Treatment of right ventricle to pulmonary artery conduit stenosis in infants with hypoplastic left heart syndrome

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Abstract

OBJECTIVES: To determine the incidence of right ventricle-to-pulmonary artery (RV-PA) conduit stenosis after the Norwood I operation in patients with hypoplastic left heart syndrome (HLHS), and to determine whether the treatment strategy of RV-PA conduit stenosis has an influence on interstage and overall survival.

METHODS: Ninety-six patients had a Norwood operation with RV-PA conduit between 2002 and 2011. Details of reoperations/interventions due to conduit obstruction prior to bidirectional superior cavopulmonary anastomosis (BSCPA) were collected.

RESULTS: Overall pre-BSCPA mortality was 17%, early mortality after Norwood, 6%. Early angiography was performed in 34 patients due to desaturation at a median of 8 days after the Norwood operation. Fifteen patients (16%) were diagnosed with RV-PA conduit stenosis that required treatment. The location of the conduit stenosis was significantly different in the patients with non-ringed (proximal) and the patients with ring-enforced conduit (distal), \( P = 0.004 \). In 6 patients, a surgical revision of the conduit was performed; 3 of them died prior to BSCPA. Another 6 patients had a stent implantation and 3 were treated with balloon dilatation followed by a BSCPA in the subsequent 2 weeks. All patients who were treated interventional for RV-PA conduit obstruction had a successful BSCPA. Patients who received a surgical RV-PA conduit revision had a significantly higher interstage \( (P = 0.044) \) and overall mortality \( (P = 0.011) \) than those who received a stent or balloon dilatation of the stenosis followed by an early BSCPA.

CONCLUSIONS: RV-PA conduit obstruction after Norwood I procedure in patients with HLHS can be safely and effectively treated by stent implantation, balloon dilatation and early BSCPA. Surgical revision of the RV-PA conduit can be reserved for patients in whom an interventional approach fails, and an early BSCPA is not an option.

Keywords: Congenital heart disease • HLHS • RV-PA conduit • Stent implantation

INTRODUCTION

As a modification of the classic Norwood I operation, a right ventricle to pulmonary artery (RV-PA) conduit is used to provide pulmonary blood flow [1–3]. Compared with the modified Blalock-Taussig-Shunt, the use of the RV-PA conduit has the advantage of eliminating the diastolic runoff and the coronary artery steal phenomenon [4]. However, the use of an RV-PA conduit is associated with a higher risk of cardiovascular complications like arrhythmias, aneurysm formation [5], negative effects on the right ventricular (RV) function [4, 6] and unintended interventions for an obstruction of the conduit [6, 7].

The purpose of this study was to determine the incidence of RV-PA conduit stenosis after the Norwood I operation in patients with hypoplastic left heart syndrome (HLHS), and to determine whether the treatment strategy of RV-PA conduit stenosis has an influence on interstage and overall survival.

PATIENTS AND METHODS

We identified all patients with HLHS, who had undergone Norwood I palliation with RV-PA conduit, between 2001 and 2011, from our institutional cardiothoracic surgical database. Angiographic data from the cardiac catheterizations between Norwood and bidirectional superior cavopulmonary anastomosis (BSCPA) were reviewed for RV-PA conduit stenosis. Conduit stenosis was defined as any conduit obstruction that required...
interventional or operative treatment. Early mortality was defined as death within 30 days after the operation. The presence and location of the stenosis and the type of treatment were noted for each patient.

Statistical analysis

Descriptive statistics are described as frequencies and percentages for categorical variables. Continuous variables are expressed as mean ± standard deviation, if normally distributed, or median and range for a non-normal distribution. Continuous variables and categorical variables were compared between groups using the two-tailed unpaired Student's t-test and Fisher's exact test, respectively. All data were analysed using SPSS software, version 20.0 (SPSS, Inc., Chicago, IL, USA).

The institutional review board approved the present retrospective follow-up study. The requirement for informed consent was waived. The authors had full access to the data and take full responsibility for the integrity of the data.

RESULTS

Ninety-six HLHS patients had a Norwood operation at age 9 ± 5 days. In all patients, a 5 mm Gore-Tex tube was implanted as an RV-PA conduit. Patients who were operated on prior to 2006 (n = 26) received a non-ringed tube; subsequently, ring-enforced tubes were used (n = 70).

Overall pre-BSCPA mortality was 17%. Early mortality after Norwood was 6% (6 patients). Three of these patients died on the day of the Norwood operation, without angiographic diagnostics. The other three underwent angiography: in one, a conduit stenosis was excluded, the second had a thrombosis of the RV-PA conduit and the third was reoperated due to RV-PA conduit stenosis and died shortly after the reoperation. Another 10 patients died prior to BSCPA (interstage mortality 10%). One patient died at home on the 100th postoperative day after the Norwood I procedure without prior diagnostics; in 7 patients, RV-PA conduit stenosis was excluded angiographically; 2 patients were reoperated due to RV-PA conduit stenosis, and died prior to BSCPA.

