HYBRID STRATEGY FOR THE MANAGEMENT OF HYPOPLASTIC LEFT HEART COMPLEX: A SINGLE CENTER EXPERIENCE

G. Perri, S.B. Albanese, A. Polito, S. Filippelli, E. Cetrano, M. Gagliardi, A. Carotti
Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children Hospital, Rome, ITALY

OBJECTIVE:
To report our experience with hybrid approach (HA) for management of Hypoplastic Left Heart Complex (HLHC) and describe the evolution up to our current policy.

METHODS:
From 2007 to date, 27 consecutive newborns underwent HA: 18 (66.6%) had aortic atresia and 9 (33.4%) HLHS-like anomalies. Up to 09/2011 technique included bilateral pulmonary artery banding (PAB) followed by percutaneous arterial duct (PDA) stenting, whereas afterwards PDA stent was delivered through a direct transpulmonary approach. The issue of atrial septal defect restrictiveness was never addressed simultaneously with HA.

RESULTS:
Twelve (44.4%) newborns underwent treatment up to 09/11. There were 4 (33.3%) hospital deaths mainly related to technical problems with PDA stent delivery. Survivors underwent comprehensive stage 2 (CS2) in 5 cases (41.6%) with no deaths, whereas in 3 (25.1%) conversion to Norwood stage 1 (NS1) was necessary. Of the latter patients, 1 died and 2 underwent subsequent bidirectional Glenn (BDG). Among 15 (55.6%) patients treated since 9/2011, neither early nor interstage mortality occurred. Six (35.2%) underwent CS2 with 1 early death, 7 (41.2%) requested conversion to NS1 with 1 late death, and 2 (23.6%) are still waiting for further treatment.

CONCLUSION:
Palliation of HLHC by HA and transpulmonary PDA stent placement can be achieved without mortality. In spite of a fair amount of patients requiring after HA an interstage conversion to NS1, this can be accomplished with satisfactory results. Such finding could lead to hypothesize a novel four-stage approach for optimal treatment of HLHC.
MID-TERM OUTCOME OF HYBRID PALLIATION IN HLHS NEWBORNS: A SINGLE CENTER EXPERIENCE

M. Pilati, M.G. Gagliardi, C. Varano, P. Guccione, A. Carotti, M. Chinali, G. Rinelli, G. Pongiglione
Ospedale Pediatrico Bambino Gesù, Roma, ITALY

Introduction
Hybrid palliative strategy (HPS) - including bilateral pulmonary bands and ductal stent- has emerged as an alternative to Norwood palliation in neonates with hypoplastic left heart syndrome (HLHS). The advantage is the avoidance in the neonatal period of cardiopulmonary bypass and circulatory arrest needed to perform the Norwood procedure, delaying arch reconstruction and Glenn procedure at the time of the comprehensive Norwood palliation. Despite promising preliminary results, some concerns are currently rising on the outcome of hybrid palliative strategy in terms of mid-term morbidity and mortality. Aim of our study is to review outcome in hybrid palliation in newborns with HLHS and its variants.

Methods
Between May 2010 and December 2012, twenty-two (n=22) newborns underwent HPS. Twenty pts were affected by HLHS (with mitro-aortic atresia in 50%), and 2 pts had an interrupted aortic arch. Four pts (18%) showed severe right ventricular dysfunction and 4 pts (18%) had severe tricuspid regurgitation. Median weight at the time of the procedure was 2.9 Kg (range, 1.6 – 3.5 Kg). Three pts (13%), who underwent HPS at the beginning of the learning curve, died during the procedure.

Results
Among the 19 pts surviving HPS, interstage reinterventions were needed to target the ductal stent in 1 (5%) and the atrial septal comunication in 7 (36%). Two pts developed reverse coarctation at 1 month of age and thus underwent Norwood I stage palliation. One pt died awaiting comprehensive Norwood palliation. At pre II-stage catheterization the median ratio of pulmonary flow to systemic blood flow (Qp/Qs) was 0.9 (range 0.75-1.4) and median pulmonary artery pressure was 16 mmHg (range, 12-22 mmHg). At present, comprehensive Norwood palliation has been performed in 10 of the 18 alive pts, with a median age of 6.3 months (range, 4.7-9.2 mos) and median weight of 5.3 Kg (range 4.0-7.3 Kg). Three pts (33%) developed severe stenosis of the left pulmonary artery and underwent a surgical plasty of the pulmonary artery. Among the 10 children who underwent comprehensive Norwood palliation, seven have been discharged with a median peripheral oxygen saturation of 83% (range, 79-89%) and are still alive at follow up.

