General Principles of the Hybrid Approach in Hypoplastic Left Heart Syndrome

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27.1 Introduction

Congenital heart disease remains the most common form of birth defect and occurs in ~1 % of all live births. Hypoplastic left heart syndrome (HLHS) occurs in 2–3/10,000 live births in the USA [1]. It was uniformly fatal through the 1970s until the Norwood surgical palliation was proposed in 1980 [2]. When done properly, this operation, and its Sano modification, achieves a tenuous but stable circulation as a first step in palliation for HLHS. Less traumatic neonatal palliation has since been pursued as an alternative to the Norwood operation. Gibbs et al. first described neonatal stenting of the arterial duct combined with pulmonary artery banding and atrial septectomy or septostomy as an alternative palliation for HLHS in 1993 [3]. Since that time hundreds of articles have been published describing variations on technique and outcomes from this so-called hybrid approach to HLHS.

Irrespective of preference or approach, successful neonatal palliation for HLHS necessitates three objectives: (1) controlling or limiting pulmonary blood flow, (2) providing reliable and adequate systemic perfusion, and (3) assuring unrestricted flow of pulmonary venous blood return from the left atrium into the right atrium. The driving principal of the hybrid Stage 1 approach to HLHS is to achieve these objectives in a less invasive way with hopefully less morbidity and mortality (Fig. 27.1).

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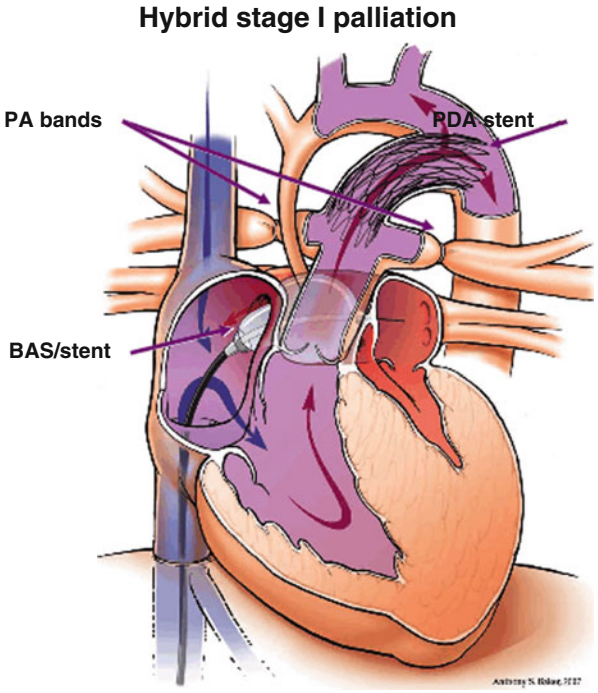

Fig. 27.1 Schematic of stage 1 hybrid palliation for HLHS. Note the pulmonary artery bands on both the left and right pulmonary arteries placed proximal to take off the upper lobe branches, the stent within the arterial duct, and the septostomy catheter representing decompression of the left atrium

27.2 Controlling or Limiting Pulmonary Blood Flow

As the newborn with HLHS transitions away from fetal circulation, pulmonary vascular resistance drops dramatically and pulmonary blood flow increases. The hybrid approach to HLHS relies on branch pulmonary artery banding to decrease and control the amount of pulmonary blood flow. While there were initial transcatheter attempts to accomplish this with internal flow restrictors, externally placed bands is the general approach in the current era. This requires a median sternotomy and dissecting free each of the branch pulmonary arteries. Surgically fashioned circumferential bands, typically made from Gore-Tex, are sewn around each of the pulmonary arteries. The band size typically varies between 3.0 and 3.5 mm depending on the patient size and goal length of palliation (Fig. 27.2). The larger the band, the less the restriction to pulmonary blood flow, at times leading to relative pulmonary overcirculation and associated clinical symptoms of tachypnea, poor feeding, and poor weight gain. The tighter the band, the lower

