

CASO CLÍNICO

Fiebre persistente en un varón de mediana edad

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R2 Medicina Interna
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ANTECEDENTES PERSONALES

Varón 54 años

Obesidad (IMC 30,5)

Fumador

Possible fiebre reumática en la infancia

No tratamiento habitual

ENFERMEDAD ACTUAL

- Fiebre 3 semanas (38,3°C)
- Amoxicilina 7 días, paracetamol
- Predominio vespertino
- Cefalea, astenia, mialgias
- Niega otra sintomatología

EXPLORACIÓN FÍSICA

- Hemodinámicamente estable
- Soplo sistólico aórtico no irradiado
- Sin otros hallazgos

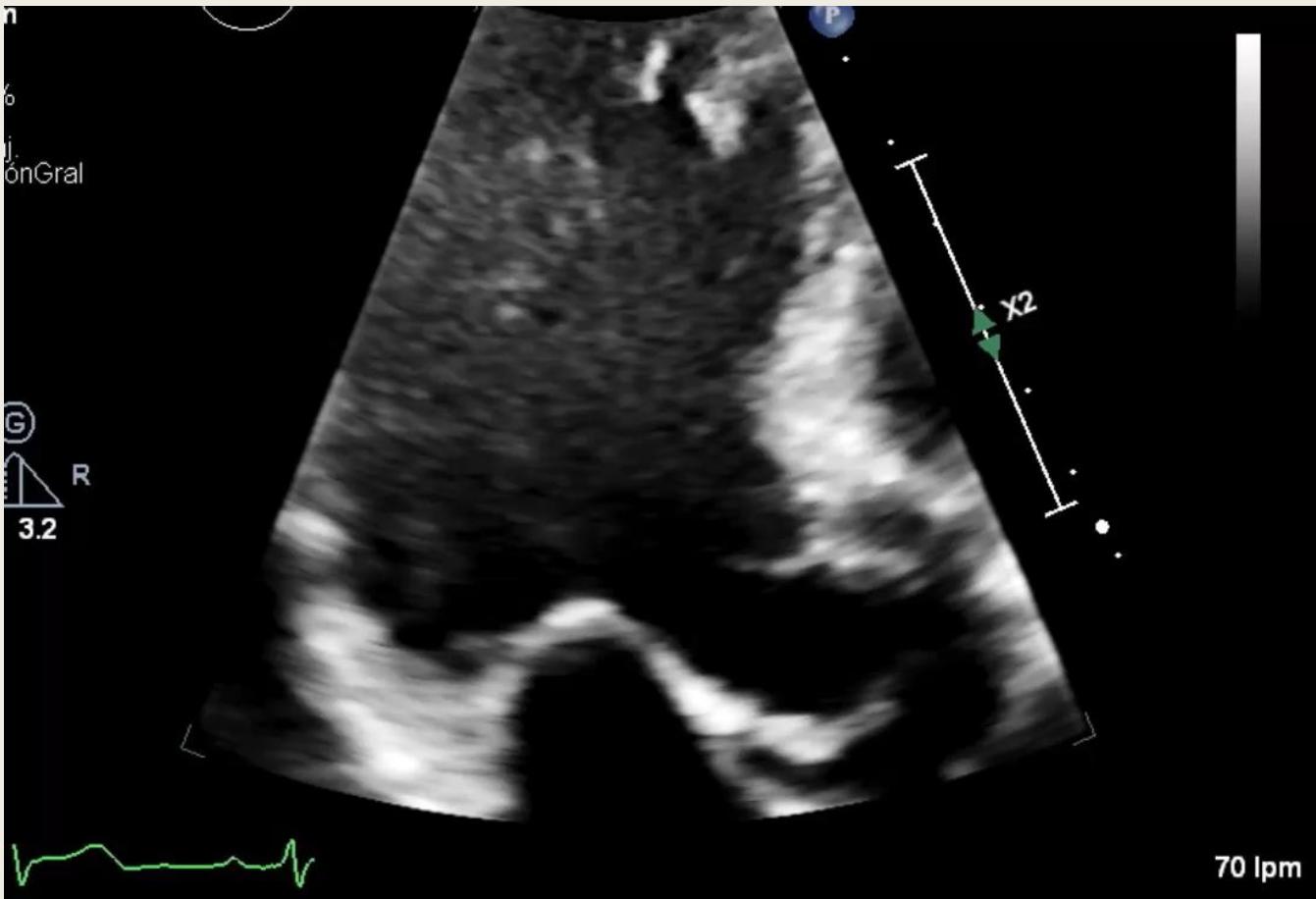
PRUEBAS COMPLEMENTARIAS

- Análisis de sangre:
 - *Bioquímica: elevación de reactantes de fase aguda PCR 156.7 mg/L (rango 0-5) PCT negativa, VSG de 58 mm (rango 1-15)*
 - *Hemograma: leucocitosis con neutrofilia (L 13,000, N 8,600)*
 - *Serologías: VHB, VHC, VIH negativos*
- Despistaje autoinmunidad:
 - *ANA + 1:80 patrón nucleolar*
 - *ENAs, antiDNA, antitiroideos, FR negativos*
- PCR SARS-CoV2 en ex. nasofaríngeo: negativa
- Radiografía de tórax: sin hallazgos



