Indications for and outcomes of interstage catheter interventions following the Norwood procedure: A single-institution study

Yousra Bouyaala, Robert Bindermann, Stefanie Wendt, Axel Kroener, Gerardus Bennink, Narayanswami Sreeram
Heart Center, University Hospital of Cologne, Cologne, Germany

ABSTRACT

Aims : **The aim of this study was to document the incidence, types, and outcome of interstage catheter interventions following the Norwood surgical palliation.**

Patients and Methods : **A retrospective single-center study of all patients surviving the Norwood operation was performed. All data concerning interstage catheter interventions up to the completion of the superior cavopulmonary shunt were collected.**

Results : **Catheter interventions were performed in 62 of 94 patients (66%; 38 males). These included interventions on the aortic arch (n = 44), the branch pulmonary arteries (PAs) (n = 17), and the Sano shunt (n = 14). Multiple interventions and repeat interventions were common. The minimum aortic arch diameter (pre- versus posttreatment) increased from median 3.1 (2.3–3.3) mm to 5.1 (4.2–6.2) mm (P < 0.001). The catheter pullback gradient decreased from 40 (36–46) mmHg to 9 (5–10) mmHg (P < 0.001), and the echocardiographic gradient from 54 (45–64) mmHg to 12 (10–16) mmHg (P < 0.001). The branch PA diameters increased from 2.4 (2.1–3.0) mmHg to 4.7 (4.2–5.1) mmHg (P < 0.001). The minimum Sano shunt diameters increased from 2.0 (1.5–2.1) mm to 5.9 (5.8–6.0) mm (P < 0.001); this was associated with an improvement in systemic saturation from 63% (60%–65%) to 80% (79–82%) (P < 0.001). Unexpected interstage death at home occurred in two patients who had received no interventions. The remainder received a superior cavopulmonary shunt palliation.**

Conclusions : **Catheter interventions were common. Systematic follow-up and a low threshold for reintervention are essential to the success of staged surgical palliation for this patient cohort.**

Keywords : Catheter interventions, Norwood operation, outcomes

INTRODUCTION

The Norwood procedure is the standard of care for the palliation of neonates with complex single-ventricle physiology and systemic outflow obstruction. Despite a relatively large worldwide experience and important technical modifications of the procedure, operative mortality and postoperative morbidity remain high.[1] In particular, interstage mortality at home seems to be highest between the completion of Stage 1 palliation, and while waiting for the Stage 2 bidirectional Glenn procedure.[2-4] A variety of clinical interventions have been proposed to ameliorate postprocedure morbidity among Norwood survivors. These include continued inpatient hospital stay until Stage 2 palliation (the bidirectional superior cavopulmonary shunt) for high-risk patients, intensive home monitoring programs, and frequent outpatient hospital visits following initial

Access this article online

Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/apc.apc_125_22

How to cite this article: Bouyaala Y, Bindermann R, Wendt S, Kroener A, Bennink G, Sreeram N. Indications for and outcomes of interstage catheter interventions following the Norwood procedure: A single-institution study. Ann Pediatr Card 2023;16:25-31.

Address for correspondence: Dr. Narayanswami Sreeram, Heart Center, University Hospital of Cologne, Kerpener Strasse 62, Cologne - 50937, Germany.
E-mail: n.sreeram@uni-koeln.de

Submitted: 11-Oct-2022 **Revised:** 15-Nov-2022 **Accepted:** 20-Nov-2022 **Published:** 04-Apr-2023

hospital discharge, to comprehensively assess residual or acquired lesions that might require interstage interventions.[2,3] This study presents the results of a single institution in the interstage management of the Norwood procedure, focusing on the incidence, type, and outcome of catheter interventions in this period.

PATIENTS AND METHODS

The entire Norwood cohort over the study period, between January 2003 (when the first Norwood procedure was done at our institution) and December 2019 consisted of 127 consecutive patients. Nine of them underwent the Giessen modification (ductal stent implantation and bilateral pulmonary artery [PA] banding, with or without stent implantation in the interatrial septum) as initial palliation. This approach was confined to patients presenting late (no antenatal diagnosis) with severe acidosis or severe systemic ventricular dysfunction. These patients were excluded from further analysis. There were 24 (20%) early deaths (occurring at <30 days after surgery) in the Norwood surgical group. Deaths were strongly associated with a gestational age of <36 weeks (P = 0.02), birth weight of <2.0 kg (P = 0.02), and weight at Norwood operation of <2.2 kg (P = 0.01).

