

Perioperative anesthetic management of patients with hypoplastic left heart syndrome undergoing the comprehensive stage II surgery—A review of 148 cases

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Abstract

Background: Patients with hypoplastic left heart syndrome undergo the comprehensive stage 2 procedure as the second stage in the hybrid approach toward Fontan circulation. The complexity of comprehensive stage 2 procedure is considered a potential limitation, and limited information is available on its anesthetic management. This study aims to address this gap.

Methods: A single-center retrospective cohort study analyzed 148 HLHS patients who underwent comprehensive stage 2 procedure, divided into Group A (stable condition, $n = 116$) and Group B (requiring preoperative intravenous inotropic therapy, $n = 32$). Demographic data, intraoperative hemodynamics, anesthetic management, and postoperative outcomes were collected.

Results: Etomidate (40%) was the most common induction agent, followed by esketamine (24%), midazolam (16%), and propofol (13%). Inhaled induction was rarely necessary (2%), occurring only in Group A patients. No statistical differences were found between groups for induction drug choice. Post-cardiopulmonary bypass management included moderate hypoventilation, inhaled nitric oxide (100%), and hemodynamic support with milrinone (97%) and norepinephrine (77%). Group B patients more frequently required additional levosimendan (20%) and epinephrine (18%). Extracorporeal membrane oxygenation was necessary in 8 patients (5%) with no between-group differences. Switching from fentanyl to remifentanyl reduced postoperative ventilation time overall. However, Group B experienced significantly longer ventilation (6.3 vs. 3.5 h) and ICU stay (22 vs. 14 days). In-hospital mortality was 5% overall (Group A: 4%, Group B: 9%). Long-term survival analysis revealed a significant advantage for Group A.

Conclusion: The use of short-acting opioids and adjusted ventilation modes enables optimal pulmonary blood flow and rapid transition to spontaneous breathing. Differentiated hemodynamic support with milrinone, norepinephrine, supplemented by levosimendan and epinephrine in high-risk patients, can mitigate the effects on

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the preoperatively volume-loaded right ventricle. However, differences in long-term survival probability were observed between groups.

Trial Registration: Local ethics committee, Medical Faculty, Justus-Liebig-University-Giessen (Trial Code Number: 216/14).

KEYWORDS

anesthesia, comprehensive stage II, congenital heart surgery, hybrid procedure, hypoplastic left heart syndrome, infants, outcome

1 | INTRODUCTION

Twenty-five years ago, the hybrid approach was introduced at the Giessen Pediatric Heart Center as first stage procedure for newborns with hypoplastic left heart syndrome (HLHS). The Giessen hybrid approach consist of bilateral surgical pulmonary artery banding followed by percutaneous arterial duct stenting.¹ From the beginning, the aim of the hybrid procedure was to postpone extensive surgery (Norwood-operation) from the vulnerable neonatal period to later infancy to decrease mortality and morbidity particularly neurological sequelae.

The comprehensive stage 2 procedure (cS2P) is the second step on this pathway to Fontan circulation and includes de-banding of the pulmonary arteries with patch augmentation if needed, removal of the stent and closure of the patent ductus arteriosus, reconstruction of the aortic arch and construction of a superior cavo-pulmonary connection.² The relative complexity of the cS2P compared to a standard superior cavo-pulmonary connection after Norwood procedure is considered as a potential limitation of hybrid strategies.³

Furthermore the interstage I period is particularly vulnerable⁴ and may lead to heart failure of the single ventricle in unfavorable cases, necessitating hospitalization, and therapy with positive inotropic drugs before cS2P. It has been proposed to avoid the cS2P in those patients⁵ although proceeding to cS2P allows unloading of the single ventricle and may therefore be useful despite a higher perioperative risk. Because only limited information is available about the perioperative anesthetic management of patients undergoing the cS2P⁶⁻⁸ we present a detailed analysis of the anesthetic management of infants with HLHS undergoing the cS2P at our institution. Furthermore, we compared patients who were decompensated during the interstage 1 period and required intravenous positive inotropic medication at the time of cS2P with standard patients.

