

Evolut Clinical Guideline 2020 for Chest Computed Tomography Angiography (CTA)

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STATEMENT

General Information

- *It is an expectation that all patients receive care/services from a licensed clinician. All appropriate supporting documentation, including recent pertinent office visit notes, laboratory data, and results of any special testing must be provided. If applicable: All prior relevant imaging results and the reason that alternative imaging cannot be performed must be included in the documentation submitted.*
- *Where a specific clinical indication is not directly addressed in this guideline, medical necessity determination will be made based on widely accepted standard of care criteria. These criteria are supported by evidence-based or peer-reviewed sources such as medical literature, societal guidelines and state/national recommendations.*
- *The guideline criteria in the following sections were developed utilizing evidence-based and peer-reviewed resources from medical publications and societal organization guidelines as well as from widely accepted standard of care, best practice recommendations.*

Purpose

Computed tomography angiography (CTA) generates images of the blood vessels (arteries and/or veins) in the chest that can be evaluated for evidence of stenosis, occlusion, embolism, dissection and/or aneurysms. Chest CTA (non-coronary) is used to evaluate the arteries outside the heart in the chest (thorax). CTA uses ionizing radiation and requires the administration of iodinated contrast agent, which is a potential hazard in patients with impaired renal function.

NOTE: Authorization for CT Angiography covers both arterial and venous imaging. The term *angiography* refers to both arteriography and venography.

INDICATIONS FOR CHEST COMPUTED TOMOGRAPHY ANGIOGRAPHY (CTA)

Known or Suspected Pulmonary Embolism (PE) ^(1–5)

- Suspected pulmonary embolism:
 - High risk for PE based on shock or hypotension
 - Documented score from a validated clinical scoring system for the prediction of pulmonary embolism indicating high probability for PE (See **Clinical Assessment of Pulmonary Embolism**) with any **ONE** of the following:
 - Wells score (original version) ≥ 7
 - Geneva score (original version) ≥ 11
 - Geneva score (simplified version) ≥ 5
 - Calculated score (when sufficient clinical data are provided) from a validated

clinical scoring system (See **Clinical Assessment of Pulmonary Embolism**) for the prediction of pulmonary embolism indicating high probability for PE

- **NOTE:** Elevated D-dimer is **NOT** needed for high-risk patients; CTA Chest is indicated for high-risk even with normal D dimer
- Intermediate and Low risk for PE requires an elevated D-dimer
- Follow-up of known pulmonary embolism with symptoms (such as dyspnea, fatigue, lightheadedness and/or edema) that recur **OR** with symptoms that are persistent at 3 months following initial diagnosis
 - **NOTE:** Follow-up imaging in asymptomatic patients to determine if an embolus has resolved or to determine cessation of anticoagulation is **NOT** indicated as imaging findings may persist and are not used to determine the duration of therapy

Thoracic Aortic Disease (6–9)

Screening for Possible Thoracic Aortic Aneurysm (TAA)

- Screening in individuals with a personal history of bicuspid aortic valve when prior TTE (Transthoracic Echocardiogram) is indeterminate or abnormal:
 - **NOTE:** Typical TTE follow-up imaging intervals for bicuspid aortic valve patients:
 - Baseline study at initial diagnosis of bicuspid aortic valve
 - Follow-up imaging is based on findings on prior imaging of a dilated aorta of > 40 mm is typically every 2-3 years thereafter
- Screening in individuals at elevated risk due to family history when TTE (Transthoracic Echocardiogram) is inconclusive or insufficient as clinically indicated with any **ONE** of the following ⁽⁸⁾:
 - First-degree relatives of individuals with a known thoracic aortic aneurysm (defined as 1.5 times (> 50%) larger than the predicted aorta size based on age, sex, body size) **OR** known aortic dissection
 - First and/or second-degree relatives of individuals with heritable thoracic aorta disease (HTAD) (HTAD comprises a clinically and genetically heterogeneous group of disorders sharing the common denominator of aneurysm or dissection of the thoracic aorta)
 - **NOTE:** Previous analogous terminology includes familial thoracic aortic aneurysm and dissection (FTAAD) and non-syndromic heritable thoracic aortic disease (NS-TAD))
 - First degree relatives of individuals with a known bicuspid aortic valve
- See Imaging in Known Genetic Conditions section for additional indications for screening

