

PALLIATIVE ARTERIAL SWITCH OPERATION AS AN ALTERNATIVE FOR SELECTED CASES: SINGLE CENTERS' EXPERIENCE AND MID TERM RESULTS

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Abstract

ABSTRACT Introduction and Objective: There are various management options for newborns with single ventricle physiology, ventriculoarterial discordance and subaortic stenosis (SOS), classically involving the early pulmonary banding and aortic arch repair, the restricted bulboventricular foramen(BVF) enlargement or the Norwood and the Damus Kaye Stansel (DKS) procedures. The aim of this study is to evaluate the midterm results of our clinical experience with palliative arterial switch operation (pASO) for this subset of patients. Method: We hereby retrospectively evaluate the charts of patients going through pASO, as initial palliation through Fontan pathway, starting from 2014 till today. Results: 10 patients underwent an initial palliative arterial switch procedure. 8 of 10 patients survived the operation and discharged. 7 of 10 patients completed stage II and 1 patient reached the Fontan completion stage and the other six of ten (6/10) patients are doing well and waiting for the next stage of palliation. There are two mortalities in the series (2/10) and one patient lost to follow-up (1/10). Conclusions: The pASO can be considered as an alternative palliation option for patients with single ventricle physiology, transposition of the great arteries and systemic outflow obstruction. It not only preserves systolic and diastolic ventricular function, but also provides a superior anatomic arrangement for following stages. Keywords: Congenital Heart Disease, Single Ventricle, Arterial Switch Operation

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Short Title: Palliative Arterial Switch Operation

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1-INTRODUCTION

Univentricular heart patients, combined with ventriculoarterial discordance and subaortic stenosis is a rare combination of anomalies and the choice between surgical options for better outcomes is still not clear in the current literature.

The options for traditional first stage of palliation for functional single-ventricle lesions with transposition of the great vessels and systemic outflow obstruction has been pulmonary artery banding and aortic arch reconstruction¹²³, enlargement of the restricted bulboventricular foramen (BVF)⁴, and the Norwood⁵ or the Damus-Kaye-Stansel (DKS)⁶ procedures.

Since there is still limited experience on the use of palliative Arterial Switch procedure (pASO)⁷⁸⁹ as the initial stage of palliation for the functional single ventricles, this study is the report of our institutions' clinical experience with this cohort of patients and their midterm results.

2-MATERIALS AND METHOD:

The study includes retrospective analysis of the institutes' clinical experience on patients with Palliative Arterial Switch operations (pASO) for single ventricle anatomy with Transposition of the Great Arteries (TGA) and systemic outflow tract obstruction (SOTO), starting from 2014.

Approval was obtained by the Institutional Review Board. The surgical database was reviewed for all arterial switch cases performed on functional single ventricles. All patients with a functional single-ventricle lesion and systemic outflow tract obstruction (SOTO) (valvar aortic stenosis, subaortic stenosis, or a restrictive

bulboventricular foramen), who underwent an arterial switch procedure as initial palliation, and had operative records available for review were included in the study. Charts were reviewed to evaluate patient demographics, pre-operative condition, cardiac anatomy, pre-operative and post-operative echocardiograms, operative data and most recent follow-up data.

SURGICAL TECHNIQUE:

