



Case report

Modified Doty repair for severe supravalvular aortic stenosis in a pediatric patient with Williams Syndrome: A case report

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None.

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ABSTRACT

A 2-year-old female patient with Williams syndrome, an imperforate anus, a right-sided cystic renal anomaly, and mild psychomotor delay presented with progressive severe supravalvular aortic stenosis, exertional dyspnea, and poor weight gain. Cardiac surgery was performed, using two expansion patches at the sinotubular junction and a third patch in the distal aorta. The systolic Doppler gradient was reduced from 87 to 20 mmHg, providing substantial early hemodynamic relief. The use of two patches for aortic enlargement has shown superiority over other techniques, providing better survival and longer intervals without reintervention. At follow-up, the patient remained asymptomatic and normotensive, with clinical and echocardiographic surveillance every 6 months.

Keywords: Heart Defect, Congenital; Aortic Stenosis, Supravalvular; Williams Syndrome (Source: MeSH-NLM).

RESUMEN

Reparación modificada de Doty para estenosis aórtica supravalvular severa en un paciente pediátrico con síndrome de Williams: reporte de caso

Una paciente de 2 años con síndrome de Williams, ano imperforado, anomalía renal quística del lado derecho y retraso psicomotor leve presentaba estenosis aórtica supravalvular grave y progresiva, disnea de esfuerzo y escaso aumento de peso. Se le practicó una intervención de cirugía cardíaca, utilizando dos parches de expansión en la unión sinotubular y un tercer parche en la aorta distal. El gradiente sistólico en Doppler se redujo de 87 a 20 mmHg, lo que proporcionó un alivio hemodinámico temprano y sustancial. El uso de dos parches para la dilatación aórtica ha demostrado su superioridad frente a otras técnicas, ya que proporciona una mejor supervivencia y intervalos más largos sin necesidad de reintervención. En el seguimiento, la paciente permaneció asintomática y normotensa, con vigilancia clínica y ecocardiográfica cada 6 meses.

Palabras clave: Cardiopatías Congénitas; Estenosis Aórtica Supravalvular; Síndrome de Williams (Fuente: DeCS - BIREME).

Introduction

In pediatric cardiac surgery, severe supravalvular aortic stenosis (SVAS) in patients with Williams syndrome (WS) represents a major clinical and surgical challenge due to anatomical complexity and the elevated risk of perioperative coronary ischemia and sudden death. First described in 1961, WS is a multisystem congenital disorder affecting approximately 1 in 8,000 live births, characterized by a microdeletion on chromosome 7q11.23 involving the elastin (ELN) gene⁽¹⁾. Elastin haploinsufficiency leads to abnormal vascular smooth muscle proliferation, non-compliant arterial walls, and widespread obstructive arteriopathy⁽²⁾. Cardiovascular abnormalities occur in over 90% of affected individuals, with SVAS being the most frequent lesion (accounting for up to 70–75% of cases), predisposing patients to severe left ventricular hypertrophy (LVH), myocardial ischemia, and sudden cardiac death if uncorrected⁽¹⁾.

Morphologically, SVAS is categorized into discrete (type I) or diffuse (type II) forms, depending on whether the luminal obstruction is confined to the sinotubular junction (STJ) or extends distally along the ascending aorta and arch branches⁽³⁾. Surgical intervention is indicated when the peak left ventricular outflow tract (LVOT) gradient exceeds 50 mmHg, or at lower gradients in the presence of progressive aortic valve insufficiency, myocardial ischemia, or severe concomitant right ventricular outflow tract obstruction⁽³⁾. The surgical approach is determined by the longitudinal extent of the obstruction and the specific coronary anatomy. The principal surgical techniques include:

- McGoon repair (single patch): Enlarges the supravalvular narrowing by extending a diamond-shaped patch into the non-coronary sinus⁽⁴⁾.

- Doty repair (inverted “Y” / pantaloon patch): Utilizes a bifurcated patch to expand both the right coronary and non-coronary sinuses, achieving a more symmetrical root reconstruction than single-sinus techniques^(3,5).
- Brom repair (three-patch aortoplasty): Reconstructs all three aortic sinuses using separate symmetrical patches, achieving complete anatomical and geometric restoration of the aortic root⁽³⁾.
- Myers' sliding aortoplasty: An autologous, patch-free reconstruction technique that mobilizes distal ascending aortic tissue into the three sinuses, preserving native tissue growth potential in pediatric patients⁽³⁾.

