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Stent Therapy for Clotted Blalock-Taussig Shunts

By Emmanouil Tsounias, MD and
P. Syamasundar Rao, MD

INTRODUCTION

Patients with large inter-ventricular communication and severe right ventricular outflow obstruction, including pulmonary atresia, require surgical intervention. Corrective surgery, when feasible, is the preferred treatment option. When total correction is not feasible, either because of the age and size of the patient at presentation or because of anatomic complexity, surgical palliation with a variety of aorto-pulmonary shunts (reviewed elsewhere¹) is undertaken. Subclavian artery to ipsilateral pulmonary artery anastomosis, first described by Blalock and Taussig² stood the test of time and became a standard mode of therapy. More recently, insertion of an interposition Gore-Tex graft (W.L. Gore & Associates, Inc., Flagstaff, AZ) between the subclavian artery and ipsilateral pulmonary artery, commonly referred to as modified Blalock-Taussig (mBT) shunt, described by deLeval et al,³ has become the procedure of choice. In patients with single ventricle physiology, palliation with first stage Norwood procedure is performed; in such patients, mBT shunt is commonly used to provide pulmonary blood flow.

Complete or partial obstruction of these shunts can occur causing acute hypoxemia or, in more severe cases, shock and

collapse. If stenosis or obstruction develops before planned surgical correction, additional palliative surgery may be necessary thus increasing morbidity and inter-stage mortality. The objectives of this presentation are to describe our experience with percutaneous implantation of stents in the Gore-Tex grafts to successfully palliate infants with blocked mBT shunts and to discuss the management issues related to this subject.

"Patients with large inter-ventricular communication and severe right ventricular outflow obstruction, including pulmonary atresia, require surgical intervention. Corrective surgery, when feasible, is the preferred treatment option."

MATERIAL AND METHODS

Between December 2004 and January 2008, three infants (2 males, 1 female; 12 days to 4 months of age; median weight of 4 Kg), two

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Table I. Summary of Cases Undergoing Stent Therapy for Thrombosis of BT Shunt

	Case #1	Case #2	Case #3
Primary Diagnosis	TOF	HLHS	HLHS
Age at Shunt Surgery	14 Days	7 Days	7 Days
Shunt (Gore-Tex graft) Diameter	5 mm	4 mm	4 mm
Time to Occlusion	28 days	5 days	3.5 months
Weight at Stent Implantation	4.9 Kg	4.0 Kg	4.0 Kg
Preceding Thrombolysis	Yes	Yes	Yes
Preceding Balloon Angioplasty	Yes	No	Yes
Stent Type Size (mm)	Express II (Scimed) Diameter: 5; Length: 16	Driver (Medtronic) Diameter: 4; Length:15	Driver (Medtronic) Diameter: 4; Length:15
Percent increase in Arterial O ₂ Saturation Following Stent Deployment	40%	40%	41%
Patency of Stent by Angiography Immediately After Stent Placement	Widely Patent (Figure 1D)	Widely Patent (Figure 2F)	Widely Patent (Figure 3D)
Follow-up Shunt Patency	Patent – 6 months – Echo	Patent – 6 months – Echo & Angiogram	Patent – 2 months – Echo & Angiogram
Follow-up Outcome	Lost to Follow-up	Bidirectional Glenn	Bidirectional Glenn

BT, Blalock-Taussig; Echo, Echo-Doppler study; HLHS, Hypoplastic Left Heart Syndrome; O₂, oxygen; TOF, Tetralogy of Fallot.

with Hypoplastic Left Heart Syndrome (HLHS) and one with Tetralogy of Fallot (TOF) with pulmonary atresia were referred for emergency transcatheter recanalization of the mBT shunts (Table I) for severe life threatening hypoxia (oxygen saturation <40%), with acute shunt occlusion not responsive to pharmacologic thrombus dissolution. Indications for the procedure were severe hypoxemia, metabolic acidosis, hypotension and shock. The technique involved initially defining the angiographic anatomy, followed by mechanical recanalization and access of the shunt beyond the stenotic region using soft tipped guide wires and right coronary artery (RCA) catheters (Cook, Bloomington, IN). Four to 5 mm-diameter (15 to 16 mm-long) premounted stents (Driver [Medtronic, Minneapolis, MN] or Express II [Scimed, Maple Grove, MN]) were positioned within the Gore-Tex graft

of the mBT shunt and the balloon inflated, thus implanting the stents. Post stent oxygen saturations and angiograms were recorded and compared with those obtained prior to stent. Immediate success is defined as improvement in arterial oxygen saturation to 75% or higher. Success at follow-up is defined as the infant reaching the age and size suitable for next stage palliation (bidirectional Glenn in HLHS patients) or corrective surgery (total correction of TOF patients).

CASE REPORTS

Case # 1. This is a six week old male infant with pulmonary atresia, a large ventricular septal defect, right aortic arch and a small patent ductus arteriosus who underwent a left mBT shunt at age two weeks. He did well in the post operative period. During fundoplication and gastric

tube placement four weeks later, the patient developed hypoxemia and hypotension with ensuing cardiac arrest; he was successfully resuscitated. Patient was started on PGE₁ infusion which resulted in minimal increase in the oxygen saturation. Echocardiogram revealed complete occlusion of the mBT shunt. The patient was taken to the Cardiac Cath Lab and a left subclavian artery cineangiogram was performed which revealed complete blockage of the mBT shunt. Mechanical thrombolysis with RCA catheter and guide wires was performed. Then balloon angioplasty of the mBT shunt with a 6-mm diameter balloon was performed, followed by local heparin infusion for 24 hours. Repeat cineangiograms the next day revealed sub-total occlusion of the shunt (Figure 1A). A 5-mm diameter, 16-mm long balloon expandable Express II stent (Scimed) was selected for implantation.

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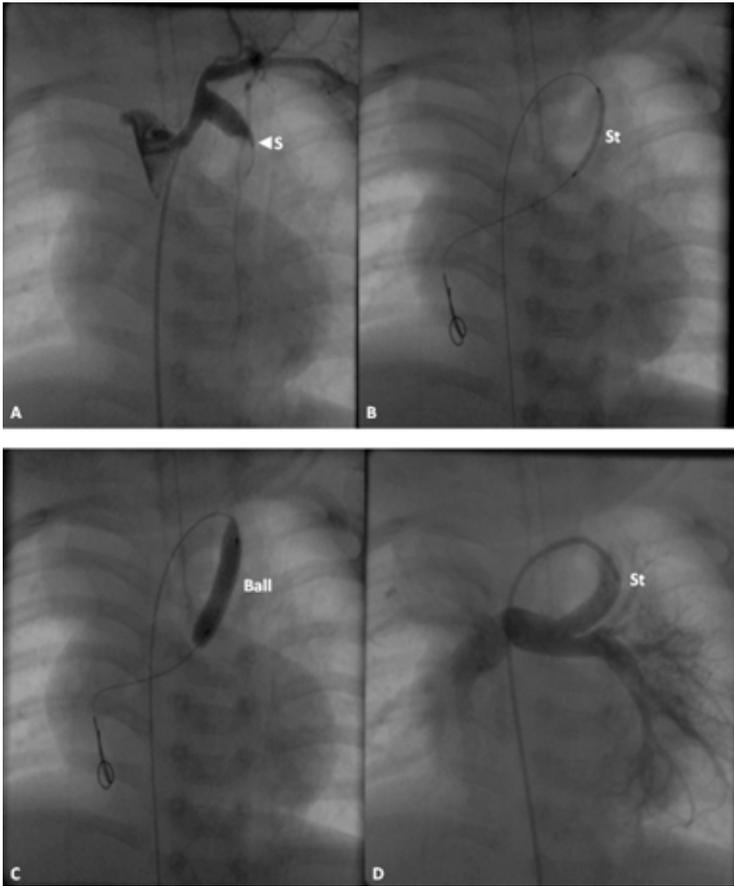


Figure 1 (Case #1). A. Selected cineangiographic frame from subclavian artery injection demonstrating stenotic and almost completely obstructed (arrowhead S) Blalock-Taussig shunt. B. A guide wire has been passed through Blalock-Taussig shunt and its tip positioned in the distal right pulmonary artery over which an unexpanded stent (St) is placed within the Blalock-Taussig shunt. C. Same as B except that the stent-covered balloon (Ball) is inflated. D. Post-stent implantation angiogram showing the well opacified, widely patent stent (St) in the modified Blalock-Taussig shunt with subsequent opacification of the branch pulmonary arteries.