We analyzed the follow-up data of the 94 consecutive patients who were discharged from the hospital following the Norwood/Sano procedure, either until the completion of the bidirectional Glenn procedure or interstage death. All patients in this cohort had undergone the Sano modification of the Norwood procedure (a right ventricle to PA shunt). A standard follow-up protocol was utilized, after discharge from the hospital. This included recording weekly weight gain, pulse oximetry for systemic saturation measurement (including home pulse oximetry measurement for infants with borderline oxygen saturation of <80%), biweekly hospital visits with comprehensive clinical examination, and focused echocardiography. Attention was paid to the three major areas where problems were anticipated: integrity of the neo-aortic arch (reflected by blood pressure recordings in the upper and lower limbs, and echocardiographic identification of stenoses either in the proximal or distal arch), and integrity of the Sano shunt and branch PAs (as assessed directly by serial measurement of their diameters on transthoracic echocardiography, and indirectly by monitoring the systemic oxygen saturation). Indications for cardiac catheterization and intervention were a persistent arm-to-leg blood pressure gradient of >20 mmHg (as measured on two separate visits spaced 1 week apart), or systemic oxygen saturation persistently below 75%, as measured on 3 consecutive days. The onset of new symptoms, such as increasing breathlessness, failure to take feeds, or absence of weight gain over a 7-day period were all considered to be indications for speedy diagnostic catheterization. Informed parental consent was obtained for all catheterization procedures.

Cardiac catheterization

All procedures were performed under general anesthesia, using the femoral or jugular venous approach. Pressure withdrawal gradients and angiograms of the sites of interest were obtained in a standard manner, using biplane imaging. Transcatheter interventions (balloon angioplasty or stent implantation) were performed as indicated by the findings. Following the procedure, the patients were routinely extubated and returned to the ward. Most patients were discharged from the hospital within 24–48 h after the procedure. Intravenous heparin (200 units/kg/24 h) was administered for 24 h in patients in whom a stent was implanted, followed by oral aspirin (5 mg/kg/day), until second-stage surgery.

Distal aortic arch

Withdrawal pressure gradients in excess of 20 mmHg systolic or a discrete stenosis with the diameter of the stenotic segment being <50% of the diameter of the descending aorta at the level of the diaphragm were considered to constitute indication for intervention. High-pressure noncompliant balloons (Emerge, Boston Scientific, Marlborough, MA, USA) equal in diameter to the descending aorta at the diaphragm were appropriately positioned and inflated up to the nominal pressures, two to three times. Repeat angiography and withdrawal gradient measurements were obtained following the dilation.

In patients requiring a second intervention of the distal arch, stent implantation was preferred to repeat balloon angioplasty (premounted Formula stent, Cook Inc. Bloomington, IN, USA). Stents were implanted over the appropriate guidewire, with or without the use of a guiding sheath.

Proximal arch

Similar criteria, as for distal arch stenosis, were used for dilation of stenoses in the proximal arch, at the level of the proximal Damus anastomosis.

Sano shunt

The indications for intervention in the Sano shunt were a stenosis of the proximal or distal anastomosis, with the minimum diameter being <50% of the nominal diameter (a 6-mm shunt was used in the majority of patients, with patients <3 kg of body weight at the time of the Norwood procedure receiving a 5-mm shunt). Proximal stenoses were routinely treated by stent implantation (premounted Cook Formula stents) 6 mm or 5 mm in nominal diameter, and 20–30 mm long.

Branch pulmonary arteries

Distal Sano shunt stenoses could not be angiographically

distinguished from discrete stenoses of the origins of the branch PAs. A reduction in diameter of >50% at the origins of the branch PAs compared to the diameter at the hilum was an indication for balloon angioplasty using high-pressure noncompliant balloons (Emerge, Boston Scientific) whose nominal diameters were equal to the diameter of the vessel at the hilum. Recurrent stenoses and some individual patients undergoing their first intervention were treated directly with stent implantation at the discretion of the operator.

