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A Heart for Life: Why Ongoing Cardiology Care Matters

PRESENTED BY

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Presented by INNERSENSE ORGANIC BEAUTY

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Session Overview

This session focuses on cardiovascular care for individuals with Williams syndrome (WS) across the lifespan.
Attendees will examine how elastin-related arteriopathy contributes to cardiovascular risk profile from infancy through adulthood.
The session explains why lifelong cardiology surveillance remains essential, even after early repair or a history of normal evaluations.
We will review age-specific screening priorities, including blood pressure management, vascular imaging, and coronary assessment.
Special attention is given to the adolescent-to-adult transition, a critical period for sustained cardiology engagement.
Presenters will close with practical strategies for shared responsibility among providers, patients, and caregivers.

Learning Objectives

ATTENDEES WILL BE ABLE TO:

- Describe how cardiovascular risk in Williams syndrome changes from childhood through adulthood
- Explain why lifelong cardiology surveillance is needed, even after early repair or normal studies
- Identify age-specific screening priorities, including blood pressure, vascular imaging, and coronary assessment
- Recognize the adolescent-to-adult transition as a high-risk period for loss to follow-up
- Describe how providers, patients, and caregivers share responsibility for long-term outcomes

PART ONE
SCREENING PRIORITIES

Age-Specific Screening and Monitoring Priorities

Blood pressure, vascular imaging, and coronary assessment across the lifespan

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Age-Specific Screening Schedule

Assessment	Birth–12 months	1–5 years	6+ years / Adulthood
Cardiology evaluation	Every 3 months	Annually	Every 2–3 years
Four-extremity blood pressure	Every 3 months	Annually	Annually
ECG (including QTc)	Annually	Annually	Annually
Echocardiogram	Every 3 months	Annually	Biennially
Ambulatory ECG	As indicated	Annually	Biennially
CT / CMR angiography	If indicated by lesion severity	If indicated by lesion severity	At least once in adulthood

PART TWO
NATURAL HISTORY

The Natural History of Cardiovascular Disease in WS

How elastin arteriopathy reshapes cardiovascular risk from infancy through adulthood

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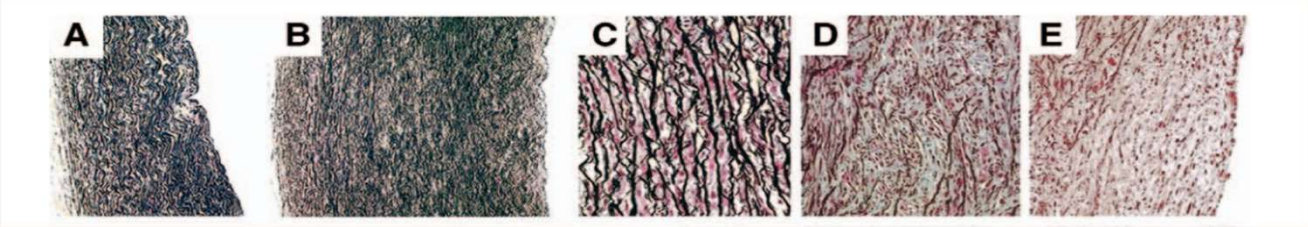
The Elastin Arteriopathy Basis

A SYSTEMIC PROCESS, NOT AN ISOLATED PROBLEM

- Williams syndrome arises from a 7q11.23 microdeletion that includes the elastin gene (ELN)
- Reduced elastin production causes smooth muscle cells to overgrow, thickening artery walls
- The result is a generalized arteriopathy, concentrated wherever elastin content is normally highest
- Cardiovascular risk should be viewed as lifelong, not resolved after childhood treatment



Normal Williams Normal Williams Williams

Infancy and Early Childhood

80%

of all patients with Williams syndrome have cardiovascular abnormalities

90%

of infants presenting in the first year of life have a cardiac finding

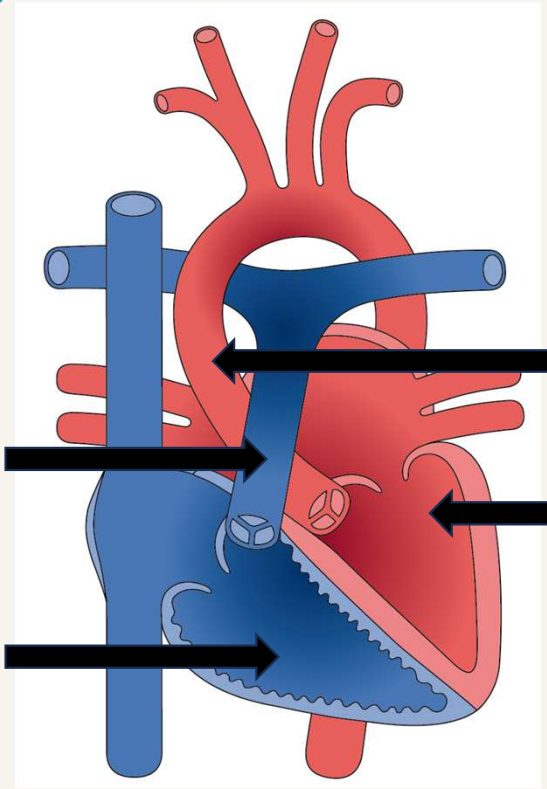
20–30%

undergo a surgical or catheter-based intervention, most by age 5

60% → 15%

peripheral pulmonary stenosis in year one, dropping sharply after infancy; SVAS, by contrast, tends to persist

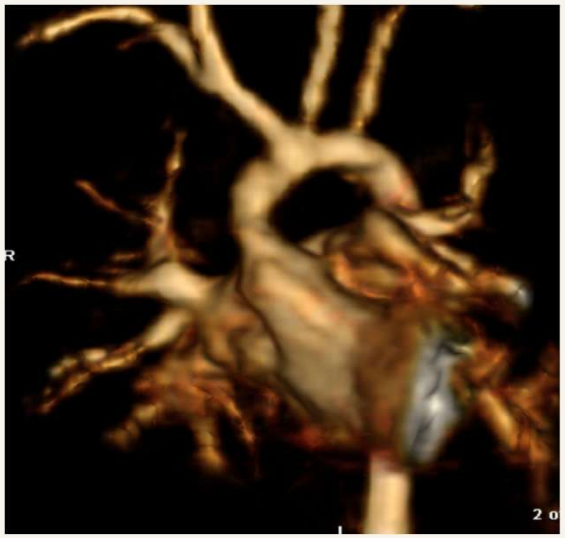
Cardiac Anatomy



Supravalvar Aortic Stenosis

Narrowing of the aorta above the aortic valve causing outflow obstruction.

Discrete "Hour-glass" Stenosis



Diffuse aortic hypoplasia

Adolescence

FEWER NEW LESIONS, RISING OTHER RISKS

- Fewer new structural cardiac lesions typically appear during adolescence
- Hypertension becomes more common, approaching rates seen in adults
- Supraventricular arrhythmias appear on ambulatory monitoring in roughly 1 in 8 children
- Ventricular tachycardia has been reported in a meaningful minority of older adolescents
- Care emphasis shifts from structural repair toward vasculopathy and blood pressure management

PART THREE
WHY SURVEILLANCE MATTERS

The Rationale for Lifelong Cardiology Surveillance

Why repaired hearts and normal evaluations still require ongoing follow-up

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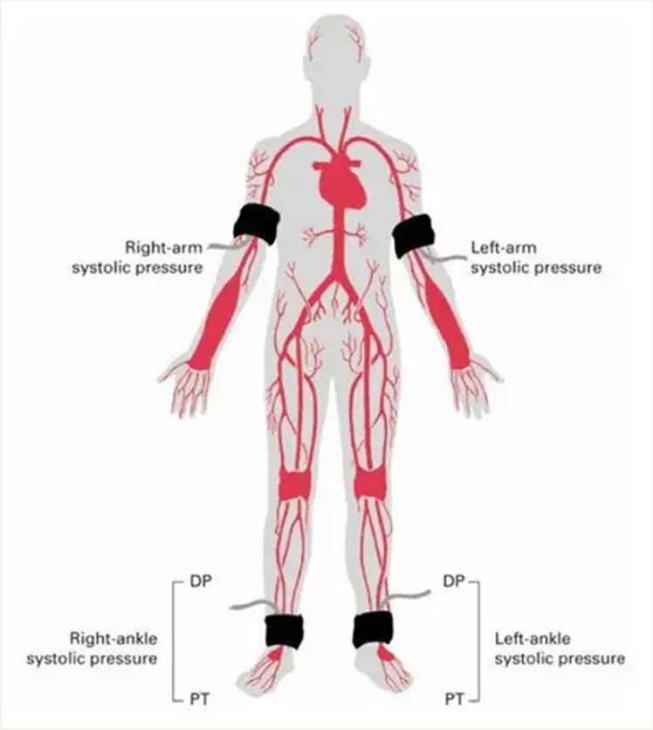
Blood Pressure Management

GETTING AN ACCURATE READING

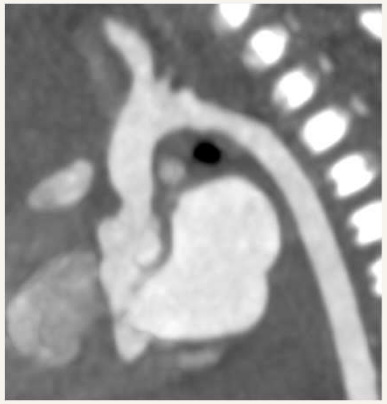
- Measure blood pressure manually in both arms and at least one leg
- Anxiety, common in WS, can falsely elevate blood pressure readings
- Severe SVAS can cause falsely elevated readings through the Coanda effect (usually R arm)
- Ambulatory blood pressure monitoring if tolerated. Many patients have elevated BP even with sleep
- No antihypertensive class is proven superior; use ACE inhibitors/ARBs cautiously until renal artery stenosis is excluded

