

CASE REPORT

Hybrid procedure in treatment of high risk hypoplastic left heart syndrome

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The hybrid approach is a less invasive initial alternative than the palliation of the hypoplastic left heart syndrome, combining surgical and interventional techniques. We present a newborn with high risk and hybrid procedure (bilateral pulmonary banding, arterial duct stenting and atrioseptostomy). Angiography and echocardiography showed unrestrictive atrial defect, unobstructed pulmonary artery and stent permeable. We will require further research to determine whether the hybrid approach will provide better results in the short and long term.

Key words: Arterial duct stenting; Hybrid procedure; Hypoplastic left heart syndrome.

El abordaje híbrido es una alternativa inicial menos invasiva en la paliación del Síndrome de ventrículo izquierdo hipoplásico, combinando técnicas quirúrgicas e intervencionismo. Presentamos un neonato con ventrículo izquierdo hipoplásico de alto riesgo y procedimiento híbrido (cerclaje bilateral pulmonar, stent en ducto arterioso y atrioseptostomía). La angiografía y ecocardiografía demostraron defecto interauricular no obstructivo, arterias pulmonares y stent permeables. Requeriremos investigaciones posteriores para determinar si la estrategia híbrida brindará mejores resultados a corto y largo plazo.

Palabras clave: Stent ducto arterioso; Procedimiento híbrido; Síndrome de ventrículo izquierdo hipoplásico.

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The results of palliative surgery (Norwood/Sano) in hypoplastic left heart syndrome (HLHS) have significantly improved over time. Nevertheless, the general results of traditional palliation remain suboptimal, with reported 1-year survival rates of 60% and 5-year survival rates of 54% [1].

The hybrid approach has been developed as an alternative strategy that involves an initial less invasive procedure. It combines surgical techniques (bilateral pulmonary artery banding) and interventional techniques (ductus arteriosus stenting and balloon atrial septostomy) to delay major surgery (cardiopulmonary bypass, aortic clamping, circulatory arrest) and its inherent risks, until the patient is older.

The goals of stage 1 of hybrid palliation are: 1) systemic output through the patent ductus arteriosus, with no obstruction; 2) improved systemic and pulmonary circulation balance; and 3) unrestricted interatrial communication [2].

With acceptable intermediate results, the technique has been reported as safe and reproducible, with a hospital survival rate of 97.5% and an interstage mortality rate of 5% [2]. Follow-up involves a weekly echocardiogram and catheterization at 6 weeks. Stage 2 is carried out when the patient is 3 to 6-months-old and consists of removal of the ductal stent and pulmonary artery bands, repair of the aortic arch and pulmonary artery branches (if necessary), division of the ascending aorta (with reimplantation into the root), atrial septectomy, and bidirectional cavopulmonary anastomosis – all carried out on cardiopulmonary bypass, with aortic clamping. The Fontan completion procedure is performed when the patient is 2 years old. We present herein the hybrid procedure as initial palliation, in a high-risk patient with HLHS (with no distal obstruction).

CLINICAL CASE

A full-term 8-day-old male neonate weighed 3.2 Kg and presented with high-risk HLHS (aortic atresia), restrictive oval foramen, mitral atresia, and diameter of the ascending aorta < 2 mm (Fig. 1). Medical treatment included postgland



Figure 1. Aortic atresia, Ascending aorta < 2 mm.

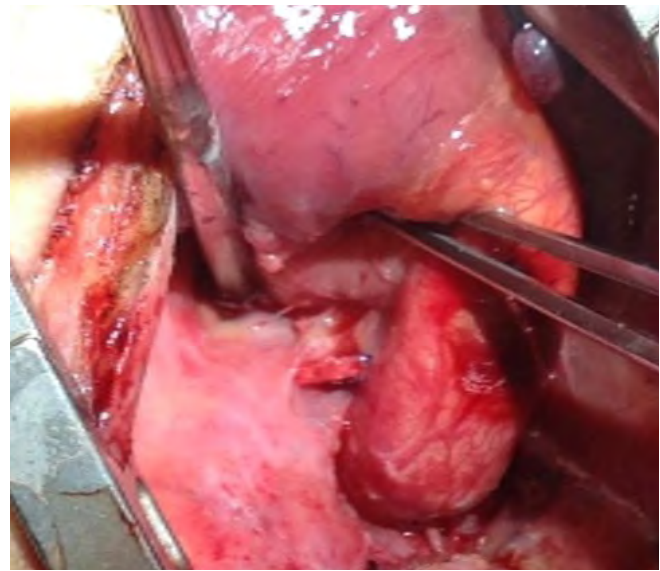


Figure 3. Placement of bilateral pulmonary artery bands.

