Background. Several surgical techniques for the treatment of congenital supravalvular aortic stenosis have been developed, yet there is no consensus about the optimal approach. We reviewed our institutional experience with 2- and 3-sinus reconstruction techniques.

Methods. Thirty-eight patients operated on for supravalvular aortic stenosis between 1987 and 2012 in our institution were analyzed retrospectively. Eight patients (21%) were infants and in 5 (13.2%) diffuse stenosis was present. Mean peak pressure gradient was 86.1 ± 28.7 mm Hg preoperatively. Surgical procedures included single-patch enlargement (McGoon, n = 3), inverted bifurcated-patch aortoplasty (Doty, n = 22), 3-sinus patch augmentation (Brom, n = 8), and autologous slide aortoplasty (n = 5). Major concomitant procedures were performed in 10 patients (26.3%).

Results. Early mortality was 2.6%. Follow-up continued for a median of 7.5 years (range 3 weeks to 22 years). Overall survival estimates were 94% and 90% and overall freedom from reoperation was 83% at 5 and 20 years, respectively. No differences were found between surgical techniques in respect to survival, clinical course, hemodynamic outcome, or freedom from reoperation rates. A significantly worse outcome in regard to survival and reoperation rates was observed in infants.

Conclusions. Our study demonstrates equally good results for the repair of supravalvular aortic stenosis with both 2- and 3-sinus reconstruction. No evidence of a superior outcome for 3-sinus reconstruction techniques was found. Operation in infancy is an important factor associated with unfavorable outcome.

Supravalvular aortic stenosis (SVAS) is a rare congenital malformation defined by an aortic narrowing at the level of the sinotubular junction. Supravalvular aortic stenosis is now known to be an elastin arteriopathy caused by mutations involving the elastin gene located on chromosome 7 (7q11.23), either as hemizygous deletion along with other genes as part of Williams-Beuren syndrome (WBS) or as localized mutations found in patients with SVAS not displaying the typical morphologic features of WBS [1, 2]. The severity and characteristics of the arteriopathy are highly variable and can involve narrowing of the entire ascending aorta, the aortic arch, and aortic arch vessels, the descending aorta, and its branches. Additionally, obstructions of the pulmonary arterial vasculature are often associated with SVAS. The first successful surgical repair was performed in 1956 by McGoon and colleagues [3] using a single diamond-shaped patch extending into the noncoronary sinus to enlarge the stenotic area. Subsequently, various surgical techniques for the relief of SVAS were developed, including the extended aortoplasty introduced by Doty and colleagues [4] which reconstructs both the non-coronary and the right coronary sinus as well as 3-sinus reconstruction techniques such as the triple-patch augmentation described by Brom [5].

Dysfunction and abnormalities of the aortic valve, including dysplastic thickening of the leaflets, bicuspid valve, and leaflet adherence to the sinotubular ridge, as well as coronary artery stenoses, are frequently observed [6]. Hence, surgical strategies aiming at restoration of the aortic root configuration with a symmetric reconstruction of all 3 sinuses are thought to improve long-term outcome of SVAS repair. However, there is no...
convincing evidence so far that the 3-sinus reconstruction techniques provide superior results compared with 1- or 2-sinus reconstructions [7–10]. We reviewed our 25-year institutional experience with various techniques for the repair of SVAS and investigated the long-term results.

Patients and Methods

Patients

A total of 41 patients undergoing surgery for the relief of congenital SVAS in our institution between 1987 and 2012 were identified, of whom 3 had had previous surgeries for SVAS in other institutions and were thus excluded from further analysis. The Institutional Ethics Committee approved the study. The need for individual consent was waived.

Patients’ characteristics are summarized in Table 1. Median age at operation was 3.3 years (range 0.1 to 51.7 years). Eight patients (21%) were less than 1 year of age at the time of operation and 5 were adults. Complete follow-up data were available for 33 (87%), incomplete data at least confirming survival and reoperation or reintervention status for 4 (10.5%), and no follow-up data for 1 patient (2.6%). Follow-up continued for a median of 7.5 years (range 3 weeks to 22 years). In 20 patients (52.6%) Williams-Beuren syndrome (WBS) was present. Five patients (13.2%) were classified as having familial SVAS; in these cases abnormalities typical for WBS were absent and at least 1 additional family member was diagnosed with SVAS. Noonan syndrome was present in 1 patient (2.6%). Twelve patients (31.6%) were found to have sporadic SVAS as none of them displayed developmental delay, dysmorphic features, or a family history of SVAS.

