	10 (23.3)	5 (10.2)	4 (7.4)	7 (12.3)	4 (6.3)	5 (8.5)
Upper respiratory tract infection	9 (20.9)	7 (14.3)	13 (24.1)	10 (17.5)	6 (9.5)	9 (15.3)
COVID-19	4 (9.3)	2 (4.1)	2 (3.7)	7 (12.3)	6 (9.5)	10 (16.9)
Nasopharyngitis	4 (9.3)	6 (12.2)	7 (13.0)	7 (12.3)	11 (17.5)	8 (13.6)
Depression	6 (14.0)	4 (8.2)	7 (13.0)	5 (8.8)	5 (7.9)	8 (13.6)
Pneumonia	6 (14.0)	6 (12.2)	6 (11.1)	10 (17.5)	5 (7.9)	7 (11.9)
Anxiety	6 (14.0)	3 (6.1)	6 (11.1)	5 (8.8)	8 (12.7)	11 (18.6)
Dyspnoea	3 (7.0)	1 (2.0)	4 (7.4)	4 (7.0)	8 (12.7)	4 (6.8)
Weight decreased	2 (4.7)	6 (12.2)	8 (14.8)	4 (7.0)	6 (9.5)	9 (15.3)
Nausea	2 (4.7)	1 (2.0)	6 (11.1)	2 (3.5)	5 (7.9)	10 (16.9)

Hoffmann-Vold AM et al. EULAR 2026 POS1207

Components of a CAR T cell[1-3]

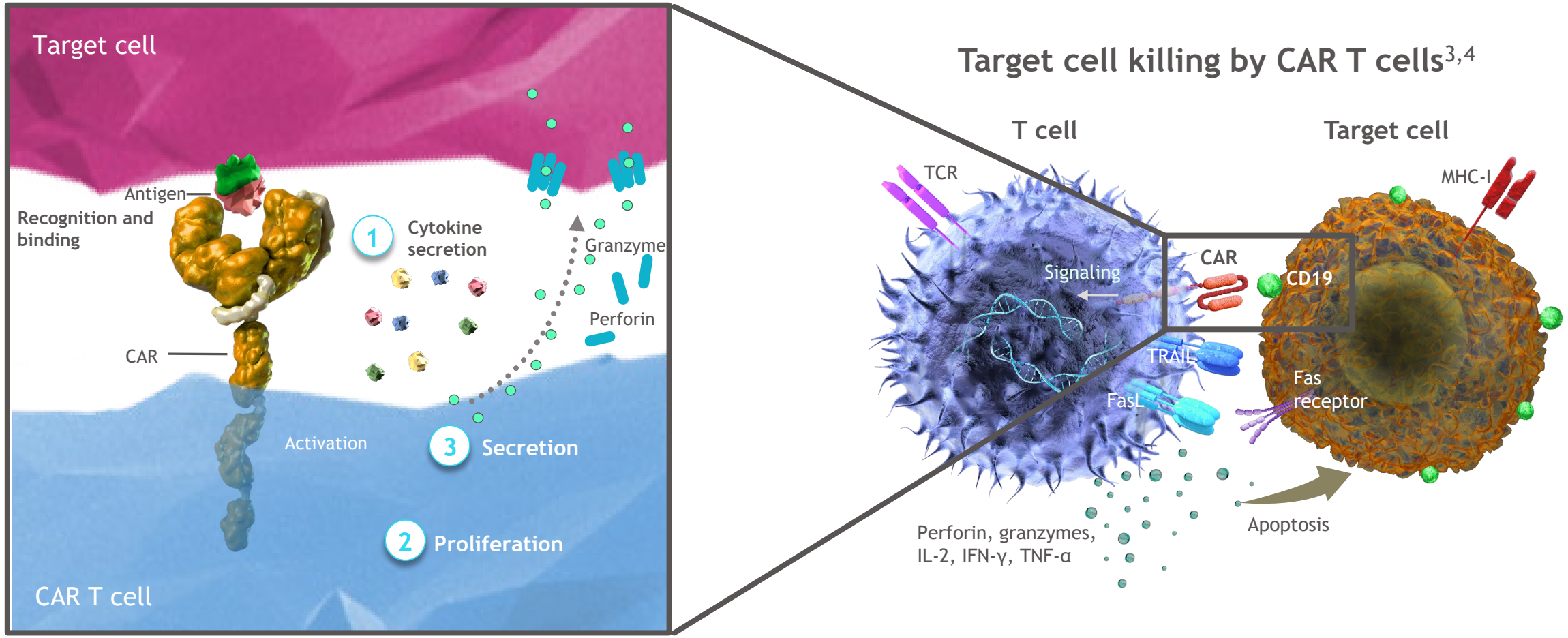
Autologous **CAR T cell therapy** equips a patient's T cells with the ability to detect and target specific cells by combining the **specificity of an antibody with the cytotoxic and memory capabilities of a T cell**[1,2]



CAR, chimeric antigen receptor.

1. Blood Cancer United. Accessed February 18, 2026. https://bloodcancerunited.org/resources/educational-resources/publications/booklet/chimeric-antigen-receptor-car-t-cell-therapy. 2. Maus MV, Levine BL. *Oncologist*. 2016;21:608–617. 3. Jayaraman J et al. *EBioMedicine*. 2020;58:102931.