Cardiac catheterization between Norwood and BSCPA was performed in 92 patients (96%). Fifty-eight of these patients underwent elective angiography at a median of 4 days (range 0–29 days) prior to BSCPA. The remaining 34 patients underwent unplanned angiography due to desaturation or unstable cardiovascular status at a median of 8 days after the Norwood procedure (range 1–88 days).

Fifteen patients (16%) had a conduit stenosis requiring treatment. Details of the conduit obstruction and the following treatment are shown in Table 1. The occurrence of conduit obstruction after the implantation of ring-enforced (10 out of 70 patients; 14%) and non-ringed conduits (5 out of 26 patients; 19%) was similar (P = 0.54). The location of the conduit stenosis was significantly different in the patients with non-ringed (proximal stenosis) and the patients with ring-enforced conduit (distal stenosis), P = 0.004.

In 6 patients, the RV-PA conduit stenosis was treated surgically, 1–14 days after Norwood I. Two of these patients underwent a successful BSCPA, while one died 2 days following BSCPA. The remaining three patients died prior to BSCPA. A patient who was reoperated 6 days after the Norwood procedure due to proximal RV-PA conduit stenosis, successfully recovered and was released to home care. The child suddenly arrested at home, 110 days after the reoperation, and passed away after an unsuccessful reanimation. Another patient was reoperated 14 days after the Norwood procedure due to distal conduit stenosis. Due to a continued unstable postoperative course, with low arterial saturation, need for haemofiltration and inotropic drugs, one more cardiac catheterization was performed 2 weeks later, and RV-PA conduit stenosis was excluded. The child died 17 days after the reoperation due to the low-output syndrome. The last of these 3 patients, who was reoperated 6 days after the Norwood procedure due to distal conduit stenosis, died 6 days afterwards due to the multiple organ dysfunction syndrome.

Six patients received a stent in the RV-PA conduit 1–44 days after Norwood I, and 3 were treated by balloon dilatation 62–88 days after Norwood I, followed by BSCPA within 14 days of the intervention. All patients who were treated interventionally for RV-PA conduit obstruction had a successful BSCPA. Patients with RV-PA stenosis who were treated surgically had significantly higher interstage (50%), and overall mortality (67%) than those who were treated interventionally (0%); P = 0.044 and P = 0.011, respectively.

BSCPA was performed in 80 patients at median age 124 days (range 64–212 days). No patient had an early BSCPA as a treatment for diagnosed RV-PA conduit stenosis without a prior interventional procedure.

DISCUSSION

RV-PA conduit obstruction is a serious complication after the Norwood I procedure, associated with increased interstage morbidity and mortality [8, 9].

In our study, an early, unplanned cardiac catheterization, performed due to desaturation or haemodynamic instability of the patient, often revealed relevant RV-PA conduit stenosis. The incidence of conduit obstruction after Norwood I varies considerably in existing studies. Photiadis et al [7] report an incidence of 26% of RV-PA conduit obstruction in their retrospective report on 38 patients undergoing the Norwood I procedure, which is similar to the report of Dahnhert et al [10], who had 23% of RV-PA conduit stenosis in 34 patients. Desai et al [11], however, reported an incidence of only 8.6% RV-PA conduit obstruction in their study population of 209 patients. In our institution, the overall incidence of RV-PA conduit obstruction after the Norwood I procedure was 16%. The introduction of a ring-enforced conduit has reduced the incidence of conduit stenosis at the proximal level, as we reported in our earlier publication [12], but not at the distal level.

Treatment options for RV-PA conduit obstruction are surgical revision of the conduit, interventional treatment (balloon dilatation, stent implantation) or early BSCPA in suitable patients. The detrimental effect of volume overload on the function of the systemic RV in patients with HLHS has led to the current consensus of favouring early unloading by BSCPA [13]. A trend towards construction of BSCPA at an earlier age can be seen over the years in our institution and was reported by other authors as well [6, 14]. Our group previously reported that clinical results after BSCPA do not depend on the age of the patient and can be
performed with good results in younger patients [15]. Early BSCPA may even be performed safely in patients under the age of 3 months [6]. In our study, in older patients who were approaching an age when BSCPA could be safely performed and who required an unplanned cardiac catheterization due to desaturation, the stenotic area was treated with balloon dilatation. All of these patients underwent a successful BSCPA in the following 2 weeks. Stent implantation can most likely be avoided at this time, since balloon dilatation alone seems to be sufficient to bridge a short time to BSCPA.