Morphologically, SVAS was defined as discrete when the supravalvular narrowing was ring-like and confined to the sinotubular junction, resulting in the typical hourglass shape of the aortic root. The SVAS was defined as diffuse if the narrowing extended to the ascending aorta, usually forming a long-segment tubular narrowing. Discrete SVAS was present in 33 patients (86.8%) while in 5 patients (13.2%) SVAS morphology was diffuse; all these patients were children (age 4 months to 9 years). Three patients (7.9%) had additional moderate stenosis of aortic arch vessels while no significant further systemic artery stenoses were observed. Associated left ventricular outflow tract anomalies and involvement of the pulmonary vasculature with presence of main, branch, or peripheral pulmonary artery stenosis were frequent, while additional cardiac malformations were rarely observed (Table 1). Preoperative diagnostic cardiac catheterization was performed in 30 patients (78.9%). In 4 patients (10.5%) catheter interventions had been performed, including balloon valvuloplasty of aortic valve stenosis (n = 1), attempted balloon dilatation of SVAS (n = 1), dilatation of pulmonary artery stenosis (n = 1), and closure of an aorto-pulmonary collateral (n = 1). Previous surgical procedures had been performed in 3 patients (7.9%) and included relief of right ventricular outflow tract obstruction (n = 1), aortic valve valvulotomy (n = 1), and closure of a ventricular septal defect together with resection of aortic coarctation (n = 1). Preoperative clinical and hemodynamic findings are given in Table 2. Left ventricular systolic function and functional capacity was well preserved in most cases. Two infant patients presented with acute cardiac decompensation requiring inotropic support and 1 patient aged 2 years experienced a sudden cardiac arrest during diagnostic catheterization. Two of these 3 patients needed continuous cardiopulmonary resuscitation during transfer to emergency surgery for SVAS relief.

Table 1. Patients’ Characteristics

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median (range)</td>
<td></td>
</tr>
<tr>
<td>Age (years)</td>
<td>3.3 (0.1-51.7)</td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>14.8 (5.5-157)</td>
</tr>
<tr>
<td>Sex</td>
<td>male to female 25/13 (65.8/34.2)</td>
</tr>
<tr>
<td>SVAS morphology:</td>
<td></td>
</tr>
<tr>
<td>Discrete</td>
<td>33 (86.8)</td>
</tr>
<tr>
<td>Diffuse</td>
<td>5 (13.2)</td>
</tr>
<tr>
<td>Associated cardiovascular anomalies:</td>
<td></td>
</tr>
<tr>
<td>LVOT</td>
<td></td>
</tr>
<tr>
<td>Aortic valve anomalies:</td>
<td></td>
</tr>
<tr>
<td>Stenosis</td>
<td>6 (15.8)</td>
</tr>
<tr>
<td>Dysplastic leaflets (without stenosis)</td>
<td>7 (18.4)</td>
</tr>
<tr>
<td>Bicuspid valve</td>
<td>2 (5.2)</td>
</tr>
<tr>
<td>Insufficiency &gt; mild</td>
<td>2 (5.2)</td>
</tr>
<tr>
<td>Subvalvular stenosis</td>
<td>2 (5.2)</td>
</tr>
<tr>
<td>Coarctation</td>
<td>2 (5.2)</td>
</tr>
<tr>
<td>RVOT</td>
<td></td>
</tr>
<tr>
<td>Pulmonary vascular anomalies:</td>
<td></td>
</tr>
<tr>
<td>Main PA stenosis</td>
<td>6 (15.8)</td>
</tr>
<tr>
<td>Branch PA stenosis</td>
<td>12 (31.6)</td>
</tr>
<tr>
<td>Peripheral PA stenosis</td>
<td>5 (13.2)</td>
</tr>
<tr>
<td>Pulmonary valve stenosis</td>
<td>2 (5.2)</td>
</tr>
<tr>
<td>Infundibular stenosis</td>
<td>1 (2.6)</td>
</tr>
<tr>
<td>Coronary anomalies:</td>
<td></td>
</tr>
<tr>
<td>Single coronary ostium</td>
<td>2 (5.2)</td>
</tr>
<tr>
<td>Dystopic coronary ostia</td>
<td>1 (2.6)</td>
</tr>
<tr>
<td>Coronary aneurysm</td>
<td>1 (2.6)</td>
</tr>
<tr>
<td>Coronary stenosis</td>
<td>3 (7.9)</td>
</tr>
<tr>
<td>Other</td>
<td></td>
</tr>
<tr>
<td>Patent foramen ovale</td>
<td>4 (10.5)</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>1 (2.6)</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>2 (5.2)</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>2 (5.2)</td>
</tr>
</tbody>
</table>

