EULAR review

Annual European Congress of Rheumatology

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#EULAReview23

Sociedad Española de Reumatología

Enfermedades Autoinmunes Sistémicas: Vasculitis, Miopatías y Otras Conectivopatías

Dra. Vanesa Calvo del Río

Servicio de Reumatología

Hospital Universitario Marqués de Valdecilla. Santander

What is new in Large-vessel vasculitis

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TCZ en ACG: ?

1. Qué pacientes se deben tratar?

2. Durante cuanto tiempo?

3. Es eficaz y seguro el tto con TCZ a largo plazo?

4. Puede TCZ prevenir las complicaciones vasculares graves?

5. TCZ puede reemplazar a los GC en el tratamiento de la ACG?

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   - **TCZ monoterapia** tras ciclo ultracorto GC eficaz y seguro **ACG no craneal**

What is new in Large-vessel vasculitis

Otras dianas en ACG


Arthritis Care & Research

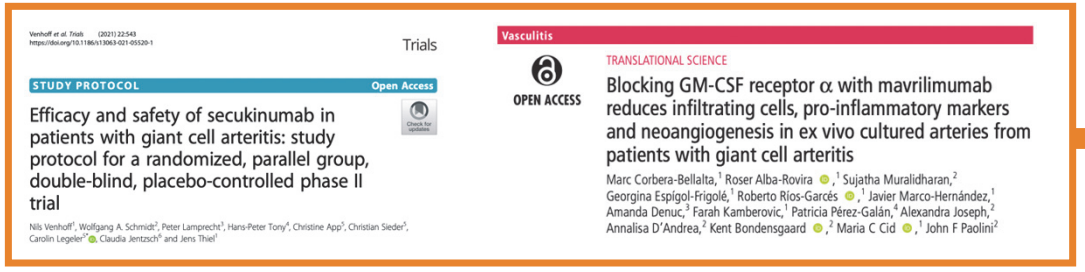
Brief Report

Ustekinumab for the Treatment of Giant Cell Arteritis

Mark A. Matza, Ana D. Fernandes, John H. Stone, Sebastian H. Unizony

First published: 05 April 2020 | https://doi.org/10.1002/acr.24200 | Citations: 12

- no muy clara eficacia


Trials

STUDY PROTOCOL Open Access

Efficacy and safety of secukinumab in patients with giant cell arteritis: study protocol for a randomized, parallel group, double-blind, placebo-controlled phase II trial

Vasculitis

OPEN ACCESS

TRANSLATIONAL SCIENCE

Blocking GM-CSF receptor α with mavrilimumab reduces infiltrating cells, pro-inflammatory markers and neoangiogenesis in ex vivo cultured arteries from patients with giant cell arteritis

- parecen eficaces


HHS Public Access

Author manuscript

Ann Rheum Dis. Author manuscript; available in PMC 2022 October 24.

Published in final edited form as:

Ann Rheum Dis. 2022 June ; 81(6): 861–867. doi:10.1136/annrheumdis-2021-221961.

Baricitinib for relapsing giant cell arteritis: a prospective open-label 52-week pilot study

Matthew J. Koster,

Department of Internal Medicine, Division of Rheumatology, Mayo Clinic, Rochester, MN, USA

U.S. National Library of Medicine

ClinicalTrials.gov

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Home > Search Results > Study Record Detail

A Study to Evaluate the Safety and Efficacy of Upadacitinib in Participants With Giant Cell Arteritis (SELECT-GCA)

- parecen efectivos
- resultados UPA?
- uso limitado por EMA (recomendaciones seguridad/edad)

LONGITUDINAL TRAJECTORIES OF RENAL FUNCTION IN ANCA-ASSOCIATED VASCULITIS: FINDINGS FROM THE EXPANDED MASS GENERAL BRIGHAM COHORT