Following median sternotomy and heparinization, brachiocephalic artery and bicaval cannulations were made to go on cardiopulmonary bypass (CPB) and the patients were cooled down to 28° C. Patent ductus arteriosus (PDA) was ligated proximally on aorta and distally on pulmonary artery and divided. Descending aorta was dissected distally. Aorta was cross (X) clamped and heart was arrested with antegrade Del Nido cardioplegia. The X clamp was moved back to the innominate artery, proximal to the arterial cannulation site, the distal ascending aorta was clamped and the left carotis and left subclavian were snared to continue with antegrade cerebral perfusion. The hypoplastic arcus aorta was reconstructed either by using autologous pericardium treated with glutaraldehyde for 2 minutes or without any patch augmentation. The coronary arteries were transferred to the neo-aorta, using the standard open technique with medially based trapdoor flaps as for the simple arterial switch operation. LeCompte maneuver is used for all patients. Pulmonary artery reconstruction was made using a generous autologous pericardium patch to prevent the upward compression of the neoconstructed aorta. And the proximal augmented pulmonary artery root with a functioning neopulmonary valve is connected to the distal pulmonary artery bifurcation. X clamp was removed after de-airing. After weaning from cardiopulmonary bypass, considering the size of the bulboventricular foramen, the pulmonary artery was left as it is, when no further interventions were needed for a balanced circulation; but when necessary, either a Blalock-Taussig shunt to provide adequate pulmonary blood flow or a pulmonary artery banding, to prevent excessive pulmonary blood flow, was used, as indicated according to peri-operative measurements.

Statistical Analysis

Standard descriptive statistical analyses were performed. Means, medians and ranges were used to describe continuous variables. Frequencies and percentages were used for categorical data. All analyses were conducted with Microsoft Excel (Microsoft Corp, Redmond, Washington).

RESULTS

From January 2014 through today, 10 patients have undergone Palliative Arterial Switch operation (pASO) for the palliation of patients with univentricular anatomy, the transposition of the great arteries and systemic outflow tract obstruction (Table1). The mean age of the patients at the time of first operation was 42.5 days (ranging 7-155). 6/10 (60%) of the patients were boys. The preoperative diagnosis through Transthoracic Echocardiography (TTE) were Tricuspid Atresia (TA) with TGA and systemic outflow obstruction (n=5, 5/10) and Double Inlet Left Ventricle (DILV) with TGA and systemic outflow obstruction (n=5, 5/10).

Average X clamp time was 108 minutes (ranging 88 to 142). All patients had aortic arch reconstruction with/without autologous pericardium, treated with glutaraldehyde. 1/10 of the patients (10%); being the first patient in the series, received a Blalock-Taussig shunt, due to low oxygen saturations when coming off bypass. 7/10 (70%) of the patients, received a loose pulmonary artery banding to balance the pulmonary artery blood flow when coming off bypass and adjusted according to oxygen saturations and blood pressure measurements. 2/10 patients (20%) in the series did not need any further interventions for adequate pulmonary blood flow management, while weaning from CPB. Mean intensive care unit (ICU) and hospital stay intervals were 9 (ranging 5-16) and 16 (ranging 8-38) days, respectively. There were 2 (2/10) mortalities in the series. The first patient was patient #3, who had a palliative arterial switch without any pulmonary banding and the post-operative course was uneventful initially, but had a persisting arrhythmia on day #4, deteriorated and did not respond to any interventions and deceased. The second patient was patient #5, who had a usual palliative arterial switch operation with a pulmonary banding, but needed a re-exploration on the first post-operative night on bedside in the ICU due to metabolic acidosis and was put on ECMO for recovery but did not respond to further interventions and died on POD#12. And the first patient in the series who had

a pASO with the BT shunt, had a long postoperative course which included a tracheostomy, but discharged after 2 months and lost follow-up after the first post-operative clinic visit, since the patient was initially accepted from a nearby country and went back home after treatment.

7 of 10 patients reached stage II and had a cardiac catheterization before the Glenn operation. The mean pulmonary artery pressures (mPAP) were acceptable, only one patient needed tightening of the previous band during the operation. And another patient underwent balloon angioplasty of the distal arch for residual gradient during the catheterization before stage II. 1/7 patients needed an aortic arch reconstruction concomitant to Glenn operation and 2/7 of patients needed pulmonary artery reconstruction as well. The AV valves and semilunar valves were functioning well, the bulboventricular foramen were also evaluated with transthoracic echocardiography and catheterization and no further interventions were needed at this stage (Picture 1). The ventricular functions were well preserved.

Only 1 patient reached the Fontan completion stage yet and preoperative cardiac catheterization showed unobstructed arch, good ventricular function and reasonable valve coaptation. mPAP was 11 with satisfactory pulmonary artery distribution and the patient had an uneventful Fontan operation with an extracardiac conduit.