* Numbers refer to patients with any of the respective anomalies; presence of more than one of the further specified anomalies is possible.

LVOT = left ventricular outflow tract; PA = pulmonary artery; RVOT = right ventricular outflow tract; SVAS = supravalvular aortic stenosis.
Table 2. Preoperative Clinical and Hemodynamic Findings

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Functional capacity, N (%)</td>
<td></td>
</tr>
<tr>
<td>NYHA or modified Ross heart</td>
<td></td>
</tr>
<tr>
<td>failure classification class</td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>24 (63.2)</td>
</tr>
<tr>
<td>II</td>
<td>11 (28.9)</td>
</tr>
<tr>
<td>III</td>
<td>-</td>
</tr>
<tr>
<td>IV</td>
<td>3 (7.9)</td>
</tr>
<tr>
<td>Left ventricular function N (%)</td>
<td></td>
</tr>
<tr>
<td>Good (LVEF &gt; 0.65)</td>
<td>31 (81.6)</td>
</tr>
<tr>
<td>Mildly impaired (LVEF 0.55-0.65)</td>
<td>3 (7.9)</td>
</tr>
<tr>
<td>Moderately impaired (LVEF 0.45-0.55)</td>
<td>1 (2.6)</td>
</tr>
<tr>
<td>Severely impaired (LVEF &lt; 0.45)</td>
<td>3 (7.9)</td>
</tr>
<tr>
<td>SVAS peak gradient (mean ± SD)</td>
<td></td>
</tr>
<tr>
<td>Echocardiography (mm Hg)</td>
<td>86.1 ± 28.7</td>
</tr>
<tr>
<td>Invasive (mm Hg)</td>
<td>66.5 ± 24.6</td>
</tr>
<tr>
<td>Hemodynamic parameters (mean±SD):</td>
<td></td>
</tr>
<tr>
<td>LVSOP (mm Hg)</td>
<td>171.3 ± 38.6</td>
</tr>
<tr>
<td>LVEDP (mm Hg)</td>
<td>14.4 ± 8.9</td>
</tr>
<tr>
<td>LAP (mm Hg)</td>
<td>6.9 ± 3.2</td>
</tr>
<tr>
<td>PA stenosis peak gradient* (mean±SD):</td>
<td></td>
</tr>
<tr>
<td>Echocardiography (mm Hg)</td>
<td>46.8 ± 27.1</td>
</tr>
<tr>
<td>Invasive RVSP (mm Hg)</td>
<td>50.1 ± 26.5</td>
</tr>
</tbody>
</table>

* Data calculated only from patients with pulmonary artery stenosis present.

Concomitant procedures included pulmonary artery enlargement (n = 3), transverse aortic arch reconstruction (n = 4), resection of subvalvular aortic obstruction (n = 2), aortic valve replacement (mechanical prosthesis n = 1, bioprosthesis n = 1), resection of a post-stenotic aneurysm (n = 1), and aortic valve commissurotomy (n = 1). Of the coronary stenoses observed in our cohort, 2 cases were mild and did not require intervention while 1 patient had multiple peripheral stenoses not amenable to surgical intervention.

Statistical Analysis

Time-related analysis of survival and freedom from reoperation or reintervention used the Kaplan-Meier method and curves were compared using the log-rank test. Continuous and categoric data were compared between groups using the Fisher exact test, Wilcoxon signed rank test, and the Kruskal-Wallis test where appropriate according to analyzed variables and the groups to be compared.