The shunt was successfully cannulated using 4 French right coronary artery (RCA) catheter and a 0.035" Benston straight guide wire. A 0.014 hi torque Spartacore guide wire (Guidant, St. Paul, MN) was positioned into the distal lower lobe branch of the right pulmonary artery and the right coronary artery catheter removed. The selected balloon-mounted stent catheter was introduced over the wire and positioned across the mBT shunt (Figure 1B). The balloon was then inflated up to 16 atm of pressure (Figure 1C), thus implanting the stent. Repeat cineangiogram revealed a wide open stent with good flow (Figure 1D) into the pulmonary arteries. The oxygen saturations improved to 80%. Following return to the observation unit, heparin, 50 units/Kg as bolus was given intravenously, followed by heparin drip of 20 units/Kg/hour and PTT maintained at 1.5 to 2X normal by adjusting heparin dose. The heparin was discontinued 48 hours later. Aspirin (40.5 mg) was continued and the baby discharged home a week later. Follow-up evaluation one and six months after discharge revealed good oxygen saturations (85%), no signs of heart failure and wide open shunt on echo-Doppler studies. The baby was lost to further follow-up.

Case # 2. A 12-day old female infant with complex congenital heart disease (forme fruste HLHS) including a single ventricle with mitral

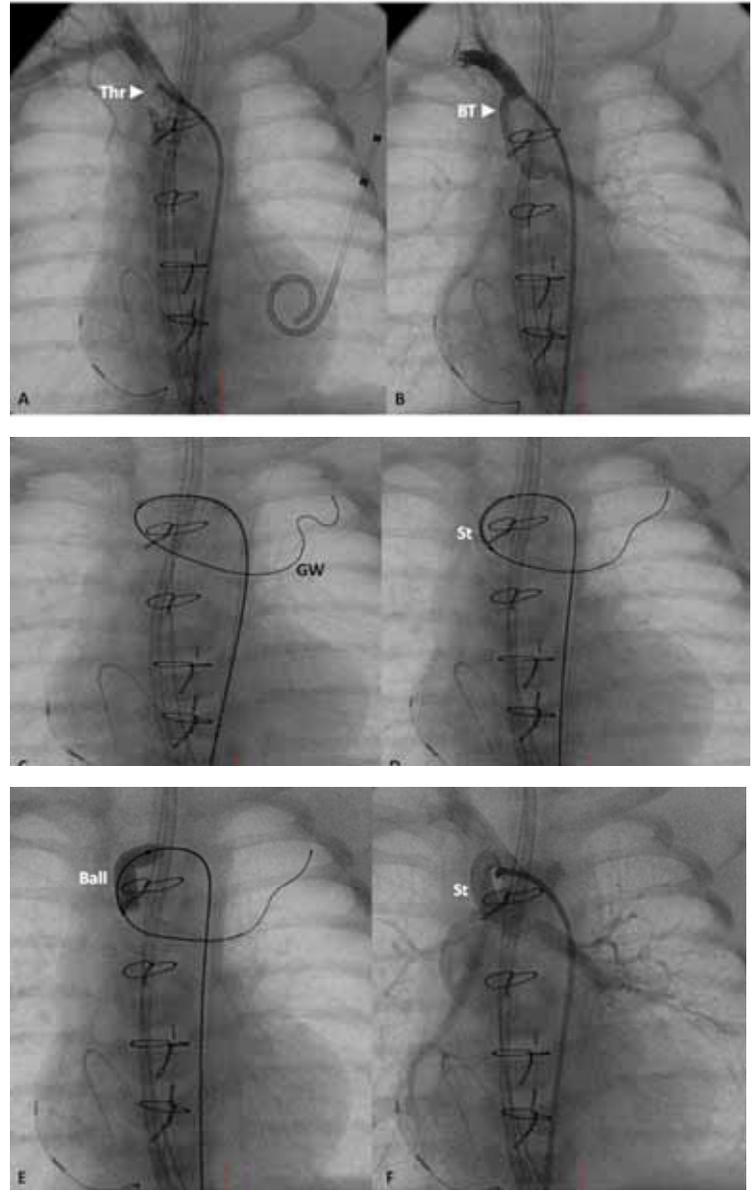


Figure 2. (Case #2) A. Selective right innominate artery cineangiographic frame demonstrating complete blockage of the BT shunt; note radiolucent mass (Thrombus - Thr) marked with arrowhead. B. Following mechanical thrombolysis, partial recanalization of the Blalock-Taussig (BT) shunt was seen. C. Selected cineradiographic frame showing a hi-torque floppy guide wire placed across the Gore-Tex graft into pulmonary artery. D. The stent (St) was advanced over the wire and positioned within the Gore-Tex graft. E. Selected cineradiographic frame demonstrating inflated stent-mounted balloon (Ball) in the Gore-Tex graft. F. Selected right innominate artery cineangiographic frame showing opacification of the stent (St) and both branch pulmonary arteries.

atresia, hemitruncus with left pulmonary artery arising from the ascending aorta and multiple aorto-pulmonary collateral arteries (MAPCAs), underwent unifocalization along with a right mBT shunt using 4-mm diameter Gore-Tex graft at age 7-days. On the 5th post-operative day, while about to be transferred to intermediate care unit, the baby suddenly developed severe cyanosis with an oxygen saturation of 40%. Echo-Doppler study did not demonstrate Doppler flow in the BT shunt. While waiting to be transferred to the Cath Lab, medical therapy including volume expanders, inotropic support, 100% oxygen and nitric oxide and recombinant tissue

plasminogen activator (r-tPA) administration increased the oxygen saturations to low 60s. Selective cineangiography revealed complete blockage of the BT shunt (Figure 2A). Mechanical thrombolysis with a RCA catheter and a 0.035" removable core straight tip guide wire was performed. Repeat cineangiogram (Figure 2B) revealed some dissolution of the thrombus. The mBT shunt graft measured 13.4 mm x 3.4 mm diameter. Based on these data, a 4 mm-diameter, 15 mm long Driver (Medtronic) stent was selected for implantation. Initially, a hi-torque floppy guide wire was placed across the Gore-Tex graft into the pulmonary artery (Figure 2C). The stent was advanced over the wire (Figure 2D) and based on fixed landmarks, the stent position was verified and the balloon was inflated (Figure 2E) twice, initially for 10 sec at 10 atm. pressure, then for 6 seconds at 13 atm. pressure. Once the stent was successfully deployed, the oxygen saturations improved to 80%. The stent delivery catheter was removed and a 4-French multipurpose catheter was positioned proximal to the stent and an angiogram was performed which revealed wide open mBT shunt with opacification of both pulmonary arteries (Figure 2F). Anticoagulation with heparin followed by aspirin, as in the first case, was instituted. Follow-up echocardiograms one and five months later, as well as an angiogram at six months of age revealed wide-open shunt. The patient had bidirectional Glenn at the age of six months.

Case # 3. A 4-month old male with forme fruste Hypoplastic Left Heart Syndrome, Shone's variant, coarctation of the aorta, mesocardia and s/p Damus-Kaye-Stansel/Norwood procedure with

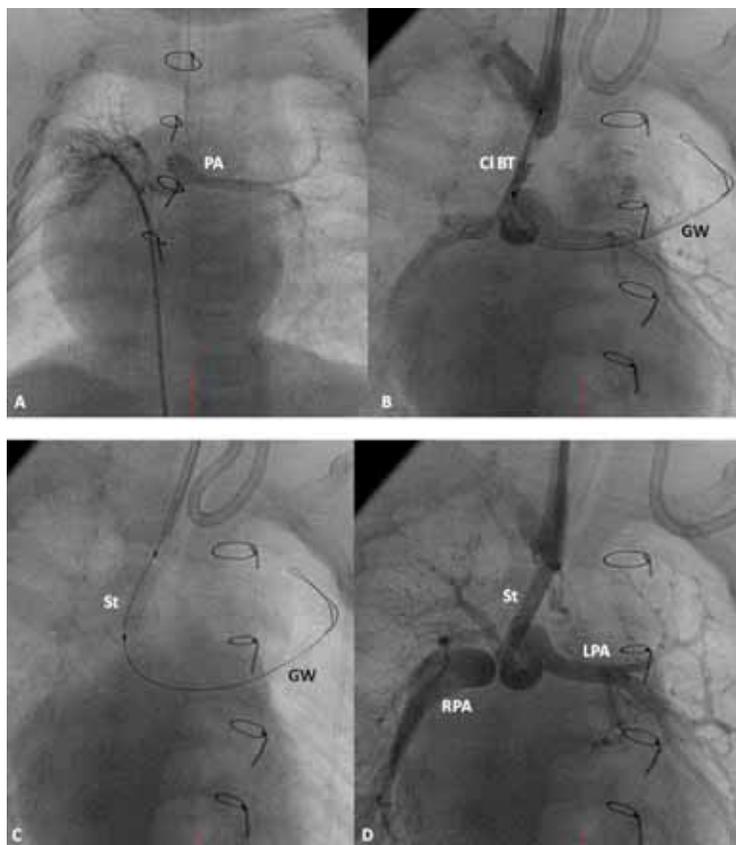


Figure 3. (Case #3) A. Pulmonary venous wedge cineangiographic frame demonstrating pulmonary arteries (PA). B. Following r-tPA for 24 hours and mechanical thrombolysis, the shunt (Cl BT) is partially open with radiolucent clots. The position of the guide wire (GW) across the BT shunt into the left pulmonary artery is also shown. C. Cineradiographic frame showing the implanted stent (St). The guide wire (GW) is still in place. D. Post-stent implantation cine angiographic frame demonstrating wide-open stent (St) with good opacification both the right (RPA) and left (LPA) pulmonary arteries.