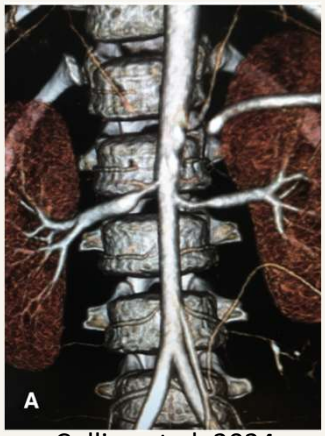


Recurrence After Repair

REPAIR IS NOT THE END OF SURVEILLANCE

- About 25% of SVAS patients require reoperation within 20 years of initial surgery
- Most SVAS reoperations occur within the first 2 years after the original repair
- Hypertension can develop later in life, independent of the original presentation
- Arterial stenosis can appear at any branch point, including renal, head, neck, and mesenteric vessels
- A successful repair still requires structured, ongoing re-imaging



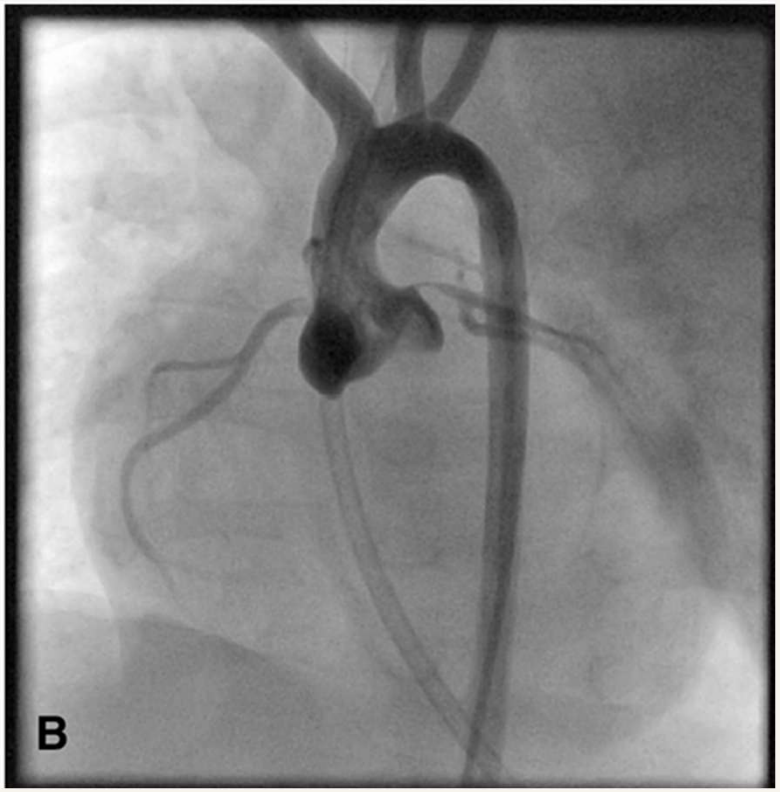


Collins et al. 2024.

Coronary Risk and Sudden Death

RISKS THAT DON'T SHOW ON A ROUTINE ECHO

- Coronary artery involvement is easy to miss on standard echocardiography
- Coronary lesions are often identified only at catheterization or surgery
- Some sudden events occur outside the periprocedural setting, without a clear trigger
- A normal echocardiogram does not rule out a clinically important coronary lesion
- Untreated hypertension can cause LVH leading to increased oxygen demand.



Collins et al. 2024.

Vascular and Coronary Imaging

WHEN TO IMAGE, AND WITH WHAT

- CT or cardiac MRI angiography is recommended for moderate or severe SVAS
- At least one adult-life study should image the aorta, coronary arteries, and pulmonary arteries
- Exercise stress testing has practical limitations due to cognitive and orthopedic factors
- CT angiography is often used instead of exercise testing to screen for ischemia in adults

Anesthesia and Procedural Risk

THE HIGHEST-RISK WINDOW

- Most sudden cardiac events in WS cluster around the periprocedural period
- Formal anesthesia risk stratification (low/moderate/high) should precede any procedure with up-to-date cardiac screening.
- Risk category determines IV placement timing and hydration protocols and is based on cardiac phenotype
- WS-experienced anesthesia and surgical teams should be engaged whenever possible
- Rapid decompensation during anesthesia may not be reversible and often requires circulatory support (i.e. ECMO)

Table 3. Risk Stratification and Prehydration Plan Before Anesthetic Administration in Patients With Williams–Beuren Syndrome

Low risk (standard anesthetic care)	Moderate risk (morning intravenous line placement and hydration ≥2h before anesthetic)	High risk* (admit preceding evening with intravenous fluid administration overnight†)
Age>10y	Hypertension	Age <3y
No cardiac involvement greater than mild SVAS or branch pulmonary artery stenosis	Moderate SVAS or branch pulmonary artery stenosis	History of adverse cardiovascular event
Normal ECG	Mild bilateral outflow tract obstruction	Preprocedural arrhythmia
No renal artery involvement	Renal artery stenosis	Bilateral outflow tract obstruction of ≥moderate severity
	Renal dysfunction	SVAS gradient of ≥40 mm Hg and the presence of left ventricular hypertrophy
	QTc on ECG >450 ms, but <500 ms	Coronary artery involvement
	Airway abnormalities, lung disease, or severe gastroesophageal reflux	Diffuse stenosis of the thoracic aorta
		Right ventricular pressure ≥75% systemic
		≥Moderate left or right ventricular hypertrophy
		Symptoms or ECG signs of ischemia
		QTc on ECG ≥500 ms

PART FOUR
TRANSITION OF CARE

Transition Periods: Adolescence to Adult Care

Recognizing the highest-risk gap and strategies to prevent loss to follow-up

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Why Transition Is Vulnerable

WHERE PATIENTS FALL THROUGH THE CRACKS

- Visit frequency naturally decreases with age, creating a false sense that surveillance is optional
- Pediatric practices often lack a built-in referral pathway to adult congenital cardiology
- Most adult cardiologists have limited experience with Williams syndrome
- Cognitive or intellectual disability can make it hard to navigate a new referral independently
- Loss of pediatric care coordination coincides with rising self-management expectations

Preventing Loss to Follow-up

BUILDING A BRIDGE, NOT A CLIFF

- Start a written transition plan in early-to-mid adolescence, not at the moment of aging out
- Arrange a warm handoff to a named adult congenital cardiologist or WS-experienced center
- Complete a baseline adult-side evaluation to establish a new reference point
- Build caregiver and family involvement explicitly into the adult care plan
- Use proactive scheduling and recall systems rather than relying on patient-initiated follow-up

PART FIVE
SHARED RESPONSIBILITY

Shared Responsibility for Cardiovascular Care

How providers, patients, and caregivers work together to sustain adherence

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A Three-Way Partnership

PROVIDERS · PATIENTS · CAREGIVERS

- Providers: maintain age-appropriate schedules and refer proactively to WS-experienced centers
- Providers: communicate clearly that feeling well is not the same as a clear cardiovascular study
- Patients: understand, to the extent of their capacity, that this is a lifelong condition
- Caregivers: track appointment history and relay symptoms the patient may not report
- Caregivers: advocate for WS-experienced anesthesia and surgical teams when procedures are needed

Adults with Williams Syndrome

Congenital Heart Disease in the Continuum of Life.

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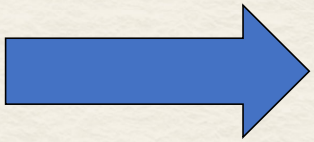
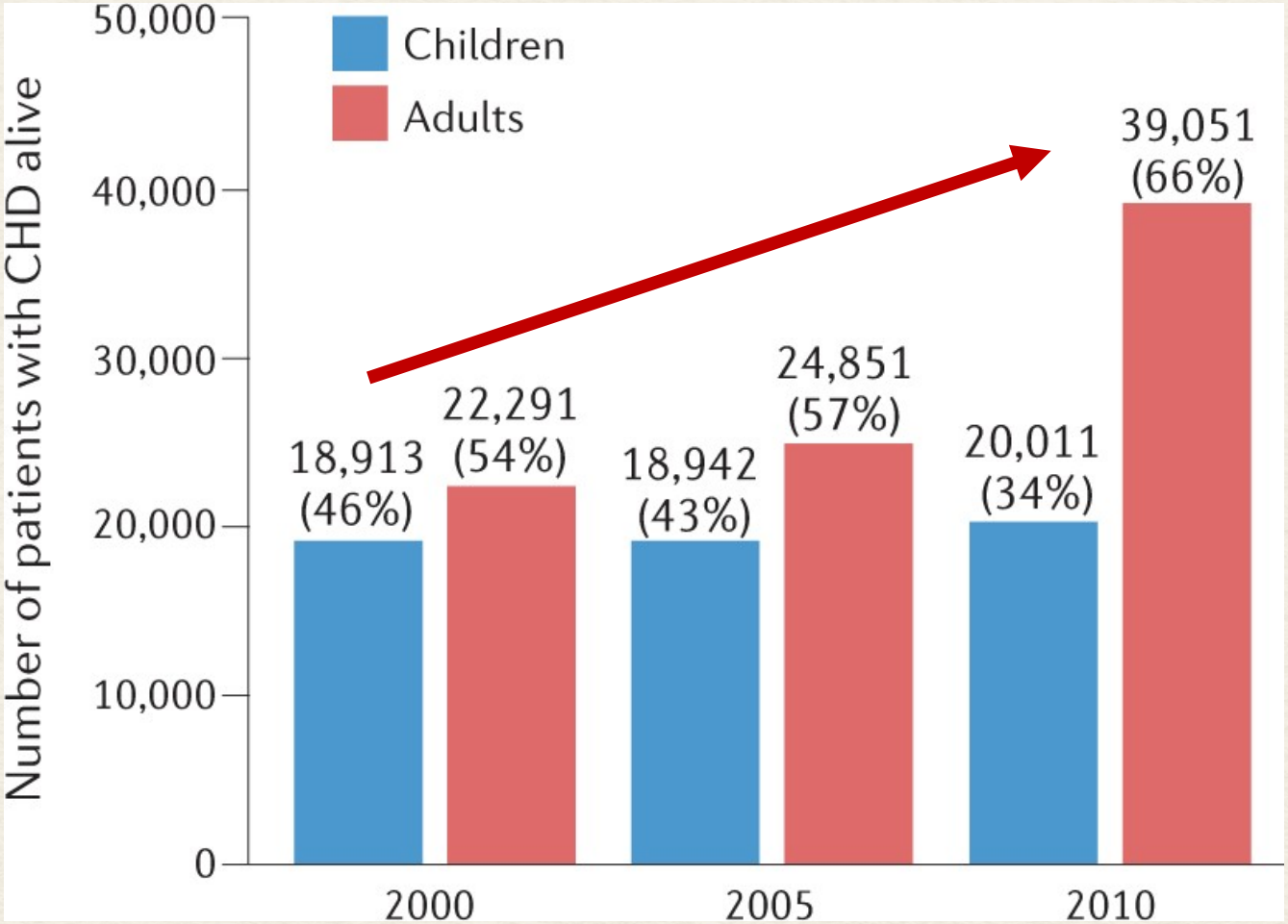
What if Mickey had Congenital Heart Disease?