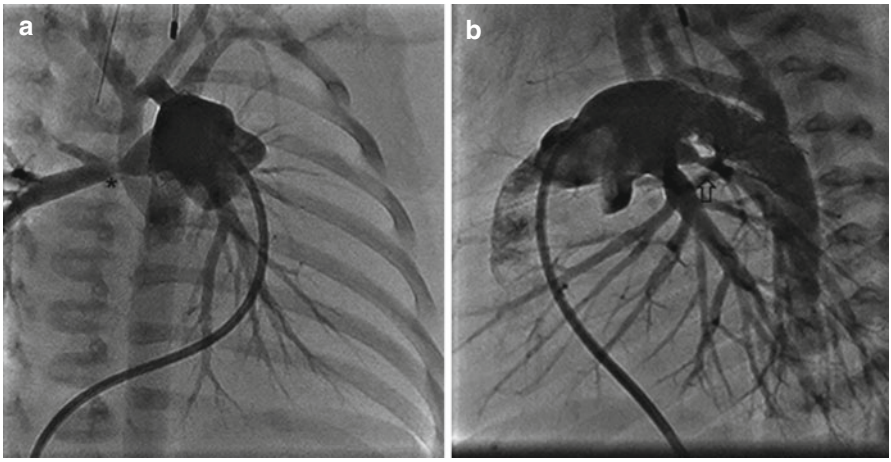

Fig. 27.2 A neonate born with HLHS undergoing catheterization 8 days after PA banding and ductal stenting for planned balloon atrial septostomy. An antegrade angiogram performed in the main pulmonary artery profiles, the RPA band (*) with RAO angulation of the frontal flat panel (**a**), and the LPA band (*arrow*) with LAO angulation on the lateral front panel (**b**)

the systemic saturation as pulmonary blood flow is limited. Finding this balance is part of the art of the hybrid procedure for neonates with HLHS.

27.3 Adequate Systemic Perfusion

Neonates born with HLHS are dependent on right to left shunting via the arterial duct for maintaining adequate systemic perfusion. When the diagnosis goes unrecognized, ductal closure ensues and profound cardiogenic shock develops as all end organs are hypoperfused. Without the initiation of prostaglandin E1 (PGE1) to maintain ductal patency, death is almost universal. Prenatal or early postnatal diagnosis of HLHS is now relatively common. As such, PGE1 is started immediately after birth or shortly thereafter as a temporizing way to maintain adequate systemic perfusion. Because long-term PGE1 infusion is associated with significant side effects, a more permanent and reliable solution is needed to maintain ductal patency. As Gibbs first described in 1993 [3], ductal stenting replaces the need for PGE1. The shunt through the arterial duct is obligatorily from right to left in systole with, at times, significant left to right "steal" into the pulmonary vasculature as the pulmonary vascular resistance continues to decrease. In the most severe forms of HLHS involving aortic atresia, both coronary artery and cerebral perfusion is completely dependent on the retrograde filling of the aortic arch (Fig. 27.3).

The timing of ductal stenting varies by institution. While some may approach this percutaneously as a separate procedure from the banding of the pulmonary

