Síndrome pulmón-riñón

Belén Safont

Sd.pulmón-riñón (SPR)

Combinación de hemorragia alveolar difusa y glomerulonefritis

- Causas:
  - vasculitis primarias ANCA+ 60-70% y enf. Ac-AMBG 10- 20%
  - Otras: LES, vasculitis ANCA negativos y asociadas a fármacos/drogas
- Mal pronóstico: diagnóstico y tratamiento precoz
  - Mortalidad (etiología): 25-70% (actual 30-40%)
  - Dependencia de diálisis: 70-80%

Rodriguez W. Crit Care Clin 2002;18:881-95
Papiris S. Crit Care 2007;1:213-24

CLASIFICACIÓN POR MECANISMOS PATOGÉNICOS

Síndrome pulmón-riñón. Clasificación de acuerdo a los mecanismos patogénicos que intervienen

1) En vasculitis sistémica ANCA positivos
   a) Granulomatosis de Wegener
   b) Poliangeítis microscópica
   c) Síndrome de Churg-Strauss
   d) Síndrome pulmón- riñón ANCA positivo idiopático — HAD y GN sin otras manifestaciones de vasculitis sistémica
2) Asociado con Ac-AMBG
   a) Síndrome de Goodpasture
3) En enfermedades autoinmunes sistémicas
   a) Lupus eritematoso sistémico — HAD 2-5.4% + nefritis lúpica
   b) Esclerosis sistémica
   c) Artritis reumatoidea
   d) Enfermedad mixta del tejido conectivo
   e) Polimiositis
4) En vasculitis sistémica ANCA negativos
   a) Púrpura de Scholein-Henoch — Vaso pequeño. Púrpura palpable, artralgias o artritis, dolor abdominal
   b) Crioglobulinemia mixta — Vasos mediano y pequeño. Púrpura, artritis, hepatitis, neuropatía
   c) Enfermedad de Behcet — Vasos grandes, medianos y pequeños. Estomatitis aftosa recurrente, úlceras genitales y uveitis.
5) En vasculitis ANCA positivos por drogas — MPO y PR3 +
   a) Propiltiouracilo
   b) D-Penicilamina
   c) Hidralazina
   d) Allopurinol
   e) Sulfasalazina
   f) Carbimazol — h) Cocaína/levamisol
   g) Fenitoína — i) Minociclina

   Vasculitis vaso pequeño. HAD 2º a capilaritis + GNproliferativa necrotizante
   Intervalo exposición y SPR: horas a años
   Menos graves
6) Asociado con Ac-AMBG y ANCA positivos.

7) microangiopatía trombótica — APS catastrófico

púrpura trombocitopénica trombótica

Santos-Ocampo AS. Chest 2000:118:1083-90
Naniwa T. Mod Rheumatol 2007;17:37-44
Martin K. Dis Mon. 2022;68:101465

Vasculitis ANCA positivos

- Causa más frecuente del SPR (60%- 70%)

Patrón IFI	Antígeno
C-ANCA: granular citoplásmico difuso	PR3: proteinasa 3
P-ANCA: perinuclear	MPO: mieloperoxidasa
Atípico: generalmente p-ANCA	No se asocian a Ag específico

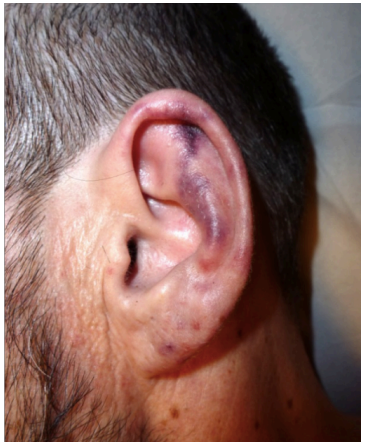
AAV	GPA	MPA	GEPA
Incidencia (millón de personas/ año)	4.9-10.5	2.6-11.6	0.5- 4.2
Edad (media)	40-55	50-60	48
Afectación pulmonar	NPs HAD 10%	HAD 30%	Asma HAD rara
Otros órganos	Cutánea, VA ocular, SNP	Cutánea SNP	Eo periférica SNP

