REVISIÓN DE MIOPATIAS AUTOINMUNES

Systemic Autoimmune Diseases Unit

VHIR

Albert Selva O'Callaghan

Vall d'Hebron Hospital

XUEC

European Reference Networks

CENTROS, SERVICIOS Y UNIDADES DE REFERENCIA (CSUR) DEL SISTEMA NACIONAL DE SALUD

UAB Universitat Autònoma de Barcelona

Accepted

Anti-cortactin autoantibodies are associated with key clinical features in adult myositis but are rarely present in juvenile myositis.

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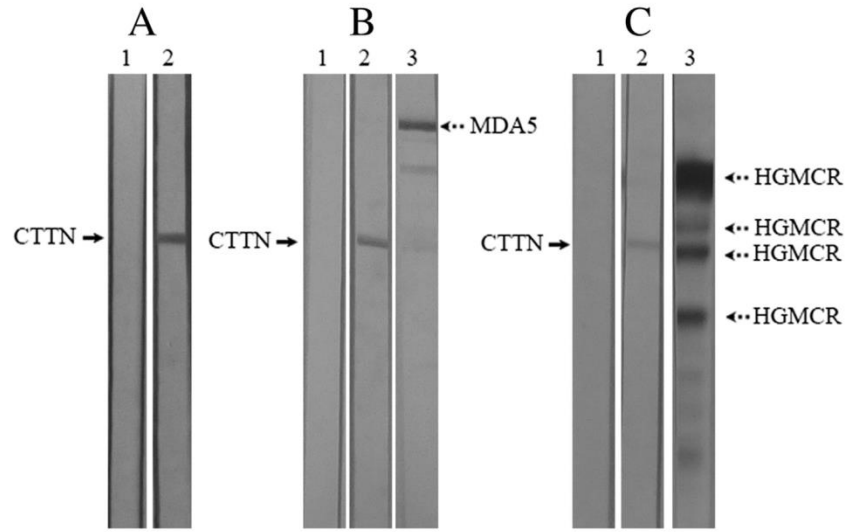

Labrador-Horrillo M, Martínez MA, Selva-O'Callaghan A, et al. Identification of a novel myositis-associated antibody directed against cortactin. Autoimmun Rev. 2014

Adultos con DM (NXP2 o Mi2)
Disfagia + ILD

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Systematic review and meta analysis

A systematic review and meta-analysis to inform cancer screening guidelines in idiopathic inflammatory myopathies

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Risk of malignancy

Dermatomyositis subtype (RR 2.21)

Older age (WMD 11.19)

Male sex (RR 1.53)

Dysphagia (RR 2.09)

Skin ulceration (RR 2.73)

Anti-TIF1γ (RR 4.66)

Protective effect
PM (RR 0.49)
CADM (0.44)
Raynaud's phenomenon (RR 0.61)
ILD (0.49)
High serum CK / LDH
Anti-Jo1 (RR 0.45)
Anti-EJ (RR 0.17)

Rheumatology key messages

- IIM cancer risk factors include dermatomyositis, older age, male sex, dysphagia, cutaneous ulceration and anti-TIF1γ.
- Cancer screening evidence is limited but CT scans may be useful.
- Findings from this study can inform IIM-specific cancer screening guidelines.

Current Rheumatology Reports (2021) 23: 63
https://doi.org/10.1007/s11926-021-01023-9

INFLAMMATORY MUSCLE DISEASE (L DIEDERICHSEN AND H CHINOY, SECTION EDITORS)

COVID-19 and Myositis: What We Know So Far

Ahmad Saud[1] · R Naveen[2] · Rohit Aggarwal[3] · Latika Gupta[2]