Statistical analysis

Comparisons of paired pre- and postintervention measurements of diameters, gradients, and oxygen saturation were performed using the Wilcoxon signed-rank test (a nonparametric test for matched samples). SPSS (IBM SPSS Statistics for Windows, version 27.0. Armonk, NY, USA: IBM Corp.) was used for statistical analysis. Oxygen saturations documented as "<60%" were coded as an exact value of 60% for the purpose of analysis. Data were presented as median (interquartile range). Demographic data were presented as median (and range).

RESULTS

Sixty-two patients (66%; 38 males) underwent an interstage catheter intervention. The most common reintervention was for aortic arch stenosis (44 interventions), of whom two patients had balloon angioplasty of the proximal arch upstream from the origin of the brachiocephalic trunk. This was followed by the branch PAs (17 interventions, including stent implantation in a critically stenosed left PA under extracorporeal membrane oxygenation in one neonate) and interventions for proximal Sano shunt obstruction (14 interventions). Several patients had an intervention at more than one site during the initial procedure. These included combined balloon angioplasty of the distal arch and branch PAs (n = 7), arch angioplasty and stent implantation in the Sano shunt (n = 2), and combined Sano stent implantation and PA branch angioplasty (n = 3). A second intervention, several weeks after the initial procedure, was performed in six patients; these included repeat distal arch angioplasty (n = 2), stent implantation in the distal arch (n = 4), and stent implantation in the proximal Sano shunt (n = 1). The median age (and range) from initial surgical palliation, weight, and the interval to completion of the bidirectional Glenn shunt from initial catheter intervention are provided in Table 1. The graphs provide data for the measured variables pre- and postintervention. For the aorta, these included the minimum diameter of the stenotic site [Graph 1], the measured catheter pullback gradient [Graph 2], and the transthoracic echocardiographic gradient prior to hospital discharge [Graph 3]. For the Sano shunt

Table 1: Demographic data relating to the first intervention following surgery

Interval from Norwood (days)	Weight (kg)	Interval to bidirectional Glenn (days)
Aortic arch interventions: Demographic data to the first intervention		
82 (39-215)	4.9 (3.4-6.8)	62 (1-309)
Branch PA interventions: Demographic data to the first intervention		
90 (2-201)	4.5 (3.1-6.8)	62 (30-126)
Sano shunt interventions: Demographic data to the first intervention		
87 (14-212)	4.2 (3.3-7.4)	85 (31-309)

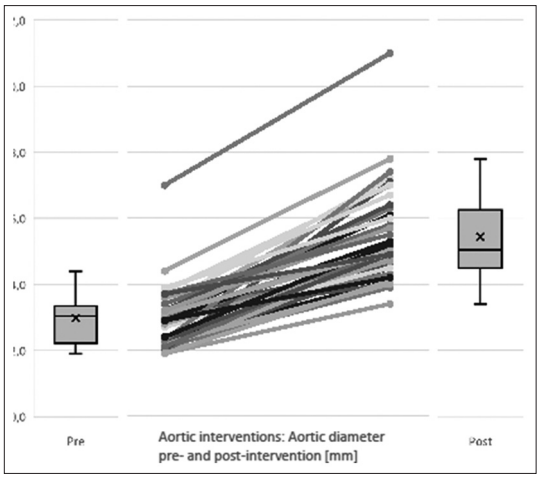
PA: Pulmonary artery

interventions, the data include the measured minimum diameter of the shunt pre- and postintervention and the change in systemic oxygen saturation. For the branch PAs, the data show the change in minimum diameter of the treated branch PA [Graph 4]. The minimum aortic arch diameter (pretreatment vs. posttreatment) of the stenotic segment increased from median 3.1 (2.3–3.3) mm to 5.1 (4.2–6.2) mm (P < 0.001). The catheter pullback gradient decreased from 40 (36–46) mmHg to 9 (5–10) mmHg (P < 0.001), and the echocardiographic gradient from 54 (45–64) mmHg to 12 (10–16) mmHg (P < 0.001). The branch PA diameters increased from 2.4 (2.1–3.0) mmHg to 4.7 (4.2–5.1) mmHg (P < 0.001). The minimum Sano shunt diameters increased from 2.0 (1.5–2.1) mm to 5.9 (5.8–6.0) mm (P < 0.001) [Graph 5]; this was associated with an improvement in systemic saturation from 63% (60%–65%) to 80% (79%–82%) (P < 0.001) [Graph 6]. The only subgroup in whom there was consistently no documented improvement were the patients (n = 2) who underwent balloon angioplasty of the proximal aorta and upstream of the brachiocephalic trunk.

