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Original article

Balloon Atrial Septostomy with Hypoplastic Left Heart Syndrome with Restrictive Atrial Septum

Short running title: Management of Hypoplastic Left Heart Syndrome

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The authors declare no conflicts of interest.

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Abstract

Background: Rashkind balloon atrial septostomy (BAS) can be challenging in infants with hypoplastic left heart syndrome (HLHS) and small atrial septal defects (ASD).

Methods: We retrospectively reviewed all infants with HLHS who underwent surgery and BAS between January 2006 and December 2015. The infants were divided into three groups: no BAS, catheter BAS, and open BAS. Infants who underwent catheter BAS were divided into two groups based on atrial septal anatomy: standard and complex.

Results: Of the 70 patients, 57 (81%) underwent Glenn surgery. Subsequently, a significant difference in survival was observed: 86% (44/51), 91% (10/11), and 25% (2/8) in the no BAS, catheter BAS, and open BAS groups, respectively (p=0.0002). No significant difference was noted between the no BAS and the catheter BAS groups (p=1.0). In the 56 patients who underwent catheterization after surgery, no intergroup differences in mean pulmonary artery pressure, pulmonary vascular resistance, or pulmonary artery index were found. We classified catheter BAS cases into standard (n=5) and complex (n=5) based on ASD location, and septum thickness. All patients in the standard group underwent complete Rashkind BAS; however, in the complex group, only 1 patient underwent complete Rashkind BAS, with the remaining requiring initial static BAS (p=0.048). Following septostomy, ASD size, ASD flow, and SpO_2 showed no significant differences between the two groups.

Conclusions: Catheter BAS is effective in infants with HLHS and a restrictive atrial septum. Infants with standard or complex atrial septum can achieve equivalent outcomes despite more patients often requiring static BAS.

Key words: Hypoplastic Left Heart Syndrome; Atrial Septum; Catheterization; Heart Septal Defects, Atrial; Prognosis

Introduction

Surgical outcomes for hypoplastic left heart syndrome (HLHS) have improved markedly in recent years. However, short-term and long-term prognoses can be unsatisfactory in patients presenting with hypoxemia or circulatory failure soon after birth, as well as in patients with restrictive atrial septal defects (ASD) or an intact atrial septum (IAS) that requires therapeutic intervention. Balloon atrial septostomy (BAS) is an important therapeutic strategy for eliminating pulmonary congestion in HLHS. However, Rashkind BAS for HLHS can be difficult in patients with small left atrial volumes, depending on the morphological characteristics and the location and thickness of the septal wall. We investigated the efficacy of BAS for HLHS at our hospital. In addition, we examined the effects of ASD morphology on the methods and efficacy of catheter BAS.

Subjects and methods

Subject patients

We reviewed the medical records to retrospectively identify patients who underwent cardiac catheterization and surgery for HLHS at Okayama University Hospital between January 2006 and December 2015. HLHS was defined as hypoplasia of the left ventricle, normal atrioventricular and ventriculo-arterial relationships, aortic atresia (AA) or aortic stenosis (AS), and mitral atresia (MA) or mitral stenosis (MS). Patients with HLHS variants, including those with similar hemodynamics and a ventricular septal defect, double-outlet right ventricle, transposition of the great arteries, and other forms of ventriculoarterial discordance, were excluded.

Items examined and methods

The subjects were divided into intervention (for interatrial communications) and non-intervention groups. The intervention group was further divided into BAS via thoracotomy (open BAS, including atrial septostomy for IAS) and catheter BAS groups. The non-intervention group, the catheter BAS group, and the open BAS group were then compared in terms of survival rates, mean pulmonary pressure, pulmonary vascular resistance, and pulmonary artery index (PAI)[1] after the Glenn procedure .

Next, we further examined the group that underwent catheter BAS. We investigated ASD morphology, BAS method, ASD size before and after BAS, peak flow velocity (m/s) through the ASD, SpO_2, and complications that accompanied BAS. The ASDs were categorized as either standard atrial anatomy or complex atrial anatomy according their morphological characteristics based on a report by Holzer et al.[2] Standard atrial anatomy was defined as (1) the presence of patency near the center of the atrial septum and (2) the absence of thickening of the atrial septal wall. Complex atrial anatomy was defined as (1) the presence of patency superior (near the pulmonary vein) or inferior (near the atrioventricular node) to the center of the atrial septum and (2) the presence of thickening of the atrial septal wall. (Fig. 1)

Regarding the BAS method, our department's approach is to initially perform the Rashkind procedure whenever possible and reserve the static procedure for technically difficult cases.

