Subaortic obstruction in univentricular heart: results using the double barrel Damus–Kaye Stansel operation

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Abstract

Objective: We review our experience with relief of subaortic obstruction in univentricular hearts following pulmonary artery banding (PAB) with double barrel Damus–Kaye Stansel procedure (DKS) and subsequent staged palliation to Fontan. The purpose was to determine if PAB alters semilunar valve function after the double barrel DKS procedure and if this staged approach negatively influences the achievement of Fontan palliation.

Methods: From January 1990 to March 2006, 27 patients underwent PAB (mean 22 days, range 1–150 days; 3.4 kg) and coarctation as corrected simultaneously in 18 (18/27) 66%. These 27 patients subsequently had PA debanding and double barrel DKS connection at a mean age of 10.2 months (range 0.3–58 months). Pulmonary flow was established with a bidirectional Glenn in 14 patients; modified Blalock in 6, Glenn with modified Blalock in 5 and completion Fontan in 2 patients.

Results: There were six early deaths (22%) following DKS: four patients receiving DKS with systemic shunt and two receiving bidirectional Glenn and systemic shunt. Patients receiving DKS with bidirectional Glenn shunt had a significantly lower mortality than patients who had a DKS with systemic shunt alone or in combination with a Glenn (p < 0.03). Single ventricle to aortic gradient was reduced from 27.5 ± 18 mmHg to 3.4 ± 2 mmHg following double barrel DKS procedure (p < 0.001). Aortic and pulmonary insufficiency was trace to mild in all patients. Nineteen of 21 survivors (90%) have completed Fontan with no early and three late deaths. Two patients are completion Fontan candidates. Conclusions: PAB (±coarctation repair) with interval double barrel DKS is effective palliation for univentricular heart and excessive pulmonary blood flow. PAB does not create significant pulmonary insufficiency and subsequent DKS effectively relieves single ventricle to aortic gradient. Optimal second stage pulmonary blood flow is usually established with a bidirectional Glenn. The need for a Blalock shunt or a Glenn plus a Blalock is associated with increased mortality.

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1. Introduction

A challenging subset of patients born with univentricular heart can have unrestricted pulmonary blood flow in the presence of distal aortic arch obstruction and a normal ascending aorta connected to a rudimentary subaortic outlet chamber. The only pathway for blood flow to enter the systemic outflow chamber is through a ventricular septal defect (VSD). Systemic ventricular outflow obstruction is caused by a restrictive VSD. When the VSD is small at birth, aortic hypoplasia, coarctation of the aorta or interrupted aortic arch may be present. Even when the VSD is nonrestrictive at birth, it may narrow over time and subaortic obstruction then becomes apparent [1].

Palliative treatment strategy for this patient group has evolved with the common goal to limit pulmonary blood flow, provide a pulmonary artery to aortic anastomosis to address future VSD narrowing and repair of the hypoplastic aortic arch. One approach consists of pulmonary artery banding (PAB) and if necessary, distal aortic arch reconstruction in the newborn period. At a later time, the band is removed and the aorta to pulmonary artery anastomosis (Damus–Kaye Stansel, DKS) performed to relieve the systemic outflow obstruction secondary to VSD narrowing in the presence of a pulmonary artery band. Pulmonary blood flow can be established with a systemic to pulmonary artery shunt or a bidirectional superior cavopulmonary connection (Glenn) [2,3].

The DKS connection can be performed using one of two techniques. The end of the divided proximal main pulmonary artery can be anastomosed to the side of the ascending aorta; alternatively, both great vessels can be completely divided and their adjacent wall joined. The resultant 'double barrel'
can be anastomosed to the distal aorta directly or by employing patch augmentation.

The goals of this 15-year analysis were to determine if PAB alters semilunar valve function after the double barrel DKS procedure and if this staged approach negatively influences the ability to achieve a successful completion Fontan.

2. Materials and methods

We retrospectively reviewed the medical and surgical records of 108 consecutive patients with functional single ventricle associated with unrestricted pulmonary blood flow that underwent PAB at Riley Hospital for Children, Indianapolis, IN and Cardinal Glennon Children’s Hospital, St. Louis, MO between January 1990 and March 2006. Among this cohort, 27 patients (25%) required debanding and a DKS procedure to relieve outflow obstruction from the single ventricle and constitute the study population. (Indiana University, 17 patients; St. Louis University, 10 patients) Pulmonary artery banding (±coarctation repair) was the initial palliation in all cases. No patient had undergone a previous operation. All patients underwent ligation or division of a patent ductus arteriosus at the time of PAB and coarctation repair if required.