DISCUSSION

The earliest presentation of the palliative arterial switch procedure in the literature was by Freedom and colleagues in 1980⁷, who described 2 patients with tricuspid atresia that underwent arterial switch and simultaneous Fontan procedure. Karl and associates later developed the technique and reported 6 patients with univentricular hearts and subaortic stenosis, 2 of them went on to BDG and 1 patient underwent a Fontan procedure⁸. Lacour-Gayet and colleagues, also reported 7 patients who underwent palliative arterial switch with good short-term results⁹, but Fontan palliation stage and suitability has not been reported.

The potential advantages of the pASO are well presented earlier¹⁰¹¹; we also believe that the basic principle of this operation is beneficial to "switch" the subaortic obstruction into a subpulmonary obstruction and create a laminar flow through the systemic outflow tract. The relief of subaortic stenosis, thus preventing myocardial hypertrophy of the single ventricle and further restriction of the BVF. pASO also prevents diastolic dysfunction of the single ventricular chamber by keeping a functioning valve in the pulmonary position. The connection of the rudimentary ventricular chamber to the pulmonary artery trunk creates a natural protection for the pulmonary vascular bed through the restrictive bulboventricular foramen¹⁰¹¹¹². And another potential advantage is using the LeCompte maneuver, which not only prevents the compression of the pulmonary arteries under the enlarged root of posteriorly dislocated neo-aorta, but also making it easier to reach the branch pulmonary arteries, to address the future complications at following stages, if necessary¹⁰¹¹. There is also decreased need for a systemic to PA shunt, as a shunt which carries the risk of shunt occlusion, distortion of PA anatomy, and decreased diastolic pressure with potentially poor coronary perfusion.

The main concern with pASO is generally the unpredictable pulmonary blood flow through the ventricular septal defect. Even if the criteria for diagnosis of SAS and restrictive bulboventricular foramen like the BVF size being less than the diameter of the aortic root either by echocardiography or cardiac catheterization or any measured ventriculo-aortic gradient (above 10-20 mmHg) was helpful but considered as unreliable by us and others to be clinically significant⁸¹³¹⁴. These preoperative measurements of the BVF and the aortic annulus circumference might estimate the likelihood of a restriction probability of the blood flow to the pulmonary bed and help to predict the need for an additional intervention for maintaining adequate pulmonary blood flow, when coming of cardiopulmonary bypass.

And another aspect to keep in mind when evaluating the bulboventricular foramen preoperatively is, if the aortic arch is hypoplastic, this might point to the BVF is going to be also restrictive after birth, since it was not providing enough flow for the arch to grow during pregnancy. It has been suggested that the signs of subaortic stenosis in utero can result in subnormal aortic flow, causing underdeveloped aortic arch and even favoring the development of coarctation.

The aim of initial palliation for infants with univentricular hearts, transposition of the great arteries and systemic outflow tract obstruction is basically to provide unrestricted systemic and adequate pulmonary blood flow. The primary objective is carrying the patient safely to the next stage after initial palliation. Either a bidirectional Glenn operation (BDG) or a Fontan procedure, both require the control of excessive pulmonary blood flow to prevent future pulmonary vascular resistance and maintain adequate pulmonary blood flow to provide satisfactory oxygen saturations. The strategy also needs to preserve the univentricular systolic and diastolic function by addressing the systemic outflow obstruction¹⁵¹⁶.

Various other management options to address these conflicts in this group of patients with single ventricle, ventriculoarterial discordance, and subaortic stenosis have included enlargement of the BVF⁴, pulmonary artery banding with arch repair¹²³, modifications of the DKS⁶ or Norwood⁵ procedure.