Echocardiography was performed before and after BAS. A Philips IE33 system (Phillips Medical Systems, Andover, MA) was used to measure the size, position, and the peak flow velocity at the ASD. The measurements were performed using the subcostal frontal view.

Statistical analysis

Continuous variables were compared using the Mann-Whitney U or chi-squared tests. Paired data were examined using the paired t-test. Probability values less than 0.05 were considered statistically significant.

Results

Cases

During the study period, 70 patients underwent cardiac catheterization and surgery for HLHS at Okayama University Hospital. Prior to the Norwood procedure, 19 patients underwent interventions for the restrictive atrial septum and 51 did not. Of the 19 patients undergoing intervention, 8 underwent open BAS (6 with an IAS, 3 with birth weight <2 kg, and 1 with both an IAS and birth weight <2 kg). The 6 patients with an IAS underwent atrial septostomy and the other 2 underwent balloon dilation. Catheter BAS was performed in 11 patients. (Fig. 2)

Glenn completion rate and Pulmonary artery findings after Glenn procedure

Of the 70 HLHS patients, the Glenn procedure was completed in 57 (81%) (Fig. 3), who underwent the procedure at a mean age of 7.0 ± 2.6 months. BAS was not performed in 51 patients, catheter BAS was performed in 11, and open BAS was performed in 8. A significant difference in survival after the Glenn procedure was observed among the 3 groups (86%, 44/51; 91%, 10/11; and 25%, 2/8, respectively; p=0.0002). When comparing 2 groups together, significant differences in the Glenn completion rate were observed between the open BAS and non-BAS groups (p=0.0008) and the open BAS and catheter BAS groups (p=0.0063), with the open BAS group having a lower rate. A significant difference was not observed between the non-BAS and the catheter BAS groups (p=1.0). The 6 patients in the open BAS group in which

the Glenn procedure could not performed included 4 with an IAS and 3 with low birth weight (1650 g, 1514 g, and 1100 g; 1 patient had both an IAS and low birth weight). Four patients died after bilateral pulmonary artery banding and two died after the Norwood procedure.

Excluding only 1 patient who died after the Glenn procedure, catheter examinations were performed in 56 patients. The mean pulmonary pressure, pulmonary vascular resistance, and PAI after Glenn completion were 11 ± 2 mmHg, 1.7 ± 1.0 Wood Units (WU)·m^2, and 216 ± 62, respectively. In the non-BAS, catheter BAS, and open BAS groups, the mean pulmonary pressures were 11 mmHg, 11 mmHg, and 12 mmHg (p=0.81); pulmonary vascular resistances were 1.6 WU·m^2, 1.5 WU·m^2, and 1.5 WU·m^2 (p=0.7); and PAIs were 204, 219, and 237 (p=0.76), respectively. There were no significant differences between the groups.

Catheter BAS group: Patients characteristics and BAS method

The characteristics of patients in the catheter BAS group are shown in Table 1. We excluded 1 patient with cor triatriatum because the intra-atrial communication was not the only area of stenosis. At the time of BAS, the patients (8 boys, 2 girls) had a median age of 3 days (0–50), mean body weight of 2.9 kg (2.2–3.5), and the following aortic valve/mitral valve subtypes: MA/AA, 6 patients; MS/AA, 3 patients; and MS/AS, 1 patient.

In terms of the BAS method, the initial Rashkind BAS was successful in 6 patients, while static BAS was performed before the Rashkind BAS in 4. A Miller Catheter (Edward-Baxter Healthcare Corporation, Irvine, CA) was used in all patients who underwent Rashkind BAS. The mean size of the angioplasty catheter balloon used for static BAS was 7 mm (4–10).

BAS efficacy

After BAS, the mean ASD size changed from 3.2 ± 1.1 mm to 4.7 ± 1.3 mm, the mean peak flow velocity (ASD flow) decreased from 1.7 ± 0.5 m/s to 0.9 ± 0.2 m/s, and the mean SPO_2 increased from 81 ± 9% to 92 ± 6%. Using transthoracic echocardiography, all patients exhibited significant dilation of the ASD ($p < 0.0001$), significant reductions in ASD flow ($p < 0.0003$), and significant increases in SpO_2 ($p=0.0015$), indicating that BAS was effective in all the patients. (Fig. 4)