a right mBT shunt (4 mm-diameter Gore-Tex graft) at age 7 days, presented to the emergency room with severe cyanosis and had an oxygen saturation of 37%. Echocardiogram revealed the mBT shunt to be completely occluded, without evidence of flow through the shunt and no flow into the pulmonary arteries. An aorto-pulmonary collateral vessel provided minimal blood supply to the lungs. The patient was started on r-tPA (initially 0.1 mcg/kg/min and was later increased to 0.3 mcg/kg/min) and was taken to the Cath Lab approximately 24 hours later to evaluate the shunt. Transvenous aortic arch angiography revealed completely occluded mBT shunt, presumably thrombosed. Pulmonary venous wedge angiography revealed small but patent distal pulmonary arteries (Figure 3A). A collateral vessel from the descending aorta to left lower lobe was seen (not shown). Despite multiple attempts, we were unable to advance a wire through or around the thrombus. The patient was restarted on r-tPA and heparin was added. Repeat cardiac catheterization was performed the next day via right common carotid artery cut-down. Cineangiograms revealed partially occluded mBT shunt which measured 13.2 mm x 1.7 mm. A 4-French multipurpose catheter was placed proximal to the shunt. Rapid to and fro movement of the Benston straight guide wire in and out of the shunt, as well as vigorous catheter manipulation to disrupt the clot to improve flow across the shunt, was undertaken. Recanalization was partially achieved by mechanical clot disruption (Figure 3B). A 4 mm-diameter, 15 mm-long Driver stent (Medtronic) was selected for implantation. The catheter was advanced into the pulmonary artery and the guide wire was removed. A Choice PT Extra Support guide wire (Boston Scientific, Natick, MA) was introduced and the catheter was removed. The selected balloon-mounted Driver stent was introduced over the wire, through the carotid arterial sheath and positioned across the mBT shunt. The balloon was inflated at 9 atm of pressure thus deploying the stent (Figure 3C). Follow-up cineangiograms revealed wide open stent with good flow (Figure 3D) and improved O₂ saturations (78%). He underwent bidirectional Glenn procedure two months later.

RESULTS

In summary, successful stent implantation into the BT shunt was performed in all cases; thus the technical success rate was 100%, with complete revascularization of all stented shunts (Table I). No procedural complications were encountered. There was no acute in-stent re-thrombosis, stenosis or stent kinking. Following stent deployment, the oxygen saturations improved to 78 to 80% and the stents implanted were wide open by angiography. No vessel injury or perforation was encountered. The patients did not require immediate or subsequent surgical revision of the stent implanted shunt. Two HLHS patients underwent subsequent bidirectional Glenn procedures, and the patient with TOF and pulmonary atresia was lost to follow-up.

DISCUSSION

As mentioned in the introduction section, surgical aorto-pulmonary shunts are performed if total surgical correction of a given cyanotic heart defect with pulmonary oligemia is not feasible. Modified BT shunts³ are most commonly used for this purpose. Similarly, mBT shunts³ are also used in Norwood palliation of HLHS patients although some surgeons use Sano shunts.⁴

Causes of Obstruction. Obstruction of such surgical stents can occur, causing hypoxemia. While causes of such obstruction are not clearly understood, it may be related to thrombosis, suture line scar formation and stricture, neo-intimal proliferation and vascular distortion/stenosis due to surgery or constriction of ductal tissue remnants or a combination thereof. Administration of platelet-inhibiting doses of aspirin to prevent thrombosis of the shunts has long been in practice and all three of our patients have been on aspirin.

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Clinical Features. Disappearance of a previously documented continuous murmur and a fall in arterial oxygen saturation are indicative of shunt occlusion, especially if the shunt is the only or the major source of pulmonary blood flow. Echo-Doppler studies using pulsed and continuous wave Doppler interrogation and color flow mapping are useful in the evaluation of the patency of the shunt. Selective innominate/subclavian artery angiography is confirmative.

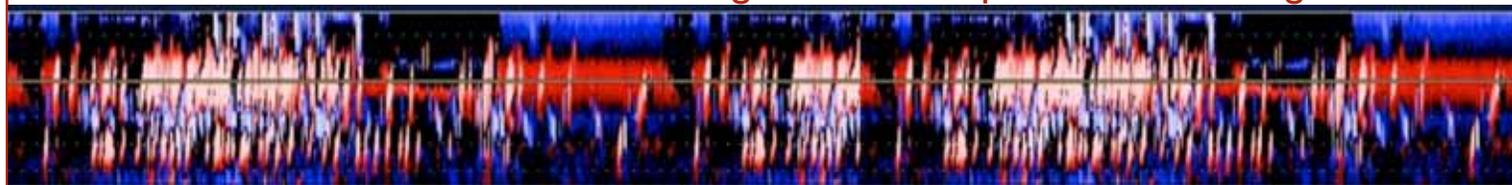
Management Options. If stenosis or occlusion of the Blalock-Taussig (BT) shunt develops prior to the planned surgical correction or bidirectional Glenn procedure, additional palliative surgery may become necessary. In such situations, transcatheter intervention to relieve the obstruction should be considered. Balloon dilatation of the narrowed BT shunt, first described by Fisher et al,⁵ is an option; they reported improvement in oxygen saturation from 68 to 80% following balloon angioplasty of BT shunt in a 4-year-old child. Subsequently, a number of cardiologists, including our group⁶ have reported on the experience with balloon angioplasty of both classic and modified BT shunts, reviewed elsewhere.⁷⁻⁹ The majority of this experience is with dilating stenotic lesions that develop months or years after the creation of BT shunts, and may not be truly applicable to acutely clotted shunts that are being addressed in this report. Furthermore, balloon angioplasty is not uniformly effective^{9,10} and some develop restenosis with time. Placement of stents across the stenosed or occluded BT shunts to ensure their patency appears to be an attractive option.

Management of acutely clotted shunts in the past was largely limited to surgical shunt revision. Less invasive methods such as pharmacological thrombolysis,¹¹⁻¹⁷ mechanical thrombolysis,^{18,19} balloon angioplasty¹⁹⁻²⁷ and placement of stents²⁷⁻³⁶ and a combination thereof are increasingly used in the initial treatment. Successful use of fibrinolytic therapy with r-tPA or streptokinase in the treatment of thrombosed BT shunts has been reported.¹¹⁻¹⁷ However, the risk of bleeding complications, especially if reoperation is required, and failure of fibrinolytic therapy, requiring reoperation suggest that fibrinolytic therapy may not be ideal for use in early postoperative period. While balloon angioplasty has been used with success,¹⁹⁻²⁷ several patients required stent placement to adequately address the shunt occlusion. Moszura et al³⁶ reviewed their experience with recanalization of shunt thrombosis/occlusion in 23 patients; 14 of these were acute occlusions. Balloon angioplasty with or without local r-tPA administration was undertaken in 22 of 23 patients with improvement. However, three of their patients required stent implantation.

Stent Therapy. Zahn and his colleagues²⁸ were the first to report stent implantation to successfully treat a thrombosed mBT shunt; they implanted a Palmaz-Schatz (Johnson & Johnson Interventional Systems Co., Warren, NJ) stent in an 8-day old baby with HLHS who developed shunt thrombosis following Norwood procedure in less than 24 hours after surgery. The oxygen saturation improved by 42% following stent deployment. No additional interventions were necessary until bidirectional Glenn procedure at the age six months. Subsequently a number of interventional cardiologists reported use of the stents to treat acutely thrombosed mBT shunts.^{27,29-36}

Deployment of stents for treating obstructions that develop shortly after surgical creation of shunt²⁷⁻³⁶ as well as those that occur during follow-up^{31,34,36,37} appears possible. Implantation of stents is feasible and effective in thrombotic, obstructive lesions that develop spontaneously^{28-31, 33-36} and those that develop after catheter manipulation within the shunt.³² Successful treatment is possible in

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obstructive lesions at proximal^{31,34,37} or distal^{27,31} anastomotic sites, as well as those in mid-portion of the graft secondary to intimal proliferation.³⁷ Stent therapy appears feasible and effective in mBT shunts,²⁷⁻³⁶ classic BT shunts,³¹ central aorto-pulmonary Gore-Tex grafts^{35,38} and Sano shunts.^{27,34,35,39} The experience with cases described in the current report supplements that described previously²⁷⁻³⁶ in relieving hypoxemia secondary to acutely clotted BT shunts.

