

Evolent Clinical Guideline ~~061-2~~2063 for Upper Extremity Computed Tomography Angiography (CTA) ~~CT Angiography A/CTV~~

Guideline or Policy Number: Evolent_CG_061-22063		Applicable Codes
<i>"Evolent" refers to Evolent Health LLC and Evolent Specialty Services, Inc.</i> © 2008 - 202 65 <u>Evolent</u> . All rights Reserved.		
Original Date: July 2008	Last Revised Date: April 2024 <u>June 2025</u>	Implementation Date: January 2025 <u>2026</u>

TABLE OF CONTENTS

STATEMENT	3
GENERAL INFORMATION	3
PURPOSE	3
SPECIAL NOTES	3
INDICATIONS FOR UPPER EXTREMITY COMPUTED TOMOGRAPHY ANGIOGRAPHY (CTA)	3
UPPER EXTREMITY PERIPHERAL VASCULAR DISEASE	<u>43</u>
DEEP VENOUS THROMBOSIS (DVT)	4
ARTERIAL THROMBOEMBOLISM	<u>54</u>
CLINICAL SUSPICION OF ANEURYSM	<u>54</u>
CLINICAL SUSPICION OF VASCULITIS	<u>54</u>
HEMODIALYSIS GRAFT DYSFUNCTION	5
VASCULAR MALFORMATION (VM)	<u>65</u>
TRAUMATIC INJURIES	<u>65</u>
EVALUATION OF TUMOR	<u>65</u>
PREOPERATIVE OR POSTOPERATIVE ASSESSMENT	<u>65</u>
FURTHER EVALUATION OF INDETERMINATE FINDINGS	<u>75</u>
IMAGING IN KNOWN GENETIC CONDITIONS	<u>76</u>
CODING AND STANDARDS	<u>86</u>
CODES	<u>86</u>
APPLICABLE LINES OF BUSINESS	<u>86</u>
BACKGROUND	<u>87</u>
CONTRAINDICATIONS AND PREFERRED STUDIES	<u>87</u>
SUMMARY OF EVIDENCE	<u>87</u>
ANALYSIS OF EVIDENCE	<u>108</u>
POLICY HISTORY	<u>109</u>

LEGAL AND COMPLIANCE	<u>1110</u>
GUIDELINE APPROVAL	<u>1110</u>
<i>Committee</i>	<u>1110</u>
DISCLAIMER	<u>1110</u>
REFERENCES.....	<u>1311</u>

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

CTA/CTV

Computed tomography angiography (CTA) generates images of the arteries and veins that can be evaluated for evidence of stenosis, occlusion, or aneurysms. It is used to evaluate the arteries of the abdominal aorta and the renal arteries. blood vessels of the upper extremities. CTA uses ionizing radiation and requires the administration of iodinated contrast agent, which is a potential hazard in patients with impaired renal function. Upper Extremity CTA is not used as a screening tool, e.g., evaluation of asymptomatic patients, without a previous diagnosis.

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

Special Notes

Imaging Request

When a separate CTA and CT exam is requested, documentation requires a medical reason that clearly indicates why additional CT imaging of the upper extremity is needed.

—Authorization for CTMR Angiography covers both arterial and venous imaging. The term angiography refers to both arteriography and venography.

INDICATIONS FOR UPPER EXTREMITY COMPUTED TOMOGRAPHY ANGIOGRAPHY (CTA)

Hand Upper Extremity Ischemia Peripheral Vascular Disease

(1,2)

- For known or suspected acute upper extremity ischemia with any **ONE** of the following ⁽³⁾: Acute symptoms (including):
 - Ischemic ulceration without segmental temperature change
 - Painful ischemic ulceration with painful ischemia
 - Acute sustained loss of perfusion with or without acral ulceration
 - Imminent loss of digit(s)
- NOTE: -Does NOT require prior ultrasound
- NOTE: Does not require prior arterial Doppler
- For known or suspected upper extremity vascular disease (including Raynaud's) ⁽⁴⁾ with ALL of the following: clinical symptoms,
 - following Prior abnormal or indeterminate ultrasound arterial Doppler, when
 - CTA results will potentially change management ⁽³⁾
- Includes Raynaud's (can be associated with scleroderma), Buerger disease, and other vasculopathies ⁽³⁾
- Clinical concern for vascular cause of ulcers with abnormal or indeterminate ultrasound ⁽⁴⁾ For upper extremity ulcers on exam from known/suspected peripheral vascular disease after prior abnormal or indeterminate ultrasound ^(2,5)
- For known predisposing conditions (sSuch as Buerger disease, cystic adventitial disease, arterial endofibrosis, fibromuscular dysplasia, segmental arterial mediolysis and/or genetic conditions such as Marfan syndrome, Loeys-Dietz syndrome, or vascular Ehler-Danlos Syndrome) and any **ONE** of the following ⁽⁶⁾:
 - Prior imaging suggestive of non-atherosclerotic peripheral vascular disease of the upper lower extremity
 - Signs or symptoms of upper extremity vascular disease (sSuch as claudication, weak pulses)
 -
- AfterAfter stenting or surgery with signs of recurrence or indeterminate ultrasound ⁽⁶⁾ AfterAfter prior stenting or surgery (arterial and/or venous) with any **ONE** of the following ⁽⁷⁾:
 - Rrecurrent symptoms
 - Ssigns of recurrent disease on examination
 - Aabnormal / indeterminate prior non-invasive testing or imaging