ASD morphology

The patients were categorized into 2 groups based on ASD morphology and subsequently compared. Following the aforementioned report by Holzer et al., 10 patients were categorized based on ASD morphology, which resulted in 2 groups comprising 5 standard ASD patients and 5 complex ASD patients. In the standard ASD group, the mean ASD size after BAS decreased from 3.4 ± 1.0 mm to 5.3 ± 1.5 mm ($p=0.0014$), ASD flow decreased from 1.6 ± 0.4 m/s to 0.8 ± 0.2 m/s ($p=0.017$), and SpO_2 increased from 83 ± 8% to 92 ± 4% ($p=0.001$). In the complex ASD group, the mean ASD size after BAS expanded from 2.4 ± 0.7 mm to 4.2 ± 0.9 mm ($p=0.0011$), ASD flow declined from 1.8 ± 0.5 m/s to 0.9 ± 0.2 m/s ($p=0.017$), and SpO_2 increased from 81 ± 12% to 90 ± 8% ($p=0.04$). All 5 patients in the standard group underwent a complete Rashkind BAS at the first attempt. However, in the complex ASD group, only 1 patient underwent a complete Rashkind BAS as the first procedure, and the remaining patients required static BAS prior to Rashkind BAS. Based on our comparison of ASD morphology, an initial Rashkind BAS was significantly more difficult to perform in patients with complex ASD than in patients with standard ASD ($p=0.048$). In addition, no significant differences were observed between the two groups in terms of ASD size, ASD flow, or SpO_2 after BAS. (Table 2)

Complications

Complications were observed in 2 patients: 1 case of paroxysmal supraventricular tachycardia and 1 case of hypoxemia. Both patients were in the standard ASD group. The complications were transient and later improved. No critical complications were observed.

Discussion

The prognosis of infants with HLHS has markedly improved recently due to developments in perioperative management[3] and surgical methods.[4] However, it remains significantly poorer in infants with HLHS with an intact/restrictive atrial septum and hypoxemia, acidosis, pulmonary congestion, high pulmonary vascular resistance, or other issues presenting soon after birth. About 6% of HLHS cases are accompanied with an intact septum, and 22% with a restrictive ASD.[5] Hoqoe et al.[6] examined 141 patients with HLHS from 2003 to 2010, dividing them into a group that underwent catheterization within 72 hours of birth and a group that did not. The rates of hospital discharge after the Norwood procedure were 80% and 94%, respectively, which was a significant difference. The long-term survival rates of patients who survived the Norwood procedure were nearly equivalent between the groups, highlighting the importance of successful postnatal atrial septostomy.

It has been suggested that an intact/restrictive atrial septum can affect the prognosis of HLHS in the perioperative period and thereafter due to structural abnormalities in the pulmonary lymphatic and venous vessels caused by increased left atrial pressure. According to Canter et al.,[7] HLHS with restrictive ASD before or after heart transplantation is associated with high pulmonary vein pressure and an increased risk of mortality. In addition, Graziano et al.[8]

compared pathological pulmonary artery and lymphatic findings between restrictive ASD and non-restrictive ASDs. They reported significantly higher frequencies of pulmonary vein thickening and lymphatic dilation in the restrictive ASD group, which increased perioperative risks. It is essential to secure sufficient interatrial communication in patients with HLHS and an intact/restrictive ASD. In addition to Norwood procedure, resolving the early stage pulmonary congestion that accompanies an intact/restrictive atrial septum is considered to be an important factor in long-term survival.

Our treatment strategy for HLHS is to perform the Norwood procedure as the first palliation, except for high-risk patients such as low birth weight infants, severe tricuspid valve regurgitation or accompanied with total anomalous pulmonary venous return. In the high-risk patients, we perform bilateral pulmonary artery banding after administering prostaglandin E1, in order not to close the ductus arteriosus. We then quickly performed the Norwood procedure. The objective of first-stage palliation for infants with HLHS, including so-called hybrid procedures such as bilateral pulmonary artery banding with ductal stenting,[9-11] is to create an outflow tract to the systemic circulation that is free of stenosis, to appropriately control pulmonary blood flow, and to secure an interatrial communication free of stenosis. Regardless of the procedure, an intact/restrictive ASD can worsen the prognosis by causing pulmonary congestion. Therefore, our policy is to quickly perform BAS. The appropriate time for the initial surgery is considered to be after pulmonary congestion improves and high pulmonary blood flow is achieved. Our hospital has been performing a modified Norwood procedure using a right ventricle to pulmonary artery shunt, which we have used to manage 123 patients with HLHS (classical HLHS, 95 patients; HLHS variants, 28 patients) since February 1998. After excluding

5 patients who underwent biventricular repair, bidirectional Glenn (BDG) procedure completion/awaiting status was achieved in 91 (77%) of 118 remaining patients and Fontan completion/awaiting status was achieved in 83 (70%). [12]

Of the 70 patients in this cohort who underwent an interatrial intervention before the Norwood procedure, 8 underwent open BAS (11.4%) and 11 underwent catheter BAS (15.7%), for a total of 19 (27%). The outcomes in the open BAS group in infants with an IAS or birth weight <2 kg were markedly poorer than those in the catheter BAS group. Our study revealed that even with restricted interatrial communication or pulmonary congestion, when catheter BAS was possible, the therapeutic outcomes were approximately equivalent to patients who did not require BAS.