Conclusions
Hybrid palliation represents a good alternative to neonatal Norwood I stage in HLHS patients. Nevertheless, comprehensive Norwood palliation after hybrid palliation carries significant morbidity. Larger studies with long term follow-up are needed to establish the superiority of HPS as compared to Norwood I stage palliation.
OBJECTIVE:
Aorto-pulmonary collaterals (APCs) are frequent in patients with uni-ventricular heart (UVH). However their clinical significance remains controversial. Quantitative assessment of APCs blood flow using cardiac magnetic resonance (CMR) have been already validated.

Aim to measure APCs flow volume (QAPCs) by CMR and evaluate their impact on early post-operative outcomes after the Fontan operation.

METHODS:
Routine CMR studies were performed in 50 patients with UVH prior to Fontan operation. In 25 of them (med: 4.5 years, range 3-12.8 years) QAPCs was calculated using through-plane phase-contrast as QAPCs = (left pulmonary veins flow + right pulmonary veins flow) - (right pulmonary artery flow + left pulmonary artery flow). Values were normalized to body surface area and indexed to aortic flow (QAPCs/aortic flow) to determine the percentage of cardiac output. No subject had embolization of APCs. A fenestrated total cavo-pulmonary anastomosis was completed in 12 of them, and non fenestrated in 13 patients. The early post-operative outcomes were recorded.

RESULTS:
QAPCs was 1.25 ± 0.6 l/min/m² constituting 39% ± 20% (range, 12-87%) of systemic blood flow. QAPCs didn’t correlates with age at MRI study (p=0.5). Meanwhile QAPCs and QAPCs/aortic flow were associated with decreased RPA diameter (respectively r=-0.44 P= 0.04, r=-0.46, P=0.02) and LPA diameter (respectively r= -0.4, P= 0.04, r=-0.49, P=0.01). Moreover QAPCs correlates with ascending aortic flow (r= 0.41, P=0.03). QAPCs/aortic flow inversely correlates with pre-Fontan 02 sat (r=-0.46, P=0.02) and was associated to overall post-operative adverse outcome (P=0.01) independently of fenestration, ventricle type and function. QAPCs/aortic flow was associated to pleural effusion only in non fenestrated Fontan patients (median 15.4 %, range: 4.3-29% vs median 43 %, range: 8.2-73%, P=0.01).

CONCLUSIONS:
New CMR techniques allow reliable quantification of QAPCs. Their flow volume is higher in pts with hypoplastic pulmonary arteries. Moreover APCs are associated to early adverse outcome after Fontan procedure. Further studies could prospectively evaluate the effect of APCs on immediate and late outcome after Fontan palliation and could help to assess the usefulness and effect of APCs embolization.
PERCUTANEOUS VERSUS SURGICAL PULMONARY VALVE IMPLANTATION. A SYSTEMATIC REVIEW AND META-ANALYSIS OF CURRENTLY AVAILABLE CLINICAL EVIDENCE

Policlinico San Donato IRCCS, San Donato Milanese, ITALY

AIMS
To summarise data from studies comparing surgical (SURG) versus percutaneous treatment (PERC) of right ventricular outflow tract (RVOT) dysfunction.

METHODS AND RESULTS
Electronic databases, journals and major international conference proceedings were systematically searched for pertinent clinical studies comparing the two methods of RVOT treatment (percutaneous (PERC) and surgical (SURG)) in patients aged >5 years, published between 2005 and 2012 and reporting on >20 patients.

Primary endpoints: occurrence of death and of total and major early complications.