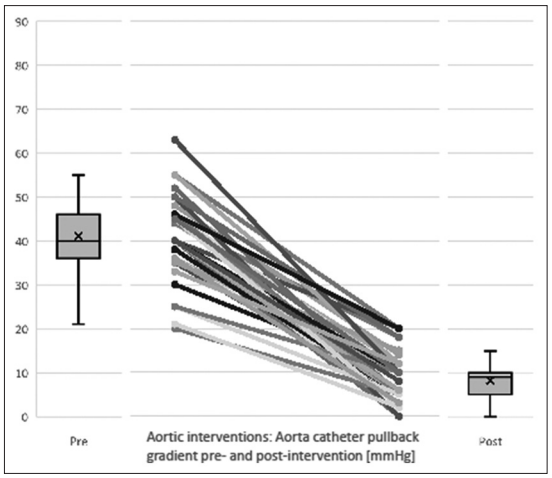
The examples of balloon angioplasty and stent implantation at the various treatment sites are shown in Figures 1-6.

Follow-up

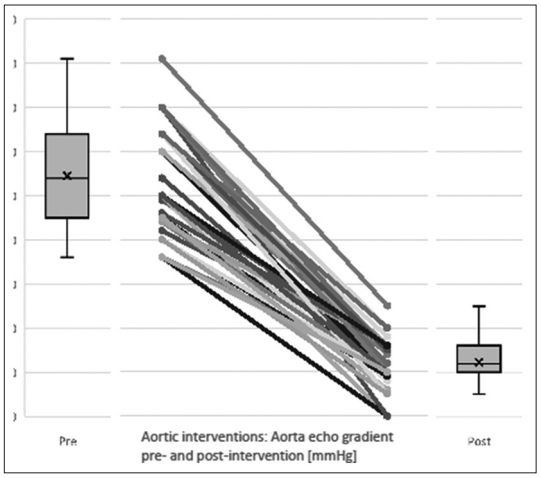
Of the 94 patients discharged from the hospital following Norwood palliation, 92 underwent the bidirectional Glenn procedure. Two infants died suddenly and unexpectedly at home; neither of them had had an unplanned catheter intervention; both families had complied with the home monitoring protocol, and had been routinely evaluated by outpatient follow-up. No permission was given for autopsy. Additional procedures required at the Glenn procedure included patch augmentation of the proximal (n = 1) or distal aortic arch (n = 3) and hybrid stent implantation in the proximal arch (n = 1) or the distal arch (n = 2). As determined by the surgeon, patch enlargement of the PA bifurcation was performed after transecting the distal Sano anastomosis. One infant



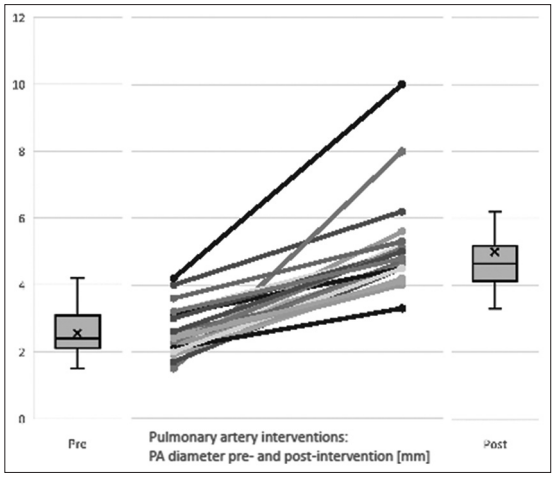
Graph 1: Change in minimum diameter of the stenotic segment of the aorta