This study was approved by the institutional review board at St. Louis University and Indiana University. The need for individual consent was waived.

2.1. Demographics

The mean age at the time of surgery (PAB ± coarctation repair and PDA ligation) was 22 days (range, 1—150 days; 20 male; 7 female). The specific anatomic lesions are listed in Table 1. The majority of patients had situs solitus, double inlet left ventricle with L-transposition of the great arteries. Five patients had unbalanced atroventricular canal with hypoplastic left ventricle and four patients had tricuspid atresia with D-transposition of the great arteries. Three patients with a VSD, mitral stenosis and coarctation were strategized to univentricular connection. The distal aorta was sewn to the posterior of the aortic root. One third of the newly joined great arteries. Anteriorly, the aorta and unobstructed pulmonary blood flow who required the Damus—Kaye Stansel connection.

2.2. Operative procedures

The technique of PAB has been described previously [4]. Extracorporeal support was used in one patient at the time of interrupted arch repair and PAB band placement. The PAB was assessed postoperatively by clinical hemodynamic status and transthoracic echocardiography. The PAB was defined as inadequate whenever there was persistence of refractory congestive cardiac failure (evaluated clinically), and/or ineffective protection of the pulmonary circulation (systolic pulmonary arterial pressure greater than 45 mmHg at follow-up). The mean interval from pulmonary artery band placement to DKS procedure was 10.2 months (range 0.3—58 months).

Coarctation repair was performed employing subclavian flap aortoplasty or resection with end-to-end anastomosis. Interrupted aortic arch was repaired using a left carotid artery swing down or direct anastomosis between the ascending and descending aorta.

The diagnosis of systemic outflow obstruction after adequate arch reconstruction was made whenever the size of the VSD (bulboventricular foramen) was less than half the diameter of the aortic valve annulus and/or the peak instantaneous gradient from the dominant ventricle to the ascending aorta was greater than 20 mmHg [5].

The double barrel DKS procedure was performed to relieve intracardiac systemic outflow obstruction from the single ventricle using conventional continuous flow cardiopulmonary bypass with antegrade and retrograde cardioplegic arrest. The innominate vein was routinely cannulated if a bidirectional superior caval pulmonary connection was planned. The aorta and main pulmonary artery were divided at the level of the PAB. The band and adjacent scar tissue were excised. The two facing sinuses of both great vessels were sewn together retaining a circular shape to avoid semilunar valve leaflet and coronary artery distortion. The distal aorta was sewn to the posterior of one-third of the newly joined great arteries. Anteriorly, the remaining defect was closed with a patch of homograft, glutaraldehyde treated autologous pericardium or Gore-Tex (Fig. 1) [6].

The choice of pulmonary blood flow at the time of DKS was based on the chronology of the operation in our series. Prior to the year 2000, patients receiving the DKS connection had a systemic to pulmonary artery shunt, while patients operated upon after 2000 had a bidirectional Glenn anastomosis.

The bidirectional Glenn, modified Blalock shunt, lateral tunnel and extracardiac Fontan operations were performed according to established techniques. Pulmonary artery patch reconstruction was performed using fresh autologous pericardium or pulmonary homograft.

The degree of pulmonary and aortic valve insufficiency was quantitated as follows: none or trace, 0; mild, 1+; moderate, 2+; severe, 3+. Pulmonary regurgitation was classified as mild if there was no retrograde diastolic flow in the pulmonary trunk and less than 1 cm regurgitant jet in the right ventricular outflow tract; moderate if retrograde diastolic flow was detected in the main PA with 1—2 cm regurgitant jet in the right ventricular outflow tract and severe if additional retrograde diastolic flow was detected in

