CONGENITAL CARDIOLOGY TODAY

Timely News and Information for BC/BE Congenital/Structural Cardiologists and Surgeons

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International Edition

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(See website for more meetings)

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Sep. 18-21, 2015; Las Vegas, NV USA
www.picsymposium.com

2015 HeartLink Gala Presented by Land O'Lakes
Sep. 19. 2015; Minneapolis, MN USA
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The Third Circulation: A New Paradigm in Congenital Heart Disease

By Maria Serratto, MD; Bruno Cortis, MD

Abstract

Recently the interaction between the lymphatic system and the cardiovascular system in pre-natal and post-natal life has come to the fore. To evaluate the role of the lymphatic system in isthmic aortic coarctation (Ao Coarc), we studied by lymphography five adults with this condition by bilateral lower extremity injection of contrast. Opacification of the lymphatic system was monitored cineangiographically for 48 hours. The angiograms showed lymphatic engorgement from the pelvic and lumbar-aortic lymphatics up to the drainage into the lymphatic duct. The picture of lymphatic congestion appears to result from the abnormal circulatory pattern of Ao Coarc. MRIs of severe Ao Coarc have shown a reversal of the normal flow pattern in the descending aorta with increased total blood flow. The lymphatic vessels engorgement and collateralization appears to be a compensatory phenomenon resulting from the increased total blood flow in the descending aorta. The lymphatic transport system seems very efficient since lymphatic circulation time was only minimally increased and venous pressure was normal.

"Recently the interaction between the lymphatic system and the cardiovascular system in pre-natal and post-natal life has come to the fore. To evaluate the role of the lymphatic system in isthmic aortic coarctation (Ao Coarc), we studied by lymphography five adults with this condition by bilateral lower extremity injection of contrast."

Introduction

The lymphatic system plays a critical role in physiology and pathology. Lymphatics regulate body fluids homeostasis, participate in immune responses, and transport dietary liquids. The interplay between the lymphatic system and the cardiovascular system has become the object of recent studies, which have identified significant interactions between the two systems.[1] To evaluate the role of the lymphatic system in isthmic Ao Coarc, we studied 5 adults with this condition using

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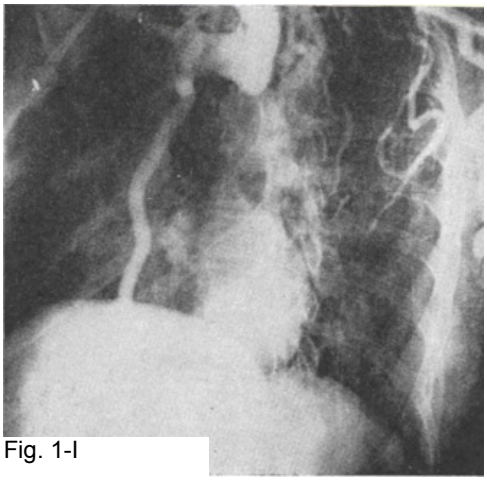


Fig. 1-I

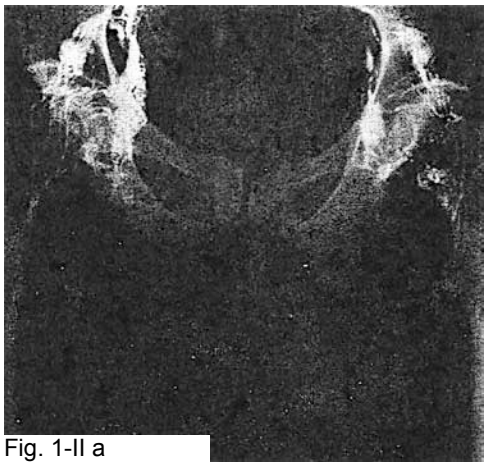


Fig. 1-II a

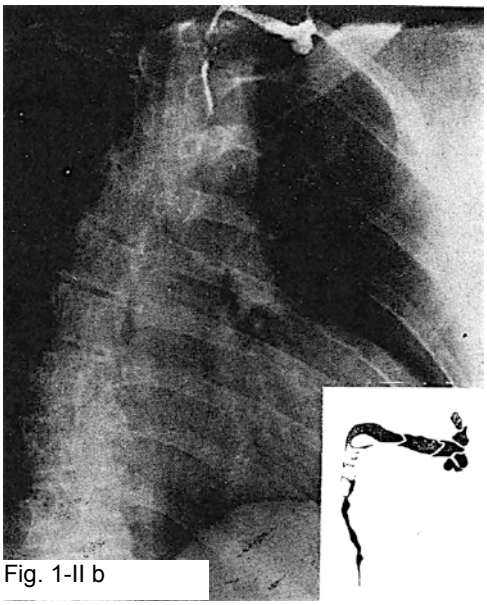


Fig. 1-II b

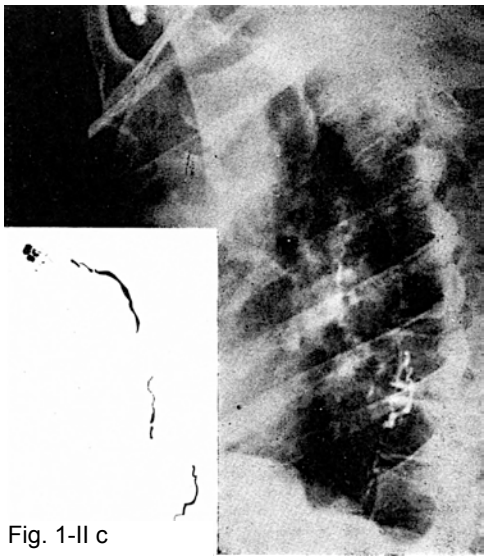


Fig. 1-II c

Case 1: Figures 1-I: Aortogramm; Figure 1-II a (Case 1: AP view: inguinal-iliac and lumbar-aortic lymphatics show some degree of ectasia more pronounced on the right and evidence of collateral circulation medially); Figure 1-II b, 1-II c (Case 1: Sagittal and LAO views: The thoracic duct is tortuous. Opacification of intercostal lymphatics with flow inversion. The terminal ampulla is dilated with valvular tension).

lymphography with bilateral lower extremity contrast injection.

Methods

Patient Population: Three of the five patients were male and two female; the patients ranged in age from 16 to 39 years old. Four of them had isolated Ao Coarc and in one the Ao Coarc was associated with Patent Ductus Arteriosus (PDA). The upper extremity systolic blood pressure ranged from 180 to 220 mmHg and the diastolic from 110 to 140 mmHg.

Laboratory Data: The electrocardiogram showed Left Ventricular Hypertrophy in four patients and Combined Ventricular Hypertrophy in the patient with associated Patent Ductus Arteriosus. The aortogram showed severe isthmic Ao Coarc in all five patients with dilatation of the aortic arch and left subclavian artery (Figures: 1-I; 2-I; 3-I;4-I; 5-I. Lymphography was carried in both

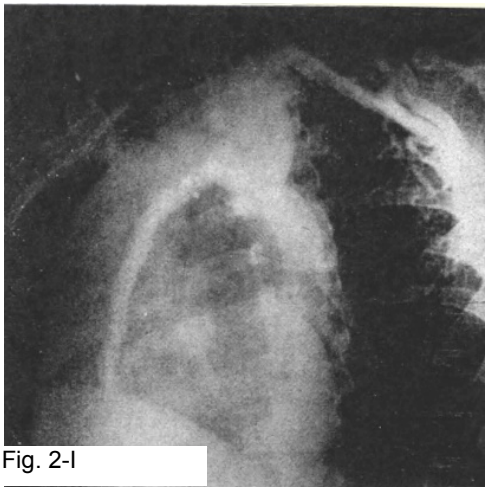
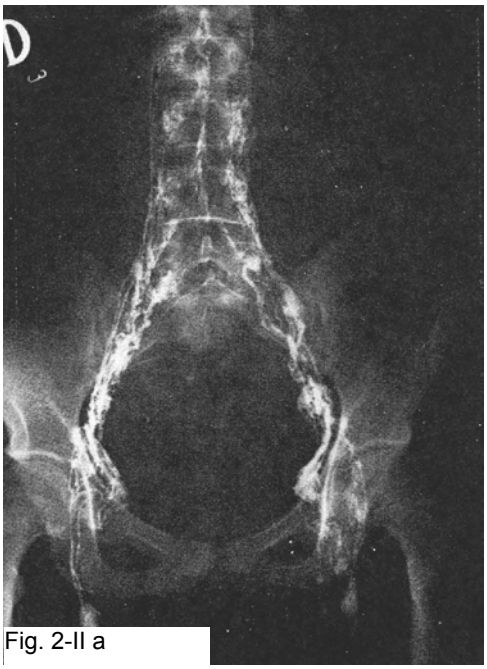


Fig. 2-I



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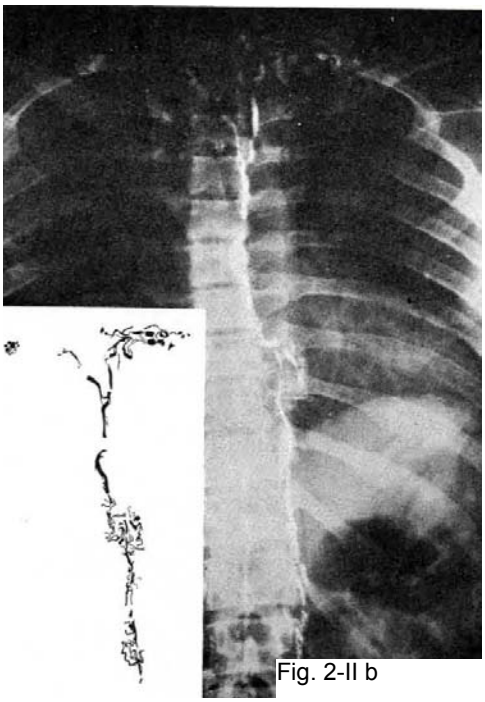