PRUEBAS COMPLEMENTARIAS (2)







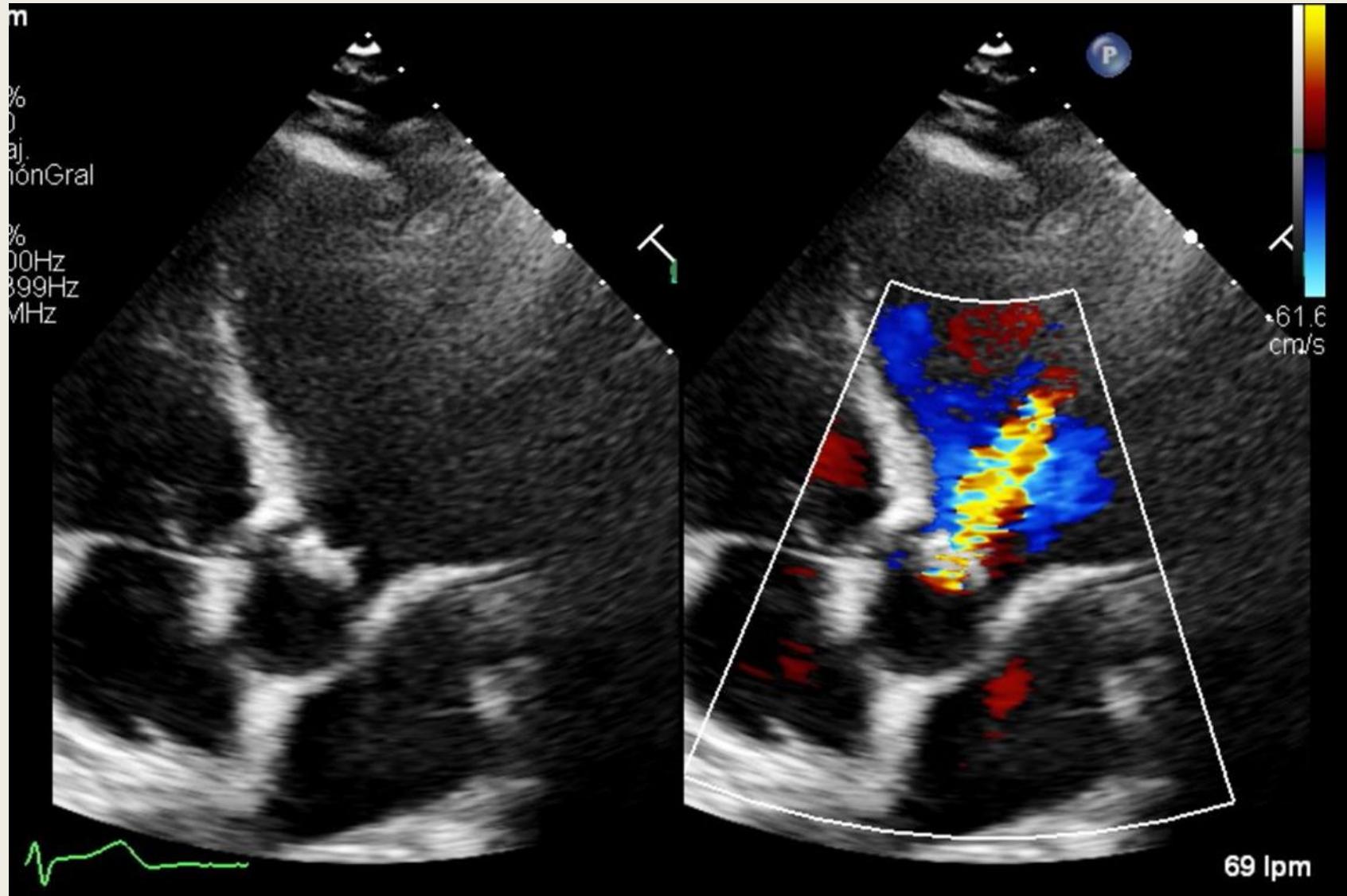
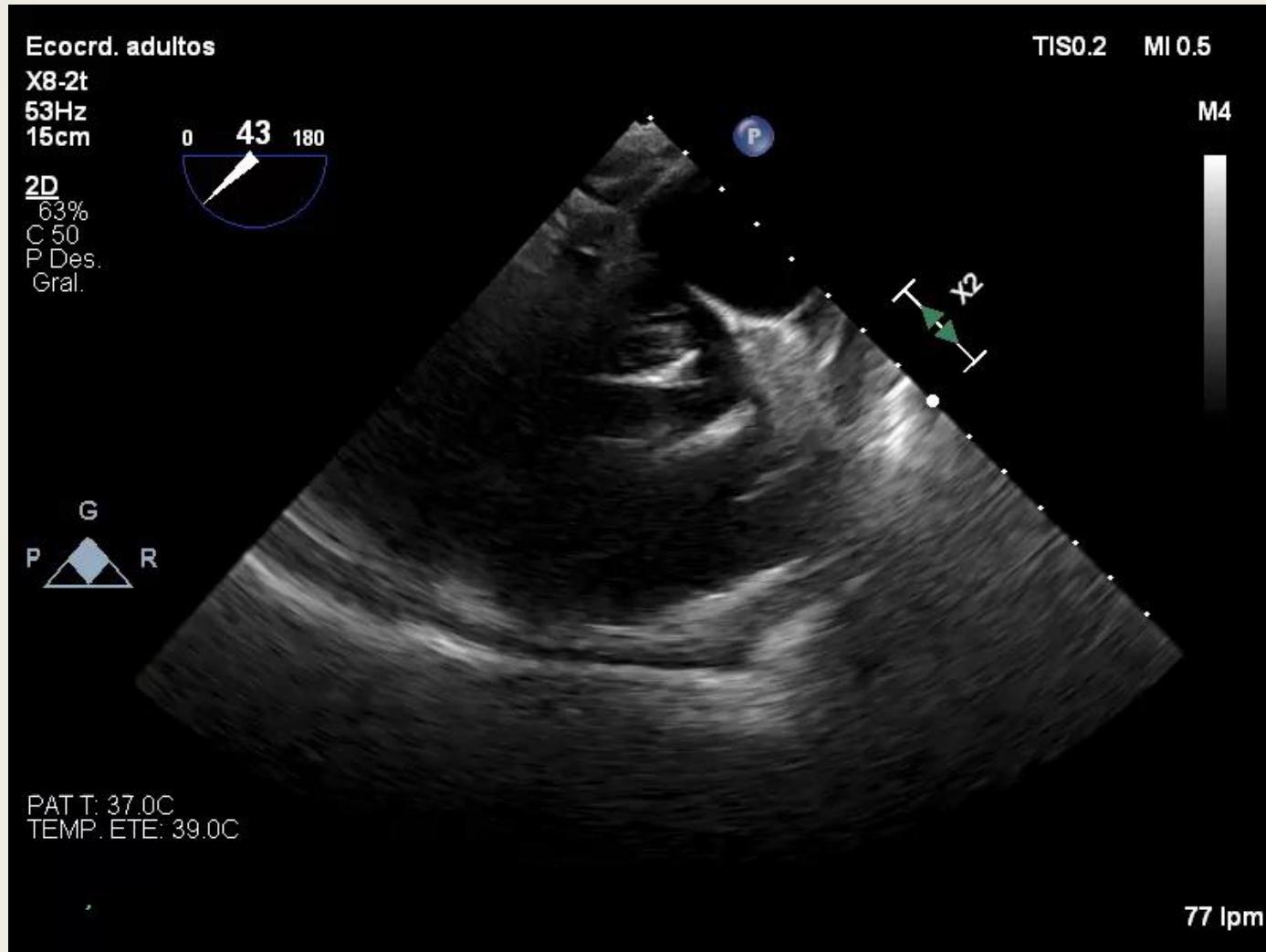