The primary outcome was to describe the anesthetic management of patients undergoing cS2P for HLHS. Secondary outcomes were postoperative use of extracorporeal membrane oxygenation, duration of mechanical ventilation, time at the intensive care unit and overall mortality. This study fills a gap in the literature by providing a comprehensive analysis of anesthetic management for cS2P, which could inform best practices and potentially improve outcomes for HLHS patients.

2 | METHODS

After positive approval by the local ethics committee (Trial Code Number 216/14), a retrospective data-analysis was performed. The examined database contains data from all surgeries performed at the Giessen Pediatric Heart Center between December 14, 1998 and December 31, 2021. We extracted and analyzed the perioperative data, including patients' characteristics, procedural times, vital data, laboratory values, drug doses, and transfusion data from the in-house patient data management system (PDMS; ICUData®, IMESO GmbH, Giessen, Germany). Furthermore the "HLHS-Database" of the Giessen Pediatric Heart Center was used for surgical details, interventional and mortality data.

Patients were categorized into two groups based on their preoperative condition:

Group A:

- Stable patients arriving for planned elective comprehensive stage 2 procedure (cS2P).

- Admitted to the operating room as scheduled.

- No intravenous inotropic support required.

Group B:

- Patients requiring intravenous inotropic support.

- Unplanned hospital admission during the interstage 1 period because of clinical deterioration at home.

- Significant right ventricular function decline and increasing tricuspid valve regurgitation, necessitating positive inotropic therapy.

2.1 | Statistics

Demographic and procedural information were analyzed by SPSS (Statistics Version 27). Categorical variables are presented as number and percentage, while continuous data are presented as mean ± standard deviation. Between-group differences for categorical data were tested using the chi-square and Fisher exact test. Continuous data were first analyzed for normal distribution using the Shapiro-Wilk test, and then either the *t*-tests or Mann-Whitney *U*-test/Wilcoxon rank test were performed. Survival analysis was performed using Kaplan-Meier curves and compared using the log rank test. Unless specifically stated otherwise, the results mentioned apply to the overall cohort of patients being discussed.

3 | RESULTS

A total of 172 patients who underwent stage II surgery after initial hybrid approach for HLHS were identified. However, five patients were excluded because they underwent a Norwood Damus–Kaye–Stansel procedure plus modified Blalock–Taussig-shunt due to absent inferior vena cava with azygos continuation. Additionally, 15 patients were excluded because of incomplete perioperative data. Ultimately 148 patients undergoing cS2P were included in the presented study (Figure 1).

Out of the 148 patients undergoing cS2P, 116 patients (78%) presented under stable conditions (Group A), while 32 patients (22%) required intravenous inotropic support after an unplanned hospital admission prior to cS2P (Group B). The two groups were comparable in terms of biometric and procedural factors, including the presence of specific risk factors like aortic atresia (Table 1).

3.1 | Anesthetic and hemodynamic management

3.1.1 | Anesthesia

In non-sedated patients, intravenous midazolam (0.1 mg/kg) was used to facilitate separation from parents when necessary. For anesthesia induction in the entire cohort, the most commonly used agents were etomidate (59/148, 40%), followed by esketamine

(36/148, 24%), midazolam (24/148, 16%), and propofol (19/148, 13%). Induction was preceded by an opioid and followed by cisatracurium before endotracheal intubation. Inhaled induction was necessary in only three patients (2%), all belonging to Group A. There were no statistical differences between the subgroups for induction drugs.

Initially, fentanyl was used as the opioid for anesthesia. However, since 2008, it has been replaced by remifentanyl, resulting in a shorter duration of postoperative mechanical ventilation time in the overall patient cohort (mean 4 h, 95% CI 3–5 h vs. 5 h, 95% CI 3–7 h, $p = .005$). The opioid-based anesthesia was supplemented with intermittent boluses of midazolam and, from 2007 onwards, additionally with central alpha-2-receptor agonists (clonidine/dexmedetomidine). In 82% of patients, isoflurane or sevoflurane was used before cardiopulmonary bypass (CPB). Although changes in anesthetic agents occurred over time, no difference was observed between the two patient groups (Table 2).