Results

Early Results

One early death occurred, resulting in an early mortality rate of 2.6%. This patient received an emergency operation for SVAS relief after experiencing cardiac arrest during preoperative catheterization. Another cardiac arrest occurred 12 hours postoperatively and resuscitation was eventually unsuccessful. Autopsy revealed extensive infarction of the interventricular septum in addition to disseminated myocardial hemorrhage.

In the survivors, median time of mechanical ventilation was 16 hours (range 7 to 244), median length of intensive care unit stay 2 days (range 1 to 11), and median length of hospital stay 9 days (range 5 to 25). The echocardiographic mean peak aortic gradient was significantly reduced by the operation from 86.1 ± 28.7 mm Hg to 22.3 ± 11.1 mm Hg (p < 0.001). No differences were observed between the surgical techniques used; at discharge the mean peak systolic gradient was 19.7 ± 9.0 mm Hg in the single-patch, 21.4 ± 11.1 mm Hg in the 2-sinus reconstruction, and 24.5 ± 12.0 mm Hg in the 3-sinus reconstruction group. In 5 patients residual obstructions with peak gradients 40 mm Hg or greater were present. Of
these, 2 occurred in the Doty group and 3 in the 3-sinus reconstruction group. Three patients were infants and 2 had diffuse SVAS. There was no statistical difference in respect to operation technique \( (p = 0.41) \) or SVAS morphology \( (p = 0.13) \). A trend toward a higher frequency of residual obstruction in the infant group did not reach statistical significance \( (3 \text{ of } 8 \text{ vs } 2 \text{ of } 29, p = 0.057) \). At discharge, good left ventricular function was documented by echocardiography in most patients; 2 patients \( (5.4\%) \) had mild and 3 \( (8.1\%) \) had moderate left ventricular dysfunction. No or trivial aortic valve incompetence was observed in 30 \( (81.1\%) \), mild in 6 \( (16.2\%) \), and moderate in 1 patient \( (2.7\%) \).

### Midterm and Late Results

Three late deaths occurred. One patient operated at 8 months of age died 1.5 months after autologous slide aortoplasty. A single coronary ostium was present in this patient and he had been discharged with a residual gradient of 40 mm Hg and mild left ventricular dysfunction. After readmission with deteriorating cardiac function eventually requiring extracorporeal membrane oxygenation (ECMO), diagnostic catheterization revealed multiple coronary stenoses not accessible for intervention. He could not be weaned from ECMO and died. The second patient died 5 years after Doty repair and additional resection of a post stenotic aneurysm performed at 3 months of age. After 2 successful reoperations for early recurrent obstruction 5 and 11 months postoperatively, the patient developed a progressive aortic valve stenosis and required valve replacement. Postoperatively she required ECMO support due to left ventricular dysfunction and died from massive cerebral hemorrhage. One patient died from a cerebral malignancy 18 years postoperatively without any cardiac events in the postoperative course; he was excluded from survival analysis. Overall survival estimates were 94% and 90% at 5 and 20 years, respectively \( (\text{Fig 1A}) \). The SVAS repair during infancy is associated with poorer survival rates \( (p = 0.038) \), while SVAS morphology and surgical technique have no significant influence on survival rates \( \text{diffuse SVAS} 88.4\%, \text{discrete SVAS} 100\% \text{ at 10 years, } p = 0.49; \text{McGoon} \ 100\%, \text{Doty} \ 89.8\%, \text{3-sinus reconstruction} 90.9\% \text{ at 10 years, } p = 0.86, \text{respectively}) \).

During follow-up, 9 reoperations were performed in 6 patients \( (17\%) \). Reoperations for recurrent obstruction of the supravalvular aorta were necessary in 4 patients between 5 months and 3.5 years postoperatively. Additional causes for reoperations included development of a coarctation \( (n = 1) \) and progressive aortic valve disease \( (n = 1) \). Freedom from reoperation in the entire cohort was 83% at 5 and 20 years \( (\text{Fig 1B}) \). Patients operated upon during infancy \( (\text{Fig. 1B}) \) and patients with diffuse SVAS \( (\text{Fig. 1C}) \) had lower freedom from reoperation rates \( (p = 0.024 \text{ and } p = 0.007, \text{respectively}) \) while rates did not differ between types of surgical repair \( (\text{McGoon} \ 66.7\%, \text{Doty} \ 89.8\%) \).

