



Neo-aortic insufficiency late after staged reconstruction for hypoplastic left heart syndrome: impact of differences in initial palliative procedures

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Abstract

The neo-aortic insufficiency in patients with hypoplastic left heart syndrome is an important sequela. We assessed the risks of the neo-aortic valve deterioration by the difference of initial palliations: Group I underwent primary Norwood (Nw) with systemic-to-pulmonary artery shunt (SPS), Group II underwent bilateral pulmonary artery banding (bPAB) and subsequent Nw with SPS (bPAB-Nw/SPS), Group III underwent bPAB and subsequent Nw with bidirectional Glenn (BDG) procedure (bPAB-Nw/BDG). The neo-aortic valve *z* score changes over time did not reach statistical significance in all groups ($p=0.43$ for Group I, 0.20 for Group II, and 0.30 for Group III). The degree of neo-aortic valve insufficiency did not change significantly over time during this period except for Group III ($p=0.34$ for Group I, 0.20 for Group II, and 0.02 for Group III). On the other hand, dimensions of the neo-aortic annulus and degrees of neo-aortic insufficiency did not differ significantly among the 3 groups at any pre-determined time. The presence or absence of incision into the sino-tubular junction at Nw did not affect the late neo-aortic valve *z* score or insufficiency. These data indicate that the difference of initial palliative procedures does not affect late neo-aortic valve insufficiency in Nw survivors. Because valve failure may develop in longer follow-up, further observation should be conducted.

Keywords Hypoplastic left heart syndrome · Late neo-aortic insufficiency · Norwood operation · Bilateral pulmonary artery banding

Introduction

In staged repair for hypoplastic left heart syndrome (HLHS), many surgical strategies have been employed [1, 2]. Norwood operation is essential in the physiological repair for HLHS. On the other hand, bilateral pulmonary artery banding (bPAB) and maintaining the patency of the ductus arteriosus in patients with HLHS were initially introduced to

treat critically ill neonates before the Norwood operation (Nw) [3, 4]; nowadays, it was subsequently developed as an alternative initial palliation [5].

Some studies raised concerns about neo-aortic root dilatation and insufficiency late after Nw [6, 7]. However, it remains to be clarified whether the difference of initial palliations affects these sequelae. In the present study, we evaluated patients' outcomes with an emphasis on the semilunar valve dimension and insufficiency.

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Patients and methods

Patient inclusion criteria

We retrospectively reviewed 66 consecutive patients with HLHS and its related variants who underwent Nw at our hospital between July 1999 and October 2015. Of these, 37 patients who completed the staged Fontan operation were

enrolled and classified into 3 surgical groups in accordance with the type of Nw: Group I underwent primary Nw (pNw) using systemic-to-pulmonary artery shunt (SPS); Group II underwent Nw using SPS after bPAB (bPAB-Nw/SPS); and Group III underwent Nw plus the bidirectional Glenn procedure (BDG) after bPAB (bPAB-Nw/BDG). The surgical strategy had some transitions in this time frame. Until 2003, conventional pNw was performed basically. Between 2003 and 2008, bPAB was performed primarily, and if possible, followed by Nw/BDG at 4 months of age and body weight of 4 kg. Since 2008, we have changed our strategy to perform Nw/SPS at 1–2 months after placing bPAB. Our Institutional Review Board approved this retrospective study, and individual patient consent was waived because of its retrospective nature. Clinical medical records and databases were reviewed in accordance with institutional guidelines.

bPAB procedure

Our operative technique and strategy for bPAB and subsequent Nw-type procedures have been described in detail elsewhere [8, 9].

bPAB was conducted at 2–24 days of age in this series, immediately after pulmonary overcirculation was detected. Through a median full sternotomy with minimal vessel dissection, bilateral branch PAs were encircled and tightened using strips of expanded polytetrafluoroethylene graft (2-mm width), expanded polytetrafluoroethylene suture, or Teflon tape (2-mm width). The tightness of bPAB was adjusted by employing direct epicardial echocardiography. An ideal rate of flow acceleration across the banding sites is that greater than about 3 m/s, while maintaining arterial oxygen tension (PaO₂) in the range of 35–40 mmHg under conditions of room air and normocapnea.