J. Hanberg[1], C. Cook[2,3], X. Fu[2,3], H. Choi[2,3,4], Y. Zhang[2,3,4], Z. Wallace[2,3,4]

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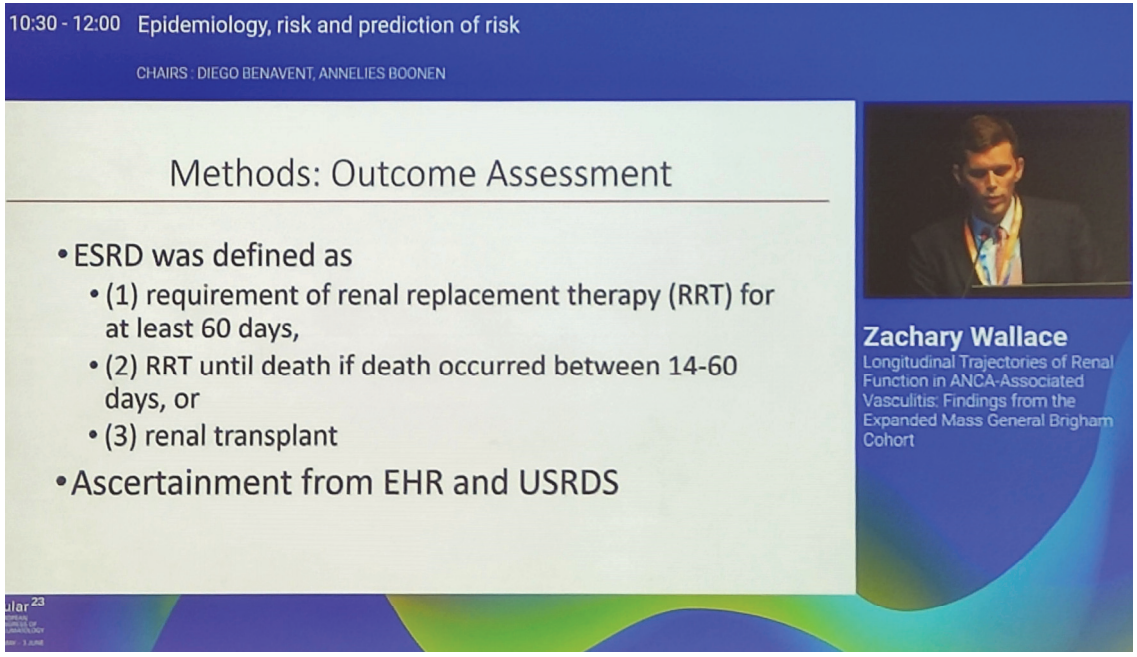
OP0273 (2023)



Objetivos:

- Confirmar y describir trayectoria longitudinal función renal en cohorte VAA.

Métodos:

- cohorte VAA Mass General Brigham (2002-2022)
- ≥ 2 mediciones función renal
- evaluación hasta mensualmente -12 m y +24 m respecto fecha índice

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OP0273 (2023)

Resultados: 375 pc

		6%	29%	58%	7%	
	Overall (N=375)	Rapid decline (N=23)	Impaired (N=109)	Preserved (N=216)	Recovery (N=27)	p-value
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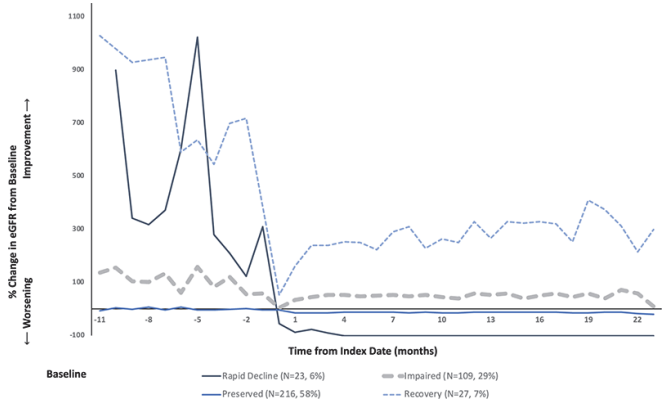