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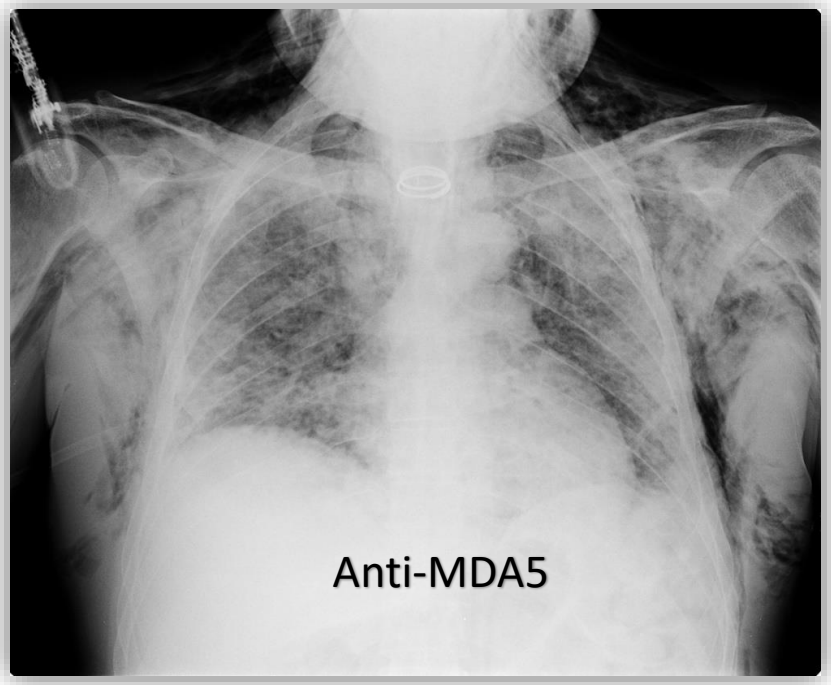
Miositis vírica por invasión del miocito

Miopatía autoinmune (IMNM)

Rabdomiolisis

Dermatomiositis…



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RESPIRATORY INVESTIGATION XXX (XXXX) XXX

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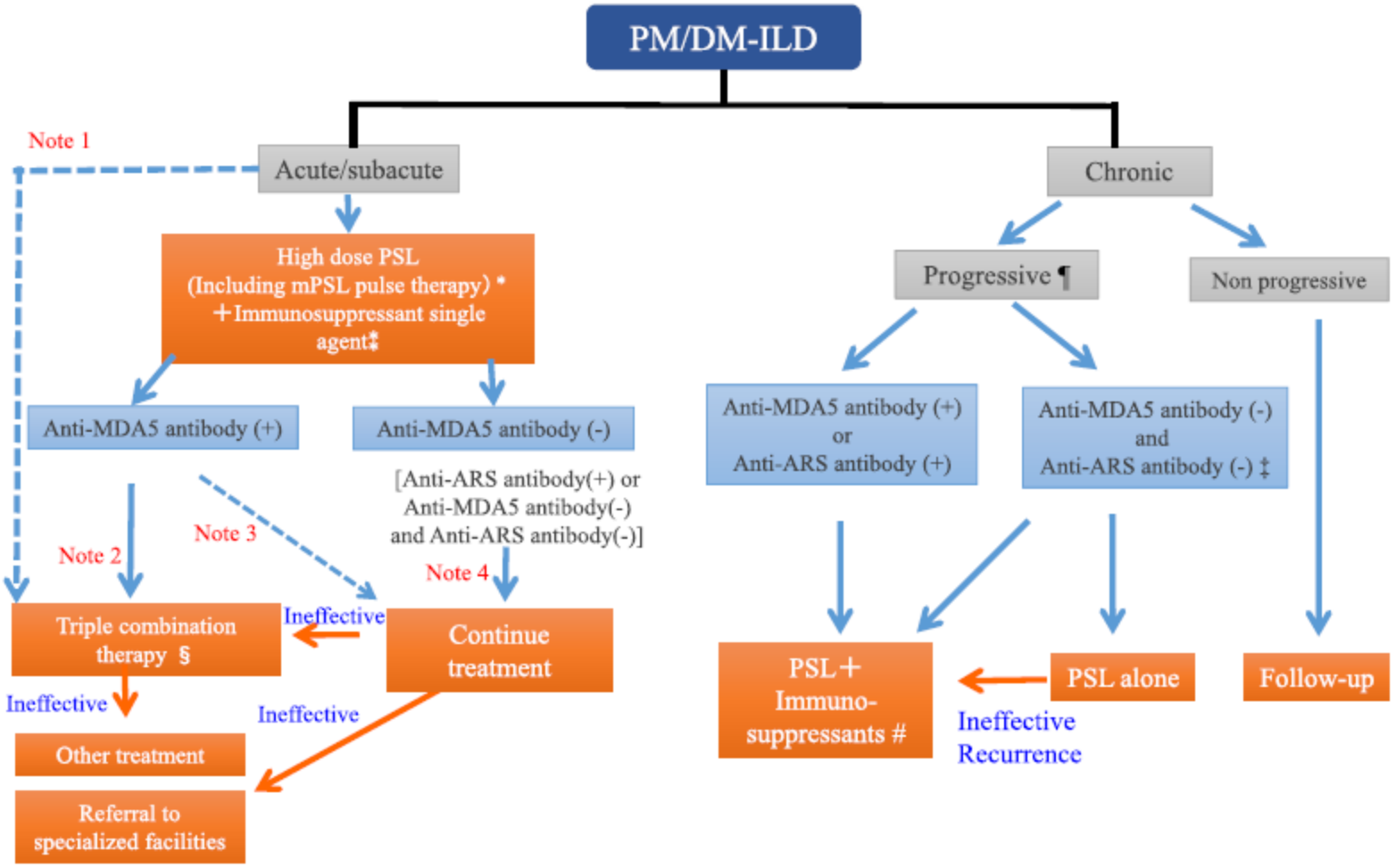
Respiratory Investigation