Suspected Thoracic Aortic Aneurysm (TAA)

- **Asymptomatic** suspected thoracic aortic aneurysm:

- With prior indeterminate or abnormal imaging (Such as echocardiogram or chest X-ray)
- **Symptomatic** known or suspected thoracic aortic aneurysm:
 - Signs and Symptoms may include:
 - Abrupt onset of severe sharp or stabbing pain in the chest, back or abdomen (could indicate aneurysm rupture)
 - Asymmetric blood pressure between limbs
 - Acute chest or back pain and at high risk for aortic aneurysm and/or aortic syndrome (risk factors include hypertension, atherosclerosis, prior cardiac or aortic surgery, underlying aneurysm, bicuspid aortic valve, and connective tissue disorder (such as Marfan syndrome, vascular form of Ehlers-Danlos syndrome, Loeys-Dietz syndrome)

Thoracic Aortic Syndromes (7,8)

- For **suspected** acute aortic syndrome (AAS) (Such as aortic dissection, intramural hematoma and penetrating atherosclerotic ulcer) with any **ONE** of the following:
 - Prior imaging (such as echocardiogram) is suggestive of AAS
 - **High risk** patient for AAS and **one** sign/symptom concerning for AAS
 - **High risk** conditions for AAS:
 - Marfan's syndrome or other connective tissue disease, family history of aortic disease, known aortic valve disease, recent aortic manipulation and/or known thoracic aortic aneurysm
 - **Signs and symptoms** concerning for AAS:
 - Chest, back or abdominal pain described as abrupt onset, severe in intensity and/or ripping or tearing in quality
 - Pulse deficit or systolic blood pressure differential
 - Focal neurologic deficit with pain
 - New heart murmur with pain
 - Hypotension or shock
 - **Non-high-risk** patient and **two** or more signs/symptoms concerning for AAS (See above)
- For follow-up of **known** aortic syndromes (Such as aortic dissection, intramural hematoma and penetrating atherosclerotic ulcer) as clinically indicated
- Suspected vascular cause of dysphagia (from vascular compression of the esophagus) or expiratory wheezing (from vascular compression of the trachea/bronchus) with prior imaging that is indeterminate or abnormal.

Follow-Up of Known Thoracic Aortic Aneurysm ^(7,8)

- Baseline study at diagnosis then every 6-24 months
 - If there is a change in clinical status or cardiac exam, then imaging sooner than 6 months is indicated

Postoperative Follow-up of Aortic Repair ^(7,8)

- Follow-up after thoracic endovascular aortic repair (TEVAR) at the following intervals if there is a reason for CTA rather than CT:
 - Baseline study at 1-month post-EVAR
 - Annually thereafter if stable
 - More frequent imaging (as clinically indicated) may be needed if there are complications or abnormal findings on surveillance imaging
 - After 5 post-operative years without complications, continuing follow-up every 5 years should be considered
- Follow-up after thoracic aorta open repair at the following intervals if there is a reason for CTA rather than CT:
 - At one year post-repair
 - Every 5 years thereafter
 - If abnormal findings are seen on any prior surveillance imaging study, imaging is then done annually