3.1.2 | Intraoperative monitoring

Perioperative monitoring consisted of standard non-invasive ASA monitoring. Two-site NIRS monitoring (cerebral and flank) was added. After inducing anesthesia, we establish comprehensive vascular access and monitoring. Two arterial catheters are inserted: a 24G catheter in the right radial or brachial artery for

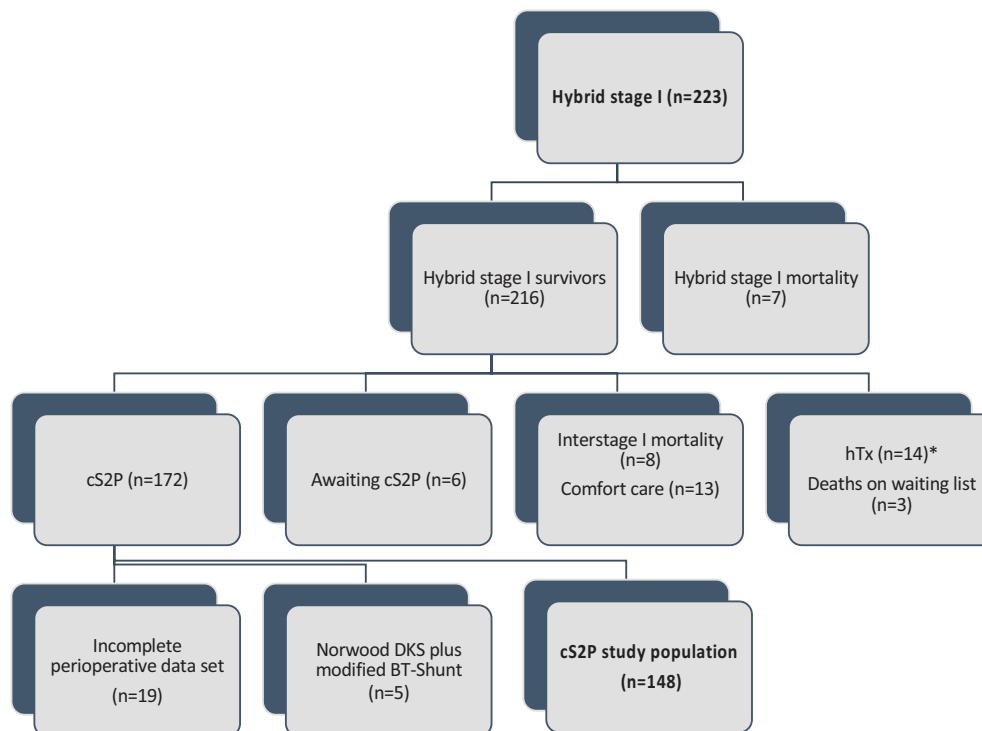


FIGURE 1 Participant flow diagram including all patients undergoing the hybrid approach for hypoplastic left heart syndrome from July 1998 to December 2021. BT, Blalock-Taussig; CS2P, comprehensive stage II surgery; DKS, Damus-Kaye-Stansel-Procedure; hTx, heart transplantation. *Thirteen patients underwent primary transplant protocol including three patients who died on the waiting list. Four additional patients originally scheduled for stage palliation underwent cardiac transplantation during interstage I.

TABLE 1 Patients characteristics and process times.

Characteristics	Group A	Group B
	(stable) (n = 116)	(unstable) (n = 32)
Mean age—months	5.0 ± 2.3	4.2 ± 1.3
Mean weight—kg	5.6 ± 1.0	5.2 ± 0.6
Male sex—no. (%)	75 (65)	20 (63)
HLHS with aortic atresia—no. (%)	50 (43)	16 (50)
Mean preoperative oxygen saturation—%	83 ± 6	85 ± 6
Mean preoperative hemoglobin—g/L	12.6 ± 2.0	12.2 ± 1.4
Mean preoperative lactate—mmol/L	0.7 ± 0.6	0.8 ± 0.7
Preoperative inotropic support—no. (%)	0 (0)	32 (100)*
Preoperative milrinone infusion—no. (%)	0 (0)	26 (81)*
Preoperative levosimendan infusion—no. (%)	0 (0)	16 (50)*
Preoperative norepinephrine infusion—no. (%)	0 (0)	1 (3)
Preoperative epinephrine infusion—no. (%)	0 (0)	2 (6)**
Mean surgical procedure time—(min)	391 ± 53	410 ± 82
Mean time on CPB—(min)	244 ± 46	263 ± 41
Mean aortic cross clamp time—(min)	64 ± 31	70 ± 40
Mean selective cerebral perfusion time—(min)	68 ± 26	71 ± 19

Abbreviations: CPB, cardiopulmonary bypass; HLHS, hypoplastic left heart syndrome.

