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Pulmonary Atresia With Intact Ventricular Septum - Part II

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Introduction

In Part I, the pathologic anatomy, pathophysiology, and clinical features of pulmonary atresia (PA) with intact ventricular septum (IVS) were reviewed along with findings in chest x-ray, electrocardiogram, echocardiogram, and cardiac catheterization with selective cineangiography.[1] In Part II, therapy of PA with IVS will be reviewed.

Therapy

In the mid-1970s, we recommended a comprehensive program to treat patients with PA with IVS[2] which was reiterated in subsequent publications.[3-6] The objective of the treatment plan is to accomplish a four-chamber, biventricular heart, with fully separate pulmonary and systemic circuits. The principles of such a comprehensive approach are: 1). To alleviate hypoxemia by urgent procedures to augment pulmonary blood flow when the baby presents first, usually during the neonatal period, 2). To encourage the growth of the right ventricle (RV) in an attempt to ultimately support pulmonary circulation, 3). To enable satisfactory egress of blood flow from the right atrium (RA), and 4). To finally separate right and left heart circuits. The treatment will be described on the basis of these principles.

1. Management of the Neonate at Initial Presentation

General Management

Management of the newborn with PA with IVS is analogous to that of any infant with cyanotic CHD.[7-9] Infusion of intravenous prostaglandin E_1 (PGE_1) to keep the ductus open[10,11] and perfuse the pulmonary circulation should be instituted similar to all babies with ductal dependent pulmonary blood flow while the diagnostic studies completed in order to confirm the diagnosis.

Restore Pulmonary Blood Flow

Since the pulmonary valve is atretic, there is no forward flow from the RV into the lungs and, consequently, PGE_1 should be stated as soon as a diagnosis of PA with IVS is suspected/made. We

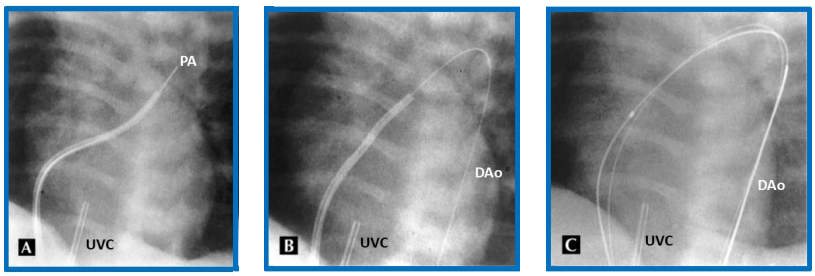

FIGURE 1 *Cine-radiographic frames illustrating passage of the blunt end of a guidewire into the pulmonary artery (PA) (A) and then into the descending aorta (DAo) following reversal (B) of the coronary guidewire. A second guidewire was similarly advanced into the DAo (C) when a balloon angioplasty catheter could not initially be advanced across the atretic pulmonary valve membrane. UVC, umbilical venous catheter. Modified from Reference 5.*

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Z-5™ Catheter vs. Z-6™ Catheter

UNDERSTANDING THE DIFFERENCES

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usually start with PGE_1 dose of 0.05 to 0.1 mcg/kg/min and once the O_2 saturation improves, the dosage is slowly, step-by-step, reduced to 0.015 to 0.02 mcg/Kg/min.

Once the baby is stabilized, alternative methods to more permanently perfuse the lung should be considered and these include: A) Transcatheter perforation of the atretic pulmonary valve along with balloon pulmonary valvuloplasty,[4,5,12,13] surgical pulmonary valvotomy/enlargement of the RV outflow tract[14-18] or hybrid opening of the pulmonary valve.[19-21] These procedures are performed also to encourage the growth of the RV so that a biventricular circulation can be established, B) Performing a modified Blalock-Taussig (BT) shunt,[22,23] or C) Implantation of stent within the ductus.[4,24,25]

2. Encourage the Growth of the RV

The RV growth may be encouraged by transcatheter, surgical or hybrid approaches:

Transcatheter Perforation of the Pulmonary Valve

Prologue

Transcatheter opening of the pulmonary valve provides most natural way of perfusing the pulmonary circulation in patients with PA with IVS and is the most frequently performed procedure around the world to address this condition. In addition, the growth of the RV is encouraged by opening the pulmonary valve.[2,3,5] Initially, blunt ends of coronary guide wires[4,26] **(Figure 1)**, laser wires[27,28] and coronary recanalization wires[29,30] were used to perforate the pulmonary valve and more recently, radio frequency wires[31-34] were utilized for this purpose. Once the guidewire is positioned across the pulmonary valve, balloon pulmonary valvuloplasty is performed with progressively larger balloons.

