Surgical Repair of Supravalvular Aortic Stenosis in Children With Williams Syndrome: A 30-Year Experience

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Background. **Williams syndrome is an uncommon genetic disorder associated with supravalvular aortic stenosis (SVAS) in childhood. We reviewed outcomes of children with Williams syndrome who underwent repair of SVAS during a 30-year period at a single institution.**

Methods. **Between 1982 and 2012, 28 patients with Williams syndrome were operated on for SVAS. Mean age at operation was 5.2 years (range, 3 months to 13 years), and mean weight at operation was 18.6 kg (range, 4.1 to 72.4 kg). Associated cardiac lesions in 11 patients (39.3%) were repaired at the time of the SVAS repair. The most common associated cardiac lesion was main pulmonary artery stenosis (8 of 28 [28%]).**

Results. **A 3-patch repair was performed in 10 patients, a Doty repair in 17, and a McGoon repair in 1 (3.6%). There were no early deaths. Follow-up was 96% complete (27 of 28). Overall mean follow-up was 11.2 years (range, 1 month to 27.3 years). Mean follow-up was 5 years (range, 1 month to 14.3 years) for the 3-patch repair patients and 14.7 years (range, 6 weeks to 27 years) for the Doty repair patients. Of the 17 Doty patients, there were 4 (24%) late deaths, occurring at 6 weeks, 3.5 years, 4 years, and 16 years after the initial operation. There were no late deaths in the 3-patch repair patients. Overall survival was 86% at 5, 10, and 15 years after repair. Survival was 82% at 5, 10 and 15 years for the Doty repair patients. Overall, 6 of 27 patients (22%) patients required late reoperation at a mean of 11.2 years (range, 3.6 to 23 years). No 3-patch repair patients required reoperation. Overall freedom from reoperation was 91% at 5 years and 73% at 10 and 15 years. Freedom from reoperation for the Doty repair patients was 93% at 5 years and 71% at 10 and 15 years.**

Conclusions. **Surgical repair of SVAS in children Williams syndrome has excellent early results. However, significant late mortality and morbidity warrants close follow-up.**

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Williams syndrome is a rare congenital disorder associated with cardiac defects in children, most commonly supravalvular aortic stenosis (SVAS). SVAS occurs in approximately 45% to 70% of children with Williams syndrome and requires a surgical repair in approximately 1 in 3 or 4 [1, 2]. Several surgical techniques exist for the repair of SVAS. In 1961, McGoon and colleagues [3] described the single-patch repair of the noncoronary sinus. Later, Doty and colleagues [4] introduced a pantaloon patch repair of the noncoronary and right coronary sinuses. Subsequently, Brom [5] described a 3-patch repair of all 3 aortic sinuses. Other authors have published their experience with slide aortoplasty [6, 7]. We previously described our current technique of a 3-patch repair with extension of the noncoronary sinus patch into the ascending aorta [8].

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Patients operated on for SVAS represent a heterogeneous group, because SVAS is also seen in patients with familial elastin arteriopathy and in sporadic cases [9]. Previous studies of SVAS outcomes have included all patients with SVAS and have not specifically looked at patients with Williams syndrome. Williams syndrome patients have comprised between 14% and 60% of patients in these studies [10–13] (Table 1). Thus, few data exist specifically on the outcomes of repair of SVAS in children with Williams syndrome. In this study we sought to determine outcomes of 28 patients with Williams syndrome who underwent surgical repair for SVAS during a 30-year period at a single institution.

Material and Methods

This retrospective study was approved by the Royal Children's Hospital Human Research Ethics Committee.