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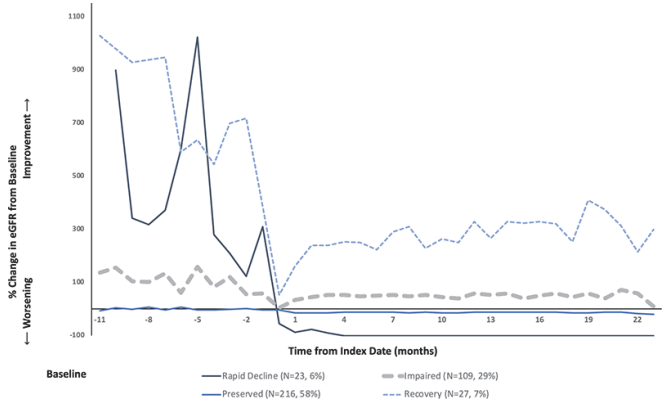

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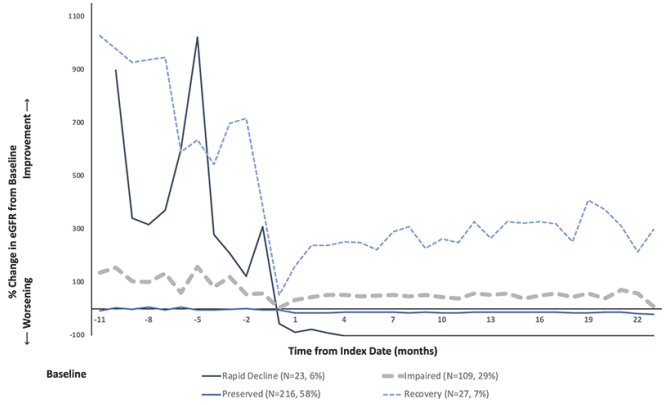

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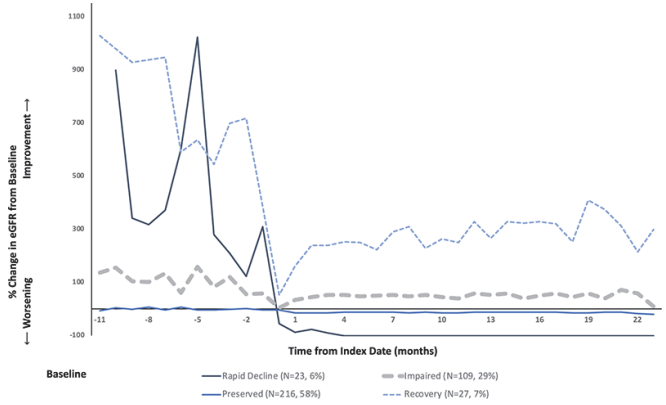

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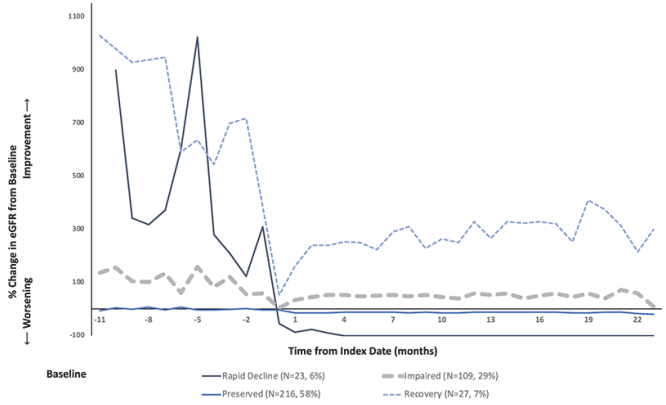