- SPR- ANCA positivos idiopático

Niles JL. Arch Intern Med 1996;154:440-5

SPR por cocaína adulterada con levamisol

- Vasculitis ANCA (PR3 y MPO dual): vasos pequeños
- Otros marcadores: Ac antifosfolípidos, ANA, anti dsDNA, Ac anti-elastasa de neutrófilos humanos (HNE) y crioglobulinas
- Revisión sistemática: 8 casos, edad 22 -58 años, 50% mujeres
- Afectación cutánea 50% (80% púrpura retiforme), ¾ lesiones cutáneas necróticas (especialmente en en hélix auricular) casi patognomónicas
- 63% SPR manifestación inicial



Bucur P. Pharmaceuticals (Basel). 2023 Jun 6;16(6):846
Vaezi Z. Am J Case Rep. 2026 Apr 17;27:e951159

Enfermedad anti-MBG o Goodpasture

- Ac anti-MBG se unen a la cadena α-3-colágeno tipo IV activando la cascada del complemento → inflamación y daño tisular
- Incidencia: 1- 1.64 millón de personas/año
- Edad: 2º a 3º década y 6ª a 7ª década
- FR: Predisposición genética, exposición tabaco, hidrocarburos e infecciones
- Biopsia: IF depósito lineal de IgG
  - nº glomerulos afectados y > afectación intersticial: necesidad de HD crónica
  - glomérulos normales (<10%) y/o 100% de glomérulos con semilunas: evolución
- Pronóstico: Supervivencia 5 años: >80%, HD: < 1/3 requieren diálisis
- 50% SPR, < síntomas constitucionales, hematuria macroscópica <25%

Martin K. Dis Mon. 2022;68:101465
Bharati J. Adv Kidney Dis Health 2024;31:206-215

Variantes de presentación

Atypical anti-GBM disease[30,56]	~5-10% of all Cases of anti-GBM disease have absent circulating anti-GBM antibodies Mild clinical and/or histopathological presentation Management: § Exclude cases of IgG4-, IgA-, and IgM-mediated anti-GBM disease using modified assays § Thorough evaluation for atypical IgG antibodies using highly sensitive assays and modified assays targeting newer epitopes/antigens
Dual anti-GBM antibody and anti-MPO antibody positive disease[21,22,37,53]	~20-40% of all cases of anti-GBM disease Older age and more systemic manifestations than classic anti-GBM disease Relapse commoner than classic disease Outcomes similar to classic disease Management: § Initial phase is similar to classic anti-GBM disease; § Maintenance immunosuppression to prevent relapse akin to AAV
Drug-induced anti-GBM disease[57–68]	1. Anti-CD52 monoclonal antibody (**alemtuzumab**) is associated with the classic anti-GBM disease after 9-10 months of administration in genetically susceptible patients 2. TNF-alpha antagonists (**etanercept, adalimumab**) are associated with classic anti-GBM disease. The exact pathogenesis is unclear as TNF-alpha blockers are known to prevent/resolve anti-GBM GN in animal models. 3. Immune checkpoint inhibitors: Anti-programmed death-1 (**nivolumab, pembrolizumab**), CTLA4 antagonist (**tremelimumab**), kinase inhibitors (**dabrafenib, trametinib**) are associated with classic and atypical forms of anti-GBM disease 4. SARS-CoV-2 vaccines: Among all other de novo primary GNs reported, anti-GBM disease is rare after this vaccine. All types of SARS-CoV-2 vaccines such as **mRNA**, **Pfizer-BioNtech**, and **AstraZeneca** are associated with classic anti-GBM disease Management: Drug withdrawal + treatment similar to classic anti-GBM disease

Bharati J. Adv Kidney Dis Health 2024;31:206-215

Vasculitis por IgA (púrpura de Henoch-Schönlein)

- Vasculitis por inmunocomplejos con depósitos predominantes de IgA: vasos pequeños.
- Incidencia: adultos 5/100.000 adultos
- Desencadenada por infecciones VAS, estreptocócicas
- Púrpura: Superficies extensoras y zonas declives
- Artralgias: rodillas y tobillos
- Dolor abdominal tipo cólico, 1/3 vómitos y HGI
- Adultos: edemas e hipertensión