Deep Venous Thrombosis (DVT) or Embolism ^(8,9)

- For known/suspected upper extremity DVT with **ALL** the following: Clinical suspicion of lower extremity DVT (when

- Prior ultrasound is abnormal or inconclusive and

-

- aAdvanced imaging positive study results would will potentially change management

- After abnormal ultrasound of arm veins if it will change management

After negative or indeterminate ultrasound to rule out other causes

- For evaluation of central veins

Arterial Thromboembolism

- Clinical findings (ssuch as pulselessness, acute limb ischemia) and / or prior imaging suggestive of upper extremity arterial thromboembolism ^(10,11)
- Clinical suspicion of upper arterial emboli ^(8,9)

NOTE: Echocardiogram and advanced vascular imaging of the chest, abdomen, and/or pelvis may also be indicated to identify the source of the emboli. Clinical Suspicion of Vascular Disease

With abnormal or indeterminate ultrasound ^(8,9), for suspicion of:

- Tumor invasion ^(10,11)
- Trauma ⁽¹²⁾
- Vasculitis ^(1,13)
- Aneurysm ⁽¹³⁾
- Stenosis/occlusions ⁽¹⁴⁾

Clinical Suspicion of Aneurysm

- With prior abnormal or indeterminate ultrasound or other imaging ⁽¹²⁾

Clinical Suspicion of Vasculitis

- With prior abnormal or indeterminate ultrasound or other imaging ⁽¹²⁾

Hemodialysis Graft Dysfunction

- If Doppler ultrasound was not adequate for treatment decisions If Doppler prior ultrasound was completed and not adequate sufficient for required treatment decisions

(13)

Vascular Malformation (VM) ^(14,15)

- For known / suspected upper extremity VM with ALL the following:
 - Prior abnormal or indeterminate ultrasound
 - MRA/MRV is contraindicated or not possible
 - Advanced imaging study results will potentially change management
 -

~~NOTE: (MRA preferred however CTA useful in delineating some high flow lesions such as an arteriovenous malformation)~~

~~After initial evaluation with ultrasound~~

~~Preoperative planning~~

- A concurrent upper extremity CT ~~of the upper extremity~~ is also ~~approvable~~ indicated for initial evaluation ~~/and/or~~ preoperative planning if MRI is contraindicated or cannot be performed, or per surgeon preference.

Traumatic Injuries

- ~~Clinical findings suggestive of arterial injury~~ Clinical findings (sSuch as bruit, hemorrhage, hematoma, pulselessness) and/or prior imaging suggestive of upper extremity vascular arterial injury ⁽¹⁶⁾

Evaluation of Tumor

- When needed for clarification of vascular invasion ~~involvement~~ from tumor after prior imaging (may be approved in combination with CT or MRI of tumor)

~~PRE-OPERATIVE~~ OR POSTOPERATIVE ASSESSMENT

~~When not otherwise specified in the guideline~~ /Procedural Evaluations

Preoperative Evaluation:

- Imaging of the area requested is needed to develop by a surgical plan ~~Pre-operative evaluation for a planned surgery or procedure~~ ⁽¹⁶⁾

~~Post-Operative/Procedural Evaluations~~

Postoperative Evaluation:

- Known or suspected complications

- A clinical reason is provided how imaging may change management follow-up study may be needed to help evaluate a patient's progress after treatment, procedure, intervention, or surgery. Documentation requires a medical reason that clearly indicates why additional imaging is needed for the type and area(s) requested.