Figura 1. Ecocardiograma transtorácico. Válvula aórtica bicúspide con nódulo de calcio y fusión de velo coronariano izquierdo y derecho. Válvula desestructurada con dudosa imagen móvil únicamente visible en plano 5 cámaras. Insuficiencia aórtica moderada (ver Doppler).

PRUEBAS COMPLEMENTARIAS (3)



Ecocrd. adultos

X8-2t

32Hz

13cm

xPlane

57%

57%

50dB

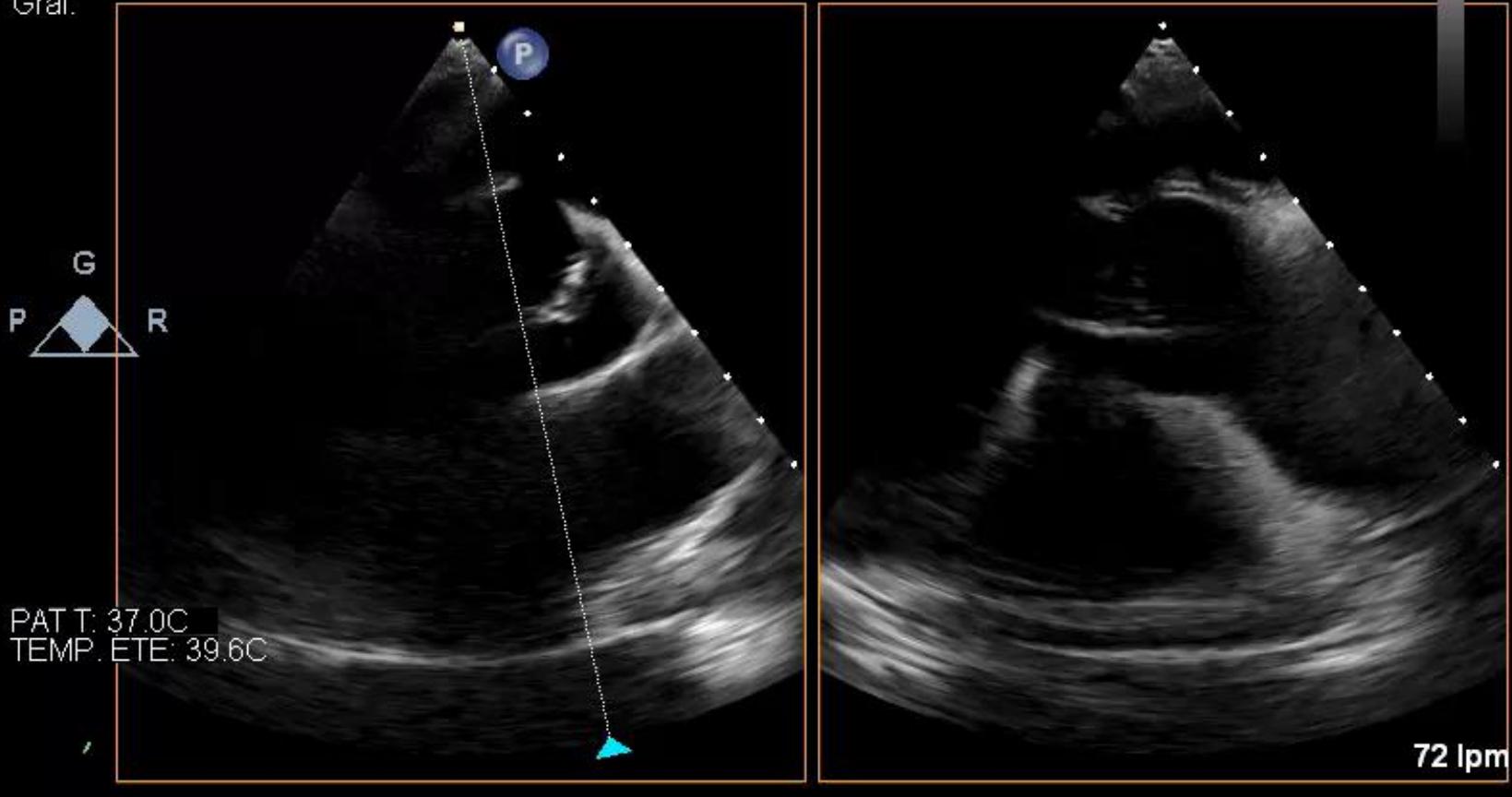
P Des.

