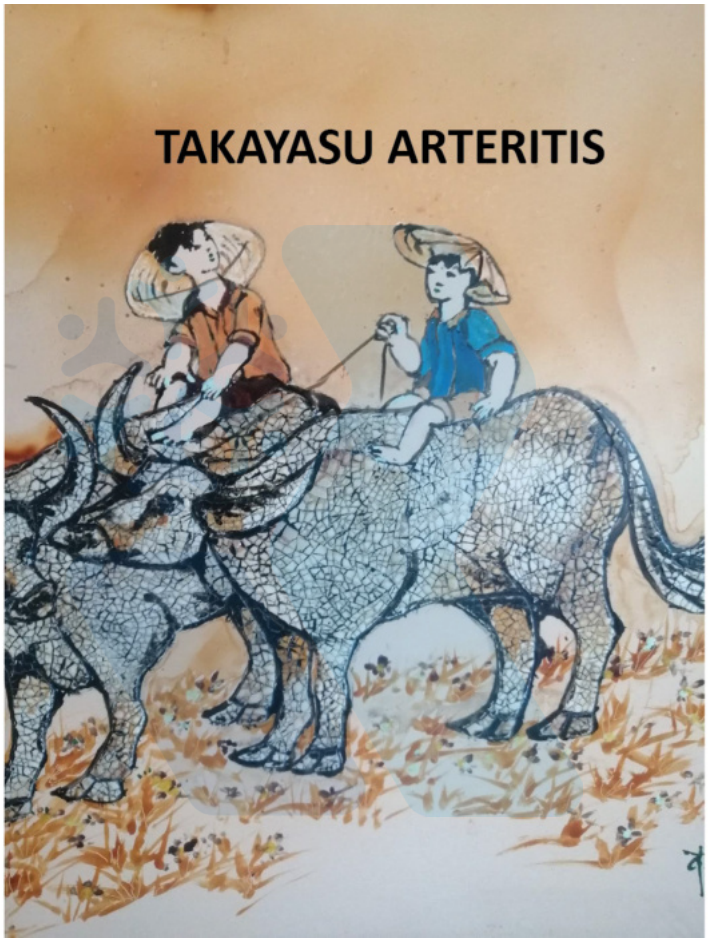

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MARSEILLE

FRANCE

Sorry not to be with you

I thought I could bike to HCM city

Like these 2 ladies

But I started too late

Could not follow them

And I am too slow!

DEFINITION

- First description by Yamamoto in 1830, but usually attributed to Takayasu in 1905
  English literature 1950: « pulseless disease » then « aortic arch syndrome »
- Chronic large vessel granulomatous vasculitis
- Affects aorta, cervical, visceral and lower limb main branches and coronary and pulmonary arteries
- Inflammation initially leads to thickening of the arterial wall
- Results in stenosis, occlusion, dilatation or aneurysms
- Ubiquitous (5-8 cases/million), but in Japan (40 cases/million).
  Mainly southern asia, mediterranean area and middle east.
- Predominately female (83-97%) before 30yo, but 15% after 40
- Morbidity+++: vascular claudication, heart failure, neurologic ischemic events
  Impaired daily activities (74%)
- Mortality: 3-27% (congestive heart failure)

ACR 1990 classification

CRITERIA
1. Age at disease onset< 40 years
2. Claudication of extremities (upper extremities++)
3. Decreased brachial artery pulse
4. Blood pressure difference>10 mmHg between arms
5. Bruit over subclavian arteries or abdominal aorta (on auscultation)
6. *Arteriogram abnormality* (narrowing, occlusion not due to atherosclerosis or fibromuscular dysplasia

At least 3 criteria or more (Sensitivity= 91% et Specificity = 98%)

Arend WP et al. Arthritis Rheum 1990

Ishikawa's criteria modified by Sharma (1996)

3 major criteria
1. Left mid subclavian artery lesion
2. Right mid subclavian artery lesion
3. Characteristic signs and symptoms of at least one month duration • Limb Claudication /pulselessness or significant blood pressure difference (>10 mmHg) • Fever • Neck pain • Transient amaurosis / blurred vision • Syncope • Dyspnea • Palpitations

Ishikawa's criteria modified by Sharma (1996)

10 minor criteria
High ESR > 20 mm/h
Carotid artery tenderness on palpation (UL or BL)
Hypertension> 140/90 mmHg brachial or popliteal> 160/90 mmHg
Aortic regurgitation or annuloaortic ectasia(auscultation or US)
Pulmonary artery lesion
Left mid common carotid stenosis or occlusion
Distal brachiocephalic trunk stenosis or occlusion
Descending thoracic aorta lesion
Abdominal aorta lesion
Coronary artery lesion before the age of 30 in the absence of dyslipidemia ou diabetes

2 major criteria or 1 major+ 2 minor criteria or 4 minor criteria are highly suggestive of TA (Sensitivity 92.5% - Specificity 95%).

Sharma BK et al. Diagnostic criteria for Takayasu arteritis. Int J Cardiol 1996

PEDIATRIC TAKAYASU ARTERITIS

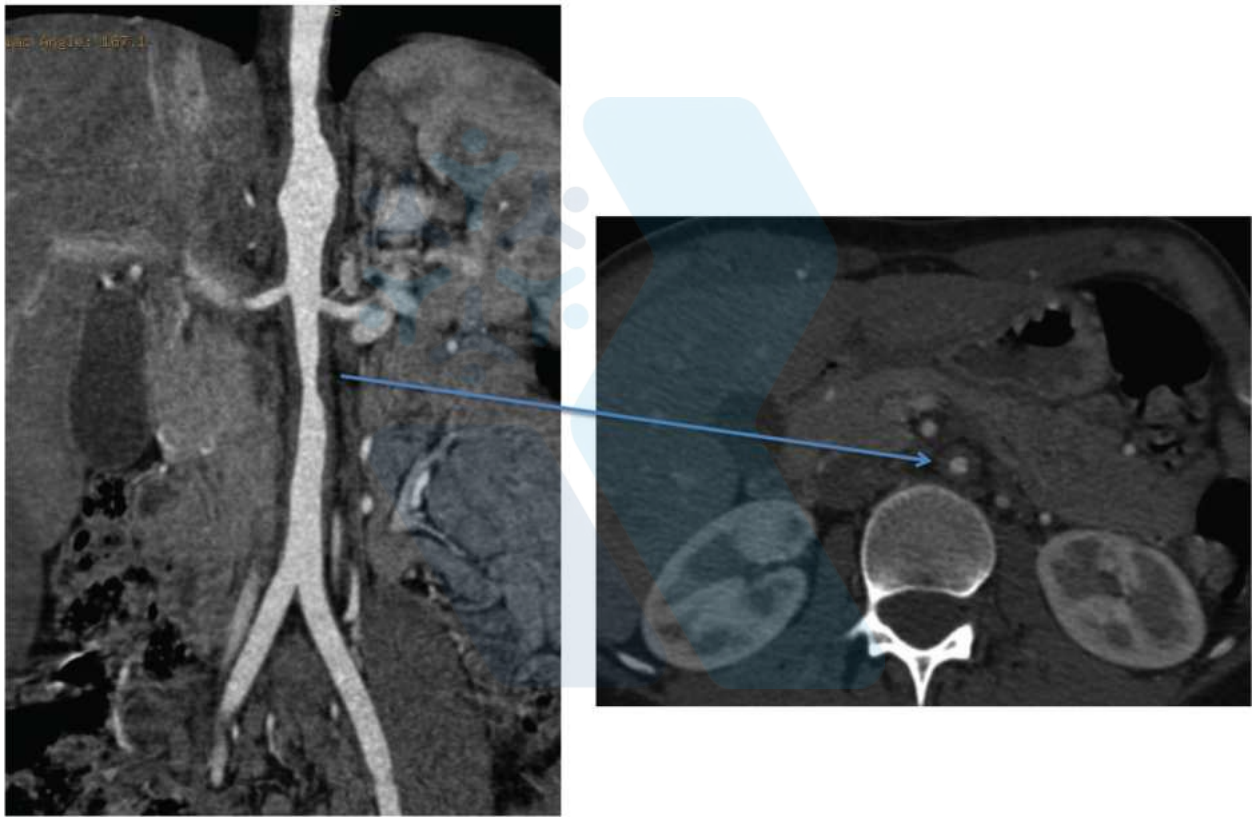
The EULAR/PRINTO/PRES criteria for childhood Takayasu arteritis [55].

