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**THE EXPERIENCE OF SUBAORTIC OBSTRUCTION REPAIR CONCOMITANTLY WITH TOTAL CAVOPULMONARY CONNECTION**

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Systemic ventricle outflow tract obstruction (SVOTO) in single ventricle patients could result in unfavorable hemodynamic conditions that might worsen the prognosis of patients with total cavopulmonary connection (TCPC).

From 2005 to 2015, 92 consecutive children with variable single ventricle pathologies underwent TCPC procedure, 10, 8% (n=10) of which was presented with SVOTO. We used two main options for surgical relief of SVOTO: the first consisted in ventricular septal defect enlargement, the second one - in modified Damus-Kaye-Stansel procedure (DKS). The patients was divided into three main groups: group 1 - 82 (89.1%) patients who underwent TCPC without performing any procedure on SVOT; group 2 - 6 (6.4%) in which the patients treated with TCPC simultaneously with modified DKS procedure (double-barrel); group 3 - 4 (4.4%) patients who underwent the restrictive VSD resection at the moment of TCPC. There was 1(1, 2%) early death in the control group and no mortality in study groups throughout the study period. Two patients in study groups underwent a permanent pacemaker implantation. The SVOT gradient decreased from 17 ± 5, 82 mm Hg preoperatively to 8 ± 4, 33 mm Hg after the procedure.

Our results indicate that the SVOTO relief at the concomitantly with TCPC does not cause significant increase in morbidity and mortality at early postoperative period. The results suggest that the DKS is the method of choice with regard to avoidance of heart block or adequate removal of SVOTO.

***Keywords:*** *total cavopulmonary connection, subaortic obstruction, single ventricle, the Damus-Kaye-Stansel procedure.*

Given the impending negative effects of SVOTO on hemodynamic conditions and future outcome of patients with SV it may poses a serious problem and is particularly important in patients with SV hemodynamic. Elevated systemic ventricle afterload leads to a ventricular hypertrophy and to increase the left atrial pressure that creates additional resistance to pulmonary blood flow [1].

Even a mild SVOTO leads to rapid decompensation and reduces contractility of the SV and subsequently produce some kinds of rhythm disturbances [2]. Therefore, SVOTO in patients with SV creates adverse hemodynamic conditions, and thereby worsens the prognosis of patient with TCPC.

The SVOTO in SV patients may occur as a primary lesion, but more often it has seen after the pulmonary artery banding (PAB) [3]. PAB produces myocardial hypertrophy with resultant decrease in VSD/bulboventricular foramen size. Limitations of pulmonary blood flow decrease the systemic ventricle linear dimensions that might further decrease VSD/bulboventricular foramen (BVF) size and accelerate the rate of SVOTO development. And subsequently the risk of SVOTO might appear late afterTCPC, especially in the presence of morphological substrate [2,4].

Therefore, besides the classic selection criteria, we must consider the obstruction factors and if available to take effective measures to eliminate it [1,2,4].

There are several single ventricle anomalies which associated with the presence of morphologic substrate to cause SVOTO: it may be persistent subaortic conus, restrictive VSD / BVF, and abnormal attachment of chords (subvalvar structures) that cross the outflow tract.  
Systemic obstruction is more likely to develop in two distinct patient groups: (1) with double inlet left ventricle (DILV) or tricuspid atresia (TA) with ventriculoarterial discordancy and (2) complex forms of double outlet right ventricle (complex DORV) [1, 2].

Several options are available to surgical relief of SVOTO. The first one include the VSD/bulboventricular foramen enlargement - associated with a high risk of AV-block, and the second one - the Damus-Kaye-Stansel operation (DKS), which consist of creation an additional pathway into the aorta through the pulmonary valve.

The DKS operation was originally described for the treatment of patients with d-transposition of the great arteries and performed by anastomosing the arteries end-to-side. Subsequently, there have been reports of successful repair of patients with SVOTO utilizing modified DKS operation with great vessels anastomosed side-by-side, so called double barrel technique. The double barrel DKS is very effective in minimizing SVOTO risk, with sustainable good semilunar valves function.

**Materials and methods**

From 2005 to 2015, 92 consecutive children with variable single ventricle pathologies underwent TCPC procedure at UCCC. In 10 (10.8%) of them at the time of TCPC revealed the SVOTO. Median age at the time of procedure was 10,4 ± 4,58 months (range 3.8 months–28 years) and the median weight was 24,3 ± 13,6 kg (range 19,8–33,3kg).

To analyze the results of hemodynamic correction, patients were divided into 3 groups depending on method of eliminating systemic obstruction:

Group 1 - 82 (89.1%) patients who underwent TCPC without performing any procedure on SVOT;

Group 2 - 6 (6.4%) in which the patients treated with TCPC simultaneously with modified DKS procedure (double-barrel);

Group 3 - 4 (4.4%) patients who underwent the restrictive VSD resection at the moment of TCPC.

**Table 1.**

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| Groups | N | Аge (mon.) | Weight (kg) | CPB time (min.) | Ао cross-clamp time (min.) |
| Group 1 | 82 | 128,8± 55,84 | 33,3±14,62 | 92±48,45 | 74,0±28,45 |
| Group 2 | 6 | 75,2± 50,53 | 19,8±15,24 | 149,6±50,1 | 77,2± 11,30 |
| Group 3 | 4 | 104,5±49,9 | 22,1±10,8 | 160,0± 46,03 | 65,0± 25,01 |

Below are described the anatomical characteristics of children with single ventricle who underwent SVOTO repair concomitant with TCPC at our institution:

DILV – 6 patients; corrected transposition of the great arteries (CC-TGA) with morphological tricuspid valve atresia (TA) – 1; mitral valve atresia (MA) - 4, a complex form of double outlet right ventricle (complex DORV) remote VSD – 1 patient.