ANKARA 2008

Criterion	Definition
Skin involvement (mandatory criterion)	▪ Petechiae or purpura, painful skin lesions of any type, maculopapular or papular rash
Abdominal pain 1)	▪ Diffuse abdominal colicky pain with acute onset assessed by history and physical examination. May include intussusception and gastrointestinal bleeding
Histopathology 2)	▪ Typically leukocytoclastic vasculitis with predominant IgA deposits *or* ▪ Proliferative glomerulonephritis with predominant IgA deposits
Arthritis or arthralgia 3)	▪ Arthritis of acute onset defined as joint swelling or joint pain with limitation on motion ▪ Arthralgia of acute onset defined as joint pain without joint swelling or limitation on motion
Renal involvement 4)	▪ Proteinuria >0.3 g/24 h or >30 mmol/mg of urine albumin/creatinine ratio on a spot morning sample *and/or* ▪ Hematuria or red blood cell casts: >5 red blood cells/high power field or red blood cells casts in the urinary sediment or ≥2+ on dipstick
Exclusion criteria	▪ Positive test for ANCA (cANCA or pANCA on IF, PR3-ANCA ELISA, MPO ANCA ELISA) *or* ▪ Positive test for blood cryoglobulins

afectación cutánea + ≥1 (1-4)

Reamy BV. Am Fam Physician. 2020 Aug 15;102(4):229-233
Bayindir Y.RMD Open. 2025 Jul 21;11(3):e005728

Crioglobulinemia mixta

- Vasculitis mediada por inmunocomplejos (vasos pequeños y medianos)
- Triada clásica: artralgias, púrpura y neuropatía periférica (debilidad)
- 5% SPR
- FR: infecciones (VHC; VHB, VIH), asociadas de ETC (LES; AR, Sjögren), enf linfoproliferativa
- Diagnóstico: identificación de crioglobulinas séricas (análisis 37ºC), FR+, ↓ C4
- Tratamiento: plasmaféresis + bolos MPD → CS + RTX 1v/semana 4 semana
- Tratamiento etiológico: VHC, ETC

Galli M. Autoinmun Rev 2019;18:778-785

Infecciones: desencadenante

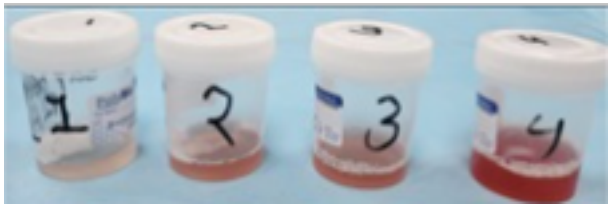
- Desencadenante: vasculitis ANCA+, crioglobulinemia mixta, vasculitis por IgA SAF, PTT
- DD complicado: infiltrados pulmonares + insuficiencia renal aguda
- Si sospecha: ATB empíricos
- Complicación en el curso de la enfermedad (IS)

Martin K. Dis Mon. 2022;68:101465

Presentación clínica y diagnóstico

- Pródromo infección viral/bacteriana
- Δ pulmonar: disnea, tos, fiebre, hemoptisis (1/3), IRAP
- Δ Renal: no oliguria inicialmente, hematuria
- Analítica: anemia, Cr, proteinuria en rango nefrítico, hematuria y cilindros urinarios

Disorder	Diagnostic investigation
ANCA-associated vasculitis	ANCA - MPO and PR3 by immunoassay and IF
Anti-GBM disease	Anti-GBM antibody by ELISA
Autoimmune connective tissue disorders	ANA, Anti ds DNA
SLE	Scl 70
Scleroderma	RA, Anti-CCP
Rheumatoid arthritis	Anti U1RNP
Mixed collagen vascular disease	
Cryoglobulinemia	Cryoglobulin titers
Antiphospholipid syndrome	Antiphospholipid antibodies



FBC: macrófagos cargados de hemosiderina >20 % (Se 100 % y Sp 91,6)

Martin K. Dis Mon. 2022;68:101465

Pronóstico

- Enfermedad rápidamente progresiva
- Mortalidad 30-40%
- Pacientes que sobreviven a la fase aguda: 70%-80% diálisis (1-2 a)

Table 2. Factors associated with severity of disease in pulmonary renal syndrome.