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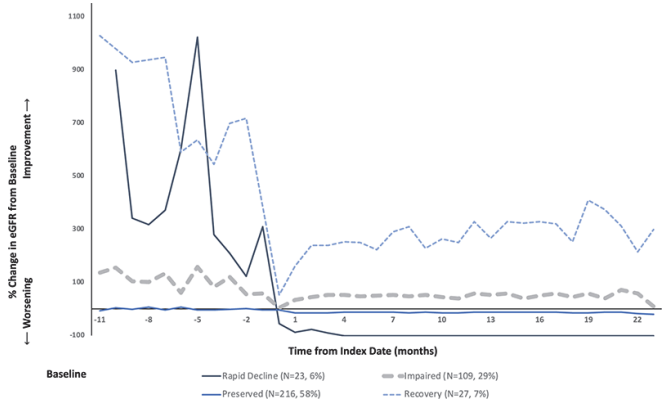
Hypertension	135 (36)	13 (57)	40 (37)	69 (32)	13 (48)	0.06
eGFR (±30d, median, IQR)	51 (21-88)	7 (6-10)	27 (18-39)	82 (56-97)	10 (8-16)	<0.001
Outcomes						
Permanent ESRD*	49 (13)	22 (96)	11 (10)	15 (7)	1 (4)	<0.0001
Due to active AAV	30 (61)	20 (91)	5 (45)	5 (33)	0 (0)	0.01
Time to ESRD (y)*	0.1 (0.0-3.5)	0.01 (0-0.1)	3.5 (0-6.8)	2.5 (0-5.6)	10 (10-10)	<0.001
CKD ≥III at 1 y	210 (56)	23 (100)	93 (85)	67 (31)	27 (100)	<0.001
CKD ≥III at 2 y	144 (49)	13 (100)	62 (70)	58 (33)	11 (69)	<0.001
CKD ≥III at 5 y	88 (48)	10 (100)	37 (69)	36 (34)	5 (38)	<0.001



Conclusiones

- Confirmamos 4 grupos distintos de trayectoria renal en VAA.
- Diferencias grupos:
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LONGITUDINAL TRAJECTORIES OF RENAL FUNCTION IN ANCA-ASSOCIATED VASCULITIS: FINDINGS FROM THE EXPANDED MASS GENERAL BRIGHAM COHORT

J. Hanberg[1], C. Cook[2,3], X. Fu[2,3], H. Choi[2,3,4], Y. Zhang[2,3,4], Z. Wallace[2,3,4]
[1]Massachusetts General Hospital, Department of Medicine, Boston, United States of America
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OP0273 (2023)

Resultados: 375 pc

		6%	29%	58%	7%	
	Overall (N=375)	Rapid decline (N=23)	Impaired (N=109)	Preserved (N=216)	Recovery (N=27)	p-value
Baseline characteristics						
Age at diagnosis (mean, SD)	62 (17)	65 (17)	64 (16)	61 (17)	64 (18)	0.22
Female (N, %)	226 (60)	13 (57)	63 (58)	136 (63)	14 (52)	0.60
BVAS/GPA (median, IQR)	5 (4-6)	6 (4-7)	5 (4-6.5)	4 (3-6)	6 (4-9)	<0.001
Charlson Comorbidity Index	3 (1-5)	5 (3-7)	4 (2-6)	2 (1-5)	3 (2-5)	<0.001
Diabetes mellitus (N, %)	50 (13)	3 (13)	16 (15)	29 (14)	2 (7)	0.80
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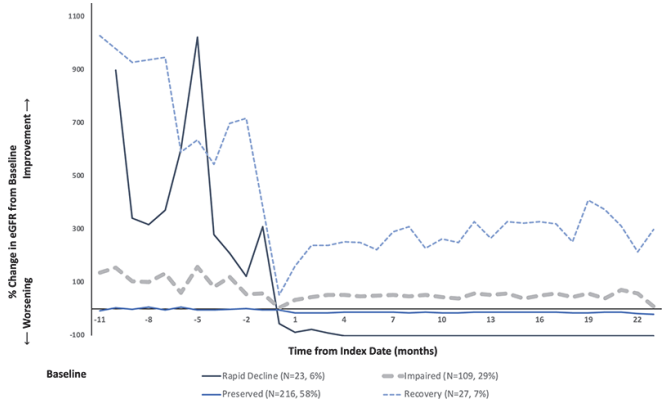