Technique. The procedure involves initially defining the site and extent of obstruction by selective cine-angiograms from the innominate or subclavian artery (Figures 1A and 2B). Probing the shunt using angled catheters (RCA, Cobra, cut-pigtail or angled glide) and soft-tipped straight guide wires (Straight Benston, removable-core straight Amplatz, glide or Woolley guide wires) to produce mechanical thrombolysis may be helpful in at least partial recanalization of the thrombosed shunt. Adequate anticoagulation should precede these maneuvers. The shunt should then be crossed with a soft-tip floppy wire over which balloon catheters (same size as the Gore-Tex graft diameter) are inserted and balloon dilatation performed. If there is not an adequate increase in oxygen saturation or angiography shows significant residual thrombus (Figures 1A, 2B and 3B), stent implantation should be undertaken. The currently available coronary stents track well over the coronary guide wires. Some interventionalists^{27,31,34} use long sheaths to position the stents; this may result in hemodynamic instability and femoral arterial complications. We have had success in delivering these stents via short 4-French sheaths in the femoral artery (Figures 1B and 2D). Stent deployment should be followed by selective cineangiography to demonstrate the patency of the stented shunts (Figures 1D, 2F and 3D). While we have utilized fibrinolytic agents such as r-tPA prior to the procedure, administering locally with a catheter left in place for an extended period of time, in some of our patients, there is no unanimity of opinion regarding their use. Based on lack of success in our cases we do not advocate their routine use. However, anticoagulation with heparin drip following stent implantation followed by aspirin is recommended by most interventionalists.

Additional Considerations. Infusion of prostaglandin E₁ (PGE₁) in neonates may help open the ductus which may improve the oxygen saturation. Addressing the proximal stenosis (of the subclavian⁴⁰ or innominate³¹ artery) and branch pulmonary artery stenosis^{10,31,34,36} either by balloon angioplasty or stent implantation is important to ensure the patency of the stented shunt and to provide adequate pulmonary blood flow.

Apart from stent implantation into the occluded shunt, alternative solutions such as stenting the stenosed aorto-pulmonary collateral vessels,^{37,41} stenting of the right ventricular outflow tract,^{26,36} balloon valvuloplasty of the pulmonary valve^{31,42} and radiofrequency or blunt guide wire perforation of the atretic pulmonary valve⁴³⁻⁴⁶ should also be considered with the intent to augment pulmonary blood flow when managing these hypoxemic infants. However, the use of such procedures should be in accordance with achieving long-term goals of biventricular correction or separation of circuits by Fontan route, as the case may be.

Recommendations. In our patient cohort, we initially infused heparin or r-tPA to dissolve the clots for 24 hours prior to going to the Cath Lab, performed mechanical thrombolysis including balloon angioplasty and administered heparin and/or r-tPA with the catheter tip placed close to the shunt and none were effective. Deployment of stents appears to have achieved the best and most lasting result. Based on this experience we would recommend mechanical thrombolysis with the catheters/guide wires used for cannulation of the mBT shunt (to reduce the clot burden) followed by balloon angioplasty, and if that is not successful, stent placement at the same sitting for treatment of hypoxemia secondary to acute thrombosis of BT shunts. Alternative ways to augment pulmonary blood flow such as stenting the stenosed

aorto-pulmonary collateral vessels, stenting of the right ventricular outflow tract, balloon dilatation of stenotic pulmonary valve and guide wire perforation of atretic pulmonary valve, as alluded to above, should also be entertained when dealing with hypoxemic infants secondary to pulmonary oligemia.

Much larger experience than is reported thus far, further miniaturization stent delivery systems, and greater flexibility of the stents to pass through tortuous courses may further improve feasibility, safety and effectiveness of this mode of therapy.

CONCLUSIONS

The experience in stenting aorto-pulmonary shunts, though limited, appears feasible, safe and effective. Improvement in oxygen saturation, the primary objective, was achieved in all our cases. The procedure, though labor intensive, is a useful technique in alleviating hypoxemia and allowing the patient to reach the next step palliation of their complex cyanotic congenital heart disease.

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TRANSCATHETER PULMONARY VALVE (TPV) THERAPY

Hybrid Procedures: Evolution of Change in Managing Congenital Heart Disease

By Sharon L. Hill, MSN, ACNP; John P. Cheatham, MD; Ralf J. Holzer, MD, MSc; Alistair Phillips, MD and Mark Galantowicz MD

Collaboration between cardiothoracic surgery and interventional cardiology has evolved over the past decade resulting in increased Hybrid procedures and ultimately improved outcomes for patients with congenital heart disease (CHD). Infants with Hypoplastic Left Heart Syndrome (HLHS), muscular ventricular septal defect (MVSD), and complex pulmonary artery stenosis are examples of patients who have benefited from Hybrid procedures. The aim of Hybrid therapy is to combine surgical and interventional techniques in an effort to

decrease morbidity and mortality and lessen the impact of multiple procedures over the lifetime of a patient. Thinking collaboratively “outside the box” has led to decreased invasive procedures (i.e. avoidance of cardiopulmonary bypass, cardioplegic arrest), increased cost effective care, and improved patient outcomes. Within our group, the idea of cardiothoracic surgical and interventional collaborative procedures was first developed over a decade ago by having to share one conventional operating room for all cardiac procedures, surgery and intervention alike, for children with congenital heart disease while a new catheterization lab was being constructed. The teams were forced into a sort of “marriage,” where each service could experience first hand the advantages and disadvantages of each discipline.



Figure 1. Hybrid Stage I palliation for HLHS in a traditional operating room with portable X-Ray imaging.



Figure 2. Same procedure performed in the Hybrid Cardiac Catheterization Suite.

HLHS and Hybrid Stage I Palliation

Collaborative cardiac procedures were initially targeted for neonates born with HLHS. Hybrid Stage I palliation was first performed in our traditional catheterization lab entirely using a percutaneous transcatheter technique to deliver a balloon-expandable PDA stent, perform balloon atrial septostomy, and for placement of intrapulmonary artery “flow restrictors” (AGA Medical, Golden Valley, MN, USA)¹ as a compassionate use device. The 6mm diameter flow restrictors were 7mm in length with two 2mm fenestrations to allow limited flow. Due to the combination of the stiff delivery cable and long sheath which stented open the right heart valves during delivery, and the acute 90 degree turn to the branch pulmonary arteries, this technique was abandoned due to the instability and hemodynamic compromise in HLHS patients. This led to transcatheter intervention involving PDA balloon expandable stent placement followed by balloon atrial septostomy (if needed) in the cardiac catheterization lab, with bilateral banding of the branch pulmonary arteries in the operating room the following day. One millimeter wide bands were cut from 3.5 or 3.0mm Gore-tex tube graft (W.L. Gore & Associates, Flagstaff, AZ, USA) to encircle the left and right branch pulmonary artery before the takeoff of the upper lobe branch. It was discovered that the previously placed PDA stent increased the technical difficulty for the surgeon applying the LPA band. Subsequently, the order of palliation was reversed and all HLHS patients underwent bilateral branch PA banding in the operative suite, followed by PDA stent and BAS in the cardiac catheterization lab. Controlling pulmonary blood flow with the PA bands placed first allowed stabilization of systemic and pulmonary flow and improved hemodynamics. Ultimately, all HLHS patients have undergone Hybrid Stage I palliation with surgical placement of bilateral PA bands followed by direct puncture and sheath placement in the main pulmonary artery for PDA stent placement (now using both self- and balloon-expandable stents) performed off cardiopulmonary bypass through a limited sternotomy incision.² This technique eliminated going through the right heart and crossing the tricuspid and pulmonary valves. A BAS was performed at a later date if needed. The palliative procedure started in the traditional operating room with portable X-Ray imaging (Figure 1), moved to specially designed Hybrid Cardiac Catheterization Suites (Figure 2), and now is equally performed in the Hybrid Cardiac Operating Suite (Figure 3). This technique has now been perfected and reproducible throughout many centers, even in high risk HLHS weighing <2.5kg.²⁻⁶ In our own center, babies as small as 1.0kg have had successful Hybrid Stage I palliation, Comprehensive Stage II repair, and Fontan completion a feat unfathomable in 1980 in Dr. Norwood’s original report (Figure 4).

Perventricular Device Closure of MVSD

A Hybrid approach for successful perventricular device closure of MVSD is dependent upon a collaborative effort between the



Figure 3. The newer Hybrid Cardiac Operative Suite is well suited for Hybrid procedures.



