Influence of surgical strategies on outcome after the Norwood procedure

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Objective: The study objective was to identify how the evolution of surgical strategies influenced the outcome after the Norwood procedure.

Methods: From 1992 to 2004, 367 patients underwent the Norwood procedure (median age, 4 days). Three surgical strategies were identified on the basis of arch reconstruction and source of pulmonary blood flow. The arch was refashioned without extra material in group A (n = 148). The arch was reconstructed with a pulmonary artery homograft patch in groups B (n = 145) and C (n = 74). Pulmonary blood flow was supplied by a modified Blalock-Taussig shunt in groups A and B. Pulmonary blood flow was supplied by a right ventricle to pulmonary artery conduit in group C. Early mortality, actuarial survival, and freedom from arch reintervention or pulmonary artery patch augmentation were analyzed.

Results: Early mortality was 28% (n = 102). Actuarial survival was 62% ± 3% at 6 months. Early mortality was lower in group C (15%) than group A (31%) or group B (31%; $P < .05$). Actuarial survival at 6 months was better in group C (78% ± 5%) than group A (59% ± 5%) or group B (58% ± 4%; $P < .05$). Fifty-three patients (14%) had arch reintervention. Freedom from arch reintervention was 76% ± 3% at 1 year, with univariable analysis showing no difference among groups A, B, and C ($P = .71$). One hundred patients (27%) required subsequent pulmonary artery patch augmentation. Freedom from patch augmentation was 61% ± 3% at 1 year, and was lower in group C (3% ± 3%) than group A (80% ± 4%) or group B (72% ± 5%; $P < .05$).

Conclusions: Survival after the Norwood procedure improved after the introduction of a right ventricle to pulmonary artery conduit, but a greater proportion of patients required subsequent pulmonary artery patch augmentation. The type of arch reconstruction did not affect the incidence of arch reintervention.

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Hypoplastic left heart syndrome (HLHS) refers to a spectrum of congenital cardiac abnormalities that are characterized by severe stenosis or atresia of the mitral and aortic valves, a diminutive ascending aorta, and left ventricular hypoplasia.[1] Coarctation is usually associated with this lesion and contributes to reduce retrograde blood flow into the ascending aorta.[2] The left ventricle is unable to support the systemic circulation, which must be maintained by the right ventricle through a patent ductus arteriosus. HLHS accounts for 2.5% of congenital heart defects but is responsible for up to 25% of all deaths from congenital heart defects within the first week of life.[3] Without surgical palliation, 95% of children with HLHS die within the first month of life.[4]

More than 20 years ago Dr Norwood reported the first successful surgical palliation of HLHS.[5,6] The first stage of this surgical palliation is now commonly

Abbreviations and Acronyms
CI = confidence interval
CPA = central pulmonary artery
HLHS = hypoplastic left heart syndrome
LR = likelihood ratio
MBTS = modified Blalock-Taussig shunt
NP = Norwood procedure
RV-PA = right ventricle to pulmonary artery

called the Norwood procedure (NP). The aim of the NP is to establish unobstructed systemic and coronary blood flow from the right ventricle, and unobstructed pulmonary venous return across the atrial septum, and ensure adequate pulmonary blood flow without causing volume overload. Over the years, there has been a substantial improvement in the outcome after surgical palliation for HLHS. This has been attributed to refinements in the surgical technique and perioperative medical management care, together with a better understanding of the postoperative physiology.[7]

The success of the NP in HLHS has allowed the application of this procedure to other different pathologies, characterized by systemic outflow tract obstruction and functionally single ventricle anatomy. The early survival after the NP in contemporary series varies between 70% and 80%,[8-12] although an increasing number of centers have reported survivals of greater than 90% in the hospital.[13-16]

The aim of this study was to determine the influence of different surgical strategies and techniques used at our institution on outcome after the NP for all patients with classic HLHS or systemic outflow tract obstruction associated with either right or left ventricular hypoplasia, focusing particularly on aortic arch reconstruction and pulmonary blood flow supply.