Today >90% of Children Born with CHD will Survive to Adulthood





https://www.nature.com/articles/s41569-022-00749-y

Congenital Heart Disease Controversy

Repaired
To restore by replacing a part or putting together what was broken to a good or sound state

Palliate
To relieve or lessen without curing

Cure
To heal, to make well, to restore to good health without a recurrence of disease

Access to HEALTH CARE for people with disabilities

Embrace Life's Simple 7® to Reduce Your Risk for Stroke Seven Small Steps to Big Changes

1. Get Active.
2. Eat Better.
3. Lose Weight.
4. Stop Smoking.
5. Control Cholesterol.
6. Manage Blood Pressure.
7. Reduce Blood Sugar.

Plus, have regular checkups and take any medications as prescribed.

American Heart Association.
EmPOWERED to Serve™

American Stroke Association.

Head to Toe Assessment of Adults with Congenital Heart Disease

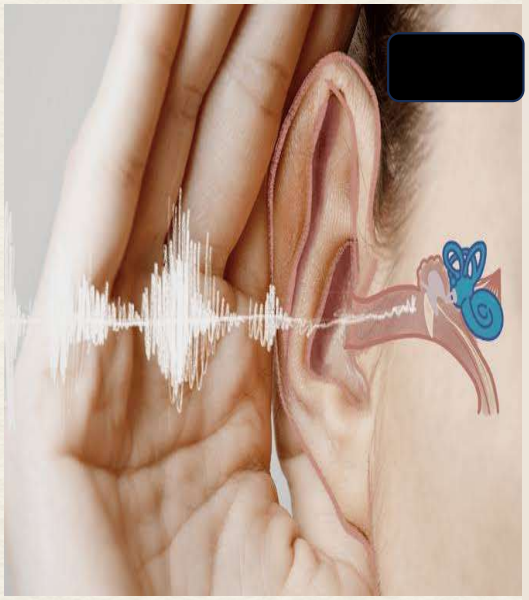
Head / Central Nervous System

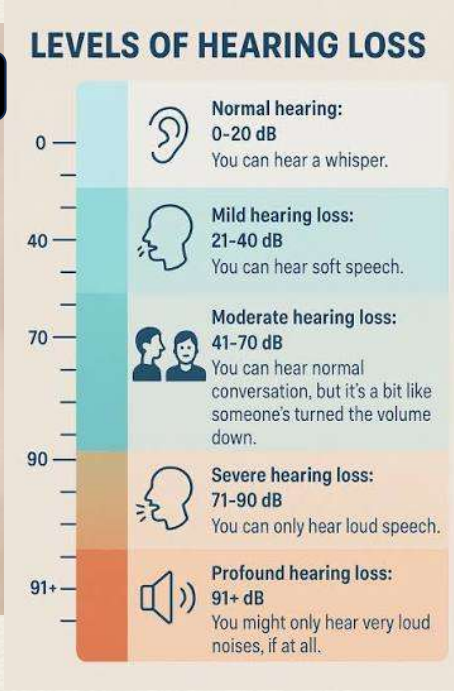
- Delayed developmental milestones
- Visual-spatial deficits
- Cognitive impairments
- Learning disabilities

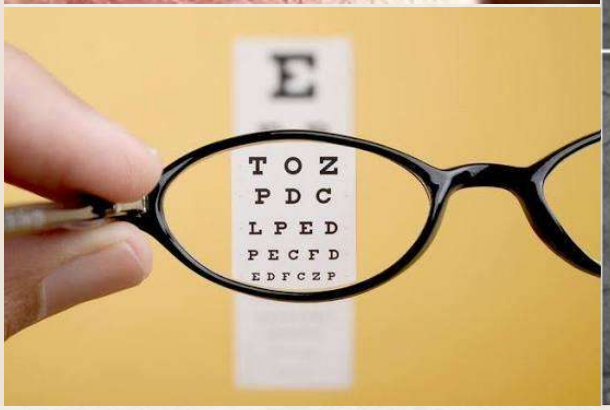
Behavioral Issues

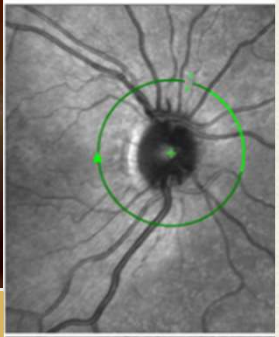
- Emotional dysregulation “outbursts”
- Confidence issues
- Phobias
- Anxiety
- Attention issues

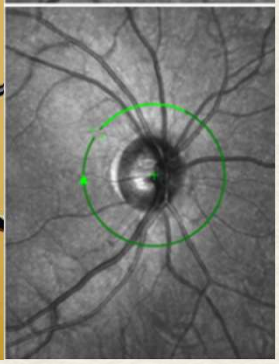
Eyes and Ears



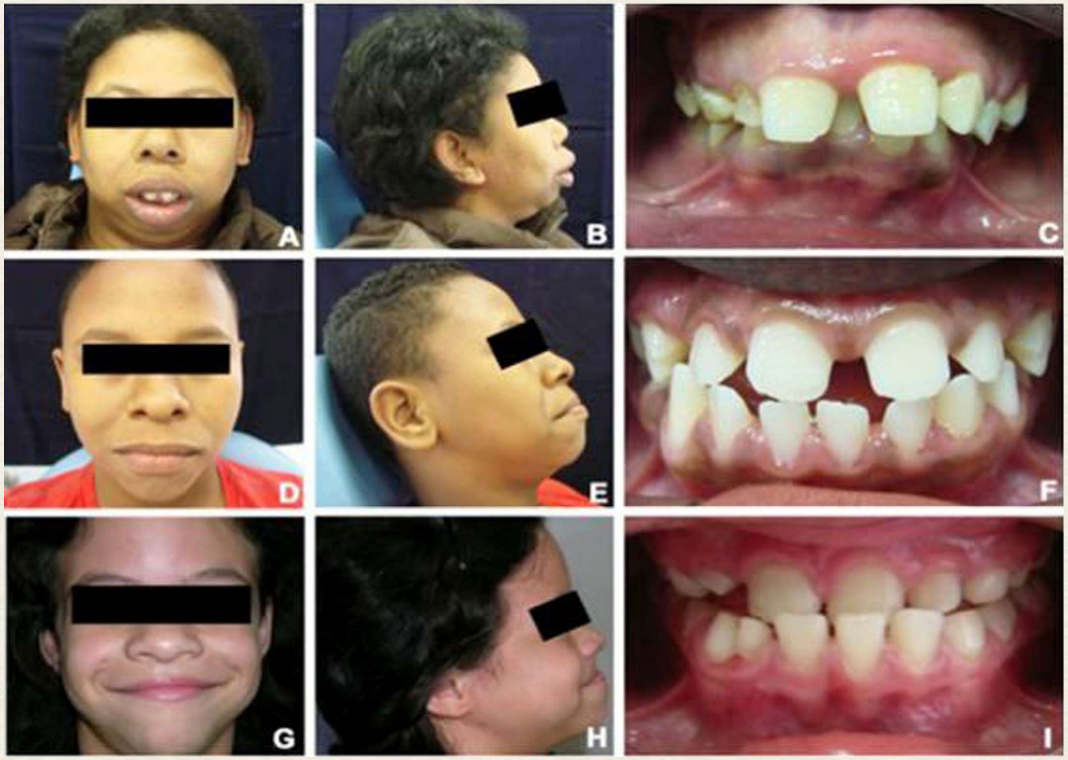








Dental Exams

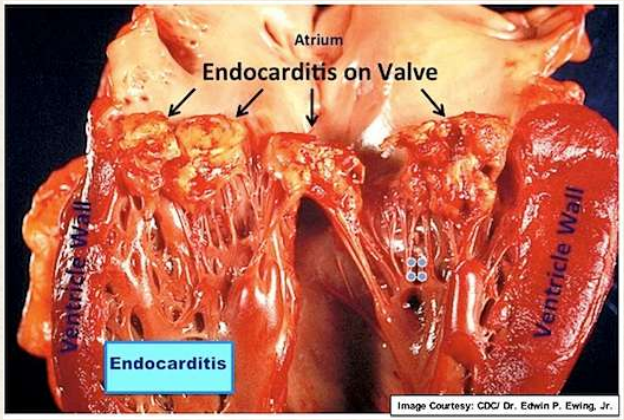


Facial and dental features of WBS. (A and B) Skeletal class II malocclusion, mandibular retrognathism, dental biprotrusion, thick lips. (C) deep bite, overjet, screwdriver shaped incisors and diastema. (D and E) thick lips, and small chin. (F) cross-bite, dental biprotrusion, and diastemas. (G and H) front and lateral pictures showing wide mouth, midface flattening. (Reprinted from Castro T, de Paula Martins Santos C, de Oliveira Lira Ortega A, Gallottini M. Oral characteristics and medical considerations in the dental treatment of individuals with Williams syndrome. Special Care in Dentistry. 2019 Mar; 39(2):108–13. DOI: https://doi.org/10.1111/scd.12361, with permission.)