Case report

A 2-year-old female patient (weight: 10.5 kg) with Williams syndrome, an imperforate anus, a right-sided cystic renal anomaly, and mild psychomotor delay presented with exertional dyspnea and poor weight gain, symptoms that preceded documented hemodynamic progression. A transthoracic echocardiogram in November 2023 revealed SVAS with a systolic Doppler gradient of 57.6 mmHg and mild LVH. By August 2024, the systolic Doppler gradient had progressed to 87 mmHg, indicating severe obstruction and left ventricular pressure overload. Preoperative computed tomography angiography (CTA) with three-dimensional reconstruction demonstrated severe narrowing localized at the STJ and marked narrowing of the ascending aorta (**Figure 1A, 1B**).

Surgical correction was performed via median sternotomy under cardiopulmonary bypass. Anatomical exposure of the heart confirmed severe SVAS at the STJ (**Figure 2A, 2B**).

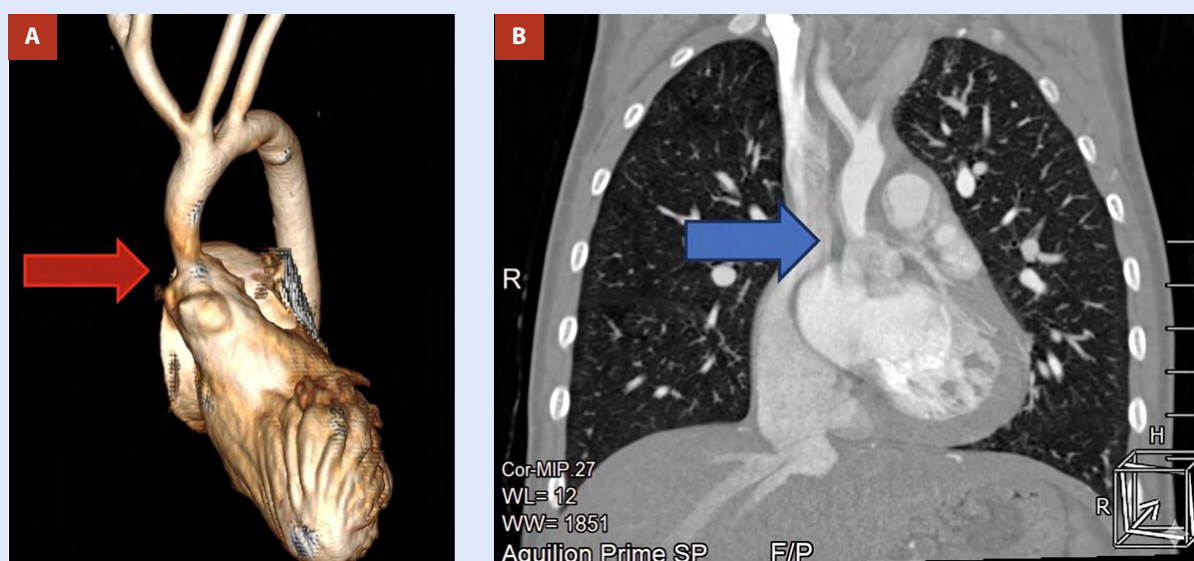


Figure 1. A. Three-dimensional CT reconstruction demonstrating severe supravalvular aortic stenosis at the sinotubular junction (red arrow). **Image B.** Coronal tomographic image showing marked narrowing of the ascending aorta at the same level (blue arrow). Images were obtained at the Instituto Nacional de Salud del Niño de Breña.