journal homepage: www.elsevier.com/locate/resinv

Guideline and Statement

2020 guide for the diagnosis and treatment of interstitial lung disease associated with connective tissue disease☆

Yasuhiro Kondoh [a,*], Shigeki Makino [b], Takashi Ogura [c], Takafumi Suda [d], Hiromi Tomioka [e], Hirofumi Amano [f], Masaki Anraku [g], Noriyuki Enomoto [h], Takao Fujii [i], Tomoyuki Fujisawa [d], Takahisa Gono [j], Masayoshi Harigai [k], Hidenori Ichiyasu [l], Yoshikazu Inoue [m], Takeshi Johkoh [n], Hideto Kameda [o], Kensuke Kataoka [a], Yasuhiro Katsumata [k], Yasushi Kawaguchi [k], Atsushi Kawakami [p], Hideya Kitamura [c], Noboru Kitamura [q], Tomohiro Koga [p], Kazuhiro Kurasawa [r], Yutaro Nakamura [d], Ran Nakashima [s], Yasuhiko Nishioka [t], Osamu Nishiyama [u], Masaki Okamoto [v], Fumikazu Sakai [w], Susumu Sakamoto [x], Shinji Sato [y], Toshimasa Shimizu [p], Noboru Takayanagi [z], Reoto Takei [a], Tamiko Takemura [aa], Tohru Takeuchi [ab], Yuko Toyoda [ac], Hidehiro Yamada [ad], Hideaki Yamakawa [ae], Yasuhiko Yamano [a], Yoshioki Yamasaki [j], Masataka Kuwana [j], on behalf of the joint committee of Japanese Respiratory Society and Japan College of Rheumatology


PM/DM-ILD
Note 1
Acute/subacute
Chronic
High dose PSL
(Including mPSL pulse therapy) *
+Immunosuppressant single agent‡
Progressive ¶
Non progressive
Anti-MDA5 antibody (+)
Anti-MDA5 antibody (-)
Anti-MDA5 antibody (+)
or
Anti-ARS antibody (+)
Anti-MDA5 antibody (-)
and
Anti-ARS antibody (-) ‡
[Anti-ARS antibody(+) or
Anti-MDA5 antibody(-)
and Anti-ARS antibody(-)]
Note 2
Note 3
Note 4
Triple combination therapy §
Ineffective
Continue treatment
Ineffective
Ineffective
Other treatment
Referral to specialized facilities
PSL+
Immuno-
suppressants #
PSL alone
Follow-up
Ineffective
Recurrence

Seminars in Arthritis and Rheumatism 50 (2020) 776–790

Contents lists available at ScienceDirect

Seminars in Arthritis and Rheumatism

journal homepage: www.elsevier.com/locate/semarthrit

Recommendations for the treatment of anti-melanoma differentiation-associated gene 5-positive dermatomyositis-associated rapidly progressive interstitial lung disease