Endocarditis

Antibiotic prophylaxis is reasonable before dental procedures that involve manipulation of the gingival tissue, manipulation of the periapical region of teeth, or perforation of the oral mucosa in patients with valvular heart disease who have any of the following:

- Prosthetic cardiac valves, including transcatheter-implanted prostheses and homografts
- Prosthetic material used for cardiac valve repair, such as annuloplasty rings, chords or clips
- Previous IE
- Unrepaired cyanotic congenital heart defect (CHD) or repaired CHD, with residual shunts or valvular regurgitation at the site of or adjacent to the site of a prosthetic patch or prosthetic device
- Cardiac transplant with valve regurgitation due to a structurally abnormal valve

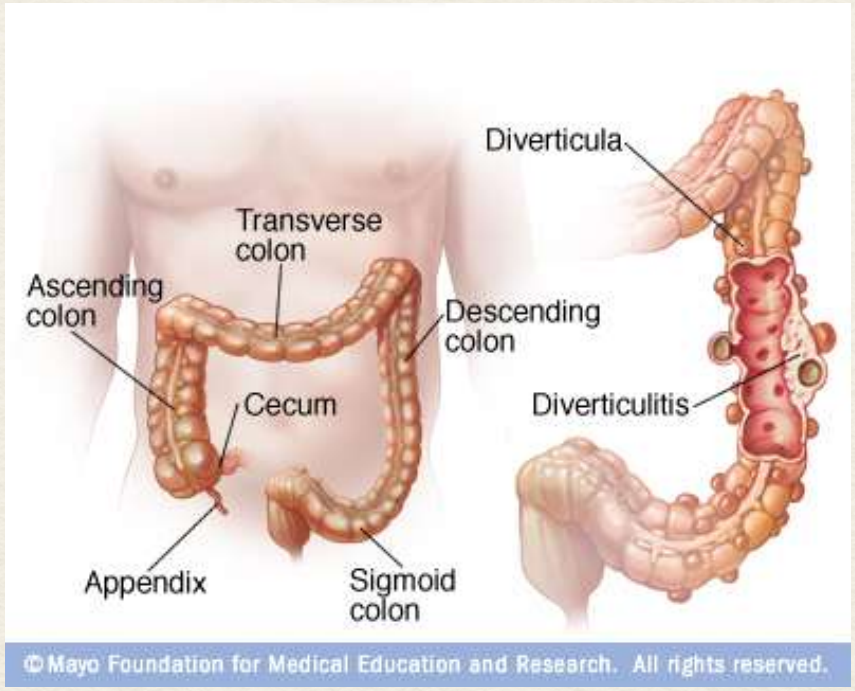


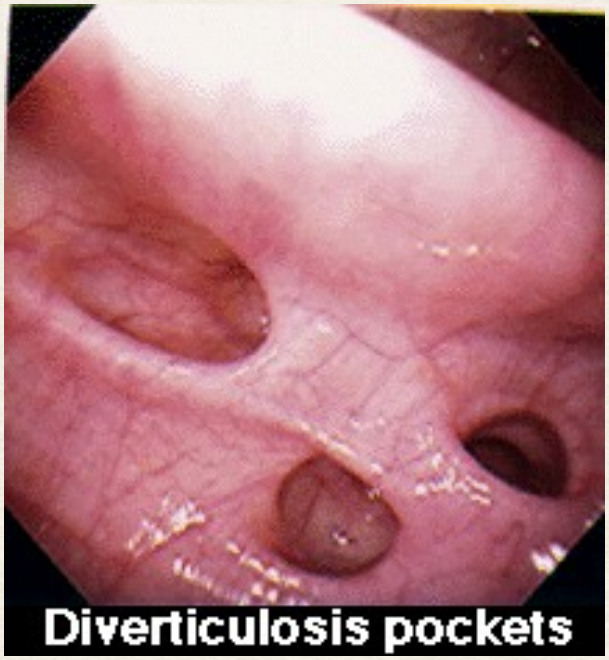
Antibiotic Prophylactic Regimens for Dental Procedures

Regimen – Single dose 30 to 60 minutes before procedure

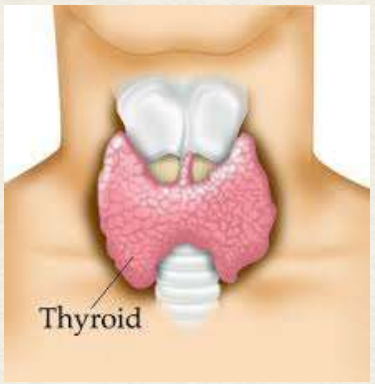
Situation	Agent	Adults	Children
Oral	Amoxicillin	2 g	50 mg/kg
Unable to take oral medication	Ampicillin **OR**	2 g IM or IV	50 mg/kg IM or IV
	Cefazolin or ceftriaxone	1 g IM or IV	50 mg/kg IM or IV

Gastrointestinal Abnormalities





Endocrine



Thyroid dysfunction
Hypercalcemia
Type 2 diabetes
Hyperlipidemia
Osteoporosis

Musculoskeletal:

Scoliosis

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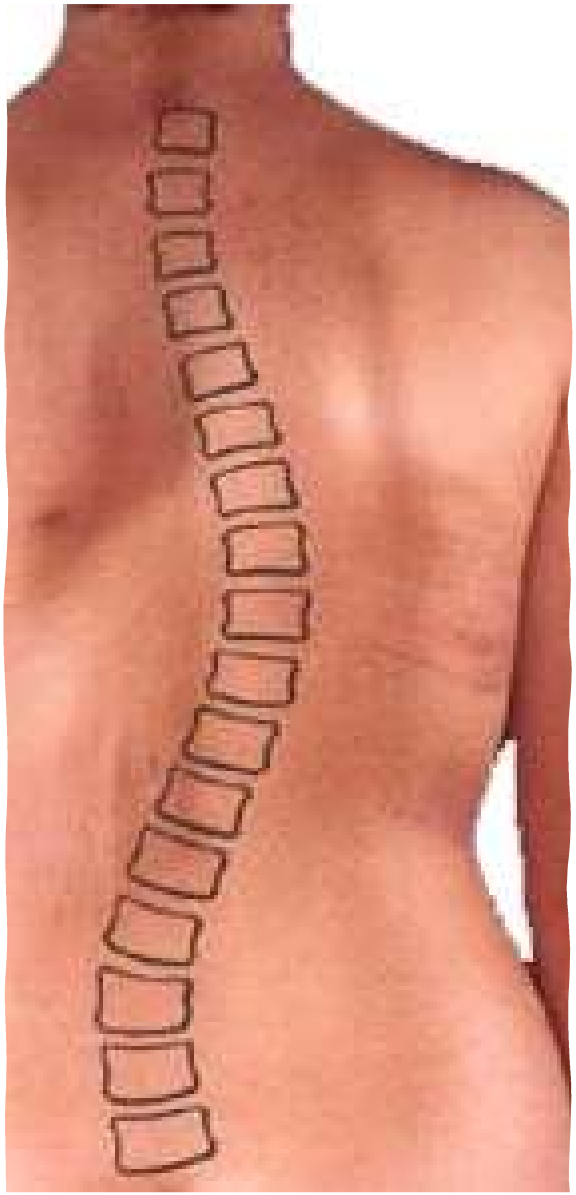