Figure 4. After initial Hybrid Stage I palliation at 1 Kg, and later Comprehensive Stage II repair, eventual Fontan completion was performed around 2 years of age.

cardiothoracic surgeon, the cardiac interventionalist, as well as the echocardiographer. Intraoperative perventricular device closure of a MVSD was first reported in 1998.⁷ Using this technique, cardiopulmonary bypass is not required. In a multi-center study, Bacha et al reported this technique in a large series of patients who underwent successful perventricular device closure of MVSD.⁸ This approach has the advantage of avoiding or reducing time on cardiopulmonary bypass, avoiding a ventriculotomy, as well as resection of right ventricular muscle bundles. This technique is not limited in terms of patient weight or size, as no vascular access for device delivery or crossing multiple cardiac valves with a stiff cable are required (Figure 5a,b). When combining perventricular MVSD closure in conjunction with concomitant surgical repair of other complicated CHD utilizing partial flow on cardiopulmonary bypass, “under filling” of the heart may result in under-sizing the defect and ultimately the device. Under-sizing of the device may potentially result in embolization. Therefore, we recommend perventricular MVSD closure either before cardiopulmonary bypass has been initiated or after it is terminated and while the heart is filled.

Hybrid Intra-operative Stent Therapy

The cooperative relationship between the cardiothoracic surgeon and cardiac interventionalist in the operating room environment has

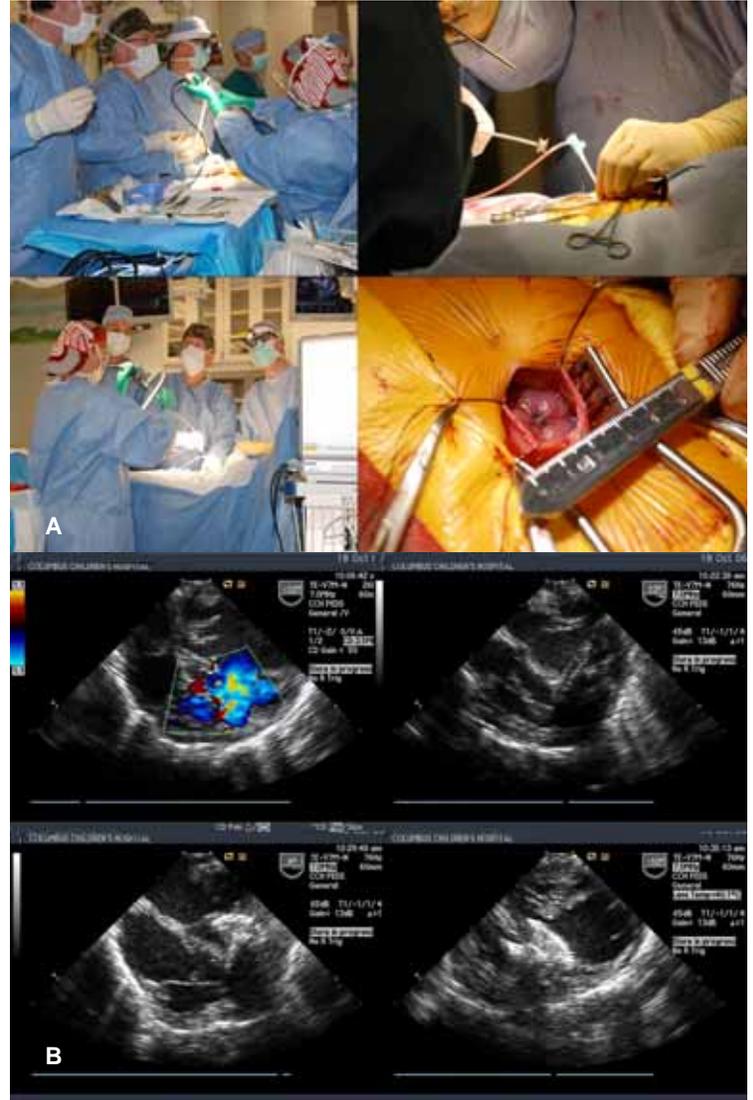


Figure 5.(a) Perventricular MVSD closure is performed in a small infant through a median sternotomy off cardiopulmonary bypass. (b) A 10mm Amplatzer MVSD Occluder is successfully deployed.

also led to Hybrid stent delivery to treat proximal and distal pulmonary artery stenoses in complex lesions such as repaired tetralogy of Fallot, repaired pulmonary atresia with VSD, repaired truncus arteriosus, and while undergoing surgical procedures such as RV-PA conduit or pulmonary valve replacement.⁹ Stents can be placed under direct visualization using endoscopic camera guidance while on cardiopulmonary bypass (Figure 6). The endoscope can be introduced into the pulmonary artery and advanced to view side branches, as well as the area of stenosis. The distance can easily be measured on the endoscope to determine the appropriate stent length necessary to treat the lesion effectively without jailing any side branching. The proximal stent struts at the origin of the MPA can easily be crimped back to allow easy access into the branches during future cardiac catheterizations. In situations where cardiopulmonary bypass is not required, direct access is provided by the surgical team through a median sternotomy, or sometimes from a transthoracic needle puncture. Fluoroscopy and angiography are used in the procedures, either using a portable C-arm or a permanent x-ray system, as found in our Hybrid Cardiac Operating Suite. The Hybrid approach to PA stent therapy is beneficial in those patients with hypoplastic PA branches and who are at increased risk for vascular injury.



Figure 6. Endoscopic camera guidance is used in the Hybrid Cardiac Operating Suite to place intraoperative pulmonary artery stents while on cardiopulmonary bypass.



Figure 8. The RadPad is very useful for the surgeon during Hybrid procedures and exit angiography in order to provide shielding from radiation.



Figure 7. Exit angiography is routinely performed after Comprehensive Stage II repair for HLHS.

Additionally, in patients without vascular access, direct access into the main pulmonary artery is possible to treat branch or distal PA stenoses. There is no need for stiff wires and long sheaths. There is no difficulty tracking over guidewires and around curves or difficult outflow tracts. Also with direct visualization, the surgeon has better control in the event of a vascular complication.

Hybrid Intraoperative Exit Angiography

Finally, in order to evaluate surgical repair or palliation of complex congenital heart disease, performing an “exit angiogram” is often helpful, especially in post-operative management or planning for future intervention. Performing an exit angiogram may identify a potential lesion which may benefit by treatment immediately in the operating suite versus just having awareness for future imaging (Figure 7). A complicated post-operative course and poor outcomes may be a result of residual structural pathology. Reports have demonstrated that patients who required cardiac catheterization and intervention during the early post-operative course while in the ICU have increased mortality compared to patients who did not require cardiac catheterization.¹⁰ Delaying cardiac catheterization post-operatively in patients with residual pathology has been identified as a significant risk factor for death.¹⁰ Although Zahn and colleagues have reported that interventional therapy can be safely performed in the immediate post-operative period,¹¹ interventional therapy via a Hybrid approach in the O.R. with cardiopulmonary bypass readily available is a safer modality as opposed to a percutaneous intervention in the cardiac catheterization suite in a hemodynamically unstable post-op patient. Exit angiography can aid in identifying residual lesions which can be treated immediately while still on cardiopulmonary bypass in the operating suite. An angiographic catheter may be introduced directly through a suture line or purse string and advanced into position. Angiograms can be performed during low flow or off cardiopulmonary bypass. Holzer et al reviewed completion angiograms performed at Nationwide Children’s Hospital in a specially designed Hybrid Cardiac Operating Suite and confirmed expected results in 47% of patients, while in 53% of patients unexpected abnormalities or residual

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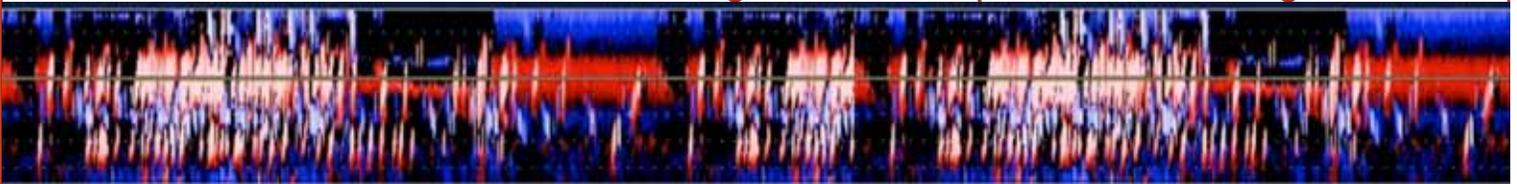




Figure 9. The new Cradle Table allows the motion necessary for the cardiothoracic surgeon and is fully integrated into the biplane FPD Hybrid Cardiac Catheterization Suite.



Figure 10. Transapical balloon pulmonary valvuloplasty is performed in this critically ill 700 gram premature baby with poor RV function, ascites, and limited vascular access.

lesions were identified.¹² As a result, 40% underwent Hybrid therapy during the surgery.

