Interstage Course With the Hybrid Procedure for Hypoplastic Left Heart Patients

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ABSTRACT

Objective: The hybrid procedure consisting of bilateral pulmonary artery bands, ductus arteriosus stent, and atrial septostomy has been proposed as an alternative for initial palliation of patients with hypoplastic left heart syndrome (HLHS). Minimal information about morbidity and mortality exists for the inter-stage (IS) period. Goal of this study was to report our IS experience with the hybrid procedure. Methods: Retrospective chart review performed on patients discharged from the hospital with the diagnosis of HLHS who underwent the hybrid procedure. Patients were excluded if they had a HLHS variant or if they had not undergone the second stage procedure. Results: Between July 2002 to December 2006, 32 patients met inclusion criteria. Four patients had a restrictive atrial septum requiring transcatheter therapy before the hybrid procedure. Age at hybrid procedure was 11.2 ± 15.3 days and weight was 3.0 ± 0.6 kg. Eleven patients had one additional catheterization intervention, two patients had two interventions, and two patients had three interventions. Two patients had one additional surgical procedure. Age at stage II was 167.4 ± 60.8 days and weight was 5.6 ± 1.1 kg. IS mortality was 12.5% (4/32). Three deaths were associated with likely infectious issues while the fourth death occurred in a patient with progressive pulmonary vein stenosis. Conclusion: The hybrid IS mortality is comparable to the reported mortality for the classic and modified Norwood procedures. Further large randomized studies are needed to determine whether these results remain consistent.

BACKGROUND

-Since Norwood first described a palliative procedure for patients with hypoplastic left heart syndrome (HLHS), morbidity and mortality for this procedure have steadily improved[1].

-A recent modification of the Norwood procedure consisting of a right ventricle to pulmonary artery conduit (NW-RVPA) instead of a modified Blalock-Taussig shunt (NW-BT) to supply pulmonary blood flow has also been documented to improve surgical as well as inter-stage (IS) mortality.

-Despite these improvements, there remains a cohort of patients with HLHS that are at high risk for mortality from either procedure[2].

-Another alternative for initial palliation of high risk patients with HLHS that has recently been advocated is a hybrid procedure[3].

-This procedure consists of placement of bilateral pulmonary artery bands, stenting of the ductus arteriosus off cardiopulmonary bypass, and performing a balloon atrial septostomy as previously reported.

-Initial procedural results have been promising, but mid- and long-term data are minimal.

GOAL

-The goal of this study was to report our IS experience, defined as time from hospital discharge after hybrid procedure to hospitalization for the comprehensive stage II operation

METHODS

-A retrospective chart review was performed on all patients who met criteria.

-Inclusion criteria included patients with HLHS, defined as aortic atresia/mitral atresia, aortic atresia/mitral stenosis, aortic stenosis/mitral atresia, or aortic stenosis/mitral stenosis, who underwent hybrid procedure and were discharged from the hospital.

-Patients were excluded if they underwent the hybrid procedure, but had an anatomic diagnosis different from those stated above or if they were awaiting their second stage operation.

-Baseline data consisting of anatomical diagnosis, gender, age and weight at the time of the hybrid procedure were recorded. Procedures performed before the hybrid procedure and any additional IS cardiac procedures were also documented.

-These procedures were defined as any interventional cardiac catheterization, excluding the initial atrial septostomy, or any additional surgical procedures.

-If two or more interventions were performed during a single catheterization procedure, it was counted as only one procedure.

-Age and weight of the patient at the second stage were recorded.

-IS mortality was documented.

RESULTS

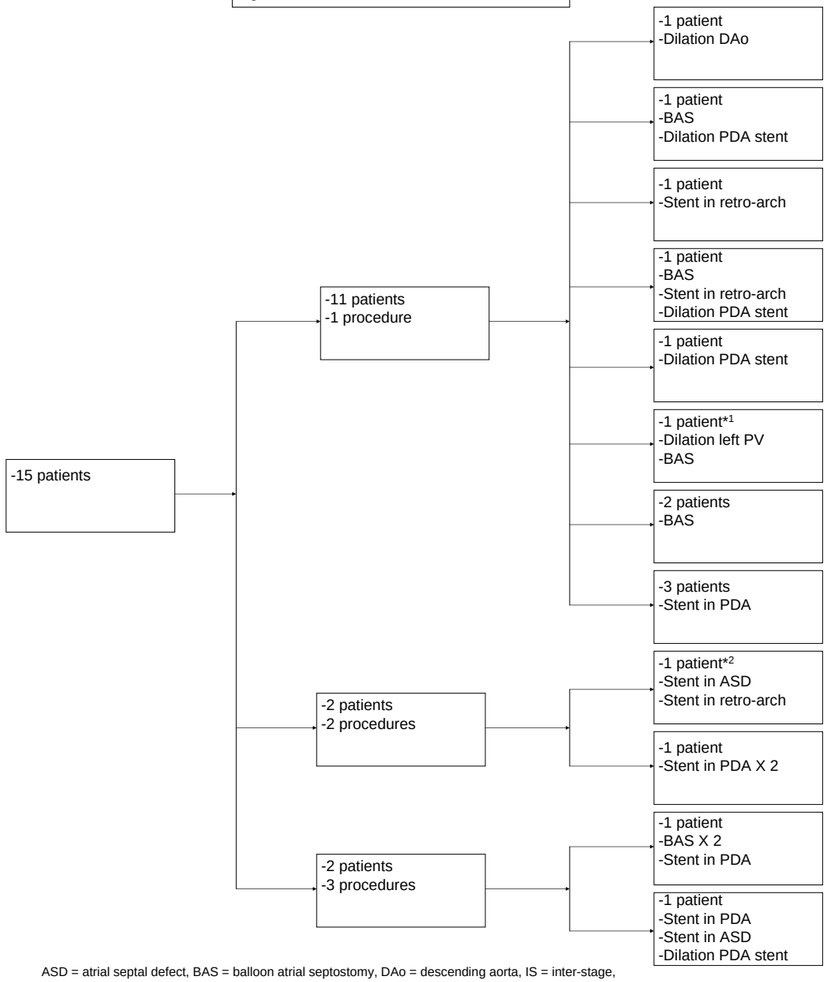
-Between July 2002 to December 2006, 35 patients with the diagnosis of HLHS who underwent the hybrid procedure were discharged from the hospital. Three patients are awaiting their second stage operation therefore this study consisted of 32 patients.