Interstage management of patients after bPAB

Although a new hybrid approach including ductal stenting has been reported, we routinely employed the continuous administration of lipo-prostaglandin E1 to maintain ductal patency. This was because of our concern that ductal stenting, which required full-length coverage between the pulmonary artery junction and passed beyond the aortic isthmus, could obstruct retrograde flow. Baba et al. previously reported that the absence of a ductal stent facilitated easier arch reconstruction at Nw [10].

Subsequently, the surgical strategy for Nw candidates varied, from bPAB followed by Nw/BDG at 4 months of age with a body weight of 4 kg to the modified Nw procedure with SPS at 1–2 months after bPAB.

Staged Fontan track

Among Nw survivors, patients who were evaluated as suitable Fontan candidates underwent a staged extracardiac Fontan procedure through BDG.

Follow-up and echocardiographic examination

All patients had their neo-aortic annular dimension and insufficiency assessed by transthoracic echocardiography on a routine basis, and five time-points (at the time of birth, pre-Nw, pre-BDG, pre-Fontan, and the most recent follow-up) were used for analysis.

All echocardiographic measurements were performed and reviewed by pediatric cardiologists in our hospital. To adjust for the effect of somatic growth and cardiac valve dimensions, the Boston Children's Hospital *z* score calculator system version 1.2.3 (available at: <http://zscore.chboston.org/> [accessed 24 November, 2016]) was used. *Z*-scores were calculated from both normative native aortic and pulmonary valves. In this study, there were no significant differences identified, and dimensions of this valve annulus are expressed as *z* scores using values for the dimensions of the native aortic valve in a normal population. Previous studies described *z* scores in the same fashion [6, 11]. The degree of neo-aortic valve insufficiency was classified as: none (0), trivial (1), mild (2), moderate (3), or severe (4), according to the features on Doppler echocardiography.

Statistical analysis

Data were reviewed retrospectively from patient records and are expressed as the mean \pm standard deviation or median (range). Comparisons among three surgical groups (pNw, bPAB-Nw/SPS, and bPAB-Nw/BDG) and follow-up data were made using one-way analysis of variance, and post hoc tests were done by Tukey's method. Two-group comparisons were performed with an unpaired *t* test. All statistical tests were two-tailed, and differences with a *p* value < 0.05 were considered as significant. Statistical analyses were conducted using GraphPad Prism 5.0 (GraphPad Software, Inc., San Diego, CA, USA).

Results

A total of 66 patients underwent Nw during the study period. Of these, 23 underwent pNw, and 42 underwent bPAB before Nw. The one remaining patient was excluded: the patient underwent coarctation repair and banding of the pulmonary trunk as the first palliation.

This patient was initially considered to have sufficient forward flow through the native aortic valve. Subsequently, the patient's treatment was converted to Nw palliation, since inadequate forward flow was proven. Among 42 who underwent bPAB, Nw/SPS was performed in 27 and Nw/BDG in 15 as second-stage palliation. Of the above, 37 Fontan-completion patients were enrolled in this study as follows: 10 of 23 pNw; 12 of 27 bPAB-Nw/SPS, and 15 of 15 bPAB-Nw/BDG. During the follow-up, 2 of bPAB-Nw/SPS underwent biventricular repair (staged-Yasui operation) and were excluded.

A summary of the clinical outcomes of these patients is presented in Fig. 1, and profiles of enrolled patients are shown in Tables 1 and 2. The median follow-up period was 8.0 years (range 4.8–16.4 years). Because surgical groupings were non-randomized, there were some expected differences between groups in preoperative demographics.

Differences in neo-aortic annular dimensions and insufficiency at each time-point

Dimensions of the neo-aortic annulus and degrees of neo-aortic insufficiency did not differ significantly among the 3 groups at any pre-determined time during the follow-up (Table 3). There was also no clear tendency for groups to show high or low values. At pNw, the dimension and insufficiency at the initial point (at birth) and Nw showed the same values. This was the same for the values at Nw and BDG in bPAB-Nw/BDG.