Fig. 2-II b

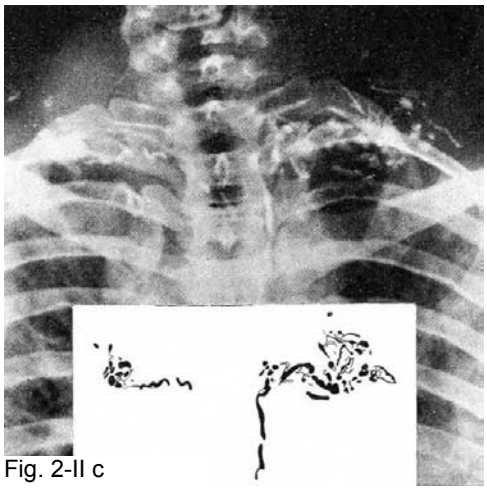

Fig. 2-II c

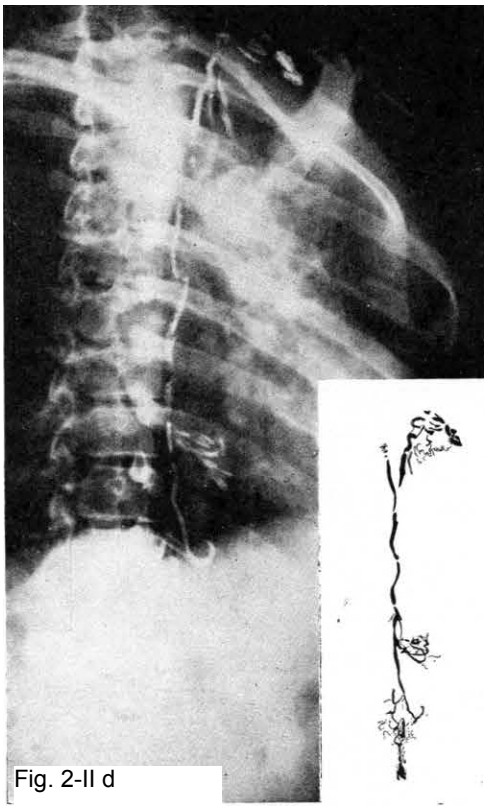

Fig. 2-II d

Case 2: Figure 2-I: Aortogram; Figure 2-II a: AP view: Normal appearance of inguinal-iliac and lumbar-aortic lymphatics) 2-II b (Case 2: Sagittal view: The thoracic duct is tortuous and originates from multiple slender channels. Opacification of intercoastal lymphatics indicates retrograde flow), 2-II c (Case 2: Sagittal view: The thoracic duct ends in separate channels on the right and on the left), 2-II d (Case 2: RAO view: Tortuosity of the thoracic duct ending in two separate channels. Opacification of collaterals).

lower extremities using the Kimmoth-Wiljasalo technique. During the injection the progression of the contrast was followed on a monitor screen. Subsequently, the opacification of the lymphatic system was monitored radiographically and recorded by cine-angiography up to 48 hours after injection.

Results

The angiograms revealed an overall picture of lymphatic plethora demonstrated by pelvic and lumbar-aortic lymphatics presenting engorgement in four of the five patients with ectasia near the lymphonodal drainage and evidence of collateral circulation. The thoracic duct showed dilation, prominent valvular apparatus, tortuousity, duplication, hyperperistalsis and dilation of the terminal ampulla and of the cysterna chili when present. The drainage of the thoracic duct often showed numerous small vessels in a reticular pattern. Opacification of collateral intercostal vessels due to flow inversion was also

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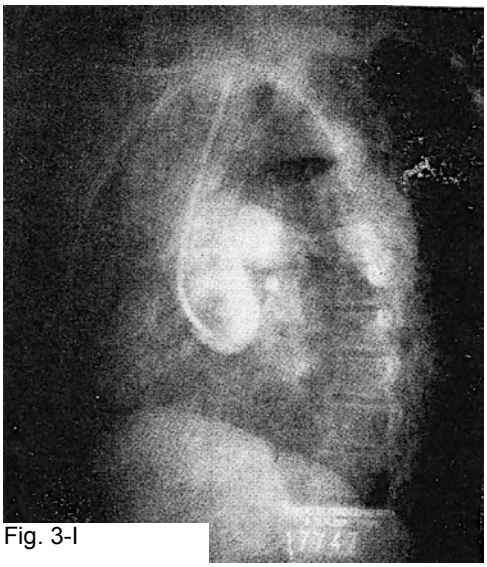

Fig. 3-I

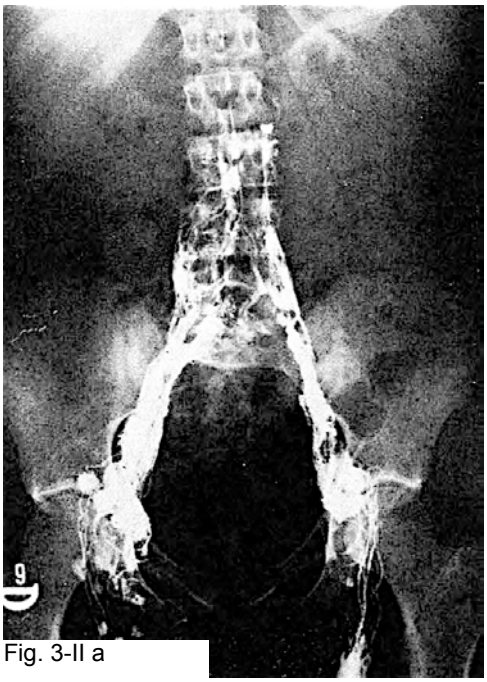

Fig. 3-II a

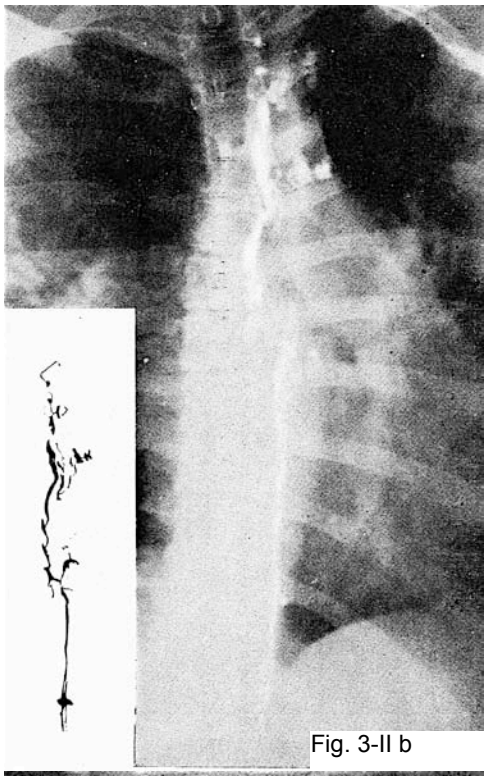

Fig. 3-II b

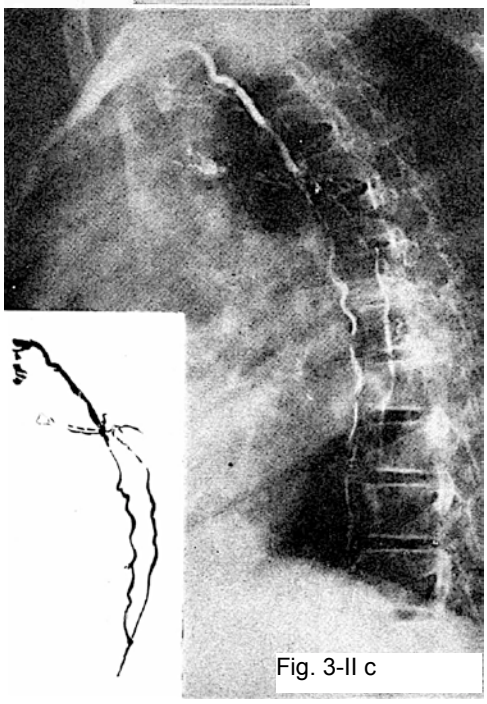

Fig. 3-II c

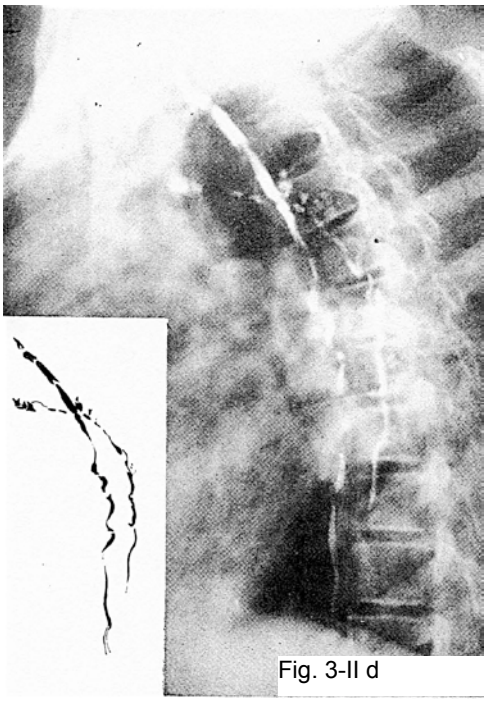

Fig. 3-II d

Case 3:Figure 3-I: Aortogram; Figure 3-II a (AP view: Ectasia of the inguinal-iliac and lumbar-aortic lymphatics), 3-III b (Case 3: AP view: Tortous thoracic duct with opacification of intercoastal lymphatics), 3-II c and 3-III d (Case 3: LOA view: Duplication of the thoracic duct. Opacification of intercoastal linfatics)

present (Figures: 1-II c; 2-II b; 3-II b, c, d; 5-II c, d).

Discussion

This pattern of lymphatic fluid congestion appears to result from the abnormal circulatory pattern of Ao Coarc. MRI studies of severe Ao Coarc demonstrated a reversal of the normal flow pattern with an increase in total blood flow in the descending thoracic aorta.[2]

"Thus, the lymphatic engorgement appears to be a phenomenon resulting from the increased total blood flow in the descending thoracic aorta in severe Ao Coarc with consequent increased production of lymphatic fluid as a compensatory mechanism."