Non-Aortic Vascular Disease

- Suspected or known Superior Vena Cava (SVC) syndrome ⁽¹⁰⁾
 - SVC syndrome is a clinical diagnosis and may be suspected when there are signs of venous congestion in the upper body (such as shortness of breath, distended neck veins and facial/upper extremity edema)
- Subclavian steal syndrome after indeterminate or abnormal ultrasound ⁽⁸⁾
- Suspected or known thoracic outlet syndrome ^(11,12)
- Suspected pulmonary hypertension with suggestive prior testing (Such as echocardiogram or CT chest) ^(7,8)
- For patients with fibromuscular dysplasia (FMD) ^(13,14):
 - Baseline vascular study indicated from brain to pelvis
- Takayasu's Arteritis ⁽¹⁵⁾:
 - At initial diagnosis
 - Every 6 months for the first 2 years while on therapy
 - Annually after the first 2 years

- Non-Central Horner's Syndrome (Secondary/preganglionic or tertiary/post-ganglionic) to evaluate for a vascular source (Such as dissection, aneurysm, arteritis) ^(16,17)
 - **NOTE:** CTA/MRA of the brain and neck may also be indicated

Congenital Vascular Disease of the Chest ^(18–20)

- Suspected thoracic malformation with suggestive prior imaging (Such as chest X-ray, echocardiogram, gastrointestinal study, chest CT)
- Congenital heart disease with pulmonary hypertension and/or extra-cardiac vascular anomalies (Such as Tetralogy of Fallot, transposition of the great vessels, truncus arteriosus)
- Suspected coarctation of the aorta with suggestive prior testing (Such as disparity in the pulsations and blood pressures in the legs versus the arms)
- Pulmonary sequestration
 - **NOTE:** Chest MRA preferred for pediatrics or for repeat imaging

Evaluation of Tumor

- When needed for clarification of vascular involvement from tumor

PREOPERATIVE OR POSTOPERATIVE ASSESSMENT

When not otherwise specified in the guideline:

Preoperative Evaluation:

- Prior to the following procedures:
 - Ablation for atrial fibrillation
 - Endovascular aneurysm repair (EVAR) ⁽⁸⁾
 - Transcatheter Aortic Valve Replacement (TAVR) ⁽²¹⁾
 - Solid organ transplantation
- Imaging of the area requested is needed to develop a surgical plan

Postoperative Evaluation:

- Evaluation of post-operative complications (Such as pseudoaneurysms) following interventional vascular procedures (Such surgical bypass grafts, vascular stents, and IVC filters) ⁽²²⁾
- Known or suspected complications
- A clinical reason is provided how imaging may change management

NOTE: This section applies only within the first few months following surgery

FURTHER EVALUATION OF INDETERMINATE FINDINGS

Unless follow up is otherwise specified within the guideline

- For initial evaluation of an inconclusive finding on a prior imaging report that requires further clarification
- One follow-up exam of a prior indeterminate MR/CT finding to ensure no suspicious interval change has occurred. (No further surveillance unless specified as highly suspicious or change was found on last follow-up exam).

IMAGING IN KNOWN GENETIC CONDITIONS

- Vascular Ehlers-Danlos syndrome (vEDS) ^(23,24):
 - Every 18 months (including at diagnosis) **OR**
 - As clinically indicated to follow known vascular abnormalities
- Marfan Syndrome ⁽²⁵⁾:
 - Every 3 years (including at diagnosis)
 - More frequently (annually) if **EITHER**: history of dissection, dilation of aorta beyond aortic root **OR** aortic root/ascending aorta are not adequately visualized on TTE (i.e. advanced imaging is needed to monitor the thoracic aorta)
- Loeys-Dietz ⁽²⁶⁾:
 - Every two years (including at diagnosis) **OR**
 - More frequent if abnormalities are found
- Williams Syndrome ⁽²⁷⁾:
 - Abdominal vascular exam or imaging findings (such as diminished pulses, bruits or signs of diffuse thoracic aortic stenosis)
- Turner Syndrome ⁽⁷⁾:
 - Screening with no known vascular abnormality at the following intervals:
 - At diagnosis
 - Every 5 years until age 18
 - Every 10 years in adults
 - Prior to pregnancy/pregnancy planning
 - Annually if any one of the following are present: coarctation of the aorta, aortic dilation, bicuspid aortic valve, hypertension
- For other syndromes and rare diseases not otherwise addressed in the guideline, coverage is based on a case-by-case basis