din-E1, dopamine, dobutamine, and norepinephrine. Hybrid procedure was as follows: under general anesthesia, through median sternotomy, bilateral pulmonary artery banding was performed, with 3.5mm polytetrafluoroethylene (PTFE) tape. The left branch band was placed just after its origin at the trunk of the pulmonary artery and the right branch band was placed between the ascending aorta and the superior vena cava (Fig. 2) (Fig. 3). A 10mm (weight + 7mm) band and an 11mm band (weight + 7.5mm), for the left and right branches, respectively, are proposed by Kitahori et al. [3]. There is an approximate increase of 10mmHg in the systolic pressure

and a decrease of 10 points in oxygen saturation [2]. With a purse-string suture at the base of the pulmonary trunk and under angiographic control, utilizing a 6 Fr introducer, the ductus arteriosus was measured (6.5mm [pulmonary side] x 6mm [aortic side] x 16 mm length). An 8 x 17 mm stent was placed (Fig. 4) (Fig. 5), completely covering the duct, going beyond the retrograde orifice of the transverse aorta. Balloon atrial septostomy was performed. Control angiography (lateral left and anterior-posterior projections, and cranial and caudal angulations) verified the patency of both branches and the adequate position of the pulmonary artery banding and the

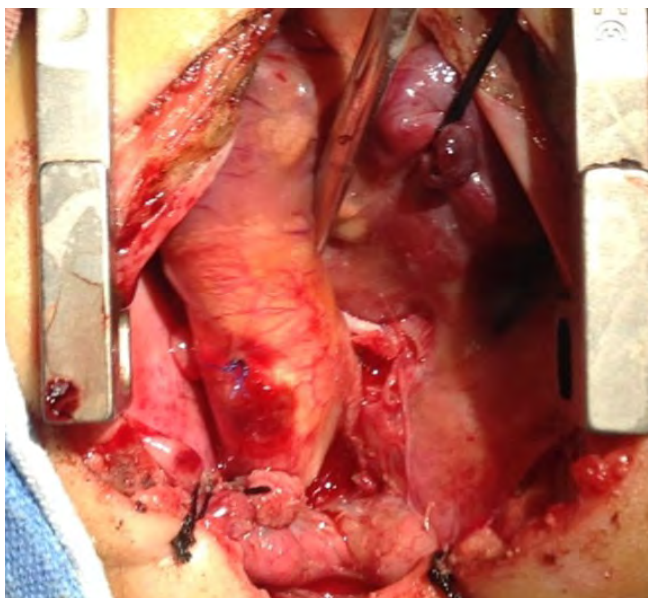


Figure 2. Placement of bilateral pulmonary artery bands.



Figure 4. A 6-F sidearm sheath is advanced into the main pulmonary artery.

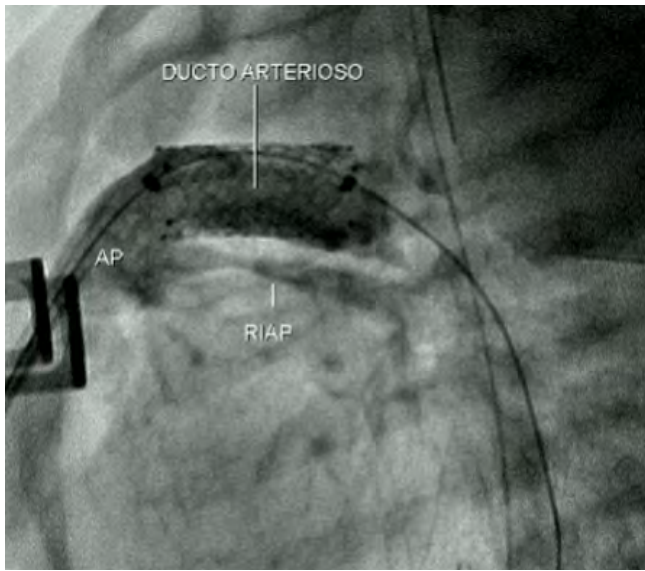


Figure 5. Anterograde flow into the descending aorta and retrograde flow into the aortic arch and ascending aorta through the stent.