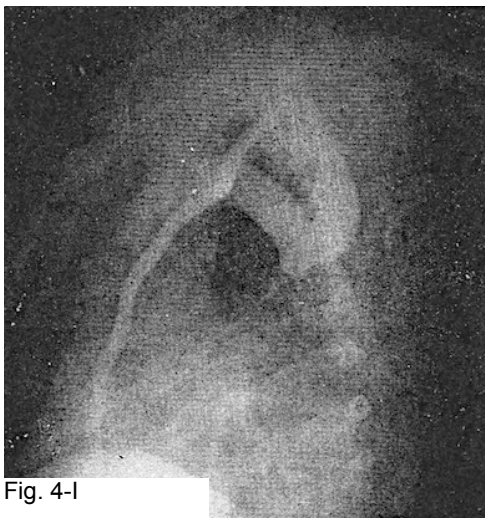

Fig. 4-I

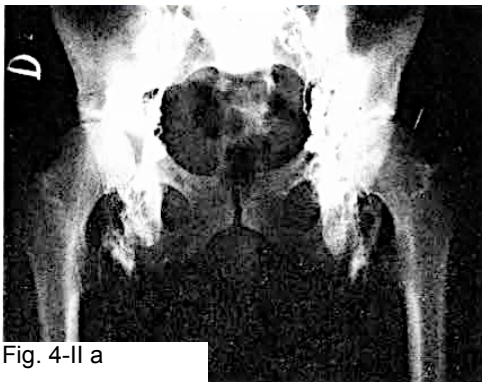

Fig. 4-II a

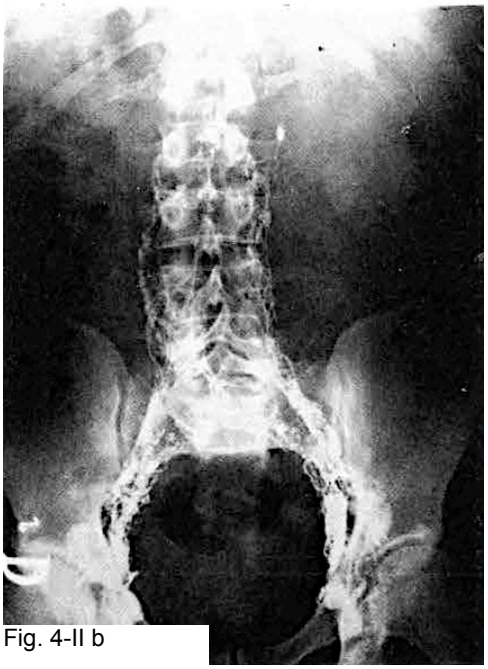

Fig. 4-II b

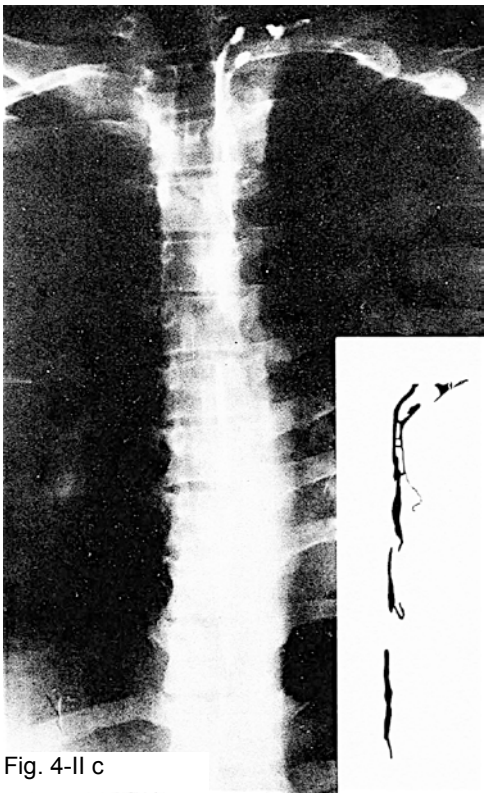

Fig. 4-II c

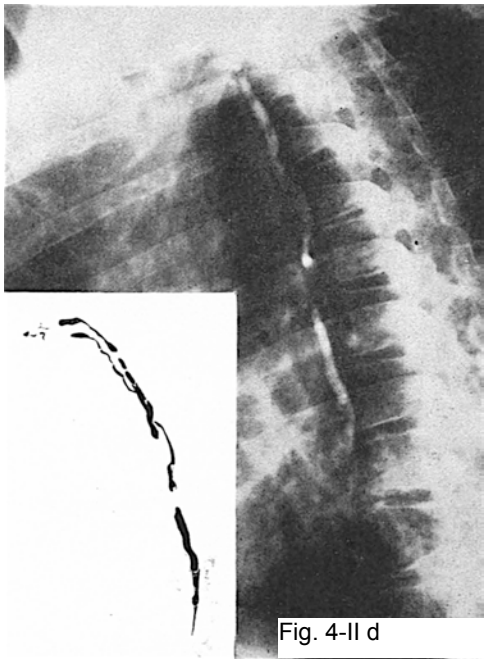

Fig. 4-II d

"In conclusion, this study shows that lymphography can demonstrate the presence of collateral flow in tight Ao Coarc, and serves as a useful method for providing a clear demonstration of the severity of coarctation of the aorta."

Case 4: Figure 4-I: Aortogram; Figure 4-II a and Figure 4-II b (AP view: Inguinal lymphatics show a reticular pattern mostly on the left. Large channel in the gluteal region); Figure 4-II c and Figure 4-II d (Case 4 - AP & LAO views: The thoracic duct shows increased peristalsis. The duct ends in two separate channels which appear dilated in the terminal portion).

Thus, the lymphatic engorgement appears to be a phenomenon resulting from the increased total blood flow in the descending thoracic aorta in severe Ao Coarc with consequent increased production of lymphatic fluid as a compensatory mechanism. The transport system appears to be very efficient since lymphatic circulation time was only minimally increased. The pattern of engorgement could not result from increased venous pressure since measurements of this parameter gave normal results.

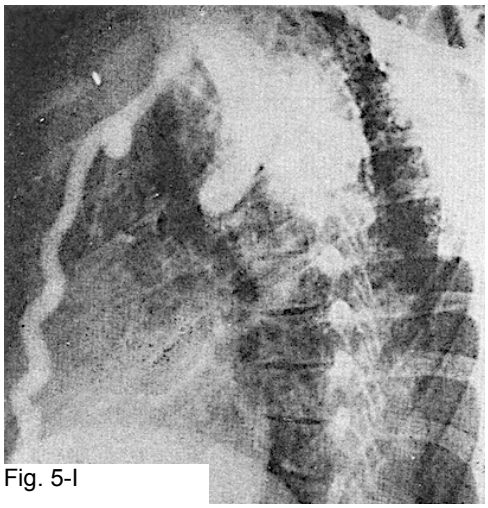

Fig. 5-I

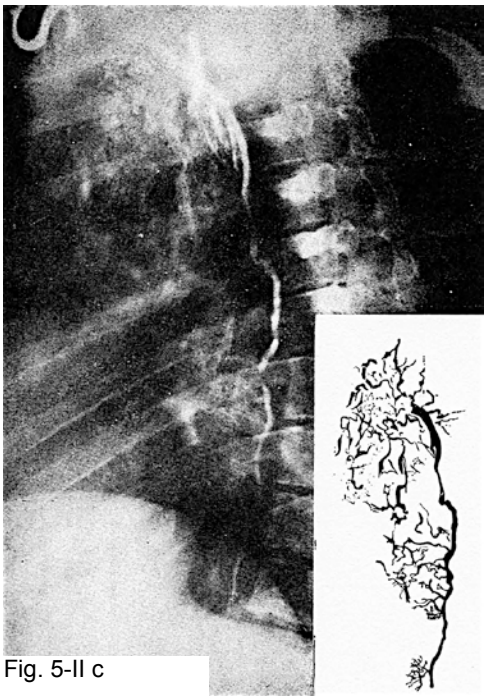

Fig. 5-II c

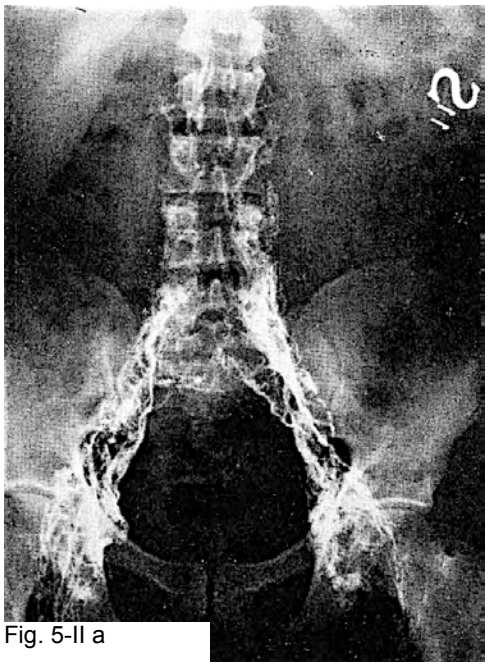

Fig. 5-II a

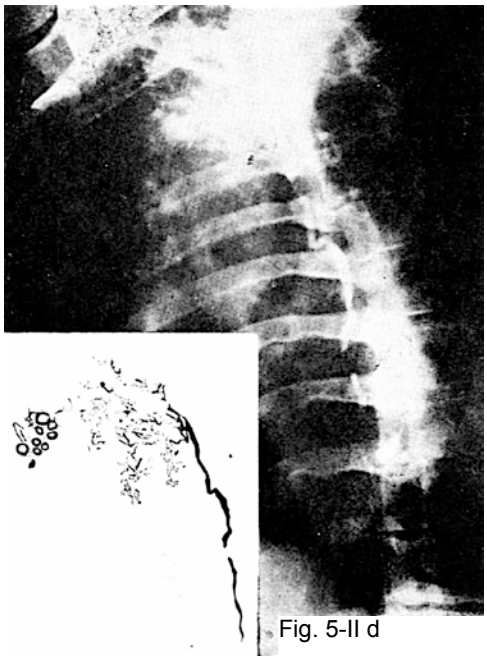

Fig. 5-II d

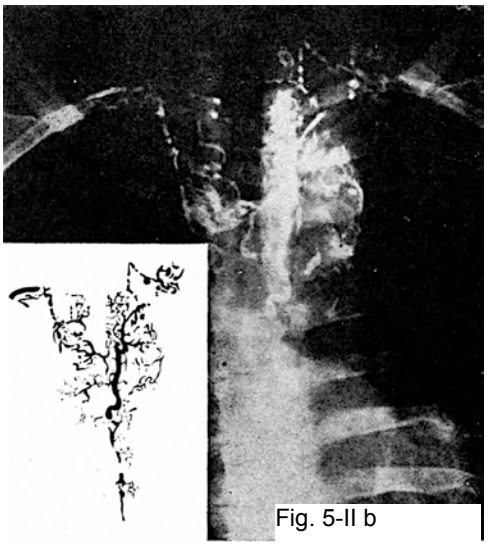

Fig. 5-II b

Case 5: Figure 5-I: Aortogram; Figure 5-II a (Case 5: AP view: inguinal-illiac and lumbar-aortic lymphatic show a net of small lymphatic vessels connecting the mid-line); Figure 5-II b, Figure 5-II c and Figure 5-II d (Case 5: Sagittal and LAO views: Tortuous thoracic duct with ectatic segments. Numerous small collateral vessels are opacified. The thoracic duct ends in multiple channels.)

In conclusion, this study shows that lymphography can demonstrate the presence of collateral flow in tight Ao Coarc and serves as a useful method for providing a clear demonstration of the severity of coarctation of the aorta.

Note: Dr. Bruno Cortis performed the lymphograms in this study.

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CHOP Cardiology 2015: A Review from the Desert

By Mitchell Cohen, MD, FACC, FHRS

In the days following the excitement of the Super Bowl in the Valley of the Sun, the Hyatt Gainey Resort in Scottsdale, Arizona hosted the *18th Annual Update on Pediatric and Congenital Cardiovascular Disease.* The Children's Hospital of Philadelphia has consistently taken the lead on providing what has become the most comprehensive annual multidisciplinary cardiology meeting in the country. The meeting provides extensive didactic and interactive sessions with cardiologists, cardiac surgeons, nurses, intensivists, anesthesiologists, and perfusionists. The focus of this year's meeting was on: "Challenges and Dilemmas." The practice of Pediatric Cardiology and Congenital Cardiovascular Medicine spans a wide spectrum of care from the fetus to the adult with Congenital Heart Disease. With so many conditions manifesting at various stages of life, many challenges and dilemmas exist in our understanding of disease and treatment. It was with this task in mind that the 71 faculty members provided, not just an update on the various conditions, but often, new paradigms of thought regarding management.