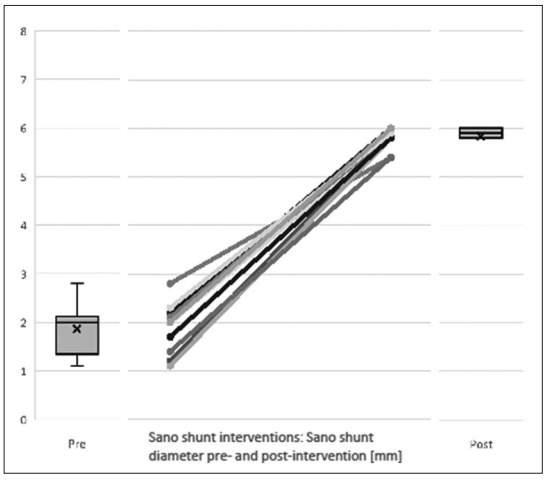
Graph 2: Change in catheter pullback gradient in the aortic arch



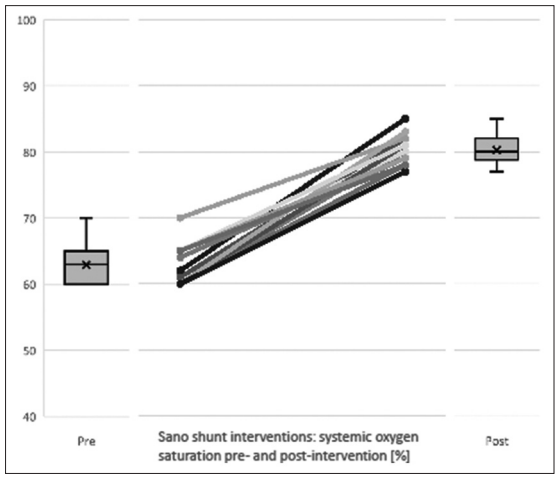
Graph 3: Change in outpatient echocardiographic (continuous-wave Doppler) gradient in the aorta



Graph 4: Change in minimum branch pulmonary artery diameter



Graph 5: Change in minimum diameter of Sano shunt following stent implantation



Graph 6: Change in systemic arterial oxygen saturation following stent implantation in Sano shunt

with a previously implanted stent in the left PA had stent excision followed by patch angioplasty. There were two inhospital deaths following second-stage palliation (at 15 days and 55 days, respectively). Both were infants who underwent combined bidirectional Glenn shunt and hybrid stent implantation in the distal aortic arch.

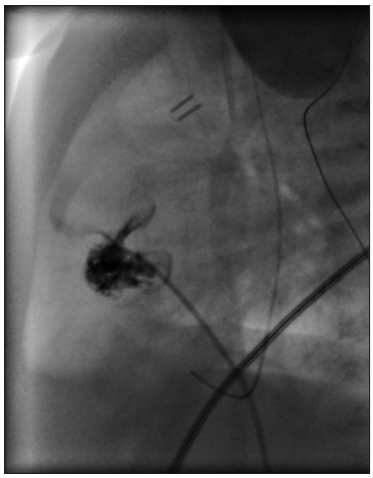


Figure 1: Lateral angiogram showing a critical proximal stenosis of the Sano shunt

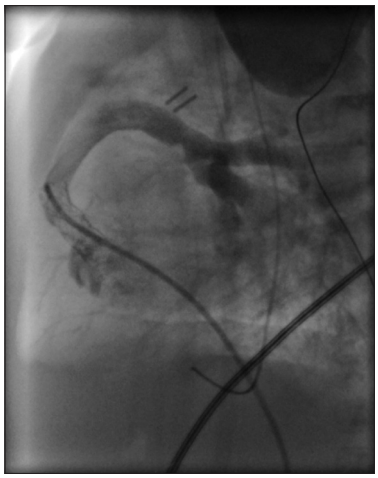


Figure 2: Lateral angiogram following implantation of a stent in the proximal stenotic segment

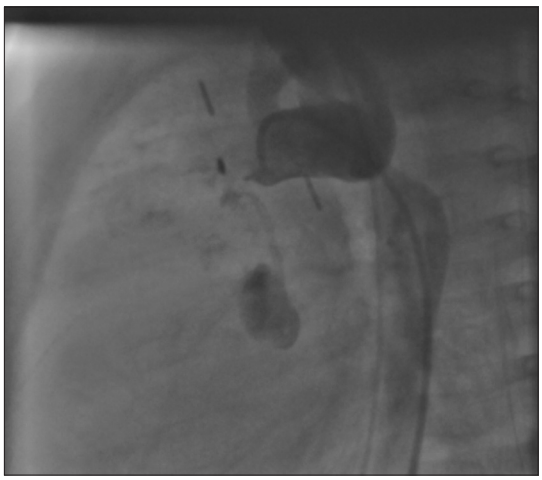


Figure 3: Recoarctation of the distal neo-aorta, following balloon angioplasty as the first intervention

