Leading articles

Hypoplastic left heart syndrome—outcome and management

In this issue of *Archives of Disease in Childhood*, the Guy's group present their experience with staged reconstructive surgery for hypoplastic left heart syndrome (HLHS).[1] This leading article reviews the management and outcome of this condition with special reference to this important paper and stresses the outcome from the perspective of antenatal diagnosis.

Management of HLHS

The active surgical management of HLHS has been established in the UK over the past 12 years. It comprises three operations (Norwood stages I, II, and III) that establish the right ventricle as the systemic chamber. The first stage is discussed in more detail below. The second stage is when a cavopulmonary shunt is established and the third stage is the completion of the Fontan circulation.

PREOPERATIVE PERIOD

Figure 1A summarises the situation in the preoperative period. The systemic and pulmonary circulations are both supplied by the right ventricle. The balance between flow to the lungs and flow to the body via the patent arterial duct is critical: too much pulmonary blood flow will result in systemic underperfusion and too little in hypoxaemia. A satisfactory balance between adequate oxygenation and perfusion is often achieved in the early newborn period without specific intervention, other than intravenous prostaglandin to maintain ductal patency. However, if signs of a low systemic cardiac output develop (low urine output and progressive acidosis) it will be necessary to increase the pulmonary vascular resistance by active respiratory management: it may seem counterintuitive to reduce inspired oxygen in a sick, shocked neonate but this is frequently necessary to achieve stability. Furthermore, the oxygen levels in inspired air may prove to be too high and further reduction with the addition of nitrogen to the headbox or ventilator circuit may be required. If mechanical ventilation is necessary, maintenance of the systemic arterial P_{CO_2} at 5–6 kpa and systemic saturations around 80% often results in an appropriate balance between the two circulations. Surgery is usually deferred until about 5 days of age or longer to allow complete resolution of any markers of organ damage; failure to achieve this by medical management is usually considered a contraindication to surgery.

POSTOPERATIVE PERIOD

Figure 1B summarises the situation in the postoperative period. The Norwood stage 1 procedure is associated with a high mortality. It comprises an atrial septectomy to allow free mixing of the systemic and pulmonary venous return, using the main pulmonary artery to establish flow from the right ventricle to aorta and providing a Goretex shunt between the innominate or right subclavian artery and the branch pulmonary arteries. The systemic circulation is now supported directly by the right ventricle and pulmonary blood flow is dependent on the shunt. The challenge of achieving an appropriate balance between the two circulations remains. This again depends on the ratio between the pulmonary and systemic vascular resistances, but now also on the physical size of the shunt. A 3 mm Goretex shunt is usually satisfactory but an appropriate sized shunt at the time of surgery may prove far too generous in the early postoperative period (as the pulmonary vascular resistance falls). Shunt revisions as well as pharmacological/respiratory management of the pulmonary circulation are frequently required.

Outcome

Traditionally, outcomes for congenital heart disease are reported according to surgical mortality. Although these papers are extremely important to promote the development of surgical technique, they can be misleading when

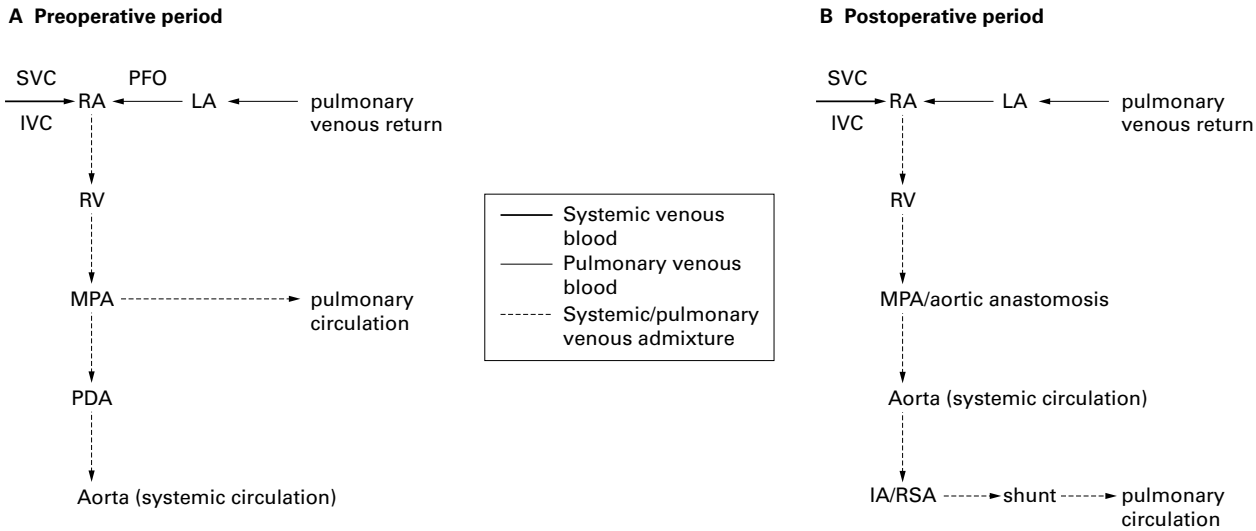

Figure 1 Preoperative and postoperative Norwood stage I circulation in hypoplastic left heart syndrome: the balance between the systemic and pulmonary circulations is crucial. The circulations have to be manipulated to achieve adequate systemic perfusion (systemic cardiac output) and adequate oxygenation (pulmonary blood flow). SVC, superior vena cava; IVC, inferior vena cava; RA, right atrium; LA, left atrium; PFO, patent foramen ovale; RV, right ventricle; MPA, main pulmonary artery; PDA, patent arterial duct; IA, innominate artery; RSA, right subclavian artery.