Fig. 1 Summary of patient outcomes for all Norwood operations. *BDG* bidirectional Glenn procedure, *bPAB* bilateral pulmonary artery banding, *BVR* biventricular repair, *PAB* pulmonary artery banding, *pNw* primary Norwood procedure, *SCF* subclavian flap aortoplasty, *SPS* systemic-to-pulmonary shunt

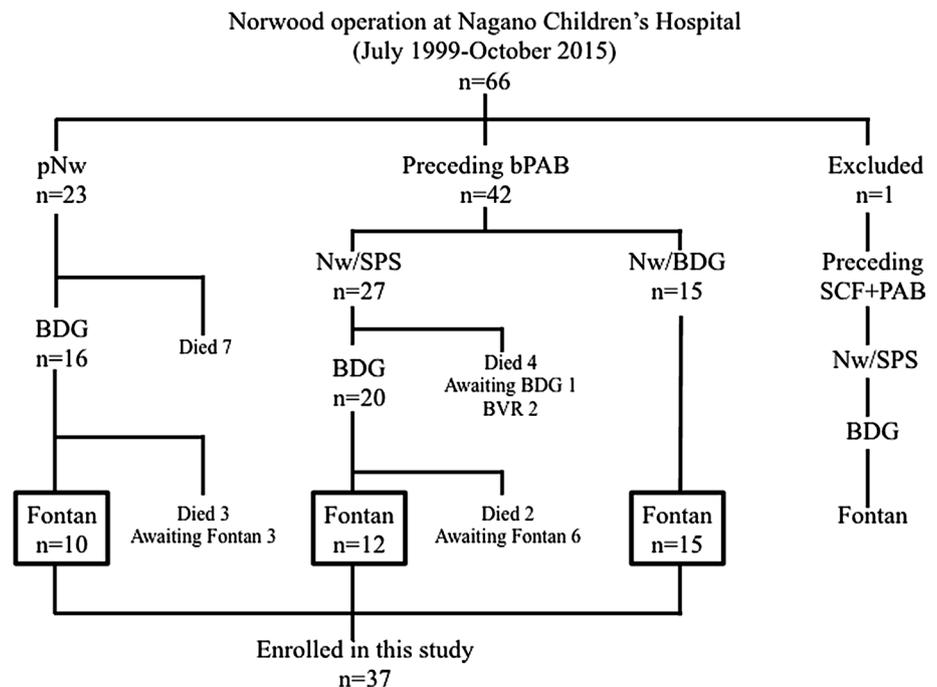


Table 1 Patients' characteristics

Group I	Group II	Group III
pNw (n = 10)	bPAB-Nw/SPS (n = 12)	bPAB-Nw/BDG (n = 15)
HLHS 9	HLHS 6	HLHS 11
DILV, TGA, IAA 1	Polysplenia, CoA, unbalanced AVSD 2	CCH, CoA, DORV 1
	IAA, SAS, small LV 2	DILV, TGA, IAA 1
	IAA, DORV, SAS 1	MA, DORV, CoA 1
	DILV, TGA, CoA 1	AA, CoA 1

AA aortic atresia, AVSD atrioventricular septal defect, CCH criss-cross heart, CoA coarctation of aorta, DILV double-inlet left ventricle, DORV double-outlet right ventricle, HLHS hypoplastic left heart syndrome, IAA interruption of aortic arch, LV left ventricle, MA mitral atresia, SAS subaortic stenosis, TGA transposition of great arteries

Secular changes of neo-aortic annular dimensions and insufficiency in each procedure

Secular changes of neo-aortic annular dimensions and insufficiency in each procedure are shown in Table 3, and the distribution of patients with various degrees of neo-aortic insufficiency is presented in Fig. 2.