Twenty-three original studies (2494 patients: PERC 10 studies: 717 patients; SURG 13 studies 1777 patients) were included. All studies were non-randomized. A total of 20 early deaths were encountered: 3 in the PERC group and 17 in the SURG group (PERC 0.4% (95% CI 0.17-0.63) vs SURG 1% (95% CI 0.77-1.23) p<0.01).

Quantitative synthesis of total major complications after procedure showed a 5.16% (95% CI 3.55-6.77%) rate in PERC patients and 9% (95% CI 8.3-9.7%) rate in SURG subjects (p<0.01).

At a follow-up, late death rate was 1.2% (95% CI 0.8-1.6%) in the PERC group and 0.8% (95% CI 0.54-1.6%) in the SURG group (p N.S.).

Redo procedures were needed in 5.4 % (95% CI 4.56-6.24%) in the PERC group and 5.1 % (95% CI 4.6-5.6%) in the SURG group (p=N.S.).

Endocarditis during follow-up was reported in only 2 subjects in the SURG group (0.11 (95% CI 0.03-0.19) and in 11 subjects in the PERC group (1.5 % (95 CI 1.05-1.95%) p<0.001.

CONCLUSIONS
The largest cohort to date of patients undergoing treatment of RVOT dysfunction shows that treatment by a percutaneous approach has a significantly lower rate of both early death and major early post-procedural complications.
HYBRID SUITE: A NEW WAY TO THE HEART

Centro Cardiologico Pediatrico del Mediterraneo-IRCCS Ospedale Pediatrico Bambino Gesù, Taormina, ITALY

Background

The field of catheter-based therapies have clearly demonstrate safety and efficacy in the area of pediatric cardiology. Recent developments in cardiac surgery and interventional cardiology have led to the installation of integrated operating rooms that allow both surgical and endovascular procedures. These units offer surgical as well as angiographic equipment and personnel and therefore require special planning and design. This technical innovation facilitates complex hybrid operations in a sterile environment. A variety of integrated procedures can be performed through different new "way" to the heart.

Methods

According to a bilateral program between Sicilian Ministry of Health and Ospedale Pediatrico Bambino Gesù – Roma, since October 2011 at Centro Cardiologico Pediatrico del Mediterraneo-Taormina, was activated an “Hybrid Suite” fully dedicated to the treatment of patients affected by congenital heart disease under 18 years of age.

All consecutive patients with diagnosis of hypoplastic left heart syndrome (HLHS) and its variants were electively treated by single stage pulmonary artery banding and anterograde transpulmonary ductal stenting. Muscular ventricular septal defect were firstly approached by beating heart perventricular closure. Relief of the obstruction in the case of pulmonary atresia with intact ventricular septum (APSI) was performed by retrograde transpulmonary to right ventricle puncture and dilatation or stenting. Elective exit angiograms were performed in all patients after comprehensive stage II while urgent control in the cases of unsuccessfull weaning from CPB with suspicion of residual post surgical lesion.

Results

49 procedures were performed under hybrid approach through full median sternotomy. Several "way" to the heart were used by direct puncture or introducer cannulation: main pulmonary artery, selective right or left pulmonary branch, ascending aorta, right ventricle, right or left superior vena cava both for diagnostic or interventional procedure. 72% (35 pts) were primary procedure - median weight 3,2 Kg (range 2,2 – 6 Kg) - : # 24 HLHS or complex; # 5 closure muscular ventricular septal defect; # 3 APSI through retrograde pulmonary dilatation or stenting; # 2 ductal stenting; # 1 stenting of the right pulmonary artery after Glenn operation. 28% (14 pts) of the cases were secondary procedure median weight 5,7 Kg (range 4,5 – 7,8 Kg): # 11 elective exit angiograms after comprehensive stage; # 2 urgent control post surgical procedure; # 1 right pulmonary rebanding. Two patients were on ECMO. No major neither minor procedure related adverse events occured and all patients completed the procedure successfully.