Criterion	Definition
Angiographic abnormality (mandatory criterion)	Angiography (conventional, computed tomography, or magnetic resonance imaging) of the aorta or its main branches and pulmonary arteries showing aneurysm/dilatation, narrowing, occlusion or thickened arterial wall not due to fibromuscular dysplasia, or similar causes; changes usually focal or segmental
Pulse deficit or claudication	Lost/decreased/unequal peripheral artery pulse(s) Claudication: focal muscle pain induced by physical activity
Blood pressure discrepancy	Discrepancy of four limb systolic blood pressure >10 mmHg difference in any limb
Bruits	Audible murmurs or palpable thrills over large arteries
Hypertension	Systolic/diastolic blood pressure greater than 95th percentile for height
Acute phase reactants	Erythrocyte sedimentation rate >20 mm per first hour or C-reactive protein any value above normal (according to the local laboratory)

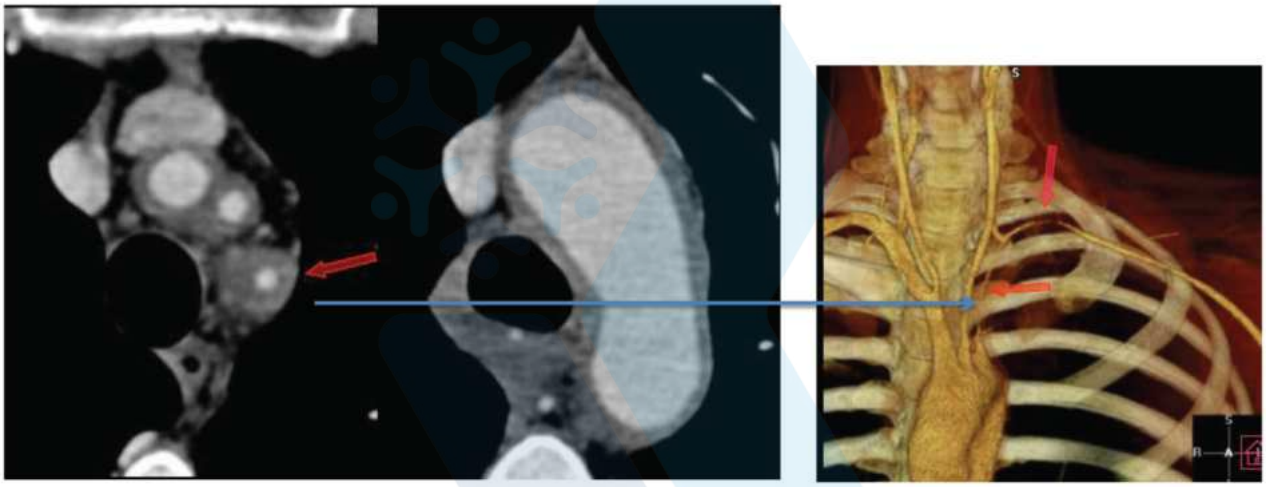
TA is classified when the mandatory criterion is present plus any other criteria.

<18 YO 100% sensitivity, 100% specificity

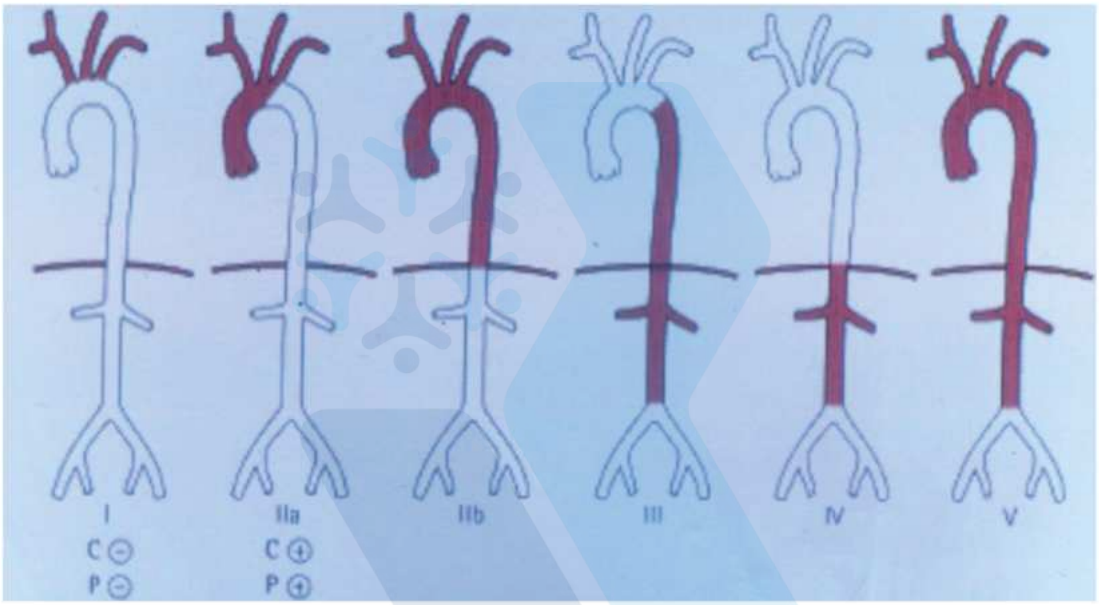
Takayasu Arteritis



Takayasu:
Supraaortic ostium thickening



Takayasu Arteritis: Numano's angiography classification



C(+) (coronaries), P(+)(Pulmonary arteries)

Grayson Ann Rheum Dis 2022

TA CLINICAL DIAGNOSIS
VARIOUS CIRCUMSTANCES

- General symptoms, ischemic symptoms, high blood pressure<50 YO
- Fortuitous discovery: absence of pulse, vascular murmur on physical examination
- Imaging tests: arterial parietal thickening, arterial stenosis or aneurysms
- Associated diseases: Crohn's , ulcerative colitis, spondylarthropathy, rarely sarcoidosis

- « **Pre-occlusive phase** »: fever, arthralgia, myalgia, erythema nodosum, pyoderma gangrenosum, carotinodynia, episcleritis
Often unnoticed, absent, or retrospectively identified

- « **Vascular phase** »: results from arterial lesions (stenosis, obliteration, aneurysms) and symptoms depend on the impaired area

Saadoun Orphanet J Rare Dis 2021

TA CLINICAL DIAGNOSIS

- **Axillo-subclavian:** asymptomatic, upper limb claudication or subclavian steal,
Blood pressure asymetry
Absence of pulses, murmur, Raynaud's phenomenon

- **Carotid:** 10-30% of cases. Neck pain, **carotidodynia**, absence of pulse, murmur

- Dizziness, lipothymia, syncope, bilateral dark vision when getting up due to transient low verebral flow

Saadoun Orphanet J Rare Dis 2021

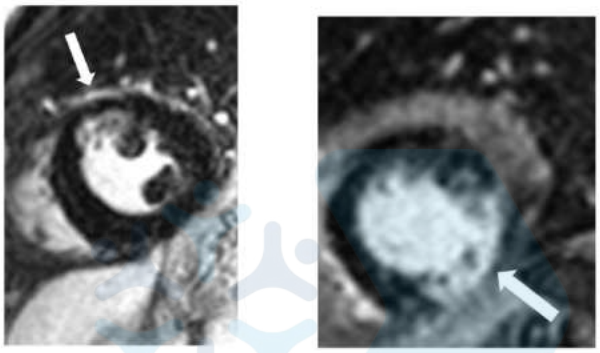
TA CLINICAL DIAGNOSIS

- **Thoraco-abdominal Aorta:** lower limb claudication
- **Renal artery or pseudo-coarctation**: renovascular hypertension+++ very common

(systolic blood pressure may be measured on **ankles** rather than arms)

Saadoun Orphanet J Rare Dis 2021

TA CLINICAL DIAGNOSIS



Cormamond Am J cardiol 2014

- **Heart:** from non specific perfusion abnormalities on MRI (26%) to angina pectoris due to coronary involvement (5-15%),
  Aortic insufficiency by dilatation of the ring (poor prognosis)++
  Hypertrophic or left heart disease due to high blood pressure
- **Lung:** 50% of patients, asymptomatic or chest pain, cough, dyspnea, hemoptysia
- **Skin:** Pyoderma gangrenosum, erythema nodosum
- **Biological signs:** no specific biomarker
  Inflammation: CRP, ESR, fibrinogen, alpha2 globulin but erratic, may be absent

Saadoun Orphanet J Rare Dis 2021

TA OCULAR MANIFESTATIONS

- **Prevalence of ocular manifestations in TA: 8-65%**
- **May be inaugural (58% of cases) or any time during the course**
- **Associated with a severe TA phenotype and activity of the disease**

- Anterior segment: ischemic uveitis
- Cataract
- **Posterior segment++ aneurysms of central and peripheral retina, then retinal ischemia**
- **Symetric involvement: 50%**

Carotid artery involvement induces an **ISCHEMIC RETINOPATHY**

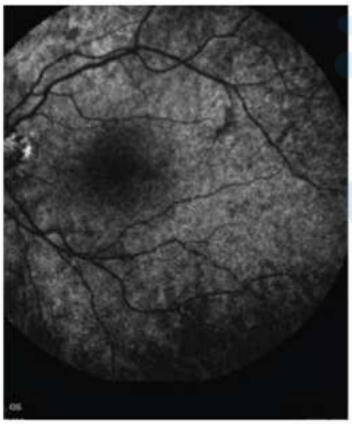
-Hypoperfusion retinopathy (70% of cases): microaneurysms, venous abnormalities, ocular ischemic syndrome,neovascular glaucoma, proliferative retinopathy

-Hypertensive retinopathy: 30%

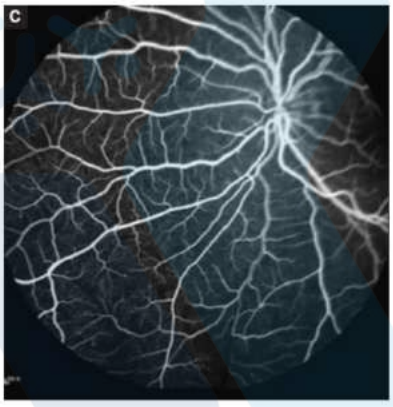
Sharma Indian J Ophtalmol 2024

TA OCULAR MANIFESTATIONS

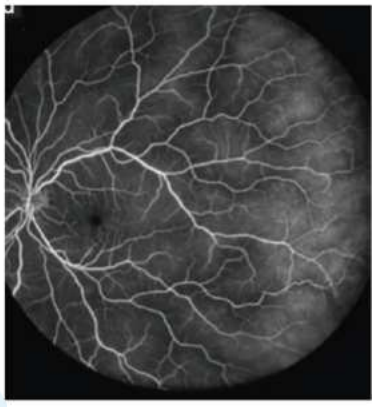
- **Stage 1: vascular dilatation**
  - **1a: with microaneurysms**
  - **1b: without**
- **Stage 2: vascular anasotomosis**
- **Stage 3: Complications (cataract, retinal ischemia, neovascularization, vitreous hemorrhage)**



