

# 2024 WSA National Convention

ANESTHESIA

ISSUES

**williamssyndrome**   
ASSOCIATION



# PANEL

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# Case 1

A 4 yr old needing sedation for non-cardiac surgery,  
e.g. eye muscle surgery, MRI.

No significant cardiovascular problems.

## Case 2

A 12 year old needs sedation for a dental procedure.  
The dentist is suggesting it can be done in the office.

## Case 3

An adult is due to have colonoscopy, routine or for a non-emergency problem.  
What are the considerations?

## Case 4

A child with WS who has not adequate imaging beforehand (for a variety of reasons) is posted for a sedated ECHO and CT of the heart.

## Case 5

An emergency procedure is required.  
What should be the pre-operative assessment to determine how to proceed?

**QUESTIONS ?**



**TABLE 1** Preprocedural management of patients with Williams syndrome

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Cardiovascular assessment within 1 mo of procedure <sup>a</sup>
Consider hospital admission the night prior to the procedure to insure intravenous access and ability to maintain fluid hydration, especially in patients <5 years of age
Preprocedural evaluation by an anesthesiologist
Limit period without fluid intake to 2 h
Begin maintenance intravenous fluid replacement if period of NPO is to be >2 h
Coordinate procedure to be the first case of the day
Electrocardiogram within 2 h of start of the procedure

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**Abbreviation:** NPO, nil per os.

<sup>a</sup>Assessment should include those steps previously published.<sup>8</sup>

**TABLE 3** Risk stratification for anesthetic administration in patients with Williams syndrome

Low risk	Moderate risk	High risk
Age >20 years	Hypertension	Age <3 years
No cardiac involvement greater than mild supra-avalvar or branch pulmonary artery stenosis	Moderate supra-avalvar 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 $\geq$ moderate severity
	Renal dysfunction	SVAS gradient of $\geq$ 40 mmHg 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
		$\geq$ Moderate left or right ventricular hypertrophy Symptoms or ECG signs of ischemia
		QTc on ECG $\geq$ 500 ms

**Abbreviations:** ECG, electrocardiogram; ms, milliseconds; QTc, corrected QT interval on ECG.

Adapted from Matisoff et al.<sup>46</sup>