Gral.

TIS0.2 MI 0.5

M4

45 -12



Ecocrd. adultos

X8-2t

53Hz

13cm

2D

57%

C 50

P Des.
Gral.



TIS0.2 MI 0.9

M4



PAT T: 37.0C
TEMP. ETE: 40.0C

75 lpm

Ecocrd. adultos

X8-2t

61Hz

7.8cm

xPlane

57%

57%

50dB

P Des.

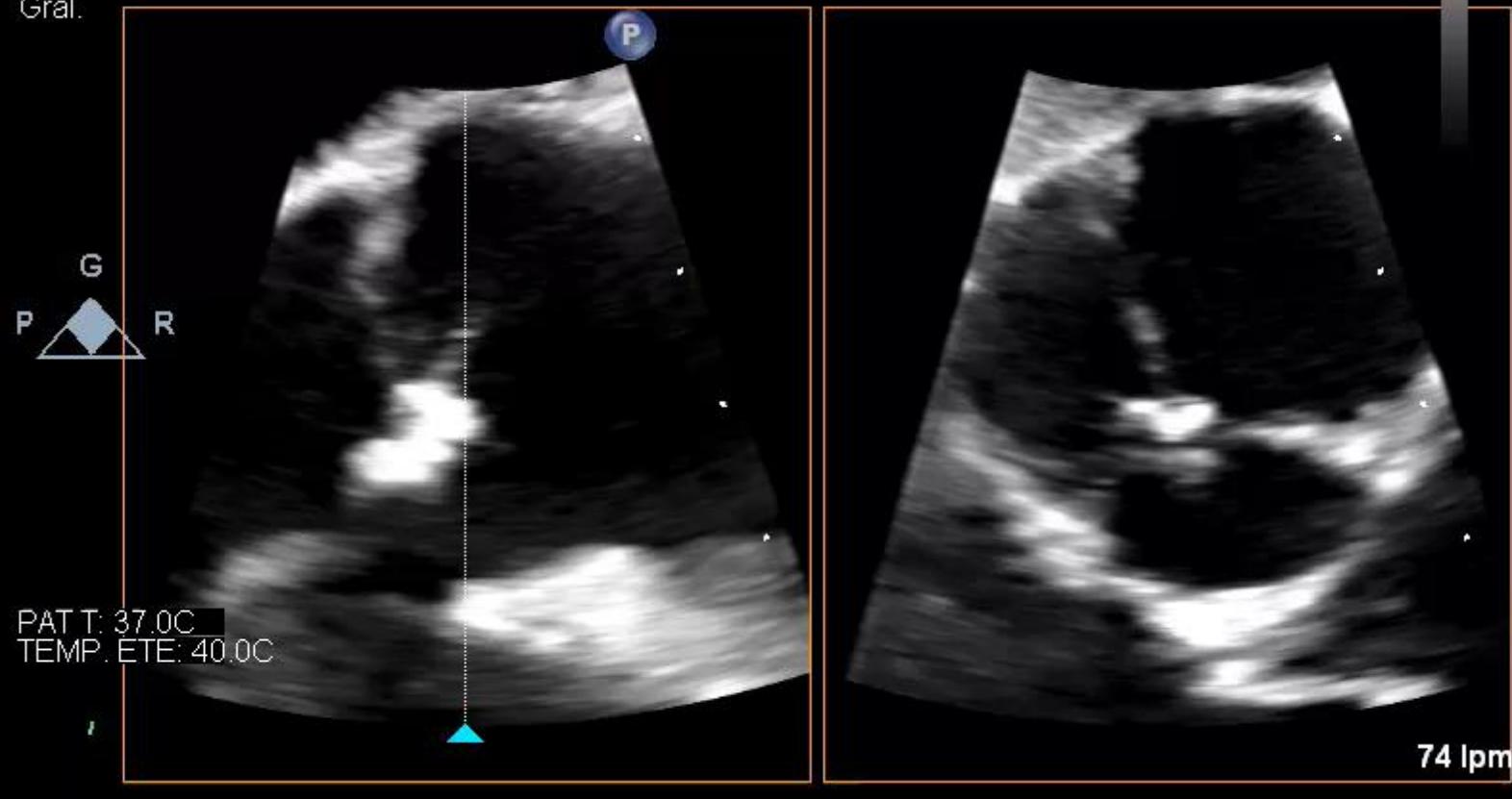
Gral.