Patchy choroidal filling Due to prolonged arm-retina time



Microaneurysms in the peripheral retina



Dye diffuse leakage in the late phase

Sharma Indian J Ophtalmol 2024

APPROPRIATE IMAGING IS THE MAINSTAY FOR DIAGNOSIS

EULAR recommendations for the use of imaging in large vessel vasculitis in clinical practice: 2023 update

4. In patients with suspected TAK, MRI to investigate mural inflammation or luminal changes should be used as the first imaging test to make a diagnosis of TAK.	3	9.5 (0.8)	96
5. FDG-PET, CT or ultrasound may be used as alternative imaging modalities in patients with suspected TAK. Ultrasound is of limited value for assessment of the thoracic aorta.	3 (CT) and 5 (PET and US)	9.7 (0.6)	100
6. Conventional angiography is not recommended for the diagnosis of GCA or TAK as it has been superseded by the previously mentioned imaging modalities.	5	9.8 (0.5)	100
7. In case of a suspected relapse of GCA or TAK, particularly when laboratory markers of disease activity are unreliable, ultrasound, FDG-PET or alternatively MRI may be considered for the assessment of vessel abnormalities. Imaging is not routinely recommended for patients in clinical and biochemical remission.	5	9.3 (1.4)	88

- **Conventional arteriography is no longer used for the diagnosis or the follow-up**
- **Other techniques such as doppler US, angio-CT, angio MRI and FDG-PET should be used**

Quinn, A&R 2022 75: 98-107 Ann Rheum Dis 2024, 83: 741-51

APPROPRIATE IMAGING IS THE MAINSTAY FOR DIAGNOSIS

- **Doppler Ultrasound (US):**
  - ***-Advantages:*** simple, easily reproducible, no radiation
    typical hypoechoic perivascular halo
    measures the thickness of carotid and axillary walls
  - ***-Inconvenients:*** no determination of activity
    Less sensitive than cross-sectional imaging for visceral arteries
    Inaccessibility of thoracic aorta
- **Angio-CT:**
  - ***-Advantages:*** parietal thickening>2-3 mm, arterial stenosis or aneurysm
    circumferential and regular sometimes enhanced by injection
    excellent spatial resolution covering the entire arterial tree
  - ***-Inconvenients:*** Radiation, iodinized contrast product injection
- **Angio-MRI:**
  - ***-Advantages:*** morphological study + parietal inflammation (T1 imaging+/-gadolinium)
    No radiation, no iodinized contrast product injection
  - ***-Inconvenients:*** Disease inflammatory activity?

Saadoun Orphanet J Rare Dis 2021 Bhandari Cureus 2023

APPROPRIATE IMAGING IS THE MAINSTAY FOR DIAGNOSIS

2-[18-F]FDG-PET/CT

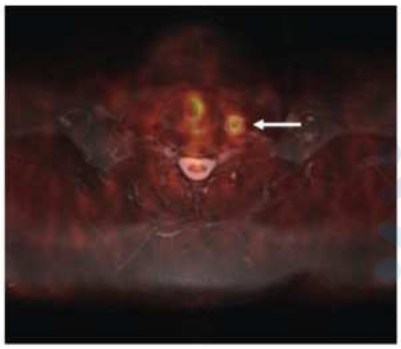
- More direct approach of the degree of vascular inflammation
- Useful for the diagnosis
- SUV max, SUV ratio (Vascular SUV max/mean standard uptake)
- Compared the aortic uptake and the liver uptake: PETVAS
  - Grade 0: no uptake
  - Grade 1: lower uptake than liver
  - Grade 2: equal uptake
  - Grade 3: Greater uptake in the aorta
- To follow inflammatory activity?

Not alone but in combination with clinical, biological and other imaging techniques

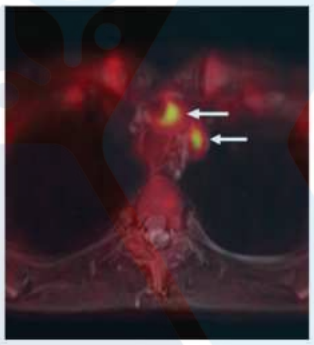
Jimenez Clin Exp Rheumatol 2021/Nassarmadji Front Med 2023

APPROPRIATE IMAGING IS THE MAINSTAY FOR DIAGNOSIS

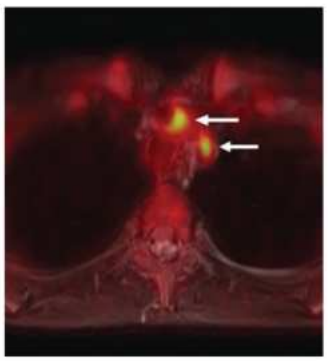
2-[18-F]FDG-PET/CT FOR DIAGNOSIS



Increased uptake in the left carotid artery in a patient with carodinodynia



Increased uptake in the ascending and descending thoracic aorta in a patient with fatigue and increased CRP



Increased uptake in the aortic arch, proximal left subclavian and carotid in a patient with left arm claudication

Luo Clin Radiol 2024

APPROPRIATE IMAGING IS THE MAINSTAY FOR DIAGNOSIS

INITIAL DIAGNOSIS

1/ MRI
2/ FDG-PET
CT or US
3/ Conventional angio
not recommended

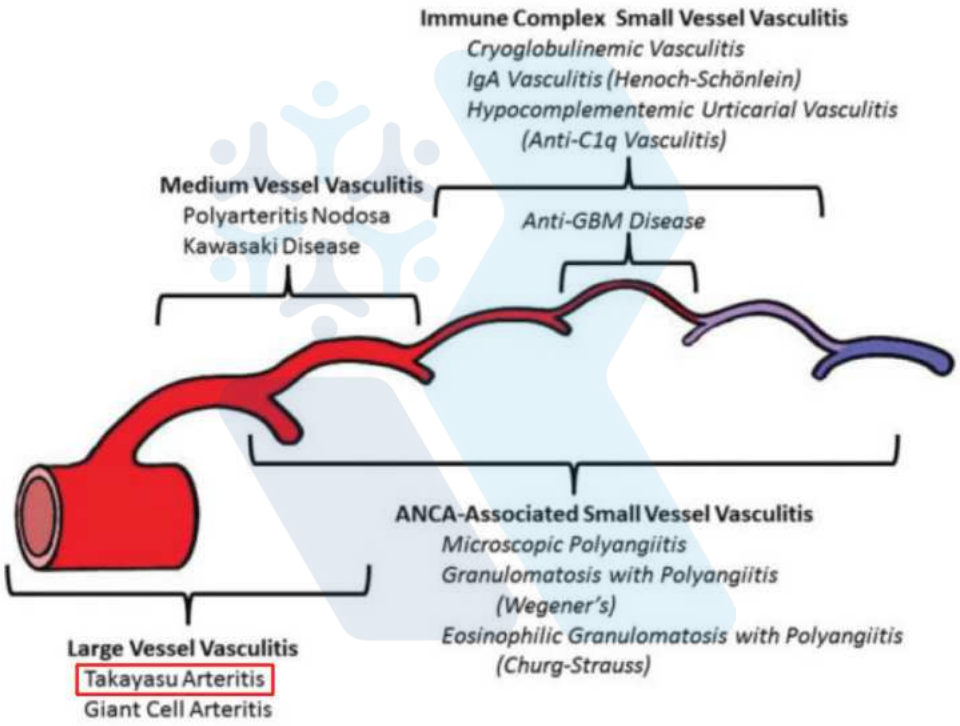
FOLLOW-UP

- No routine imaging if remision
- If suspected relapse without biomarkers
- Or long-term monitoring

FDG-PET (if SUV>1.3)
US, CT

Ann Rheum Dis 2024, 83: 741-51

Chapel Hill Classification 2012



Jennette JC et al. Arthritis Rheum 2013

DIFFERENTIAL DIAGNOSIS

- **INFECTIONS:**

mainly **tuberculosis, syphilis,**
staphylococcus, streptococcus, salmonella

- **INFLAMMATORY DISEASES:**

Behçet, Cogan, **relapsing polychondritis,**
Rheumatoid arthritis
Granulomatosis with polyangeitis

- **NON-INFLAMMATORY DISEASES:**

Atheroma
Genetic (Marfan, Ehlers-Danlos)
Post-radiation
Fibromuscular dysplasia

- **PERIAORTITIS:**

Erdheim-Chester
IgG4 disease
Perianeurysmal retroperitoneal fibrosis

- **PEDIATRICS:** Kawasaki (coronary, polyarteritis nodosa)

Saadoun Orphanet J Rare Dis 2021 Watanabe Curr Rheumatol Rep 2022

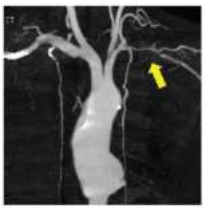
TA AND GIANT CELL ARTERITIS NOT THE SAME DISEASES