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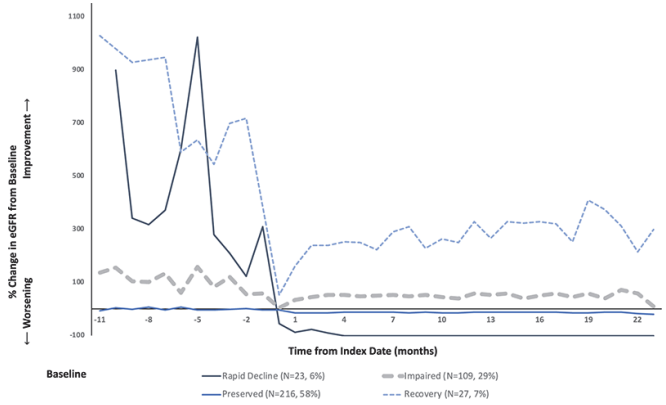

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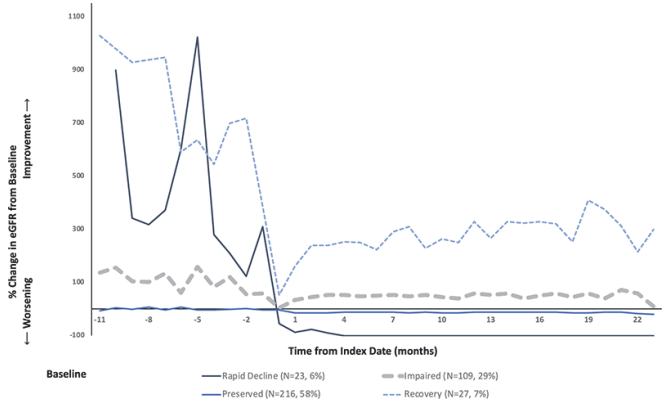

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EFFICACY AND SAFETY OF ABATACEPT IN MYOSITIS ASSOCIATED INTERSTITIAL LUNG DISEASE

R. Aggarwal[1], S. Moghadam-Kia[1], D. Koontz[2], D. Saygin[1], S. Bae[3], D. Sullivan[4], G. Marder[5], S. Venuturupalli[6], P. Dellaripa[7], S. Danoff[8], T. Doyle[9], G. Hunninghake[9], J. S. Lee[10], A. Fischer[11], J. Falk[12], C. R. Kang[13], Y. Lin[14], C. Johnson[15], D. Ascherman[1], C. V. Oddis[1]

POS1240 (2023)

Efficacy and Safety of Abatacept in Myositis Associated Interstitial Lung Disease

University of Pittsburgh

Rohit Aggarwal[1], Siamak Moghadam-Kia[1], Diane Koontz[1], Didem Saygin[1], Sangmee Bae[2], Daniel Sullivan[1], Galina Marder[3], Swamy Venuturupalli[4], Paul Dellaripa[5], Sonye Danoff[6], Tracy Doyle[7], Gary Hunninghake[7], Joyce S. Lee[8], Aryeh Fischer[9], Jeremy Falk[10], Chae Ryon Kang[11], Yan Lin[12], Cheilonda Johnson[13] Dana Ascherman[1], Chester V Oddis[1]

[1]University of Pittsburgh, Department of Medicine, Division of Rheumatology, Pittsburgh, PA, [2]University of California Los Angeles David Geffen School of Medicine, Department of Medicine, Division of Rheumatology, Los Angeles, CA, [3]Northwell Health, Department of Rheumatology, Great Neck, NY, [4]Cedars- Sinai Medical Center, Department of Medicine, Division of Rheumatology, Beverly Hills, CA, [5]Brigham and Women's Hospital BWH, Department of Rheumatology, Boston, [6]John Hopkins Medicine, Department of Medicine, Division of Pulmonary and Critical Care Medicine, Baltimore, [7]Brigham and Women's Hospital BWH, Division of Pulmonary and Critical Care Medicine BWH, Boston, [8]University of Colorado Denver , Department of Medicine, Aurora, CO, [9]Bristol Myers Squibb, Lung Fibrosis - Clinical Development Lead, Lawrenceville, NJ, [10]UCLA Medical Center, Department of Rheumatology, Los Angeles, CA, [11]University of Pittsburgh, Department of Biostatistics, Pittsburgh, [12]University of Pittsburgh, Department of Biostatistics, Pittsburgh, PA, [13]University of Pennsylvania, Department of Medicine, Philadelphia, PA