Factors associated with severity of disease in pulmonary renal syndrome	>50% crescents on kidney biopsy[140]
	Proteinuria >1g/day
	Creatinine on presentation
	Increased interstitial infiltrate on kidney biopsy [26]

Martin K. Dis Mon. 2022;68:101465

MANEJO DE SOPORTE

- Soporte respiratorio: O2, OAF, IOT-VMI
- ECMO en centros expertos
- Soporte renal:

Terapia de reemplazo renal

Table 3. Indications for kidney replacement therapy in pulmonary renal syndrome.

Indications for kidney replacementtherapy in pulmonary renalsyndrome	Refractory volume overload
	Uremia (pericarditis, encephalopathy, altered mental status)
	Hyperkalemia ([K+] >6.5 mEq/L, or rapidly rising [K+])
	Metabolic acidosis (pH <7.1)

- TP renal:
  - AAV ≥ 6m del inicio del tto, Goodpasture: 6m con Ac.anti-MBG −
  - Recaída: 1.9-4% Goodpasture, <10% AAV

Martin K. Dis Mon. 2022 Dec;68(12):101465
Koirala A. J Clin Med 2025 Sep 22;14(18):668

VASCULITIS ANCA +: TRATAMIENTO DE INDUCCIÓN (EULAR 2022)

- Objetivo: controlar rápidamente la inflamación y prevenir el daño orgánico irreversible

GC + rituximab o CYC

- Bolos MPD 1-3d: acumulado 1-3g (FG <50 ml/min/1.73m2 y/o HAD)
- Dosis inicial 50-75mg de prednisolona, ↓gradual hasta 5mg a los 4-5m

- Avacopan (inhibidor del R-C5a): 30 mg dos veces al día como estrategia para reducir la exposición a GC (6-12m) ensayo ADVOCATE

- No se recomienda el uso rutinario de plasmaféresis (PLEX)
  - Considerar Cr sérica > 3,4 mg/dL (> 300 µmol/L)
  - Superposición de anti-MBG/AAV
  - KDIGO: Cr > 4mg/dl, ER rápidamente progresiva o HD o HAD con hipoxemia

- Inducida por fármacos /cocaína: Terapia de inducción y cese de consumo. No necesario tratamiento de mantenimiento

GEPA: GC + CYC (alternativa rituximab)

Chung SA. Arthritis Rheumatol. 2021 Aug;73(8):1366-1383
Hellmich B. Ann Rheum Dis. 2024 Jan 2;83(1):30-47
KDIGO. Kidney Int. 2024 Mar;105(3S):S71-S116

REDUCCIÓN DE CORTICOIDES

- Ensayo PEXIVAS: GC en dosis bajas no inferiores al régimen estándar de esteroides

	Body weight (kg)	Metilprednisolona	
Weeks	<50	50–75	>75
1*	50	60	75
2	25	30	40
3–4	20	25	30
5–6	15	20	25
7–8	12.5	15	20
9–10	10	12.5	15
11–12	7.5	10	12.5
13–14	6	7.5	10
15–18	5	5	7.5
19–52	5	5	5
>52	Individual taper	Individual taper	Individual taper