Over 800 attendees participated in this 4-½ day meeting with nearly equal representation by physicians and nursing. There were over 100 participants from outside the United States, with 18 alone coming from China. The first day of the meeting there were three optional pre-conferences to choose from:

- "The Fetus with Heart Disease"
- "Mechanical Circulatory Support in Pediatric and Congenital Heart Disease," and
- "A Primer of Neonatal CHD for Nurses and Frontline Providers"

The meeting officially kicked-off Wednesday afternoon and introduced some of the challenges and dilemmas that face the pediatric cardiology community. Dr. Betsy Goldmuntz provided an update on the genomic revolution and the vast understanding of single nucleotide polymorphisms and copy-number variations that exist within CHD. However, the future is likely going to move towards using whole exome sequencing and challenges will arise as we bring the complexity of genomic analysis into phenotype and outcomes measurements. Dr. Jack Rychik, the Course Director, discussed the challenges fetal echocardiographers face moving forward. As a paradigm of thought, Dr. Rychik discussed the challenges of fetal intervention to alter the physiology and modify outcomes in the first real "9 months" leading up to delivery. Dr. Thomas Spray gave an insightful talk on the limitations of databases and registries and how

Figure 1. Chair of the Program Organizing Committee, Dr. Jack Rychik of CHOP, led off the conference.

Figure 2. New this year was a 5K Heart Run!

Figure 3. Panelists for the Plenary Session on "The Borderline Left Ventricle" included from left-to-right Dr. Paul Weinberg (CHOP), Dr. Charles Fraser (Texas Children's), Dr. Meryl Cohen (CHOP), Dr. James Tweddell (Milwaukee Children's), and Drs. Sitaram Emani and Wayne Tworetzky (Boston Children's).

innovation for the surgeon will likely not arise from new surgical techniques, but through modifying risks. Databases need to move away from strictly looking at mortality and should focus on longitudinal outcomes over the life of the child rather than a simple 30-day window. Dr. Andrew Redington gave a provocative talk on how disruptive behavior can lead to inspirational change and improvements in CHD outcomes will arise from "shooting for the moon" and not accepting average. Wednesday evening ended with emotionally paired-talks. Meghan Roswick, a young adult with Hypoplastic Left Heart Syndrome, presented her inspirational journey through Fontan and beyond. Her surgeon, Dr. Marshall Jacobs, touched on his role as a surgeon and as a student constantly learning and adapting from his patients.

The 2nd day of the meeting focused on heart failure in the young. Dr. Joe Rosanno presented the current epidemiology of heart failure in children and the tremendous personal and financial dollars needed to care for these children. The session continued with discussions on the medical management, utilization of novel biomarkers, mechanical circulatory support, and most importantly, the role of the nurse in the care of children with ongoing heart

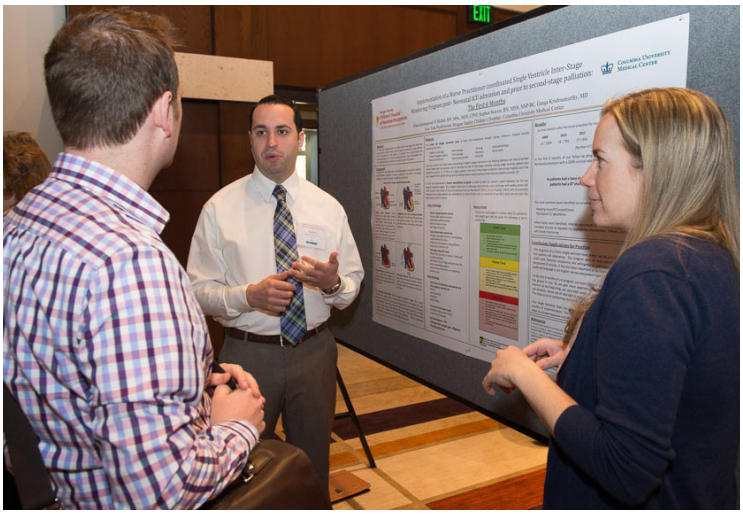

Figure 4. Over 150 abstracts as posters and oral presentations were offered.

Figure 5. Moderated panel of experts on controversial topics. Left to right: Dr. David Goldberg (CHOP), Dr. Girish Shirali (Mercy Children's, Kansas City) and Dr. Jack Rome (CHOP).

failure. As traditionally occurs at the CHOP meeting, a number of breakout sessions took place following the plenary. These breakout sessions, allowed for a more focused discussion on challenges within the sub-disciplines of Interventional Cardiology, Electrophysiology, Perfusion, etc. One of the more interesting sessions, led by Drs. Stephen Paridon and Julie Brothers featured a team debate between Drs. Chris Calderone and Welton Gersony versus Drs. Marshall Jacobs and Silvana Molossi regarding controversies surrounding clinical decision making in children with anomalous right coronary arteries. How do we best manage an asymptomatic 15 year-old with an anomalous right coronary? While "no official winner" was declared, the challenges that care providers are faced with will require more advanced and novel imaging strategies as well as ongoing registry data collection. The afternoon session began with a Moderated Panel of Experts, led by Dr. Jack Rome, regarding the timing of pulmonary valve replacement in Tetralogy of Fallot. This casual "laid-back" session had cardiologists, surgeons, and catheterization physicians from around the country provide insight into the challenges and the "unknowns" that prompt decision making into the timing of pulmonary valve replacement. The afternoon concluded with a session on the: Challenges and Dilemmas in Caring for the Patient with Single Ventricle. Dr. Rick Ohye reviewed the outcomes of the SVR Trial and provided a scorecard comparing Norwood-to-Hybrid-to-Sanno and that advances and differences between surgeries will likely depend on pushing centers of excellence. Dr. Shirali introduced a novel app that families of single ventricle patients could use to help facilitate care, interact with care providers, and hopefully improve outcomes between Stage I and II palliation. Dr. Yoav Dori presented some fascinating imaging modalities of the lymphatic system and novel aims to preserve central lymphatic flow with early rerouting in patients with plastic bronchitis.

The 3rd day of the meeting began with a breakfast session specifically for nurses and advanced nurse practitioners in Pharmacology. The main plenary session of the morning addressed the challenges we face in identifying borderline left ventricles that are destined to a single ventricle preparation versus those hearts that have a truly identifiable two-ventricle repair. One of the most identifying features which may help the clinician deciding between which infants are likely to have a single ventricle or a two-ventricle repair is the presence or absence of endocardial fibroelastois. Dr. Wayne Tworetzky discussed the evolution of aortic stenosis to HLHS in-utero and the challenges in selecting fetuses with borderline left ventricles that might benefit from a fetal interventional strategy. One of the challenges we face as a pediatric cardiology community, especially in the borderline left ventricle, is the limited statistically-significant randomized data that facilitate our ability to make probable decisions as opposed to anecdotal possible decisions.

Following the morning plenary, a number of breakout sessions addressed the Challenges and Dilemmas in: "Non-invasive Imaging", and Tetralogy of Fallot with Pulmonary Atresia." One of the unique breakout sessions included involvement of healthcare administrators and the challenges that children's hospitals face related to cost, delivery of care through dedicated service lines, and the future role of standardized care pathways. The ability to have healthcare administrators present, not just for their particular breakout sessions, but involved in all of the sessions, is unique, and adds an excellent opportunity to hear a different perspective. The afternoon plenary session discussed the problems of neurocognitve impairment in repaired or palliated CHD. The session highlighted what we have learned about identifying children already with neurocognitve impairment and those at risk and how we can modify certain outcomes by interacting with neurologists, psychologists, and parents.

The 4th day celebrated Valentine's Day and started with over 100 participants enjoying the sunrise with a 5K Heart Run. The historical Saturday morning line-up at the CHOP meeting revolves around key invited lectures. William Brawn MD, cardiothoracic surgeon at UK Birmingham, gave the 15th Annual C. Walton Lillehi Memorial Lecture in Cardiovascular Surgery on the double switch procedure for patients with congenital corrected transposition of the great vessels. The second invited speaker was Dr. George Lister,

Figures 6 and 7. Over 800 attendees and faculty participated in the educational and stimulating 5 day conference.

"Next year's meeting will be held February 22nd-26th, 2016 at the Loews Royal Pacific Resort at Universal Studios in Orlando. We hope to see you there!"

Physician-in-Chief at Yale New Haven Children's Hospital, who gave the 7th Annual Jack Downes Lecture in Cardiac Anesthesia and Critical Care. Dr Lister retrospectively looked at his lengthy career and the lessons learned from his patients, the real teachers. Deepak Srivastava, MD, gave one of the most "cutting edge" talks on the use of the induced pluripotent stem cell and its potential future as a rejuvenated cardiomyocte in pediatric heart failure and CHD. Such technology is exciting and inspirational, but will no doubt elicit additional challenges and dilemmas to the basic scientist and, potentially, the clinical physician in the future. The final lecture, the 15th Annual T. Garrett Rauch Nursing Lecture, was given by Nancy Rudd, RN, MS, APNP, on how home monitoring for children with single ventricle all began. Nancy presented not just the success of interstage monitoring, but more importantly, how a single idea empowered nurses to initiate, implement, and complete a project. The afternoon plenary session dealt with the ethical and social issues that abound as a pediatric cardiology community. The team of experts discussed how to deal with social media, define quality of life, define the boundaries of care, and how to deal with the ethical challenges that face all providers in a busy cardiac ICU. Saturday evening provided a respite from the didactic lectures as Jeopardy Host, Matthew Gillespie, MD, entertained the audience as varying programs across the country competed in Quiz Bowl. The team from Phoenix Children's Hospital, the local host sponsor, won the annual Quiz Bowl. Congratulations to all participants.

The meeting concluded Sunday morning in a very well-attended session on the challenges and dilemmas in adult congenital heart disease. As Adrienne Kovacs, PhD discussed, the start of any great adult congenital heart program begins during the transitional adolescent time-period. Curt Daniels, MD followed with a discussion on building the ideal ACHD program. Dr. Daniels acknowledged that there is no single way to build an ACHD program, but rather the focus needs to be on building a program for the patients not for the hospital's own self-interest. Phillip Moons PhD, RN, presented interesting data from Europe on varying parental styles and how they might impact health care and perception of illness in the adolescent with CHD. Linda Tobin MSN, CRNP, discussed the ever-expanding role of the nurse practitioner in an ACHD program. Dr. Gruschen Veldtman presented new information on the evaluation of liver disease and vascular reserve in the ACHD Fontan patient. Dr. Daniels followed up with a talk on the challenges posed by the 4%-6% of ACHD patients with pulmonary hypertension and right ventricular failure. Dr. Maully Shah presented the recently published ACHD Arrhythmia Guidelines (Heart Rhythm. 2014 Oct;11(10):e102-65.) Yuli Kim, MD, who discussed the challenges faced by managing pregnancy and contraception in our ACHD population, delivered the final talk. Pregnancy creates an atypical physiologic milieu with alterations in cardiac output and systemic vascular resistance. Managing these patients during the pregnancy and in the post-partum period can be challenging and requires a true team effort with involvement from ACHD specialists and maternal-fetal-medicine colleagues. Dr. Rychik and the organizing team should be credited for putting together a spectacular meeting that was equally enhanced by 70°, perfect weather. Next year's meeting will be held February 22nd-26th, 2016 at the Loews Royal Pacific Resort at Universal Studios in Orlando. We hope to see you there!