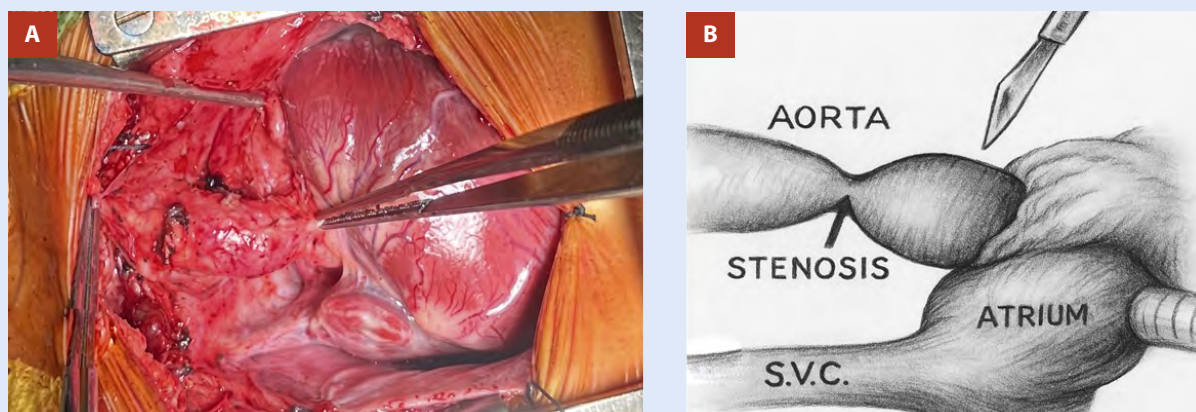


Figure 2. A. Intraoperative view demonstrating the anatomical exposure of the heart, with evident supravulvar aortic stenosis at the sinotubular junction. (blue arrow). **B.** Charcoal-style schematic drawing illustrating the anatomical relationship of the ascending aorta, supravulvar stenosis, right atrium, and superior vena cava (SVC), as well as the location of the initial surgical incision. This illustration provides a visual correlation to the intraoperative findings. Image B: Original illustration.

Following transverse aortotomy, marked LVH and severe type I SVAS were identified, with a narrowed aortic lumen measuring 2 mm in diameter (compared against a 1 mm dissection tip) and thickened aortic walls (**Figure 3A, 3B**). The aortic valve cusps were structurally intact (**Figure 4A**). Following complete resection of the thickened supravulvar tissue, aortic enlargement was performed using pericardial patches. Longitudinal incisions were made in the right and non-coronary sinuses, extending to approximately 1 mm from the aortic valve bed (**Figure 4A, 4B**). The left coronary sinus was deliberately not enlarged due to the close proximity of the left coronary artery ostium to the commissures, which prevented sufficient space for patch placement without compromising coronary flow (**Figure 4B**). An additional third pericardial patch

was placed to enlarge the distal aorta above the STJ, and aortic reanastomosis was completed with maximal symmetry to avoid altering the position of the coronary arteries (**Figure 5A, 5B**). Due to the patient's condition in the immediate postoperative period, delayed sternal closure was performed 24 hours later.

Extubation was successfully accomplished 24 hours after delayed sternal closure, and the patient was transferred from the intensive care unit on the fifth postoperative day without neurological or renal complications. Postoperative echocardiography demonstrated an immediate reduction in the systolic Doppler gradient from 87 to 20 mmHg, confirming substantial relief of the obstruction. The patient was discharged in stable condition on oral calcium channel blocker therapy for postoperative hypertension. At subsequent follow-up, she