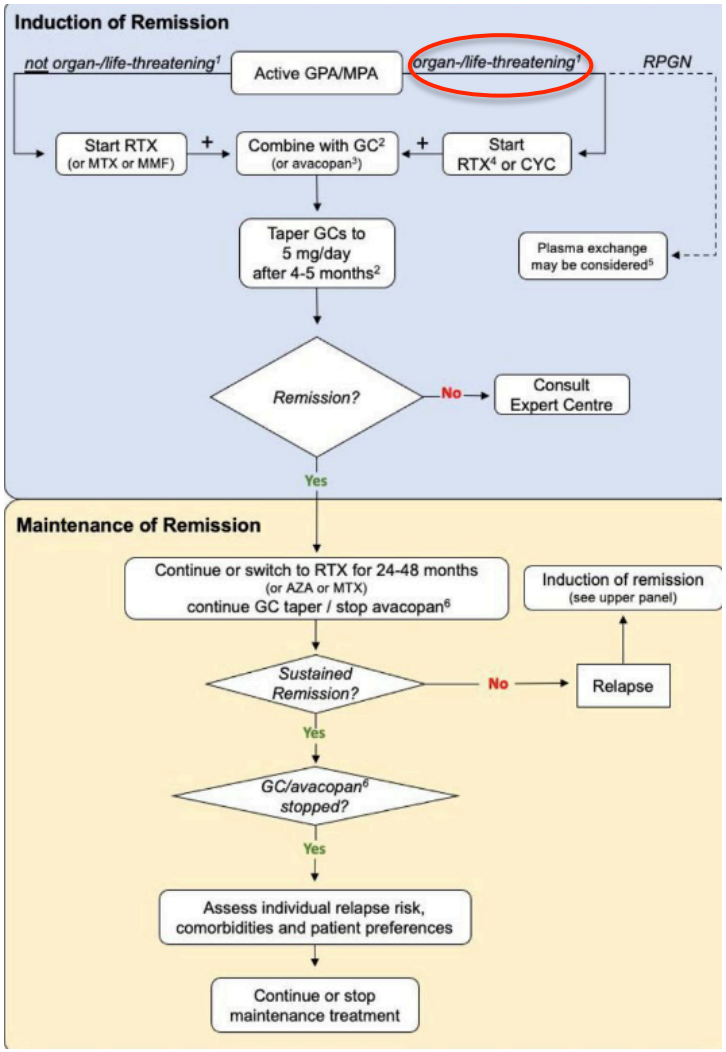
Hellmich B. Ann Rheum Dis. 2024 Jan 2;83(1):30-47
Antonelou M. Scand J Rheumatol 2022;51:135–41

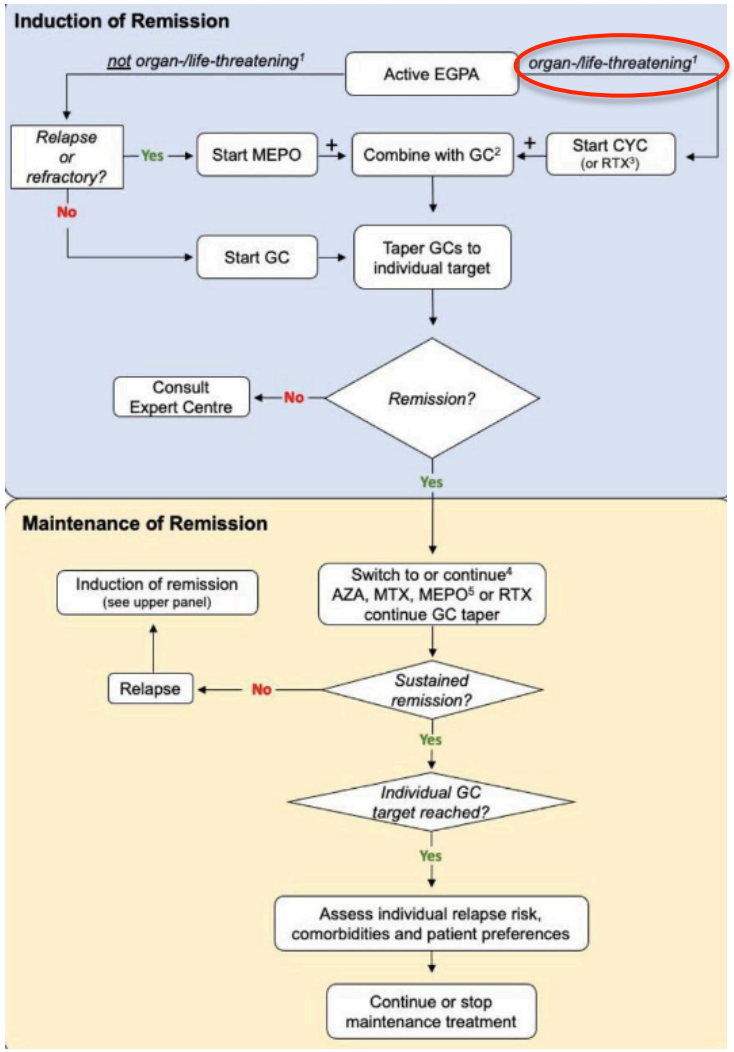
TRATAMIENTO DE MANTENIMIENTO

- Objetivo: mantener la remisión
- Mantenimiento: rituximab (< riesgo de recaída)
  - Alternativas: azatioprina, metotrexato
- Duración de la terapia: 24-48m tras la inducción de la remisión (48m// 24m < R. de recaída)
- GEPA: azatioprina, MTX, mepolizumab o RTX

- Medición de Igs séricas antes de cada ciclo de RTX para detectar ID 2º
- Profilaxis Pneumocystis jirovecii
- Si CYC: análisis periódicos de orina (hematuria)

EULAR recommendations for the management of ANCA-associated vasculitis: 2022 update.