Gossett et al. reported that among patients with HLHS who underwent pre-Norwood interventions for ASD, 30 with an intact/restrictive ASD that required catheter therapy exhibited a BDG operation rate of 43% and a Fontan operation rate of 32%. Of the 7 patients with an IAS, BDG operation was achieved in 3 (43%) and Fontan operation was achieved in 2 (33%).[13]

Vlahos et al. reported that among 33 patients with intact/highly restrictive atrial septa who underwent interatrial communication interventions before the Norwood procedure, the Norwood operation rate was 38% and the survival rates at the ages of 1 month, 6 months, and 12 months were 52%, 42%, and 34%, respectively. These rates were significantly lower than those in a control group that did not require pre-Norwood interventions (91%, 79%, and 72%, respectively). Moreover, post-Glenn catheter examinations showed mean pulmonary pressures of 12.7 ± 6.5 mmHg and mean pulmonary vascular resistances of 1.9 ± 0.9 WU in the intact/highly restrictive atrial septum group. In the control group, the mean pulmonary pressure

was 12.6 ± 3.6 mmHg and the mean pulmonary vascular resistance was 1.9 ± 1.0 WU, with differences not being statistically significant.[14]

In our study, cardiac catheterization after the Glenn operation showed similar results in the non-BAS and BAS groups with respect to pulmonary pressure, pulmonary vascular resistance, and PAI. Moreover, pre-Norwood catheter interventions secured interatrial communications in restrictive ASD cases, which likely contributed to maintaining the survival rate and helping to achieve Glenn operation rates and post-Glenn catheter examination results that were similar to those of the non-BAS group.

It has been suggested that successful interventions for an intact/restrictive atrial septum in the fetal or neonatal period may improve long-term prognoses, although the morphology of the atrial septum in HLHS often limits the therapeutic options. In our results, 4 of the 11 patients (36%) who underwent catheter BAS needed to undergo static BAS prior to Rashkind BAS. Excluding the patient with cor triatriatum, 5 of these 10 patients (50%) were categorized as standard ASD morphology and 5 (50%) were categorized as a complex ASD. These ratios resemble those reported by Holter et al. Of the 67 patients they examined, 39 (58%) had standard ASDs and 28 (42%) had complex ASDs.[2] In our study, the initial Rashkind BAS was difficult in 4 of the 5 patients with complex ASDs. However, in all 4 patients, performing static BAS before the Rashkind BAS achieved Fontan operation or awaiting, which produced safe and sufficient interatrial communication.

There has been significant advancement in fetal catheter interventions in recent years. Vida et al. reported a 64% 6-month survival rate for patients who underwent interventions for an intact or highly restrictive atrial septum in the fetal period.[15] The pulmonary artery pressure and

pulmonary vascular resistance of the patients who underwent fetal therapy and survived the neonatal period were equivalent to those of the control group. This suggests that therapeutic interventions in the fetal period can promote the development of pulmonary vessels and lung tissue, and improve both short-term and long-term prognoses.

Although different institutions use a variety of devices for BAS, Andrew et al. reported that in 16 HLHS patients with restrictive ASD, the Brockenbrough atrial septostomy (including stent placement) effectively increased SpO_2 from 50% to 83% and lowered the mean interatrial pressure gradient from 16 mmHg to 1 mmHg. Hoqoe et al.[6] reported 20 cases of atrial septostomy, including 11 where radio frequency (RF) wires were used. They reported that the left atrial pressure went from 21 mmHg to 11 mmHg. After performing static BAS, Rashkind BAS, and stent implantation, significant improvements in mean interatrial pressures (17 mmHg to 4 mmHg) and SpO_2 levels (72% to 85%) were observed.

RF wires were not used in any of the patients in the present study partly because they were only recently approved in Japan (in 2014). Nevertheless, expanding the range of available therapeutic techniques is a promising development and further studies will be needed to determine how to appropriately utilize them.