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

FURTHER EVALUATION OF INDETERMINATE FINDINGS ~~ON PRIOR IMAGING~~

Unless follow-up is otherwise specified within the guideline

- For initial evaluation of an inconclusive finding on a prior imaging report (i.e., x-ray, ultrasound or CT) 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 SYNDROMES AND RARE DISEASES

- ~~Known vascular EDS (vEDS) with acute extremity pain and concern for dissection/rupture (18,19)~~
- Vascular Ehlers-Danlos Syndrome (vEDS) ~~surveillance imaging:~~ With inconclusive ultrasound or ultrasound suggestive of vascular pathology OR acute extremity pain and concern for dissection/rupture ^(17,18)
- ~~Known Williams Syndrome: when there is concern for vascular disease based on~~ Abnormal vascular exam or imaging findings (such as diminished pulses, bruits or signs of diffuse thoracic aortic stenosis) ⁽¹⁹⁾
- For other syndromes and rare diseases not otherwise addressed in the guideline, coverage is based on a case-by-case basis using societal guidance
- For known predisposing conditions (such as Buerger disease, cystic adventitial disease, arterial endofibrosis, fibromuscular dysplasia, segmental arterial mediolysis and/or genetic conditions such as Marfan syndrome, Loeys-Dietz syndrome, or vascular Ehler-Danlos Syndrome) and any ONE of the following ⁽⁶⁾:
 - Prior imaging suggestive of non-atherosclerotic peripheral vascular disease of the upper extremity
 - Signs or symptoms of upper extremity vascular disease (such as claudication, weak pulses)

CODING AND STANDARDS

Coding

CPT Codes

73206

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

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

CT Angiography of the Upper Extremity Arterial System: Part 2—Clinical Applications Beyond Trauma Patients ⁽¹¹⁾

Study Design: This study focuses on the clinical applications of CT angiography for evaluating the upper extremity arterial system beyond trauma patients. It is a review article published in the American Journal of Roentgenology (AJR) in October 2013.

Target Population: The study primarily targets patients with non-traumatic vascular pathologic abnormalities of the upper extremity arterial system. This includes patients with atherosclerotic disease, thromboembolic disease, vasculitis, and other vascular conditions.

Key Factors:

- Atherosclerotic Disease: The study discusses the disproportionate impact of atherosclerotic disease on the upper extremities compared to the lower extremities. It highlights risk factors such as hypertension, dyslipidemia, diabetes mellitus, age, and smoking history.
- Thromboembolic Disease: The study notes the decreased incidence of arterial embolism to the extremities due to increased use of anticoagulation. It emphasizes the higher mortality rate associated with peripheral arterial emboli compared to arterial thrombosis.
- Vasculitis: The study describes the imaging characteristics of vasculitis affecting the upper extremity vessels, including Takayasu arteritis, giant cell arteritis, and thromboangiitis obliterans.
- Other Conditions: The study also covers conditions such as thoracic outlet compression, hypothenar hammer syndrome, and radiation arteritis.

ACR Appropriateness Criteria Nonatherosclerotic Peripheral Arterial Disease ⁽⁶⁾

Study Design: This document outlines the ACR Appropriateness Criteria for nonatherosclerotic peripheral arterial disease. It is a guideline developed by the American College of Radiology (ACR) and published in the Journal of the American College of Radiology in 2019.

Target Population: The guidelines are intended for patients with nonatherosclerotic diseases affecting the peripheral arteries. This includes conditions such as popliteal entrapment syndrome, external iliac artery endofibrosis, inflammatory vasculitides, and vascular trauma.

Key Factors:

- Diagnostic Imaging: The guidelines recommend various imaging modalities for initial evaluation, including MRA, CTA, and US duplex Doppler.
- Clinical Presentation: The document discusses the clinical presentation of different nonatherosclerotic peripheral arterial diseases and provides recommendations for appropriate imaging studies based on the suspected condition.
- Treatment Recommendations: The guidelines offer evidence-based recommendations for imaging and treatment procedures, emphasizing the importance of accurate vascular imaging and nonvascular findings in guiding surgical management.

2024 ESC Guidelines for the management of peripheral arterial and aortic diseases ⁽²⁾

Study Design: This document presents the 2024 ESC Guidelines for the management of peripheral arterial and aortic diseases. It is developed by the task force of the European Society of Cardiology (ESC) and endorsed by several European associations.

Target Population: The guidelines target patients with peripheral arterial and aortic diseases, including atherosclerotic and nonatherosclerotic conditions. It covers a wide range of topics, from epidemiology and risk factors to diagnostic work-up and treatment strategies.