Background

- Interstitial Lung Disease (ILD) is the most common cause of mortality and morbidity in myositis
- There is lack of randomized clinical trials for myositis-associated ILD (MA-ILD)
- A T-cell mediated pathogenesis has been postulated for MA-ILD
- Abatacept (ABA) is a T-cell co-stimulatory modulator that prevents T-cell activation

Objective

- Evaluation of the efficacy, safety and tolerability of ABA (125 mg SQ weekly) combined with standard of care (SOC) vs. placebo (PBO) with SOC in MA-ILD patients in a multi-center, double- blind, randomized placebo-controlled proof of concept clinical trial

Methods

- 20 patients (pts) with anti-synthetase antibody (Anti-Syn Ab) were enrolled across 5 centers for 24 weeks in a double blind randomized placebo-controlled phase (DB-RCT) followed by an open label (OLE) for 24 weeks.
- Active myositis was not required but pts must have had active ILD (new onset or worsening requiring treatment) and previously failed ≧ 1 SOC drug for ILD.
- Pts were required to be on stable SOC [glucocorticoids (GC)/1 immunosuppressive (IS) agent (azathioprine or mycophenolate) prior to the trial as well as stable SOC IS throughout the trial]. A recommended GC taper was instituted.
- Weekly 125 mg SQ ABA vs. PBO was given for 24 weeks, followed by weekly 125 mg SQ ABA from 24-48 weeks.
- Primary endpoint was the change in Force Vital (FVC) (in ml) from baseline to week 24.
- Secondary endpoints included FVC changes over 48 weeks, patient reported outcomes (PRO) of dyspnea (UCSD-shortness of breath questionnaire; range 0-120, MCID of 8) and DLCO corrected % predicted (DLCO) at 24 and 48 weeks.

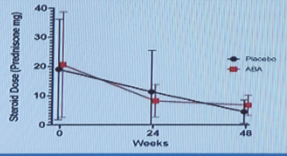
Results

- 20 Ab positive pts were randomized to ABA (n=9) and placebo (n=11)
- All but 1 pts completed 48 weeks of the trial
- 3 and 4 pts required rescue above baseline immunosuppression in PBO vs. 0 and 1 in ABA group 0-24 weeks and 0-48 weeks, respectively.
- There were 3 adverse events (1 in RCT, 2 in OLE) in 2 pts requiring hospitalization in the ABA group for progressive respiratory failure and 1 died (at 24 weeks)
- There were 36 and 23 adverse events amongst 11 placebo and 9 treatment group pts. ABA was well tolerated

Table 1. Baseline demographics and clinical variables by treatment groups

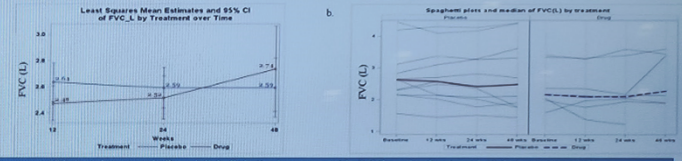
Clinical variable	Placebo (N=11)	Abatacept (N=9)
Age	57.7 [47.8-64.5]	49.7 [46.6-59.3]
Female	4 (36)	5 (56)
Non-Hispanic	11 (100)	9 (100)
Caucasian	10 (91)	7 (78)
Anti-synthetase antibody		
Jo-1	7	4
Non-Jo1	4	5
ILD status		
New onset	3 (27)	2 (22)
Chronic	8 (73)	7 (78)
Myositis status		
Inactive	5 (45)	2 (22)
Active	4 (37)	5 (56)
Not present	2 (18)	2 (22)
FVC%	61 [53-78]	66 [49-69]
DLCO%	49 [41-64]	47 [38-52]
Prednisone	11 (100)	8 (88)
Mycophenolate	8 (73)	8 (88)
Azathioprine	0 (0)	1 (11)

Figure 4. Change in steroid dose (prednisone mg) by treatment over time



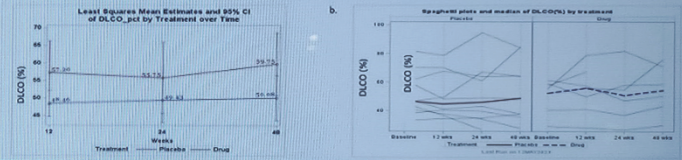
No significant difference was seen in steroid dose reduction between the 2 groups