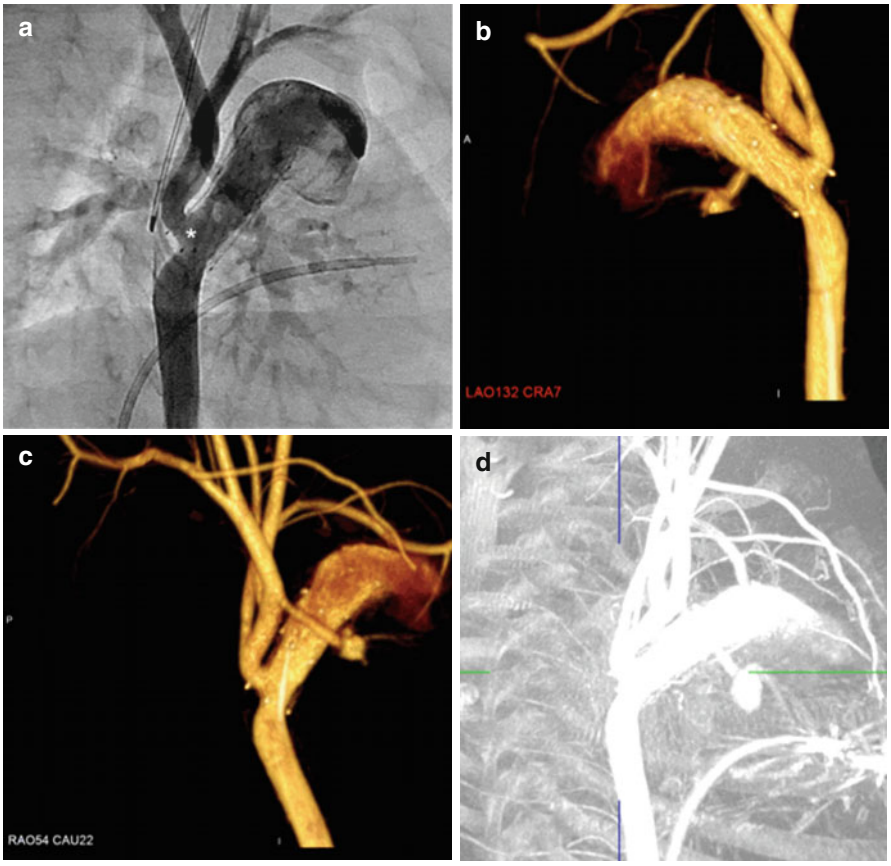

Fig. 27.3 While the stented arterial duct can typically be seen adequately with an antegrade angiogram in the main pulmonary artery, if concerns arise for retrograde filling of the hypoplastic aortic arch, a retrograde angiogram (**a**) or 3D rotational angiogram (**b**, **c**) with CT-like tomographic images (**d**), can be utilized to help profile this area. Note the angle and take off the retrograde arch (*)

arteries, it can easily be done in the same setting of the pulmonary artery bands via the median sternotomy and in concert with one's surgical colleagues. In either scenario, it is essential that the entire length of the arterial duct be covered appropriately by the stent(s). Uncovered ductal tissue on either end of the duct will constrict once off PGE1, increasing afterload on an already stressed right ventricle as well as compromising systemic perfusion both to the retrograde arch (brain and coronary arteries) and to the descending aorta. The deleterious effects of this may not be recoverable depending on the timing of recognition and severity of the insult.

Needless to say, a successful first stage hybrid palliation for HLHS depends on appropriate stenting of the arterial duct and close surveillance of retrograde arch

perfusion during the interstage period. This is never truer when dealing with the severest forms of HLHS, namely, the aortic atresia type.

27.4 Assuring Unrestricted Flow of Pulmonary Venous Return into the Right Atrium

Neonates born with HLHS typically have a relatively unrestricted atrial communication, allowing oxygenated pulmonary venous return to reach the right atrium and sufficiently mix with systemic venous return. Rarely, the atrial septum is highly restrictive at birth and even more rarely completely intact. These scenarios require urgent and at times emergent intervention and thankfully deviate from the norm.

When dealing with a stable newborn on PGE1, the systemic saturation and mean Doppler gradient across the atrial septum will inform the degree of true restriction at the atrial level. While the echo mean Doppler gradient may increase over the first several days of life, this is often in association with and due to the decreasing pulmonary vascular resistance and increasing amount of pulmonary blood flow and subsequent return to the left atrium. An increasing echo Doppler gradient in conjunction with decreasing systemic saturations should alert the clinician to increasing the restriction to flow at the atrial septal level.

Part of a successful hybrid stage 1 palliation is creation of a reliable unrestrictive communication that will remain so until stage 2 reconstruction. While this may have required surgical atrial septectomy, in some of the original descriptions of this approach, with current technologies, a durable and reliable atrial septal communication can be created with a balloon atrial septostomy. For a number of reasons, including size of the left atrium, location of the defect within the atrial septum, and initial size of the atrial septal defect, this step of hybrid stage 1 palliation can be technically challenging. Specific techniques and timing of this portion of stage 1 palliation are discussed separately and reflect the experience and knowledge gained over the many years of taking on this approach to HLHS palliation.

Conclusion

All three objectives for successful palliation of the vulnerable newborn with HLHS can be accomplished less invasively via the hybrid stage 1 procedure. No single approach, either hybrid or Norwood, eliminates the fragile interstage period where the slightest change to a tenuously balanced circulation can lead to potentially irreversible myocardial and other end organ damage. Interstage monitoring is essential for this patient population to help minimize these risks. Hybrid stage 1 palliation offers an acceptable method for managing the fragile newborn with HLHS. With appropriate outpatient monitoring, it delays the inevitable complex semilunar amalgamation and aortic reconstruction to the outside of the neonatal period where it is combined with pulmonary artery band removal, ductal stent removal, atrial septectomy, and cavopulmonary anastomosis into the so-called comprehensive stage 2.

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