In our study group, the patients with RV-PA conduit stenosis who were not suitable for BSCPA were treated surgically or underwent stent implantation in order to provide a satisfactory longer-term solution. Surgical revision of the conduit was associated with high mortality prior to BSCPA. On the other hand, all patients who received a stent in the stenotic area underwent a successful BSCPA. In cases of proximal stenosis, the stent was removed prior to suturing the proximal end of the conduit at the connection to the right ventricle. In cases of distal stenosis, the stent was removed from the pulmonary artery at the time of BSCPA, and the pulmonary artery was reconstructed using a patch. In our experience, the benefit of stent implantation outweighs the potential morbidity associated with patch angioplasty for stent removal. Altogether, the patients who received a surgical revision of the RV-PA conduit had significantly higher interstage and overall mortality than those who were treated interventionally.

Current data on interventional therapy in case of RV-PA conduit obstruction show good results [6-10], and surgical therapy is being increasingly replaced by interventional procedures [6]. Gray et al. [9] reported a need for stent implantation in 24% of patients undergoing Norwood I procedure with an RV-PA conduit. Muyksens et al. [16] had an incidence of 22% of stent placement after the Norwood I procedure. In the last 5 years, with the increasing experience of the interventional cardiologists, all patients in our institution with relevant conduit stenosis were treated interventionally: younger patients by stent implantation; older patients by balloon dilatation followed shortly by BSCPA.

In summary, if a relevant RV-PA conduit obstruction is found in patients in whom an early BSCPA cannot be performed safely, stent implantation seems advisable. In cases of conduit obstruction in older patients, a BSCPA should be considered. A balloon dilatation in these patients may improve the clinical status prior to surgery. Surgical revision of the RV-PA conduit should be reserved for patients in whom an interventional approach fails, and an early BSCPA is not an option. Using this protocol of treatment, in the last 5 years, there was no interstage or early post-BSCPA mortality after treatment for RV-PA conduit stenosis in our institution.

LIMITATIONS

The present study was retrospective. Although the study group was relatively large, the number of the patients with relevant RV-PA conduit stenosis was small. This might have limited the value of the statistical evaluation of the groups.

CONCLUSION

RV-PA conduit obstruction after Norwood I procedure in patients with HLHS can be safely and effectively treated by stent implantation, balloon dilatation and early BSCPA. Surgical revision of the RV-PA conduit should be reserved for patients in whom an interventional approach fails, and an early BSCPA is not an option.

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REFERENCES


APPENDIX. CONFERENCE DISCUSSION

Dr B. Maruszewski (Warsaw, Poland): Dr Horer and colleagues reviewed their experience of a group of 96 hypoplastic left heart patients who underwent the Norwood operation with the Sano modification and RV-PA conduit during the last decade. The aim of this study was to determine the incidence of conduit stenosis and whether the treatment strategy, redo surgery versus interventional cardiology, has an impact on interstage and overall survival. The authors concluded that introduction of the ring reinforced 5 mm PTFE graft neutralized the issue of proximal stenosis, and distal stenosis remains an issue, but the results of balloon dilatation, stenting, and then second stage done earlier, are superior to redo surgery. I fully agree with the statement based on our experience.

The manuscript that I received did not describe the technique of the conduit connection with the pulmonary arteries. In my opinion it is crucial for avoiding future stenosis and for promoting development of the pulmonary bifurcation. The way we address this issue is to anastomose first the distal end of the PTFE graft with a set of autologous tissue, pulmonary homograft or homograft pericardium, and then to reconstruct the pulmonary bifurcation avoiding specifically going to the left main PA too much. Could you please describe your technique and comment on that.

Dr Horer: Our technique for placement of the distal connection is to pull the central pulmonary artery to the right side of the neoaoic arch. Then we close the pulmonary artery with a patch. We use the same material as we use for arch reconstruction. This is usually a pulmonary homograft or glutaraldehyde-fixed autologous pericardium. In very rare cases we used a Contegra graft or femoral artery. We insert the conduit within the created hood. Sometimes we tend to place the conduit more towards the RPA, to avoid tension on the left pulmonary artery. We analysed our results with regards to conduit stenosis at the distal anastomosis stratified by different patch material, but found no significant differences. Personally I think that the pulmonary homograft is the best graft for reconstructing the main pulmonary artery. We saw peel formation in pericardium and in Contegra grafts, so this may contribute to the incidence of distal shunt stenosis.

Dr Maruszewski: I just suggest that it would be nice if you could add this description to the manuscript, because it probably will add some extra information about your experience and problems and the issue of the peripheral stenosis.

Now, the second issue is the age at the second stage. You have shown very clearly that bidirectional cavopulmonary anastomosis can be performed very early, before the age of three months, in patients who develop conduit stenosis and had balloon dilatation or stenting. You reported no early mortality in this group treated after 2008. So having in mind the importance of early unloading of the single ventricle, what is your current strategy for the age at the second stage in patients who do not develop any complications demanding early reintervention?

Dr Horer: We schedule our patients for elective angiography at 3 months. However, in patients with an RV-PA conduit, we will aim to perform the examination at around 10 weeks after the Norwood procedure. In patients with an initial 3 mm BT shunt, we tend to perform the angiography at an age of 8 to 10 weeks.