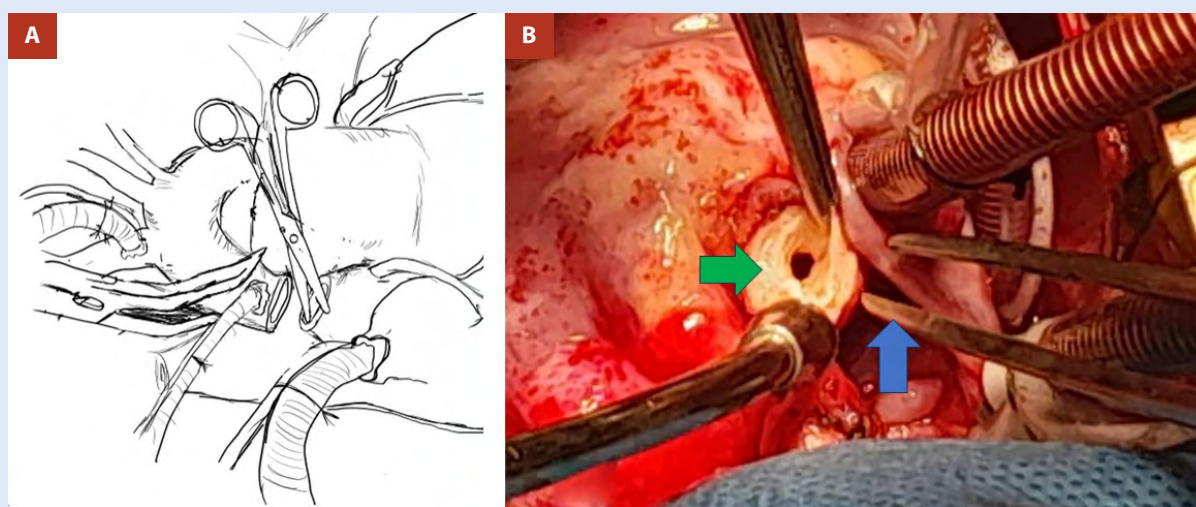


Figure 3. A. Schematic drawing showing the initial incision site and the complete removal of the thickened tissue. **B.** Anatomical image of the patient demonstrating the aortic narrowing with a lumen of 2 mm in diameter (Green Arrow) ; the width of the dissection tip is 1 mm (Blue Arrow). Image A: Original creation

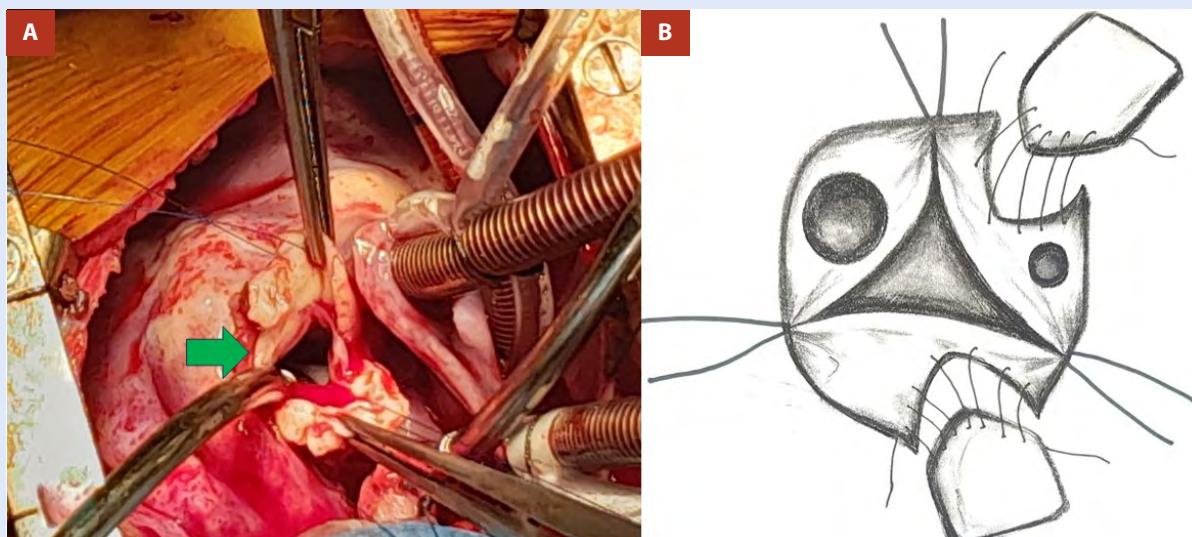


Figure 4. A. Intraoperative photograph demonstrating the aortic root following complete resection of the thickened supralvalvular tissue, with longitudinal incisions performed at the right and non-coronary sinuses to enlarge the sinotubular junction. (Green Arrow). **B.** Schematic charcoal drawing illustrating the left-dominant coronary ostium, the location and direction of the longitudinal incisions, and the anatomical placement of the enlargement patches and suture lines. Image B is original art work.

remained asymptomatic and normotensive, with clinical and echocardiographic surveillance conducted every 6 months.