Among patients who enforced the removal of CO, prior palliative procedures include banding of the pulmonary artery - in 9 cases, systemic-pulmonary anastomosis - 1, bidirectional cavopulmonary anastomosis Glenn (BDCPA) - 10, the coarctation of the aorta repair - 2, rebanding pulmonary artery (re PAB) - in the 4 pulmonary valve function in this case was not broken. Data catheterization of heart cavities are presented in Table 2 (averages).

**Table 2. The catheterization data**

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| Groups | N | PA pressure (mm Hg) | Pulmonary vascular resistance (mmHg) | SV end-diastolic pressure (mm Hg) |
| Group 1 | 82 | 17±5,21 | 1,9±0,92 | 17±5,14 |
| Group 2 | 6 | 18±4,42 | 2,1±0,73 | 16±4,12 |
| Group 3 | 4 | 14±3,32 | 1,0±0,65 | 15±3,21 |

Indications for surgical repair of SVOTO was the presence of the pressure gradient between the SV and ascending aorta (> 10mmHg), the presence of anatomical substrate for the development of SVOTO (above mentioned anatomical diagnoses) and reducing bulboventricular foramen area index at time of initial presentation (<2sm2 / m2 or less than the diameter of the aortic valve ). The latter index is not always possible to exactly set due to the complexity of most forms of SV. The degree of SVOT on preoperative stage was assessed by echocardiography (ECHO) and by cardiac catheterization (Cath) (Table 3).

In first group, there was not carried out any surgical SVOTO relief procedure because at that time of TCPC the pressure gradient between the systemic ventricle and the aorta was absent.

The TCPC was performed in its extracardiac modification to all of patients. There were implanted extracardiac conduit of polytetrafluoroethylene (Gore-Tex®), the diameter of which is chosen depending on the diameter of the inferior vena cava, patient body weight was between 18 to 22 mm.

The TCPC was performed using conventional cardiopulmonary bypass methods with moderate hypothermia (26–28°C). Mean cardiopulmonary bypass and ischaemic times were 284 ± 6,4 and 75 ± 6,4 min., respectively.

**Results**

We assessed the short- and mid-term clinical (max 11±3, 03 mo.) outcomes with a focus on the evolution of SVOT haemodynamics with time (Table 3).

**Table 3. The evolution of SVOT haemodynamics**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| Groups | Δp SVOT1  before surgical treatment (Cath2) | Δp SVOT  before surgical treatment (ECHO3) | Δp SVOT immediately after surgical treatment  (ТЕЕ4) | Δp SVOT  before discharge (ECHO3) | Δp SVOT  the last survey (ECHO3) | The average time after (mon.) |
| Group 1 | 18±7,92 | 16 ±5,12 | 6,5 ±1,97 | 9,16 ±1,72 | 9,83 ±0,98 | 11 ±3,03 |
| Group 2 | 13±7,32 | 11±4,57 | 9,3 ±0,57 | 9,6 ±0,58 | 12,3 ±2,51 | 8 ±2,80 |

ΔpSVOT1 – the pressure gradient at systemic ventricle outflow tract, mm Hg ; Cath2- data of catheterization; ECHO3- echocardiography data; ТЕЕ4 – transesophageal echocardiography data

Over the study period there was one early death and no late death in group 1(1.2% ), due to coagulopathy and acute cerebrovascular accident. There was no one early and late death in the 2nd and 3rd groups.

Two patients required permanent pacemaker insertion postoperatively. One of them who underwent concomitant enlargement of the VSD/BVF had immediate heart block while the other patient developed sinus node dysfunction following TCPC and DKS.

No case of cerebrovascular accident was noted. One patient (16.7%) of 2nd group postoperatively develops the phrenic nerve paresis. The patient was subsequently underwent the diaphragm plication. One patients of 3rd group has prolonged pleural effusion of hilous nature which eliminated conservatively.

We have founded that inotropic support for more than 72 hours require 14 (17%) patients in group 1, 3 (50%) patients in the second group and 1 (25%) patients - in the 3rd group. All patients are extubated during the first 6 hours. The mean duration of pleural effusion, ICU stay and the length of stay in the hospital are shown in Table 4.

**Table 4. Postoperative data (mean)**

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| Groups | N | ICU stay (day) | Inotropic support >3mcg>3day | Pleural effusion (day) | Exudate volume by day (ml/day) |
| Group 1 | 82 | 9±6,51 | 14(17%) | 8,0±5,79 | 206,5±96,71 |
| Group 2 | 6 | 12,0± 6,67 | 3(50%) | 10,0± 4,50 | 207,7±99,3 |
| Group3 | 4 | 9,0± 3,23 | 1(25%) | 12,0± 4,21 | 272,5± 109,2 |

At last follow-up none of the patients developed any SVOT gradient (by Cath). All of patients had zero or trivial neo-aortic valve regurgitation.

**Conclusion**

One of the key points during the TCPC procedure is to create an unobstructed blood flow from the SV to aorta. Our results indicate that the SVOTO relief at the concomitantly with TCPC does not cause significant increase in morbidity and mortality at early postoperative period. The results suggest that the DKS is the method of choice with regard to avoidance of heart block or adequate removal of SVOTO.

Our results are consistent with recent literature data [4,6,8,9,10,11,12].

DKS can be safely performed in conjunction with TCPC without added significant morbidity and mortality risk. It is very effective in eliminating SVOTO risk, with sustainable good semilunar valves function.  
There is a concern that prior PAB could result in compression of the left main bronchus, especially in patients with a GA orientated not side-by-side, may cause pulmonary valve distortion with consequent regurgitation at time of DKS. We have no any mentioned complication incidence in our series.

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