Conclusion

Although infants with HLHS with a restrictive atrial septum are considered to have poor prognoses, the group that underwent catheter BAS had an acceptable prognosis and no significant differences in post-Glenn catheter data compared with the non-BAS group. Compared with infants with a standard atrial septum, infants with a complex atrial septum can

achieve equivalent BAS outcomes despite a larger proportion of them needing static BAS prior to Rashkind BAS.

Conflicts of interest

The authors declare no conflicts of interest.

Author contributions

Y.F. designed the study and wrote the manuscript; K.B. designed the study; M.K., Y.K., T.K., Y.S., K.H., K.T., T.I., S.K., Y.K., and S.O. gave technical support. All authors read and approved the final manuscript.

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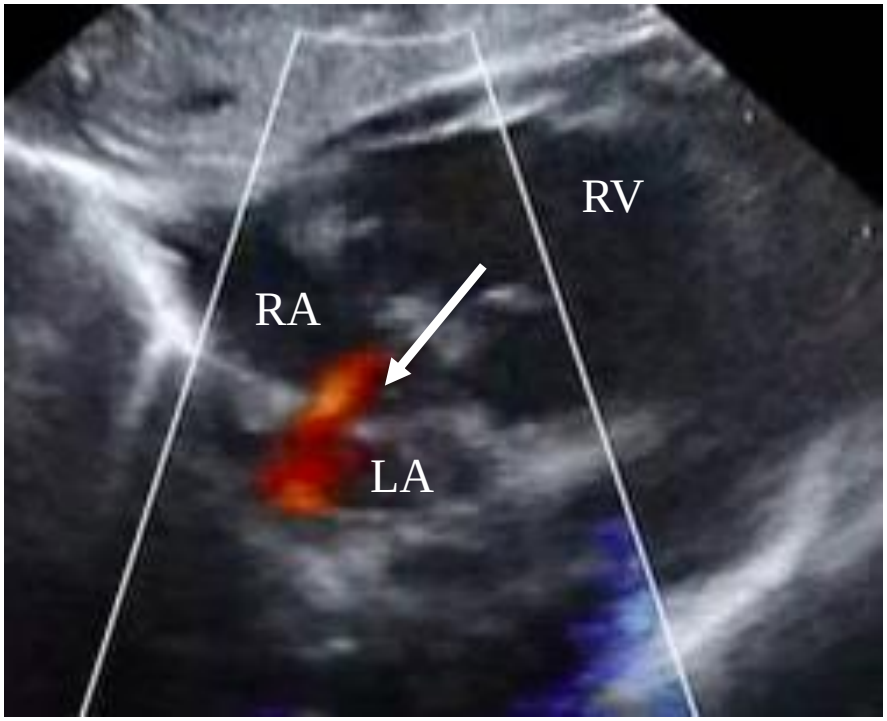
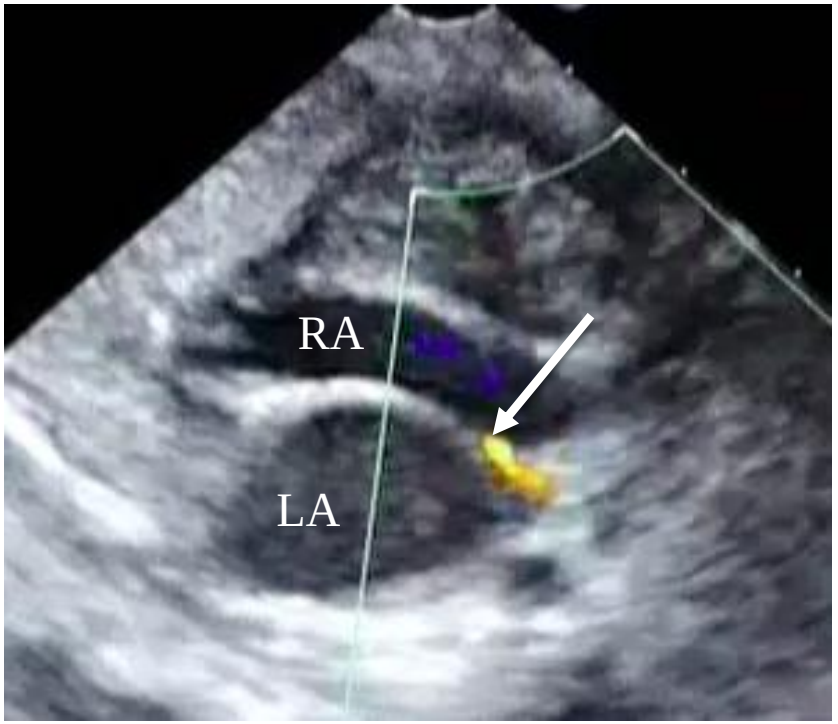
Standard ASD