The neo-aortic valve z score change over time did not reach statistical significance in all groups ($p = 0.43$ for pNw, 0.20 for bPAB-Nw/SPS, and 0.30 for bPAB-Nw/BDG). The degree of neo-aortic insufficiency did not change significantly over time during this period except for group

Table 2 Demographic data

	Group I pNw (n = 10)	Group II bPAB-Nw/SPS (n = 12)	Group III bPAB-Nw/ BDG (n = 15)	p value
bPAB				
Age, days	–	6.9 ± 3.6	7.7 ± 5.1	0.67
Interval (bPAB-Nw), days	–	57.1 ± 30.0	115.7 ± 18.1	<0.01
Nw				
Age, days	11.7 ± 3.0*,**	64.2 ± 30.7 [#]	123.3 ± 16.9	<0.01
BW, kg	2.9 ± 0.5**	3.3 ± 0.7 [#]	5.3 ± 0.6	<0.01
AAo size, mm	3.4 ± 2.0	4.8 ± 1.3	4.3 ± 2.4	0.46
ST-J incision +	4/10	2/12	8/15	
BDG				
Age, months	5.1 ± 2.5*	10.4 ± 4.0 [#]	4.1 ± 0.6	<0.01
Fontan				
Age, months	40.4 ± 9.0	33.6 ± 8.2	32.1 ± 8.9	0.07
Follow-up				
Age, months	160.0 ± 38.2*,**	77.0 ± 16.3	94.9 ± 21.3	<0.01

Continuous data are shown as mean ± standard deviation and categoric data as number

AAo ascending aorta, BDG bidirectional Glenn operation, bPAB bilateral pulmonary artery banding, BW body weight, Nw Norwood operation, SPS systemic-to-pulmonary artery shunt, ST-J sino-tubular junction

* $p < 0.05$ versus Group II. ** $p < 0.05$ versus Group III. [#] $p < 0.05$ versus Group III

Table 3 Changes in neo-aortic valve dimension and insufficiency

	Group I pNw (n = 10)	Group II bPAB- Nw/SPS (n = 12)	Group III bPAB- Nw/BDG (n = 15)	p value
Neo-aortic valve z score				
Initial (bPAB or Nw)	4.3 ± 0.8	3.4 ± 2.1	4.5 ± 1.3	0.29
Nw	4.3 ± 0.8	4.0 ± 1.4	5.3 ± 2.0	0.14
BDG	5.2 ± 2.6	5.3 ± 2.1	5.3 ± 2.0	0.99
Fontan	5.3 ± 2.4	4.5 ± 2.0	5.8 ± 2.4	0.36
Follow-up	4.0 ± 1.7	4.8 ± 2.1	4.4 ± 1.8	0.60
p value	0.43	0.20	0.30	
Degree of neo-aortic valve insufficiency				
Initial (bPAB or Nw)	0.4 ± 0.5	0.3 ± 0.6	0.1 ± 0.4*	0.43
Nw	0.4 ± 0.5	0.5 ± 0.7	0.4 ± 0.5	0.88
BDG	0.7 ± 0.8	0.8 ± 0.8	0.4 ± 0.5	0.37
Fontan	0.9 ± 0.9	0.6 ± 0.7	0.7 ± 0.8	0.64
Follow-up	1.0 ± 1.2	0.9 ± 0.7	0.8 ± 0.7	0.84
p value	0.34	0.20	<u>0.02</u>	

Continuous data are shown as mean ± standard deviation. Data comparisons between 3 surgical groups and 5 defined times were made

Underline value indicates the degree of neo-aortic insufficiency significantly deteriorated over time in bPAB-Nw/BDG group ($p = 0.02$)

BDG bidirectional Glenn operation, bPAB bilateral pulmonary artery banding, Nw Norwood operation, SPS systemic-to-pulmonary artery shunt

* $p < 0.05$ versus follow-up in group-III

bPAB-Nw/BDG. ($p = 0.34$ for pNw, 0.20 for bPAB-Nw/SPS, and 0.02 for bPAB-Nw/BDG).

Diameter of native ascending aorta with/without incision into sino-tubular junction at Nw

Relationships between the neo-aortic valve with/without incisions into the sino-tubular junction at Nw are shown in Fig. 3. Because large native ascending aorta did not need incision into the sino-tubular junction at the time of Nw operation, the group without incision showed a significantly larger ascending aorta at Nw, as expected (Fig. 3a). However, comparison among the groups did not reveal a significant difference in the neo-aortic annular dimension or degree of insufficiency (Fig. 3b, c).