<table>
<thead>
<tr>
<th>Anatomic lesion</th>
<th>Number of patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Double inlet LV</td>
<td>12 (44)</td>
</tr>
<tr>
<td>Unbalanced AVC; hypoplastic LV</td>
<td>5 (18)</td>
</tr>
<tr>
<td>Tricuspid atresia with TGA</td>
<td>4 (15)</td>
</tr>
<tr>
<td>Mitral stenosis, VSD, hypoplastic LV</td>
<td>3 (11)</td>
</tr>
<tr>
<td>DORV: mitral atresia</td>
<td>2 (7)</td>
</tr>
<tr>
<td>Heterotaxy; dextrocardia; single ventricle</td>
<td>1 (4)</td>
</tr>
</tbody>
</table>

AVC, atrioventricular canal; DORV, double outlet right ventricle; LV, left ventricle; GA, transposition of the great arteries; VSD, ventricular septal defect.
the branch pulmonary arteries with greater than 2 cm jet in the right ventricular outflow tract. The peak systolic gradient across the semilunar valve was measured using the modified Bernoulli equation.

2.3. Repair of associated defects

At the time of PAB, 15 patients (55%) underwent repair of aortic coarctation employing the subclavian flap aortoplasty.
or resection with end-to-end anastomosis. Three patients presented with interrupted aortic arch type B. Two underwent left carotid artery turndown, while one patient had direct anastomosis between the ascending and descending aorta. At the time of the DKS operation, the most common associated procedures performed were atrial septectomy in 17 patients (17/27; 63%), patch augmentation of the branch pulmonary arteries in 15 patients (15/27, 55%) and patch augmentation of a hypoplastic transverse aortic arch in two patients (2/27, 7.4%).

3. Follow-up

Follow-up was complete in all patients with a mean of 71 months (range 12—157 months) from PAB to last patient contact. The mean follow-up from the DKS procedure to the latest patient contact was 60 months (range 9—141 months). Data were obtained in all patients from outpatient records and by communication with primary care physicians.

3.1. Statistical analysis

Measured and calculated data are expressed as mean ± SD. Comparison between groups was performed using Wilcoxon sign rank test or analysis of variance as indicated. The degree of semilunar valve insufficiency was compared using binary logistic regression analysis.

Early mortality was defined as death during initial hospitalization or within 30 days of operation. All other deaths were defined as late mortality. A p value of less than 0.05 was considered significant. Specific statistical software SPSS for Windows version 10 (SPSS, Inc., Chicago, IL) was used for data analysis.

4. Results

4.1. Surgical strategies and clinical outcomes

The DKS procedure was performed at a mean of 10.2 months (range 0.3—58 months) following PAB. In 14 patients (14/27; 52%) a superior cavopulmonary connection was the only source of pulmonary artery blood flow. Two patients had DKS, Glenn and completion lateral tunnel Fontan procedures performed concurrently.

Six patients early in the series (6/27; 22%) underwent a DKS with a systemic to pulmonary artery shunt as the only source of pulmonary blood flow.

The remaining five patients (5/27; 18.5%) initially had a bidirectional Glenn performed at the time of the DKS procedure. After separation from cardiopulmonary bypass and completion of modified ultrafiltration, the arterial PO2 was less than 30 mmHg and the systemic saturation was less than 55% on 100% oxygen. For this reason, a 3.5 mm (one patient), 4 mm (two patients), or 5 mm (two patients) Gore-Tex graft was interposed centrally from the ascending aorta to the left or right pulmonary artery.

A subsequent completion Fontan has been performed in 19 of 21 hospital survivors (90%) at a mean of 28 months (range 0—55 months). A fenestrated lateral tunnel was performed in nine patients and a non-fenestrated extracardiac conduit in 10.

Two patients are currently awaiting completion Fontan. The time interval from the DKS procedure to the latest follow-up is 15.2 months. Echocardiographic analysis demonstrated a peak single ventricle to aortic gradient of 4.4 mmHg. The degree of pulmonary and aortic valve insufficiency ranged from trace to mild (aortic valve, 0.60; pulmonary valve, 0.20). The remaining two patients are Fontan candidates.

5. Mortality

5.1. Early mortality

There were a total of 17 deaths (17/108, 16%) with eight early deaths occurring after the initial PAB ± arch reconstruction and nine late deaths before definitive repair in infants who presented with single ventricle and unrestricive pulmonary blood flow. The DKS procedure resulted in six early deaths (6/27; 22%). Two patients required a Blalock shunt to supplement their bidirectional Glenn. They expired at 7 and 21 days postoperatively from a cerebral hemorrhage post-ECMO placement and low cardiac output with multi-organ system failure, respectively. The four remaining early deaths following DKS procedure occurred suddenly and unexpectedly post-extubation at 4, 7, 8 and 9 days postoperatively. In each case, the sole source of pulmonary blood flow was a 4 or 5 mm modified Blalock-Taussig shunt. In three of these patients, the cardiac arrest followed nasotracheal suctioning. The remaining sudden death was secondary to bleeding resulting in cardiac tamponade.