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Mustard Transcatheter Pulmonary Venous Baffle Intervention – A Pictorial Essay

Abhay Divekar, MBBS, MD

Introduction

Patients with Systemic (SVB) and Pulmonary Venous Baffle (PVB) Stenosis after the atrial switch procedure (Mustard or Senning) for Transposition of the Great Arteries (D-TGA), or as a component of the double switch procedure for Congenitally Corrected Transposition of the Great Arteries (cc-TGA) continue to present for transcatheter interventions in the present era.[1-3]

Stenosis of the PVB is a well-described complication after the atrial switch procedure. PVB stenosis can occur early or late at an incidence of 1-10%.[4] PVB stenosis has traditionally been addressed by surgery and is associated with high mortality.[5] Multiple approaches for transcatheter relief of PVB stenosis, including prograde (trans-baffle), retrograde (transarterial), as well as a hybrid approach, have all been suggested.[2, 5-10]

In this report we utilize a physical patient-specific 3D model to create a pictorial essay and discuss the interventional options and challenges of various transcatheter approaches for PVB stenosis and review the published literature.

Case Report

A 35-year-old man was born with D-TGA and palliated with balloon atrial septostomy and then underwent Mustard repair as an infant. Postoperatively he developed severe SVC baffle stenosis which was treated surgically by using a Dacron patch. He presented at age 35 years with palpitations and lower extremity edema. His symptoms were treated with furosemide and metoprolol. Echocardiography showed mild-to-moderately depressed systemic ventricular function, and caused suspicion for systemic and pulmonary venous baffle stenosis. A gated 3D Computed Tomography (CT) was performed to plan transcatheter intervention. The CT showed systemic ventricular EF of 35%, severely stenotic SVC baffle, and moderate-to-severe PVB stenosis (Figure 1). Based on the CT, a retrograde approach was deemed to be suitable for PVB intervention (use Figure 2, 3, 4 to follow along with text). Diagnostic right and left heart catheterization showed a 3 mmHg mean gradient across the SVC baffle and a 10-12 mmHg mean gradient across the PVB. There was no shunt by oximetry and the pulmonary vascular resistance (PVR) was elevated. The systemic ventricular end-diastolic pressure was 9 mmHg. An Amplatz super-stiff wire was placed from the RIJ across the SVC baffle into the IVC. A pigtail catheter was advanced retrograde into the systemic

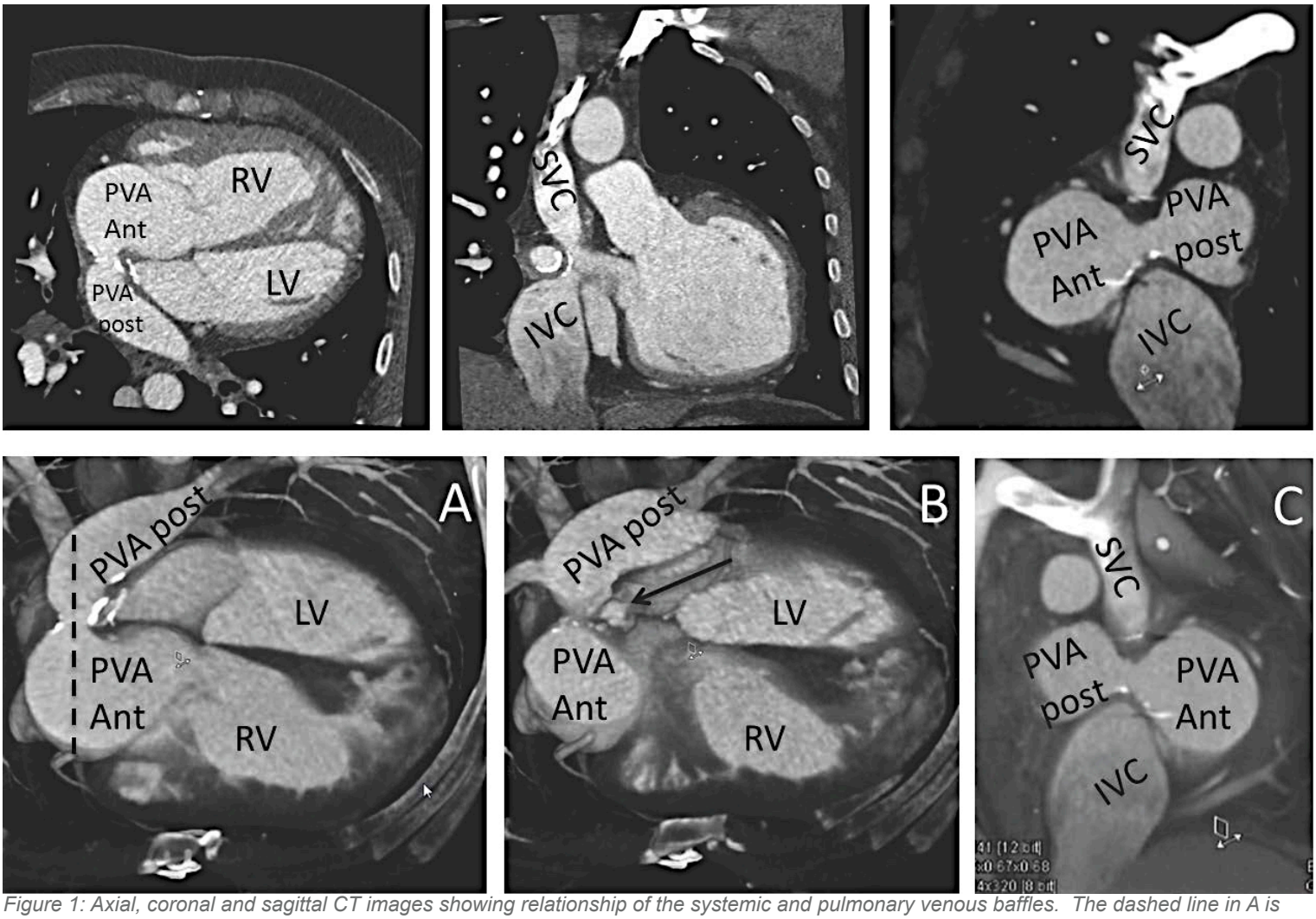

Figure 1: Axial, coronal and sagittal CT images showing relationship of the systemic and pulmonary venous baffles. The dashed line in A is the plane of reference of the sagittal image in C. Images A, B, C represent 3D reconstructed axial (A, B) and sagittal (C) views and the arrow in B points to the close relationship between the SVC (arrow) and the pulmonary venous baffle.

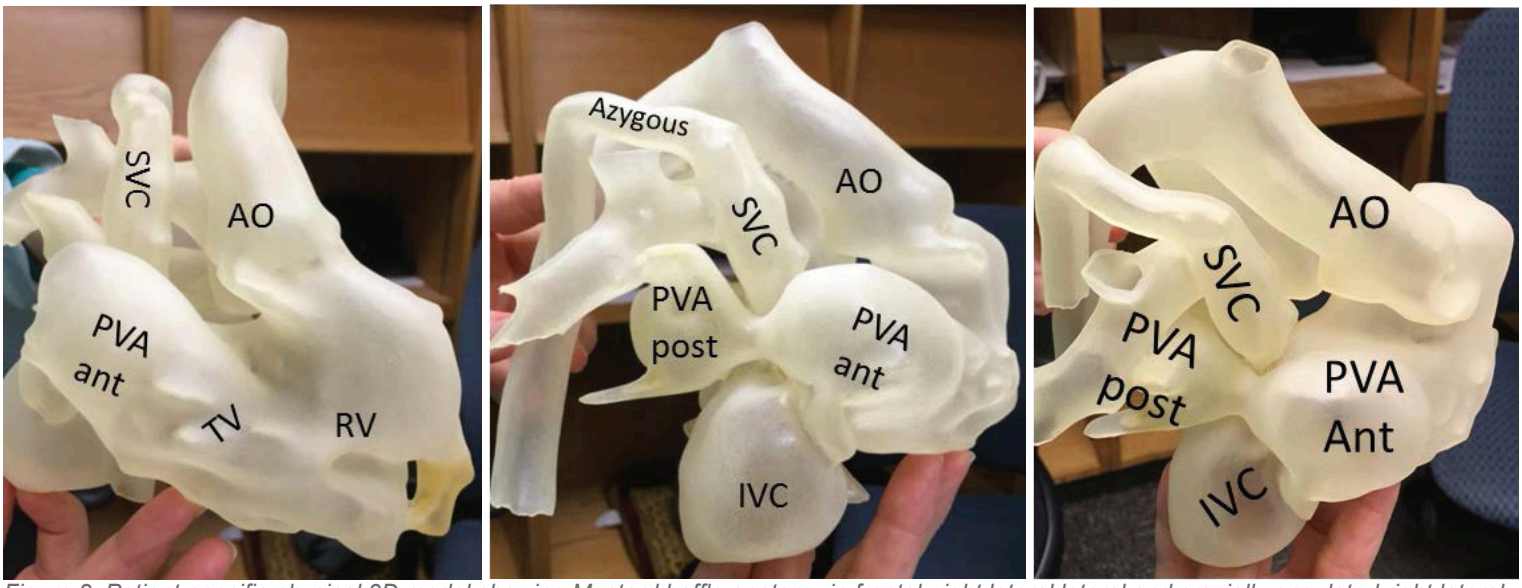

Figure 2. Patient-specific physical 3D model showing Mustard baffle anatomy in frontal, right lateral lateral and cranially angulated right lateral projection.

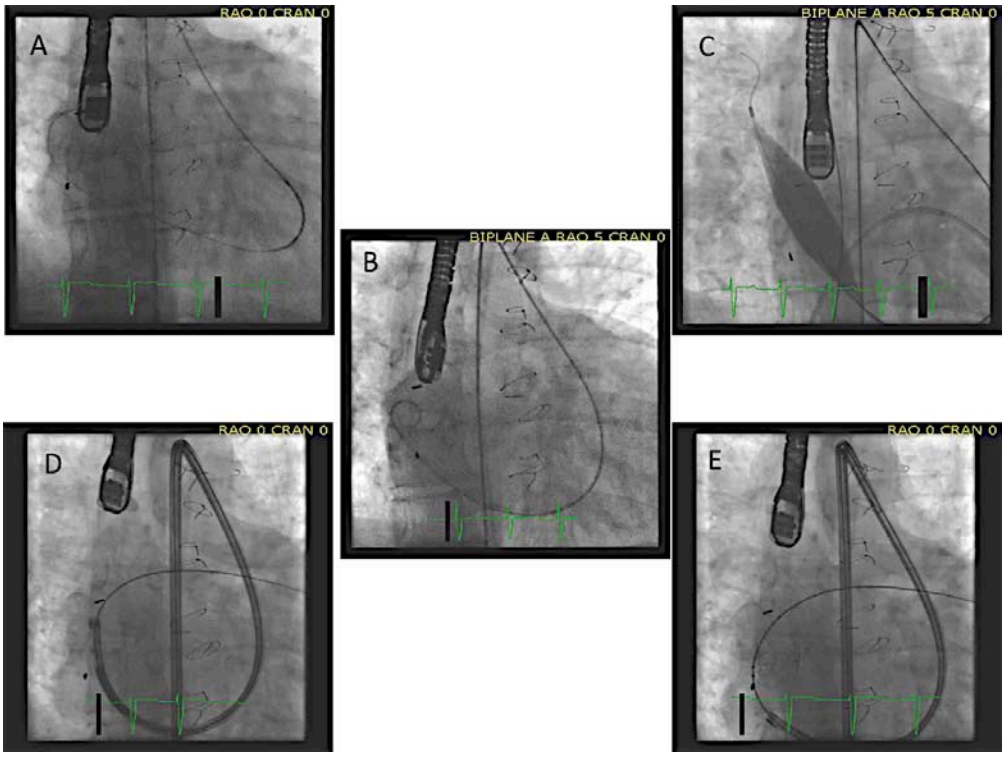

Figure 3. Fluoroscopic images (frontal projection) showing retrograde access to the PVA, (A) guidewire directed by opening pigtail, (B) pigtail in PVA, (C) balloon angioplasty with wire anchored in RUPV, (D) sheath positioned in the posterior compartment of the PVA, (E) stent deployed to effectively treat PVB stenosis.

right ventricle. In the frontal projection the pigtail was pointed towards the right-sided atrioventricular (AV) valve. A 0.035" Glidewire (Terumo) was use to "open" the pigtail and then directed across the right-sided AV valve into the pulmonary venous atrium (PVA) with minimal difficulty. The pigtail catheter was then advanced over the wire into the anterior compartment of the (towards the right AVV) PVA. The pigtail catheter was then exchanged over a 260cm 0.035" J guidewire for a 5 Fr glide catheter. A glide wire was then used to cross the stenotic PVB stenosis into the right upper pulmonary vein. This catheter was then exchanged over a 0.035" Amplatz super-stiff guide wire. There was no sustained atrial or ventricular ectopy and there was no hypotension related to a wire across the systemic semilunar and AV valves.