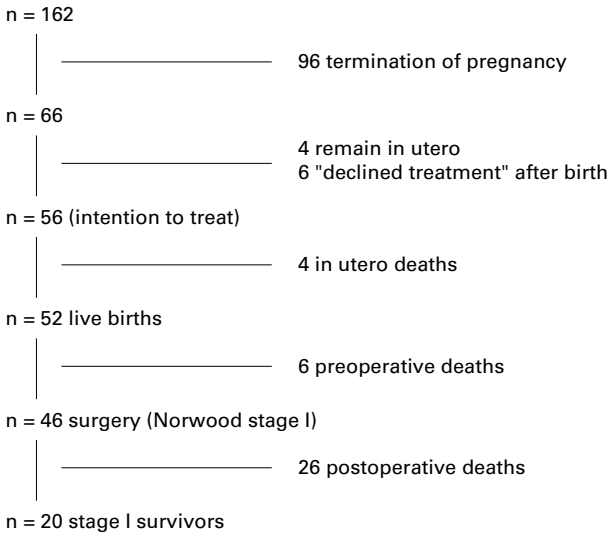

Figure 2 Guy's Hospital antenatal diagnosis.

considering overall outcome. This is particularly so when considering a condition diagnosed in utero that is associated with a high preoperative attrition.

SURGICAL MORTALITY

When only the babies who undergo surgery are included, the published surgical results indicate a mortality of approximately 30–50% for the Norwood stage 1, depending on patient selection and era.[2 3] The surgical mortality for stage I in the Guy's group is 51% and this is likely to reflect the fact that the diagnosis in the majority was made in utero. The evidence suggests that those babies diagnosed in the postnatal period may be favourably preselected by reaching a surgical centre. The small number in the postnatal diagnosis group in the Guy's series renders comparison with their antenatal group difficult.

MORTALITY AND IN UTERO DIAGNOSIS

When antenatal data are analysed on an intention to treat basis, the results are somewhat different. A total of 162 babies were diagnosed in utero at Guy's Hospital, with only two cases diagnosed elsewhere and nine cases diagnosed and treated in the postnatal period. If we study the Guy's antenatal diagnosis group alone, to avoid selection bias, we can assess outcome on an intention to treat basis (see fig 2).

The survival when the diagnosis of HLHS is made in utero is therefore 20/56 or 36% and this is a key figure when counselling families. It is also worth noting that this figure could be lower if the "parents' decision" not to operate on six of the babies was in any way influenced by a poor preoperative state or other specific risk factors that were identified during the preoperative period. These figures are remarkably similar to those of Allan *et al* who noted a 37.5% survival following antenatal diagnosis and a 50% surgical mortality.[4]

The causes of early death following the stage I Norwood procedure have been studied[5] and include impairment of coronary perfusion, excessive pulmonary blood flow, obstruction of pulmonary blood flow, neoaortic arch obstruction, right ventricular failure, tricuspid valve dysfunction, and other causes. It is expected that continued improvements in surgical technique and perioperative management will lead to reduced mortality and better functional outcome in the survivors.

LONG TERM OUTCOME

The published data and the Guy's experience for the stage II and III Norwood procedure confirm a low operative mortality. However, there is known to be an ongoing attrition: two of the Guy's patients died within one year of the stage II and a third patient has undergone cardiac transplantation. A review[3] of 840 patients who underwent stage I surgery for HLHS showed a 1, 2, 5, 10, and 15 year survival of 51%, 43%, 40%, 39%, and 39%, respectively. Late death occurred in 14 of 291 patients (4.8%) discharged home after the Fontan procedure, although only one patient had died beyond 5 years of age. Heart transplantation after stage I reconstruction was performed in five patients.

A number of patients are not deemed suitable for a Fontan procedure because of adverse haemodynamics and so the functional status of those who have proceeded to a Fontan is usually good. However, it is also known that poor functional status following the Fontan procedure is more common with a longer duration of follow up.[6] Detailed studies of the neurodevelopmental outcome of patients are limited,[7] but there are concerns and this is an area that now requires detailed study.

The future

The Norwood procedure has resulted in the survival of a large number of patients with HLHS who would previously have died. There continues to be a high morbidity and mortality and future developments have to be aimed at achieving perfect single ventricle haemodynamics (normal right ventricular and tricuspid valve function, normal pulmonary artery anatomy and vascular resistance, and an unobstructed systemic ventricular outflow) to ensure the best possible neurodevelopmental and functional outcome with the Fontan circulation (the "perfect Fontan"). Furthermore, until we know the long term outcome for the "perfect Fontan" in these patients we will not know what these children can expect as adults. Counselling of families and provision of balanced information on the benefits and uncertainties of entering a Norwood programme remain fundamental to management of HLHS.

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