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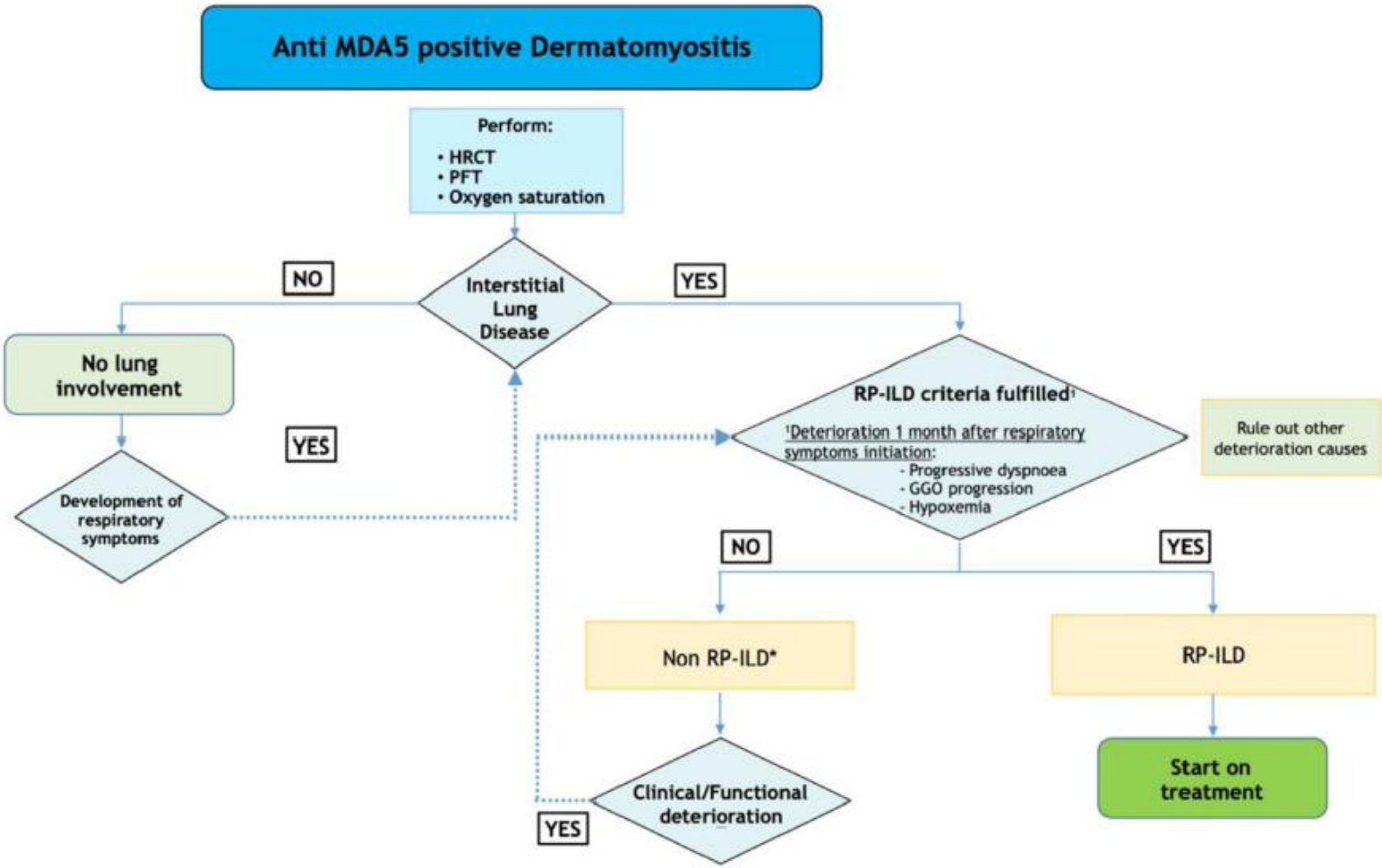
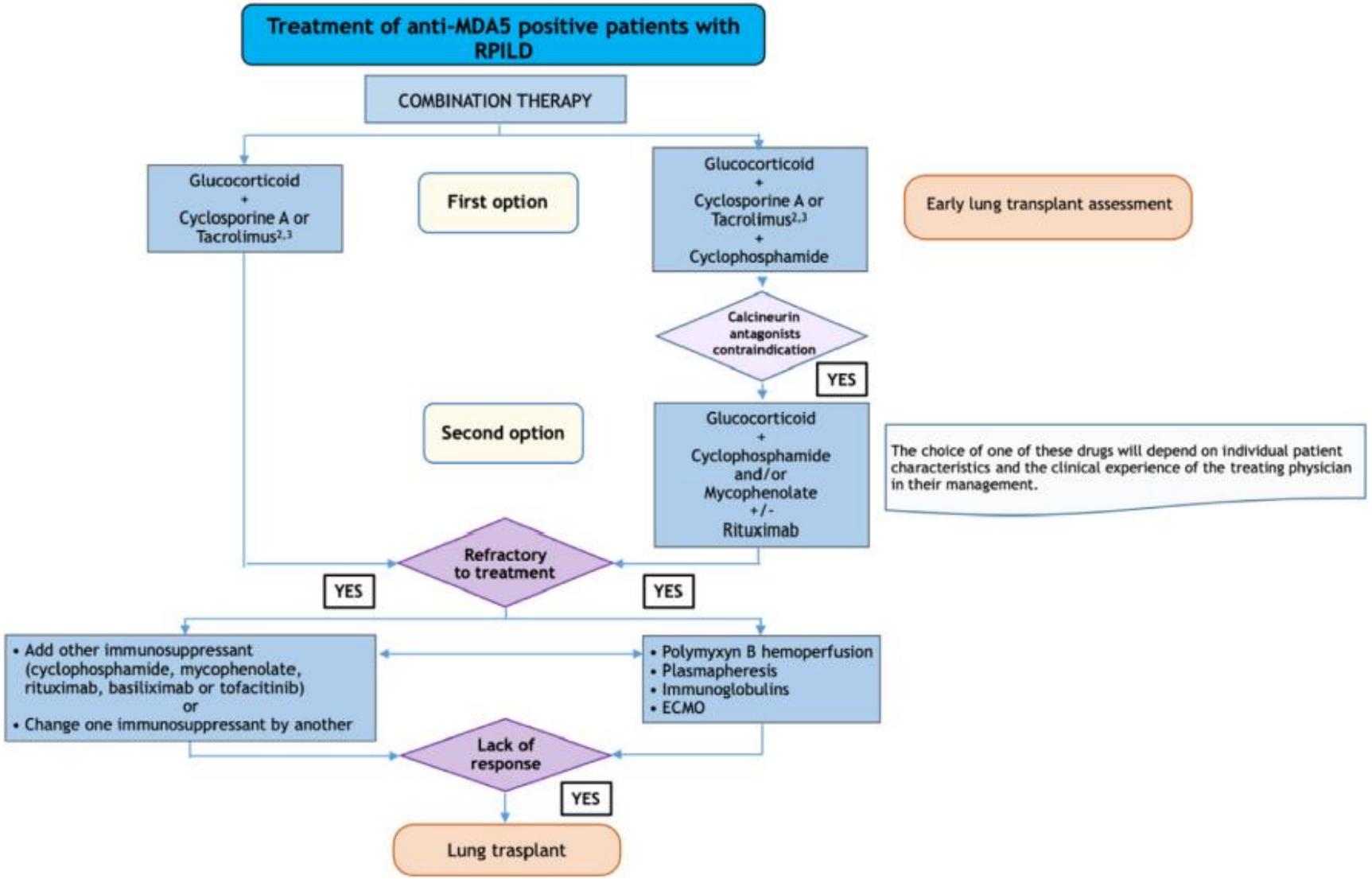