Radiation protection must be taken into consideration and available for the O.R. staff, especially for the surgeon who must remain at the table during fluoroscopy and digital acquisition. Sterile radiation protection pads are commercially available and can easily be clipped to the surgeon's sterile gown during the exit angiography procedure¹³ (Figure 8). A limiting factor for the surgeon in a Hybrid Cardiac Catheterization Suite is the integrated table and the inability to perform all of the functions of a surgical table, including cradle roll to the right or left and reverse and trendelenberg motion. New technology has helped overcome this problem using the Toshiba[®] cradle table (Figure 9). This allows tilting of the table to accommodate the surgeon's field of vision, while also allowing steep RAO/LAO and

AUGUST 2010 MEDICAL MEETING FOCUS

The Fifth International Symposium on the Hybrid Approach to Congenital Heart Disease (ISHAC)

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Overview: A two-day Symposium with a one-day skills Workshop. Attendees will learn the latest clinical and research developments in Hybrid therapies from Nationwide Children's Hospital's international faculty. Past ISHAC Symposiums have had faculty and participants from over 34 states, 50 countries, and 6 continents. ISHAC has something for surgeons, cardiologists, anesthesiologists, nurses and nurse practitioners, perfusionists, allied health professionals, and research scientists.

There will be didactic lectures, moderated discussions, live case demonstrations, special lunch presentations, tours of the Hybrid Suites at Nationwide Children's Hospital, and direct access to the faculty for Q & A. This year, procedures will be performed from Hybrid Suites at Rush Medical Center in Chicago, Miami Children's Hospital, and Nationwide Children's Hospital... with special Hybrid procedures broadcast from the University Laboratory Animal Resources Experimental Surgical Suites (ULAR).

Course Directors: John P. Cheatham, MD & Mark Galantowicz, MD

Keynote Speakers: Philipp Bonhoeffer, MD & John G. Byrne, MD

Workshop Directors: Ralf J. Holzer, MD and Alistair Phillips, MD

Program Co-Director: Sharon L. Hill, MSN, ACNP-BC

Program Coordinator: Karen E. Heiser, PhD

Perfusion Directors: Vincent Olshove, CCP; Daniel Gomez, CCP & Thomas J. Preston, CCP

ISHAC will enable participants to:

- Compare clinical outcomes between Hybrid and traditional approaches to CHD.
- Implement innovative evidence-based approaches to CHD management.
- Utilize research and multi-center outcomes data to determine care options for infants, children and adults with CHD.
- Acquire clinical skills with new Hybrid treatments and devices (Skills Workshop).

Selected List of Faculty: Valerie K. Bergdall, DVM; Philipp Bonhoeffer, MD; John G. Byrne, MD; Christopher Caldarone, MD; Mario Carminati, MD; John P. Cheatham, MD; Curt Daniels, MD; Timothy F. Feltes, MD; Mark Galantowicz, MD; Daniel Gomez, CCP; Daniel Gruenstein, MD; Ziyad M. Hijazi, MD; Sharon L. Hill, MSN, ACNP-BC; Ralf J. Holzer, MD; Shengshou Hu, MD; John P. Kovalchin, MD; Pamela A. Lucchesi, PhD; William T. Mahle, MD; Doff McElhinney, MD; Richard G. Ohye, MD; Vincent Olshove, CCP; Carlos Pedra, MD; Simone Fontes Pedra, MD; Thomas J. Preston, CCP; Alistair Phillips, MD; Mark D. Rodefeld, MD; Carlos Ruiz, MD, PhD; Dietmar Schranz, MD; Nikolay V. Vasilyev, MD; Zhen Xu, PhD; Evan Zahn, MD; David Zhao, MD

even cranial projections where normally the C-arm would come in contact with the standard surgical table.

The Future

This spirit of Hybrid collaboration has led to performing cardiac procedures that were thought impossible years ago. Unusual cases include a transapical right ventricular approach to treat critical pulmonary valve stenosis in a 700gm, 24 week gestation neonate with RV failure,



Figure 11. A 550 gram newborn with complete AV block and complex CHD presents challenges that are addressed in a Hybrid setting.

ascites, and no vascular access¹⁴ (Figure 10); a placement of a ventricular pacemaker in a 550gm neonate with complex CHD and congenital atrioventricular heart block (Figure 11); and delivery of a customized covered stent to close a traumatic communication between the right innominate artery and the trachea in a 9-year-old presenting with life threatening hemoptysis (Figure 12 a,b). Hybrid therapy has even been transferred from the human population to “man’s best friend” with equally outstanding results (Figure 13). All it takes is a spirit of collaboration, innovation, and team work. Oh yeah...and it doesn’t hurt to have specially designed Hybrid Cardiac Catheterization and Operative Suites!

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Figure 12. (a),(b) When a 9 year old presented with massive hemoptysis from erosion of a tracheostomy collar into the right innominate artery, the Hybrid team customized a covered stent to successfully close the erosion and control the bleeding.

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Figure 13. After a call from the OSU Veterinary School Cardiology Department concerning a 7 month old Cavalier King Charles Spaniel that was in heart failure with a large MVSD, the combined Hybrid teams successfully placed a perventricular 18mm Amplatzer MVSD Occluder. "Sparky" was discharged within a few days and rejoined his family.

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Abstracts from “Evolving Concepts in the Management of Complex Congenital Heart Disease II” - Part IV

“Abstracts from ‘Evolving Concepts in the Management of Complex Congenital Heart Disease II’ - Part IV” includes the following topics and presenters:

- When MRI, When CT by Beth Feller Printz, MD, PhD
- Indications for AICD, Anti-Tachycardia PM, Bi-V Pacing by Kevin Shannon, MD
- Is the Fontan Procedure Different in HLHS versus Other Single Ventricles? by Thomas L. Spray, MD

Read Parts I, II and III in the April, May and June issues of *Congenital Cardiology Today*.



Abstract Title: When MRI, When CT
Presenter: Beth Feller Printz, MD, PhD
Medical Director, Non-Invasive Cardiac Imaging, Division of Cardiology, Rady Children’s Hospital - San Diego, San Diego, CA USA

Objectives

1. Review the strengths and weaknesses of cardiac MRI (CMR) when compared with cardiac and thoracic CT for the evaluation of children and young adults with congenital and acquired heart disease.
2. Discuss relative ‘costs’ of CT vs. CMR.
3. Review potential future applications of these imaging modalities for the evaluation and treatment of congenital heart disease.

Abstract

Although echocardiography has become the mainstay of non-invasive imaging for children and young adults with congenital and acquired heart disease, there are many instances when echocardiography is insufficient to fully characterize cardiac and thoracic anatomy and physiology. Cardiac MRI (CMR) and CT have both become important imaging techniques as adjuncts not only to echocardiography, but also to cardiac catheterization and radionuclide imaging in these patients.^{1,2} For example, both CMR and CT can evaluate complex anatomic relations in any imaging plane, as they are both 3-D tomographic techniques. CMR phase-contrast flow imaging can be used to accurately measure pulmonary and systemic blood flow for shunt assessments, to determine valvar regurgitation (for example, to quantify pulmonary regurgitation following Tetralogy of Fallot repair), and to assess differential pulmonary perfusion as an alternate to radionuclide perfusion imaging. CMR can also be used to assess myocardial viability, both at rest and with stress, which may be particularly helpful in evaluating patients who have undergone surgical repair of congenital heart disease, or in those patients with ‘acquired’ cardiac pathology such as cardiomyopathy or myocarditis. Hybrid CMR technology is also being pioneered for interventional cardiac procedures, in part to decrease exposure to ionizing radiation, but also to take advantage of CMR’s strengths.³

CMR, however, can be a time-consuming procedure and requires sedation or anesthesia in most young children. At present, cardiac

MRI cannot be performed in patients with pacemakers; the presence of ferro-magnetic coils or stents can also preclude diagnostic-quality imaging. Cardiac CT can be obtained in much less time, potentially obligating the need for sedation or anesthesia; also, CT is not as sensitive to artifacts from metallic implants and can be performed in patients following pacemaker insertion. Another advantage of CT is that its spatial resolution can be superior to what can be obtained using many commercially-available CMR platforms, leading to improved coronary artery imaging. Cardiac CT may also be a less ‘expensive’ test vis a vis monetary charges billed. These advantages of cardiac CT are tempered by the risk of radiation exposure, particularly in young children who are more sensitive to the effects of radiation or in those subjects where serial scans are required.^{4,5} Recent advances in multi-detector CT technology can significantly decrease radiation exposure and make cardiac CT a feasible alternative to CMR in select cases.

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Abstract Title: Indications for AICD, Anti-Tachycardia PM, Bi-V Pacing
Presenter: Kevin Shannon, MD; Director, Pediatric Electrophysiology Program; Mattel Children’s Hospital, Division of Pediatric Cardiology; Los Angeles, CA, USA

Objective

- Discuss published guidelines and controversies in device therapy for congenital heart disease.
- Review indications for ICD placement in patients with congenital heart disease.
- Review efficacy and complication data in patients with congenital heart disease who receive ICD’s.
- Review available data on safety and efficacy of antitachycardia pacing in congenital heart disease.
- Review indications for biventricular pacing.