Complex ASD



RA: right atrium; LA: left atrium; RV: right ventricle

Fig. 1

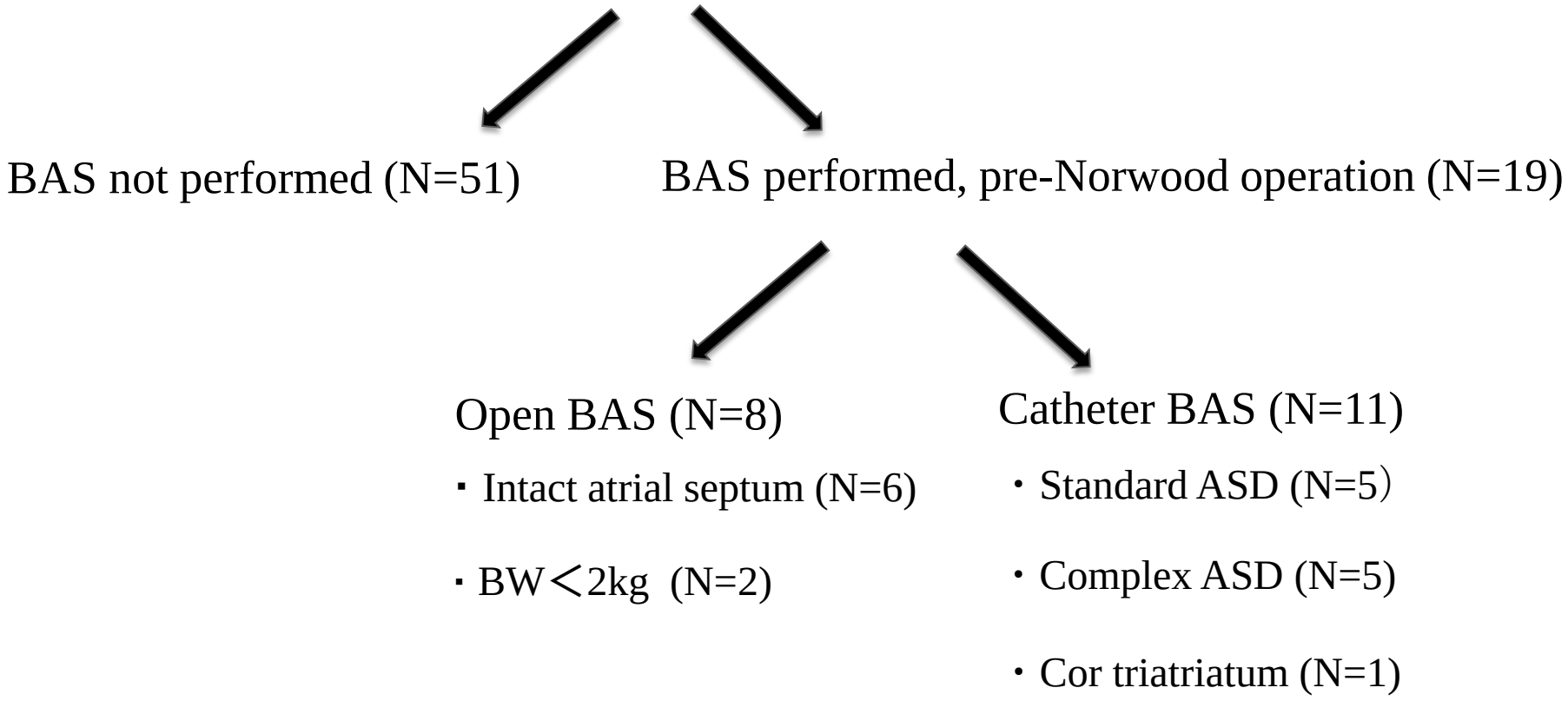

Infants enrolled in the study (N=70)
BAS not performed (N=51)
BAS performed, pre-Norwood operation (N=19)
Open BAS (N=8)
・Intact atrial septum (N=6)
・BW＜2kg (N=2)
Catheter BAS (N=11)
・Standard ASD (N=5)
・Complex ASD (N=5)
・Cor triatriatum (N=1)