Combination Studies for Known Genetic Conditions

NOTE: When medical necessity is met for an individual study **AND** conscious sedation is required (such as for young pediatric patients or patients with significant developmental delay), the entire combination is indicated)

Abdominal Aorta CT Angiography with Lower Extremity Runoff and Chest CTA

- Williams Syndrome: Abnormal vascular exam or imaging findings (such as diminished pulses, bruits or signs of diffuse thoracic aortic stenosis) ⁽²⁷⁾

Brain/Neck/Chest/Abdomen/Pelvis CTA

- Vascular Ehlers-Danlos syndrome (vEDS): Every 18 months (including at diagnosis) **OR** as clinically indicated to follow known vascular abnormalities ^(23,24)
- Loeys-Dietz: Every two years (including at diagnosis) **OR** more frequently if abnormalities are found ⁽²⁶⁾

Chest/Abdomen/Pelvis CTA

- Marfan syndrome ⁽²⁵⁾:
 - Every 3 years (including at initial diagnosis)
 - More frequently (annually) if **EITHER**: history of dissection, dilation of aorta beyond aortic root **OR** aortic root/ascending aorta are not adequately visualized on TTE (i.e. advanced imaging is needed to monitor the thoracic aorta)
- Williams Syndrome ⁽²⁷⁾
 - Abnormal vascular exam or imaging findings (such as diminished pulses, bruits or signs of diffuse thoracic aortic stenosis)

OTHER COMBINATION STUDIES WITH CHEST CTA

NOTE: When medical necessity is met for an individual study **AND** conscious sedation is required (such as for young pediatric patients or patients with significant developmental delay), the entire combination is indicated)

Abdominal Aorta CT Angiography with Lower Extremity Runoff and Chest CTA

- To evaluate for an embolic source of lower extremity thromboembolic vascular disease.
 - **NOTE:** Echocardiogram is also indicated as the heart is the most commonly reported source of lower extremity emboli

Brain/Neck/Chest/Abdomen/Pelvis CTA

- For patients with fibromuscular dysplasia (FMD), a one-time vascular study from brain to pelvis is indicated ^(13,14)
- For assessment in patients with spontaneous coronary artery dissection (SCAD), (SCAD is a common initial diagnostic event for underlying fibromuscular dysplasia (FMD)). ⁽²⁸⁾
 - **NOTE:** Body vascular imaging for SCAD can be performed at the time of coronary angiography
- Takayasu's Arteritis ⁽¹⁵⁾:
 - At initial diagnosis
 - Every 6 months for the first 2 years while on therapy
 - Annually after the first 2 years

Chest/Abdomen CTA

- Evaluation of extensive vascular disease involving the chest and abdominal cavities when pelvic imaging is not needed
- Significant post-traumatic or post-procedural vascular complications when pelvic imaging is not needed

Chest/Abdomen/Pelvis CTA

- Evaluation prior to endovascular aneurysm repair (EVAR) when thoracic involvement is present
- Evaluation prior to Transcatheter Aortic Valve Replacement (TAVR) ⁽²¹⁾
- Acute aortic dissection ⁽²⁹⁾
- Significant post-traumatic or post-procedural vascular complications reasonably expected to involve the chest, abdomen and pelvis

Chest CTA and Chest CT (or MRI)

- When needed for clarification of vascular involvement from tumor

Brain/Neck/Chest CTA

- Non-Central Horner's Syndrome (Secondary/preganglionic or tertiary/post-ganglionic) to evaluate for a vascular source (Such as dissection, aneurysm, arteritis) ^(16,17)