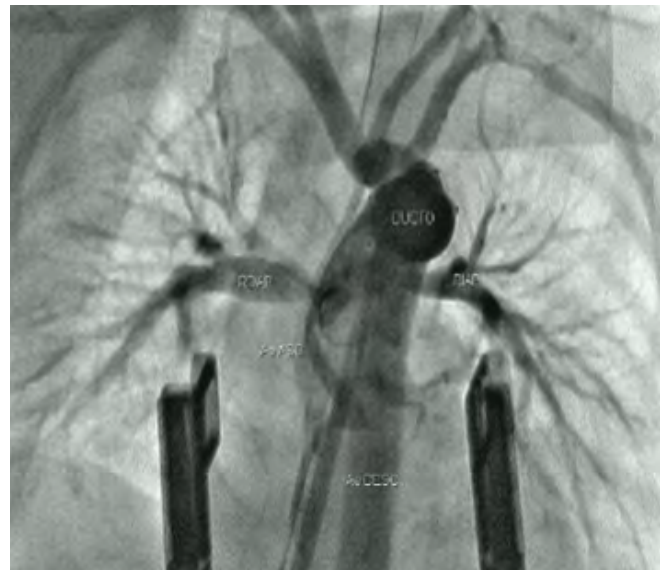


Figure 7. Balloon atrial septostomy.

stent (Fig. 6) (Fig. 7). Conventional sternal closure was carried out with a 0.6 mm PTFE interface. Interstage monitoring: follow-up has been carried out with weekly echocardiogram, observing nonobstructive interatrial communication, anterograde flow into the descending aorta and retrograde flow into the aortic arch and ascending aorta through the stent. Right ventricular function was preserved, with no tricuspid insufficiency, and flow alterations were within acceptable parameters (2-3m/s, according to Kitahori et al. [3]): 2 m/s in the left branch (24mmHg maximum gradient) and 2.9 m/s in the right branch (15 mmHg maximum gradient). (Fig. 8) (Fig. 9)

(Fig. 10). Stage 2 (removal of the stent and pulmonary artery bands, reconstruction of the aortic arch, bidirectional cavopulmonary diversion, atrial septectomy) is planned to be carried out in 4 to 6 months.

COMMENT

The short-term and long-term results with traditional HLHS treatment remain suboptimal. In its report covering the period of 1994 to 2000, the Congenital Heart Surgeons' Society, which includes 29 centers, showed a 54% 5-year sur-

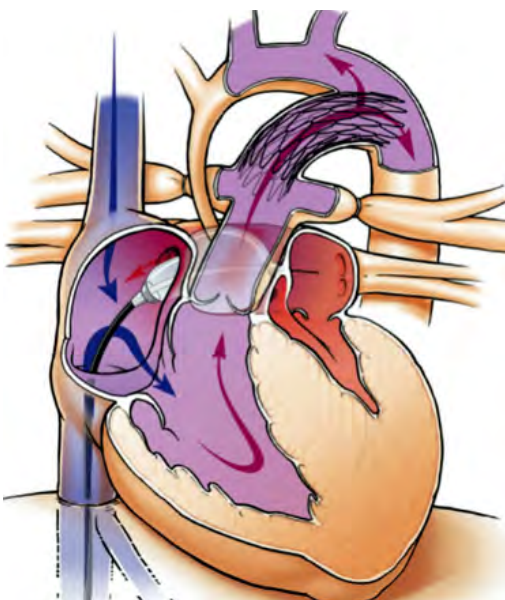


Figure 6. Hybrid approach for hypoplastic left heart syndrome.

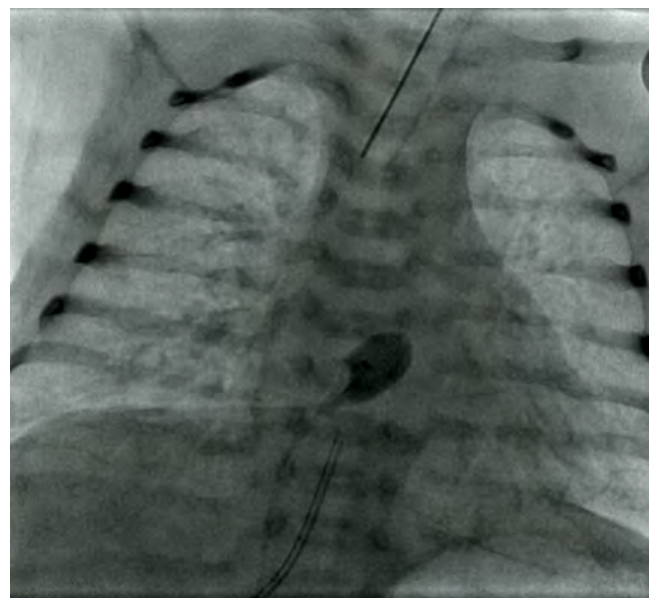


Figure 8. Nonobstructive interatrial communication.