Fig. 2

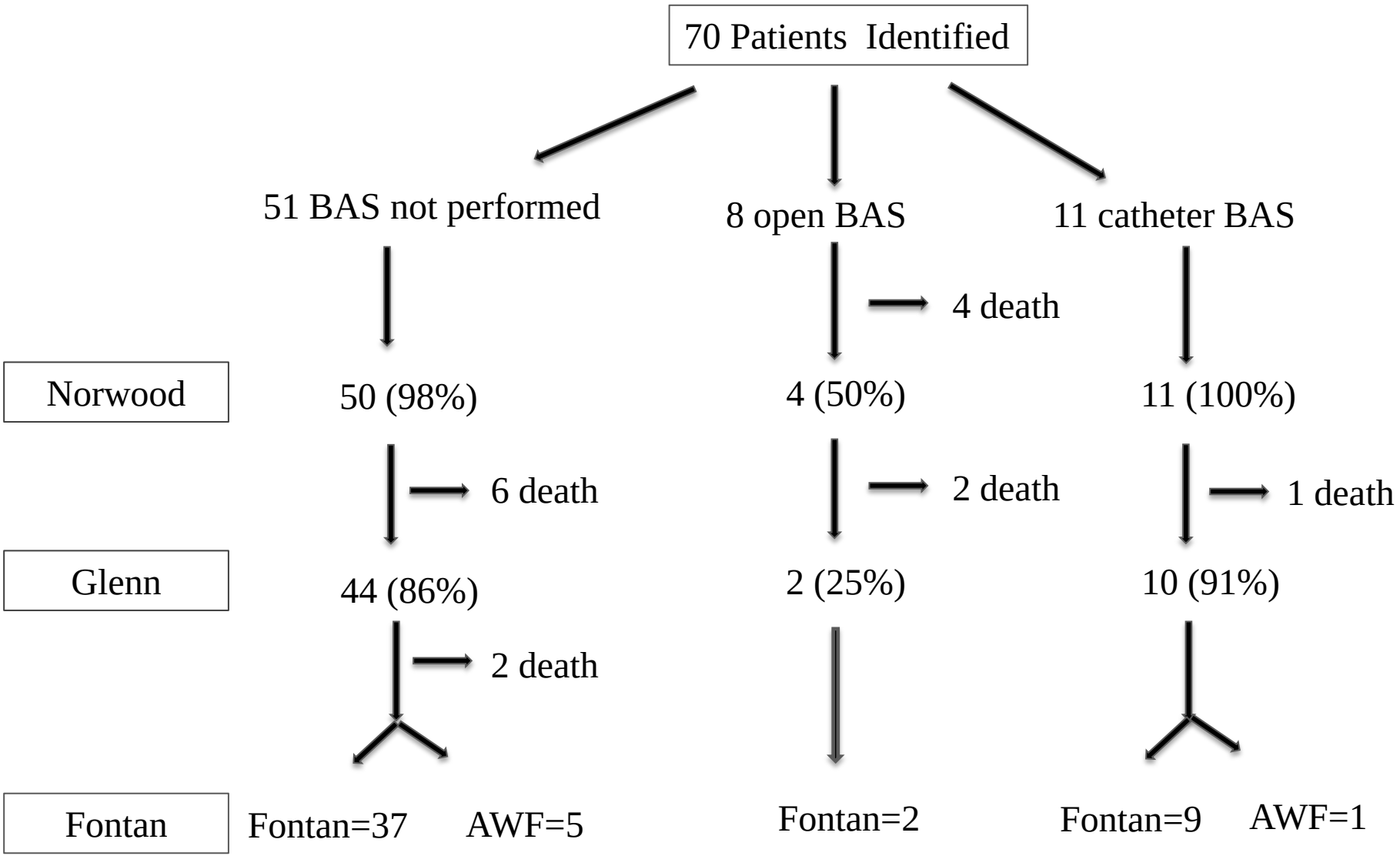

70 Patients Identified
51 BAS not performed
8 open BAS
11 catheter BAS
4 death
Norwood
50 (98%)
4 (50%)
11 (100%)
6 death
2 death
1 death
Glenn
44 (86%)
2 (25%)
10 (91%)
2 death
Fontan
Fontan=37
AWF=5
Fontan=2
Fontan=9
AWF=1

Fig. 3

	Sex	Subtype	Age (days)	Weight (kg)	ASD type	BAS
1	f	MA/AA	5	2.5	standard	Rashkind
2	m	MS/AA	10	3.4	standard	Rashkind
3	m	MS/AA	1	2.7	standard	Rashkind
4	m	MA/AA	41	2.2	complex	Rashkind
5	m	MS/AS	7	2.3	standard	Rashkind
6	m	MA/AA	1	3.5	standard	Rashkind
7	m	MS/AA	50	3.1	complex	S→R
9	f	MA/AA	1	2.5	complex	S→R
9	m	MA/AA	0	3.1	complex	S→R
10	m	MA/AA	0	3.5	complex	S→R