Indications and Contraindications

Patients with bipartite or tripartite ventricles with mild to moderate hypoplasia are candidates for transcatheter opening of the pulmonary valve. However, one must first ensure that there is no RV-dependent coronary circulation (RV-DCC). Contraindications for performing the procedure are infundibular atresia and evidence for RV-DCC. Of course, the procedure should not be undertaken if the tip of the catheter could not be positioned directly beneath the atretic pulmonary valve apparatus.

Procedure

The technique of percutaneous treatment of PA with IVS was described in the past[4-6,12,13,35-37] and will be reviewed here briefly. The usual hemodynamic data are secured and appropriate angiography performed to confirm the diagnosis of PA with IVS, with particular attention to exclude RV-DCC. Selective cineangiography of the RV in sitting-up (15° left anterior oblique and 35° cranial) and straight lateral projections **(Figure 2A)** is performed to evaluate the size and partite status of the RV and to establish the membranous nature of the atretic pulmonary valve membrane. If necessary, simultaneous RV and pulmonary artery angiograms **(Figure 3)** are secured. These images also serve as roadmaps during the procedure.

A #4- or 5F right coronary artery catheter (Cook, Bloomington, IN) or a similar catheter is advanced into the RV outflow tract and the catheter tip is manipulated so that it is apposed against the center of the atretic pulmonary valve membrane **(Figure 4)**. If necessary, main pulmonary artery cine-angiogram **(Figure 5)** may be performed to ensure accurate positioning of the catheter tip. The techniques of usage of blunt ends of coronary guide wires,[4,5,26] laser wires[27,28] and coronary recanalization

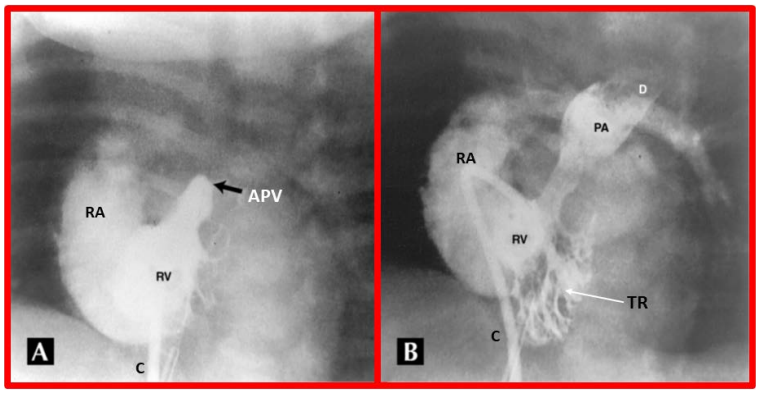

FIGURE 2A. *Cine-angiographic images of the right ventricle (RV) in a sitting-up view secured before (A) and 15 minutes after (B) perforation of the atretic pulmonary valve (APV) membrane (thick arrow) as shown in Figure 1 and balloon pulmonary valvuloplasty. A small and heavily trabeculated RV is seen without anterograde flow into the pulmonary artery.*
FIGURE 2B. *After the procedure the pulmonary artery (PA) is opacified. Pulmonary end of the ductus arteriosus (D) is also seen. A larger proportion of the trabecular (TR) component of the RV is also seen following the opening of the pulmonary valve. Significant tricuspid regurgitation, opacifying the right atrium (RA) is seen both before and after opening the pulmonary valve. (C) catheter. Modified from Reference 4.*

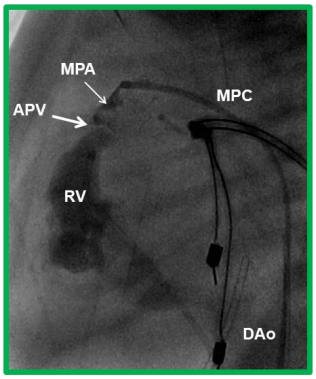

FIGURE 3 *Cine image from lateral view of simultaneous injection of contrast material into right ventricle (RV) and main pulmonary artery (MPA) (thin arrow) in a newborn baby with pulmonary atresia with intact ventricular septum illustrating thin atretic pulmonary valve (APV) (thick arrow). MPA angiogram was performed via a multipurpose catheter (MPC) introduced into the MPA from the descending aorta (DAo) through a patent ductus arteriosus (not marked). Unlabeled artifacts are seen and are related to radiofrequency equipment placed beside the baby. Modified from Reference 36.*