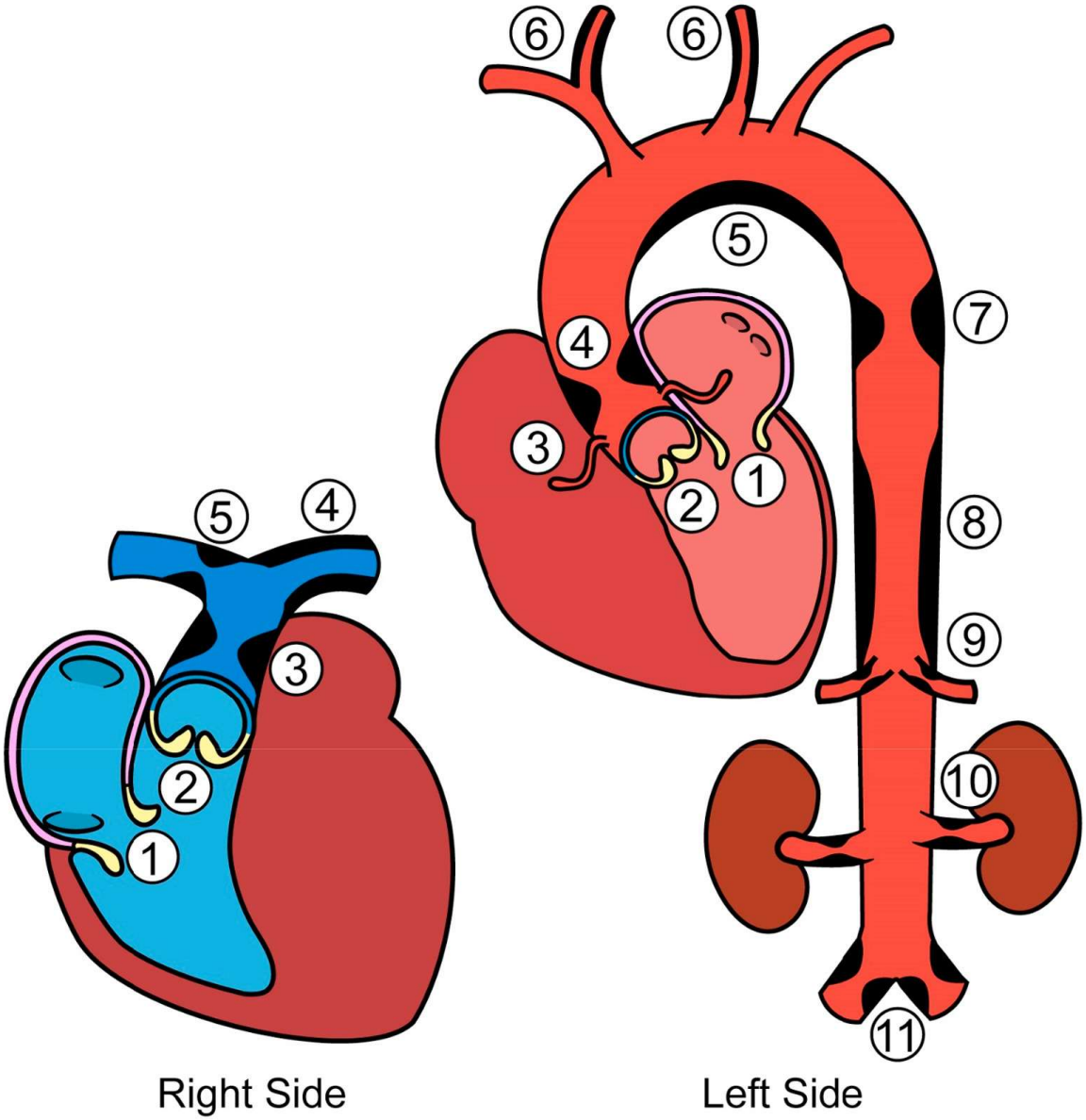

Progress in Pediatric Cardiology
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Review

Cardiovascular screening in Williams syndrome

Gul H. Dadlani [a b], Coralis Mercado [c], Val Roberts [c], Holiday Blackwelder [c], Kaye Erickson [c], Gabrielle Shrimpton [c], Jennifer Stein [c], Chloe Morrison [c], Stacey Stoner [c], Karen Bender [a b], Brian Gronert [c], Peace Madueme [a b]

Common Cardiovascular Issues in Adults with Williams syndrome

Valve deterioration (regurgitation / stenosis / need for valve replacement)

- Anticoagulation / blood thinners

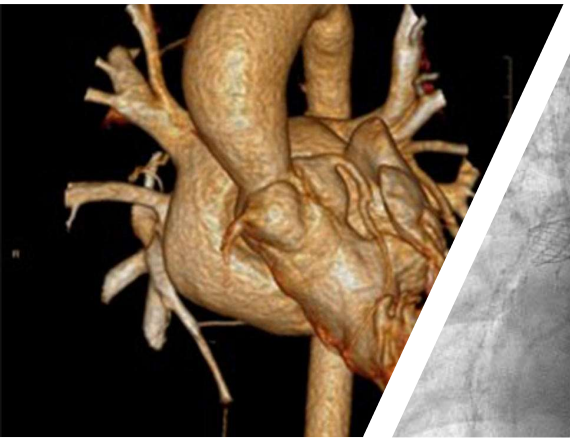
Supravalvar aortic stenosis and residual aortic hypoplasia

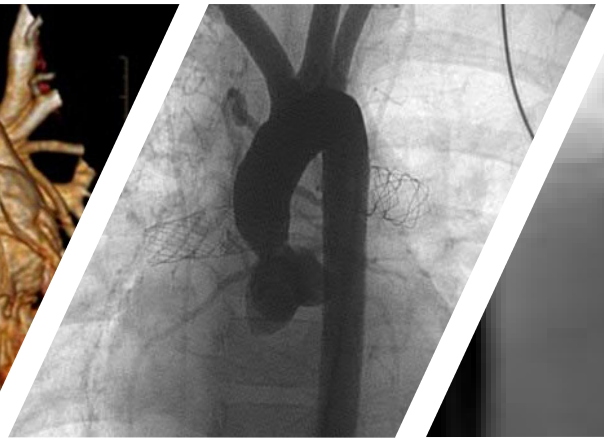
Coronary artery anomalies: hypoplasia, stenosis and development of atherosclerosis with age

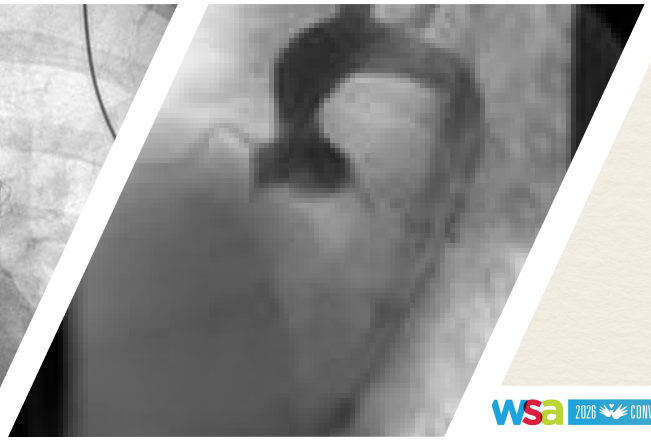
Peripheral vasculopathy (mid aortic syndrome, renal artery stenosis)

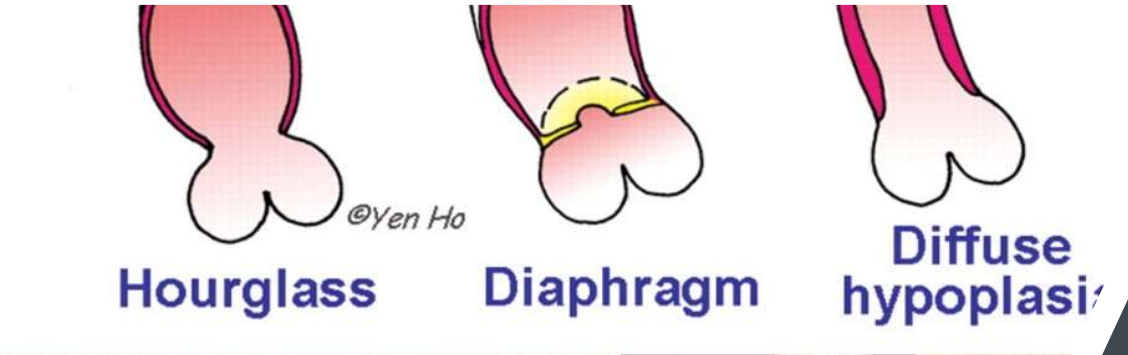
Arrhythmias (abnormal heart rhythms) = Long QTc, premature ventricular contractions and ventricular tachycardia

