Current Advances In The Treatment For Inflammatory Aneurysms: How Effective Are EVAR And TEVAR

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I have **no financial relationships** to disclose.

The most common inflammatory diseases causing aortic aneurysms;

Large Size Vessel Vasculitis:Giant Cell Disease (GCD),Takayasu Disease (TAK)

Medium Size Vessel Vasculitis:Kawasaki Disease(KD),Periarteris Nodosa(PAN)

Small Size Vessel Vasculitis(SVVs):

ANCA-ascoiated Vasculitis(AAVss);GPA,MPA,EGPA

IgA vasculitis(Henoch-Schoenlein Purpura)

Hypocomplementaemc vasculitis (HUV)

Behcet's Disease(BD)

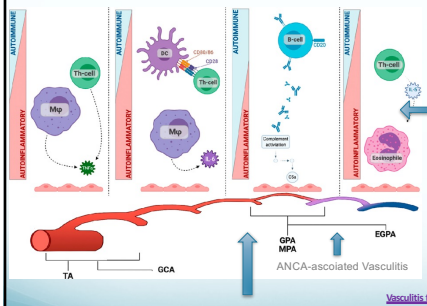
Systemic Lupus Erythamatousis (SLE)

Rheumatid Artiris (RA)

Classification and epidemiology of vasculitis: Emerging concepts
Ecclestone T, Watts RA.Best Pract Res Clin Rheumatol. 2023 Jul 17:101845.doi:10.1016/j.berh.2023.101845. Online ahead of print.PMID: 37468418 Review

The American College of Rheumatology (ACR)-European Alliance of Associations for Rheumatology (EULAR) classification;

The differentiation of TA from GCA (LVVs) and EGPA from GPA/MPA (AAVs) is important.



The central immune cells within the pathology of vasculitis subtypes and an estimates weighing of **autoinflammatory-autoimmune disease mechanisms**

Chapel Hill classification of vasculitis according to vessel size.

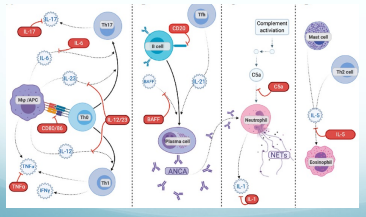
Vasculitis therapy refines vasculitis mechanistic classification
Torp CK, Brüner M, Keller KK, Brouwer E, Hauge EM, McGonagle D, Kragstrup TW.
Autoimmun Rev. 2021 Jun;20(6):102829. doi: 10.1016/j.autrev.2021.102829. Epub 2021 Apr 16.

Drugs for inflammatory diseases are;

Glucocorticoids,

Conventional synthetic disease modifying anti-rheumatic drugs (csDMARDs);
Methotrexate,cyclophosphamide,
azathioprine, cyclosporine and mycophenolate.

Targeted biological disease modifying anti-rheumatic drugs (bDMARDs) ;
Tocilizumab,Infliximab,Etanercept, ,Adlimumab,Secukinumab,Ustekinumab,Abatacept,
Mepolizumab ,anti-TNF) and interferon-α (IFNα) Jak1,JAK3



Immunologic pathways targeted by the drugs;
LVVs ;GCA and TAK
AAVs ;GPA, MPA, and EGPA.

Giant Cell Disease (GCD);

Abnormal maturation of vascular dendritic cells (DC) in the adventitia of the large-vessel walls.

Genes:Human leucocyte antigen (HLA) class II.

Proinflammatory cytokines can increase the patient's risk of ischaemic complications, PMR and relapsing disease.

Ann Rheum Dis. 2022 Dec;81(12):1647-1653.doi: 10.1136/ard-2022-223480. Epub 2022 Nov 9.
2022 American College of Rheumatology/EULAR classification criteria for giant cell arteritis

Giant Cell Disease (GCD);

2022 ACR-EULAR Classification Criteria for Giant Cell Arteritis

Criteria absolute requirement	
Age ≥ 50 years at the time of diagnosis	
Criteria Items	
Clinical Features	
Morning stiffness in shoulders/neck	+2
Sudden visual loss	+3
Jaw or tongue claudication	+2
New temporal headache	+2
Scalp tenderness	+2
Abnormal examination of the temporal artery[1]	+2
Investigations	
Laboratory	
Maximum ESR ≥ 50 mm/hour or maximum CRP ≥ 10 mg/L[2]	+3
Biopsy/Imaging Findings	
Positive temporal artery biopsy or halo sign on temporal artery ultrasound[3]	+5
Bilateral axillary involvement[4]	+2
FDG-PET activity throughout aorta[5]	+2