TIS0.2 MI 0.5

M4

125

10



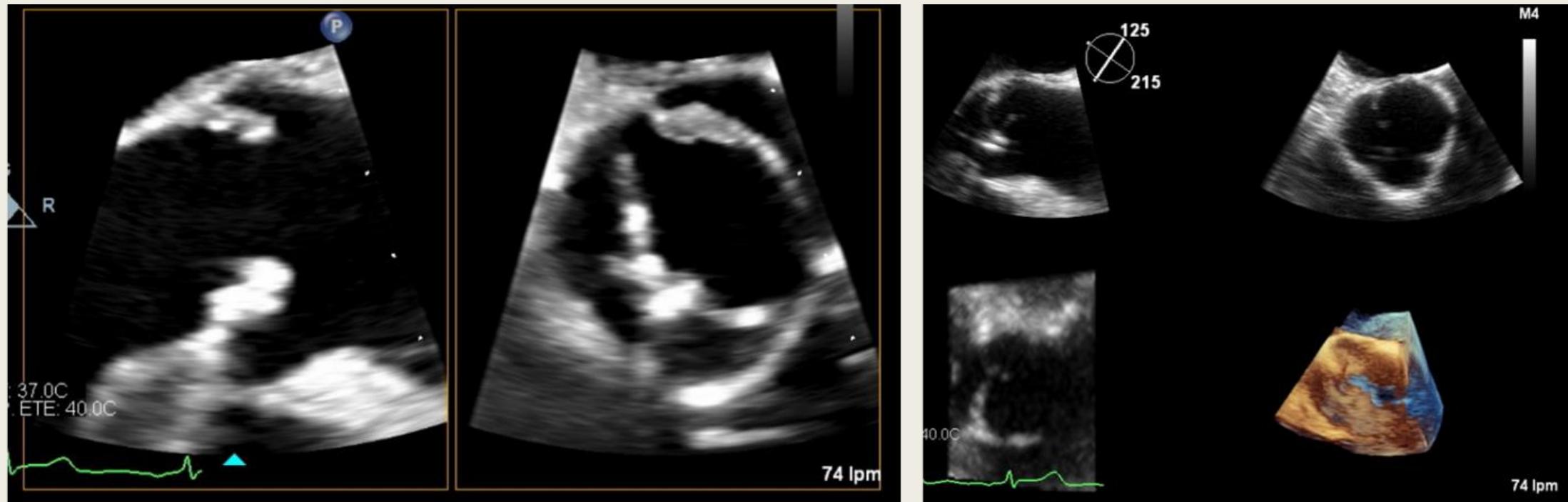


Figura 2. Ecocardiograma transesofágico: Válvula aórtica bicúspide por fusión del velo coronario derecho e izquierdo. Este velo coronario está calcificado y fijo. Insuficiencia aórtica moderada. Se objetiva imagen filiforme en cara aórtica del velo coronario de 0,73 cm con escasa movilidad.

Major criteria
1. Blood cultures positive for IE
a. Typical microorganisms consistent with IE from 2 separate blood cultures:
• <i>Viridans streptococci, Streptococcus gallolyticus (Streptococcus bovis), HACEK group, Staphylococcus aureus</i> ; or
• Community-acquired enterococci, in the absence of a primary focus; or
b. Microorganisms consistent with IE from persistently positive blood cultures:
• ≥2 positive blood cultures of blood samples drawn >12 h apart; or
• All of 3 or a majority of ≥4 separate cultures of blood (with first and last samples drawn ≥1 h apart); or
c. Single positive blood culture for <i>Coxiella burnetii</i> or phase I IgG antibody titre >1:800
2. Imaging positive for IE
a. Echocardiogram positive for IE:
• Vegetation;
• Abscess, pseudoaneurysm, intracardiac fistula;
• Valvular perforation or aneurysm;
• New partial dehiscence of prosthetic valve.
b. Abnormal activity around the site of prosthetic valve implantation detected by ¹⁸ F-FDG PET/CT (only if the prosthesis was implanted for >3 months) or radiolabelled leukocytes SPECT/CT.
c. Definite paravalvular lesions by cardiac CT.