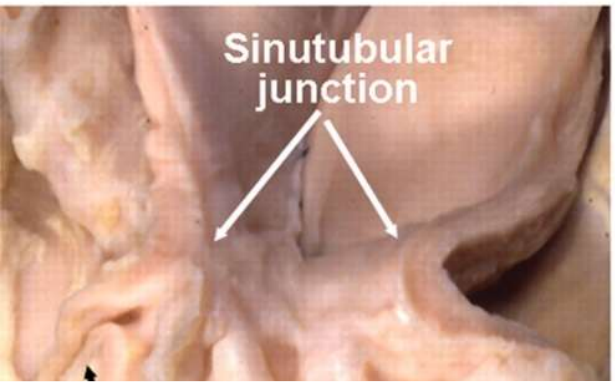
Hypertension

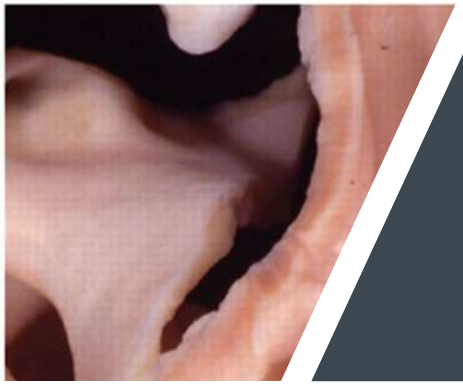












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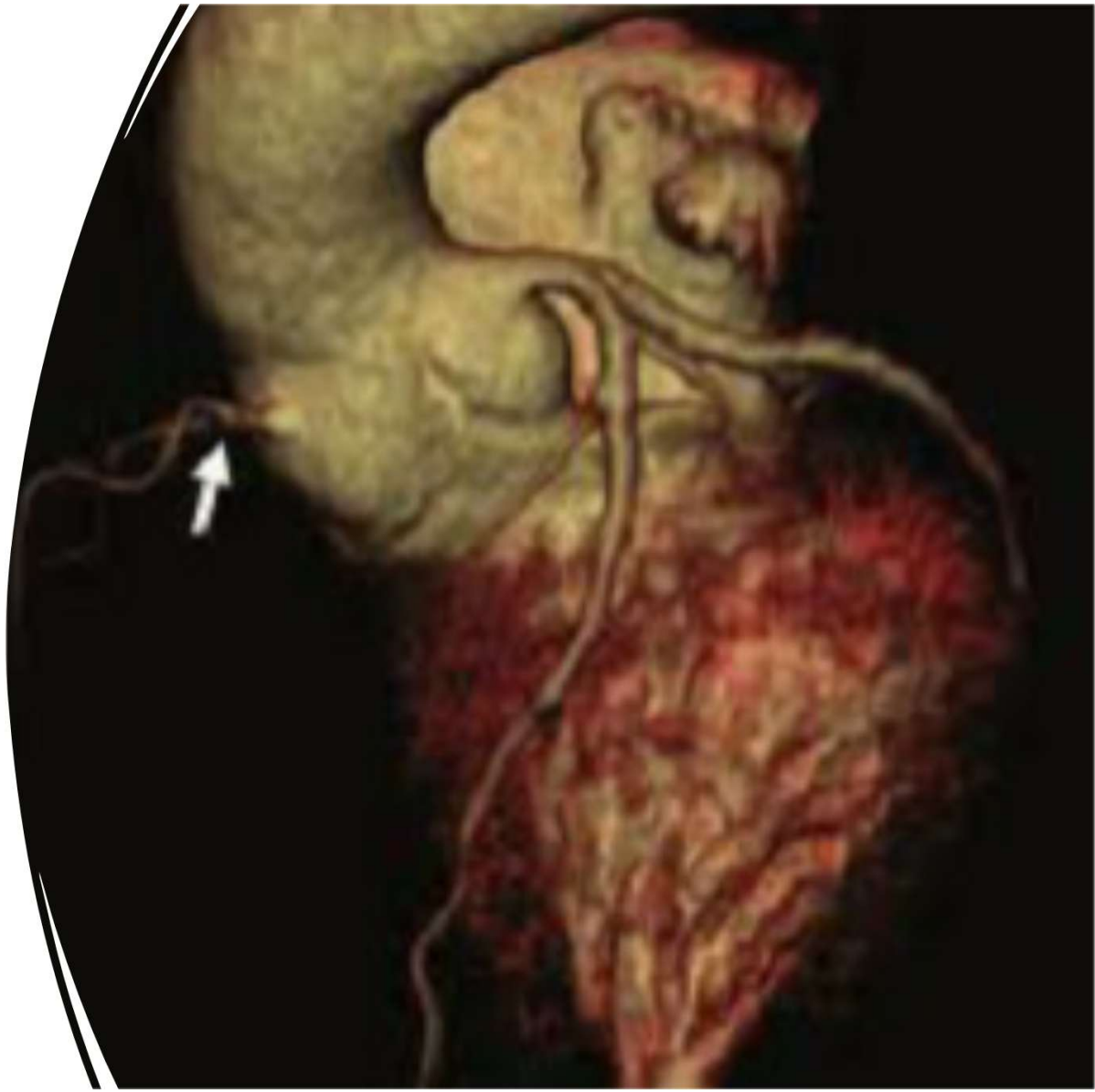
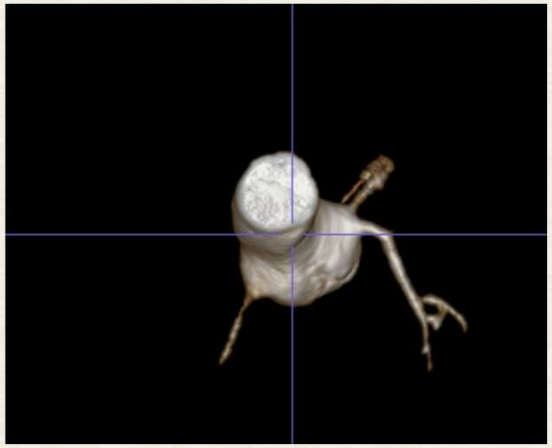
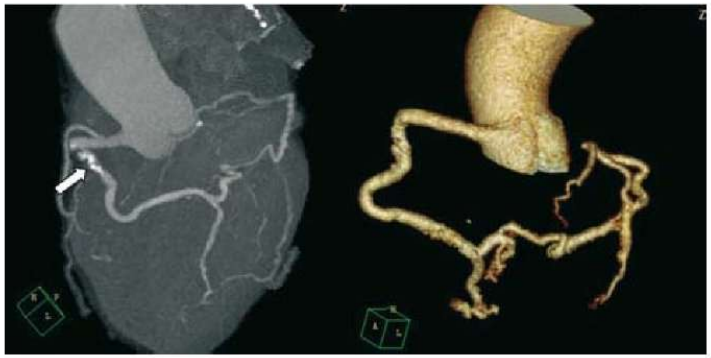
Supravalvar Aortic Stenosis

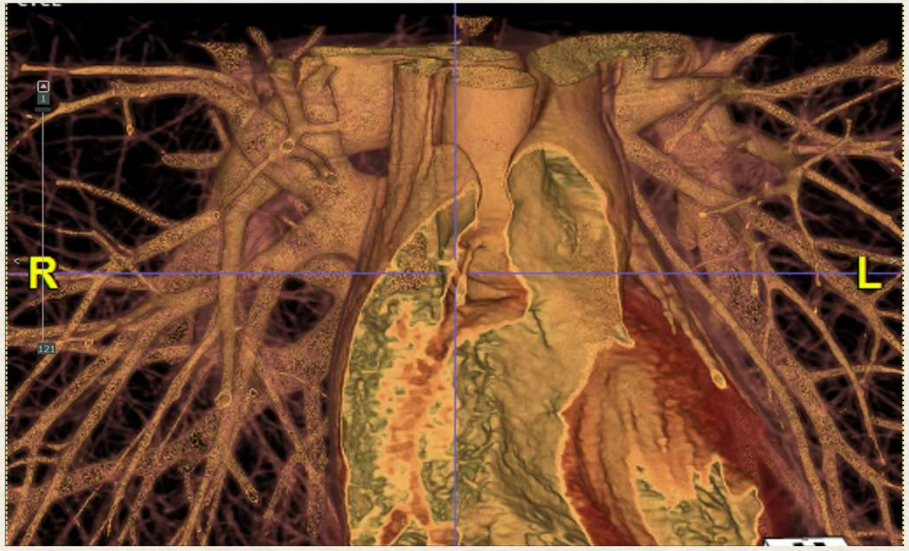
- **Coronary artery stenosis: the coronary arteries arise from the aorta and carry oxygenated blood to the heart muscle. These are the arteries that may develop blockages in later adult life and cause heart attacks. In Williams syndrome - patient's may be born with small or narrowed coronary arteries. Coronary artery stenosis may predispose to sudden cardiac arrest especially with anesthesia. This example shows a very small right coronary artery and a normal left coronary artery.**