Balloon angioplasty of the systemic and pulmonary venous baffles was performed sequentially under transesophageal echocardiographic (TEE) guidance monitoring if there was any compromise of the adjacent baffles during intervention on the other. The stenotic SVC baffle was sequentially dilated with a 16 and then a 20 mm high pressure angioplasty catheter at 10 and 14 atmospheres. There was no encroachment into the PVB. Post intervention angiography showed excellent results without any residual gradient. We then turned our attention to the PVB. Again, under TEE guidance, the PVB was sequentially dilated with a 12, 16, 18, 20 mm high pressure angioplasty catheter at 14 atmospheres.

There was no encroachment into the systemic venous baffle. The narrowest dimension by TEE increased from 7 -14 mm and there was a decrease in the mean echocardiographic gradient from 12 to 4 mmHg. Stenting of both baffles was considered, but the need for systemic anticoagulation after PVB stenting and an adequate result prompted us to consider reevaluation. Total fluoroscopy time was 35.45 min and radiation dose was 1245 mGy. Transthoracic echocardiogram the next day showed improvement, but the mean gradient across the PVB increased to 6 mmHg. As his PVR was already increased, and since heart transplant is the only option for progressive symptomatic systemic ventricular dysfunction and the reported high recurrence with angioplasty alone, we decided to reevaluate the patient in 6 weeks and consider stent implantation if necessary. At repeat catheterization 6 weeks later the PVR was still elevated. There was sustained hemodynamic and angiographic relief of SVC baffle stenosis. However, there was still a 6 mmHg mean gradient across the PVB. Since previous retrograde intervention was easily performed, we chose the same approach. The arterial access in the right femoral artery was pre-closed with two Perclose sutures. A 12-Fr long Cook introducer was advanced into the posterior compartment of the PVA (use Figure 3 to follow along with text).

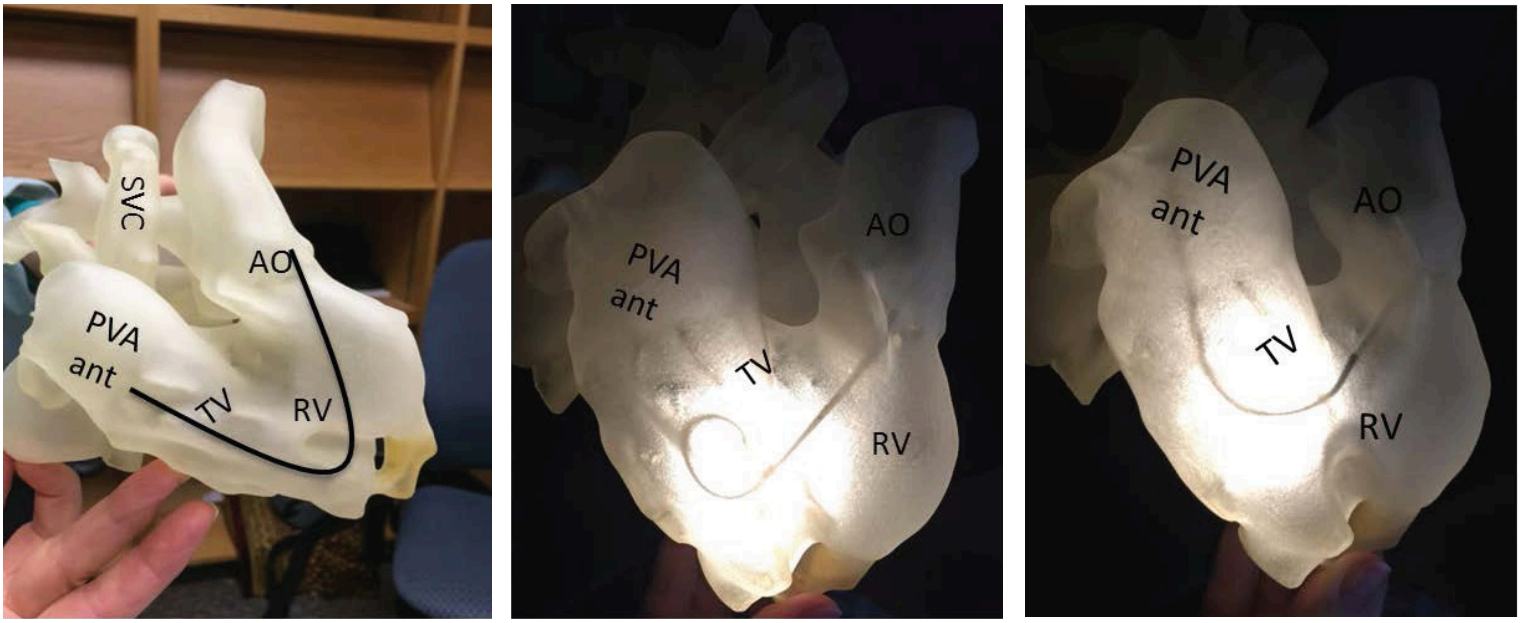

Figure 4. Patient-specific 3D model showing interventional strategy using the tripartite nature of the right ventricle to the interventionalist's advantage.

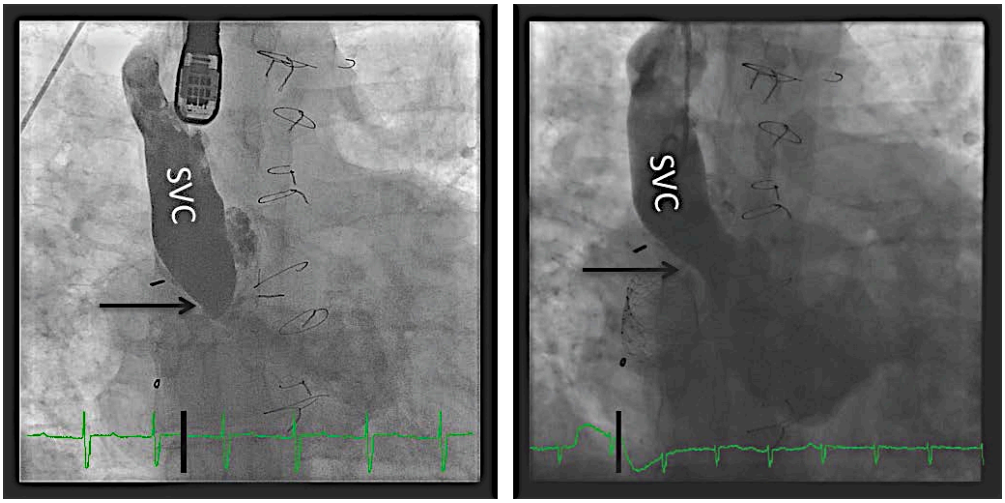

Figure 5. Frontal angiographic image showing severe SVC baffle stenosis (arrow) and successful relief at 2-year follow-up with angioplasty alone.

Initially, with the dilator in the sheath, was just above the AV valve. As the dilator was removed and tension on the wire adjusted, the sheath advanced just beyond the stenosis into the posterior compartment.

There was no sustained ectopy or hemodynamic compromise from the wire or the long sheath across the semilunar and systemic AV valve. A 26 mm EV3 LD Max stent was mounted on a 20 mm BIB catheter. To avoid difficulty in advancing the stent related to sheath kinking, the sheath was pulled back across the AV valve into the ventricle once the mounted stent was across the aortic valve. The balloon was then advanced to the tip of the sheath, and the entire unit was advanced as a unit using the balloon as the dilator. The stent was then centered across the stenosis, unsheathed and then deployed by sequential inflation of the inner and outer balloons. The stent was post-dilated with a 22 mm high-pressure angioplasty catheter. Post-intervention there was a 1 mmHg mean gradient and excellent relief of stenosis. Total fluoroscopy time was 19.43 min and radiation dose was 236 mGy. The SVC baffle remains patent without recurrence of stenosis before and after PVB stent placement (Figure 5). At the most recent follow-up (2 years), there has been sustained hemodynamic relief and decrease in PVR; the results of cardiac catheterization are summarized in Table 1.

Discussion

The baffles that redirect systemic and pulmonary venous flow after the atrial switch repair have a complex 3D arrangement. The advent of 3D imaging and 3D printing of physical models greatly increases our ability to understand these complex pathways and plan interventional procedures.

To understand the 3D relationship of the baffles, one can consider an analogy to a pair of trousers (use Figure 1 and 2 to follow along with text). The limbs are connected to the SVC and IVC and the waist to the left AV valve (mitral). The PVA is formed by two components. The pulmonary veins enter into a posterior compartment (native left atrium) which then wraps around the "crotch" of the trouser to enter the anterior compartment (native right atrium), and then across the right-sided AV valve (tricuspid) into the systemic right ventricle. This anatomy is relevant for and dictates several interventional techniques. The 3D model nicely illustrates why, during transbaffle puncture, the needle is pointed anterior and to the right rather than the posterior and to the left, as is typical for standard transseptal puncture. As a result, one usually enters into the anterior compartment of the PVA. The confluence of the PVB and SVB tends to be the site for PVB stenosis. This close relationship between the SVB and PVB underscores the potential for encroachment and iatrogenic narrowing of one of the baffles when the other baffle is treated with stenting. Therefore, especially during SVC baffle stenting, one must ensure that there is no compromise of the PVB during balloon angioplasty prior to stent implantation.