Key Factors:

- Epidemiology and Risk Factors: The guidelines provide detailed information on the prevalence and risk factors associated with peripheral arterial and aortic diseases.
- Diagnostic Work-Up: The document outlines the recommended diagnostic tests and imaging modalities for evaluating peripheral arteries and the aorta.
- Treatment Strategies: The guidelines offer comprehensive recommendations for medical and interventional treatment of various peripheral arterial and aortic conditions, including chronic limb-threatening ischemia, carotid artery stenosis, and aortic aneurysms.
- Follow-Up: The guidelines emphasize the importance of regular follow-up and monitoring for patients with peripheral arterial and aortic diseases.

ANALYSIS OF EVIDENCE

Shared Findings ^{(2,6,11):}

- **Accuracy and Utility of Angiography:** All three articles emphasize the accuracy and utility of angiography in diagnosing vascular pathologies. Bozlar et al 2013 highlights the use of CT angiography (CTA) for evaluating non-traumatic vascular abnormalities in the upper extremities. Francois et al 2019 discusses the appropriateness of various imaging modalities, including CTA and MRA, for diagnosing nonatherosclerotic peripheral arterial diseases. Mazzolai et al 2024 also underscores the importance of CTA and MRA in evaluating peripheral arterial and aortic diseases.
- **Non-invasive Nature:** The non-invasive nature of CTA and MRA is a common theme. Bozlar et al 2013 mentions the evolution of CTA into a highly accurate non-invasive diagnostic tool. Francois et al 2019 highlights the non-invasive nature of MRA and CTA in their appropriateness criteria. Mazzolai et al 2024 also discusses the non-invasive benefits of these imaging techniques.
- **Clinical Applications:** All three articles discuss the clinical applications of extremity angiography imaging. Bozlar et al 2013 focuses on the use of CTA in non-traumatic vascular pathologies. Francois et al 2019 provides guidelines for the initial evaluation of various nonatherosclerotic peripheral arterial diseases. Mazzolai et al 2024 offers comprehensive guidelines for managing peripheral arterial and aortic diseases, including the use of imaging.

Conclusion ^{(2,6,11):}

In summary, while all three articles highlight the importance and accuracy of extremity angiography imaging, they differ in their focus on specific conditions, imaging modalities, and recommendations. Bozlar et al 2013 provides a detailed analysis of CTA's clinical applications in the upper extremities, Francois et al 2019 offers appropriateness criteria for various imaging modalities, and Mazzolai et al 2024 presents comprehensive guidelines for managing peripheral arterial and aortic diseases.

POLICY HISTORY

Date	Summary
<u>June 2025</u>	<ul style="list-style-type: none"> ● <u>Guideline number changed from 061-2 to 2063</u> ● <u>Guideline name changed from Upper Extremity CTA/CTV to Upper Extremity Computed Tomography Angiography (CTA)</u> ● <u>Added in general information statement regarding guideline criteria development by reputable sources, standard of care, and best practices</u> ● <u>Added non-atherosclerotic PVD to Peripheral Vascular Disease and Genetics Syndromes and Rare Diseases sections</u> ● <u>Broke down clinical suspicion of vascular disease section for clarity</u> ● <u>Standardized preoperative and postoperative assessment and Genetic Syndrome and Rare Diseases sections</u> ● <u>Edited text for clarity and consistency</u> ● <u>Updated references</u> ● <u>Added a Summary of Evidence and Analysis of Evidence</u>
April 2024	<ul style="list-style-type: none"> ● Updated references ● Added Evaluation of Tumor, Genetics Syndromes and Rare Diseases, and Contraindications and Preferred Studies sections.
April 2023	<p>Modified background section</p> <p>Added vascular malformations</p> <p>Added indeterminate prior imaging findings</p> <p>General Information moved to beginning of guideline with added statement on clinical indications not addressed in this guideline</p>

LEGAL AND COMPLIANCE

Guideline Approval

Committee

Reviewed / Approved by Evolent Specialty Services Clinical Guideline Review Committee

Disclaimer



Evolent Clinical Guidelines do not constitute medical advice. Treating health care professionals are solely responsible for diagnosis, treatment, and medical advice. Evolent uses Clinical Guidelines in accordance with its contractual obligations to provide utilization management. Coverage for services varies for individual members according to the terms of their health care coverage or government program. Individual members' health care coverage may not utilize some Evolent Clinical Guidelines. Evolent clinical guidelines contain guidance that requires prior authorization and service limitations. A list of procedure codes, services or drugs may not be all inclusive and does not imply that a service or drug is a covered or non-covered service or drug. Evolent reserves the right to review and update this Clinical Guideline in its sole discretion. Notice of any changes shall be provided as required by applicable provider agreements and laws or regulations. Members should contact their Plan customer service representative for specific coverage information.