CAR T cell mechanism of action involves recognition, binding, and activation of the CAR[1,2]



CAR, chimeric antigen receptor; CD, cluster of differentiation; FasL, Fas ligand; IFN, interferon; IL-2, interleukin 2; MHC, major histocompatibility complex; TCR, T cell receptor; TNF, tumor necrosis factor; TRAIL, tumor necrosis factor-related apoptosis-inducing ligand.

1. June CH, Sadelain M. *N Engl J Med*. 2018;379:64-73. 2. Dotti G et al. *Immunol Rev*. 2014;257:107-126. 3. Cartellieri M et al. *J Biomed Biotechnol*. 2010;2010:956304.
4. Schett G et al. *Lancet*. 2023;402:2034-2044.

Disclaimer: CAR T cell therapy is not approved for the treatment of autoimmune diseases in any country and is an investigational therapy undergoing evaluation

CAR T cell therapy trials in SSc (phase 2 and phase 3)

Sponsor	Therapy	Trial	NCT	Phase
Bristol Myers Squibb[1]	Zolacaptagene autoleucel (BMS-986353)	Breakfree-SSc	NCT07335562	Phase 3
Novartis[2]	Rapcabtagene autoleucel (YTB323)		NCT06655896	Phase 2
Cabaletta Bio[3]	Resecabtagene autoleucel (CABA-201)	RESET-SSc	NCT06328777	Phase 1/2
Kyverna Therapeutics[4]	KYV-101	KYSA-5	NCT06400303	Phase 1/2

CAR, chimeric antigen receptor; SSc, systemic sclerosis.
1. ClinicalTrials.gov. Accessed February 2026. https://clinicaltrials.gov/study/NCT07335562. 2. ClinicalTrials.gov. Accessed February 2026. https://clinicaltrials.gov/study/NCT06655896. 3. ClinicalTrials.gov. Accessed February 2026. https://clinicaltrials.gov/study/NCT06328777. 4. ClinicalTrials.gov. Accessed February 2026. https://clinicaltrials.gov/study/NCT06400303.

Disclaimer: CAR T cell therapy is not approved for the treatment of autoimmune diseases in any country and is an investigational therapy undergoing evaluation

Safety and Early Efficacy of Rapcabtagene Autoleucel (YTB323), an Autologous CD19 Directed Chimeric Antigen Receptor T-cell Therapy, in Severe Refractory Idiopathic Inflammatory Myopathies and Diffuse Cutaneous Systemic Sclerosis: Preliminary Analysis of the Open-label AUTOGRAPH-IIM and -SSc Studies

Dinesh Khanna[1], Xavier Andrade-Gonzalez[2], Mark Vercel[2], Merav Lidar[3], Avichai Shimoni[3], Alexander Pfeil[4], Ulf Schnetzke[5], Gianluca Moroncini[6], Francesco Saraceni[7], Yves Allenbach[8], Sylvain Choquet[8], Andrea HL Low[9,10], Yunxin Chen[9], Takashi Matsushita[11], Akiyo Yoshida[11], Marie-Elise Truchetet[12], Edouard Forcade[12], Ran Nakashima[13], Yasuyuki Arai[13], John Moore[14], Laila Girgis[14], Lidia Sanchez-Riera[15], Angeliki Giannelou[16], Yuejia Xu[17], Alexandra Kuznetsova[17], Fernando Takeshi Kawakami[18], Bibha Das[18], Chih-Yung Sean Lee[19], Ewa Gatlik[18], Muhsen Alani[16,20], Vineeth Varanasi[21], Susanne Diehl[18], Anubhav N Mathur[16], Rohit Aggarwal[22]

[1]University of Michigan, Ann Arbor, United States of America; [2]Avera Cancer Institute, Sioux Falls, United States of America; [3]Chaim Sheba Medical Center, Ramat Gan, Israel; [4]Jena University Hospital, Friedrich Schiller University Jena, Germany; [5]Universitätsklinikum Jena, Jena, Germany; [6]Marche University Hospital, Ancona, Italy; [7]Azienda Ospedaliero Universitaria Delle Marche, Ancona, Italy; [8]Pitié-Salpêtrière University Hospital, Paris, France; [9]Singapore General Hospital, Singapore; [10]Duke-National University of Singapore Medical School, Singapore; [11]Kanazawa University Hospital, Kanazawa Ishikawa, Japan; [12]CHU Bordeaux GH Pellegrin, Bordeaux, France; [13]Kyoto University Hospital, Kyoto, Japan; [14]St. Vincent's Hospital, Sydney, Australia; [15]Novartis Farmacéutica, S.A., Barcelona, Spain; [16]Novartis Pharmaceuticals, East Hanover, New Jersey, United States of America; [17]Novartis Pharma AG, London, United Kingdom; [18]Novartis Pharma AG, Basel, Switzerland; [19]Novartis Pharmaceuticals, Massachusetts, United States of America; [20]University of Washington, Seattle, United States of America; [21]Novartis Healthcare Pvt Ltd, Hyderabad, India; [22]University of Pittsburgh School of Medicine, Pittsburgh, United States of America.