Although surgical approach was initially preferred, it was associated with high morbidity and mortality and there was a growing trend towards transcatheter interventions.[5,9] However, recurrence or insufficient relief of PVB stenosis after

Table 1. Serial Data from Cardiac Catheterizations Before and After Balloon Angioplasty, Stent Implantation and at 2-Year Follow-up						
Timeline	SBP mmHg	PAP mmHg	PCWP mmHg	PVRi Wood Units indexed	CI l/min/m2	Gradient
Baseline	100/62,75	60/32,45	21	8.05	2.5	12
Post-balloon 6 weeks	100/61,76	47/19,33	16	5.9	2.8	6
Post-stent 6 months	86/45,60	33/10,20	11	3.21	2.8	1
Post-stent 2 years	86/50,61	34/12,21	12	3.19	2.8	1
BP = systemic blood pressure (S/D, mean), PAP = pulmonary artery pressure (S/D, mean), PCWP = Pulmonary capillary wedge pressure (mean), PVRi = indexed pulmonary vascular resistance, CI = cardiac index.						

balloon angioplasty is high.[9] Inability to achieve a sufficient balloon to stenosis ratio has been suggested to be a major factor and stent implantation is suggested to provide long-term relief of stenosis.[6,9] Three approaches have been proposed and used for transcatheter treatment of PVB stenosis: namely, transvenous (transbaffle), transarterial (retrograde) and hybrid.[1, 2, 9]

Most reports during the earlier era favored the retrograde transarterial approach both for balloon angioplasty, but also for stent implantation.[5-9] The 3D model allows insight into the potential merits of the retrograde transarterial approach (Figures 2, 3, 4). The tripartite RV anatomy, using a series of catheter exchanges, aids in creating a gentle curve for transcatheter intervention. Once the pigtail is positioned in the RV, it is rotated so that the catheter is en-face in the frontal projection and the pigtail opens to the patient's right. Advancing an angled Glidewire opens the pigtail and naturally directs the wire to the orifice of the right AV valve. The pigtail catheter can then be advanced into the anterior compartment of the PVA. It may be difficult to advance the catheter any further, and exchanging it for an end-hole catheter allow further manipulation of a guidewire into the posterior compartment, and then into the pulmonary veins to anchor the interventional exchange length guidewire. The potential

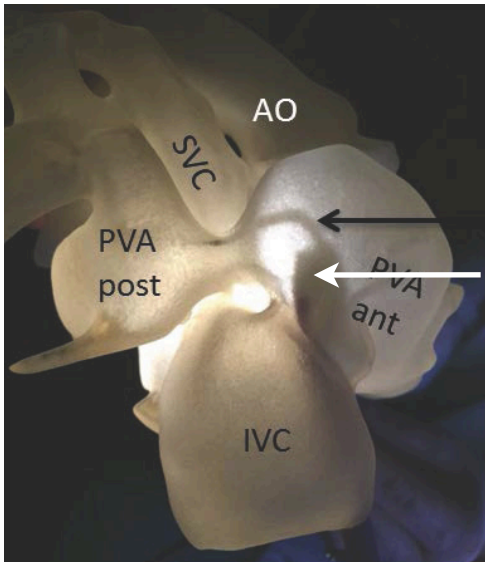

Figure 6. Patient specific right lateral view showing the transbaffle puncture into the anterior compartment of the PVA and the limited "real estate" available to make the acute angle to enter into the posterior compartment of the PVA (arrow).

disadvantages include: the larger introducer in the artery, potential for hemodynamic perturbation by crossing the semilunar and systemic AV valve, catheter entrapment in the AV valve apparatus and finally, the length of the sheath to advance the hand-mounted bare metal stent. Based on published reports and our experience (three separate procedures performed reliably and

reproducibly using retrograde access to the PVA for diagnostic and interventional purposes), these technical challenges are not commonly encountered, and the retrograde approach lends itself nicely for both balloon angioplasty and stent implantation.

A major advantage of the transvenous trans-baffle approach is that it allows the large introducer to be placed in the vein rather than the artery. Based on the model, the transbaffle puncture results in access to the anterior compartment of the PVA (Figures 1, 2, 3, 6). If the PVB stenosis is in the typical location, an acute almost 90 degree bend is needed to cross the PVB stenosis into the posterior compartment. This then leaves relatively little real estate to maneuver the stiff stent/balloon complex. Other considerations include thick calcified baffles, which can make the transbaffle puncture challenging, and the potential of having a residual defect. Therefore, the transbaffle puncture needs to be positioned to maximize the real estate available, and minimize the acute bend if possible. Sometimes the stenosis in the PVA may be in the posterior compartment as described by Mullins et al and in this case the transbaffle approach may be ideal.[11] Therefore, careful pre-procedure planning is critical to determine the best approach. Olivieri et al recently demonstrated utility of patient specific 3D models to plan transvenous intervention for PVB stenosis.[1]

Sareyyupoglu et al have recommended the Hybrid approach.[2] The Hybrid approach has the obviou as advantage of direct access, avoids large introducer in the artery or vein, and other challenges for transvenous and retrograde approach discussed above. However there is some morbidity related to surgical access and the potential for having to deal with dense adhesions.

In conclusion, although the atrial switch operation is no longer commonly performed, patients with a double switch or older patients previously palliated with atrial switch for D-TGA still present for transcatheter interventions.[1-3] It is important to be conversant with the complex 3D anatomy to minimize technical challenges, plan the procedure and ultimately improve patient care. Patient-specific 3D models are extremely useful in planning intervention in patients with complex anatomy. Each approach has its own merit and finally a decision should be made based on experience, patient anatomy and physician or institutional preference.

Acknowledgment

The author would like to acknowledge the contribution of Mark Ginsberg and Bounnak Thammavong from MCG ADAM Product Development and Prototyping Services, Iowa City, IA for their contribution in generating the physical 3D models for illustrations.

"In conclusion, although the atrial switch operation is no longer commonly performed, patients with a double switch or older patients previously palliated with atrial switch for D-TGA still present for transcatheter interventions.1-3 It is important to be conversant with the complex 3D anatomy to minimize technical challenges, plan the procedure and ultimately improve patient care."

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Medical News, Products & Information

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Study in Animal Model Paving Way Forward for Tissue Repair

Newswise - The heart tissue of mammals has limited capacity to regenerate after an injury such as a heart attack, in part due to the inability to reactivate a cardiac muscle cell and proliferation program. Recent studies have indicated a low level of cardiac muscle cell (cardiomyocytes) proliferation in adult mammals, but it is insufficient to repair damaged hearts.

A team led by Ed Morrisey, PhD, a Professor of Medicine and Cell and Developmental Biology and the Scientific Director of the Institute for Regenerative Medicine in the Perelman School of Medicine at the University of Pennsylvania, has now shown that a subset of RNA molecules, called microRNAs, is important for cardiomyocyte cell proliferation during development and is sufficient to induce proliferation in cardiomyocytes in the adult heart. MicroRNAs, which do not generate proteins, repress gene expression by binding messenger RNAs, which do generate proteins, and promote their degradation. The findings appeared the March 17th issue of *Science Translational Medicine.*

The team found that the loss of the microRNA cluster miR302-367 in mice led to decreased cardiomyocyte cell proliferation during development. In contrast, increased expression of the microRNA cluster in adult hearts led to a reactivation of proliferation in the normally non-reproducing adult cardiomyocytes.

This reactivation occurred, in part, through repression of a pathway called Hippo that governs cell proliferation and organ size. “The Hippo pathway normally represses cell proliferation when it is turned on. The cluster miR302-367 targets three of the major kinase components in the Hippo pathway, reducing pathway activity, which allows cardiomyocytes to re-enter the cell cycle and begin to regrow heart muscle,” explains Morrisey. “This is a case of repressing a repressor.”

In adult mice, re-expression of the microRNA cluster reactivated the cell cycle in cardiomyocytes, resulting in reduced scar formation after an experimental myocardial infarction injury was induced in the mice. There was also an increase in the number of heart muscle cells in these same mice.

However, long-term expression of more than several months of the microRNA cluster caused heart muscle cells to dedifferentiation and become less functional. “This suggested to us that persistent reactivation of the cell cycle in adult cardiomyocytes could be harmful and causes the heart to fail,” says Morrisey. The investigators surmised that cardiomyocytes likely need to de-differentiate to divide, but they may lose their ability to contract over time.

“We overcame this limitation by injecting synthetic microRNAs with a short half-life called mimics into the mice,” says Morrisey. Mimic treatment for seven days after cardiac infarction led to the desired increase in cardiomyocyte proliferation and regrowth of new heart muscle, which resulted in decreased fibrosis and improved heart function after injury.

Importantly, the team found that the transient seven-day treatment did not lead to the progressive loss of cardiac function as seen in the genetic models of increased microRNA expression. Overall, these results suggested that any treatment that promotes cardiomyocyte proliferation to improve cardiac regeneration will likely need to be transient to avoid the deleterious effects of maintaining a high level of proliferation and de-differentiation in a tissue that is normally non-proliferative.

“The next stage in this study is to determine whether miRNA mimics will work in a larger animal model and to collaborate with bioengineers to create a local delivery system for the heart, rather than giving it systemically,” notes Morrisey.

Coauthors are: Ying Tian (previously a postdoctoral fellow in the Morrisey Lab who is now an assistant professor at Temple University), Ying Liu, Tao Wang, Ning Zhou, Jun Kong, Li Chen, Melinda Snitow, Michael Morley, Deqiang Li, Nataliya Petrenko, Su Zhou, Minmin Lu, and Kathleen M. Stewart, all from Penn. Erhe Gao and Walter J. Koch are from Temple University.

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First One-Year Report Of Outcomes With Edwards SAPIEN 3 Valve Demonstrate High Survival And Low Stroke Rate

PRNewswire -- Edwards Lifesciences Corporation (NYSE: EW), the global leader in the science of heart valves and hemodynamic monitoring, today announced that the first high-risk European patients to receive the advanced Edwards SAPIEN 3 transcatheter aortic valve via transfemoral delivery had a one-year survival rate of 91.6%, as well as low rates of stroke and paravalvular leak. The independently adjudicated data from the CE Mark study were presented at EuroPCR 2015 by John Webb, MD, Director of Interventional Cardiology and Cardiac Catheterization Laboratories at St. Paul's Hospital, Vancouver, and Professor of Cardiology at the University of British Columbia.

Among the outcomes reported from the study, transfemoral SAPIEN 3 transcatheter aortic valve replacement (TAVR) was associated with a disabling stroke rate of 1.1%. In addition, only 2% of patients had moderate paravalvular leaks and there were no reports of severe leaks. There were also no observations of structural valve deterioration.

"The one-year survival rate in the transfemoral cohort is the highest reported in a multicenter, fully adjudicated TAVR study to date. These results set a new standard of care for patients at high risk for surgical aortic valve replacement. The results of the SAPIEN 3 trial via the transfemoral approach also raise the question of whether transcatheter valve replacement should be considered for approval for all elderly patients with severe aortic stenosis," said Webb, who served as a principal investigator in the trial.

The SAPIEN 3 Trial is a prospective, multicenter, non-randomized study. The one-year analysis documented outcomes of the first 150 patients treated with the SAPIEN 3 valve between Jan. 2013 and Nov. 2013 at 16 centers in France, Germany, Italy, United Kingdom and Canada. Access approaches included transfemoral (n=96) and transapical/transaortic (n=54), as determined by the Heart Team. At baseline, the patients in the alternative access group were significantly sicker than the transfemoral group. All-cause mortality in the alternative access group was reported as 24.3% in the as-treated population. The SAPIEN 3 Trial is designed to evaluate patients annually for five years.