* $p = .000$ versus Group A. ** $p = .046$ versus Group A.

upper body monitoring, and a 22G/20G catheter in a femoral artery for lower body monitoring. Additionally, we place two double lumen 4F/5F central venous catheters, one each for the upper and lower body. These are inserted percutaneously if not already present preoperatively.

We generally try to avoid surgically placed intracardiac catheters. However, the catheter positioning is crucial for monitoring key pressures. The catheter in the superior vena cava measures pulmonary artery pressure after constructing the superior cavopulmonary connection, while the one in the inferior vena cava measures ventricular filling pressure. The difference between these pressures indicates the transpulmonary pressure gradient, which ideally should be less than 10 mmHg. The arterial access points serve specific monitoring purposes. The right-sided upper extremity measurement is essential for assessing blood pressure beyond the aortic arch and during periods of antegrade cerebral perfusion. When combined with the femoral artery pressure reading, this setup allows us to detect any preoperative or postoperative obstructions in the aortic arch.

This comprehensive monitoring approach enables us to closely track the patient's hemodynamic status throughout the procedure and in the immediate postoperative period.

TABLE 2 Intraoperative drug therapy, transfusion therapy, coagulation management, and ECMO support.

Characteristics	Group A	Group B
	(stable) (n = 116)	(unstable) (n = 32)
Anesthesia		
Fentanyl—no. (%)	25 (22)	8 (25)
Remifentanyl—no. (%)	92 (79)	25 (78)
Mean total fentanyl dose— $\mu\text{g}/\text{kg}$	111 ± 41	123 ± 66
Mean total remifentanyl dose— $\mu\text{g}/\text{kg}/\text{min}$	0.6 ± 0.3	0.6 ± 0.3
Mean total midazolam dose— mg/kg	0.7 ± 0.3	0.8 ± 0.4
Clonidine—no. (%)	83 (72)	18 (56)
Dexmedetomidine—no. (%)	21 (18)	10 (31)
Volatile anesthetics—no. (%)	97 (84)	25 (78)
Hemodynamic support at the end of surgery		
Milrinone—no. (%)	113 (97)	31 (97)
Milrinone mean dose— $\mu\text{g}/\text{kg}/\text{min}$	0.7 ± 0.2	0.8 ± 0.2
Norepinephrine—no. (%)	92 (79)	22 (69)
Norepinephrine mean dose— $\mu\text{g}/\text{kg}/\text{min}$	0.1 ± 0.1	0.1 ± 0.1
Epinephrine—no. (%)	15 (13)	12 (38)*
Epinephrine mean dose— $\mu\text{g}/\text{kg}/\text{min}$	0.02 ± 0.07	0.04 ± 0.07*
Levosimendan—no. (%)	9 (8)	20 (63)**
Levosimendan mean dose— $\mu\text{g}/\text{kg}/\text{min}$	0.0 ± 0.04	0.1 ± 0.08**
Vasopressin/terlipressin—no. (%)	3 (2.6)	1 (3.1)
Inhaled nitric oxide mean dose—ppm	25 ± 9	28 ± 10
ECMO—no. (%)	5 (4.3)	3 (9.4)
Transfusion therapy (including priming volume of cardiopulmonary bypass)		
Mean packed red cells—mL/kg	139 ± 47	150 ± 50
Mean fresh frozen plasma—mL/kg	61 ± 25	66 ± 36
Mean platelet infusion—mL/kg	31 ± 13	33 ± 13
Coagulation management		
FC—no. (%)	86 (74)	26 (81)
PCC—no. (%)	75 (65)	23 (72)
FXIII—no. (%)	26 (22)	9 (28)
rFVIIa—no. (%)	12 (10)	7 (22)

Abbreviations: ECMO, extracorporeal membrane oxygenation; FC, fibrinogen concentrate; FXIII, factor XIII concentrate; PCC, 4 factor prothrombin complex concentrate, rFVIIa, recombinant activated factor VII.