- Review safety and efficacy data on biventricular pacing in patients with congenital heart disease.

Abstract

Implantable Cardioverter-Defibrillator therapy in congenital heart disease.

Data from studies in adult patients with ischemic cardiomyopathy and idiopathic dilated cardiomyopathy have consistently shown a survival benefit in patients undergoing ICD implantation, if they met symptom and ejection fraction criteria. These findings have led to the development of the following criteria for implantation of ICD in this patient population:

- ICD therapy is indicated in patients with a LVEF <35% due to prior myocardial infarction who are 40 days post-myocardial infarction and in New York Heart Association (NYHA) functional class II or III.
- ICD therapy is indicated in patients with a LVEF <35% due to nonischemic dilated cardiomyopathy who are in NYHA functional class II or III.
- ICD therapy is indicated in patients with a LVEF <30% due to prior myocardial infarction who are 40 days post-myocardial infarction and in NYHA functional class I.

The annual sudden death-rate in the patient populations studied were 6-11 %. Sudden death rates in patients with congenital heart disease and ventricular dysfunction are substantially lower, and thus, the efficacy of ICD placement may not be as high. The incidence of inappropriate shocks is as high as 20% in most studies of patients with congenital heart disease, with a similar incidence of complications resulting in the need for re-operation. These factors suggest that the criteria for ICD implantation in younger patients with congenital heart disease should be more stringent than the current guidelines for adult patients with ischemic or idiopathic cardiomyopathy.

Anti-tachycardiac pacing.

Atrial antitachycardia pacing has limited availability, and is of limited utility. Cardiac catheter ablation has become the treatment of choice for most patients with a single tachycardia circuit and uncomplicated atrial tachycardia. Patients with complex heart disease tend to have multiple atrial tachycardia substrates, and as a consequence, have limited efficacy with antitachycardia pacing. Patient activated therapies and antitachycardia pacing on an ICD platforms may improve the utility of this therapy.

Biventricular pacing

Several retrospective studies have shown that biventricular or multisite pacing can improve cardiac function and alleviate heart failure symptoms in patients with congenital heart disease, including patients with univentricular heart disease and systemic morphologic right ventricles. The patients most likely to benefit appear to be those with a deterioration in cardiac function related to single site pacing. Criteria for identifying likely responders and or non-responders need to be improved, as do techniques for identifying and accessing ideal pacing sites.

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CCT



Abstract Title: Is the Fontan Procedure Different in HLHS versus Other Single Ventricles?

Presenter: Thomas L. Spray, MD; Chief, Division of Cardiothoracic Surgery; Children's Hospital of Philadelphia, Philadelphia, PA USA

Objective

To analyze the results with Fontan surgery for single ventricle malformations and the potential long-term effects of a right ventricle as the systemic ventricle.

Abstract

As larger numbers of patients are undergoing the Fontan operation for variants of Hypoplastic Left Heart Syndrome (HLHS) with a single right ventricle as the systemic ventricular mass, there is increasing interest in the longevity of the right ventricle as the systemic ventricle in the Fontan circulation. Most infants undergo a series of staged reconstruction operations beginning in the neonatal period and culminating at an eventual Fontan procedure. The goals of neonatal management include creation of unobstructed systemic outflow and pulmonary venous pathways, limitation of pulmonary blood flow and avoidance of pulmonary artery distortion. A variety of technical modifications have been introduced over the years, including staging with an interim superior cavopulmonary connection, use of an intra-atrial baffle to create a lateral tunnel Fontan, creation of an intentional right-to-left shunt (fenestration), and introduction of the extracardiac conduit Fontan. Because of improved management in the neonatal period in early infancy an increasing number of patients previously thought to be at high risk, especially those with HLHS, are now being referred for the Fontan operation. Despite this very dramatic change in patient population there has been a dramatic reduction in mortality and morbidity after the Fontan operation.

Although reports from some centers have suggested that Hypoplastic Left Heart Syndrome is a risk factor for survival after the Fontan procedure, our own experience at The Children's Hospital of Philadelphia has not been able to identify the presence of a systemic right ventricle as a significant risk factor for Fontan

survival. In a series of 332 Fontan patients from January 1, 1992 to December 31, 1999, including a large percentage of patients with HLHS, systemic ventricular morphology had no effect on overall outcome. Similar results have been reported from the group at The University of Michigan where anatomy also was not a significant factor in overall survival. At interim staging with the bidirectional cavopulmonary anastomosis there has been no obvious influence of ventricular anatomy on survival, although A-V valve regurgitation seems to be an independent risk factor for death or transplantation. Whether A-V valve regurgitation is associated with a single right ventricle versus a left ventricle was not independently evaluated. Even when performed at elevated altitude where pulmonary resistance might effect overall survival, no influence of ventricular morphology on survival was identified in a series from Denver Children's Hospital. Ventricular morphology also did not turn out to be a significant predictor of exercise performance in the first two decades of life after the Fontan operation.

Evaluation of neurodevelopmental outcome after the Fontan operation also has not consistently shown a difference between right and left ventricular morphology or that patients with HLHS drome was an independent risk factor for low achievement scores, however, other studies from Michigan have not identified any difference in patients with HLHS as have the studies at The Children's Hospital of Philadelphia.

In summary, a review of the reported series of Fontan operations in patients with single ventricle anatomy have not been able to identify ventricular anatomy as an independent risk factor for outcome. However, the increasing number of patients with HLHS now coming to the Fontan operation with now longer survival times may ultimately show a decrease in longevity in patients with a systemic right ventricle as has been seen in patients with the atrial switch operation for transposition. It is conceivable however that the late problems with the Fontan circulation are less related to ventricular morphology than to primary problems with the pulmonary vascular bed or diastolic function of the ventricles rather than purely ventricular morphologic anatomy. An increasing attention to methods for treating late Fontan failure will be necessary to deal with these patients as they age.

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Read Part V - "Abstracts from 'Evolving Concepts in the Management of Complex Congenital Heart Disease II' in the August issue.



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Medical News, Products and Information

Edwards Lifesciences Announces Receipt of CE Mark for Pulmonic Transcatheter Valve

IRVINE, CA--(Marketwire - May 26, 2010) - Edwards Lifesciences Corporation, a global leader in the science of heart valves and hemodynamic monitoring, announced on May 26, 2010, the receipt of CE Mark for the Edwards SAPIEN pulmonic transcatheter heart valve. The valve is designed to be an alternative to surgical valve replacement for patients suffering from congenital heart disease of the pulmonic valve and leverages the company's transcatheter valve technology, which has been used in more than 5,000 cases around the world.

"The Edwards SAPIEN pulmonic transcatheter heart valve gives clinicians the potential to eliminate one of the multitude of open-chest procedures that their young congenital heart patients will face in their lifetimes," said Ziyad M. Hijazi, MD, MPH, Director of the Rush Center for Congenital and Structural Heart Disease, Chief of Pediatric Cardiology and Professor in the Departments of Pediatrics and Internal Medicine at Rush University, Chicago. "As every re-operation increases a patient's risk of infection, illness and death, it is a gift to have a minimally invasive way to treat these fragile patients."

"We are proud to apply our extensive experience in heart valve innovation and leadership in transcatheter technology to provide a high-quality solution for patients with so many unmet needs," said Larry L. Wood, Edwards' Corporate VP, Transcatheter Valve Replacement.

The Edwards SAPIEN pulmonic transcatheter valve's leaflet design is modeled after Edwards' clinically proven aortic tissue valves and its stainless steel frame provides high radial strength. Prior to delivery, the bovine pericardial tissue valve is compressed onto a balloon to the approximate diameter of a pencil. It is then threaded through the patient's circulatory system using the RetroFlex 3 transfemoral delivery system, which enables accurate deployment of the valve across the patient's pulmonary valve.

In the US, this valve is an investigational device currently being studied in the COMPASSION (COngenital Multicenter trial of Pulmonic vAlve regurgitation Studying the SAPIEN InterventIOnal THV) clinical trial, which will assess the safety of the valve. The company completed its feasibility study of the valve and is progressing into the pivotal portion of the clinical trial with the goal of receiving a Humanitarian Device Exemption from the US Food and Drug Administration.

Dr. Hijazi is the principal investigator of the COMPASSION clinical trial and a consultant to Edwards for education and research.

Additional company information can be found at www.edwards.com.

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Scientists Discover Key Step for Regulating Embryonic Development

New on-off switches: SUMO protein silences developmental genes, SENP2 snips SUMO to allow gene expression

HOUSTON – Deleting a gene in mouse embryos caused cardiac defects and early death, leading researchers to identify a mechanism that turns developmental genes off and on as an embryo matures, a team led by a scientist at The University of Texas M. D. Anderson Cancer Center reported today in *Molecular Cell*.