Minor criteria
1. Predisposition such as predisposing heart condition, or injection drug use.
2. Fever defined as temperature >38°C.
3. Vascular phenomena (including those detected by imaging only): major arterial emboli, septic pulmonary infarcts, infectious (mycotic) aneurysm, intracranial haemorrhage, conjunctival haemorrhages, and Janeway's lesions.
4. Immunological phenomena: glomerulonephritis, Osler's nodes, Roth's spots, and rheumatoid factor.
5. Microbiological evidence: positive blood culture but does not meet a major criterion as noted above or serological evidence of active infection with organism consistent with IE.

Habib G et al; ESC Scientific Document Group. 2015 ESC Guidelines for the management of infective endocarditis. Eur Heart J. 2015



INGRESO

- Reexploración: sin estigmas de endocarditis
- Hemocultivos
- Se inicia tratamiento empírico:
 - *Cloxacilina + ampicilina + gentamicina*

EVOLUCIÓN

- Persiste fiebre y elevación de reactantes
- Hemocultivos iniciales: sin aislamiento
- Larga incubación
- Nuevos hemocultivos: estériles
- Bacterias de difícil incubación:
 - Serologías: sífilis, brucella, fiebre Q, borrelia, *M. pneumoniae*, *C. pneumoniae* y *Legionella* negativas
 - PCR *T. whipplei* y *Bartonella* negativa
- IGRA negativo.

PET-TC

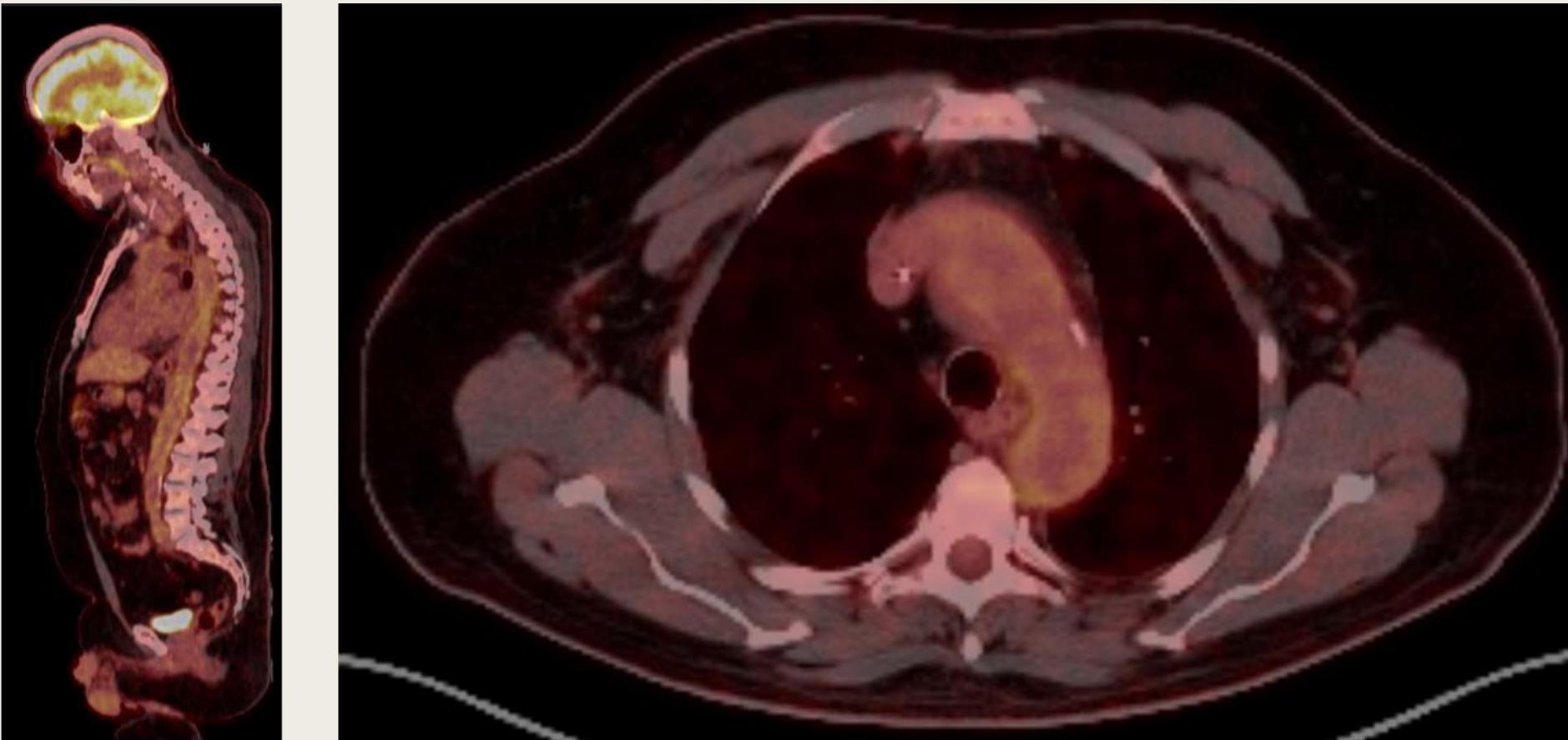


Figura 3. PET-TC. Aumento de captación de morfología lineal, irregular y parcheado a nivel de paredes vasculares que se extiende desde raíz aórtica hasta bifurcación iliaca, compatible con aortitis difusa.

