Objective: Congenital supravalvular aortic stenosis (SVAS) is an uncommon obstructive arteriopathy of varying severity with frequent association with stenoses of systemic and pulmonary arteries. Several techniques for symmetric reconstruction of the aortic root in SVAS have been developed, but it remains unclear what is the optimal surgical procedure. We reviewed our experience with surgical management of SVAS.

Methods and results: Forty patients underwent operations to treat congenital SVAS at our institution between 1987 and 2012. Twenty-six patients were male (65%) and nine under 1 year old (22%). Twenty-five patients had associated Williams-Beuren syndrome (62%) and 49% had pulmonary artery involvement. The mean preoperative gradient pressure was 79.85 ± 27 mmHg. Surgical procedures included patch enlargement of the noncoronary sinus only (McGoon’s technique) (n = 4), inverted bifurcated patch plasty (Doty’s technique) (n=24), triple-sinus reconstruction of the aortic root (Brom’s technique) (n=6) and sliding aortoplasty (Myers-Waldhausen’s technique) (n=6). Nine patients had associated procedures (22.5%). The mean postoperative pressure gradient was 16 ± 7.1 mmHg. There was one early death. Among those who survived the early postoperative period, 95% were alive at 5 years, 86% were alive at 10 years, and 72% were alive at 20 years. According to time-related analysis there was no difference in terms of survival and reoperation between different surgical techniques (p=NS). Patients under 1 year old have worse prognosis for survival, freedom from reoperations and re-interventions (p<0.05).

Conclusions: In our cohort, the most important prognostic factor was the age at the first operation. Surgical treatment is palliative and requires careful follow-up examinations. Good surgical outcome can be achieved with the appropriate method of treatment in patients with both localized and diffuse SVAS, but because each case is different, the surgeon should individualize the approach based on the anatomic findings.