"Our study focused on regulation of two genes that are critical to the healthy development of the heart, but many other genes are regulated in this way," said senior author Edward T.H. Yeh, MD, Professor and Chair of M. D. Anderson's Department of Cardiology. "This novel pathway marks an advance in our understanding of how developmental genes are turned on and off."

All cells in an embryo contain the same DNA. Different genes are turned off and on in different cells at different times to form specific tissues and organs as the embryo develops. This gene regulation is accomplished by epigenetic processes that control gene expression without altering DNA. Instead, epigenetic processes attach chemical groups to genes or to histones, proteins that are intertwined with DNA to form chromosomes, to activate genes or to shut them down.

"Our findings provide a new window through which to look at epigenetic control," Yeh said, "and how epigenetics and development are unexpectedly tied together by the SUMO/SENP2 system."

The key actors are members of two tightly associated families of proteins that Yeh and colleagues discovered and continue to study. The first, Small Ubiquitin-related Modifier, or SUMO, attaches to other proteins to modify their function or physically move them within the cell (SUMOylation). The second, Sentrin/SUMO-specific protease 2, or SENP2, snips SUMO off of proteins (de-SUMOylation).

This line of research started when Yeh and colleagues knocked SENP2 out of mouse DNA and found that the embryos died at about day 10. Their hearts had smaller chambers and thinner walls. Through a series of experiments, the team worked backward from this observation to show:

- A group of proteins called the polycomb repressive complex 1 (PRC1) that silences genes must first bind to a particular methylated address on a histone and,
- A key component of the complex must be SUMOylated to make this connection, which results in
- the silencing of Gata4 and Gata6, genes that are essential for cardiac development.
- In early development, SENP2 works as a switch to turn on Gata4 and Gata6
- "When SENP2 is turned on, it peels SUMO off of PRC1, which then falls off the histone, and when that happens, the lock is removed and

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genes are transcribed," Yeh said. Gata4 and Gata6 are free to properly develop the heart.

In short, SUMO helps the PRC1 complex repress genes, and SENP2 reverses this repression, allowing gene transcription and expression.

"By understanding how development unfolds, we can better control this process, which includes cell proliferation and organ development," Yeh said. "This will help us to better understand cancer.

"SUMO and SENP are important in cancer development, neurological diseases and heart development. Everything under the sun can be regulated by this system," Yeh said. "Here we've established a new role for SUMOylation, mediating the interaction between protein and protein methylation in epigenetic regulation."

Funding for this research was provided by from the National Natural Science Foundation of China, National Basic Research Program of China and grants from the U.S. National Cancer Institute. Yeh also is the McNair Scholar of the Texas Heart Institute / St. Luke's Episcopal Hospital.

Co-authors with Yeh are co-first author Yitao Qi, PhD, and Robert Schwartz, PhD, both of the Texas Heart Institute / St. Luke's Episcopal Hospital, and co-first author Xunlei Kang, MD, PhD, Yong Zuo, Ph.D., Qi Wang, Yanqiong Zou and Jinke Cheng, DVM, all of the Key Laboratory of Cell Differentiation and Apoptosis of the Chinese Ministry of Education, Shanghai Jiao Tong University School of Medicine in Shanghai.

Newer Heart Surgery for Infants Offers First-Year Survival Benefit Over Traditional Procedure

Infants born with a severely underdeveloped heart who undergo a newer surgical procedure are more likely to survive their first year and not require a heart transplant than those who have a more traditional surgical procedure, according to a report by researchers supported by the National Heart, Lung, and Blood Institute (NHLBI), which is part of the National Institutes of Health. The study of 549 newborns, however, suggests that after the first year, the two surgical procedures for the relatively rare condition yield similar results.

The Single Ventricle Reconstruction (SVR) Trial is the largest clinical trial to compare treatments for congenital heart disease, and the first North American, multi-center, randomized trial of surgical therapy for congenital heart disease patients. Results were published in the May 27, 2010, issue of the *New England Journal of Medicine*. An editorial accompanies the article.

Congenital heart disease is the most common birth defect. Every year, about 1% of babies are born with abnormally formed hearts. The normal heart has two pumping chambers called ventricles. The right ventricle pumps blood to the lungs, and the left ventricle pumps blood to the body. This trial studied babies born with a severe form of congenital heart disease in which babies are born with a functioning right ventricle and a small, underdeveloped, nonfunctioning left ventricle. The condition is sometimes referred to as Hypoplastic Left Heart Syndrome. Without treatment, these babies usually die shortly after birth.

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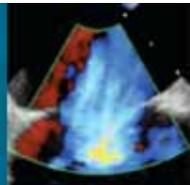
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The SVR Trial compared for the first time two surgical procedures that are commonly used to treat babies born with only the functioning right ventricle to determine whether one procedure improves outcomes more than the other. The SVR Trial was conducted at 15 North American clinical sites that are part of the NHLBI's Pediatric Heart Network.

"Rigorous comparison of patient outcomes is critical to fully understanding the risks and benefits of different interventions," said NHLBI Acting Director Susan B. Shurin, MD, a board-certified pediatrician. "To conduct meaningful clinical research on rare conditions, however, we need collaboration among multiple study sites. This study demonstrates that through consortia such as the NHLBI's Pediatric Heart Network, we can accelerate our ability to provide needed evidence on the best ways to care for some of our most vulnerable patients."

In general, three surgeries are needed to treat a single right ventricle. The first procedure, called the Norwood procedure, is usually performed within the first two weeks of life and is one of the highest risk procedures in congenital heart surgery. A shunt, or small tube, is implanted to provide a connection for blood to flow from the heart to the blood vessels in the lungs, or pulmonary arteries, so that blood can pick up oxygen and release carbon dioxide. Children later undergo a second surgery at 4 to 6 months of age, and a third procedure, known as the Fontan procedure, at 18 to 36 months. The operations are staged to allow the child to grow large enough that the corrective procedures can be performed. Heart transplantation may be required for children with single ventricles when surgery and other treatments fail.

In the SVR trial, participants were randomly assigned shortly after birth to receive one of two types of shunts for their initial surgery, as part of the Norwood procedure. About half of the newborn participants received a modified Blalock-Taussig (MBT) shunt, the traditional approach, which places the shunt from a branch off of the aorta, the major blood vessel that takes blood from the heart to the rest of the body, to the pulmonary artery. The other participants received a newer type of shunt, called an RVPA shunt, which is placed between the right ventricle and the pulmonary arteries.

Each type of shunt has advantages and disadvantages. A few small studies of the RVPA shunt have suggested that it provides better survival and other outcomes than the MBT shunt. However, randomized clinical trials have not been conducted to demonstrate whether one procedure is better than the other.

Researchers followed all study patients for at least 14 months. They evaluated the

number of deaths and heart transplantations in each group at one year, as well as the number of complications linked to each type of shunt.

"We found that the right ventricle-to-pulmonary artery shunt improved the chances of being alive without a heart transplant one year after surgery. However, the benefit appears to be limited to the first 12 months, as the two shunts showed similar results after about two years," said Richard G. Ohye, MD, Head, Pediatric Cardiovascular Surgery Division, University of Michigan, Ann Arbor, and lead author of the study.

"We are continuing to follow these children, and this longer follow-up will be important to determine which shunt is truly superior in the long run," Ohye added. A follow-up study is analyzing the effects of the procedures in children 2 to 6 years of age.

The researchers report that after 12 months, 74% of infants with the RVPA shunt survived and didn't need a heart transplant, compared to 64% of infants with the MBT shunt. The newborns with the RVPA shunt, however, had significantly more complications requiring additional interventions, for example, insertions of stents or balloons to keep the shunt open. Other outcomes, such as the size and pumping ability of the right ventricle at 14 months, were similar in the two groups of participants.

In a preliminary analysis of participants after an average follow up of 32 months, researchers found similar outcomes in the group of participants with the RVPA shunt compared to the MBT shunt.

"This study shows great promise for helping not only this high-risk group of patients, but also for improving the health and well being of many more babies and children with heart problems," noted Gail D. Pearson, MD, ScD, a pediatric cardiologist, Director of the NHLBI Adult and Pediatric Cardiac Research Program, and a coauthor of the paper.

The Pediatric Heart Network (PHN) (www.PediatricHeartNetwork.org) is a multi-center clinical research consortium supported by the NHLBI since 2001 to study congenital and acquired heart diseases that occur in childhood. The PHN also provides training in clinical research to young investigators, and is the clinical cornerstone of the NHLBI's comprehensive Bench to Bassinet program in translational pediatric cardiovascular research.

Further information about this trial (NCT00115934) can be found at: www.clinicaltrials.gov.

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