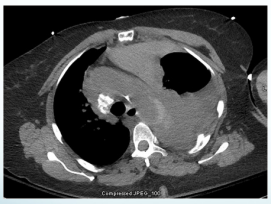
Sum the scores for all items, if present.
A score of ≥ 6 is needed for the classification of giant cell arteritis

1. Examination of the temporal artery showing absent or diminished pulse, tenderness, or hard 'cord-like'.
2. Maximum erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) values prior to the initiation of treatment for vasculitis.
3. Presence of either definitive vasculitis on temporal artery biopsy or halo sign on temporal artery ultrasound. There are no specific histopathologic criteria to define definitive vasculitis on temporal artery biopsy. The presence of giant cells, mononuclear leukocyte infiltration, and the fragmentation of the internal elastic lamina were independently associated with histopathologic interpretation of definite vasculitis in the Diagnostic and Classification Criteria in Vasculitis Study (DCVAS) cohort [24]. Halo sign is defined by the presence of a homogenous, hypoechoic wall thickening on ultrasound [25].
4. Bilateral axillary involvement is defined as luminal damage (stenosis, occlusion, or aneurysm) on angiography (computed tomography, magnetic resonance, or catheter based) or ultrasound, halo sign on ultrasound, or fluorodeoxyglucose uptake on positron emission tomography.

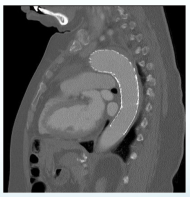
Abnormal fluorodeoxyglucose (FDG) uptake in the arterial wall (e.g., greater than liver uptake by visual inspection) throughout the descending thoracic and abdominal aorta on positron emission tomography (PET).

Giant Cell Disease (GCD);

Salaskar et al. CVIR Endovascular (2019) 2:9 https://doi.org/10.1186/s42155-019-0052-CVIR Endovascular
Thoracic endovascular repair of a rare case of leaking aortic arch intramural hematoma secondary to Giant cell arteritis
Abhijit Salaskar*, Farzad Najamz, Elizabeth Pococks and Shawn Sarins



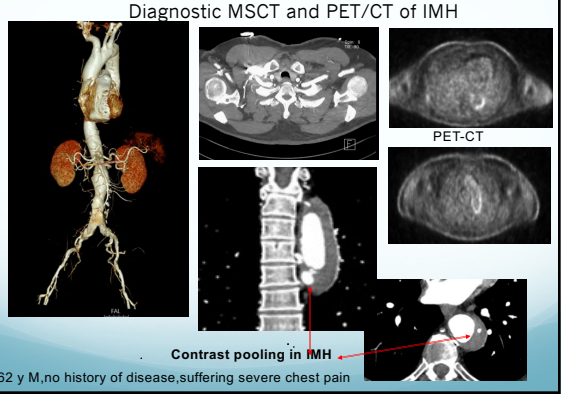
Pre-TEVAR CTA (axial view of non-contrast phase) showing crescentic aortic IMH originating in the distal aortic arch near the takeoff of the left subclavian artery and large left sided hemothorax



Follow up CTA (sagittal view) one month after TEVAR showing widely patent aortic stent graft without any evidence of extravasation or endoleak

Giant Cell Disease (GCD);

Diagnostic MSCT and PET/CT of IMH

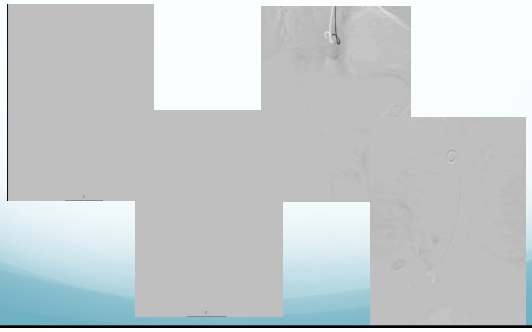


62 y M,no history of disease,suffering severe chest pain

Giant Cell Disease (GCD);