Characteristic Features of IgG4-related CVD, Takayasu Arteritis, Giant Cell Arteritis, and Atherosclerosis

Feature	IgG4-related CVD	Takayasu Arteritis	Giant Cell Arteritis	Atherosclerosis
Sex	Affects substantially more males than females	Affects substantially more females than males	Affects more females than males	Affects approximately the same numbers of males and females
Age	Typically affects those older than 60 years	Typically affects those aged 20–30 years	Typically affects those older than 50 years	Typically affects those older than 60 years
Most common site	Abdominal aorta	Thoracic aorta	Temporal artery, thoracic aorta	Abdominal aorta
Involved region of vessel wall	Adventitia	Adventitia and media	Media	Intima
Other organ involvement	Retroperitoneal fibrosis, lymph node swelling, pancreatitis, etc	Generally no, but pulmonary artery may be involved	No	No
Associated coronary aneurysm	Yes	Yes	No	Yes, but mainly iatrogenic

AMPLIACIÓN ESTUDIO

- Autoinmunidad:
 - *ANCA negativos*
 - *Anti fosfolípido negativos con anticardiolipina IgG+*
- Proteinograma sin alteraciones
- IgG4 en rango

Noninfectious causes of aortitis

✗ Rheumatic diseases (e.g., rheumatoid arthritis, systemic lupus erythematosus, or HLA-B27-related spondyloarthropathies)

Vasculitides (e.g., giant-cell arteritis, Takayasu's arteritis, Cogan's syndrome, relapsing polychondritis, Behçet's disease, polyarteritis nodosa, or antineutrophil cytoplasmic antibody-associated vasculitis) ?

Cancer

IgG4-related disease ?

Sarcoidosis

Histiocytic disorders (e.g., Erdheim–Chester disease)

Idiopathic disease ?

Infectious causes of aortitis ✗

Common bacteria (e.g., staphylococcus, salmonella, enterococcus, *Streptococcus pneumoniae*, *Coxiella burnetii*, or listeria)

Fungi (e.g., candida, cryptococcus, aspergillus, or histoplasma)

Viruses (e.g., human immunodeficiency virus or varicella–zoster virus)

Mycobacteria (e.g., *Mycobacterium tuberculosis* or *M. bovis*)

CAMBIO TERAPÉUTICO

- Se suspende antibioterapia
- Se inician bolos de metilprednisolona 125 mg 3 días

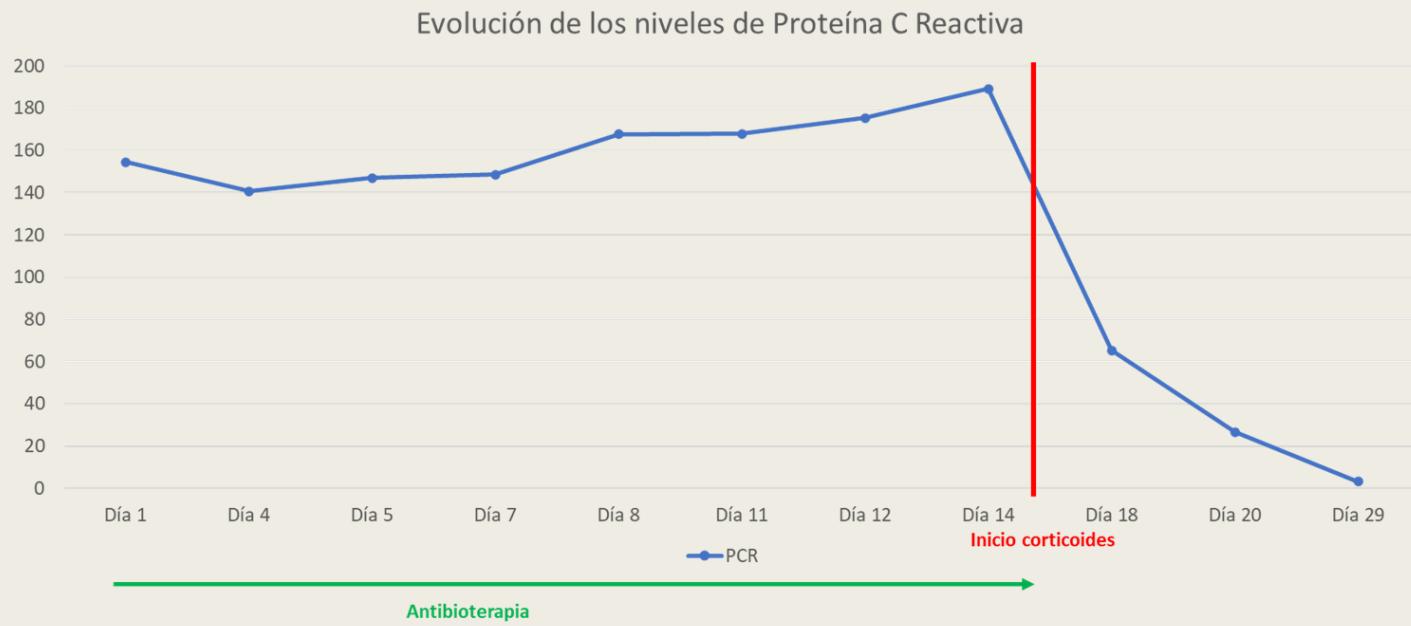


Figura 4. Evolución de los niveles de reactantes de fase aguda (Proteína C-reactiva) a lo largo del ingreso y en relación al tratamiento antibiótico y el inicio de la corticoterapia tras los hallazgos del PET-TC. Se observa un claro descenso tras el inicio de los corticoides hasta normalizarse en el día 29.

