

# Surgical Correction of Congenital Supravalvular Aortic Stenosis



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Supravalvular aortic stenosis is a rare congenital cardiac anomaly involving stenosis of the sinotubular junction that can extend to the ascending aorta and the aortic arch. It is an elastin arteriopathy that is classically associated with Williams syndrome. Many surgical techniques have been developed over time, most commonly involving pericardial patches to relieve the stenotic area. The 1-patch repair techniques (McGoon and Doty repairs) have low mortality but do not restore the normal morphology of the aortic root. Herein we describe the extended 3-patch technique that can symmetrically relieve supravalvular aortic stenosis.

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## Introduction

Congenital supravalvular aortic stenosis (SVAS) was first described by Chevers in 1842.<sup>1</sup> SVAS is a rare systemic arteriopathy caused by mutations in the elastin gene on chromosome 7q11.23. Defective elastin production results in arterial stiffening and proliferation of the vascular smooth muscle, causing luminal obstruction in the large systemic arteries.<sup>2,3</sup> The ascending aorta and branches of the pulmonary and coronary arteries are most commonly affected. SVAS can be localized to the sinotubular junction, where it causes a classic hour-glass appearance, or diffuse, involving the ascending aorta and the aortic arch (Fig. 1). SVAS is strongly associated with Williams syndrome, occurring in 45%-75% of these children,<sup>4,6</sup> in conjunction with other cardiovascular, neurobehavioral, craniofacial, and metabolic abnormalities. Different mutations in the elastin gene can cause a familial form of SVAS, or a form without other concomitant abnormalities (nonsyndromic SVAS).

SVAS does not always progress to require surgery.<sup>7</sup> Patients with lower left ventricular outflow tract (LVOT) gradients can be observed, with surgery indicated only in children with severe (LVOT >50 mmHg) or symptomatic lesions. Clinical presentation is similar to that of valvular aortic stenosis with left ventricular hypertrophy and congestive cardiac failure.

The first successful surgical repair of SVAS occurred in 1956, when McGoon and colleagues<sup>8</sup> used a diamond-shaped patch extending into the noncoronary sinus to relieve the stenotic aortic area. This technique became the standard surgical repair until more extensive aortoplasties were developed, aimed at restoring the physiological morphology of the aortic root. In 1977, Doty and colleagues<sup>9</sup> described their method of using an inverted Y-shaped patch to reconstruct the noncoronary sinus and the right coronary sinus. This method, however, leaves the potential for stenosis above the left coronary sinus that could narrow or obstruct the left coronary artery, with catastrophic consequences. In 1988, Brom<sup>10</sup> described a technique using 3 small patches to provide a symmetrical enlargement of the aortic root. Other authors have advocated against the use of a foreign material. In 1993, Myers and colleagues<sup>11</sup> described an autologous slide aortoplasty using flaps of the ascending aorta to enlarge the 3 sinuses without using pericardial patches.

Although an effective enlargement of the ascending aorta can be achieved with the above-mentioned techniques, the technique of choice at the Royal Children's Hospital is a modified 3-patch repair, where all 3 sinuses are incised and enlarged with glutaraldehyde-treated autologous pericardial patches (Figs. 2-7).

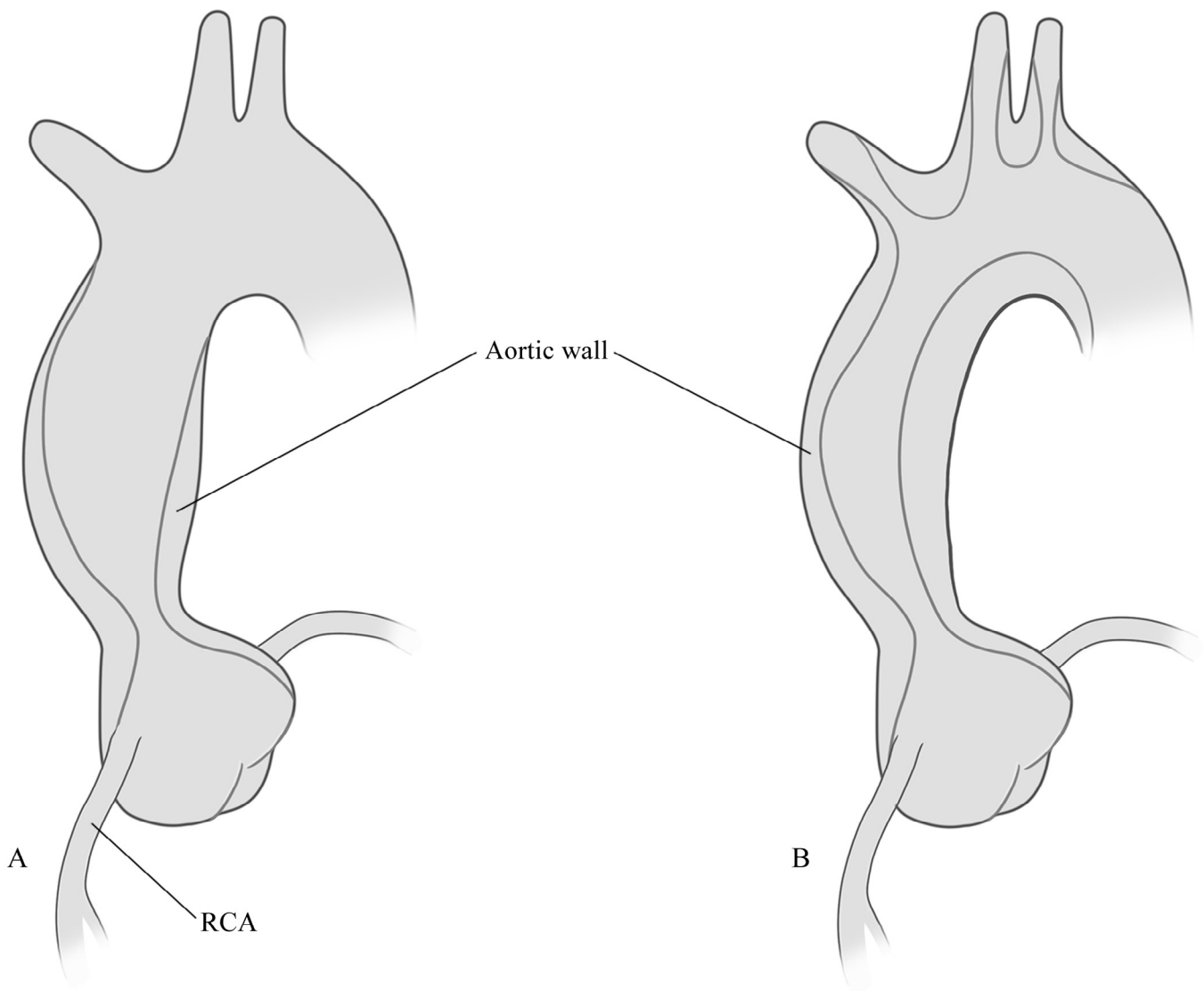
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**Figure 1** (A) One should keep in mind that supravulvar aortic stenosis is almost always associated with an excessive thickness of the aortic wall typically in the proximal ascending aorta at the level of sinotubular junction. RCA = right coronary artery. (B) However, the same process may involve the entire aortic arch and the aortic arch branches. Thus, preoperative evaluation of the extent of supravulvar aortic stenosis is crucial as the concomitant patching of the aortic arch and the aortic arch branches may be required.

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