![Graphs](image.png)
A total of 11 catheter interventions were performed in 6 patients (17%) during follow-up. In 2 patients unsuccessful dilatation of the re-obstructed supravalvular aorta was performed; both patients were successfully reoperated on. Additional interventions included stent implantation with subsequent re-dilatations due to development of aortic coarctation (n = 3), aortic valve balloon valvuloplasty (n = 1), stent implantations and re-dilatations of aortic arch vessels (n = 1), and dilatation of a supravalvular pulmonary stenosis (n = 1). Overall freedom from catheter intervention was 84% and 79% at 5 and 20 years, respectively. It differed significantly in patients operated on in infancy (53.6% vs 84.5% at 10 years, p = 0.014) and in patients with diffuse SVAS (20.0% vs 92.6% at 10 years, p = 0.001) while type of surgery did not have a significant influence (McGoon 66.7%, Doty 77.8%, 3-sinus reconstruction group 88.9% at 10 years, p = 0.8).

At the last examination, most surviving patients had unlimited functional capacity corresponding to New York Heart Association (NYHA) functional class I (n = 31, 91.1%) while three (8.9%) had mild incapacities and were in NYHA class II. Left ventricular function was good in 30 (88.2%), mildly impaired in three (8.9%) and moderately impaired in one patient (2.9%). Mean peak systolic aortic gradient including patients reoperated on was 20.3 ± 10.0 mm Hg. It was 18.0 ± 3.5 mm Hg after single-patch repair, 18.7 ± 9.2 mm Hg after 2-sinus reconstruction and 22.8 ± 12.0 mm Hg after 3-sinus reconstruction (Fig 2). A residual gradient 40 mm Hg or greater was present in 1 patient after Doty repair and in 2 patients after 3-sinus reconstruction (Brom n = 1, Souza/Myers n = 1).

Aortic valve function was well preserved in most patients and regurgitation generally did not show significant progression during follow-up. At the last examination, no or trace regurgitation was present in 23 (65.7%), mild in 11 (31.4%), and moderate in 1 (2.9%) patient. One patient who had received a bioprosthetic valve replacement concomitantly with SVAS repair developed a moderate stenosis currently not requiring reoperation. Two patients developing severe stenosis required reoperation. In the remaining patients, no relevant aortic valve stenosis occurred during follow-up.

**Comment**

We reviewed our institutional results with different surgical techniques for the repair of congenital SVAS during a 25-year period. Our study demonstrates low operative and late mortality as well as low reoperation rates for all surgical procedures applied.

Several surgical techniques for the treatment of SVAS have been described (reviewed in references [13] and [14]) including the single-patch augmentation by McGoon and colleagues [3], the inverted bifurcated-patch aortoplasty by Doty and colleagues [4], 3-sinus augmentation by Brom [5], and autologous slide aortoplasty by Souza and colleagues [11] and Myers and colleagues [12]. Presently, there is no consensus about the optimal technique for the treatment of SVAS. While some studies argue that symmetric 3-sinus repair results in improved long-term outcome with reduced risk of reoperation, most authors consistently conclude that clear superiority of one technique over another has hitherto not been demonstrated [7, 8, 10, 14]. The primary surgical goal, relief of the supravalvular obstruction, can be adequately achieved with all approaches. The main theoretic advantage of a symmetric 3-sinus reconstruction is that a more natural geometry of the distorted aortic root can be restored, which might result in improved preservation of aortic valve function [6].