Khanna D et al. EULAR 2026 OP077

Baseline characteristics: Cohort 1 (safety lead-in)

Characteristics	IIM (N=6)	dcSSc (N=6)
Age (years), median (range)	56 (43–61)	49 (32–56)
Sex: Female, n	2	5
Race: White / Asian / not reported, n	2 / 2 / 2	3 / 1 / 2
Diagnosis: ASyS / DM / IMNM, n	3 / 2 / 1	-
Time since diagnosis (years), median (range)	4.0 (0.3–8.9)	1.9 (0.8–3.0)
Positive MSA / anti-Scl-70 / anti-RNA polymerase 3, n	6 / - / -	- / 5 / 2
Number of prior background non-GC systemic immunosuppressive and immunomodulatory therapies, median (range)	3.5 (3–5)	2.5 (2–4)
History of ILD, n	**3**	**3***
FVC% predicted, median (range)	**68.1 (57.5–81.7)**	**88.9 (75–114)**
mRSS score, median (range)	-	36.5 (34–48)
MMT-8, median (range)	129 (98–133)	-
CDASI score, median (range)	28 (18–38)**	-
PhGA, median (range)	6.85 (4.5–10.0)	9.0 (7.0–10.0)
HAQ-DI, median (range)	1.5 (0.1–2.6)†	1.9 (1.5–2.75)
Follow-up duration since rap-cel infusion (weeks), median (range)	38 (32–54)	38 (34–58)

As of the latest data cut-off, 4/6 **IIM** patients and 6/6 **dcSSc** patients completed assessments up to Week 38±

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Data show a favorable safety profile consistent with the established profile of CD19-targeted CAR-T cell therapies

As of the data cut-off (**≥Week 24**):

- Adverse events were **manageable** and consistent with the established safety profile of CAR-T cell therapies
- All cases of **CRS resolved without sequelae**
- All events of infection were **responsive** to clinical management and **recovered without sequalae**
- **No instances of ICANS** or immune effector cell-associated hemophagocytic lymphohistiocytosis-like syndrome were reported
- **No deaths were reported**

	IIM (N=6)		dcSSc (N=6)	
	All Grs, n	**Gr ≥3, n**	**All Grs, n**	**Gr ≥3, n**
Number of patients with ≥1 AESI	6	3	6	5
Infections	4	1	5	1
Hematological disorders	3	3	4	4
Neutropenia	3	3	4	3
Hypogammaglobulinemia	0	0	1	0
Received IVIG	-	-	0	-
CRS	5	1	6	0
Received tocilizumab	4	1	6	-
Median time to onset (range), days	7 (4–8)		6 (5–14)	
Median duration (range), days	11 (9–14)		6 (1–14)	

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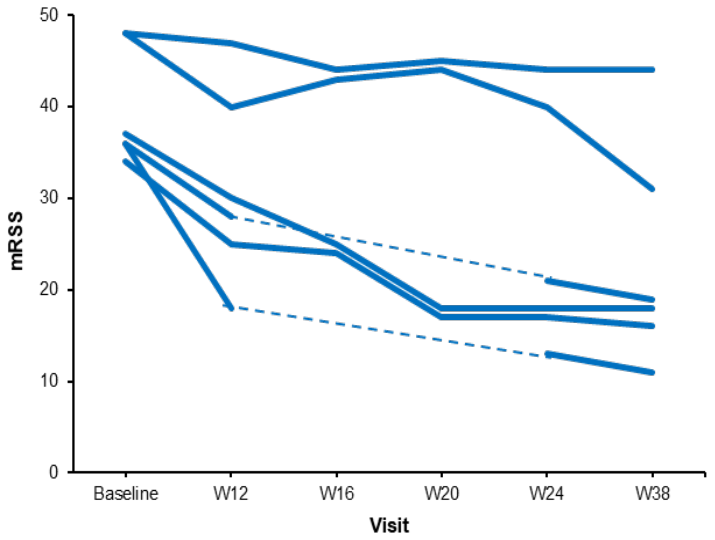
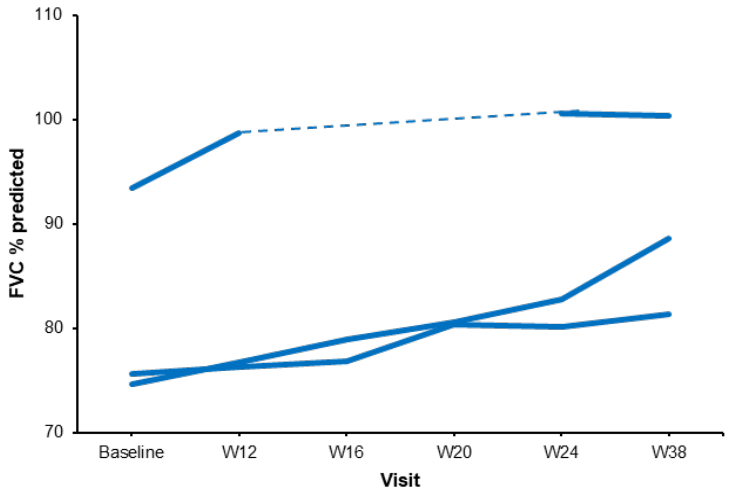
Improvements in mRSS and FVC % predicted were observed in patients with dcSSc

- At data cut-off, median (range) reduction in mRSS score from BL was **17.5 (4–25) points (49.3% [8–69] relative reduction)**
- Among patients with history of ILD (n=3), median (range) absolute **improvement from baseline in FVC % predicted was 6.9% (6–14) (7.5% [7–19] relative improvement)**
- The majority of patients **achieved a rCRISS response at Week 38**; **all patients remained treatment-free from immunomodulatory therapy at last follow-up**