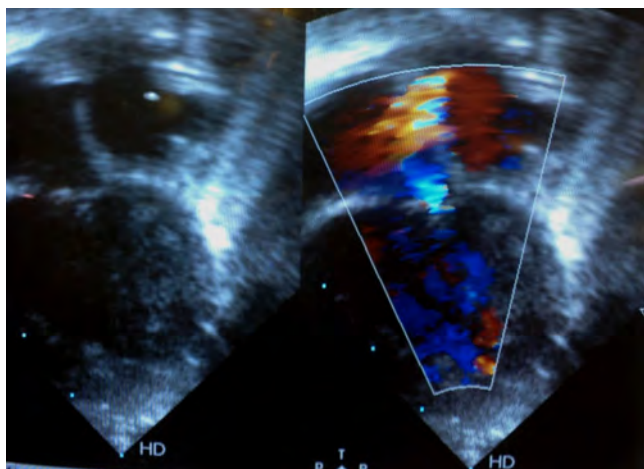


Figure 9. Patent stent with retrograde flow.

vival rate with conventional palliation in cases of HLHS. Only 28% of patients reached the Fontan completion procedure and another 20% are possible candidates [1]. Recent enthusiasm for the use of a conduit from the right ventricle to the pulmonary artery, in place of the modified Blalock-Taussig shunt during the Norwood procedure, has not significantly changed survival. Those data reveal the lack of improvement in the results, despite advances in the understanding of the anatomy and physiology involved in HLHS, in surgical techniques, and the formation of multidisciplinary teams, even in centers that specialize in these types of procedures.

Since 1993, some institutions have employed the hybrid alternative [4,5]. The primary aim is to create balanced cir-

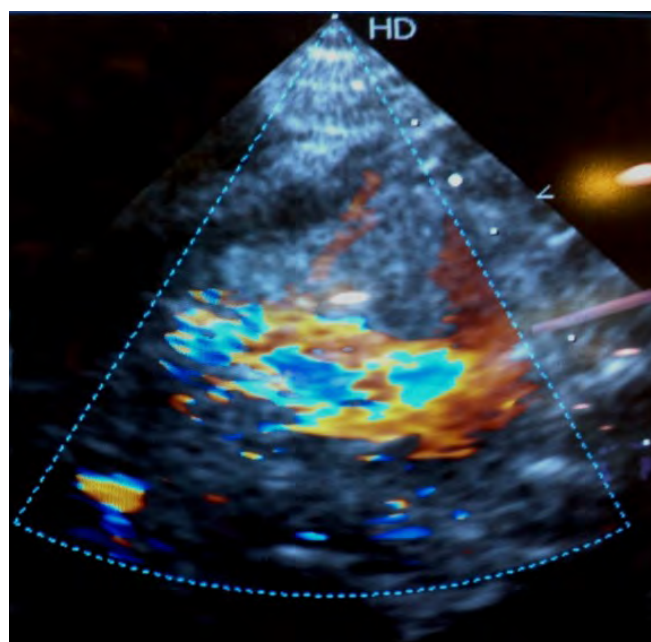


Figure 10. Velocity across the right pulmonary artery.

ulation, with no need for cardiopulmonary bypass and its risks in the neonatal stage. Therefore, major heart surgery is delayed until the patient is older and circulation in series can be established through cavopulmonary anastomosis.

Initial reports on the hybrid procedure described limited results due to the learning curve involved, the small number of patients, the inclusion of different diagnoses and risk stratifications, and short-term follow-up periods. Subsequent studies have been focused on high-risk patients with HLHS: weight under 2 Kg, gestational age <36 weeks, intracerebral bleeding, genetic alterations, and aortic valvular atresia [6,7]. To evaluate the real risks involved in the hybrid procedure, the Norwood surgery should be compared with the hybrid procedure's stage 1 and stage 2, as well as with the interstage period. Galantowicz et al. [2] concluded that the hybrid approach is a safe and reproducible procedure, based on its intermediate results. Theirs is the largest case series conducted at a single institution that offers the hybrid alternative to all patients with HLHS, and it included 40 cases treated within the time frame of July 2002 and June 2007. The only exclusion criterion was echocardiographic evidence of coarctation of the aorta, and those patients were referred for the traditional Norwood procedure. Hybrid stage 1 results indicated low surgical risk, with 80% of the patients extubated within the first 24 hours, occasional inotropic support, no delayed sternal closure, no need for extracorporeal membrane oxygenation (ECMO), short intensive care unit (ICU) stay, and hospital survival of 97.5%. The 5% interstage mortality rate and the 36% reintervention rate are similar to those reported for the Norwood procedure [2].

