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CASE REPORT

"Rapid Two-Stage" Norwood Operation in a Child with Multiorgan Failure

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Abstract The Norwood I operation continues to be a procedure with significant operative mortality. One well-accepted risk factor for death after the first step of the Norwood operation is critical preoperative status. We describe herein a new concept for the treatment of patients with hypoplastic left heart syndrome (HLHS) in very poor preoperative condition. This is a case report of a child who was born in a rural hospital. On the second day of life he was referred to our center in multiorgan failure. There were signs of liver dysfunction and the child was anuric. Therapy was started immediately with prostaglandin and vasodilators as well as diuretics, milrinone, and dobutamine. However, systemic perfusion continued to be insufficient. Finally, the child was placed on a ventilator. On the fourth day of life, bilateral pulmonary artery (PA) banding was performed and circulation stabilized immediately. Two hours after the operation urine output started. Liver function stabilized over the next couple of days. Two days after PA banding the child was weaned from the ventilator. On the 12th day of life a Norwood operation with PA debanding and a right ventricle-PA conduit was performed, and 2 days postoperatively the child was weaned from the ventilator. Twenty days after the operation he was discharged home. When the boy was 4 months old a bidirectional cavopulmonary anastomosis was performed. In selected cases of patients with HLHS with very poor hemodynamic conditions, a rapid two-stage approach with bilateral banding followed by a Norwood operation after cardiac stabilization can be recommended.

Keywords Congenital heart disease · Hypoplastic left heart syndrome · Norwood operation

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The Norwood I operation continues to be a procedure with significant operative mortality [4, 6]. One well-accepted and well-documented risk factor for death after the first-stage Norwood palliation is critical preoperative status [3]. Previously there have been two options to address this issue: either accepting the increased risk of the Norwood I operation or starting a multistep approach with bilateral pulmonary artery banding combined with patent ductus arteriosus (PDA) stenting [2, 4, 5, 8]. But both strategies seem to result in similar median-term survival rates [8].

We describe herein a new concept for the treatment of patients with hypoplastic left heart syndrome (HLHS) that present in poor preoperative condition.

Case Report

The child was born in a rural outlying hospital. First diagnosis of HLHS was made on the second day of life, and

the child was immediately referred to our center. The patient presented in multiorgan failure with signs of liver dysfunction (increased transaminases), and he was anuric. Immediate therapy was started with prostaglandin and vasodilators as well as diuretics, milrinone, and dobutamine. However, systemic perfusion continued to be insufficient, as indicated by an increase in serum lactate (up to 2.8 mM) and serum creatinine (up to 2.3 mg/dl) as well as development of metabolic acidosis (minimum pH, 7.32; base excess, −9). Finally, the child was intubated and ventilated. However, despite maximal medical support, he deteriorated further.

On the fourth day of life, emergent bilateral pulmonary artery banding was performed (Fig. 1). Circulation stabilized in the operating room, and directly after the operation considerably improved skin perfusion was noted. The child received dobutamine (8 μg/kg/min), milrinone (0.25 μg/kg/min), and urapidil (0.1 mg/kg/h), and blood pressure was stable at about 70/45 mm Hg, with a heart rate of 120 bpm. Oxygen saturation measured by pulsoximetry was 84%. The arterial duct was kept open by prostaglandin infusion (10 ng/kg/min). A few hours after the operation urine output started, and serum lactate dropped to 1.3 mM. Liver function stabilized over the next couple of days. Two days after bilateral pulmonary artery banding the child was weaned from the ventilator.

On the 12th day of life, 8 days after the bilateral pulmonary artery banding, a Norwood operation with a right ventricle-pulmonary artery conduit and pulmonary artery debanding was performed. The operative course was uneventful and 2 days after the operation the child was weaned from the ventilator. On the 20th postoperative day the child was discharged home in good health on captopril and aspirin (acetylsalicylic acid). Feeding was without problems.

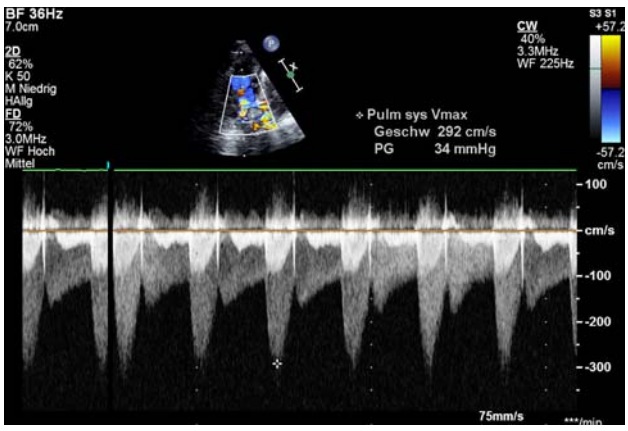

Fig. 1 CW Doppler of the left pulmonary artery after bilateral pulmonary artery banding with a peak gradient of 35 mm Hg. Systemic blood pressure was 55 mm Hg