Conclusion

By maximizing the use of existing technologies while developing new approaches to treating these challenging cases, we hope that our experience would lead to improve overall clinical outcomes and further reduce the mortality and morbidity rates associated with managing the complex cardiovascular congenital patient. It is hoped that as these new fields develop and with increasing experience with these new hybrid methods, we may well be able to maximize the applicability of minimally invasive endovascular and hybrid technology to treat a larger cohort of patients.
Background
Use of percutaneous pulmonary valve implantation (PPVI) is currently limited to patients with right ventricle-pulmonary artery conduit. Few reports have described PPVI in patients with severe pulmonary disease following repair of tetralogy of Fallot (TOF) with right ventricular outflow tract (RVOT) patch. In the present study we evaluated feasibility and safety of PPVI in native RVOT.

Methods
Eight pts (n=8) with failure of native outflow tracts underwent PPVI between October 2011 and May 2013 and followed up for a median of 18m months (3 to 24 months). Primary diagnosis was corrected TOF in all pts. Eligibility criteria were: weight > 28 Kg, RVOT diameter > 20mm and< 26mm, measured by either MRI or CT scan. Median age was 18 years (range, 9-30yrs) and median weight was 56Kg (range, 30-86 Kg). Indication for PPVI was severe pulmonary insufficiency.

Results
RVOT measurements obtained by non invasive diagnostic tools were confirmed in all patients by angiography before the procedure. Six 23- and one 26-Sapien Edwards valve were implanted successfully (n=7; 87%). In one pt during pre-stenting for PPVI right pulmonary artery was occluded, thus surgical removal of the occlusion with subsequent valvolization was performed. All patients showed fully competent pulmonary valves at end procedure. In all pts pre-stenting (7 bare metal stents, 8 covered stents) was performed. In four pts pre-stenting was performed 2 to 3 months before the procedure in order to allow stent endothelial ingrowth. At follow up all pts had improvement in symptoms with no evidence of stent fracture or valve failure.

Conclusions
Adequate selection criteria using non invasive diagnostic tools, allow successful PPVI using the Sapien Edwards Valve in TOF patients with native RVOT.
HEARTWARE HVAD AS BRIDGE TO TRANSPLANT IN CHILDREN AND ADOLESCENTS: THE BEST OPTION IN “GREATER” KIDS

M. Padalino 1, T. Bottio 2, G. Bortolussi 1, A. Cerutti 3, O. Milanesi 3, G. Gerosa 2, G. Stellin 1

1 Pediatric and Congenital Cardiac Surgery, Department of Cardiac Thoracic and Vascular Sciences, Padua, 2 Cardiac Surgery and Heart Transplantation Unit, Department of Cardiac, Thoracic and Vascular Sciences, Padua, 3 Pediatric Cardiology, Department of Women and Children’s Health, Padua, ITALY

Ventricular assist device (VAD) support is a well established therapy for treatment of adult patients with advanced heart failure. Children with end stage heart failure refractory to conventional therapy have much fewer mechanical support options. We describe our preliminary experience with Heartware LVAD, a third generation continuous flow intracorporeal VAD which was successfully employed in 3 pediatric patients < 16 years of age with dilative cardiomyopathy.

Methods
The HeartWare System is a small centrifugal flow pump with a displacement volume of 50 ml, weighs 140 g, and an output capacity from 2 up to 10 L/min, at 1,800 to 4,000 rpm. A unique wide-blade impeller is suspended by hybrid passive magnets and hydrodynamic forces. An integrated inflow cannula is inserted into the left ventricle and it is held in position by an adjustable sewing ring; the pump is positioned in the pericardial space, avoiding the need for an abdominal device pocket. The 10-mm outflow graft is anastomosed to the ascending aorta.