The early mortality of those patients receiving a DKS connection with a Blalock shunt alone or in conjunction with a bidirectional Glenn was significantly higher than patients who underwent a DKS with a bidirectional Glenn shunt as their sole source of pulmonary blood flow (p < 0.03).

5.2. Late mortality

There were three late deaths following the Fontan procedure (3/19; 16%). In two patients operated upon early in our series, the DKS, Glenn and completion Fontan (one each, fenestrated and non-fenestrated) were performed concurrently at one operative sitting 21 and 32 months following PAB. Both patients expired at 2.5 and 30 months from respiratory failure and low cardiac output. The remaining patient underwent a non-fenestrated completion Fontan at 23 months. Late death occurred at 4 months secondary to low cardiac output and multi-organ system failure.

5.3. Hemodynamic analysis

Cardiac catheterization was performed prior to the DKS in 27 patients and in 19 patients prior to completion Fontan. The results are summarized in Table 2. The DKS procedure significantly reduced the gradient across the single ventricle and in the main pulmonary artery at the time of the Fontan operation. The single ventricle end diastolic pressure, an
indirect measure of ventricular function, was at the lower limit of normal.

The hemodynamic parameters in the six non-survivors of DKS procedure were not significantly different from the 21 survivors (non-survivors; SV-AO gradient, 33.8 ± 19; RA, 7.4 ± 3.1; PA 17.4 ± 6; EDP, 10.2 ± 4; SAO₂, 79.5 ± 3.0).

No significant differences were noted when the pre-Fontan hemodynamics in those patients having the DKS operation with bidirectional Glenn were compared to those patients receiving the DKS procedure with a systemic shunt alone or in combination with a bidirectional Glenn.

5.4. Echocardiographic analysis

Echocardiographic analysis was performed in 27 patients prior to the DKS operation and in 21 patients at latest follow-up (Table 3).

The pulmonary artery band was of adequate tightness as judged by a peak gradient of 93 mmHg. The gradient from the single ventricle to the ascending aorta derived echocardiographically was similar to that observed at cardiac catheterization (31.0 mmHg vs 27.5 mmHg) and remained significantly reduced at the time of last follow-up.

Prior to the DKS operation, the presence of a PAB was associated with trace to mild degrees of aortic and pulmonary valve insufficiency. At follow-up, the degree of aortic insufficiency rose slightly, but no change in pulmonary insufficiency was noted. The differences were not significant. The echocardiographic parameters in the six non-survivors of DKS were not significantly different from the 21 survivors (non-survivors PAB gradient, 109 ± 21; SV-AO gradient, 42 ± 16; AI, 0.33 ± 0.2; PI, 0.33 ± 0.23 ± 0.1).

6. Discussion

In this report PAB was used to control excessive pulmonary blood flow in 108 patients born with univentricular heart. In the majority of patients, the systemic outflow traversed a bulboventricular foramen to reach the aorta. In 27 patients (25%) narrowing at this site occurred following PAB causing single ventricle systemic outflow obstruction, which was effectively relieved with the double barrel DKS procedure. Our intermediate follow-up of 71 months demonstrated that debanded patients receiving the double barrel DKS operation did not develop significant semilunar valve dysfunction and 19 of 21 hospital survivors to date (90%) achieved a Fontan operation with no early deaths. The two remaining patients in this series awaiting Fontan have been followed for a mean of 15 months. The pressure gradient across the DKS anastomosis has remained low (mean 4.42 mmHg) and no patient has more than trivial to mild semilunar valve insufficiency. Two late deaths occurred early in the series in patients having the DKS, Glenn and lateral tunnel Fontan concurrently. Experience with this approach is limited and has been associated with increased Fontan mortality [7]. We recommend an interval of at least 12—15 months following DKS and superior caval pulmonary anastomosis to allow single ventricle hypertrophy to resolve and diastolic compliance to improve prior to performing the completion Fontan.