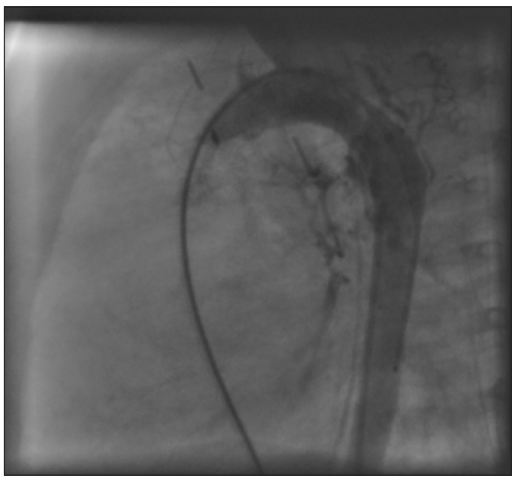


Figure 4: Repeat aortic angiogram, following stent implantation

DISCUSSION

Interstage mortality for the staged Fontan palliation is highest in the period between the Norwood operation and the superior cavopulmonary (bidirectional Glenn) shunt. Assuming that the majority of deaths at home are likely to be caused by residual or recurrent obstruction affecting several critical components of the Norwood-type circulation, several home monitoring strategies have been developed to identify at-risk patients early, which in turn allows remedial therapeutic procedures to be performed in a timely fashion. Some programs even advocate inpatient hospital care until the bidirectional Glenn for post-Norwood survivors deemed to be at high risk.[3] Home monitoring programs have evolved with time. The infant single-ventricle management and monitoring program proposes that patients be discharged from the hospital with a pulse oximeter and weighing scale. It involves appropriate parental education to undertake daily measurement of various physical and clinical parameters, home visits by dedicated nurse practitioners, and routine telephone communication between parents and caregivers.[2,4] Crucial to the follow-up are routine clinic visits and focused echocardiographic examination. Shortening the interstage interval, with a view to performing the bidirectional Glenn shunt at between 3 and 5 months of age, has been proposed to reduce the "time at risk." A simplified version approximating this protocol was followed at our institution; institutional policy dictated that no patient would remain in the hospital until the completion of Stage 2 surgery. As mentioned previously, two infants died suddenly and unexpectedly at home; neither had undergone an interstage intervention, and both had apparently been doing well at the last follow-up.

Recurrent aortic arch obstruction in particular is associated with progressive ventricular dysfunction, the new onset of atrioventricular valve regurgitation, and death.[5] A recent multicenter retrospective study demonstrated a 17% unplanned reintervention rate in a large cohort of patients.[6] A variety of pre- and

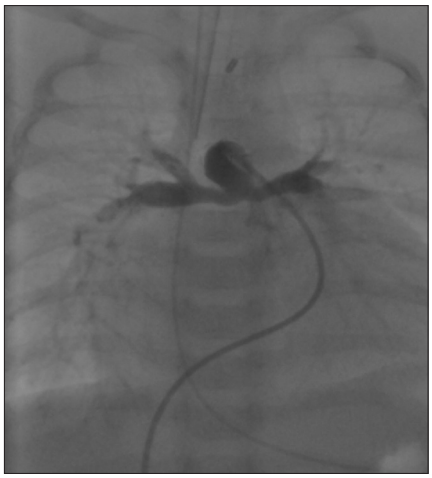


Figure 5: Bilateral proximal branch pulmonary artery stenoses following the Norwood procedure

Figure 6: Repeat angiogram, following balloon angioplasty of the branch pulmonary arteries

perioperative factors during initial Norwood palliation were associated with an increased risk of an unplanned interstage catheter intervention. Although there was no difference in interstage mortality between the cohort undergoing an interventional procedure and those who did not (6% in both subgroups), a worrying finding was that patients who required an interstage intervention were unlikely to proceed to Stage 2 surgery (the bidirectional Glenn shunt), and more likely to receive undergo heart transplantation as the second procedure. In contrast, all infants in the current cohort who had a catheter intervention post-Norwood surgery went on to the completion of the bidirectional Glenn shunt, regardless of the number and type of interventions. In a time of increasing scarcity of resources, in particular for pediatric heart transplantation, it is imperative that all Norwood programs undertake careful and routine interstage screening of survivors, to rapidly identify the development of recurrent hemodynamic lesions that may contribute to higher morbidity or death. There were two post-Glenn inhospital deaths: both patients had a stent implantation in the aortic arch concomitant with surgery. Neither was documented to have severe ventricular dysfunction prior to Glenn palliation.