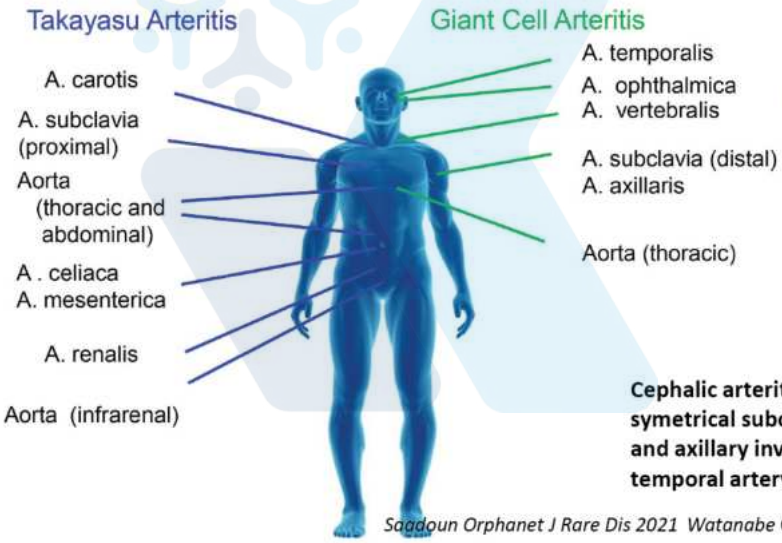
	Takayasu	GCA
Age	< 50 yo	> 50 yo
Sex Female ratio	9/1	3/1
Geographic Origin	Asia, North Africa, India	Caucasian
Genetics	HLA B52	HLA DR4
Associated symptoms		
• **Erythema nodosum**	+	-
• **Scleritis**	+	-
• **Ischemic Optic neuropathy**	-	+
• **Retinopathy**	+	-
Associated diseases	IBD/SA/Sarcoïdosis	PMR

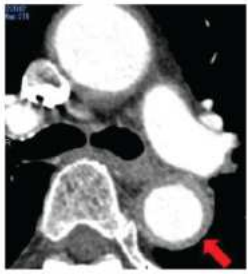
Saadoun Orphanet J Rare Dis 2021

TA AND GIANT CELL ARTERITIS

Vascular involvement	100%	≈30-60%
Carotid artery tenderness	+	Exceptional
Subclavian arteries	+	Symetrical
Renal arteries	+	Exceptional
Mesenteric/carotid arteries	++	-
Axillary arteries	+	++



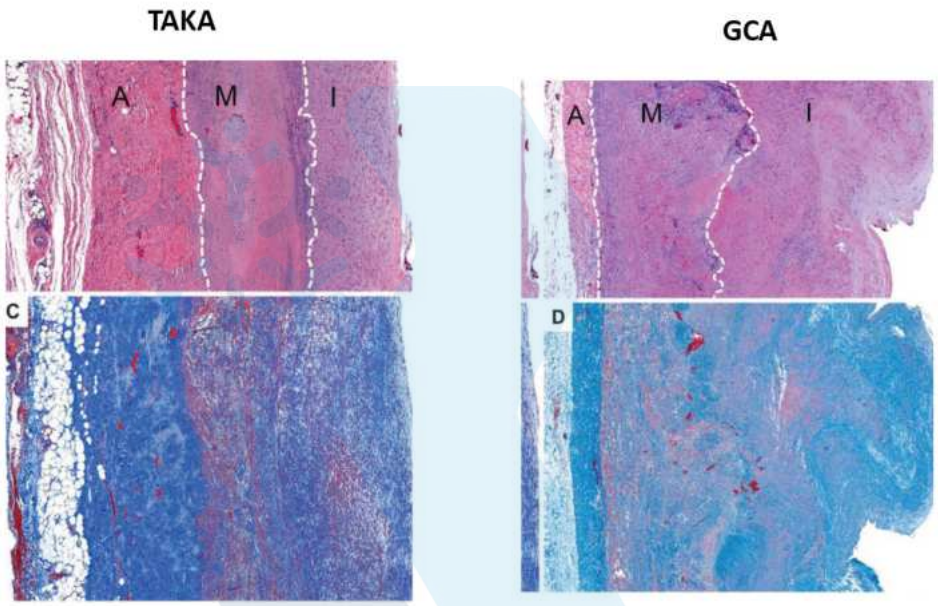




Cephalic arteritis, symetrical subclavian and axillary involvement temporal artery biopsy++

Saadoun Orphanet J Rare Dis 2021 Watanabe Curr Rheumatol Rep 2022

TA AND GIANT CELL ARTERITIS



- (M)edia damage
- (I)ntima thickening

Inflammation + fibrosis

- (A)dventitia fibrous

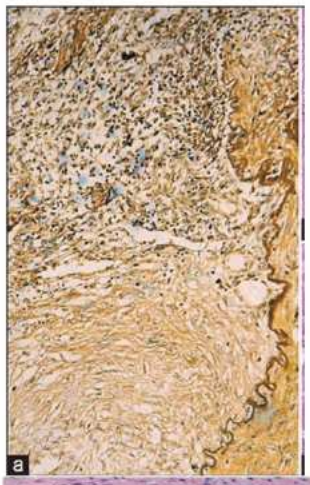
Expansion and remodeling

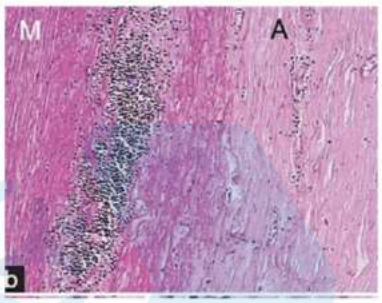
- (M)edia damage
- (I)ntima thickening

Inflammation + fibrosis

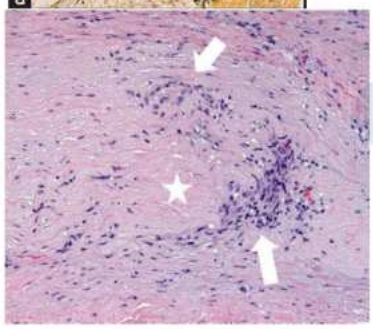
Watanabe Curr Rheumatol Rep 2022

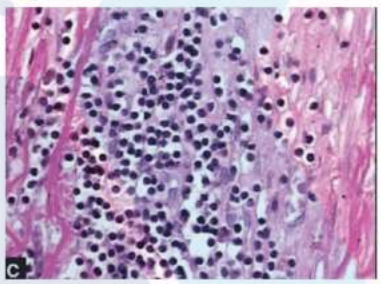
TA AND GIANT CELL ARTERITIS





Cluster of mononuclear cells in the media and fibrosis of the adventitia





Infiltrate:
lymphocytes
few Plasma cells
histiocytes

Granulomatous inflammation of descending aorta media-adventitial junction

TA histologic lesions are close from GCA but.....

Watanabe Curr Rheumatol Rep 2022/Bhandari Cureus 2023

TA AND GIANT CELL ARTERITIS

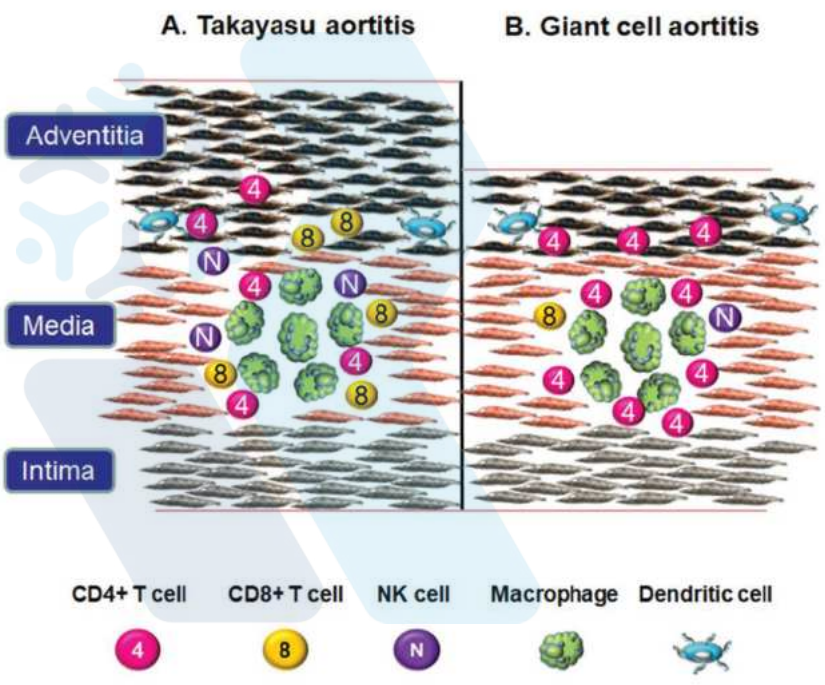
- IL-12B susceptibility gene
- Increased IL-12

↓

CD4 TH-1 and TH-17

CD8 perf+ NK

B Lympocytes: Pro-inflammatory anti-EC Ab



CD4/CD8/NK/ B — **CD4 disease**

Saadoun A&R 2015/Watanabe Curr Rheumatol Rep 2022/Bhandari Cureus 2023

TA INFLAMMATORY ACTIVITY

Difficult to evaluate!! **Combination of clinical, biological and imaging criteria**