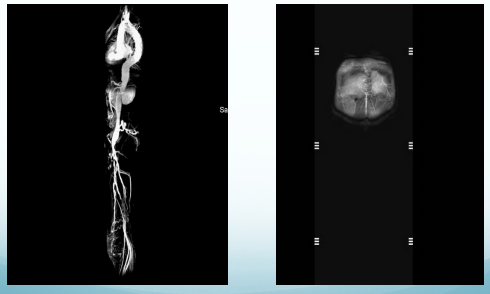
After diagnosis and having medication

DSA /TEVAR



Giant Cell Disease (GCD);

Post-TEVAR and Post-op Abdominal Aneurysm MR



Takayasu Disease (TAK);

Inflammation in TA is around the **vasa vasorum** and at **the medio-adventitial junction**, leading to **panarteritis** causing stenosis or aneurysms.

Inflammatory infiltrates are ; macrophages and various lymphoid cells, including CD4+ and CD8+ T cells, GD T cells, natural killer (NK) cells, and B cells.

Genes:Human leukocyte antigen (HLA) class I and class II.

Vascular remodeling;

-**Stenosis:**Three stages: the "prepulseless" phase (stage I), the "pulseless" phase (stage II), and "fibrotic" phase (stage III).

-**Aneurysm formation :**
Severe or **rapid inflammation** cause the destruction of smooth muscle cells in the media result in the weakening of the arterial wall, leading to vascular dilatation and **aneurysm formation**

Takayasu Disease (TAK);

2022 ACR-EULAR Classification Criteria for Takayasu's Arteritis

Criteria Absolute Requirements	
Age ≤ 60 years at the time of diagnosis	
Evidence of vasculitis on imaging[1]	
Criteria Items	
Clinical Features	
Female sex	+1
Angina or ischemic cardiac pain	+2
Arm or leg claudication	+2
Vascular bruit[2]	+2
Reduced pulse in upper extremity[3]	+2
Carotid artery abnormality[4]	+2
Systolic blood pressure difference in arms ≥ 20 mm Hg	+1

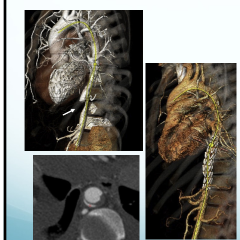
Sum the scores for all items, if present.
A score of ≥ 5 is needed for the classification of Takayasu arteritis

1. Evidence of vasculitis in the aorta or branch arteries must be confirmed by vascular imaging (e.g., computed tomographic/catheter-based/magnetic resonance angiography, ultrasound, or positron emission tomography).
2. Bruit detected by the auscultation of a large artery, including the aorta, carotid, subclavian, axillary, brachial, renal, or iliofemoral arteries.
3. Reduction or absence of pulse by the physical examination of the axillary, brachial, or radial arteries.
4. Reduction or absence of pulse of the carotid artery or tenderness of the carotid artery.
5. Number of arterial territories with luminal damage (e.g., stenosis, occlusion, or aneurysm) detected by angiography or ultrasonography from the following nine territories: thoracic aorta, abdominal aorta, mesenteric, left or right carotid, left or right subclavian, and left or right renal artery.
6. Bilateral luminal damage (stenosis, occlusion, or aneurysm) detected by angiography (computed tomography, magnetic resonance, or catheter based) or ultrasonography in any of the following paired vascular territories: carotid, subclavian, or renal arteries.
7. Luminal damage (stenosis, occlusion, aneurysm) detected by angiography or ultrasonography involving the abdominal aorta *and* either the renal or mesenteric arteries.

Classification and epidemiology of vasculitis: Emerging concepts
Ecclestone T, Watts RA.
Best Pract Res Clin Rheumatol. 2023 Jul 17:101845. doi: 10.1016/j.berh.2023.101845. Online ahead of print.PMID: 37468418 Review

Takayasu Disease (TAK);

JACC: Cardiovascular InterventionsVolume 11, Issue 12, 25 June 2018, Pages ee101
Endovascular Management of Acute Aortic Dissection in Takayasu Arteritis
Author links open overlay panelSanjay Tyagi MD, D



Rheumatol IntDOI 10.1007/s00296-012-2598-7
Multiple endovascular stent-graft implantations in a patient with aortic thoracic and abdominal aneurysms due Takayasu arteritisF.
Bonilla-Abad́ıa • A. F. Echeverri • J. P. Carbonell • C. A. Cañas

Behcet's Disease(BD);

Behçet syndrome(BD) is one of the **systemic vasculitis** which can affect almost every organ.