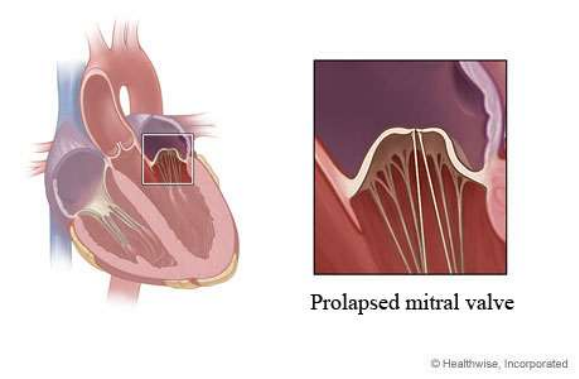


Coronary Anomalies

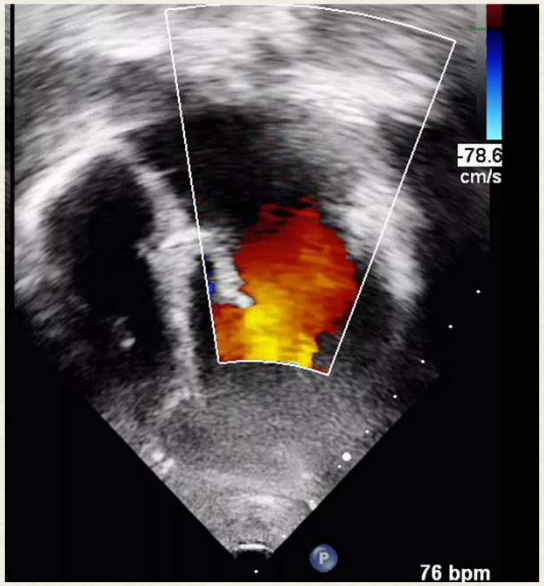


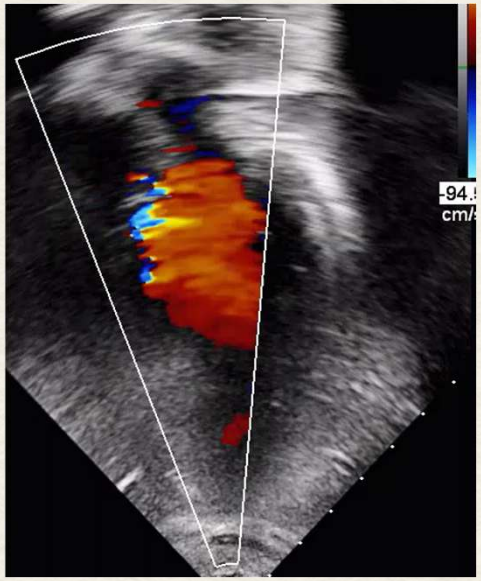


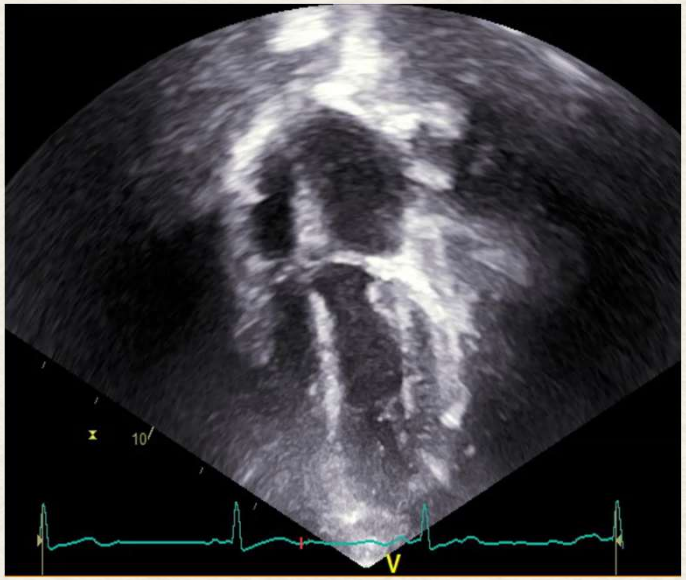




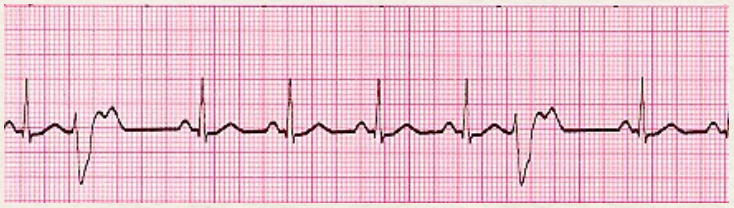
Valve Deterioration

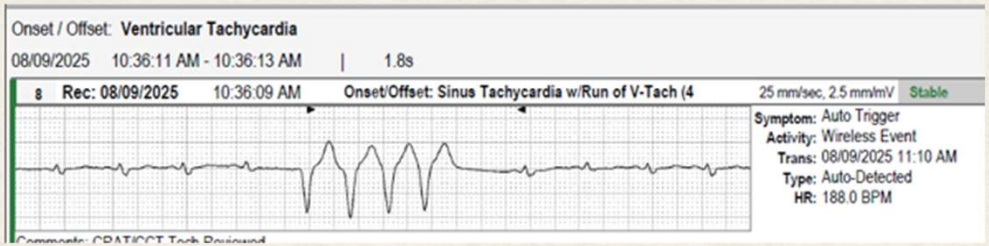


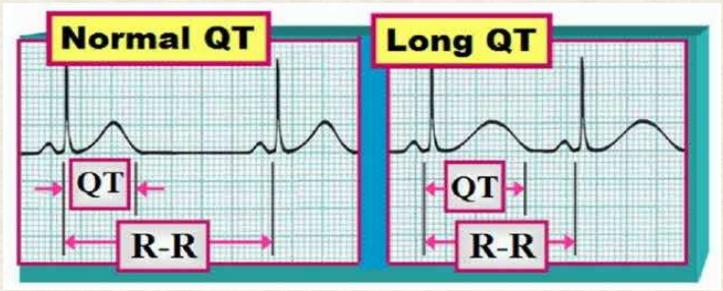


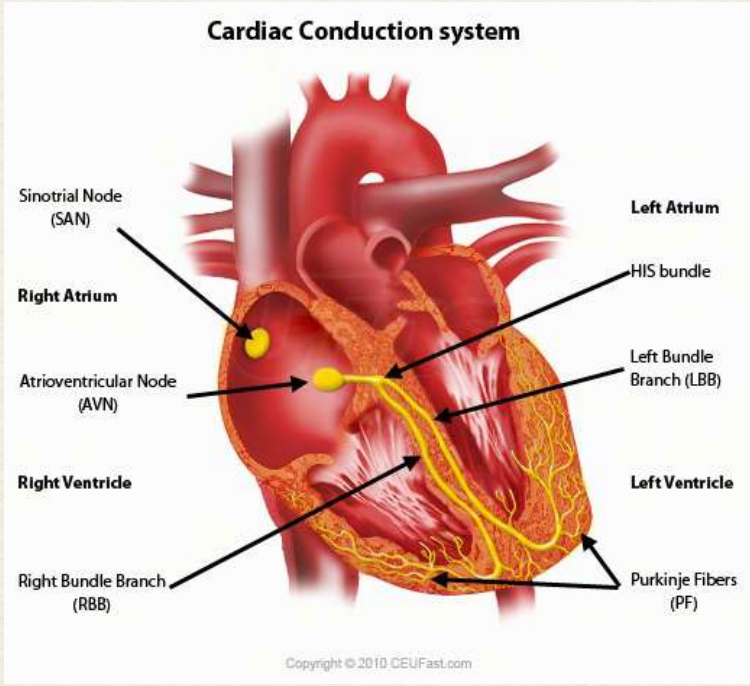


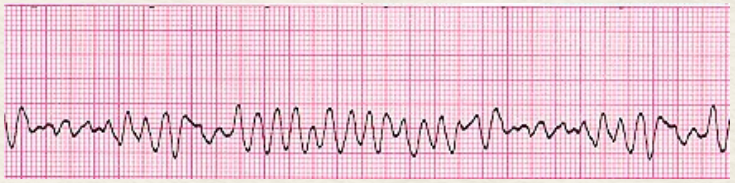
Cardiac Arrhythmias





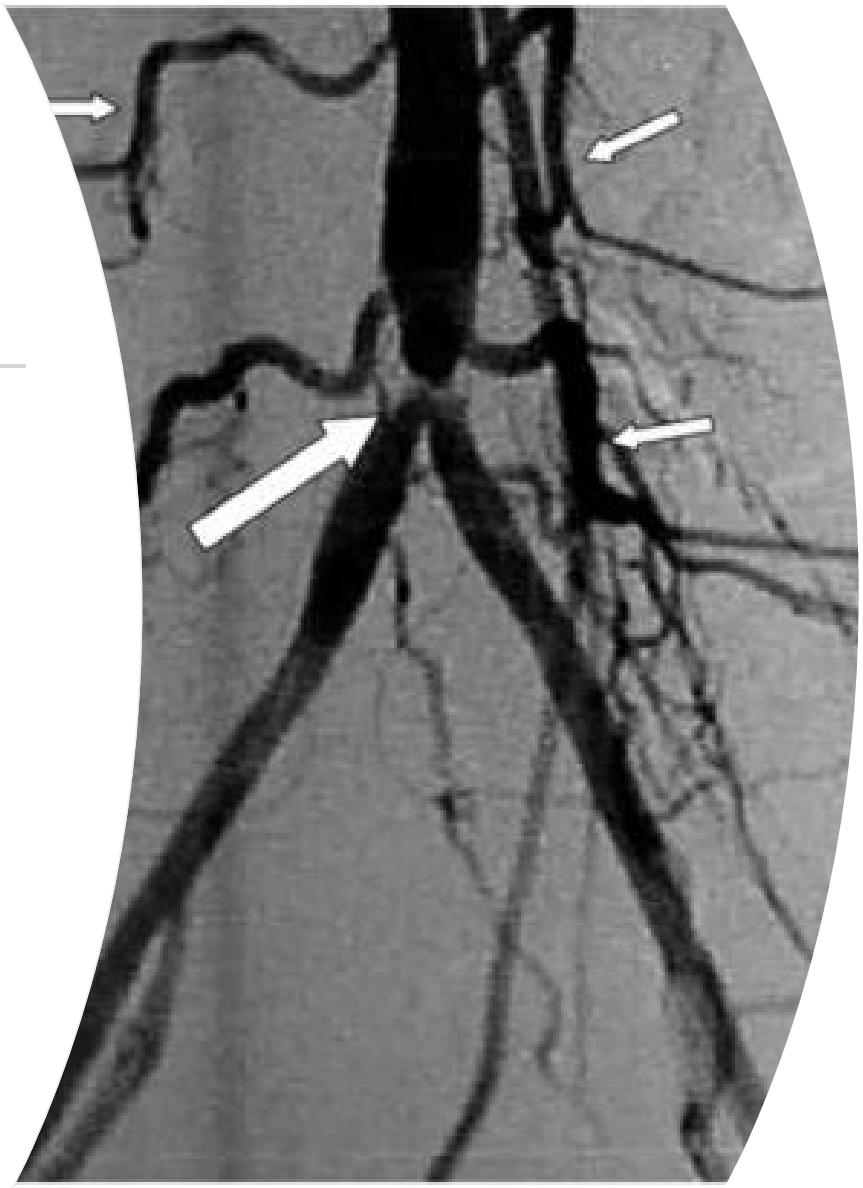


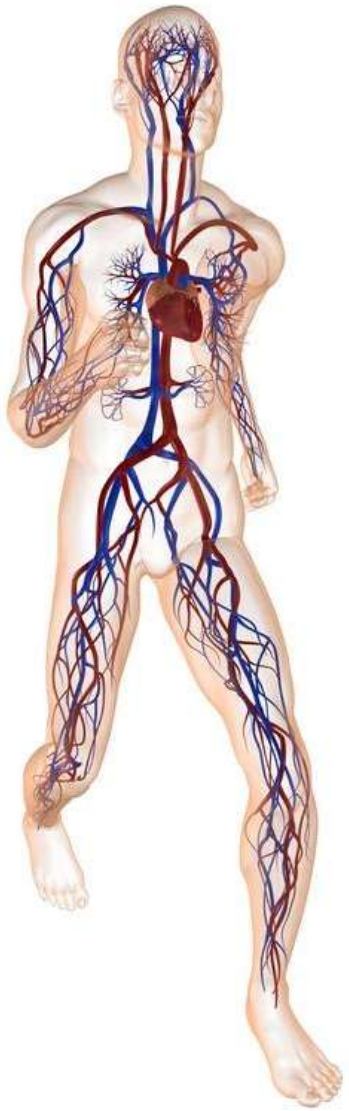


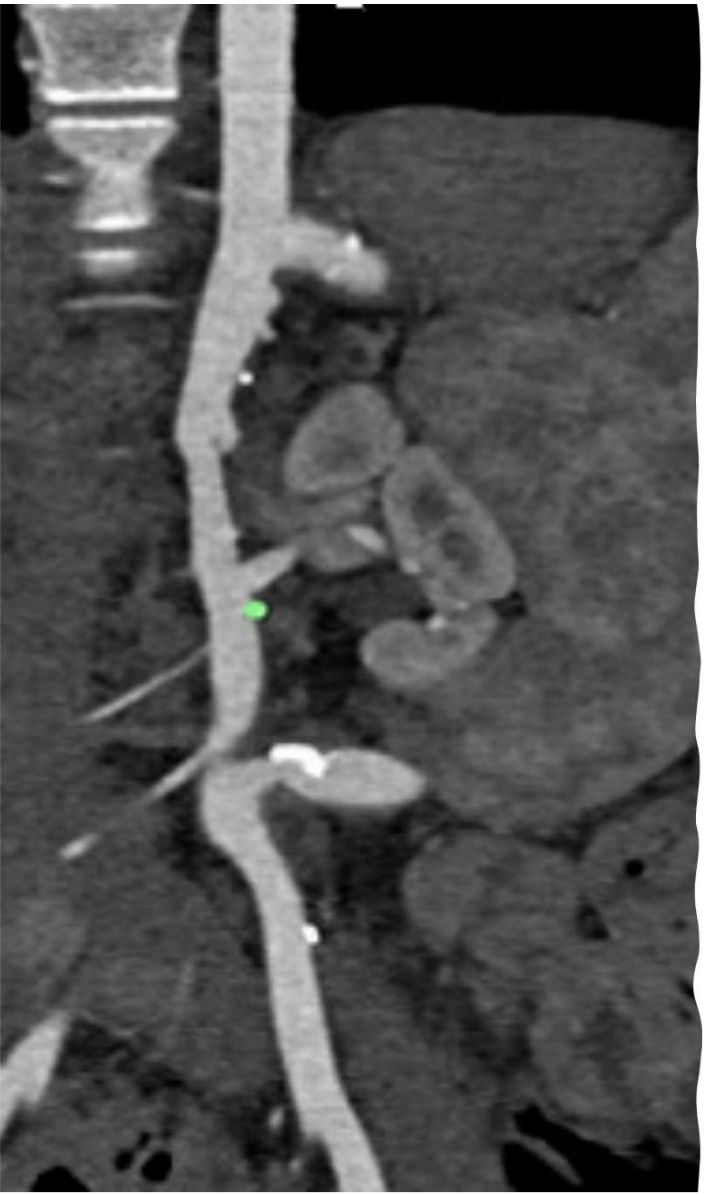


Peripheral vascular disease

- Narrowing of the arteries supplying the arms and legs.
- Risk of injury to the vessels during a cardiac catheterization
- Clots from prior lines

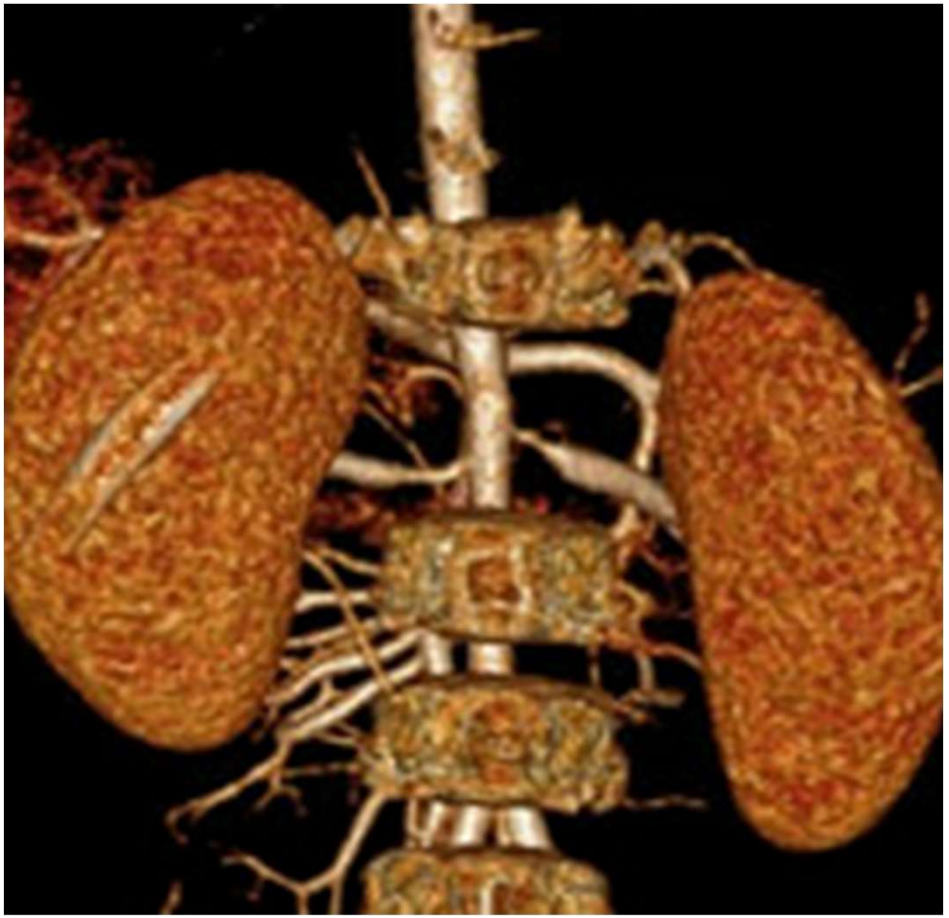


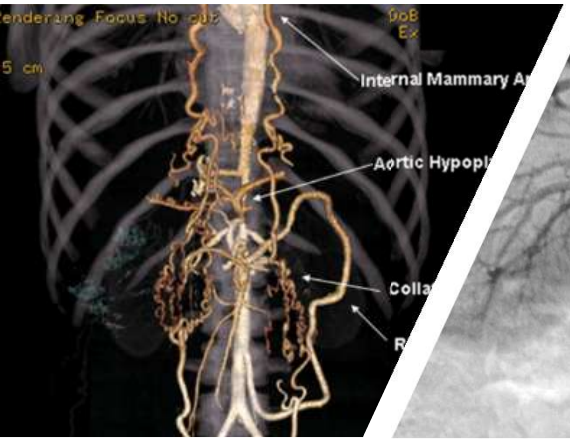


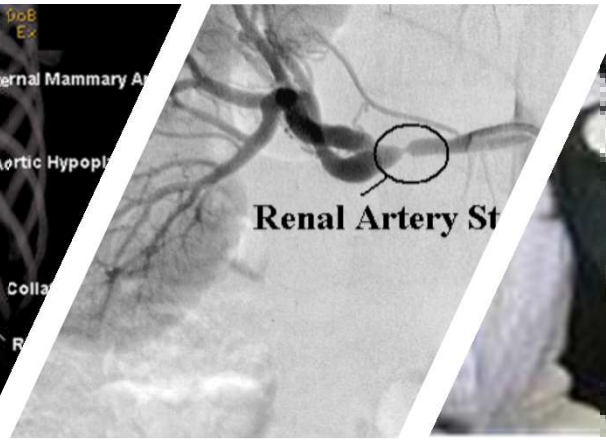


- **Mid-aortic syndrome: this is a narrowing of the aorta in lower chest or abdomen. This also causes hypertension or high blood pressure in the upper extremities and lower blood pressure in the legs. Patients may have a nausea after eating fatty foods because they have limited blood supply to the stomach and intestines or may have "claudication = pain in the legs when walking long distances" from limited blood flow to the legs.**