- **NIH: Recent onset or aggravation of at least 2 criteria among 4**

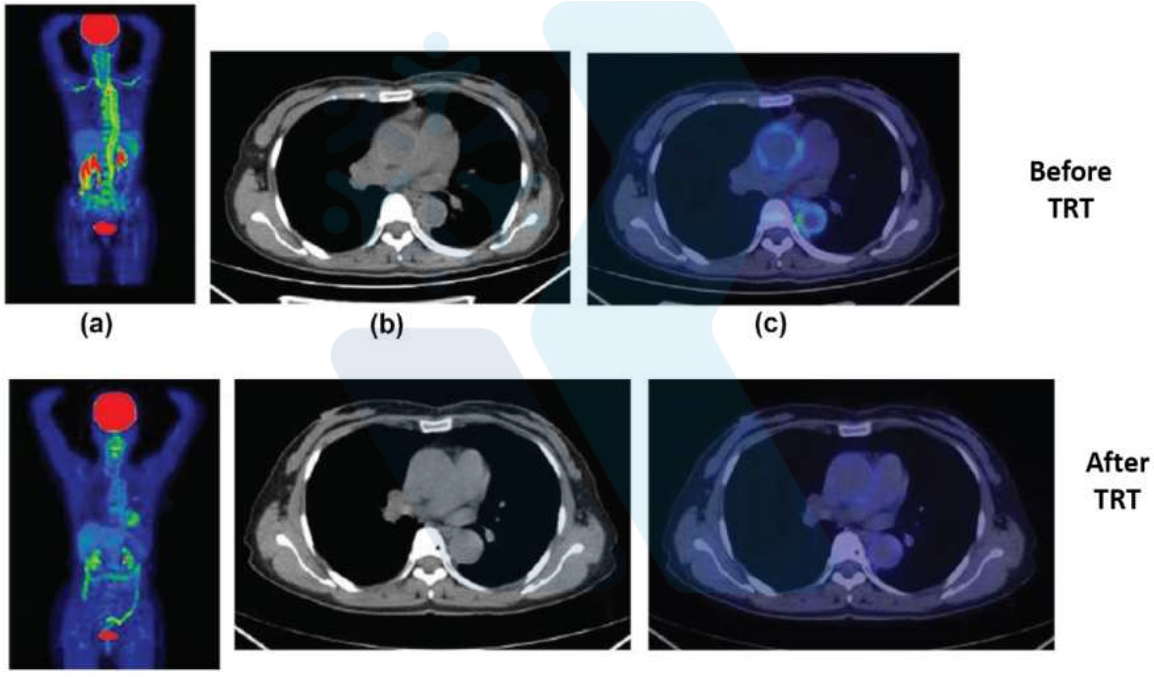
  -Signs of ischemia or vascular inflammation
  -Systemic symptoms not attributable to other events
  -Angiographic abnormalities (CT, MRI)
  -Increased inflammatory markers (CRP, fibrinigen, ESR) (but may be unadapted in 30%)

- **Turkish Disease Extend Index- Takayasu (DEI-T)**
- **Indian Takayasu (ITAK)**

Need for more precise tools.....

TA INFLAMMATORY ACTIVITY

2-[18-F]FDG-PET/CT

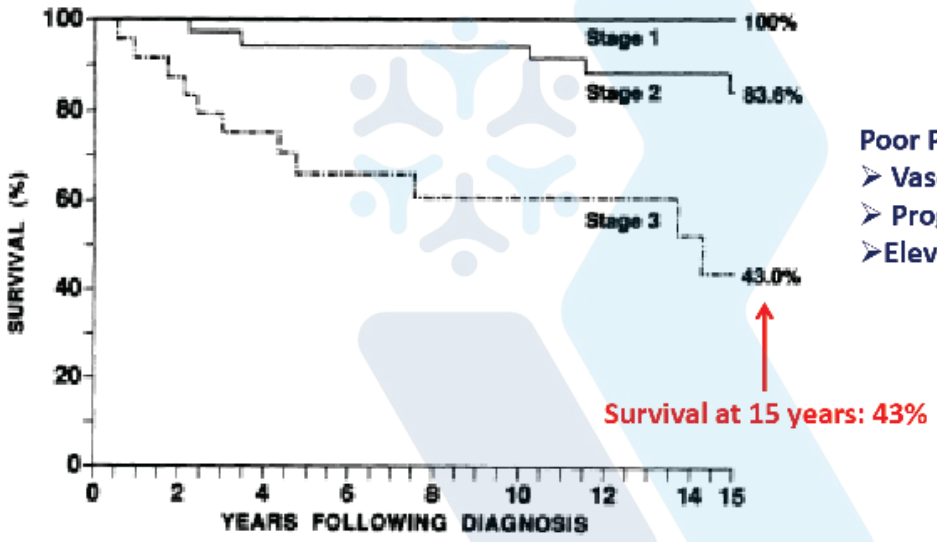


NOT YET ESTABLISHED.....

Barra AutoImm Rev 2018/Nassarmandji Front Med 2024/Luo Clin Radiol 2024

PROGNOSIS
HIGHLY VARIABLE

N= 120
Retrospective
14 year
follow-up



Poor Prognosis factors
- Vascular Complications
- Progressive disease
- Elevated ESR

Ishikawa K et al. Circulation 1994

TA AND DEATH

Death (n=16/300; 5%), survival 96.1% at 10years

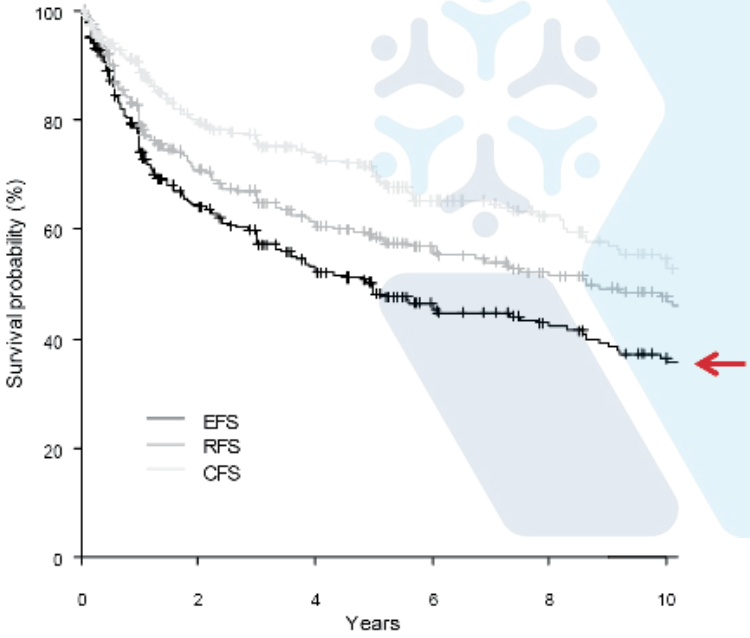
Aneurysm rupture/dissection	**25%**
Mesenteric ischemia	**25%**
Septic shock	19%
Cardiac arrest	19%
Pulmonary emboli	6%
Cardiogenic shock	6%

Matsuura	76,1% at 15 y	69 patients
Endo	86,5% at 5 y 81,4% at 10 y	31 patients
Sheikhzadeh	92,4% at 6 y	78 patients
Miyata	73,5% at 20 y	106 patients
Kerr	2 deaths	60 patients

PROGNOSIS
HIGHLY VARIABLE

178 (56%) patients had ≥ 1 event:
Vascular Complication and/or relapse, and/or death

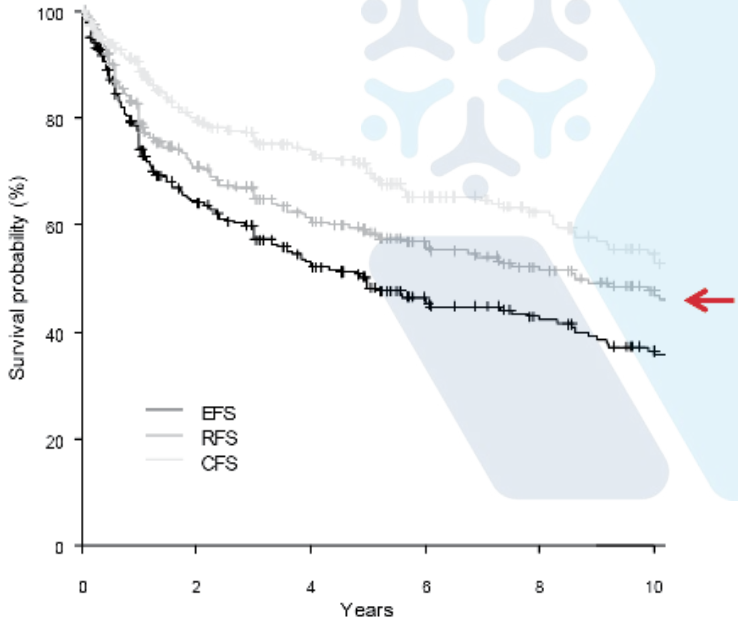
Retrospective
N= 318
Mean age 36
87% female
6 year follow-up



Event-free survival:
1 year 75.6% (70.7–80.8)
3 years 59.9% (54.–66.2)
5 years 48.2% (42.2–54.9)
10 years 36.4% (30.3–43.9)
15 years 27.1% (20.9–35.3)
20 years 17.3% (11.5–26.2)