Hellmich B. Ann Rheum Dis. 2024 Jan 2;83(1):30-47

Estudios relevantes:

	Tested therapies	Key exclusion criteria	Remission rates (%) or endpoints	Serious side-effect rates	Daily glucocorticoid* exposure (g)	Conclusions
CYCLOPS (n=149)[65]	Intravenous vs oral cyclophosphamide	Serum creatinine ≥ 5·7 mg/dL	88·1% vs 87·7% at 9 months	25·0% vs 42·5%	7·6 vs 7·6	Similar remission rates, fewer side-effects with less cyclophosphamide exposure, and more relapses in patients with PR3-ANCA-associated vasculitis and those receiving intravenous cyclophosphamide
CORTAGE (n=104)†[92]	Low-dose intravenous cyclophosphamide vs standard dose	NR	89% vs 86% (after induction)	60% vs 78%	5·2 vs 8·3	In patients with an average age of 75 years, a reduced dose cyclophosphamide and glucocorticoid regimen is safer and similarly effective to a standard dose regimen
RAVE (n=197)[3,71]	Rituximab vs oral cyclophosphamide followed by azathioprine	Serum creatinine ≥ 4·0 mg/dL; alveolar haemorrhage requiring intubation	64% vs 53% at 6 months; 39% vs 33% at 18 months	42% vs 38%	4·6 vs 5·1	Non-inferiority of rituximab at 6 months, more effective in patients with relapsing disease (especially PR3-ANCA-associated vasculitis vs MPO-ANCA-associated vasculitis)
RITUXVAS (n=44)[76]	Rituximab plus cyclophosphamide vs intravenous cyclophosphamide‡	Previous use of cyclophosphamide	76% vs 82% at 12 months	42% vs 36%	0·071 vs 0·082§	Rituximab plus cyclophosphamide was not superior to intravenous cyclophosphamide
LoVAS (n=134)[73]	Reduced dose (0·5 mg/kg body weight) glucocorticoids vs high dose (1·0 mg/kg body weight) glucocorticoids	eGFR <15 mL/min per 1·73 m²; alveolar haemorrhage (oxygen inhalation >2 L/min)	71·0% vs 69·2% at 6 months	13% vs 24%	1·3 vs 4·2¶	Rituximab with a reduced dose glucocorticoid regimen is non-inferior in patients with MPO-ANCA-associated vasculitis and eGFR ≥45 mL/min per 1·73 m² (in Japan); reduces risk of severe adverse events
ADVOCATE (n=330)[77]	Avacopan with reduced dose (20 mg/day tapered off over 4 weeks) glucocorticoids vs standard dose (60 mg/day starting dose) glucocorticoids	eGFR <15 mL/min per 1·73 m²; alveolar haemorrhage requiring invasive ventilation	72·3% vs 70·1% at 6 months; 65·7% vs 54·9% at 12 months	23·5% vs 25·0%	1·3 vs 3·7	Avacopan is non-inferior to prednisone at 6 months, and superior at 12 months

