The intensive care of infants with hypoplastic left heart syndrome

U Theilen, L Shekerdemian

Until a little over two decades ago, hypoplastic left heart syndrome was considered an inoperable and fatal condition, with most deaths occurring in early infancy, and almost all of those affected dying before their first birthday. However, the advent of surgical palliation and advances in peri-operative care, have offered hope to these patients and their families.

Hypoplastic left heart syndrome (HLHS) is a continuum which can affect all left sided cardiac structures, from the mitral valve to the aortic arch. Since Norwood’s first description of surgical palliation in 1981,1 HLHS has been managed either by staged palliation in the majority of cases, or, in a minority, by primary cardiac transplantation. In this article we will discuss the early medical and surgical management of the neonate with HLHS, focusing on the evolution of new surgical strategies, and on the changing emphasis of peri-operative circulatory management on the intensive care unit.

INTENSIVE CARE OF THE INFANT WITH HLHS

The primary goal of peri-operative care of the neonate undergoing stage 1 palliation (Norwood operation or its modifications) is to optimise systemic oxygen delivery and organ perfusion. This must be addressed as early as possible, and in most cases the critical period begins before transfer to a cardiac centre.2

PREOPERATIVE STABILISATION

The arterial duct

In the preoperative period, systemic perfusion is critically dependent on a patent arterial duct, a non-restrictive inter-atrial communication, and good right ventricular function. The arterial duct is the main source of systemic blood flow, and provides retrograde filling to the aortic arch, diminutive ascending aorta, and coronary arteries. Babies with HLHS require prostaglandin E to maintain ductal patency. This should be commenced in the delivery room if the diagnosis has been made prenatally, or otherwise as soon as the diagnosis is suspected.2 Early intervention with prostaglandin maximises systemic perfusion, and optimises the condition of the infant before surgery.

The inter-atrial communication

In contrast to the majority of HLHS patients, in whom low systemic perfusion is in the context of excessive pulmonary blood flow, an important subgroup may have inadequate inter-atrial mixing due to a restrictive atrial septal defect (ASD). This results in global hypoperfusion, with profound hypoxaemia and acidosis, severe pulmonary venous hypertension, and overwhelming pulmonary venous congestion on chest x ray. These infants are generally unresponsive to measures aimed at improving pulmonary flow such as aggressive mechanical ventilation with high inspired oxygen fractions or nitric oxide, and if left untreated they rapidly develop progressive pulmonary venous hypertension. This is associated with increased mortality in infants with HLHS.3•4

A restrictive ASD requires urgent intervention. If this has been diagnosed antenatally, delivery should be planned in a centre able to urgently institute cardiopulmonary bypass. Decompression can be achieved by open surgical septectomy, or by the transcatheter route. In a recent series of 355 neonates with HLHS, 21 required interventional cardiac catheterisation for creation or enlargement of restrictive or absent ASD.5

A number of transcatheter techniques have been used. The Rashkind balloon atrial septostomy (often unsuccessful) and blade septectomy (carrying a high risk of tamponade) have been more recently superseded by techniques using transatrial needle puncture followed either by serial balloon dilatation (septoplasty) or stent implantation (see fig 1). Following relief of inter-atrial restriction, a few days should be allowed before stage 1 palliation. This allows for resolution of organ damage secondary to severe hypoxia, and for improvement in lung function.3

Respiratory management

All neonates with HLHS should have continuous monitoring of their oxygen saturation. In the preoperative setting, the systemic, pulmonary, and coronary circulations run in parallel, and are all supplied by the right ventricle.6 The balance between pulmonary flow (Qp) and systemic flow (Qs) is critically important: an excess of one will by definition compromise the other. The balance between pulmonary and systemic flow depends on the relative resistances in the two circulations. Deliberate or inadvertent manipulation of the pulmonary resistance (primarily with changes in respiratory management), or of the systemic resistance (using pharmacotherapy), will result in respiratory failure.