Comarmond C et al. Circulation 2017

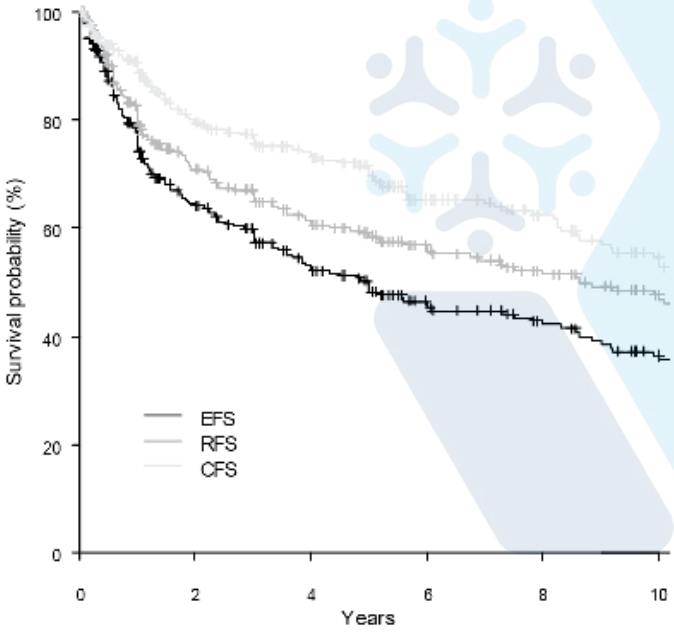
PROGNOSIS
RELAPSE



Relapse-free survival:
1year 80.1% (75.5–85.0)
3 years 66.6% (61.1–72.6)
5 years 58.6% (52.7–65.1)
10 years 47.7% (41.2–55.1)
15 years 39.6% (32.6–48.2)
20 years 32% (24.4–42.0)

Comarmond C et al. Circulation 2017

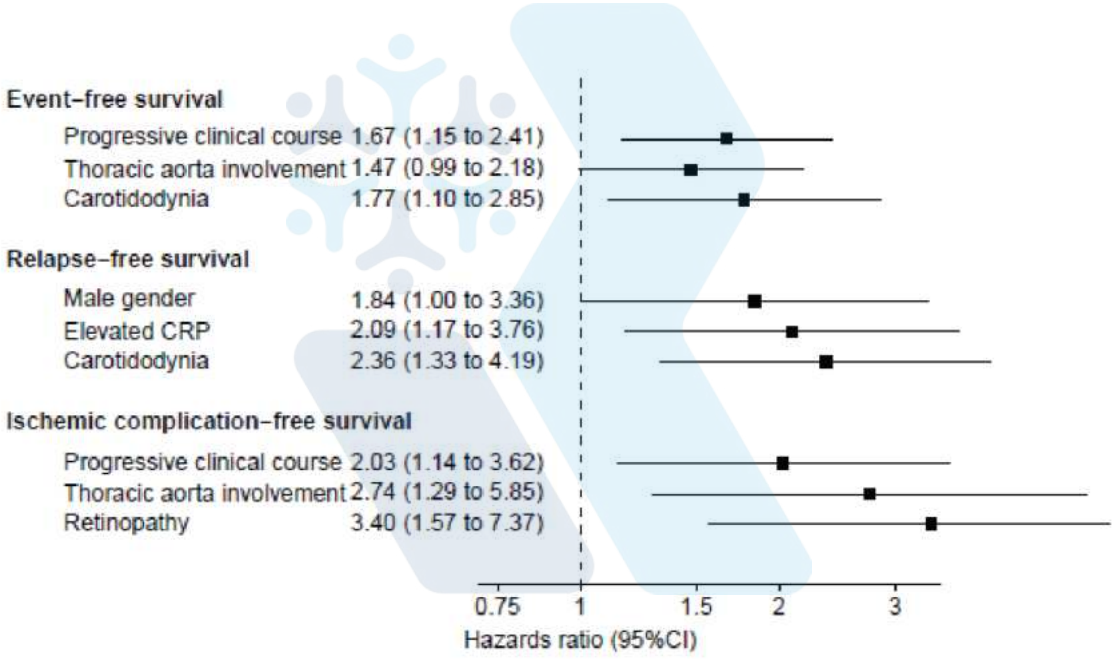
PROGNOSIS
VASCULAR COMPLICATIONS



Vascular complication-free survival:
1 year 90% (95%CI: 86.6–93.6)
3 years 77.4% (72.4–82.7)
5 years 69.9% (64.3–76.0)
10 years 53.7% (46.8–61.7)
15 years 47.0% (39.5–56.1)
20 years 28.2% (19.9–39.8)

Comarmond C et al. Circulation 2017

PROGNOSIS FACTORS

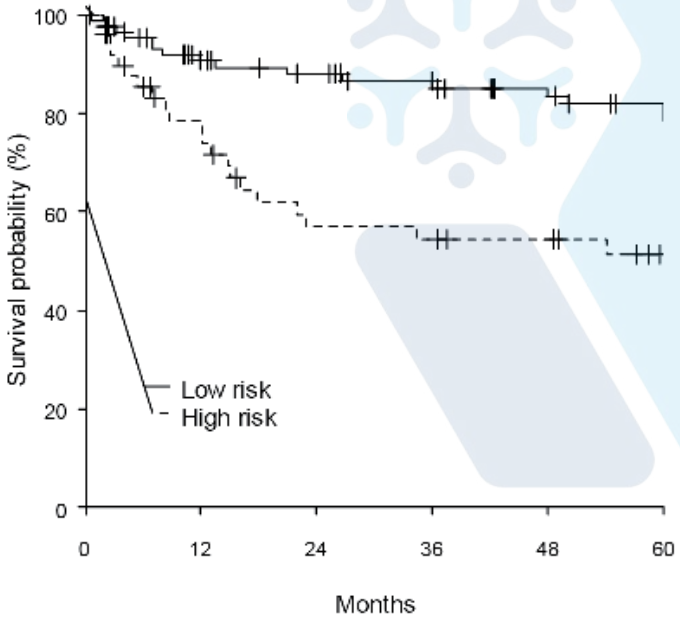


Comarmond C et al. Circulation 2017

PROGNOSIS SCORE

Patients are at high risk of death and vascular complications si ≥ 2 following factors

- **Progressive evolution**
- **Thoracic aortic involvement**
- **Retinopathy**



Low risk group:
21.6% complications at 5 years

High risk group:
48.5% complications at 5 years

Comarmond C et al. Circulation 2017

TREATMENT

OBJECTIVES:

- **Limit vascular complications and revascularization and control vascular inflammation**
- **Avoid relapses**
- **Limit corticosteroid side-effects**
- **Reduce cardiovascular risk factors, such as tobacco use and high blood pressure**
- **Anti-platelet antiaggregant drugs if no contraindication (aspirin 75-300 mg/d if severe stenosis)**
- **Anti-coagulant: not recommended routinely**
- **Statins: often but no evidence-based data**

INFLAMMATORY PHASE: steroids, oral DMRDs, bio-DMRDs

VASCULAR COMPLICATION: Revascularization only when inflammation is under control

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TREATMENT

STEROIDS

- Gold-standard TRT for the active inflammatory phase
- **Prednisone: 0.5 mg/kg (mild forms)**
  0.7 to 1 mg/kg (<70mg/d in case of vital organ damage, coronaritis, multiple or progressive arterial lesions, reno-vascular hypertension, intestinal ischemia, cerebral infarction, aortic insufficiency)
- Avoid methylprednisolone bolus except in life-threatening forms
- Progressive reduction until 5mg/d, then 1mg/month, then attempt to stop at 24 months of quiescent disease
- May achieve **remission in 25-50% of cases**
- But **relapses in 30-40% of cases**
- **Side effects: 20-50% of patients**

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TREATMENT

STEROIDS

Corticosteroid therapy started at 0.5 mg/kg				
Weight (kg)	≥ 70	60 to 70	50 to 60	40 to 50*
	Prednisone at mg/day			
D1 to D14	35	30	25	20
M1	17.5	15	12.5	10
M3	10	7.5	7.5	7.5
M6	≤ 0.1 mg/kg			
M12	5			

Corticosteroid therapy started at 1 mg/kg				
Weight (kg)	≥ 70	60 to 70	50 to 60	40 to 50*
	Prednisone at mg/day			
D1 to D14	70	60	50	40
M1	35	30	25	20
M3	20	17.5	15	12.5
M6	≤ 0.1 mg/kg			
M12	5			

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TREATMENT

ORAL DMARDs

- **METHOTREXATE:**
  - -0.3mg/kg/wk (20 mg/wk for 70 kg)
  - -Most experts recommend its use as a first-line to reduce steroids
  - -only limited open studies
  - -50-80% remission in combination with steroids
  - -Teratogenic
- **AZATHIOPRINE:**
  - -2-3mg/kg/d (max: 150mg/d)
  - -Remission and steroid reduction but non-anatomical improvement
- **MYCOPHENOLATE MOFETIL:**
  - -2g/d
  - -Clinical remission and steroid reduction
- **CICLOPHOSPHAMIDE:**
  - -No more used

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TREATMENT

Bio-DMARDs

- **Anti-TNF**
  - -Mainly **INFLIXIMAB** (5-6mg/kg 0-2-4 wk and every 4 wks)
  - -**ADALIMUMAB** (40 mg/2wks)
  - -Especially in case of association with spondylarthropathy or inflammatory bowel diseases
- **Anti-IL-6r**
  - -**TOCILIZUMAB** (8mg/kg/4wks)