Arterial involvement affects **3–5%** of the patients

.
Arterial aneurysms are the most common vascular lesion **(47%)** and the result of disrupted medial elastic fibres , and inflammatory cell infiltration, around the **vasa vasorum.**

Genes; HLA-B*51, HLA-I .

Post-op pseudo-aneurysms occur 30%-50% of cases after surgical repair at the anastomotic site .

EVAR/TEVAR seems to provoke **less(wall stress) cytokine release** and **inflammatory response.**

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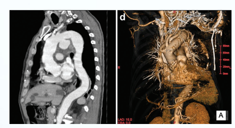
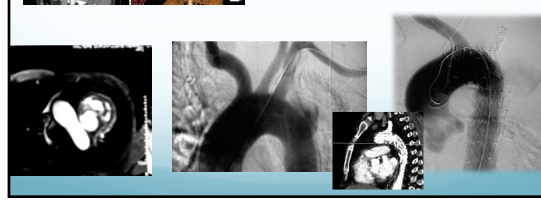
Nature reviews rheumatology Review article Check for updates Review Article Published: 21 December 2022
Vascular Behcet syndrome: from pathogenesis to treatment
Alessandra Bettiol, Fatma Alibaz-Oner, Haner Direskeneli , Gulen Hatemi, David Saadoun, Emire Seyahi, Domenico Prisco & Giacomo Emmi

Behcet's Disease(BD);

Interactive CardioVascular and Thoracic Surgery 12 (2011) 502–504
Rapidly growing aortic arch aneurysm in Behcet's disease
Nozomi Kojima, Yasuhito Sakano, Shin-ichi Ohki, Yoshio Misawa*Division of Cardiovascular Surgery, Jichi Medical University,

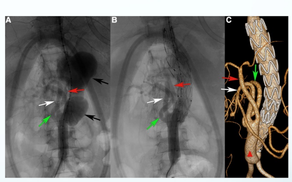




Behcet's Disease(BD);

Front. Cardiovasc. Med., 01 September 2023,Sec. Aortic Surgery and Endovascular Repair,Volume 10 - 2023
| https://doi.org/10.3389/fcvm.2023.1097129 Case Reports in Aortic Surgery and Endovascular Repair: 2023
Hybrid surgery management challenges of a Behcet's disease patient with recurrence of aortic neurysms: a case report
Guo Xin Chen Jiang Hong Wan Chu Wen Chen Bin Huang

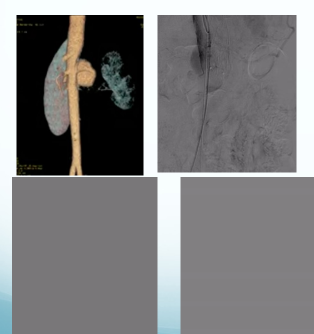
A- abdominal aortic pseudoaneurysm (black arrow).
B- CT Angiography after **pseudoaneurysm resection** and abdominal aorta reconstruction.



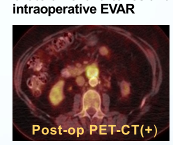
A,B- Angiography after completion of splanchnic artery branch reconstruction. andafter completion of stent placement.
C-After 5 years of follow-up, CTA showed that the pseudoaneurysm cavity completely disappeared, and the stent and branch arteries were patent.

Behcet's Disease(BD);

Case:50y,F,BD,Juxtra-renal saccular aneurysm at the level of visceral arteries



By-passes of CT,SMA and bilateral renal arteries and intraoperative EVAR



Secondary intervention after 2 year ;
L renal artery oclusion, distal endoleak,distal aortic cuff extension

Systemic lupus erythematosus(SLE);

The **vasculitis involving the vaso vasorum** of the aorta leads to **aneurysm formation**.

Triad of antiphospholipid antibodies ,**chronic steroid use** and **cystic medial degeneration (CMD)** associated with SLE may predispose to **premature atherosclerosis**.

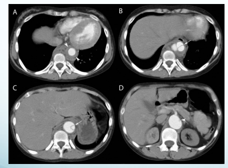
History **of hypertension**, associated **AA** and **prolonged steroid** medication **accelerate atherosclerosis** disease pattern.