CODING AND STANDARDS

Codes

71275

Applicable Lines of Business

<input checked="" type="checkbox"/>	CHIP (Children's Health Insurance Program)
<input checked="" type="checkbox"/>	Commercial
<input checked="" type="checkbox"/>	Exchange/Marketplace
<input checked="" type="checkbox"/>	Medicaid
<input checked="" type="checkbox"/>	Medicare Advantage

BACKGROUND

Clinical Assessment of Pulmonary Embolism

Original Wells Score for Prediction of Pulmonary Embolism ⁽²⁾

Clinical Criteria	Score
Clinical Symptoms of DVT (deep vein thrombosis)	3
Other diagnosis (less likely than pulmonary embolism)	3
Heart rate > 100 bpm	1.5
Surgery in previous 4 weeks or Immobilization (≥ 3 days)	1.5
Previous DVT/PE (pulmonary embolism)	1.5
Hemoptysis	1
Malignancy	1
Clinical Probability	
Low probability of pulmonary embolus	0 – 1
Intermediate probability	2 – 6

High probability	≥ 7
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Original and Simplified Geneva Score for Prediction of Pulmonary Embolism ^(2,5)

Clinical Criteria	Original Version	Simplified Version
Age ≥ 65 years	1	1
Previous DVT (deep vein thrombosis) or PE (pulmonary embolism)	3	1
Recent surgery or fracture (within 1 month)	2	1
Active malignant condition	2	1
Unilateral lower limb pain	3	1
Hemoptysis	2	1
Heart rate 75-94 bpm	3	1
Heart rate ≥ 95 bpm	5	2
Unilateral edema of lower limb and pain on deep palpation	4	1
Clinical Probability		
Low probability of Pulmonary Embolus (PE)	0 – 3	0 – 1
Intermediate probability	4 – 10	2 – 4
High probability	≥ 11	≥ 5

Contraindications and Preferred Studies

- Contraindications and reasons why a CT/CTA cannot be performed may include: impaired renal function, significant allergy to IV contrast, pregnancy (depending on trimester).
- Contraindications and reasons why an MRI/MRA cannot be performed may include: impaired renal function, claustrophobia, non-MRI compatible devices (such as non-compatible defibrillator or pacemaker), metallic fragments in a high-risk location, patient exceeds weight limit/dimensions of MRI machine.

SUMMARY OF EVIDENCE

2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease ⁽⁷⁾

Study Design: This document is a clinical practice guideline developed by the American Heart Association (AHA) and the American College of Cardiology (ACC) Joint Committee on Clinical Practice Guidelines. It includes recommendations based on a comprehensive literature review conducted from January 2021 to April 2021, with additional relevant studies considered through June 2022.

Target Population: The guidelines are intended for clinicians diagnosing and managing patients with aortic disease, including asymptomatic, stable symptomatic, and acute aortic syndromes.

Key Factors:

- **Diagnosis and Management:** Recommendations cover genetic evaluation, family screening, medical therapy, endovascular and surgical treatment, and long-term surveillance.
- **Imaging Techniques:** Various imaging techniques such as computed tomography, magnetic resonance imaging, echocardiography, and intravascular ultrasound are discussed.
- **Multidisciplinary Teams:** Emphasis on the importance of multidisciplinary aortic teams and shared decision-making.
- **Pregnancy:** Special considerations for managing aortic disease in pregnant patients.

2019 ESC Guidelines for the diagnosis and management of acute pulmonary embolism developed in collaboration with the European Respiratory Society (ERS) ⁽²⁾

Study Design: This document is a guideline developed by the European Society of Cardiology (ESC) in collaboration with the European Respiratory Society (ERS). It includes recommendations based on a thorough review of the literature and expert consensus.

Target Population: The guidelines are aimed at clinicians diagnosing and managing patients with acute pulmonary embolism (PE).