Figure 1. FVC (L) by treatment over time; a. Least Square Mean controlling for age, gender & baseline FCV, b. Spaghetti plots (median)



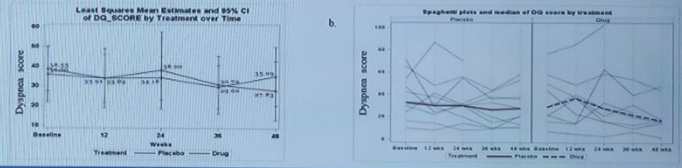
ABA group has numerically and clinically better improvement in FVC than placebo at week 48, but not at week 24.
Median change at week 24: ABA (-40 ml) vs. PBO (30 ml), p 0.97; at week 48: ABA (140 ml) vs. PBO (-40 ml), P = 0.15

Figure 2. a. DLCO (%) by treatment over time: Least Square Mean controlling for age, gender & baseline FVC, DLCO(%); b. Spaghetti plot -(median)



Drug group has numerically more improvement in DLCO (ml) at week 48, but not week 24
Median change at week 24: ABA (-4.5 ml) vs. PBO (-10.5); at week 48: ABA (6.5) vs. PBO (-2.7)

Figure 3. UCSD Dyspnea (DQ_score) by treatment over time, a. Least Square Mean controlling for age, gender & baseline FVC; b. Spaghetti plot



Drug group has numerically and clinically more decrease in UCSD Dyspnea (DQ_score) than placebo
Median change at week 24: ABA (1) vs. PBO (3); at week 48: ABA (13) vs. PBO (5)

Conclusions

- MA-ILD ABA-treated subjects showed similar FVC, DLCO and UCSD-dyspnea trends compared to placebo at week 24, but clinically and numerically meaningful trends were observed in ABA-treated subjects at week 48
- These results suggest the need for a larger randomized study of ABA in MA-ILD with outcomes at 1 year
- Abatacept was relatively safe and well tolerated in the cohort

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POS1240 (2023)

Objetivos:

- evaluar eficacia, seguridad y tolerabilidad ABA/SOC vs placebo/SOC en MA-ILD

Métodos:

- multicéntrica, doble ciego, aleatorizado, controlado con placebo
- 5 centros/24 s (RCT→OLE)
- ILD activa + fallo ≥ 1 SOC
- Endpoint 1º: cambio CVF inicio → s24

Resultados:

- 20 pc Ac anti-Syn
- edad media 57 años, 45% ♀ y 55% Jo-1

Table 1. Baseline demographics and clinical variables by treatment groups:

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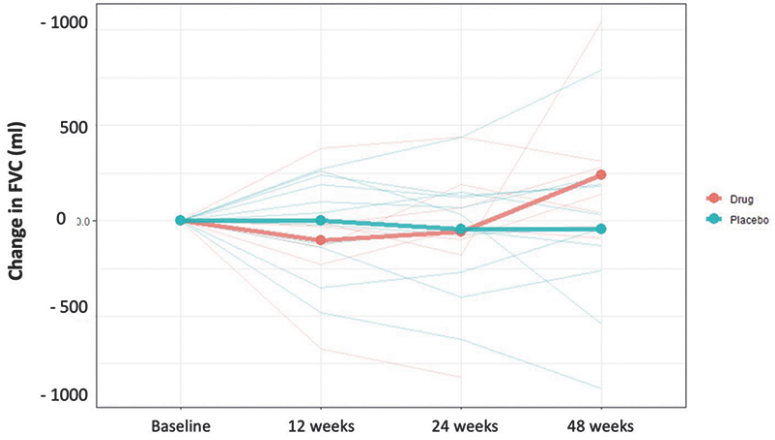
Figure 1: Change from baseline FVC in ml by treatment groups.