-Age at hybrid operation was 11.2 ± 15.3 days and weight was 3.0 ± 0.6 kg. Twenty-four patients were male.

-Eighteen patients had the anatomic diagnosis of aortic atresia/mitral atresia, four patients had aortic atresia/mitral stenosis, and ten patients had aortic stenosis/mitral stenosis. No patient had the diagnosis of aortic stenosis/mitral atresia. Four patients had a highly restrictive atrial septal defect that required balloon atrial septostomy before undergoing the hybrid procedure.

-During the IS period, 46.9% (15/32) of the patients underwent an additional intervention.

Figure 1 IS Cardiac Catheterization Procedures



ASD = atrial septal defect, BAS = balloon atrial septostomy, DAo = descending aorta, IS = inter-stage, PDA = patent ductus arteriosus, PV = pulmonary vein, retro-arch = retrograde aortic arch
* = surgical procedure: 1 = sutureless enlargement of left pulmonary veins, 2 = replacement of left pulmonary artery band

Table 1 Cardiac Catheterization Interventions

Procedure	Stent in PDA	BAS	Dilation PDA stent	Stent in retro-arch	Stent in ASD	Dilation DAo	Dilation left PV
n	7	7	4	3	2	1	1

ASD = atrial septal defect, BAS = balloon atrial septostomy, DAo = descending aorta, PDA = patent ductus arteriosus, PV = pulmonary vein, retro-arch = retrograde aortic arch

-Age at time of the second surgical palliative procedure was 167.4 ± 60.8 days and weight was 5.6 ± 1.1 kg.

-Time between the two procedures was 155.8 ± 53.8 days and weight gain was 17.2 ± 5.2 grams/day.

-Twenty patients had a cardiac catheterization procedure within 80 days of the second stage surgery and data are presented in Table 2.

Table 2 Cardiac Catheterization Data

SBP (mmHg)	DBP (mmHg)	mean BP (mmHg)	SVC (%)	Systemic (%)	LA (%)	Qp:Qs	RV SBP (mmHg)	RV EDP (mmHg)
81.5 ± 12.0	30.4 ± 6.7	49.9 ± 7.5	49.1 ± 7.5	75.2 ± 4.0	93.5 ± 5.6	1.6 ± 0.8	85.6 ± 13.4	7.3 ± 1.9

DBP = diastolic blood pressure, EDP = end diastolic pressure, LA = left atrium, RV = right ventricle, Qp:Qs = pulmonary to systemic blood flow ratio, SBP = systolic blood pressure, SVC = superior vena cava

-Four patients died during the IS period for an overall mortality of 12.5% (4/32).

Table 3 Mortality Characteristics

Diagnosis	Associated Diagnosis	Peri-mortality status
-Aortic stenosis/Mitral Stenosis		Vomiting and poor oral intake 3 days prior to death
-Aortic atresia/Mitral Atresia	Turner's syndrome	Upper respiratory congestion 2 days prior to death
-Aortic atresia/Mitral Atresia	30 EGA, 1.4 kg neonate when hybrid procedure performed	Documented Influenza B bronchiolitis
-Aortic atresia/Mitral Stenosis	Left pulmonary vein stenosis	Post-operative day 4 from sutureless repair of vein

CONCLUSION

-The hybrid IS mortality is comparable to the reported mortality for the NW-BT and NW-RVPA procedures[4, 5].

-Further large, randomized, long-term studies are needed to determine whether these results remain consistent and to evaluate the potential advantages of avoidance of cardiopulmonary bypass in the neonatal period that the hybrid procedure offers.

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