Although pulmonary artery banding has been advocated as initial palliation in the neonatal period to protect the pulmonary vasculature in patients with functional single ventricle, there is a tendency for spontaneous narrowing of the BVF by itself¹⁷ or accelerated by the immediate reduction in diastolic heart volume after an effective PAB. In the longer term there is also further reduction in BVF size due to PAB-related myocardial hypertrophy³. Subaortic obstruction may complicate the future Fontan palliation by inducing ventricular hypertrophy¹⁵¹⁶, leading to subendocardial ischemia and diastolic dysfunction and this has led some groups to abandon pulmonary artery banding as palliation when subaortic stenosis is present or anticipated.

The key anatomical feature is the BVF in the UVH patients with subaortic stenosis, which is mostly a slit-like or buttonhole like opening with a completely muscular rim and estimation of the size of the ventricular septal defect carries utmost importance. Preoperative echocardiography is often unreliable due to the mentioned anatomic features above, but when the size of the VSD is less than the size of the aortic annulus with a measurable gradient from the dominant ventricle to aortic annulus by Doppler echocardiography(>10-20 mmHg)⁸¹³ and when accompanied with an underdeveloped hypoplastic aortic arch; then the BVF can be considered as restrictive and needs to be addressed accordingly.

The surgical enlargement of the BVF and subaortic resection can be technically quite difficult in the young infant or neonate; risking injury to the conduction system which appears to be close to the inferior rim of the BVF or aortic valve might also be in jeopardy, if the surgical enlargement is carried on through the aortic annulus. Another surgical option that was employed for SAS with enlargement of the BVF is the division and resection of additional subaortic muscle bands⁴. Despite potential complications of heart block, coronary injury, and recurrent SAS this approach has been used extensively in many centers.

The DKS procedure was originally described for biventricular repair of transposition of the great arteries¹⁸. The indications were then extended for use in single-ventricle palliation. The variations of this technique¹⁹ and the Norwood operation have been used to address the sub-aortic obstruction in this subset of patients with single ventricle and transposed great arteries as the primary palliation to end up with Fontan completion⁶.

The operative mortality for the Norwood and the DKS procedures vary in the current literature between institutes, depending on the era of reporting, the weight and the anatomy like factors related to the patient undergoing the procedure, and other factors etc.¹³²⁰ but considering the overall Norwood procedures' mortality being reported as high as 10-30%, the palliative arterial switch procedure may be considered as an alternative for the management of these high risk ventriculo-arterial discordant single ventricle patients with a systemic outlet obstruction.

In summary; pASO is a complex operation, with a long bypass run and remarkable ischemic period but these patients are easier to manage postoperatively than most of the other palliative procedures, since they mostly have banded physiology because of the bulboventricular foramen when coming out of the operating room. The post-operative banded physiology with a harmonious systemic outflow tract simplifies their early post-operative management and offer long-term benefits of systolic and diastolic function preservation that make patients with single ventricles good candidates for the Fontan procedure. We think that our study will help to support an early aggressive approach to rule out potential late complications and facilitate successful palliation of these complicated subset of patients.

CONCLUSION

The ultimate palliation for infants with univentricular hearts, transposition of the great arteries and systemic outflow tract obstruction creates a safe passage through the neonatal period but also prepare them for the long term. pASO provides a natural alignment between the systemic outflow tract and the dominant ventricle and maintains an unrestricted laminar systemic flow pattern to preserve systolic or diastolic function for the future settings. The prevention of further myocardial hypertrophy offers less tendency for further BVF restriction. Thus, considerably easier medical management of pulmonary banding physiology prevents the fluctuations in systemic and pulmonary vascular resistance postoperatively. LeCompte maneuver in addition to the advantages of pASO, by pulling the pulmonary arteries anteriorly, excludes the probability of left pulmonary artery entrapment, which is a possible complication of DKS procedure. Having a functioning pulmonary valve in place also prevents the probable future diastolic dysfunction which is possible with procedures involving shunts, resulting with increase in ventricular volume load.

Limitations

This study is limited because of its retrospective pattern, but mostly the patients were decided to go with pASO in the operating room, which creates a selection bias. The follow-up is midterm for the time being, and 1 patient lost f/u for the given reasons above. The long-term results need to be reevaluated for future discussions.

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