* $p = 0.003$ versus Group A. ** $p = 0.000$ versus Group A.

3.1.3 | Concomitant medication

In all patients, dexamethasone was administered at a total dose of 1.0–1.5 mg/kg. A single dose of phentolamine (0.25 mg/kg) was

given to facilitate cooling to 28°C, whereas a continuous infusion of sodium nitroprusside (0.5–1.0 µg/kg/min) was used during rewarming.

3.1.4 | Ventilation therapy

After carefully suctioning the airway during rewarming, the surgeon restores blood flow through the superior cavo-pulmonary connection once the temperature reaches 35°C. Ventilation is then initiated with 80% oxygen and 20–40 ppm of inhaled nitric oxide (NO). The ventilation is adjusted to maintain a mean airway pressure of a maximum of 10 mmHg, and moderate, compensated hypoventilation is achieved after discontinuation of CPB (Table 3).

3.1.5 | Inotropic, vasoactive, and ECMO support

After weaning off cardiopulmonary bypass, all patients required inotropic and vasoactive support. Intravenous milrinone was administered to 97% of patients, and norepinephrine to 77% of patients, with no difference between the subgroups. Levosimendan was used in 20% of patients and epinephrine in 18% of patients with both being used more frequently in Group B patients. Vasopressin or terlipressin was given to 4 patients (3%) and extracorporeal membrane oxygenation (ECMO) support was required in 8 patients (5%) with a 90-days survival of 50%. However, no difference was noted between the subgroups with regarding the use of vasopressin/terlipressin, or ECMO support.

The intraoperative transfusion therapy and coagulation management are shown in Table 2, with no observed difference between the subgroups.

3.2 | Hemodynamic and oxygenation data

The hemodynamic and oxygenation data, including cerebral and somatic near-infrared spectroscopy (NIRS) and lactate levels at the end of surgery, are presented in Figure 2 and Table 3. Except for somatic NIRS values, where patients in Group B had lower readings, no significant differences were observed between the subgroups.

3.3 | Outcome data

Table 3 shows the main outcome data. No differences were found between the subgroups regarding in-hospital survival. A total of eight patients (5%) died during the hospital stay, with five patients (4%) belonging to Group A and three patients (9%) belonging to Group B. However, Group B patients experienced significantly prolonged duration of mechanical ventilation and intensive care unit

TABLE 3 Postoperative hemodynamic, oxygenation, and outcome data.

Characteristics	Group A (stable)	Group B (unstable)
	(n = 116)	(n = 32)
Hemodynamic data at the end of surgery		
Mean heart rate—beats/min	146 ± 16	142 ± 15
Mean arterial pressure—mmHg	52 ± 8	54 ± 7
Mean pulmonary artery pressure—mmHg	16 ± 4	16 ± 5
Central venous pressure—mmHg	8 ± 4	8 ± 4
Oxygenation parameter at the end of surgery		
Mean hemoglobin—g/L	13.1 ± 2.2	13.2 ± 2.0
Mean arterial pCO ₂ —mmHg	52 ± 12	53 ± 13
Mean arterial oxygen saturation—%	79 ± 10	74 ± 15
Mean venous oxygen saturation—%	47 ± 15	46 ± 15
Mean cerebral oxygen saturation—%	57 ± 11	56 ± 10
Mean somatic oxygen saturation flank—%	69 ± 11	63 ± 12*
Mean lactate—mmol/L	3.5 ± 2.1	3.0 ± 2.1
Outcome data		
Mean duration of mechanical ventilation—(h)	3.5 ± 5.6	6.3 ± 6.5**
Mean intensive care stay—(days)	14 ± 12	22 ± 20***
In-hospital survival—no. (%)	111 (96)	29 (91)

p* = .018 versus Group A. *p* = .003 versus Group A. ****p* = .030 versus Group A.

stay. Furthermore, overall survival after cS2P showed a significant survival advantage for Group A patients (Figure 3).