EFFICIENT
70-80% response
Steroid-sparing effect
Even in patients refractory to oral DMRDs

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Anti-TNF in Takayasu

	Total	Infliximab	Etanercept	Adalimumab
Hoffman et al. 2004	15	8	7	0
Della Rossa et al. 2005	2	2	0	0
Karageorgaki et al. 2007	4	4	0	0
Filocamo et al. 2008	4	4	0	0
Molloy et al. 2008	25	21	4	0
Nunes et al. 2010	3	3	0	0
Buonuomo et al. 2011	2	2	0	0
Osman et al. 2011	2	1	0	1
Comarmond et al. 2012	5	5	0	0
Mekinian et al. 2012	15	15	0	0
Schmidt et al. 2012	20	17	1	2
Quartuccio et al. 2012	15	15	0	0
Bonilla-Abadía et al. 2013	3	3	0	0
Novikov et al. 2013	9	8	0	1
Youngstein et al. 2014	8	8	0	0
Serra et al. 2014	5	2	0	3
Mekinian et al. 2015	35	28	6	1
Vinicki et al. 2016	4	4	0	0

+ 8 reported cases

Retrospective or small case series

Anti-IL6r in Takayasu

Case series

Nishimoto et al. 2008	1
Seitz et al. 2011	2
Salvarani et al. 2012	2
Bredemeier et al. 2012	1
Salvarani et al. 2012	1
Unizony et al. 2012	2
De Kruif et al. 2012	1
Bravo Mancheño et al. 2012	1
Goel et al. 2013	10
Xenitidis et al. 2013	2
Abisror et al. 2013	5

Tombetti et al. 2013	4
Nakaoka et al. 2013	4
Cañas et al. 2014	8
Loricera et al. 2014	7
Youngstein et al. 2014	3
Schiavon et al. 2014	1
Mekanian et al. 2015	14
Osman et al. 2015	3
Vinicki et al. 2016	1
Risse et al. 2016	1
Deniz Batu et al. 2017	4

Efficacy and safety of tocilizumab in patients with refractory Takayasu arteritis: randomised, double-blind, placebo-controlled, phase 3 trial in Japan (the TAKT study)

Hors AMM

- **Inclusion:**

Patients with TAK who had relapsed within the previous 12 weeks were induced into remission with oral glucocorticoid therapy.

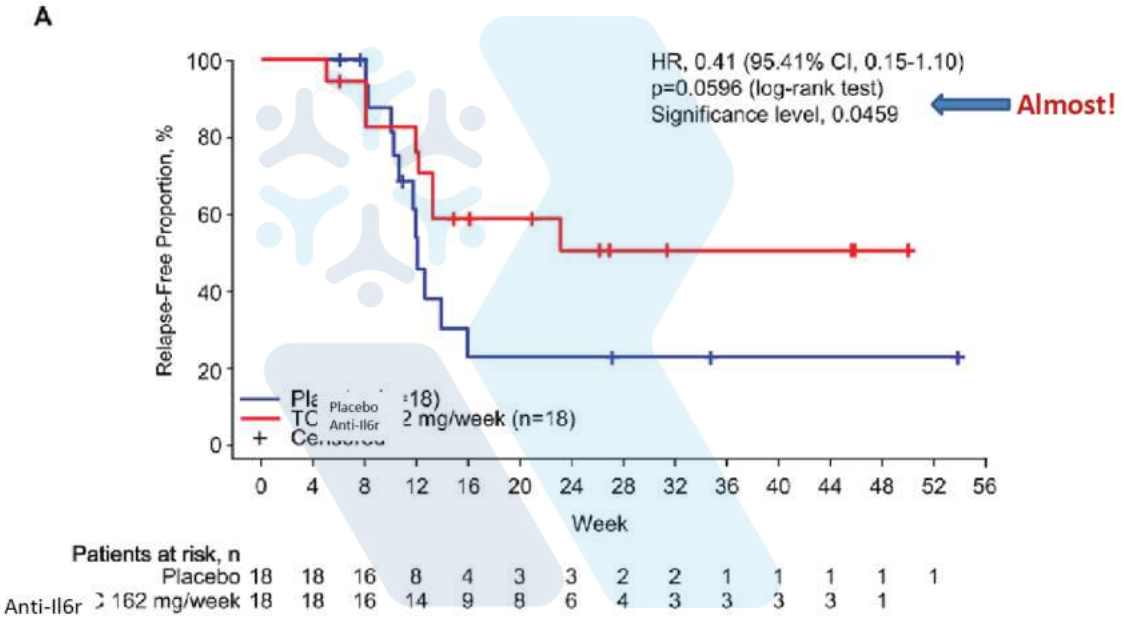
- **Design:** Double-blind, placebo-controlled trial, patients were randomly assigned 1:1 to receive **anti-Il6r or placebo subcutaneously** and **oral glucocorticoids were tapered 10 %/week from week 4 to a minimum of 0.1 mg/kg/day** (until 19 patients relapsed).

The first dose of study treatment was administered after remission from TAK for ≥1 week.

- **The primary endpoint** was the time to relapse of TAK, defined as ≥2 of the following:
  - objective systemic symptoms,
  - subjective systemic symptoms, **without imaging evaluations**
  - elevated inflammation markers,
  - vascular signs
  - ischaemic symptoms.

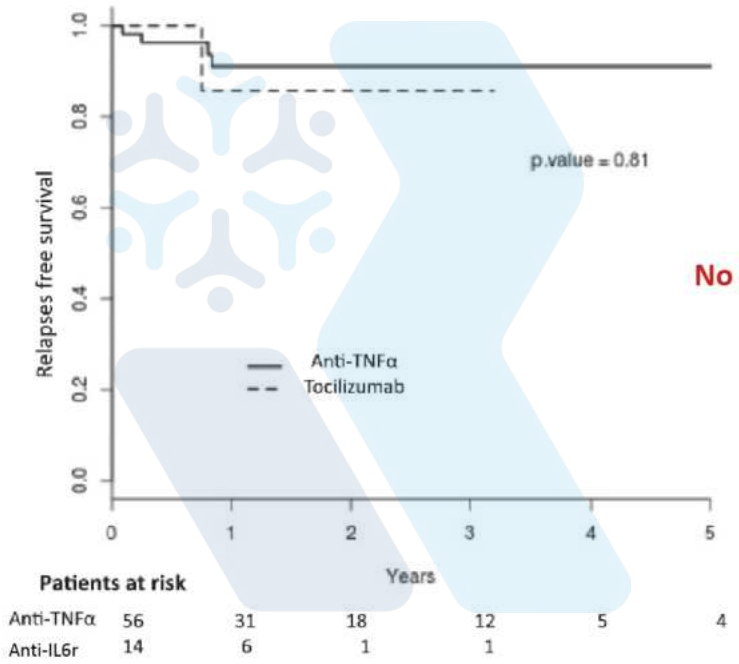
Nakaoka Y et al. Ann Rheum Dis 2018

Patients with refractory Takayasu arteritis: randomised, double-blind, placebo-controlled, phase 3 trial in Japan (the TAKT study)