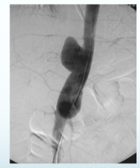
Aortic dissection should be considered in young patients having sharp chest,back or abdominal pain.

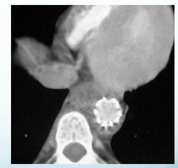
Systemic lupus erythematosus(SLE);

Endovascular stent-graft placement for ruptured dissecting aortic aneurysm in an adolescent patient with systemic lupus erythematosus: case report.

Emerg Radiol. 2011 Dec;18(6):499-502. doi: 10.1007/s10140-011-0978-z. Epub 2011 Aug 19.
Gulsen F, Cantasdemir M, Ozluk E, Arisoy N, **Numan F**

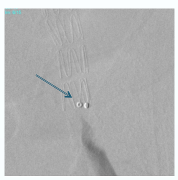


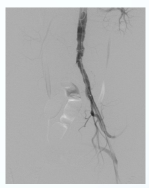




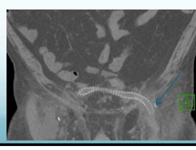
Systemic lupus erythematosus(SLE);

Case:64y,M,SLA.During f-u and monitoring had ulcerated plaque at AA.





Spontaneous dissection during EVAR procedure which was treated via self-expandable **stent**



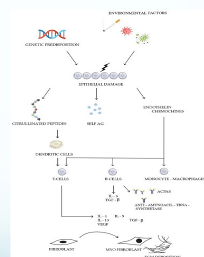
Post-op **suture failure** twice at anostomosis of the fem-fem by-passes.

Rheumatoid Artiris;

[illegible]

RV is the most **serious extra-articular complication** and can cause **high rates of morbidity and mortality.**

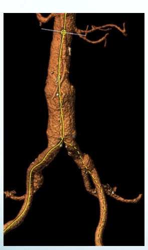
Aortic aneurysms/dissection formation is very rare.

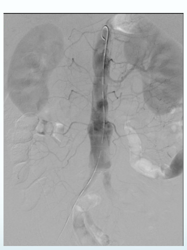


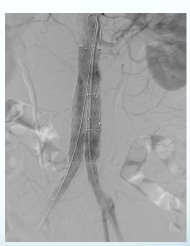
Cardiovascular manifestations of RA include predilection for **accelerated atherosclerosis and endothelial dysfunction** resulting in coronary artery disease (CAD), stroke, congestive heart failure, and peripheral arterial disease.

Rheumatoid Artiritis(RA);

Case:60y ,M, rheumatoid arthritis,AAA having thick thrombosis was montoring for a while, at diameter of 65 mm EVAR had been applied.







Periarteritis Nodosa (PAN);

PAN is a necrotizing vasculitis.

Infectious ;The reduction in the incidence of PAN may be related to the decrease in hepatitis B virus **(HBV)** infection due to widespread vaccination.

Genetic; **DADA2** (Deficiency of adenosine deaminase 2) is resposable **early-onset PAN.**

Diagnosis:
Negative ANCAs (Antineutrophil cytoplasmic antibodies)

DSA;1- 5-mm diameter **m**icroaneurysms , ectasia, or occlusive disease in celiomesenteric and renal arteries

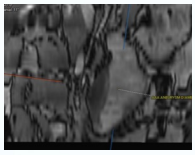
Treatment:
Non–HBV-PAN: Corticosteroids and immunosuppressants

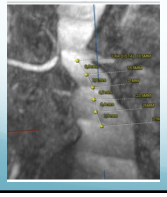
HBV-PAN: Antiviral agents, plasma exchanges and initial corticosteroids

DADA2:TNF inhibitors

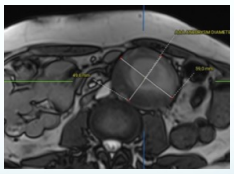
Medium- and Large-Vessel Vasculitis
David Saadoun, Mathieu Vautier and Patrice Cacoub
Originally published19 Jan 2021https://doi.org/10.1161/CIRCULATIONAHA.120.046657Circulation.2021;143:267–282

Periarteritis Nodosa (PAN);





45 y,F;During f-u of PAN rapid growth of AAA



Conclusion

EVAR/TEVAR treatment of inflammatory diseases is an effective and life saving procedure,but also have some limitations and complications .

Monitoring closely is mandantory during life time due to the nature of these diseases.