The efficacy of various types of catheter interventions in the interstage period has been documented by prior studies. Chessa *et al.* documented recurrent arch obstruction in 21 of 95 survivors of the Stage 1 Norwood procedure.[7] Their definition of recurrent obstruction (a peak catheter pullback gradient of >10 mmHg) associated with angiographic evidence of the lumen of the arch was used for this study, as were their criteria for successful balloon angioplasty. The incidence of recurrent arch obstruction in our series compares well with those reported previously.[7,8]

Successful stent implantation for the treatment of proximal stenoses of the Sano shunt, and PA bifurcation angioplasty, have also been previously reported in a smaller series of patients.[9]

CONCLUSIONS

Interstage catheter interventions are frequent following the Norwood palliation. The increased frequency of interventions seen in the current series reflects increased awareness of the adverse consequences of residual hemodynamic lesions, their potential role in interstage home deaths, and the advent of sophisticated home monitoring programs. The financial implications of home monitoring, frequent outpatient visits, hospital admissions, and catheter interventions have to be considered when starting the Norwood program, particularly in developing countries. In addition to the costs associated with Stage 1 surgical palliation and the often prolonged hospital stay, adequate resources and development of expertise in providing optimal home management (sometimes involving the use of pulse oximetry), the ease of arranging and carrying out frequent hospital outpatient checks, and the ability to rapidly program and perform necessary catheter interventions are all resource intensive but, at the same time, important components that contribute to the success of a Norwood program.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Ohye RG, Sleeper LA, Mahony L, Newburger JW, Pearson GD, Lu M, *et al.* Comparison of shunt types in the Norwood procedure for single-ventricle lesions. N Engl J Med 2010;362:1980-92.

2. Gardner MM, Mercer-Rosa L, Faerber J, DiLorenzo MP, Bates KE, Stagg A, *et al.* Association of a home monitoring program with interstage and stage 2 outcomes. J Am Heart Assoc 2019;8:e010783.
3. Hill GD, Tanem J, Ghanayem N, Rudd N, Ollberding NJ, Lavoie J, *et al.* Selective use of inpatient interstage management after Norwood procedure. Ann Thorac Surg 2020;109:139-47.
4. Rudd NA, Ghanayem NS, Hill GD, Lambert LM, Mussatto KA, Nieves JA, *et al.* Interstage home monitoring for infants with single ventricle heart disease: Education and management: A scientific statement from the American Heart Association. J Am Heart Assoc 2020;9:e014548.
5. Larrazabal LA, Selamet Tierney ES, Brown DW, Gauvreau K, Vida VL, Bergersen L, *et al.* Ventricular function deteriorates with recurrent coarctation in hypoplastic left heart syndrome. Ann Thorac Surg 2008;86:869-74.
6. Castellanos DA, Ocampo EC, Gooden A, Wang Y, Qureshi AM, Heinle JS, *et al.* Outcomes associated with unplanned interstage cardiac interventions after Norwood palliation. Ann Thorac Surg 2019;108:1423-9.
7. Chessa M, Dindar A, Vettukattil JJ, Stumper O, Wright JG, Silove ED, *et al.* Balloon angioplasty in infants with aortic obstruction after the modified stage I Norwood procedure. Am Heart J 2000;140:227-31.
8. Zellers TM. Balloon angioplasty for recurrent coarctation of the aorta in patients following staged palliation for hypoplastic left heart syndrome. Am J Cardiol 1999;84:231-3, A9.
9. Moszura T, Dryzek P, Goreczny S, Mazurek-Kula A, Moll JJ, Sysa A, *et al.* A 10-year single-centre experience in percutaneous interventions for multi-stage treatment of hypoplastic left heart syndrome. Cardiol Young 2014;24:54-63.