See end of article for authors’ affiliations

Correspondence to: Dr L Shekerdemian, Intensive Care Unit, Royal Children’s Hospital, Flemington Road, Parkville, Victoria 3052, Australia; lara.shekerdemian@rch.org.au

Accepted 7 June 2004

Abbreviations: ASD, atrial septal defect; ECLS, extracorporeal life support; HLHS, hypoplastic left heart syndrome; PVR, pulmonary vascular resistance
Ventilation and target blood gases

- \( \text{PaCO}_2: 35-45 \text{ mm Hg} \)
- \( \text{pH}: 7.35-7.40 \)
- \( \text{PaO}_2: 30-45 \text{ mm Hg} \)
- \( \text{So}_2: 70-85\% \)

†Ventilate initially using FiO\(_2\) 0.21, PEEP 4-5 cm H\(_2\)O
Haemoglobin should be maintained at 140-160 g/l
survival is variable. Infants diagnosed prenatally are certainly less likely to experience the sequelae of pulmonary overcirculation or acidosis. Prenatal diagnosis is associated with improved neurological outcome and with a lower incidence of preoperative acidosis and ventricular dysfunction, and the need for less inotropic support. For the minority with a restrictive ASD, prenatal diagnosis allows for immediate postnatal intervention. These data undoubtedly reinforce the importance of early pre-emptive intensive care of neonates with HLHS.

**SURGERY FOR HLHS**

Staged surgical palliation begins with the Norwood operation, or one of its modifications, typically performed during the first week of life. The aim of stage 1 palliation is to enlarge the aorta and to establish its continuity with the right ventricle; to provide free mixing of the pulmonary and systemic venous blood through an atrial septectomy; and to enlarge the aorta and to establish its continuity with the right ventricle; to provide free mixing of the pulmonary and systemic venous blood through an atrial septectomy; and to provide the lungs with flow through a systemic-to-pulmonary artery modified Blalock-Taussig (BT) shunt, or latterly via a conduit between the right ventricle and pulmonary arteries.

**Right ventricle to pulmonary artery conduit: a recent modification of stage 1 palliation (fig 2)**

Survival of infants born with HLHS has steadily improved since the first description of staged palliation by Norwood and colleagues. Historically, the use of an RV-PA conduit as the source of pulmonary flow had been abandoned in the early 1980s, due to high mortality. However, in retrospect this outcome was probably related to the generous diameter of the conduits (8 mm or more), resulting in massive pulmonary flow and systemic hypoperfusion, rather than the concept itself.

Recently, this approach has been revived, using smaller (4–6 mm) and therefore more limiting conduits, with encouraging results. The so-called “Sano” modification has become increasingly popular in major institutions around the world. Though still early in its evolution, the overwhelming impression from major centres is that the early postoperative course of infants with a conduit is smoother than for those with a BT shunt. A prospective randomised comparison of the two surgical techniques may never be feasible, but a number of retrospective cohort comparisons have now been published. While recognising their limitations in design and size, and relatively short follow up period, results have been remarkably uniform, and encouragingly in favour of the use of a conduit rather than a shunt.

Systemic perfusion and diastolic blood pressure are generally better maintained after the conduit procedure, and active manipulation of pulmonary blood flow (beyond standard measures) is rarely necessary. An important advantage of the conduit over the shunt is the maintenance of diastolic coronary perfusion, avoiding the “run-off” which can potentiate myocardial ischaemia in the latter group. This is supported by a number of observational studies which have reported a higher diastolic pressure in patients with a conduit and echocardiographic data showing that infants with conduit palliation have no diastolic flow reversal in the aortic arch, in contrast to those with a BT shunt. We have recently shown that these encouraging early signs may be translated into improved right ventricular performance in the convalescent phase.

Clearly, as with any new technique where long term results are not yet available, one must be cautious in over-interpreting the current data. Indeed, there are theoretical concerns regarding the long term implications of a right ventriculotomy in the conduit modification. However, right ventricular failure or arrhythmias have failed to materialise so far. Furthermore, our tissue Doppler data, catheterisation data of others, and increased survival to stage 2 palliation would all suggest improved, rather than diminished right ventricular function in patients with a conduit.

**POSTOPERATIVE INTENSIVE CARE AFTER STAGE 1 PALLIATION**

Stage 1 palliation, whether with a classical Norwood operation or with the conduit modification, is a complex procedure, which imposes profound physiological changes on a precarious substrate. Some infants have been in a suboptimal condition preoperatively, and all are subjected to a long period of cardiopulmonary bypass and aortic crossclamp, with deep hypothermia and, in the majority of patients, a period of circulatory arrest. The early postoperative period therefore imposes on these infants the major, immediate demands of adapting to a new circulation, while recovering from a long open heart procedure.