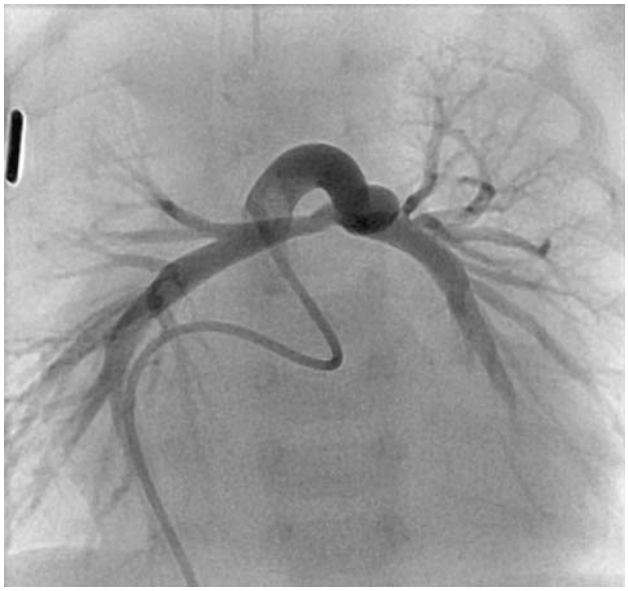

Fig. 2 Pulmonary angiography 3 months after the Norwood procedure with a right ventricle-pulmonary artery conduit, showing stenosed right and left pulmonary arteries after removal of both pulmonary artery bands

When the child reached the age of 4 months elective bidirectional cavopulmonary anastomosis was performed. During this operation the right pulmonary artery was enlarged with a homograft patch, the left pulmonary artery was dilated under direct vision with a balloon (Fig. 2), and a recoarctation was stented via an antegrade approach from the ascending aorta (CP stent 8Z16; pfm, Cologne, Germany). The child was discharged home on the 15th postoperative day.

At the age of 11 months the child weighed 9 kg and was beginning to walk. Currently the patient is awaiting Fontan completion.

Discussion

The first-stage Norwood operation still carries a significant operative morbidity and mortality in patients with critical preoperative hemodynamic status [3]. In some cases, preoperative medical stabilization alone is not effective. When pulmonary artery resistance drops, there are only limited medical options to reduce pulmonary and increase systemic blood flow. In this situation, the first-stage Norwood operation must be performed despite the increased operative risk, or alternatively, the so-called Giessen procedure [1, 2].

The Giessen procedure consists of a bilateral pulmonary artery banding combined with a PDA stent, providing similar improvements in hemodynamics as the classical first-stage Norwood operation with a modified Blalock-

Taussig shunt [1]. However, the operative risk of the Giessen procedure seems to be reduced compared to the classical Norwood operation [2, 8]. The second operative step in the Giessen procedure, which is performed 3 to 6 months later, is typically a Norwood-type arch reconstruction where pulmonary blood flow is provided by a bidirectional cavopulmonary connection [2].

In hemodynamically critically ill patients the Giessen procedure has been shown to reduce morbidity and mortality of the first surgical procedure [1]. However, the interstage mortality, as well as the operative risk of a second operation at 3 to 6 months of life, appears to increase, and result in similar overall survival after the first two operations [8].

The increased operative risk of the second stage of the Giessen procedure is mainly a result of increased surgical difficulties. First, the reconstruction of the aortic arch is a redo operation; second, the PDA stent has to be removed. It has not been fully determined whether complete removal of the stent, complete resection of the PDA including the stent, or cutting of the stent is the best option. In many cases left and right pulmonary arteries must be enlarged in order to treat residual stenoses after pulmonary artery debanding. But regardless of the surgical approach, the operative mortality is increased.

In comparison to what has been published previously, the rapid two-stage Norwood procedure may be a better surgical option for selected, medically unstable children with HLHS. Prior publications have recommended postponing palliative surgery for several months [2, 5, 7]. However, even after short-term pulmonary artery banding, residual stenoses may require surgical enlargement with a Glenn procedure or by pre- or intraoperative balloon dilatation. In contrast, when rapid bilateral pulmonary artery banding is performed, hemodynamics can be stabilized, resulting in subsequent improvement of pulmonary, renal, and liver function. A further advantage is that the Norwood operation can be performed shortly after the first procedure, before major adhesions occur.

Conclusion

In selected cases of patients with HLHS in very poor hemodynamic condition, a rapid two-stage approach with bilateral pulmonary artery banding followed by a Norwood operation after cardiac stabilization can be recommended.

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