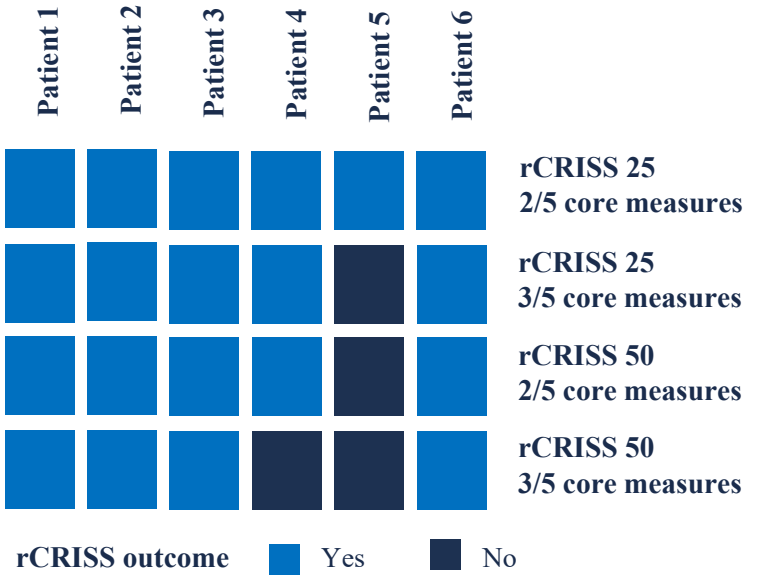
mRSS over time (N=6)



FVC % predicted in patients with history of ILD over time (n=3)



rCRISS at Week 38 (n=6)

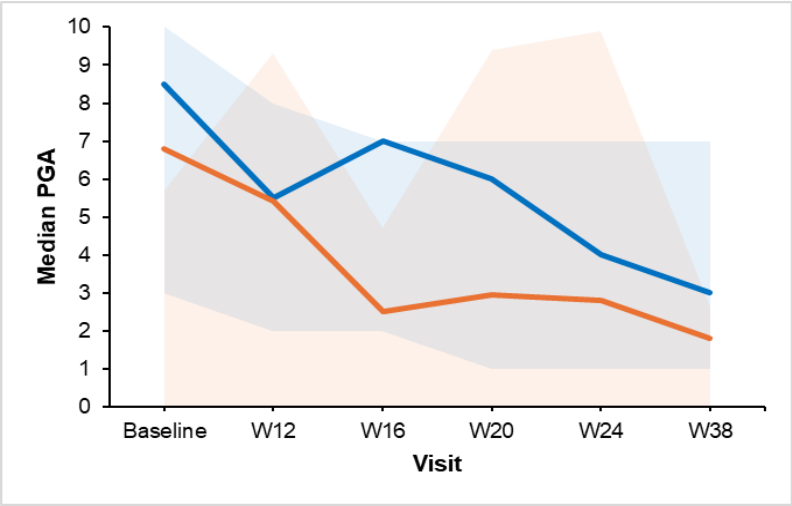


Khanna D et al. EULAR 2026 OP077

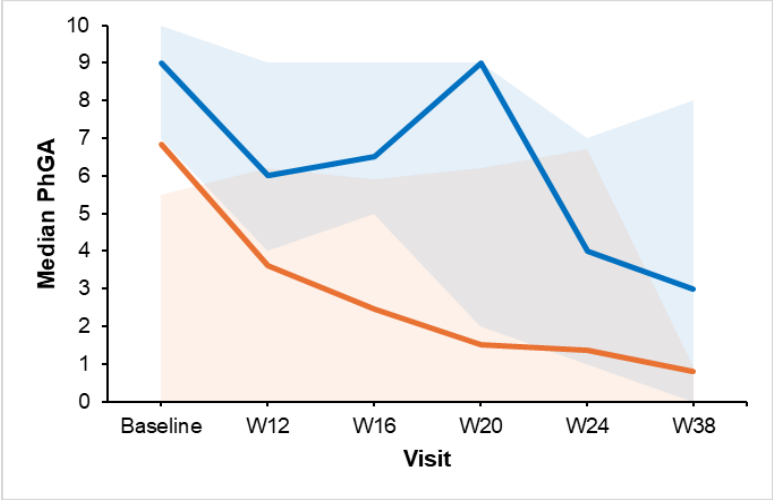
Improvements in PGA, PhGA and HAQ-disability index scores were observed over time in patients with IIM or dcSSc

- In patients with **IIM**, median relative decreases from baseline in PGA, PhGA and HAQ-disability index were 73.3%, 85.9% and 81.7% (N=6) at Week 38, respectively
- In patients with **dcSSc**, median relative decreases from baseline in PGA, PhGA and HAQ-disability index were 66.7%, 60.3% and 62.6% (N=6) at Week 38, respectively

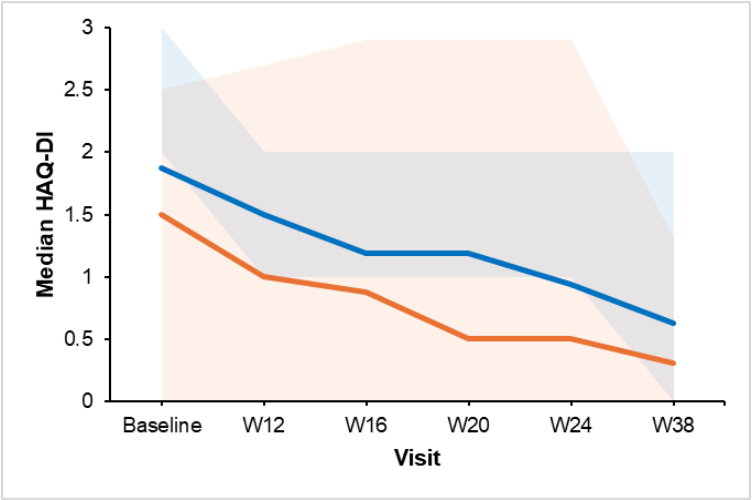
Median PGA in patients with IIM (n=5) and dcSSc (N=6) over time