Discussion

Supravalvular aortic stenosis (SVAS) in patients with Williams syndrome is an inherently progressive lesion that substantially increases the risk of left ventricular strain, myocardial

ischemia, and sudden cardiac death ^(1,3). In this patient, the rapid hemodynamic progression of the peak Doppler systolic gradient from 57.6 to 87 mmHg within a nine-month interval underscored the severity of the obstruction and mandated urgent surgical intervention.

The surgical strategy executed in this case was a customized modification of the Doty technique, employing two independent pericardial patches to enlarge the right and non-coronary sinuses, combined with an additional third patch placed along the distal ascending aorta ^(5,6).

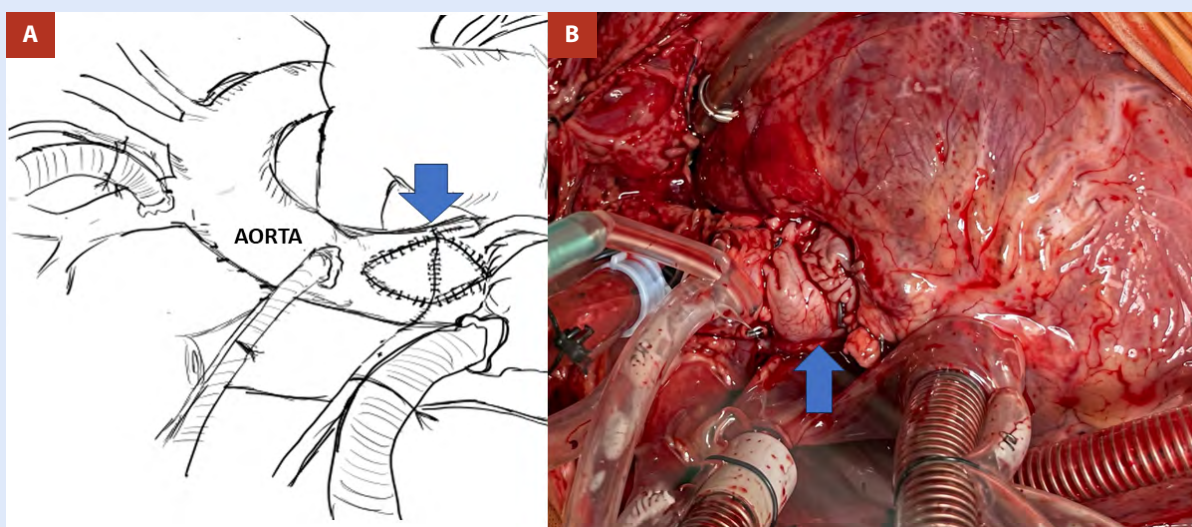


Figure 5. A. Schematic drawing showing the completed repair, with patches at the sinotubular junction and an enlargement patch in the distal aorta (Blue Arrow). **B.** Anatomical image of the patient displaying the aortic enlargement (Blue Arrow), the positioned patches, and the suture lines used. The patient is ready to be weaned off the cardiopulmonary bypass machine. Image A: Original creation.

This distal extension adheres to the principles of extended patch aortoplasty, where augmenting the vessel beyond the sinotubular junction ensures an unobstructed anatomical transition, relieves distal aortic narrowing, and prevents late anastomotic restenosis⁽⁶⁾.

Although surgical correction provides immediate hemodynamic relief, long-term outcomes following SVAS repair demonstrate a persistent cumulative burden of late complications⁽⁷⁾. In a recent meta-analysis and microsimulation study involving 1,472 patients, Meccanici *et al.* estimated a 30-year reintervention risk of 31.3% (with 27.2% directly attributable to repair dysfunction) and a 30-year risk of myocardial infarction of 8.1%, with an overall relative life expectancy of 90.7% compared to the matched general population⁽⁷⁾. Late adverse events typically stem from residual or recurrent supraaortic obstruction, progressive aortic valve insufficiency secondary to commissural distortion, and coronary ostial narrowing^(3,7). In our patient, the prompt reduction of the systolic Doppler gradient from 87 to 20 mmHg achieved effective early decompression of the left ventricle. Nevertheless, given the chronic elastopathy inherent to Williams syndrome, lifelong surveillance comprising clinical evaluation, serial Doppler echocardiography, and cross-sectional imaging every six months is essential to monitor sinotubular dimensions and aortic valve competence⁽⁷⁾.