Key Factors:

- **Diagnosis:** Recommendations for various diagnostic tests, including D-dimer testing, computed tomographic pulmonary angiography (CTPA), and echocardiography.
- **Risk Assessment:** Detailed risk assessment strategies, including clinical parameters, imaging, and laboratory biomarkers.
- **Treatment:** Guidelines for acute-phase treatment, including anticoagulation, thrombolysis, and mechanical circulatory support.
- **Pregnancy:** Specific recommendations for managing PE in pregnant patients.
- **Long-term Management:** Strategies for chronic treatment and prevention of recurrence.

2024 ESC Guidelines for the management of peripheral arterial and aortic diseases ⁽⁸⁾

Study Design: This document is a guideline developed by the European Society of Cardiology (ESC) for the management of peripheral arterial and aortic diseases. It includes recommendations based on a comprehensive review of the literature and expert consensus.

Target Population: The guidelines are intended for clinicians managing patients with peripheral arterial and aortic diseases.

Key Factors:

- **Diagnosis:** Recommendations for various diagnostic tests, including duplex ultrasound, computed tomography angiography (CTA), and magnetic resonance angiography (MRA).
- **Screening:** Guidelines for screening for carotid, peripheral arterial, and aortic diseases.
- **Medical Treatment:** Recommendations for lifestyle changes, exercise, and pharmacological therapy.
- **Interventional Treatment:** Guidelines for revascularization and surgical interventions.
- **Follow-up:** Recommendations for follow-up after treatment of aortic aneurysms and acute aortic syndromes.
- **Genetic Diseases:** Specific recommendations for managing genetic and congenital diseases of the aorta.

ANALYSIS OF EVIDENCE

Shared Findings ^(2,7,8):

- All three guidelines emphasize the importance of accurate diagnosis and comprehensive management of cardiovascular diseases, including the use of advanced imaging techniques and multidisciplinary teams.
- They all provide specific recommendations for managing cardiovascular conditions in pregnant patients, highlighting the need for special considerations in this population.
- The guidelines stress the importance of long-term management and follow-up to prevent recurrence and manage chronic conditions.

Conclusion ^(2,7,8):

In summary, while all three guidelines share common themes of diagnosis, management, and long-term care, they each have a unique focus and provide specific recommendations tailored to their respective areas of cardiovascular disease.

POLICY HISTORY

Date	Summary
June 2025	<ul style="list-style-type: none"> ● Guideline number changed from 022-1 to 2020 ● Guideline name changed from Chest CTA to Chest Computed Tomography Angiography (CTA) ● Added in general information statement regarding guideline criteria development by reputable sources, standard of care, and best practices ● Updated Pulmonary Embolism and Thoracic Aortic Disease sections ● Broke down Suspected Thoracic Aortic Disease section into screening criteria and abnormal imaging result or signs/symptoms ● Added Takayasu's Arteritis and Non-Central Horner's Syndrome indications in the Vascular Disease and Combination Studies sections ● Moved fibromuscular dysplasia (FMD) indication from Imaging in Known Genetic Condition section to Non-Aortic Vascular Disease section ● Standardized Preoperative and Postoperative Assessment section ● Separated combination studies section for Genetic and Nongenetic conditions and updated accordingly ● Edited text for clarity and consistency ● Updated Background section and references ● Added Summary and Analysis of Evidence
May 2024	<ul style="list-style-type: none"> ● Updated references ● Added Genetics and Rare Diseases, Evaluation of Tumor, Contraindications and Preferred Studies sections ● Reorganized section Thoracic Aortic Disease



LEGAL AND COMPLIANCE

Guideline Approval

Committee

Reviewed / Approved by Evolent Specialty Services Clinical Guideline Review Committee

Disclaimer

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Evolent Clinical Guidelines are comprehensive and inclusive of various procedural applications for each service type. Our guidelines may be used to supplement Medicare criteria when such criteria is not fully established. When Medicare criteria is determined to not be fully established, we only reference the relevant portion of the corresponding Evolent Clinical Guideline that is applicable to the specific service or item requested in order to determine medical necessity.