Patients and Methods

Between November 1992 and August 2004, 367 patients with functionally single ventricle anatomy and systemic outflow tract obstruction underwent the NP at the Diana Princess of Wales Children's Hospital, Birmingham, United Kingdom. Most of the patients had a right ventricle-dependent systemic circulation (n = 333, 91%; Table 1). The median age at operation was 4 days (range, 0-217 days). The majority (n = 283, 77%) underwent operation within the first 7 days, and only 19 (5.2%) underwent operation at more than 30 days of age.

All operations were performed using deep hypothermic cardiopulmonary bypass with periods of circulatory arrest for arch reconstruction. Myocardial protection was provided using a single dose of cold crystalloid cardioplegia (St Thomas Hospital solution type 1, 30 mL/kg^{-1}) administered through the side arm of the arterial cannula before circulatory arrest. Antegrade cerebral perfusion, introduced in September 2002, was used during arch reconstruction in all patients with head and neck vessels of adequate size to accommodate the arterial cannula. The remainder of the operative strategy remained unchanged throughout the study period, as previously described.[17] In particular, neither modified ultrafiltration nor aprotinin was used during surgery. The median duration of cardiopulmonary bypass, aortic crossclamp, and deep hypothermic circulatory arrest times were 71 minutes (range, 17-323 minutes), 51 minutes (range, 0-109 minutes), and 55 minutes (range, 0-121 minutes), respectively. The median period of cardiopulmonary support (ie, cumulative duration of cardiopulmonary bypass and deep hypothermic circulatory arrest) was 121 minutes (range, 20-414 minutes). The atrial septum was excised through the atrial cannulation site. Arch reconstruction was performed using 1 of 2 established techniques. The original technique involved arch reconstruction without the use of additional patch material, as previously reported by this institution[18,19] and described by Fraser and Mee.[20]

TABLE 1. Primary cardiac anatomy

RV-dependent circulation	333	90.7%
Classic hypoplastic left heart syndrome	290	
Unbalanced atrioventricular septal defect	16	
Double-outlet RV with mitral atresia	15	
Critical aortic stenosis with hypoplastic LV	6	
Double-inlet, double-outlet RV	3	
Mitral atresia with aortic arch hypoplasia	3	
LV-dependent circulation	34	9.3%
Double-inlet LV with D-TGA	20	
Tricuspid atresia with D-TGA	8	
D-TGA, VSD with hypoplastic RV	4	
Other	2	

RV, Right ventricle; *LV,* left ventricle; D-TGA, dextro-transposition of great arteries; VSD, ventriculoseptal defect.

All the duct tissue was excised from the aorta, disconnecting the descending aorta from the arch. The aortic arch was opened along the inner aspect of the ascending aorta, down to the level of the transected proximal pulmonary artery. A complex Damus-Kaye-Stansel anastomosis was then constructed between the aortic arch, proximal pulmonary artery, and descending aorta (Figure 1, *A*).

The second technique, which has been used exclusively since April 1999, involved arch reconstruction with a pulmonary homograft patch, as originally described by Jonas and colleagues[21] with some variations. The duct tissue was completely excised only in the presence of severe coarctation, leaving the aortic back wall in continuity in all the other cases.

The arch was open in the inner aspect of the ascending aorta. The incision extended as proximally as possible to allow optimal coronary perfusion, and distally well beyond the coarctation area. The arch was then reconstructed with pulmonary homograft material cut to a teardrop shape. The proximal pulmonary artery was then anastomosed to a longitudinal incision in the allograft patch on the underside of the reconstructed neoaorta (Figure 1, *B*). In those patients in whom the coarctation tissue was excised, the back wall of the distal aortic arch and proximal descending aorta are joined directly, and then the underside of the neoaortic arch is augmented with allograft material.

Pulmonary blood flow was established using a modified Blalock-Taussig shunt (MBTS) (n = 293, 80%) or a right ventricle–pulmonary artery (RV-PA) conduit (n = 74, 20%). The MBTS consisted of a polytetrafluoroethylene tube conduit (Gore-

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