Discussion

Many therapeutic strategies have been proposed for patients with HLHS and associated anomalies [1, 2, 4, 12–14]. In our hospital, the strategy has also undergone some modifications. Conventional primary Nw/SPS was initially performed as stage-1 palliation. In 2001, we introduced bPAB, especially for high-risk HLHS neonates. Since 2003, we have basically performed bPAB for most HLHS neonates excluding patients with pre-determined conditions, such as pulmonary venous obstruction or poor retrograde cerebral blood flow. This strategy requires the followings: appropriate

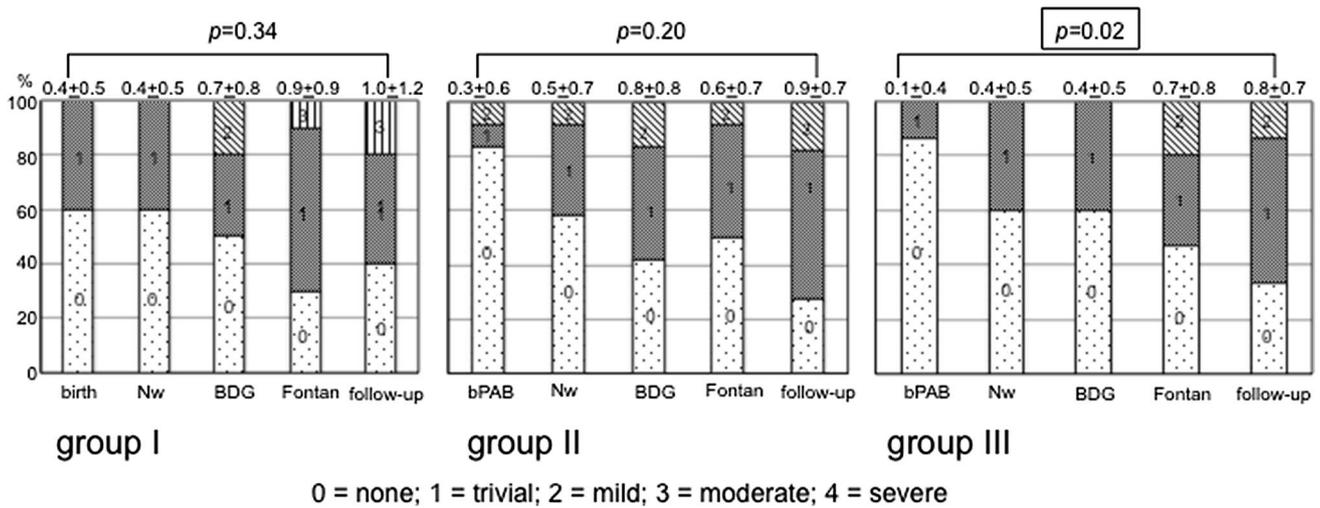


Fig. 2 Degree of neo-aortic insufficiency in patients undergoing pNw (a), bPAB-Nw/SPS (b), and bPAB-Nw/BDG (c). Histogram showing the percentage of patients with neo-aortic regurgitation at the pre-determined times. Insufficiency was classified as follows: 0=none;

0.5 = tiny; 1 = trivial; 2 = mild; 3 = moderate; 4 = severe. *Nw* Norwood procedure, *BDG* bidirectional Glenn procedure. Note that same data are at birth and Nw for pNw, and at Nw and BDG for bPAB-Nw/BDG