The SAPIEN 3 valve can be delivered through a low-profile 14 French expandable sheath (eSheath). It also has an outer skirt – a cuff of fabric that provides a seal at the bottom of the frame that is designed to reduce paravalvular aortic regurgitation.

The SAPIEN 3 valve was approved in Europe in January 2014 for the treatment of high-risk and non-operable patients with severe aortic

stenosis. The valve is an investigational device not yet available commercially in the United States. It is currently being evaluated in the U.S. in The PARTNER II Trial.

All percents cited in the press release are Kaplan-Meier estimates. Dr. Webb is a consultant to Edwards Lifesciences.

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

Philips Launches Anatomically Intelligent Quantification Tool for Cardiac Ultrasound Imaging

Philips announced the launch of HeartModel$^{A.I.}$, a new Anatomically Intelligent Ultrasound (AIUS) tool that brings advanced quantification, automated 3D views and robust reproducibility to cardiac ultrasound imaging. It was unveiled today on the Philips EPIQ 7 ultrasound system, Philips' first ultrasound with Anatomic Intelligence capabilities, during the *American Society of Echocardiography* (ASE) annual June meeting in Boston, MA.

Some key takeaways from June's unveiling include:

- HeartModelA.I. is Philips' fastest 3D AIUS ultrasound measurement method;
- HeartModelA.I. enables clinicians to quickly, easily and confidently assess disease states, determine treatment, and guide related therapies;
- In a recent comparison, exams with HeartModel$^{A.I.}$ were shown to be three to six times faster than conventional 2D exams in gathering left ventricular and atrial dimensions and volumes (LV and LA), while offering the many benefits of 3D imaging.
- HeartModel$^{A.I}$ has access to advanced clinical information that automatically adapts to variability in patient anatomy.
- HeartModel$^{A.I}$'s knowledge-base identification and patient-specific adaptation provides proven quantification of the left ventricle and atrium, and display of routine apical views.

Two Treatments Yield Similar Results for Children after Cardiac Arrest

NIH-funded research finds therapeutic hypothermia no more effective than normal temperature control.

A large-scale, multicenter study has shown that emergency body cooling does not improve survival rates or reduce brain injury in infants and children with out-of-hospital cardiac arrest more than normal temperature control.

Therapeutic hypothermia, or whole body cooling, can improve survival and health outcomes for adults after cardiac arrest and also for newborns with brain injury due to a lack of oxygen at birth. But, until now, this treatment has not been studied in infants or children admitted to hospitals with cardiac arrest.

The research findings were presented at the *Pediatric Academic Societies Annual Meeting* in San Diego and published simultaneously in the *New England Journal of Medicin*e. The study was funded by the National Heart, Lung, and Blood Institute (NHLBI) of the National Institutes of Health.

"Our results show that therapeutic hypothermia is no more effective for treating children after out-of-hospital cardiac arrest than maintaining body temperature within the normal range," said co-principal investigator Frank W. Moler, MD, a professor in the Department of Pediatrics and Communicable Diseases at the University of Michigan, Ann Arbor. "Both treatments help to control fever and result in similar outcomes for patients."

More than 6,000 children suffer out-of-hospital cardiac arrest in the United States each year, according to the American Heart Association's 2015 heart disease and stroke statistics. During cardiac arrest, the heart stops pumping effectively, and blood stops flowing to the brain and other vital organs. In many cases, the outcome is death or long-term disability.

The study included 295 participants between 2 days and 18 years old who were admitted to children's hospitals for cardiac arrest, required chest compressions for at least two minutes and remained dependent on mechanical ventilation to breathe.

After their parents or guardians provided consent, children were randomly assigned to one of the two treatment groups. One group received body cooling for two days followed by three days of normal temperature control. Another group received normal temperature control for five days.

During the treatment, study participants lay between special blankets. Pumps circulate water through tubes in the blankets to maintain specific body temperature ranges: either a lower range of 89.6°-93.2° F or a normal range of 96.8°-99.5° F.

One year after treatment, researchers observed no difference in survival or cognitive function between groups.

Out-of-hospital cardiac arrest in infants and children typically results from causes such as strangulation, drowning, or trauma. A separate study by the same researchers is examining body cooling in hospitalized patients who suffer cardiac arrest, typically as a complication of a medical condition. A goal of both studies is preventing fever, which commonly occurs after cardiac arrest and can lead to more severe outcomes.

"The findings from these studies may well lead to evidence-based guidelines that will improve the quality and rates of pediatric cardiac arrest survival by using better treatments," said co-principal investigator J. Michael Dean, MD, Professor of Pediatrics and Chief of the Division of Pediatric Critical Care Medicine at the University of Utah School of Medicine, Salt Lake City. "Our hope is to identify the most effective treatment for preventing neurological damage or death in infants and children who suffer cardiac arrest."

The studies are part of the Therapeutic Hypothermia after Pediatric Cardiac Arrest (THAPCA) trials, a six-year effort that is the largest examination of therapeutic hypothermia in children other than newborns for any health condition to date. The trials are funded through NHLBI cooperative agreements U01-HL-094339 and U01-HL-094345.

The THAPCA trials are conducted in partnership with the Collaborative Pediatric Critical Care Research Network, established in 2004 by the NIH's Eunice Kennedy Shriver National Institute of Child Health and Human Development, and the Pediatric Emergency Care Applied Research Network, established in 2001 by the Health Resources and Services Administration's Maternal and Child Health Bureau.

"Partnerships with these federally funded pediatric clinical research networks have been essential to the trials," said Victoria Pemberton, RNC, clinical trials specialist and THAPCA project scientist at NHLBI. "Through the networks, we have been able to mobilize researchers and clinicians throughout North America to answer important questions about a population with a rare health condition."

Supplemental Information:

- THAPCA Trials Home Page: www.thapca.org

- ClinicalTrials.gov THAPCA-OH (out-of-hospital) Trial Page: http://www.clinicaltrials.gov/show/NCT008786444
- ClinicalTrials.gov THAPCA-IH (in-hospital) Trial Page: www.clinicaltrials.gov/show/NCT00880087

The National Heart, Lung, and Blood Institute (NHLBI), part of the National Institutes of Health, plans, conducts, and supports research related to the causes, prevention, diagnosis, and treatment of heart, blood vessel, lung, and blood diseases; and sleep disorders. For more information: www.nhlbi.nih.gov.

The Eunice Kennedy Shriver National Institute of Child Health and Human Development (NICHD), part of the NIH, sponsors research on development, before and after birth; maternal, child, and family health; reproductive biology and population issues; and medical rehabilitation. For more information: www.nichd.nih.gov.
The National Institutes of Health (NIH), the nation's medical research agency, includes 27 Institutes and Centers and is a component of the U.S. Department of Health and Human Services. For more information: www.nih.gov.

LVADs May Lead to Declines in Health, Cognitive Thinking in Some Heart Failure Patients

Left Ventricular Assist Devices (LVADs) are life-prolonging devices for many patients with advanced heart failure, but they also may leave some patients in poor health or with declines in brain function, according to two studies presented at the April *American Heart Association's Quality of Care and Outcomes Research 2015 Scientific Sessions.*

LVADs are battery-operated, mechanical devices surgically implanted to help restore the heart's pumping ability in patients with severe heart failure. The device can be placed temporarily in patients awaiting heart transplants or permanently in patients who are not candidates for heart transplantation.

"Patients with end-stage heart failure often have reduced blood flow to their brains and other organs. Those who receive LVADs should, in theory, have a better ability to think, make decisions and remember things because the device restores blood flow to the brain," said Timothy Fendler, MD, lead author and research fellow at St. Luke's Mid America Heart Institute in Kansas City, MO. "Unfortunately, that is not always the case, due to some common complications that stem from the device itself, such as stroke."

In a large, multi-center study of cognitive function in 1,173 LVAD patients (abstract 401) researchers found:
- More than one in four patients experienced notable cognitive decline in the year following LVAD placement.
- Risk factors for cognitive decline included older age and having devices placed as permanent therapy.

"The study showed that, while patients often experienced the expected result of improved cognitive abilities after receiving an LVAD, cognitive decline events were also unfortunately common," Fendler said. "These results should help clinicians in assessing patients' prognoses and educating potential LVAD candidates, based on risk for this important poor outcome that has not been previously well-described."

In another study (abstract 413), Fendler and colleagues looked at 164 patients who received an LVAD from January 2012 to October 2013 at a single hospital and analyzed how those patients fared for up to one year. Poor overall results included not only death, but also severe, disabling stroke; poor patient-reported quality of life; or multiple readmissions for continued heart failure after the LVAD was implanted.

They found that 35.4% experienced poor overall health. Among these:

- 63.8% died.
- 29.3% reported poor quality of life related to their health.
- 5.2% had two or more heart failure readmissions to the hospital.
- 1.7% had a disabling stroke.

Compared to patients with good results in the year following LVAD placement, the group of patients with poor overall results had longer hospital stays for device placement; had been given LVADs as permanent therapy more often; and had more bleeding events, a common occurrence in patients with this device.

"These studies are important, because although LVAD therapy is a powerful, life-prolonging option that frequently improves heart failure related quality of life in very sick end-stage heart failure patients, they may also be at risk of experiencing device-related complications that could impair their overall quality of life," Fendler said. "While a third of patients with LVAD therapy experienced a poor outcome using our definition, the likelihood of death within one year had these patients not received an LVAD is much higher."

"LVADs represent an important and beneficial therapy that patients should have the option of pursuing, though these patients should be appropriately informed regarding life with LVAD support in order to make individual decisions," Fendler said.

Co-authors of abstract 401 are: John A. Spertus, MD, MPH; Kensey L. Gosch, MS; Philip G. Jones, MS; Jared M. Bruce, PhD; Michael E. Nassif, M.D.; Kelsey M. Flint, M.D.; Shannon M. Dunlay, MD, MS; Larry A. Allen, MD, MHS; and Suzanne V. Arnold, MD, MHA

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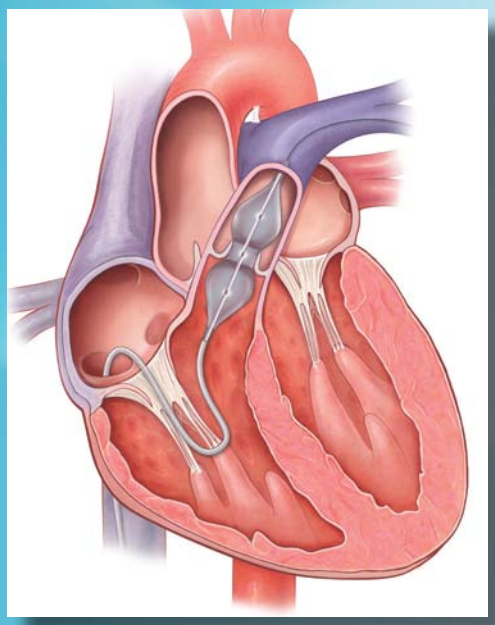

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