4 | DISCUSSION

To the best of our knowledge there is only limited information on the anesthesiologic management in patients undergoing the cS2P.

Our intraoperative management has evolved over time, focusing on optimizing systemic oxygen delivery and perfusion pressure during anesthesia induction and in the pre-cardiopulmonary bypass period. Initially, we used high-dose fentanyl in combination with a muscle relaxant and midazolam. Subsequently, we switched to remifentanyl in combination with an induction dose of etomidate, propofol, or esketamine to ensure adequate depth of anesthesia and allow for more timely tracheal extubation after surgery. Interestingly, there was no significant difference between the subgroups in the choice of induction drugs, supporting the view that the choice of hypnotic agent is less critical than careful attention to the hemodynamic consequences of the selected technique.

The physiological state immediately after cS2P is unique, and it requires the anesthesiologist to have a thorough understanding of the effects of prolonged cardiopulmonary bypass (CPB) time

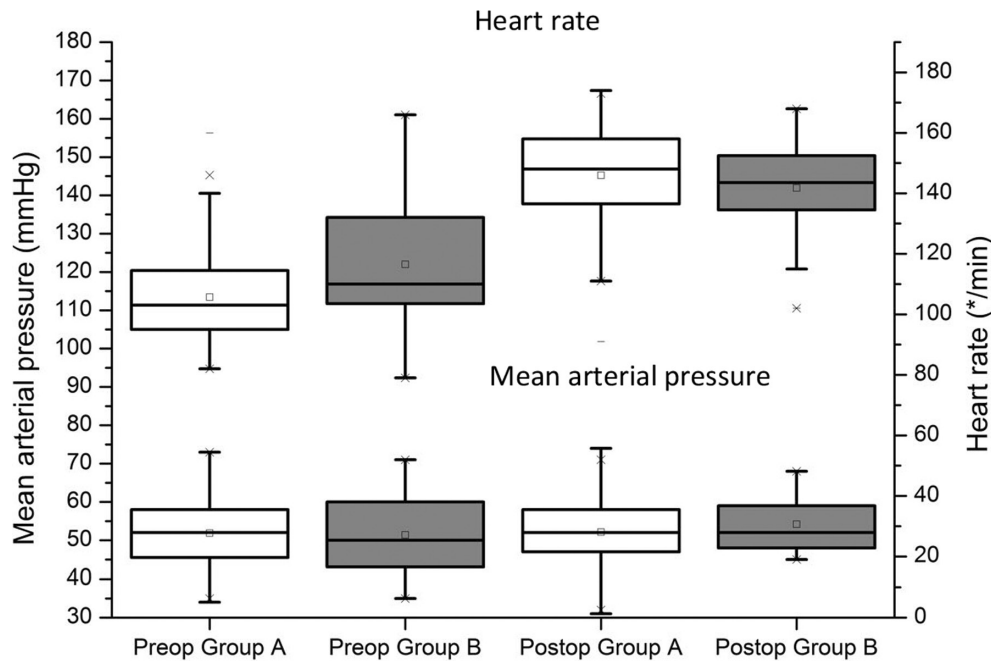


FIGURE 2 Heart rate and mean arterial blood pressure after induction of anesthesia (preop) and before transfer to the intensive care unit (postop).