No inferioridad RTX // CYC, > eficaz tras recaída PR3

RTX + dosis GC reducidas: misma eficacia y < efectos adversos

Avacopan (reducción GC) // GC estándar: no inferior 6m, superior 12m

Kronbichler A. Lancet 2024; 403: 683–98

PLEX: no ↓ la mortalidad ni la progresión a ERT

GC dosis reducidas = eficaz

	Tested therapies	Key exclusion criteria	Remission rates (%) or endpoints	Serious side-effect rates	Daily glucocorticoid* exposure (g)	Conclusions
MEPEX (n=137)[80]	Plasma exchange vs intravenous methylprednisolone	Haemodialysis >2 weeks before admission; alveolar haemorrhage requiring invasive ventilation	69% vs 49% at 3 months‖	50% vs 48%	NR	The addition of plasma exchange in patients with a serum creatinine >5·7 mg/dL has a beneficial effect on dialysis independence at 3 months
PEXIVAS (n=704)[81]	Plasma exchange vs control; reduced-dose glucocorticoids vs standard-dose glucocorticoids	Haemodialysis ≥21 days before randomisation	28·4% vs 31·0%; 27·9% vs 25·5%	39% vs 32%; 34% vs 37%**	1 year reduced dose 3·1;†† 1 year standard dose 4·9††	The addition of plasma exchange has no effect on a composite of end-stage kidney disease and death at 2·9 years of follow-up; reduced-dose glucocorticoids are safe and reduce serious infections
NORAM (n=100)[87]	Oral methotrexate vs oral cyclophosphamide	Organ-threatening or life-threatening disease (ie, serum creatinine concentrations ≥1·7 mg/dL)	89·8% vs 93·5%	17·6% vs 12·2%	8·8 vs 6·2	Excess relapse rate in the methotrexate group, and higher cumulative glucocorticoid exposure
MYCYC (n=140)[33]	Mycophenolate mofetil vs intravenous cyclophosphamide	eGFR <15 ml/min per 1·73 m^2; life-threatening disease	67% vs 61%	50% vs 40%	6·2 vs 5·8	Excess relapse rate in the mycophenolate mofetil group, driven by patients with PR3-ANCA-associated vasculitis

Summaries of trials done to induce a state of remission. Definitions of remission differ between trials and might partly explain the observed differences as reported by these trials. ANCA=antineutrophil cytoplasmic antibody. eGFR=estimated glomerular filtration rate. MPO=myeloperoxidase. NR=not reported. PR3=proteinase 3. *Prednisone or prednisolone dose. †CORTAGE also included patients with eosinophilic granulomatosis with polyangiitis (13·5%) and polyarteritis nodosa (9·6%). ‡Four infusions of rituximab were combined with two pulses of cyclophosphamide. §Weight adjusted prednisolone doses at 12 months (mg/kg per day). ¶Steroid administration before each rituximab infusion was not required. ||Dialysis independence at 3 months was the primary endpoint. **Major serious adverse events are given independently, the percentage of serious infectious complications as the most common side effect is given. ††Doses calculated on the basis of body weight, here based on body weight between 50 and 75 kg.

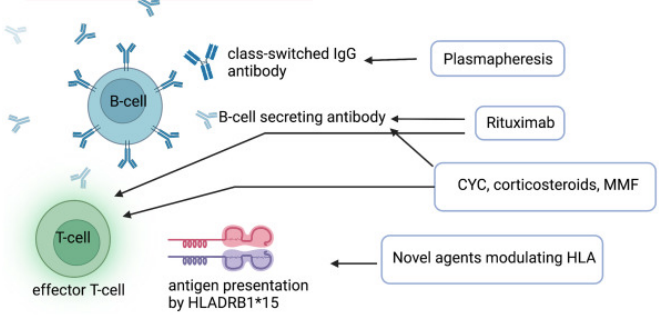
Kronbichler A. Lancet 2024; 403: 683–98

CONTROVERSIAS EN EL TRATAMIENTO

Question	What we know	Practical decision
Fixed vs. biomarker-guided RTX?	Fixed shows fewer relapses overall	Use **fixed q6 months** for most; reserve tailored for special cases
Avacopan for all?	Reduces GC exposure; benefits largest in GC-toxic risk	Use in patients where GC minimization is key and severe RPGN
PLEX in RPGN?	No mortality benefit; infection risk ↑	Consider in selected cases: SCr ≥ 3.4 mg/dL (> 300 µmol/L), RPGN/dialysis, and/or DAH with hypoxemia, after individualized risk–benefit discussion
When to stop RTX?	Some low-risk phenotypes safe to stop	Consider after **18–24 months** quiescence and close follow-up

Dal Pozzolo R. Clin Rev Allergy Immunol. 2026 Mar 23;69(1):22

Tratamiento de la enf. Ac-AMG



MPD bolos 500-1000mg/día 3 días → PDN 1mg/kg/d máx 60mg 4-8 semanas, desescalar 6-9m + plasmaféresis + CYC después de la plasmaféresis