Median PhGA in patients with IIM (N=6) and dcSSc* (N=6) over time

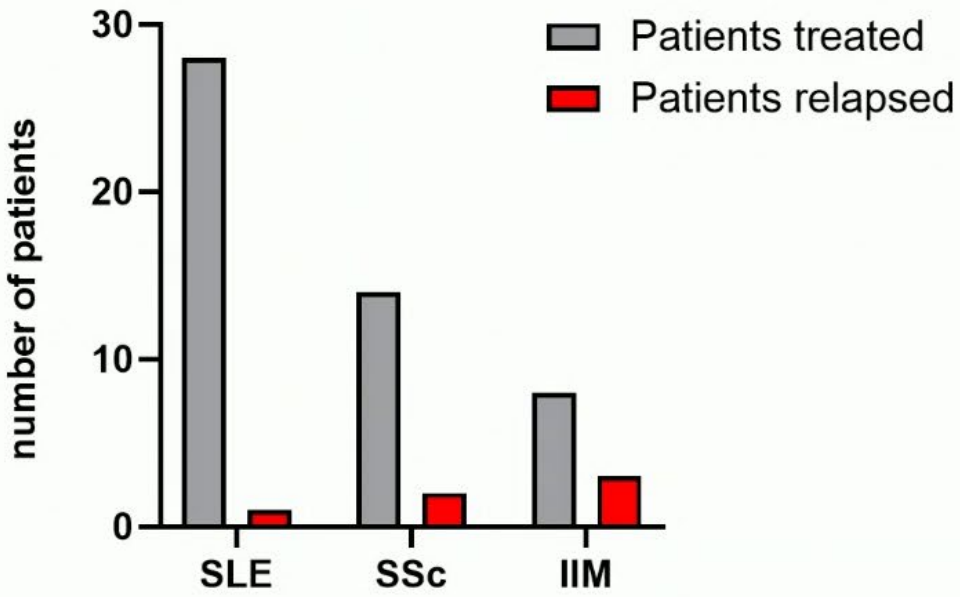


Median HAQ-DI scores in patients with IIM (n=5) and dcSSc (N=6) over time



Khanna D et al. EULAR 2026 OP077

Overview of relapses

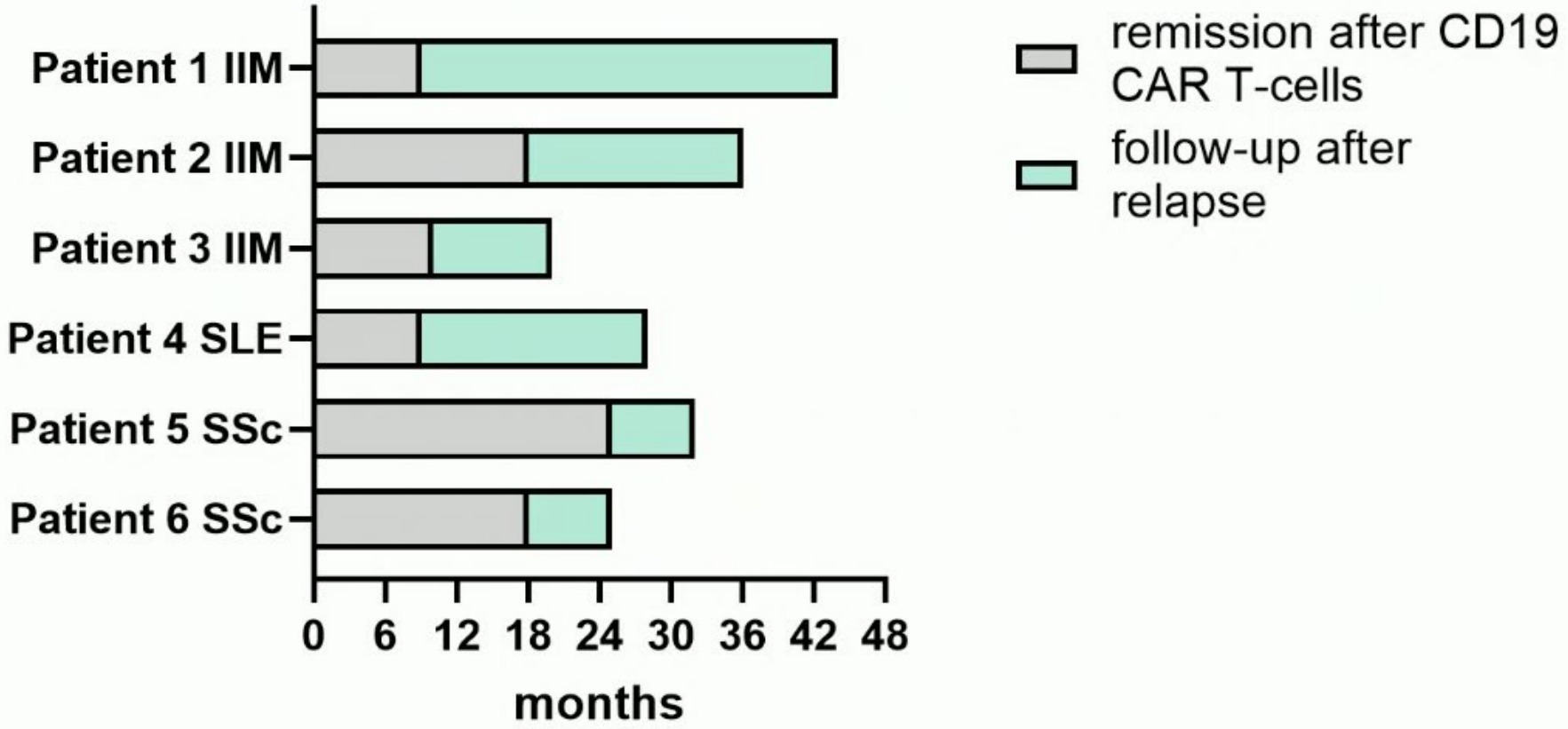


- N=50 treated with autologous CD19 CAR T-cells having a follow-up of >1 year
- Overall relapse rate 12%
- Higher probability of relapse in IIM (3/8, 37%) than SSc (2/14, 14%) and SLE (1/28, 3%)

Andreas Wirsching

Management of relapses after autologous CD19 CAR T-cell therapy in SLE, SSc and IIM – a case series

Overview of relapses



- Mean overall follow-up period (n=50): 2.5 years
- Median time of relapse (n=6): 14.8 months (range: 9-25 months)

Patient 6 – 24 year old female with Scl70-positive **systemic sclerosis**
manifestations: interstitial lung disease, skin

CD19-
CAR T-cells
Zorpo-Cel
(Miltenyi)

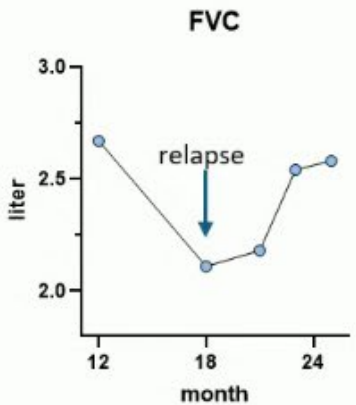
relapse of ILD,
skin, myocardial

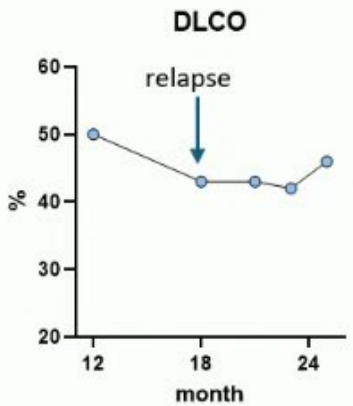
Teclistamab
step-up dosing
(0.06, 0.3, 1.5 mg/kg)