Patients

Between April 1982 and March 2012, 28 patients with Williams syndrome underwent operations for SVAS at

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Abbreviations and Acronyms

AVR = aortic valve replacement
CABG = coronary artery bypass graft
LAD = left anterior descending artery
LITA = left internal thoracic artery
LVAD = left ventricular assist device
MPA = main pulmonary artery
MV = mitral valve
PA = pulmonary artery
SVAS = supravalvular aortic stenosis

the Royal Children's Hospital; of these, 3-patch repair was performed in 10 patients, a Doty repair in 17 , and a McGoon repair in 1. Nine of the last 10 SVAS repairs were 3-patch repairs (Table 2). Table 3 summarizes the preoperative data. Eighteen patients (64%) were boys and 10 (36%) were girls. Mean age at operation was 5.2 years (range, 3 months to 13 years), and mean weight at operation was 18.6 kg (range, 4.1 to 72.4 kg). The SVAS was localized in 24 patients (86%) and was diffuse in 4 (16%). The 4 patients with diffuse disease had involvement of the aortic arch. In 3 of them the aortic arch was patched, including 1 patient who had concomitant patching extending into the left common carotid artery, and an end-to-side coarctation repair was performed in 1 patient. One patient (3.6%) was operated on before the SVAS repair. This patient underwent pericardial patch repair of the main pulmonary artery (MPA) and right pulmonary artery, infundibulectomy, and transannular patching at age 4 months age before undergoing a 3-patch repair at age 5 years.

Associated cardiac lesions in 11 patients (39.3%) were repaired at the time of the SVAS repair. The most common associated cardiac lesion was MPA stenosis (28%). Eleven patients (39.3%) had concomitant procedures with the SVAS repair. The most common concomitant procedure was pericardial patch repair of MPA stenosis (17.9% [5 of 28]). The mean preoperative peak gradient was 71 mm Hg (range, 30 to 144 mm Hg).

Definitions

Early death or reoperation was defined as death or reoperation occurring before hospital discharge or within 30 days of the operation. Late death or reoperation was defined as death or reoperation occurring after discharge and more than 30 days after the operation.

Table 1. Number of Williams Syndrome Patients in Supravalvular Aortic Stenosis Outcome Studies

Study First Author	Study Period	SVAS Repair Patients No.	Williams Syndrome Patients No. (%)
Brown [10]	1962–2000	101	14 (14)
Hickey [11]	1976–2006	47	28 (60)
Kaushal [12]	1990–2008	20	10 (50)
Metton [13]	1995–2007	34	14 (41)

SVAS = supravalvular aortic stenosis.

Surgical Technique

SVAS was repaired using one of three techniques (Fig 1 and Fig 2). Standard techniques were used to establish cardiopulmonary bypass. A Doty repair in 17 patients was performed using an inverted Y-shaped incision into the noncoronary and right coronary sinuses and insertion of a pantaloon patch. An extended 3-patch repair with extension of the noncoronary sinus patch [8] was used in 10 patients. A McGoon repair with a single incision into the noncoronary sinus and insertion of a single patch was used in 1 patient.

Statistical Analysis

Data were imported into Stata 10 software (StataCorp LP, College Station, TX). Continuous variables are reported as a mean with an accompanying range. Kaplan-Meier curves were constructed to display freedom from the study's outcomes.

Results

Early Deaths

There were no early deaths. Table 4 summarizes the intraoperative data. There was 1 (4%) early reoperation. This patient underwent a McGoon repair with mitral valve repair for a prolapsed anterior leaflet at age 6 months and underwent a redo mitral valve repair 16 days later.

Late Outcomes

Follow-up was 96% complete (27 of 28). Overall mean follow-up was 11.2 years (range, 1 month to 27.3 years). Mean follow-up was 5 years (range, 1 month to 14.3 years) for the 3-patch repair patients, 14.7 years (range, 6 weeks to 27 years) for the Doty repair patients, and 9.3 years for the patient who had a McGoon repair. The mean postoperative echocardiographic peak gradient was 19 mm Hg (range, 4 to 78 mm Hg), and no patient had

Table 2. Type of Supravalvular Aortic Stenosis Repairs by Era

Era	SVAS Repairs	3-Patch Repairs	Doty Repairs	McGoon Repairs
1982–1992	12	0	12 (7 Dacron,[a] 1 GoreTex,[b] 1 Tascon,[c] 1 homograft)	0
1992–2002	9	4 (1 GoreTex)	5 (2 homograft)	0
2002–2012	7	6	0	1
1982–2012	28	10	17	1

[a] DuPont, Wilmington, Delaware. [b] W.L. Gore and Associates, Flagstaff, Arizona. [c] Tascon Medical Technology Corporation, Los Angeles, California.

SVAS = supravalvular aortic stenosis.