- Plasmaféresis (PLEX): hasta que los Ac-AMG indetectables durante 14d
- Rituximab: si contraindicación CYC o enfermedad refractaria
- Imlifidasa: enfermedad refractaria. ECA FIII, GOOD-IDES-02
- Enfermedad doble positiva: + terapia de mantenimiento = AAV

Bharati J. Adv Kidney Dis Health 2024;31:206-215
KDIGO. Kidney Int. 2021 Oct;100(4S):S1-S276

RESUMEN DE TRATAMIENTO SPR

Table 3: Treatment protocol for pulmonary-renal syndrome as per etiology

Disease	Induction phase	Maintenance phase
ANCA AAV[37]	Pulse MPS + rituximab > cyclophosphamide	Rituximab > methotrexate/MMF/azathioprine/leflunomide
Anti GBM disease[19]	Pulse MPS + cyclophosphamide + plasmapheresis	Usually not required
Double positive disease[29]	Pulse MPS + cyclophosphamide + plasmapheresis	Rituximab/methotrexate/MMF/azathioprine/leflunomide
APS[45,46]	Steroids + plasmapheresis or IVIG. Rituximab in refractory cases. Anticoagulation once DAH resolves	Immunosuppression on case-to-case basis (no consensus available)
SLE[19]	Pulse MPS + cyclophosphamide Consider plasmapheresis in refractory cases	Cyclophosphamide/MMF/azathioprine
Cryoglobulinemia[27,28]	Pulse MPS + cyclophosphamide/rituximab + plasmapheresis. treatment of the precipitating condition	Immunosuppression on case-to-case basis (no consensus available)

Table 4: Common doses of drugs used in the management of pulmonary-renal syndrome

Drug	Dose
Iv pulse steroids	IV methylprednisolone 500–1000 mg/day for 3–5 days
High-dose oral steroids	Prednisone 1 mg/kg/day (generally up to 80 mg/day)
Methotrexate	Up to 25 mg/week (SC or oral)
Azathioprine	Up to 2 mg/kg/day
MMF	Up to 1500 mg (oral) twice per day
Cyclophosphamide	Oral: 2–3 mg/kg/day
	Remission induction-IV: 15 mg/kg (IV) every 2 weeks for 3 doses, followed by 15 mg/kg (IV) every 3 weeks for at least 3 doses or 0.5–1 g/m^2 monthly for 6 months
Rituximab	Remission induction: 375 mg/m^2 (IV) weekly for 4 doses or 1000 mg on days 1 and 15
	Remission maintenance: 500 mg (IV) every 6 months or 1 g (IV) every 4 months
Mepolizuamb in EGPA	300 mg (SC) every 4 weeks
Plasma exchange	40–50 mL/kg ideal body weight daily against 5% albumin
	Add fresh frozen plasma at the end of plasmapheresis in patients with DAH

Oral cyclophosphamide	Intravenous cyclophosphamide
2 mg/kg/d for 3 months, continue for ongoing activity to a maximum of 6 months	15 mg/kg at weeks 0, 2, 4, 7, 10, 13 (16, 19, 21, 24 if required)
Reduction for age: • 60 yr, 1.5 mg/kg/d • 70 yr, 1.0 mg/kg/d Reduce by 0.5 mg/kg/day for GFR <30 ml/min/1.73 m^2	Reduction for age: • 60 yr 12.5 mg/kg • 70 yr, 10 mg/kg Reduce by 2.5 mg/kg for GFR <30 ml/min/1.73 m^2

Kedia Y. Journal of Advanced Lung Health 2024. DOI: 10.4103/jalh.jalh_14_24
KDIGO. Kidney Int. 2024 Mar;105(3S):S71-S116

CONCLUSIONES: SPR

- Enfermedad grave que amenaza la vida: diagnóstico y tratamiento precoz
- Ante paciente con HAD es imprescindible descartar afectación renal: SPR
- Identificar la causa: elección del tratamiento y decisión de mantenimiento
- Minimizar efectos secundarios de los GC (regímenes reducidos, avacopan)
- Plasmaféresis si mediada por inmnunocomplejos o GNRP/HAD → SPR
- Vasculitis 2º a fármacos/drogas: no requiere tratamiento de mantenimiento, pero esencial eliminar la exposición