Again, the key to the intensive care of infants after stage 1 palliation is to optimise systemic oxygen delivery, and to act early on any disturbances of this. This is achieved with the combination of careful monitoring, avoidance of factors which lead to excessive pulmonary flow, maintenance of haemoglobin level, optimisation of ventricular performance, and systemic afterload reduction. Historically, early postoperative care of the Norwood patient focused on ventilatory manipulation of PVR to limit pulmonary blood flow. This often subjected the infants to a degree of acidosis which we now recognise as being undesirable. Over recent years, smaller and more limiting BT shunts, the advent of the conduit, and the more routine and liberal use of systemic vasodilators, have somewhat obviated the need for such aggressive control of PVR while appropriately optimising systemic perfusion.

**Haemoglobin**

The important contribution of haemoglobin to systemic oxygen delivery should not be underestimated. In the hypoplastic, where systemic saturation is kept relatively low, haemoglobin should be maintained around 140–160 g/l to maximise tissue oxygenation.
Monitoring of mixed venous oxygen saturation

The adequacy of systemic oxygen delivery, which is so critical, and so variable, is paradoxically difficult to assess. An increasing or persistent metabolic acidosis, or an increased serum lactate, may simply confirm that perfusion is under threat or already compromised. Furthermore, these parameters are not continuously monitored, and therefore give only periodic information. Mixed venous oxygen saturation provides a useful, continuous surrogate measure of the efficiency of systemic oxygen delivery in the functionally univentricular circulation, and can be used to guide therapy with systemic vasodilators.

In the infant after stage 1 palliation, mixed venous saturation can be continuously monitored using a co-oximeter placed directly into the SVC at the time of surgery. Although the absolute numbers obtained by this method are not always accurate, the trend in data generated by a well-positioned co-oximeter can be useful, with subtle changes often preceding changes in other haemodynamic parameters. A falling saturation would suggest increased tissue oxygen extraction, towards the anaerobic threshold, and most importantly should stimulate us to search for a cause.

Ventricular performance

Systemic oxygen delivery after stage 1 palliation can be compromised by two components of right ventricular performance: impaired contractility, and tricuspid regurgitation. Impaired systolic function is an inevitable consequence of bypass related myocardial injury, and can be exacerbated by coronary ischaemia. Ventricular contractility and competence of the tricuspid valve should be assessed using echocardiography. Right ventricular performance can then be optimised with inotropic therapy and appropriate afterload reduction using vasodilators, while avoiding factors which may result in excessive pulmonary blood flow and reduced coronary perfusion.

Pharmacotherapy

Most patients after stage 1 palliation require inotropes to maintain cardiac contractility. Dobutamine and dopamine are both appropriate for this role, though the additional vasodilatation afforded by dobutamine would make this the drug of choice in some centres. Phosphodiesterase-3 inhibitors such as milrinone are “inotodilators”, which improve systemic oxygen delivery by producing a combination of inotropic and vasodilatory effects. Milrinone has recently been shown to prevent the development of a low cardiac output state in selected children after cardiac surgery, and has been established, an oral vasodilator such as captropil is introduced.

Ventilation

All infants require mechanical ventilation postoperatively, and initial target blood gases are in general similar to those preoperatively—with a normal pH and CO₂, and an arterial PO₂ of less than 40 mm Hg. The pulmonary vasculature after surgery is often labile, and factors which may give rise to unwanted increases in pulmonary blood flow, such as hyperventilation, should be routinely avoided. If however, despite all of the above measures, there is persisting oversaturation with evidence of inadequate systemic oxygen delivery, then the addition of nitrogen to give hypoxic inspiratory gas mixtures can be considered. There is little, if any evidence to show that this manoeuvre has any more than a cosmetic effect—in other words, hypoxic gas effectively reduces the arterial oxygenation, without any convincing effect on systemic oxygen delivery.

Extracorporeal life support

Extracorporeal life support (ECLS) has been used to support many children with refractory cardiopulmonary dysfunction after surgery for congenital heart disease, with a hospital survival of around 40%. Historically, the survival for the subgroup requiring ECLS after stage 1 palliation for HLHS has been much lower, at less than 30%. In fact many centres, until very recently, considered HLHS to be a relative contraindication for ECLS. However, it is becoming increasingly recognised that the period of myocardial “rest”, and optimal systemic perfusion afforded by ECLS may have advantages over high dose inotrope therapy in borderline patients. In some centres this approach is becoming much more routine, with very encouraging outcomes. Therefore, although carrying a significant risk, ECLS can, if initiated promptly, improve the outcome in infants with impaired cardiopulmonary function after stage 1 palliation.