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. Hotchkiss R, Marks T. Management of acute and chronic vascular conditions of the hand. *Curr Rev Musculoskelet Med*. 2014;7(1):47-52. doi:10.1007/s12178-014-9202-6
2. Mazzolai L, Teixeira-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
3. Björck M, Earnshaw JJ, Acosta S, et al. Editor's Choice – European Society for Vascular Surgery (ESVS) 2020 Clinical Practice Guidelines on the Management of Acute Limb Ischaemia. *European Journal of Vascular and Endovascular Surgery*. 2020;59(2):173-218. doi:10.1016/j.ejvs.2019.09.006
4. Belch J, Carlizza A, Carpentier PH, et al. ESVS guidelines – the diagnosis and management of Raynaud's phenomenon. *Vasa*. 2017;46(6):413-423. doi:10.1024/0301-1526/a000661
5. Kakkos SK, Gohel M, Baekgaard N, et al. Editor's Choice – European Society for Vascular Surgery (ESVS) 2021 Clinical Practice Guidelines on the Management of Venous Thrombosis. *European Journal of Vascular and Endovascular Surgery*. 2021;61(1):9-82. doi:10.1016/j.ejvs.2020.09.023
6. Francois CJ, Skulborstad EP, Kalva SP, et al. ACR Appropriateness Criteria® Nonatherosclerotic Peripheral Arterial Disease. *Journal of the American College of Radiology*. 2019;16(5):S174-S183. doi:10.1016/j.jacr.2019.02.026
7. 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
8. Heil J, Miesbach W, Vogl T, Bechstein WO, Reinisch A. Deep Vein Thrombosis of the Upper Extremity. *Dtsch Arztebl Int*. 2017;114(14):244-249. doi:10.3238/arztebl.2017.0244
9. Desjardins B, Hanley M, Steigner ML, et al. ACR Appropriateness Criteria® Suspected Upper Extremity Deep Vein Thrombosis. *Journal of the American College of Radiology*. 2020;17(5):S315-S322. doi:10.1016/j.jacr.2020.01.020
10. Bozlar U, Ogur T, Norton PT, Khaja MS, All J, Hagspiel KD. CT Angiography of the Upper Extremity Arterial System: Part 1—Anatomy, Technique, and Use in Trauma Patients. *American Journal of Roentgenology*. 2013;201(4):745-752. doi:10.2214/AJR.13.11207
11. Bozlar U, Ogur T, Khaja MS, All J, Norton PT, Hagspiel KD. CT Angiography of the Upper Extremity Arterial System: Part 2— Clinical Applications Beyond Trauma Patients. *American Journal of Roentgenology*. 2013;201(4):753-763. doi:10.2214/AJR.13.11208
12. Seitz L, Seitz P, Pop R, Lötscher F. Spectrum of Large and Medium Vessel Vasculitis in Adults: Primary Vasculitides, Arthritides, Connective Tissue, and Fibroinflammatory Diseases. *Curr Rheumatol Rep*. 2022;24(11):352-370. doi:10.1007/s11926-022-01086-2
13. Richarz S, Isaak A, Aschwanden M, Partovi S, Staub D. Pre-procedure imaging planning for dialysis access in patients with end-stage renal disease using ultrasound and upper extremity computed tomography angiography: a narrative review. *Cardiovasc Diagn Ther*. 2023;13(1):122-132. doi:10.21037/cdt-21-797

14. Obara P, McCool J, Kalva SP, et al. ACR Appropriateness Criteria® Clinically Suspected Vascular Malformation of the Extremities. *Journal of the American College of Radiology*. 2019;16(11):S340-S347. doi:10.1016/j.jacr.2019.05.013
15. Schmidt VF, Masthoff M, Czihal M, et al. Imaging of peripheral vascular malformations — current concepts and future perspectives. *Mol Cell Pediatr*. 2021;8(1):19. doi:10.1186/s40348-021-00132-w
16. Lateef Wani M, Gani Ahangar A, Ahmad Ganie F, Nabi Wani S, Wani N ud din. Vascular Injuries: Trends in Management. *Trauma Mon*. 2012;17(2):266-269. doi:10.5812/traumamon.6238
17. 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
18. Byers PH. Vascular Ehlers-Danlos Syndrome. *GeneReviews*®. Published online February 21, 2019. <https://www.ncbi.nlm.nih.gov/books/NBK1494/>
19. Morris CA. Williams Syndrome. *GeneReviews*®. Published online April 13, 2023. <https://www.ncbi.nlm.nih.gov/books/NBK1249/>