The rarity of this lesion limits the individual institution’s experience and accordingly only few studies with large cohorts of patients exist; most comparative studies comprise a limited number of patients and an unbalanced distribution of the surgical techniques. Two reports with large cohorts which almost exclusively employed the single-patch technique conclude that the single-patch enlargement sufficiently relieves the supravalvular obstruction and provides good long-term results without symmetric restoration of aortic root geometry [15, 16]. One of the few studies with both a large number of patients and a relatively even distribution of different surgical techniques compares the single-patch repair with a combined group with 2- and 3-sinus reconstruction [17]. Multiple-sinus reconstruction resulted in lower mean pressure gradients during follow-up. Additionally, it was associated with lower mortality and reoperation rates; however, these differences were not statistically significant in time-related analysis. Two studies with smaller cohorts compared Brom’s technique with 1- and 2-sinus reconstruction [7, 8]. One of them reported no significant differences in terms of survival and reoperations rates; with no late deaths and only 1 reoperation, respective differences
between the techniques were also not evident in the second study. However, a lower incidence of residual obstructions and aortic valve insufficiency after Brom repair led the authors to argue that it may provide a superior long-term outcome [8]. Various additional reports with smaller patient numbers comparing different techniques of SVAS repair did not find any particular technique to provide a significantly improved outcome [9, 10, 18].

In the present study, the surgical techniques used for SVAS repair did not have a significant influence on the results in terms of survival rates, freedom of reoperation rates, peak supravalvular gradient, frequency of residual obstruction, and aortic valve function. It has to be mentioned, though, that single-patch repair is under-represented in our cohort and results concerning this group have to be viewed with caution. In addition, the follow-up period is shorter in the 3-sinus reconstruction group because these techniques were only introduced more recently at our institution. However, equally good mid-term results were obtained by 2- and 3-sinus reconstruction techniques. Our findings indicate rather that the necessity of operation in infancy, as well as a diffuse SVAS morphology, are associated with higher reoperation rates and, more importantly, that surgery in infants is associated with higher mortality rates. We agree with Mitchell and Goldberg [14] that the influence of age on the results of SVAS repair is still underappreciated in most studies. Only 2 of the more recent studies analyzed survival rates stratified for age at all. One of them reports a reduced survival rate for children operated on at less than 2 years of age, while the other found a reduced event-free survival rate for this age group [16, 19]. These results are consistent with our findings in infants. Most of the recent studies did not specifically analyze the outcome of SVAS repair in infants but from the few reports that describe their mortality in detail, it is striking that cases of early death often affect small children [8–10]. The higher risk for unfavorable outcome may be owing to the fact that surgery in this age group is usually performed only in symptomatic children with more severe forms of SVAS.

We believe that no particular technique is optimal for all cases of SVAS and that the choice for any method has to be made with careful consideration of the individual patient’s characteristics. As Doty himself put it: “I have always thought that more symmetric and anatomically correct methods of repair of the aortic root would not only provide better early results relieving obstructing gradients, but also allow the aortic valve to last longer. It is likely, however, that even the best efforts may not be enough and most, if not all, patients with repair of supravalvular aortic stenosis will eventually require reoperation.” [20]. Autologous slide aortoplasty has the theoretic advantage of improved preservation of growth potential by avoiding prosthetic material and can be performed with good results [10–12, 17, 21]. However, it offers no advantages for diffuse forms of SVAS because long-segment supravalvular narrowing will not be augmented. Even in discrete SVAS, we and others found additional patch enlargement of the ascending aorta to be necessary in some cases [10, 17]. This seems to relativize the putative advantage of preserved growth potential offered by this technique. Brom’s technique is also performed with good results and can easily be modified for adequate repair of diffuse forms of SVAS [7–9, 22]. However, we conclude from our results that the Doty procedure is equally capable of effectively and sustainably reducing the supravalvular obstruction and preserving the aortic valve function also in small children and with diffuse SVAS.

Limitations

Limitations of this study are inherent to its retrospective design, for which consistency of follow-up examinations and data collection are not given. The study cohort is relatively small, which emphasizes the rarity of this congenital malformation and constitutes a general difficulty in comparing different surgical approaches to its repair. Partially owing to the small cohort size, the surgical techniques employed are unevenly distributed and this limits the possibilities of drawing confident conclusions and might impede the recognition of potential risk factors of adverse outcome associated with a particular technique.

Conclusions

This study demonstrates that equally good early and midterm results after SVAS repair can be achieved with 2- and 3-sinus reconstruction methods. Our data do not support the proposed superiority of symmetric 3-sinus reconstruction; however, further experience with longer follow-up is necessary to draw more confident conclusions. Additionally, we demonstrate that operation in infancy is associated with lower rates of survival and freedom from reoperation.

References