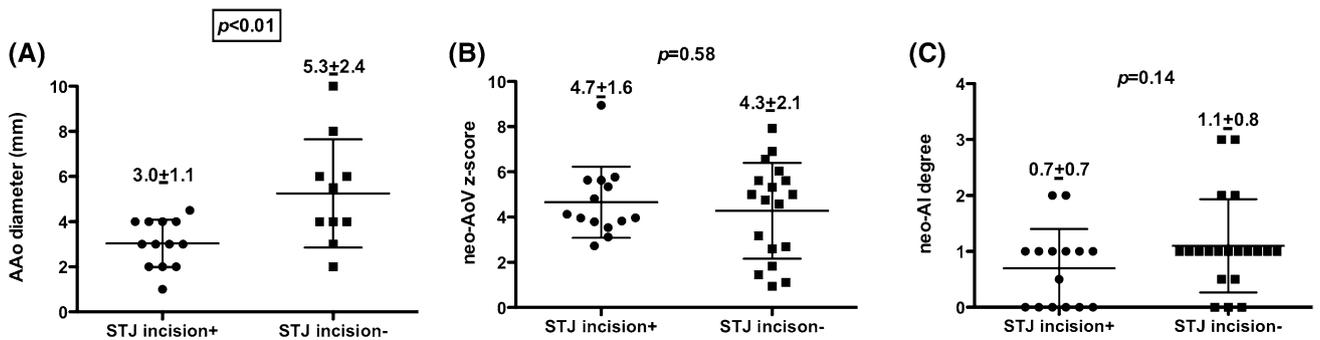


Fig. 3 Relationships between with/without incision into sino-tubular junction during Norwood operation and: diameter of native ascending aorta (a), neo-aortic valve z-score (b), and neo-aortic valve insufficiency (c). *AAo* ascending aorta, *AI* aortic insufficiency, *AoV* aortic valve, *STJ* sino-tubular junction

iciency (c). *AAo* ascending aorta, *AI* aortic insufficiency, *AoV* aortic valve, *STJ* sino-tubular junction

pulmonary blood flow controlled by bPAB; maintenance of ductal patency; and sufficient inter-atrial communication. To meet these requirements, Nw can be postponed from the early neonatal period until infancy. There are some options regarding the concomitant procedure with Nw, determined by the patients’ characteristics: age; body weight; pulmonary artery configuration and resistance; atrioventricular valve insufficiency; systemic ventricular function; and extracardiac disorders. From the perspective of systemic ventricular preload, Nw/BDG is a suitable procedure. However, it is applicable only under limited conditions: reasonable pulmonary artery configuration and low pulmonary vascular resistance. Therefore, a prolonged waiting period between bPAB and subsequent Nw/BDG is usually necessary. Moreover, hybrid procedure (bPAB and ductal stenting) is considered to have no advantage on ventricular energetics compared

to the Nw with right ventricle-to-pulmonary artery shunt [15]. In addition, the most important advantage of bPAB is to avoid the invasive operation in the first few days of life. Recently, we have conducted early conversion from bPAB circulation into Nw/SPS, as reported previously [8].

Cohen et al. described progressive neo-aortic root dilation and neo-aortic valve insufficiency over time after staged reconstruction for HLHS [6]. In the present study, the neo-aortic valve z score change over time did not reach statistical significance in all groups. The degree of neo-aortic insufficiency did not change significantly over time during this period except for group bPAB-Nw/BDG. The interpretation of this result is difficult. Since the post hoc tests showed significance between “initial” and “follow-up”, initial degree of neo-aortic insufficiency in bPAB-Nw/BDG is considered to be too small (0.1 ± 0.4). In the meanwhile, dimensions of the

neo-aortic annulus and degrees of neo-aortic insufficiency did not differ significantly among the 3 groups at any pre-determined time. Hence, the difference of initial procedure itself does not affect late neo-aortic insufficiency.

In fact, the severity of neo-aortic regurgitation steadily tend to progress in each group during the follow-up. Nw operation for HLHS may have inevitable destiny of late neo-aortic insufficiency.