Even though hybrid stage 2 is a long procedure that involves all the steps of the traditional two-stage Norwood surgery, plus cavopulmonary anastomosis and the removal of the pulmonary artery bands and the ductal stent, the results of Galantowicz et al. showed that the postoperative course was similar only to that of the stage 2 Norwood procedure. Eighty-five percent of the patients were extubated within the first 24 hours, inotropic support other than milrinone was infrequent, right ventricular function, lactate levels, and renal function were normal, and there were no neurologic complications or ECMO requirements. That could be explained by the fact that the hybrid procedure minimizes organ ischemia because most of the procedure is performed on cardiopulmonary bypass, and the resulting circulation in series with a cavopulmonary anastomosis is more stable than the circulation in parallel that occurs in stage 1 of the Norwood surgery [2].

After completing the three stages (stage 1, stage 2, and the Fontan completion), the significant complications were neurocognitive alterations, partially attributable to the anomalous brain structures inherent in patients with HLHS. However, in neonates that undergo major heart surgery, 34% have a postoperative increase in periventricular leukomalacia, a nonspecific sign of cerebral white matter injury. Thus, the benefit of stage 1 of the hybrid procedure delays the use of cardiopulmonary bypass, until the brain is more developed. That delay can have long-term benefits on neurologic results [2].

Centers in Columbus, Ohio, and in Giessen, Germany, offer the hybrid approach to all usual-risk or high-risk patient. Both centers have reported survival of up to 90%, combining stages 1 and 2 in usual-risk patients [1]. Ventricular function is preserved, there is adequate pulmonary vasculature, and results are excellent in the stage 3 Fontan completion [1], with the hybrid procedure.

Fewer resources are used with the hybrid approach for HLHS management. Galantowicz et al. [1,2] described a comparative study of the Norwood and Sano surgeries, reporting mean hospital/ICU stay of 24 and 14 days, respectively, 75% delayed sternal closure, and 10% ECMO support. In contrast, the groups from Columbus, Ohio, and Toronto, Canada [1], have shown lower hospital resource use with the hybrid approach, compared with the traditional Norwood surgery, even when the two hybrid stages (stages 1 + 2) are combined.

Other applications of the hybrid procedure are: improving the hemodynamic profile in patients with extreme forms of HLHS (late diagnosis or multiple organ failure), facilitating kidney and/or liver recovery, stabilizing other comorbidities, such as intracerebral bleeding or sepsis, and the surgical repair of other alterations (diaphragmatic hernia or omphalocele) [1,6].

Hybrid palliation is effective in high-risk patients with HLHS. Pizarro et al. [7] reported a hospital mortality rate of 21% (7/33) and an interstage mortality rate of 23% (6/26), with no significant differences, compared with the traditional Norwood procedure. Of the 33 study patients, only 48.5% reached stage 2, compared with 47.5% of the patients that had the Norwood surgery. Those results confirm the fact that ne-

onates with HIV are a high-risk group with poorer survival. Studies have shown that the hybrid approach reduces the surgical impact, there is important interstage morbidity and mortality, and survival is similar to that of conventional palliation [6,7].

The hybrid procedure has also been reported to be a bridge to transplantation because it does not significantly alter the anatomy, it optimizes hemodynamic status, enables enteral feeding, reduces the risk for infection, does not utilize transfusions, and requires less monitoring and fewer drugs [1,6]. Even though transplantation is a surgical option, transplants were not performed as a first intention within the study period reported by Galantowicz et al., and only 8 Norwood procedures were performed, reflecting the institution's systemic bias for the hybrid approach in HLHS management [2].

The initial experience at our hospital center is a first step towards further studies that are needed to determine if the hybrid strategy provides better short-term and long-term results and to evaluate the procedure in usual-risk patients, as well. Whether the hybrid approach provides a lower cumulative impact, resulting in better quality and duration of life, also needs to be assessed.

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