AA = aortic atresia; AS = aortic stenosis; MA = mitral atresia; MS = mitral stenosis
S→R = Static→Rashkind

Table 1. Clinical Characteristic

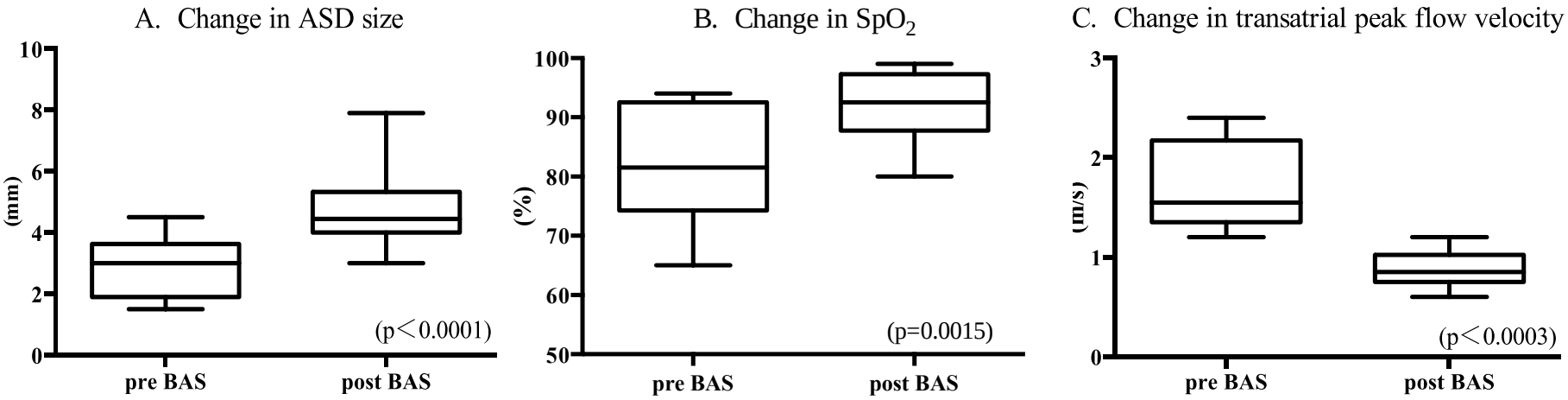

A. Change in ASD size
(mm)
10
8
6
4
2
0
(p＜0.0001)
pre BAS
post BAS
B. Change in SpO2
(%)
100
90
80
70
60
50
(p=0.0015)
pre BAS
post BAS
C. Change in transatrial peak flow velocity
(m/s)
3
2
1
0
(p＜0.0003)
pre BAS
post BAS

Fig. 4

	standard ASD group (N=5)	complex ASD group (N=5)	p-value
weight (kg)	2.9	2.9	＞0.99
pre ASD size (mm)	3.4	2.4	0.071
pre flow (m/s)	1.6	1.8	0.44
pre SPO2 (%)	83	81	0.88
Rashkind BAS	5	1	0.048
Static→Rashkind BAS	0	4	
post ASD size(mm)	5.3	4.2	0.31
post flow (m/s)	0.8	0.9	0.47
post SPO2 (%)	93	90	0.76

Table 2. Comparison data between standard ASD group and complex ASD group

1

Figure legends

Fig. 1. Two examples of standard atrial septal anatomy and complex atrial septal anatomy.
Note the central ASD location and the thin atrial septum in the standard ASD in the left panel. The ASD is marked by an arrow (LA→RA flow; red color flow) Note the superior ASD location in the right panel. The ASD is marked by an arrow (LA→RA flow; red color flow)

Fig. 2. Flow chart for infants' selection in this study (2006 – 2015). BAS = balloon atrial septostomy

Fig. 3. Procedures performed

Flow chart of outcomes for HLHS: no BAS performed, open BAS performed, and catheter BAS performed.

(BAS = balloon atrial septostomy, AWF = awaiting Fontan procedure)

Fig. 4. Procedural results after BAS

A: ASD size changed from 3.2 ± 1.1mm to 4.7±1.3mm ($p < 0.0001$)

B: Peripheral oxygen saturation changed from 81 ± 9% to 92 ± 6 % ($p < 0.0015$)

C: Peak velocity across the ASD seen on transthoracic echocardiography changed from 1.6 ± 0.4m/s to 0.8 ±0.2m/s ($p < 0.0003$)