The choice of surgical approach significantly impacts hemodynamic durability and reintervention rates^(3,8). Multi-sinus reconstruction has consistently demonstrated superiority over the traditional McGoon single-patch repair^(3,8). In a comparative series of 89 pediatric patients reported by Ibarra *et al.*, the Doty multi-sinus repair achieved a significantly greater expansion in the sinotubular junction z-score (median change: +2.5 vs. +0.8; $P=0.001$) and higher 10-year freedom from reoperation (96% vs. 67%; $P=0.008$) compared to the single-patch technique⁽⁹⁾. Single-patch augmentation often creates an asymmetrical aortic root geometry that concentrates shear stress on the non-coronary sinus and leaves untreated narrowing in the remaining sinuses, predisposing to recurrent gradients and late aortic valve dysfunction^(3,8).

Tailoring the reconstruction to individual coronary anatomy remains the primary determinant of surgical safety^(3,9). While the Brom three-patch technique offers complete geometric symmetry, it requires extensive dissection in all three sinuses and carries a documented risk of coronary ostial distortion, kinking, and acute myocardial ischemia if an ostium is displaced by adjacent patch suture lines^(3,9). As demonstrated by Lv *et al.*, multi-sinus patch modifications are frequently required to prevent ostial compression, particularly in small pediatric aortic roots⁽³⁾. In our patient, intraoperative exploration revealed an anatomical variant where the left coronary ostium arose immediately adjacent to the commissure, leaving virtually no margin for patch incorporation without compromising ostial patency. The decision to omit left sinus incision and instead enlarge the right and non-coronary sinuses, combined with a separate patch in the distal ascending aorta, successfully alleviated the left ventricular outflow tract gradient while

ensuring uninterrupted coronary perfusion^(5,6,9).

The clinical success and reproducibility of Doty-based reconstructions have also been underscored in contemporary South American pediatric experience⁽¹⁰⁾. As reported by Yagual-Gutiérrez *et al.* in a regional referral center, two-sinus enlargement provides excellent early relief of severe sinotubular obstruction with low postoperative residual gradients and favorable recovery in pediatric patients with Williams syndrome⁽¹⁰⁾. Thus, a customized multi-patch repair that avoids hazardous coronary dissection represents a reproducible, safe, and effective surgical strategy for pediatric patients presenting with complex root anatomy^(8,10).

The surgical management of supraaortic aortic stenosis in this case was successful, with postoperative results showing significant improvement in the patient's hemodynamics. The use of three patches—two in the right and non-coronary aortic sinuses and a third for distal aortic enlargement—was an effective surgical strategy that relieved the obstruction without compromising left coronary flow. This technique allowed for adequate anatomical expansion of the aorta while preserving coronary artery integrity. Despite the initial improvement, long-term follow-up is crucial to monitor the risk of restenosis or patch dysfunction, as the postoperative residual gradient still requires vigilance. The planned six-month surveillance will also assess aortic valve function and possible dilation of the reconstructed segment.

By not intervening in the left aortic sinus or modifying the left coronary ostium, this technique could represent a safe and effective surgical option for future patients, minimizing the risk of coronary ischemia. Although early hemodynamic outcomes are favorable, the progressive nature of elastin arteriopathy in Williams syndrome necessitates structured, lifelong echocardiographic and tomographic surveillance every six months to monitor sinotubular dimensions, aortic valve integrity, and potential late patch-related complications.

In conclusion, the modified Doty repair for severe supraaortic aortic stenosis in pediatric patients raises the possibility that it could become a standard surgical procedure to avoid the complications associated with intervention in all three aortic sinuses.

Ethical aspects

Informed consent for publication of the case was obtained in accordance with institutional regulations.

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Author's Contribution

JGP: conceptualization, supervision, critical review of the manuscript, and approval of the final version. **CJA:** data collection, literature review, manuscript drafting, data analysis, and approval of the final version. **MCB:** collection of clinical information, review of the manuscript, and approval of the final version.

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