- **Renal artery stenosis: the aorta gives rise to a right and left renal artery, which are blood vessels that carry blood flow to the kidneys. The kidneys balance our fluids and filter the blood, taking out byproducts and making urine. In Williams syndrome - patients can develop narrowing of the renal arteries which can lead to high blood pressure. This example has narrowing of the right and left renal arteries.**







WSA 2026 CONVENTION

Blood Pressure Categories

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BLOOD PRESSURE CATEGORY	SYSTOLIC mm Hg (upper number)		DIASTO (low
NORMAL	LESS THAN 120	and	L
ELEVATED	120 – 129	and	
HIGH BLOOD PRESSURE (HYPERTENSION) STAGE 1	130 – 139	or	
HIGH BLOOD PRESSURE (HYPERTENSION) STAGE 2	140 OR HIGHER	or	
HYPERTENSIVE CRISIS (consult your doctor immediately)	HIGHER THAN 180	and	

03

Hypertension

Key Takeaways

WHAT TO REMEMBER

- Williams syndrome cardiovascular risk evolves continuously; it does not resolve after childhood
- A normal echo or a past repair does not close the book on cardiac surveillance
- Screening priorities shift with age: structural repair, then blood pressure, then coronary and vascular imaging
- The transition to adult care is the single highest-risk gap in lifelong follow-up
- Sustained outcomes depend on shared responsibility across providers, patients, and families

References

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