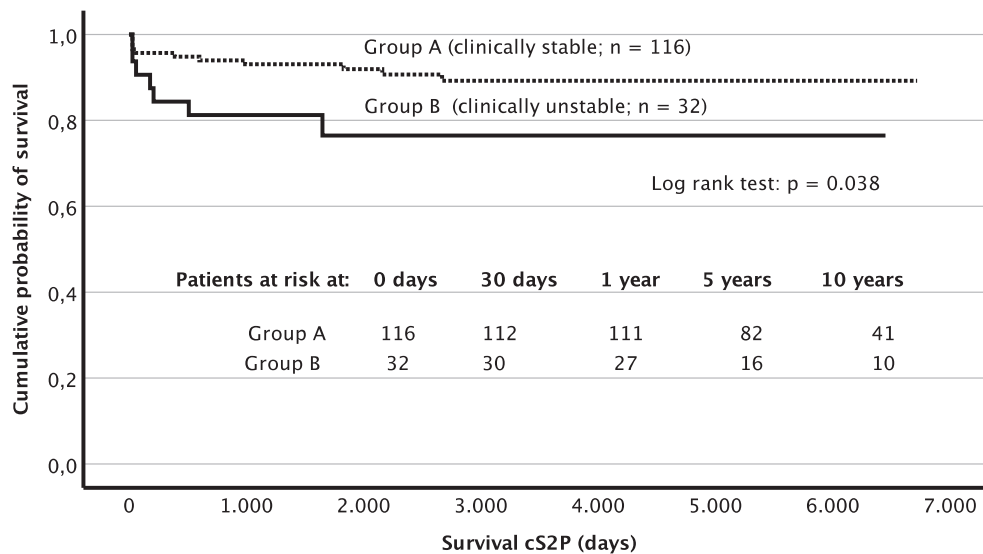


FIGURE 3 Kaplan-Meier survival curves for comprehensive stage 2 surgery. CS2P, comprehensive stage 2 surgery.

during deep hypothermia for aortic arch repair, in conjunction with passive pulmonary perfusion via the superior cavo-pulmonary connection at the end of surgery. Measures that increase upper body blood flow and decrease pulmonary vascular resistance (PVR) can improve oxygenation after weaning off CPB. To limit the negative effects of mechanical ventilation on pulmonary blood flow, moderate hypoventilation was targeted. This involved maintaining PaCO₂ values around 50 mmHg and a positive base excess. Additionally, to keep the mean airway pressure no higher than 10 mmHg, larger tidal volumes (10–12 mL/kg) were used. These tidal volumes, along with lower respiratory rates, differ from those

typically recommended for neonates and infants.⁹ Additionally, inhaled nitric oxide (NO) was administered preventively to minimize the negative effects of non-pulsatile pulmonary blood flow and hypercapnia on PVR.¹⁰ While this ventilation strategy causes cerebral oxygen saturation to overestimate central venous oxygen saturation,¹¹ somatic oxygen saturation measured in the renal cortex may gain significance. In contrast to the brain, the kidney is a high-flow, low-extraction organ under intense sympathetic control.¹² A decrease in somatic oxygen saturation may therefore indicate an early shock state and is associated with worse outcome after pediatric cardiac surgery.¹³ Comparable to classical Norwood surgery,

maintaining somatic tissue oxygen saturation >60% could serve as a reasonable target value following cS2P.¹⁴ Remifentanyl combined with dexmedetomidine or clonidine can accelerate the transition to spontaneous breathing, which also has a favorable effect on the passive pulmonary blood flow.¹⁵

A standardized approach to inotropic therapy is prudent, given the frequent single ventricle dysfunction due to chronic volume overload and extended bypass and cross-clamping times. Group B patients, required more intensive hemodynamic support post-CPB, including levosimendan and low-dose epinephrine in addition to the standard milrinone and norepinephrine protocol. We initiate a milrinone infusion at 0.5 µg/kg/min after anesthesia induction, even for standard patients (Group A), to achieve adequate drug levels without bolus administration before weaning off CPB. Administering a phosphodiesterase type III inhibitor like milrinone before cross clamping of the aorta increases cAMP in the myocardium aiding in bypass weaning.¹⁶ Norepinephrine is added before weaning off CPB if the perfusion pressure during full flow is lower than 40 mmHg, despite hemoglobin levels exceeding 12 g/L. Right ventricular contractility is continuously monitored using transthoracic echocardiography. If reduced contractility is evident, milrinone is increased first, up to a maximum dose of 1.0 µg/kg/min. Levosimendan at 0.1 µg/kg/min and epinephrine are the second and third-line inotropic agents, respectively. Given the down-regulation of beta-1 receptors in the right ventricle in hypoplastic left heart syndrome, combining positive inotropic substances with different molecular mechanisms of action is a rational approach.¹⁷ When right ventricular contractility remains inadequate despite maximum epinephrine dosage of >0.05 µg/kg/min and a norepinephrine dosage of >0.15 µg/kg/min, we consider this an indication for ECMO support. This strategy acknowledges the complex pathophysiology of the condition and provides a clear threshold for escalating to mechanical circulatory support when pharmacological interventions prove insufficient.