Nakaoka Y et al. Ann Rheum Dis 2018

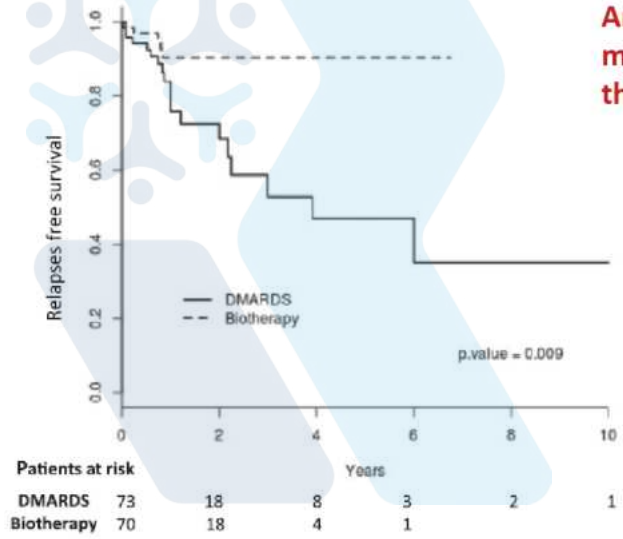
Anti-TNF vs anti-IL6r



No difference

Mekinian et al Circulation 2015

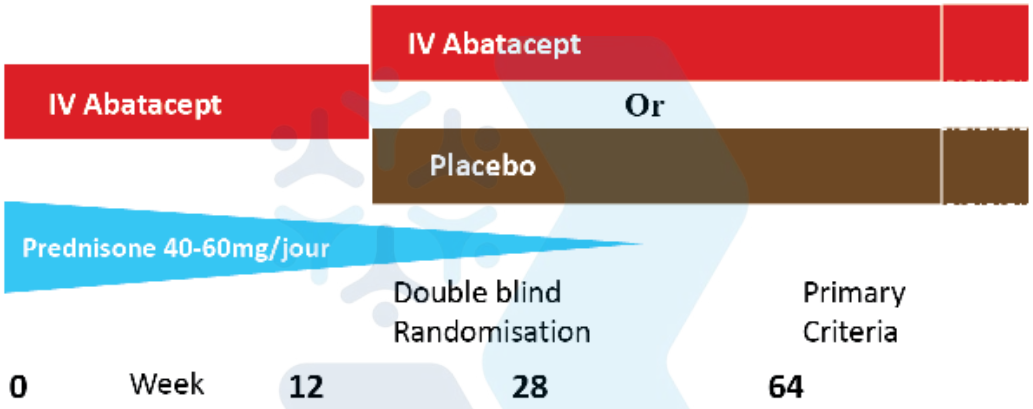
Bio-DMARDs vs DMARDs



Anti-IL-6r appears much better than oral-DMARDs

Mekinian et al Circulation 2015

Abatacept (CTLA4-Ig) in Takayasu Arteritis



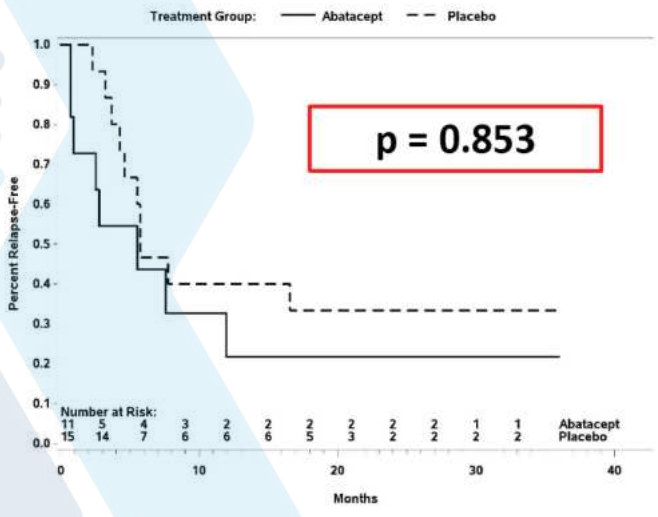
- **Primary criteria: Remission duration (relapse-free survival)**
- **Secondary criteria: Tolerance**

Langford CA et al. Arthritis Rheumatol 2017

Abatacept (CTLA4-Ig) in Takayasu Arteritis

34 treated patients
26 randomized patients
11 abatacept, 15 placebo

Relapse-free survival at 12 months
22% abatacept
40% placebo (p=0.853)



Abatacept does not reduce relapse rate in TA

Langford CA et al. Arthritis Rheumatol 2017

OTHER bio-DMARDs in Takayasu

Ustekinumab

- *Terao et al. Scand J Rheumatol 2015;27:1*

– 3 patients

- *Yachoui et al. Scand J Rheumatol 2017:Mar 3:1*

Too limited

Rituximab

- *Hoyer et al. Ann Rheum Dis 2012; 71:75*

- 3 patients with refractory TAK

- *Galarza et al. Clin Rev All Immunol 2008; 34:124*

- 2 patients

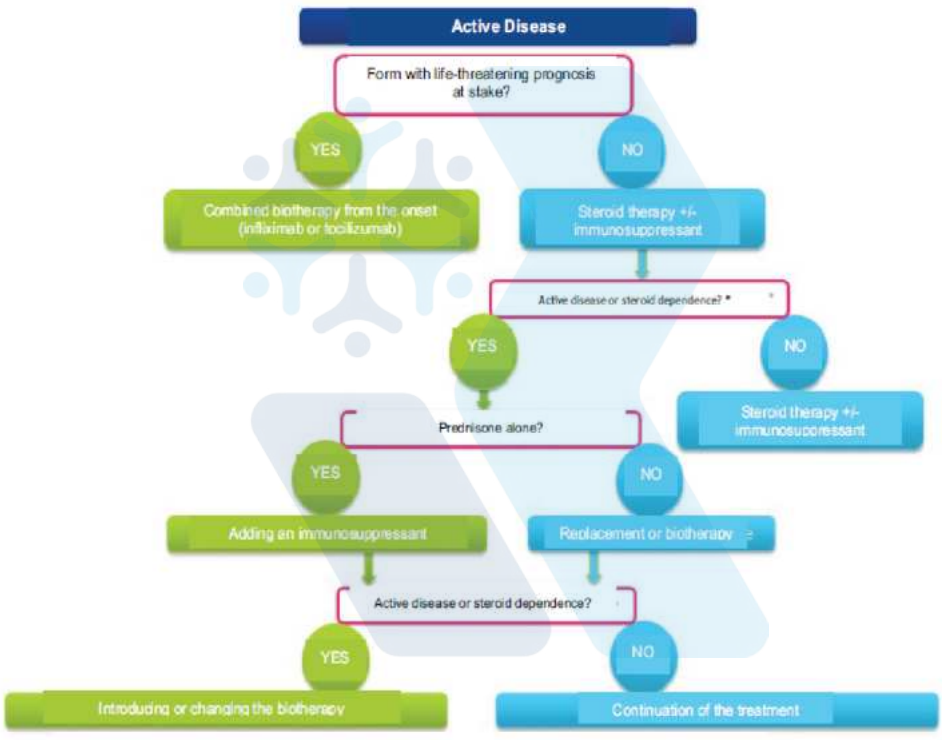
- *Ernst et al. Case Rep Rheum 2012: Epub*

- 1 patient

- *Salvarani et al. Rheumatology* 2017

- 7 patients

FRENCH GUIDELINES TO TREAT TAKAYASU ARTERITIS



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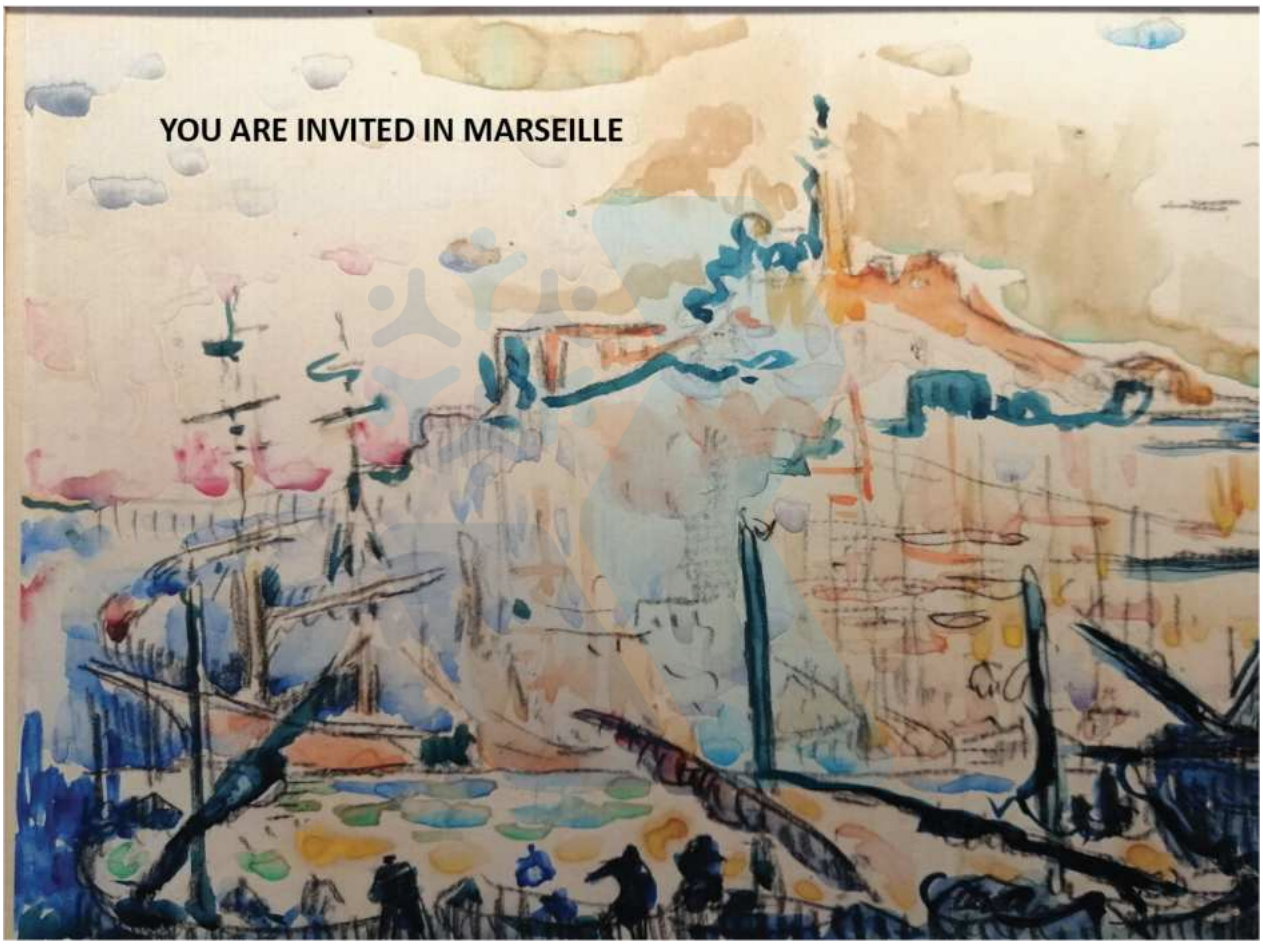
CONCLUSIONS

- TA is different from GCA
- TA is a more severe disease than GCA
- Appropriate vascular imaging is the mainstay of diagnosis
- Evaluate the inflammatory activity is not easy and relies on clinical, biological and imaging
- Steroids are the first-line therapy, possibly in association with methotrexate
- 50% of patients relapse and have vascular complications 10 years after diagnosis
- High risk patients are those with progressive course, retinopathy, thoracic aortic involvement
- High risk patients may be treated more agressively with early bio-DMARDS



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