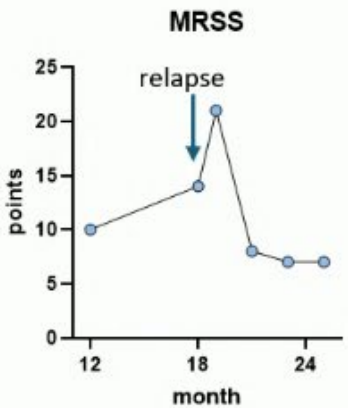
Baseline

Month 18

Month 25





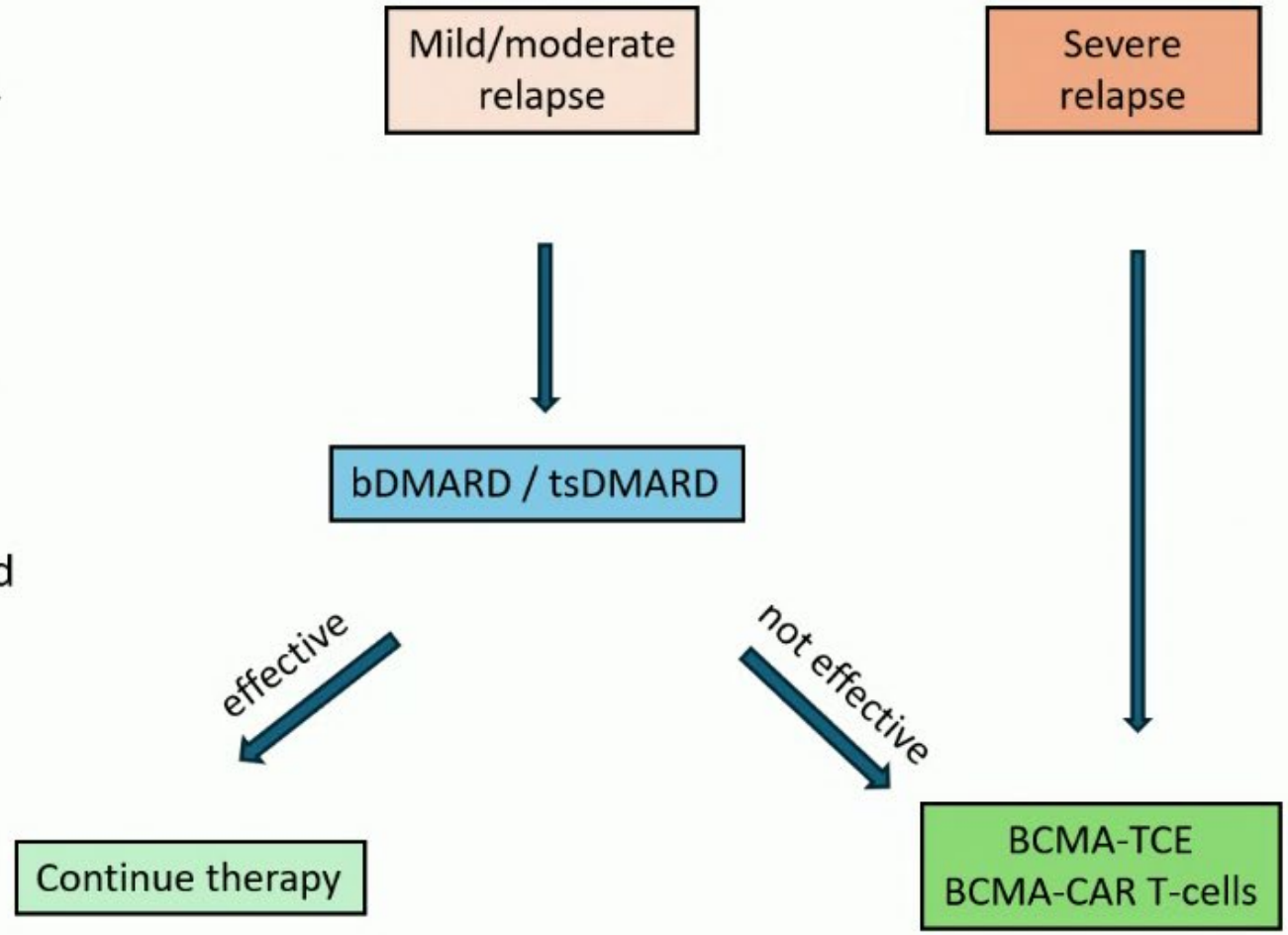


active disease

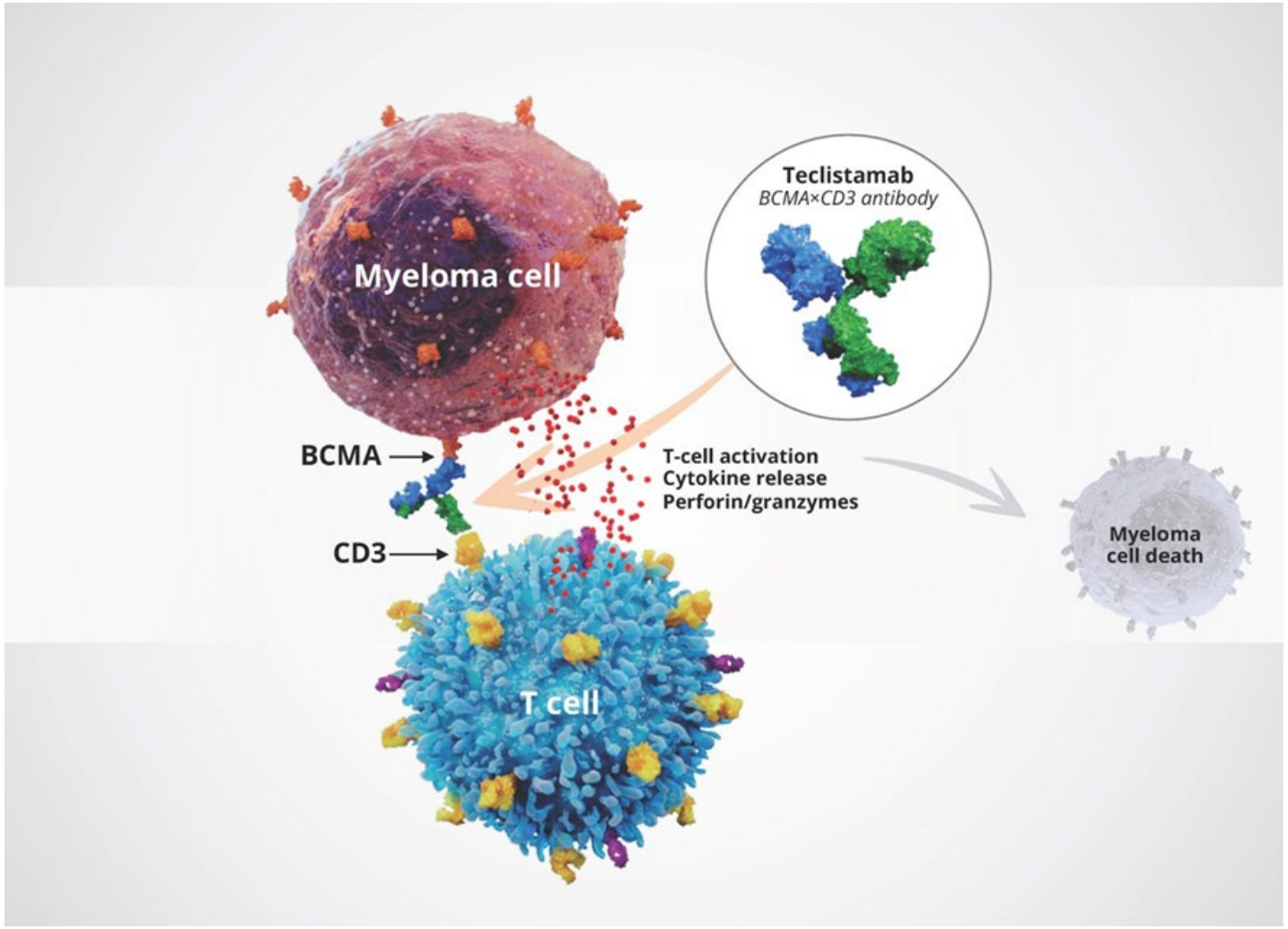
ACR CRISS 50
response 3/5 items

Algorithm of management in relapses after CD19 CAR T-cells

- Relapses are found in a minority of CD19-CAR T-cell treated patients and seem to occur slightly more frequent in myositis and systemic sclerosis than SLE
- BCMA-CAR and BCMA-TCEs can successfully reinstall drug-free remission in patients with relapsing autoimmune disease after CD19-CAR T-cells
- Administration of the same cryopreserved CAR T-cell product may fail due to antigenicity requiring the switch to another CAR T-cell product