Clinical cases
All patients were clinically followed for chronic congestive heart failure at our Department of Pediatrics. They were all admitted in hospital because of severe and progressive deterioration of their clinical conditions and heart failure, despite full anticongestive therapy. Two of them had an AICD positioned a few months before admission and were already on a transplant list. All children were first admitted to the wards, but because of progressive clinical deterioration they were transferred to the intensive care unit (ICU) where they were all supported with inotropic iv drips (milrinone, dopamine, dobutamine iv infusion), until they were scheduled for LVAD. After LVAD surgery, they were all treated with Nitric Oxide (NO) inhalation therapy (starting with 30 ppm), and epinephrine, norepinephrine, dopamine, and milrinone iv infusion. As long as clinical conditions and RV function were improving, inotropic iv support was gradually weaned off. After extubation, all but patient 3 were treated with oral therapy with Sildenafil 1mg/kg TID. All detailed patients’ characteristics, preoperative conditions and outcomes are described in Table 1. At a mean follow up of 10 months, all children are in very good clinical conditions, with excellent quality of life, on immunosuppressant therapy with prednisolone, cyclosporine and mycofenolate. Patients 1 and 3 experienced one episode of acute rejection grade 3A a few months after discharge, and were treated successfully with prednisone boluses.

Conclusions
The HeartWare system is a novel third generation, implantable VAD, which creates continuous flow to support even children in end-stage cardiac failure. It offers an attractive and safe alternative to paracorporeal systems for children and adolescents with ESHF. Despite HVAD implant is feasible in smaller patients, we recommend it especially in children with a BSA > 1.0 m2 or body weight > 20 kg. As our experience suggest, in this subgroup, HVAD has demonstrated to be a safe and effective VAD system with a very low rate of complications.
PDA STENTING IS SUPERIOR TO SURGICAL SHUNT IN PATIENTS WITH SINGLE VENTRICLE PHYSIOLOGY AND DIMINISHED PULMONARY BLOOD FLOW

N. Maschietto 1, M. Padrini 1, B. Castaldi 1, V.L. Vida 2, M. Padalino 2, G. Stellin 2, O. Milanesi 1
1 Dipartimento della Salute della Donna e del Bambino, Unità di Cardiologia Pediatrica, Università di Padova, Padova, Italy
2 Dipartimento di Scienze Cardiovascolari, Unità di Cardiochirurgia Pediatrica, Università di Padova, Padova, Italy

Introduction
Duct dependent pulmonary blood flow in univentricular heart circulation is still a challenge for both the cardiologist and cardiac surgeons. Mortality rate for surgical shunt procedures are still very high ranging from 7.7 to 11.6%. Given the lower mortality and the less invasivity, PDA stenting is becoming an attractive alternative.

Aim of the present study is to compare the outcomes of patients born with a single ventricle physiology and a reduced pulmonary blood flow (PBF) who underwent either a surgical shunt or a PDA stenting.

Methods
Between 2002 and 2013, every patient with a single ventricle physiology requiring an intervention to increase the PBF was included in the study.
Variables analyzed were: hospital mortality, length of stay in the ICU and in the hospital following either surgical shunt or PDA stenting, inotropic score in the first 48 hours and hours of mechanical ventilation following one of the two different operative strategies and incidence of major complications (defined as cardiac arrest, major arrhythmias, stroke, need for mechanical circulatory support, sepsis).

Results
Between 2002 and 2013, 38 patients with a single ventricle physiology and PGE dependent PBF underwent either a surgical shunt or a PDA stenting. 25 patients underwent surgical shunt and 13 patients underwent PDA stenting.
There was one death in the stent group (7.6%) compared to 5 (19%) in the surgical group (p 0.00001).
In the first 48 hours the mean inotropic score in the stent group was 0.6 +/- 1.6 compared to 11.6 +/- 6.1 in the surgical group (P 0.00001)
In the stent group mechanical ventilation was required for 32.4 +/- 42 hours post intervention compared to 178.1 +/- 146 hours in the surgical group (p 0.002)
In the stent group patients were discharged from the ICU 3.5 +/- 4.6 days after the procedure compared to 10.7 +/- 8.2 days after surgery (p 0.018). Hospital discharged occurred 11 +/- 4 days after stent placement compared to 21.9 +/- 9.9 days after surgery (p 0.003).
Major complications occurred more frequently in the surgical than in the stent group (p 0.0001)

Conclusion
PDA stenting is a safe and effective alternative to surgical shunts given the lower mortality and morbidity, the better hemodynamic stability and the shorter hospital stay. PDA stenting can be a valid alternative to surgical shunt in selected cases.