REFERENCES

1. Kirsch J, Wu CC, Bolen MA, et al. ACR Appropriateness Criteria® Suspected Pulmonary Embolism: 2022 Update. *Journal of the American College of Radiology*. 2022;19(11):S488-S501. doi:10.1016/j.jacr.2022.09.014
2. Konstantinides S V, Meyer G, Becattini C, et al. 2019 ESC Guidelines for the diagnosis and management of acute pulmonary embolism developed in collaboration with the European Respiratory Society (ERS). *Eur Heart J*. 2020;41(4):543-603. doi:10.1093/eurheartj/ehz405
3. Moore AJE, Wachsmann J, Chamrathy MR, Panjikaran L, Tanabe Y, Rajiah P. Imaging of acute pulmonary embolism: an update. *Cardiovasc Diagn Ther*. 2018;8(3):225-243. doi:10.21037/cdt.2017.12.01
4. Rivera-Lebron B, McDaniel M, Ahrar K, et al. Diagnosis, Treatment and Follow Up of Acute Pulmonary Embolism: Consensus Practice from the PERT Consortium. *Clinical and Applied Thrombosis/Hemostasis*. 2019;25:1076029619853037. doi:10.1177/1076029619853037
5. Tak T, Karturi S, Sharma U, Eckstein L, Poterucha JT, Sandoval Y. Acute Pulmonary Embolism: Contemporary Approach to Diagnosis, Risk-Stratification, and Management. *International Journal of Angiology*. 2019;28(02):100-111. doi:10.1055/s-0039-1692636
6. Borger MA, Fedak PWM, Stephens EH, et al. The American Association for Thoracic Surgery consensus guidelines on bicuspid aortic valve–related aortopathy: Full online-only version. *J Thorac Cardiovasc Surg*. 2018;156(2):e41-e74. doi:10.1016/j.jtcvs.2018.02.115
7. Isselbacher EM, Preventza O, Hamilton Black J, et al. 2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease: A Report of the American Heart Association/American College of Cardiology Joint Committee on Clinical Practice Guidelines. *Circulation*. 2022;146(24):e334-e482. doi:10.1161/CIR.0000000000001106
8. Mazzolai L, Teixido-Tura G, Lanzi S, et al. 2024 ESC Guidelines for the management of peripheral arterial and aortic diseases. *Eur Heart J*. 2024;45(36):3538-3700. doi:10.1093/eurheartj/ehae179
9. Mariscalco G, Debiec R, Elefteriades JA, Samani NJ, Murphy GJ. Systematic Review of Studies That Have Evaluated Screening Tests in Relatives of Patients Affected by Nonsyndromic Thoracic Aortic Disease. *J Am Heart Assoc*. 2018;7(15):e009302. doi:10.1161/JAHA.118.009302
10. Azizi AH, Shafi I, Shah N, et al. Superior Vena Cava Syndrome. *JACC Cardiovasc Interv*. 2020;13(24):2896-2910. doi:10.1016/j.jcin.2020.08.038
11. Jones MR, Prabhakar A, Viswanath O, et al. Thoracic Outlet Syndrome: A Comprehensive Review of Pathophysiology, Diagnosis, and Treatment. *Pain Ther*. 2019;8(1):5-18. doi:10.1007/s40122-019-0124-2
12. Zurkiya O, Ganguli S, Kalva SP, et al. ACR Appropriateness Criteria® Thoracic Outlet Syndrome. *Journal of the American College of Radiology*. 2020;17(5):S323-S334. doi:10.1016/j.jacr.2020.01.029