The optimal timing of debanding is unknown, but in order to avoid prolonged ventricular hypertrophy and concomitant diastolic dysfunction, our current practice is to proceed with DKS and band removal between 3 and 6 months of age.

Based on our current experience, we favor a bidirectional Glenn shunt as the source of pulmonary blood flow with the DKS procedure. In the first 5 years of this series, four of six early deaths following the DKS occurred in patients who received either a 4 or 5 mm systemic shunt. The DKS operation with a modified BT shunt is physiologically similar to a modified Norwood procedure. In this earlier era, we were not cognizant of the negative clinical effects that larger systemic shunts may have on lowering single ventricle diastolic blood pressure, coronary insufficiency and systemic under perfusion. Important postoperative management strategies that can help prevent sudden unexplained death in this setting such as continuous mixed venous oxygen saturation monitoring, serial lactate measurements, use of alpha blockade, Milrinone and early paralysis with delayed sternal closure were not fully appreciated at that time. Furthermore, Jaquiss and associates have recently demonstrated that a bidirectional Glenn shunt can be performed successfully as early as three months of age [8]. In the unlikely event that the superior caval pulmonary anastomosis does not provide adequate pulmonary blood flow in this setting, we prefer Glenn takedown and placement of a small (3.5 mm) systemic to pulmonary artery shunt. Currently in this report, higher mean pulmonary artery pressure secondary to inadequate band tightness may have accounted for the additional need of a systemic shunt with the Glenn connection. The use of nitric oxide or Sildenafil may prevent the need for an additional shunt.

The improved results with the bidirectional Glenn shunt performed as early as 3 months of age and the use of
postoperative management strategy as described above has changed our rationale to use a smaller shunt when needed and to deband these patients as early as 3–4 months of age for the DKS with superior cavo pulmonary Anastomosis.

The technique of performing the DKS Anastomosis does affect long-term competency of the semilunar valves. Chen and associates observed that mild pulmonary regurgitation was recognized in one-fourth of their patients having the end-to-side DKS connection. More concerning was their observation that this frequency of regurgitation increased in patients long after the DKS operation [9].

Daenen and associates reported that the double barrel DKS Anastomosis performed in nine patients was superior to the end-to-side connection (four patients) because it avoids unbalanced traction on the sinotubular junction of both great vessels and decreases the risk of obstruction and/or regurgitation [10].

More recently, Fuji and associates compared the double barrel with the end to side DKS in 39 patients. After a mean follow-up of 68 months, the end-to-side DKS demonstrated greater pulmonary regurgitation than the double barrel technique (4/12 vs 1/27; \( p < 0.03 \)). No DKS stenosis was observed in either group [11].

Our results support these observations. The added advantage of the double barrel DKS is that it can be constructed in any morphologic arrangement of the great vessels. In this report, the use of PAB (right arch reconstruction) followed by double barrel DKS and Glenn Anastomosis does not deter patients from achieving successful Fontan palliation. This has been confirmed by Clark and associates who recently reported their results in 25 patients with double inlet left ventricle who underwent aortic arch palliation. This has been confirmed by Clark and associates who recently reported their results in 25 patients with double inlet left ventricle who underwent aortic arch repair, PAB and delayed DKS procedure [12]. All patients, received the double barrel DKS with an early and late mortality of 4% and 8%, respectively. Fontan completion was achieved in 20 of their 22 survivors. There were no cases of DKS obstruction or semilunar valve insufficiency greater than mild.

The Norwood operation is an alternative approach that is commonly employed in this patient population. The DKS connection, arch obstruction and unrestrictive interatrial communication can be addressed at one operation and a controlled source of pulmonary blood flow established. We believe this is the procedure of choice in neonates with classic hypoplastic left heart syndrome. However, in the subset of patients described in this report, we favor initial PAB and delayed DKS connection especially if the patient is believed to be a high risk Norwood candidate (e.g. chromosomal anomaly, co-existing non-cardiac malformation or birth weight below 2.5 kg).

If the DKS operation is chosen to relieve subaortic obstruction in patients with univentricular heart, then the double barrel technique should be employed. The procedure is readily reproducible, can be applied to any great vessel relationship and adequately preserves long-term semilunar valve function. The bidirectional Glenn shunt as the source of pulmonary blood flow is associated with lower mortality.

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References