Our standard drug combination allows for a certain inotropic support without a pronounced chronotropic effect. A mean heart rate of 140–150 beats per minute after weaning off bypass is acceptable, but further increases should be avoided in a single ventricle working with limited arterial oxygen saturation values. Especially under these pathophysiological conditions, when oxygen delivery cannot be further increased, additional chronotropic effects should be minimized to prevent myocardial ischemia.¹⁸ Under such pathophysiological conditions, a reduction in oxygen consumption by achieving an optimal heart rate, often referred to as the “best heart rate,” seems to be a reasonable approach. Ultra-short-acting, highly selective beta-1 receptor blockers may play a pivotal role in this regard in the future. This potential role is particularly relevant since selective beta-1 receptor blockers are already part of the standard therapy during the interstage 1 period at our center.⁴

Employing an opioid-based anesthetic regimen in conjunction with central alpha-2 receptor agonists may aid in heart rate modulation. However, our observations did not reveal a statistically significant difference in mean heart rate before and after incorporating

central alpha-2 receptor agonists into our standard anesthetic protocol. This finding could be partly attributable to the impairment of autonomic nervous control that is known to occur in patients with congenital heart disease especially after cardiac surgery involving prolonged CPB times.^{19,20}

Coagulation therapy after cS2P presents a challenge due to the high risk of bleeding complications from the complex surgical procedure and the significant risk of thromboembolic events caused by reduced blood flow velocities in the superior cavo-pulmonary connection and left main pulmonary artery.² To achieve sufficient hemostasis, most patients received coagulation factor concentrates alongside platelets and fresh frozen plasma. Fibrinogen concentrate was the most common (76%), followed by 4-factor prothrombin complex concentrate (66%), and factor XIII concentrate (24%). Activated factor VII was used in 13% of patients. Overall, no difference was observed between the groups.

Our retrospective single-center analysis demonstrates that cS2P can achieve a 95% in-hospital survival rate, even for patients in unstable preoperative conditions requiring continuous inotropic support before surgery. However, these patients experienced more complicated postoperative courses, with longer ventilation times and intensive care unit stays. The difference in long-term outcomes between the subgroups suggests that preoperative cardiac decompensation may result from both functional causes (e.g., ductus-stent stenosis) and structural myocardial damage due to ischemia (e.g., isthmus coarctation).² This hypothesis is supported by the observation that 12 of 18 (67%) deceased patients post-cS2P had aortic atresia, a condition associated with the highest risk of myocardial ischemia during previous treatment stages due to retrograde blood flow in the ascending aorta. A recent multicenter study analyzing data from The Society of Thoracic Surgeons Congenital Heart Surgery Database reported a surgical mortality rate of up to 12% in 141 patients undergoing cS2P, with no obvious modifiable variables noted between survivors and non-survivors.²¹ However, this study provided limited information about preoperative physiological status. Other research suggests that cS2P outcomes can be improved by avoiding surgery on patients requiring hospitalization for heart failure beforehand.⁵ Nonetheless, the superior cavo-pulmonary connection should theoretically allow volume unloading in a single ventricle for the first time.

5 | LIMITATIONS

This study has several limitations. First, its retrospective nature may introduce bias. Second, changes in anesthetic practices over the study period may confound results. Finally, we did not specifically investigate postoperative thromboembolic events, which could be an important outcome measure for future studies. However, our research group recently published a study² that addresses some of these limitations. This study examined the risk of left pulmonary artery stenosis or thrombosis following comprehensive cS2P, a condition requiring postoperative catheter interventions.

6 | CONCLUSION

The described anesthetic management also allows cS2P to be performed in patients with impaired preoperative cardiac function without increased surgical mortality. However, differences in survival probability were found in the long-term course. This emphasizes the vulnerability of patients with HLHS during the interstage I phase. Future research should focus on prospective studies to validate these findings and explore strategies to improve outcomes for patients requiring inotropic support prior to cS2P. Additionally, investigating long-term neurodevelopmental outcomes in relation to anesthetic management could provide valuable insights for optimizing care for these complex patients.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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