Bispecific T cell engagers: Mode of Action

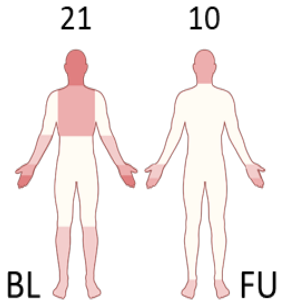


Guo et al, Clin Transl Sci. 2024;17:e13717.

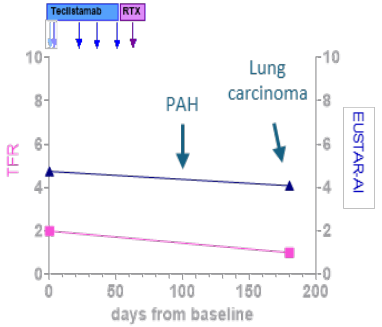
BCMA-TCE (teclistamab) in treatment refractory SSc

SSc patient 1

Previous treatments:
Azathioprine
Cyclophosphamide
Mycophenolate

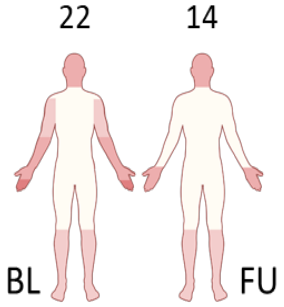


mRSS: 0 1 2 3

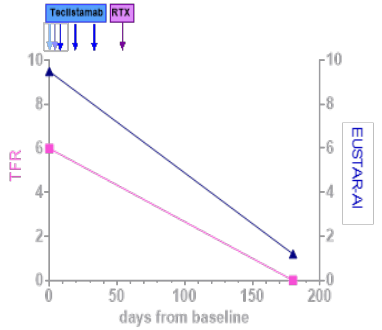


SSc patient 2

Previous treatments:
Hydroxychloroquine
Methotrexate
Mycophenolate

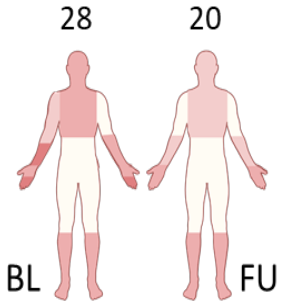


mRSS: 0 1 2 3

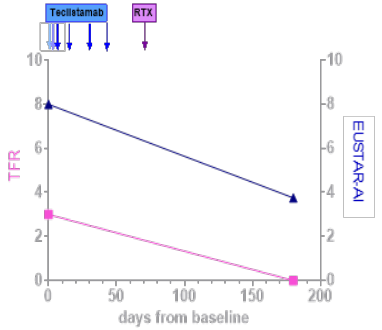


SSc patient 3

Previous treatments:
Methotrexate
Mycophenolate
Nintedanib
RTX

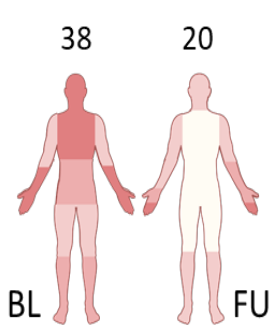


mRSS: 0 1 2 3

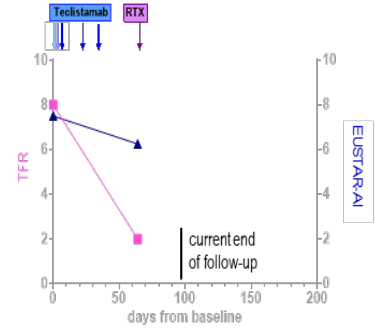


SSc patient 4

Previous treatments:
Methotrexate
Cyclophosphamide
RTX

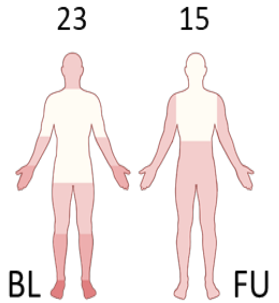


mRSS: 0 1 2 3

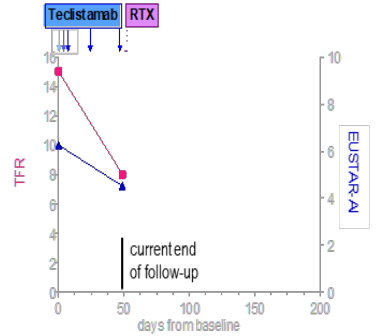


SSc patient 5

Previous treatments:
Mycophenolate
Cyclophosphamide
Nintedanib
RTX



mRSS: 0 1 2 3



--- Tendon friction rubs

---- EUSTAR Activity Score

Düsing et al., Nat Med. 2026 Feb 19.