13. Gornik HL, Persu A, Adlam D, et al. First International Consensus on the diagnosis and management of fibromuscular dysplasia. *Vascular Medicine*. 2019;24(2):164-189. doi:10.1177/1358863X18821816
14. Kesav P, Manesh Raj D, John S. Cerebrovascular Fibromuscular Dysplasia – A Practical Review. *Vasc Health Risk Manag*. 2023;Volume 19:543-556. doi:10.2147/VHRM.S388257
15. Joseph G, Goel R, Thomson VS, Joseph E, Danda D. Takayasu Arteritis. *J Am Coll Cardiol*. 2023;81(2):172-186. doi:10.1016/j.jacc.2022.09.051
16. Davagnanam I, Fraser CL, Miszkiel K, Daniel CS, Plant GT. Adult Horner's syndrome: a combined clinical, pharmacological, and imaging algorithm. *Eye*. 2013;27(3):291-298. doi:10.1038/eye.2012.281
17. Maamouri R, Ferchichi M, Houmane Y, Gharbi Z, Cheour M. Neuro-Ophthalmological Manifestations of Horner's Syndrome: Current Perspectives. *Eye Brain*. 2023;Volume 15:91-100. doi:10.2147/EB.S389630
18. Baumgartner H, De Backer J, Babu-Narayan S V, et al. 2020 ESC Guidelines for the management of adult congenital heart disease. *Eur Heart J*. 2021;42(6):563-645. doi:10.1093/eurheartj/ehaa554
19. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease. *J Am Coll Cardiol*. 2019;73(12):e81-e192. doi:10.1016/j.jacc.2018.08.1029
20. Krishnamurthy R, Suman G, Chan SS, et al. ACR Appropriateness Criteria® Congenital or Acquired Heart Disease. *Journal of the American College of Radiology*. 2023;20(11):S351-S381. doi:10.1016/j.jacr.2023.08.018
21. Hedgire SS, Saboo SS, Galizia MS, et al. ACR Appropriateness Criteria® Preprocedural Planning for Transcatheter Aortic Valve Replacement: 2023 Update. *Journal of the American College of Radiology*. 2023;20(11):S501-S512. doi:10.1016/j.jacr.2023.08.009
22. Zierler RE, Jordan WD, Lal BK, et al. The Society for Vascular Surgery practice guidelines on follow-up after vascular surgery arterial procedures. *J Vasc Surg*. 2018;68(1):256-284. doi:10.1016/j.jvs.2018.04.018
23. Bowen JM, Hernandez M, Johnson DS, et al. Diagnosis and management of vascular Ehlers-Danlos syndrome: Experience of the UK national diagnostic service, Sheffield. *European Journal of Human Genetics*. 2023;31(7):749-760. doi:10.1038/s41431-023-01343-7
24. Byers PH. Vascular Ehlers-Danlos Syndrome. *GeneReviews*®. Published online April 10, 2025. <https://www.ncbi.nlm.nih.gov/books/NBK1494/>
25. Dietz H. FBN1-Related Marfan Syndrome. *GeneReviews*®. Published online February 17, 2022. <https://www.ncbi.nlm.nih.gov/books/NBK1335/>
26. Loeys BL, Dietz HC. Loeys-Dietz Syndrome. *GeneReviews*®. Published online September 12, 2024. <https://www.ncbi.nlm.nih.gov/books/NBK1133/>
27. Morris CA. Williams Syndrome. *GeneReviews*®. Published online April 13, 2023. <https://www.ncbi.nlm.nih.gov/books/NBK1249/>

28. Teruzzi G, Santagostino Baldi G, Gili S, Guarnieri G, Montorsi P, Trabattoni D. Spontaneous Coronary Artery Dissections: A Systematic Review. *J Clin Med*. 2021;10(24):5925. doi:10.3390/jcm10245925
29. Kicska GA, Hurwitz Koweek LM, Ghoshhajra BB, et al. ACR Appropriateness Criteria® Suspected Acute Aortic Syndrome. *Journal of the American College of Radiology*. 2021;18(11):S474-S481. doi:10.1016/j.jacr.2021.09.004