Progressive neo-aortic root dilatation and subsequent semilunar valve insufficiency in the aortic position also remain major concerns in patients after the arterial switch and Ross operations. In the literature, many mechanisms are considered to be responsible for these sequelae [16–19]. Although there are similarities and differences between Nw and these procedures, one reasonable explanation for neo-aortic root dilatation is that the histologically and morphologically native pulmonary valve must tolerate a high systemic pressure in the aortic position. In this respect, the same is equally true of both Nw and the Damus-Kaye-Stansel operation (DKS), whereby the native pulmonary valve functions in the systemic circulation. Fujii et al. reported that the type of DKS affected the long-term outcome: double-barrel DKS was superior to end-to-side DKS for the prevention of postoperative native pulmonary valve regurgitation [20]. The types of material used for Nw have been associated with the development of neo-aortic root aneurysm and semilunar valve regurgitation in HLHS patients [21]. Consequently, the type of Nw reconstruction can affect neo-aortic insufficiency: anastomosis fashion between the native ascending aorta and pulmonary trunk; with/without patch augmentation with root and/or arch reconstruction; relationship of the native great vessels regarding their position and size discrepancy.

A recent study reported that the indexed neo-aortic annular dimension significantly decreased at the pre-Fontan study compared with 14-month study in Nw survivors [22]. The construction of BDG reduced the systemic right ventricular volume load in HLHS patients [23]. Thereby, the systemic ventricular preload is considered to influence the neo-aortic valve (systemic semilunar valve) dimension. Our data were inconsistent with that study. The neo-aortic valve z score unchanged over time in all groups. In contrast, neo-aortic insufficiency showed a deteriorating trend over time in all groups in this study, although it did not reach significance. We suggest that neo-aortic insufficiency in Nw survivors is not just simply the result of neo-aortic root dilatation.

Incision of the sino-tubular junction when performing the arterial switch operation causes late neo-aortic root dilatation (geometrical changes in sino-tubular junction) and semilunar valve insufficiency [24, 25]. In Nw candidates with a small ascending aorta, it is inevitable for an incision to be made facing sinuses between the native-aortic and native-pulmonary root to maintain an adequate coronary blood

supply [26, 27]. Regarding the hemodynamic effects, an equivalent outcome for the neo-aortic root can be expected with an incision and suture line in the sino-tubular junction in arterial switch and Nw. However, we did not identify any differences between groups with/without incision of the sino-tubular junction in Nw patients.

Banding of the pulmonary trunk has been reported as a risk of pulmonary valve insufficiency in patients undergoing the DKS operation based on mid- and long-term follow-ups [28]. However, it remains unclear whether banding of the bilateral branch pulmonary artery has any effect on the semilunar valve in Nw candidates. There was no evidence of change in blood flow through semilunar valves after bPAB. With respect to banding of the pulmonary trunk in DKS patients, the semilunar valve and blood flow across the banding site lie on the same straight line. Moreover, the banding site is very near to the neo-aortic valve. In patients with bPAB, flow across the semilunar valve occurs on a different axis from flow across the banding site of the branched pulmonary artery. Hence, the differences in the banding site may lead to different effects on the semilunar valve. This hypothesis warrants testing with a computational fluid dynamic model.

In Nw patients with progressive neo-aortic root disease, the indication of surgical aortic root repair remains unclear. Some reported a challenging case of valve-sparing neo-aortic root replacement late after Nw [11, 29]. This technically demanding operation must include not only the standard valve-sparing maneuver involving the neo-aortic root, but also temporary division and subsequent reattachment between the original ascending aorta and main pulmonary trunk. Furthermore, it is necessary to remove all affected tissue and provide circumferential support below the annulus, thereby excluding recurrent neo-aortic root dilatation and valve insufficiency.

In this current “congenital heart disease carrying-over into adulthood” era, the number of patients requiring neo-aortic root surgery late after Nw will increase steadily.

This study had some limitations. This was a retrospective, single-center, and relatively small-cohort observational study conducted over a long period of time. Changes in surgical and perioperative managements and the non-randomized nature might have influenced the results. In addition, because some patients with HLHS variants were enrolled, the native aortic and pulmonary valve dimensions ranged widely.

Conclusions

The difference of initial palliative procedures does not affect late neo-aortic valve insufficiency in Nw survivors. Dimensions of the neo-aortic annulus remained unchanged during

the follow-up. We speculate that neo-aortic insufficiency in Nw survivors is not just simply the result of neo-aortic root dilatation.

These findings raise concerns about the long-term semi-lunar valve function after Nw. Therefore, further observation into adulthood should be conducted.

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Compliance with ethical standards

Conflict of interest None declared.

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