	24 S		48 S	
	ABA (n=9)	**Placebo** (n=11)	**ABA** (n=9)	**Placebo** (n=11)
cambio medio de DLCO	-4.5	-10.5	6,5	-2,7
Mejoría disnea mediana	1	3	13	5

Resultados:

- **Eventos adversos:**
  - 36 en 9 pc placebo
  - 23 en 9 pc ABA
  - 3 graves en 2 pc ABA: 1 falleció 24 s

Conclusión

- MA-ILD tto ABA tendencias similares FVC, DLCO y disnea vs placebo a 24 s pero tendencias clínica y numéricamente significativas en ABA a 48s.
- Necesidad estudio aleatorizado más grande de ABA en MA-ILD.
- ABA fue relativamente seguro y bien tolerado.

CLINICAL FEATURES AND TREATMENT-RELATED OUTCOMES OF IGG4-RELATED DISEASE FROM A LARGE EUROPEAN STUDY COHORT

M. Lanzillotta[1], K. Overbeek[2], J. Poulsen[3], O. Vinge Holmquist[4], P. Macinga[5], P. Prescraip Study Group[6], M. Lohr[7], J. Rosendahl[8], E. Della Torre[9]

Objetivos:

- Caracterizar epidemiología, clínica y respuesta tto de los pacientes con IgG4-RD en Europa.

Métodos:

- **Registro paneuropeo** (PrescrAIP) (**42 hospitales**), análisis retrospectivo adultos IgG4-RD 2005-2020.
- Criterios diagnósticos integrales + específicos órganos
- Datos: registros médicos, formulario registro REDCap.
- **Índice respuesta IgG4-RD**
- **predictores recaída**: análisis multivariable regresión logística

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Resultados:

- 1079 sospecha IgG4-RD → **735** dco **IgG4-RD** (69% ♂; mediana edad 57 años; 85% caucásicos).
- **45%** afectación **multiorgánica** (≥2)(páncreas, gl salivales y vía biliar)
- **Tto esteroides** 634 pc (**86,25%**)
- **dosis diaria +** ↑ (PD>0,4 mg/kg) **=** eficaz **dosis +** ↓ (<0,4 mg/kg) (OR 0,428; IC95% 0,054-3,387) **inducir** respuesta.
- **inducción + prolongada** (>2 s) **=** eficaz terapia **+ corta** (<2 s) (OR 0,908; IC95 %: 0,818-1,009).
- niveles ↑ **IgG4** asociación independiente **< probabilidad RC** (OR 0,639; IC 95% 0,427-0,955)
- **30% recaidas**:
  - **6m post-inducción** independientes de: **duración tto inducción, duración** ↓ gradual **GC, dosis** total **acumulada**.
- ↑ **tamaño parénquima** (OR 0,390, IC 95% 0,167-0,910) y terapia mantenimiento **IS** asociación independiente con **< recaídas 6 m** (OR 0,299, IC 95% 0,120-0,740). No
- **No ≠ GC** y **RTX mantenimiento** respuesta

Table 1. Characteristics of IgG4-RD patients at diagnosis. *ULN, upper limit of normal.*

Patient characteristics	Total (N=735)
Male sex	509 (69)
Age, median (IQR), y	57 (27)
Diabetes mellitus	213 (29)
Pancreatic exocrine insufficiency	190 (26)
Symptoms	
Incidental finding	32 (4)
Obstructive jaundice	381 (52)
Abdominal pain	471 (64)
Weight loss	270 (37)
Acute pancreatitis	76 (10)
Other organ involvement	329 (45)
Orbit	12 (2)
Bilateral salivary glands	54 (7)
Thyroid	13 (2)
Pulmonary	38 (5)
(Peri)aorta	17 (2)
Retroperitoneal fibrosis	24 (3)
Sclerosing cholangitis/ biliary tree	262 (36)
Renal	49 (7)
IgG4 > 1x ULN	440 (60)

	Tratamiento (625)	NO tratamiento (95)
RC	79%	61%
RP	18%	19%
NO R	3%	10%

Conclusión

- 1º estudio paneuropeo epidemiología y clínica IgG4-RD en Europa.
- niveles ↑ IgG4: control + estricto durante inducción
- estrategia + eficaz inducir remisión: PD 0,4 mg/kg/día/ 2 s

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