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ORIGINAL ARTICLE

The hybrid approach for palliation of hypoplastic left heart syndrome: Intermediate results of a single-center experience

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KEYWORDS
Hypoplastic left heart syndrome;
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Abstract
Introduction: Hypoplastic left heart syndrome (HLHS) is a major cause of cardiac death during the first week of life. The hybrid approach is a reliable, reproducible treatment option for patients with HLHS. Herein we report our results using this approach, focusing on its efficacy, safety and late outcome.
Methods: We reviewed prospectively collected data on patients treated for HLHS using a hybrid approach between July 2007 and September 2014.
Results: Nine patients had a stage 1 hybrid procedure, with seven undergoing a comprehensive stage 2 procedure. One patient completed the Fontan procedure. Five patients underwent balloon atrial septostomy after the hybrid procedure; in three patients, a stent was placed across the atrial septum. There were three deaths: two early after the hybrid procedure and one early after stage two palliation. Overall survival was 66%.
Conclusions: In our single-center series, the hybrid approach for HLHS yields intermediate results comparable to those of the Norwood strategy. The existence of dedicated teams for the diagnosis and management of these patients, preferably in high-volume centers, is of major importance in this condition.

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PALAVRAS-CHAVE
Síndrome do coração esquerdo hipoplásico;
Abordagem híbrida;
Stent canal arterial

Abordagem híbrida de paliação de síndrome de coração esquerdo hipoplásico – resultados intermédios: experiência de um centro

Resumo
Introdução: A síndrome do coração esquerdo hipoplásico (SCEH) é uma das principais causas de morte durante a primeira semana de vida. A abordagem híbrida é uma opção de paliação para doentes com SCEH. Reportamos os nossos resultados com esta abordagem, com foco particular na sua eficácia, segurança e resultado final.
Métodos: Trabalho prospetivo. Revisão dos dados clínicos de doentes com SCEH, submetidos a abordagem híbrida de paliação entre julho de 2007 e setembro de 2014.
Resultados: Nove doentes foram submetidos a primeiro estadio híbrido de paliação. Destes, sete completaram segundo estadio e um doente foi submetido a cirurgia de Fontan. Cinco doentes foram submetidos a atriosseptostomia com balão. Em três procedeu-se a implantação de *stent* no septo interauricular. Verificaram-se três óbitos: dois logo após o primeiro estadio híbrido e um após o segundo estadio. A sobrevida global foi de 66%.
Conclusões: Na nossa experiência, a abordagem híbrida para SCEH produz resultados comparáveis aos da estratégia de Norwood. A necessidade de uma equipa dedicada para o diagnóstico e manejo destes doentes, de preferência em centros de alto volume, é de grande importância nesta condição particular.

Introduction

Hypoplastic left heart syndrome (HLHS) represents 1.4–3.8% of congenital heart diseases but is responsible for 23% of cardiac deaths during the first week of life.[1–3] HLHS is characterized by variable degrees of underdevelopment of left heart structures.[4–6] Traditional management of HLHS, either with staged surgical palliation (Norwood, Glenn, Fontan)[7,8] or cardiac transplantation,[9] remains a challenge in most centers, particularly in high-risk patients such as those with low birth weight (<3 kg) and diminutive aortas (<2 mm).[10–12] For this subgroup of patients, a hybrid approach – combining transcatheter and surgical techniques – was devised in the last decade, aiming to combine the best characteristics of surgical and interventional cardiology techniques.[13–15]

Originally reported in 1993,[16] hybrid stage 1 palliation consists of bilateral branch pulmonary artery banding and stenting of the ductus arteriosus, without cardiopulmonary bypass, during the neonatal period. Assurance of a nonrestrictive atrial septal defect is the next step, through either balloon atrial septostomy or stenting. Later in infancy, a more complex comprehensive stage 2 is performed at approximately six months of age, in the form of a Norwood-type reconstruction combined with a bidirectional cavopulmonary (Glenn) anastomosis. Although initially suggested for high-risk candidates for the classic Norwood stage 1 and 2 palliation,[14,17] it has since been adopted as the preferred first option by several centers.[14,18,19]

In 2007, our center started a hybrid program for HLHS palliation in high-risk cases. Our aim is to report our initial results with this technique, focusing on its efficacy, safety and late outcome, with particular emphasis on morbidity and mortality, need for unplanned reintervention, and the current status of the patients.

Methods

Patient population

Our cohort comprised patients who underwent a hybrid stage 1 procedure between July 2007 and September 2014. All had typical HLHS (aortic atresia or critical stenosis with mitral atresia or stenosis). Initially, our center chose only high-risk patients for the hybrid palliation, defined by the presence of low birth weight (<2.5 kg), prematurity, severe aortic arch hypoplasia, poor right ventricular function, more than mild tricuspid regurgitation, highly restrictive atrial septal defect, and the presence of non-cardiac malformations. Later on, we widened our criteria and opted for a hybrid approach for all HLHS patients. A review of prospectively collected data including information from all planned staged procedures, any unplanned reinterventions, and interstage morbidity and outcomes are discussed herein. Follow-up was complete in all patients (Figure 1).

Technique

Hybrid stage 1

The procedure was performed in the catheterization laboratory, adapted as a hybrid suite, under general anesthesia. The technique was based on reports by Galantowicz et al.[18] Briefly, through a median sternotomy, and without cardiopulmonary bypass, bilateral branch pulmonary artery (PA) bands were placed using a 3.5-mm Gore-Tex tube graft (WL Gore & Associates, Flagstaff, AZ, USA); the band was first placed on the left PA. The circumference of the band was calculated according to the caliber of the branch PA and the degree of band tightening was adjusted according to the