Anti MDA5 positive Dermatomyositis
Perform:
• HRCT
• PFT
• Oxygen saturation
Interstitial Lung Disease
NO
YES
No lung involvement
YES
Development of respiratory symptoms
RP-ILD criteria fulfilled[1]
[1]Deterioration 1 month after respiratory symptoms initiation:
- Progressive dyspnoea
- GGO progression
- Hypoxemia
Rule out other deterioration causes
NO
YES
Non RP-ILD*
RP-ILD
Clinical/Functional deterioration
YES
Start on treatment


Treatment of anti-MDA5 positive patients with RPILD
COMBINATION THERAPY
Glucocorticoid + Cyclosporine A or Tacrolimus[2,3]
First option
Glucocorticoid + Cyclosporine A or Tacrolimus[2,3] + Cyclophosphamide
Early lung transplant assessment
Calcineurin antagonists contraindication
YES
Second option
Glucocorticoid + Cyclophosphamide and/or Mycophenolate +/- Rituximab
The choice of one of these drugs will depend on individual patient characteristics and the clinical experience of the treating physician in their management.
Refractory to treatment
YES
YES
• Add other immunosuppressant (cyclophosphamide, mycophenolate, rituximab, basiliximab or tofacitinib) or • Change one immunosuppressant by another
• Polymyxyn B hemoperfusion • Plasmapheresis • Immunoglobulins • ECMO
Lack of response
YES
Lung trasplant

JUNTOS SOMOS MEJORES MEDICOS

…ORGANIZADOS SOMOS INSUPERABLES