Vascular access

Most infants require central venous access in the first days or weeks after stage 1 palliation, for drug delivery, and in some cases for parenteral nutrition. Line related venous thrombosis is a well recognised complication of central venous catheters, and this can have disastrous consequences in infants with HLHS. In particular, thrombosis of the superior caval vein can preclude any further surgery. Many units have now moved away from using internal jugular central venous catheters, in favour of direct atrial lines. Although there is little evidence base in the current literature, it would be worth considering the routine use of low dose intravenous heparin in infants with HLHS who have central venous lines.

INTERSTAGE MORTALITY

This term refers to deaths occurring after hospital discharge and prior to stage 2 palliation, the bi-directional cavopulmonary shunt. Most institutions quote an interstage mortality of 10–15%, though early data would suggest that this figure may be lower with the conduit modification.

Most interstage deaths are sudden and apparently unpredictable. Infants after stage 1 palliation require close and regular monitoring, with regular medical review. Ghanayem et al reported a reduction of interstage mortality from 16% to 0% in 81 consecutive infants with HLHS, following the introduction of systematic daily monitoring following discharge; parents maintained a log of daily weight and saturations. Oxygen saturations below 70% and/or failure to gain at least 20 g weight within three days triggered review by a physician. In our institution, all infants have weekly echos, and remain hospitalised, or in hospital accommodation until stage 2 palliation.

Stage 2 palliation is associated with very low mortality, and following this procedure patients show improved growth, and are low risk candidates for their Fontan completion.
TRANSLATION

In some centres, the option of heart transplantation in early infancy is offered as an alternative to stage 1 palliation. Centres routinely performing transplantation have previously reported impressive early survival figures which would favour this option over stage 1 palliation. For the single reason of limited donor organ supply, primary transplantation is not a realistic option in most institutions outside North America. Transplantation is not an easy option. Most infants awaiting transplantation require long term therapy with prostaglandin, may need repeated septal decompressions, and potentially other extracardiac procedures. Also, transplantation introduces a new set of considerations: the need for lifelong immune suppression, the accelerated onset of coronary artery disease, and the unknown question of the long term viability of a neonatal donor heart. Moreover, with improving survival rates for staged palliation, the relative benefits of heart transplantation have narrowed over the last decade. Thus, a study from the USA, of 2264 infants diagnosed with HLHS, showed a reduction in the proportion being transplanted during the past decade.

CONCLUSION

Survival of patients with hypoplastic left heart syndrome has improved dramatically over the past 20 years, with many centres now quoting a survival after stage 1 palliation of over 80%.17-19 Moreover, mortality for these infants remains higher than for other congenital heart defects requiring surgical intervention in the neonatal period. Three main reasons for this have been identified: preoperative instability, mainly caused by inadequate systemic oxygen delivery; mortality following stage 1 palliation; and out-of-hospital mortality whilst waiting for stage 2 palliation. In this article, we have emphasised the need to optimise systemic perfusion in the peri-operative period, the particular challenge caused by a restrictive ASD, and the benefits of prenatal diagnosis and early optimisation of medical management. We have also highlighted recent advances in the surgical technique of stage 1 palliation by using a right ventricle to pulmonary artery conduit, the role of heart transplantation, and the use of ECLS.

All in all, over recent years, the refinement of surgical techniques, with pre-emptive management and improved intensive and convalescent care, have dramatically changed the outlook for infants with HLHS and their families.

ACKNOWLEDGEMENTS

We would like to thank Dr Tom Karl, Director of Pediatric Cardiac Surgery, University College San Francisco, USA for kindly allowing us to include his illustrations for figs 1 and 2.

Authors’ affiliations
U Theilen, L Shekerdemian, Intensive Care Unit, Royal Children’s Hospital, Parkville, Australia

Competing interests: none declared

REFERENCES


doi: 10.1136/adc.2004.064337

Congenital absence of the left sternomastoid muscle

This baby was brought to medical attention at 1 hour of age. The birth was a normal spontaneous vaginal delivery, after an uneventful pregnancy. Examination revealed an absent left sternomastoid muscle with paradoxical movement of the soft tissues in the left supraclavicular space. This was more pronounced on crying, as illustrated in figs 1 and 2. The rest of the clinical examination was unremarkable. The swelling seen superior to the clavicle was confirmed as being the herniating apex of the left lung.

A literature search found only one paper1 that describes a similar infant.

S Haroon
25 Magdalin Drive, Stanningley, Leeds LS28 6LD, UK
D Beverley
York District Hospital, Wigginton Road, York YO31 8HE, UK; david.w.beverley@york.nhs.uk

The child’s parents have consented to the publication of this article.

REFERENCE