ALTA

- Metilprednisolona 80 mg vo
- Pulsos de ciclofosfamida
- Buena evolución
- Actualmente con corticoides orales en pauta descendente

DISCUSIÓN

- Fiebre de origen desconocido
- Ecocardiograma
 - Válvula bicúspide no conocida
 - Strand (excrecencia de Lamb)
- PET-TC: utilidad en procesos vasculares



Figure I: An example of a typical strand on the aortic valve.

- Leitman M. Clinical significance and prevalence of valvular strands during routine echo examinations. European Heart Journal – Cardiovascular Imaging (2014).
- Buchrits S et al. The yield of F18 FDG PET-CT for the investigation of fever of unknown origin, compared with diagnostic CT. Eur J Intern Med. 2021

Aortitis no infecciosa

- Principalmente autoinmune
 - *Arteritis de células gigantes*
 - *Arteritis de Takayasu*
 - *AR, EA, Cogan, Behçet, LES, Sjögren, GPA, sarcoidosis, IgG4.*
 - *Aortitis clínicamente aislada*
- MORBIMORTALIDAD
 - *Aneurismas, roturas, disecciones, trombosis*
 - *50 % en 10 años*

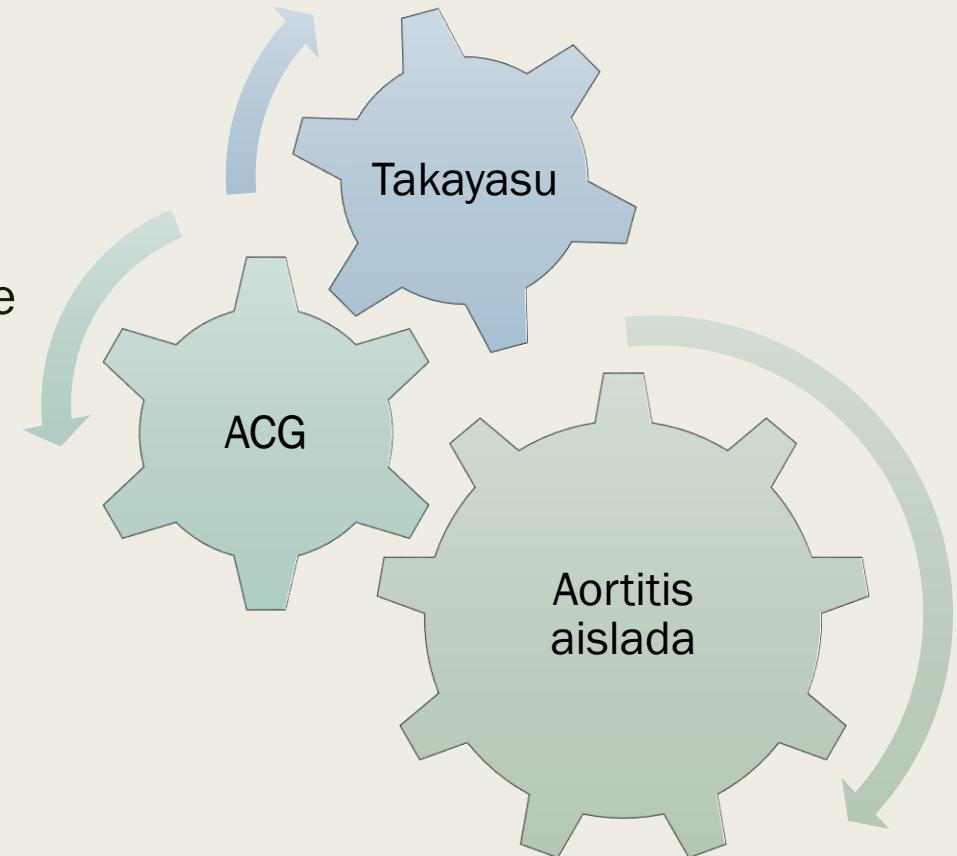
Ferfar Y et al. Spectrum and outcome of noninfectious aortitis. J Rheumatol. 2021.

Pugh D et al. Aortitis: recent advances, current concepts and future possibilities. Heart. 2021;

Espitia O et al. Comparison of idiopathic (isolated) aortitis and giant cell arteritis-related aortitis. A French retrospective multicenter study of 117 patients. Autoimmun Rev. 2016.

Tratamiento:

- Corticoides a dosis altas con pauta descendente
- Ahorradores de corticoides
- Adaptarlo a cada caso individual



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CONCLUSIONES

Importancia de replantear el diagnóstico

Utilidad del PET-TC en FOD

Espectro de la aortitis no infecciosa

Importante morbimortalidad

Necesidad de tratamiento agresivo