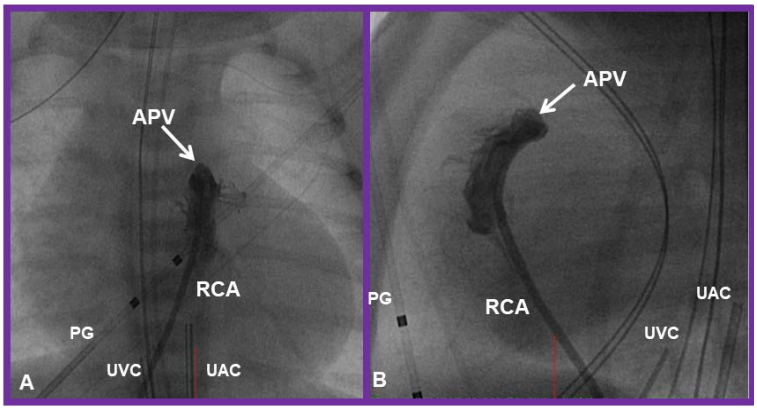

FIGURE 4 *Cine-angiographic images of the right ventricular outflow tract in postero-anterior* **(A)** *and straight lateral* **(B)** *projections in a baby with pulmonary atresia with intact ventricular septum illustrating the location of the tip of the right coronary artery (RCA) catheter beneath the atretic pulmonary valve (APV) (arrows in A and B). PG, pigtail catheter used for calibration purposes; UAC, umbilical artery catheter; UVC, umbilical venous catheter. Reproduced from Reference 36.*

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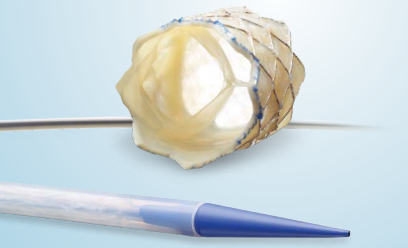

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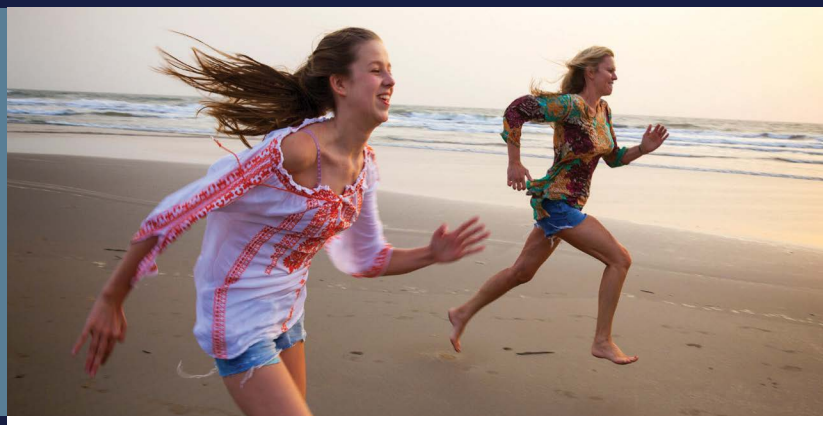

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- **DO NOT implant in the aortic or mitral position. Pre-clinical bench testing of the Melody valve suggests that valve function and durability will be extremely limited when used in these locations.**
- DO NOT use if patient's anatomy precludes introduction of the valve, if the venous anatomy cannot accommodate a 22 Fr size introducer, or if there is significant obstruction of the central veins.
- DO NOT use if there are clinical or biological signs of infection including active endocarditis. Standard medical and surgical care should be strongly considered in these circumstances.
- Assessment of the coronary artery anatomy for the risk of coronary artery compression should be performed in all patients prior to deployment of the TPV.
- To minimize the risk of conduit rupture, do not use a balloon with a diameter greater than 110% of the nominal diameter (original implant size) of the conduit for pre-dilation of the intended site of deployment, or for deployment of the TPV.
- The potential for stent fracture should be considered in all patients who undergo TPV placement. Radiographic assessment of the stent with chest radiography or fluoroscopy should be included in the routine postoperative evaluation of patients who receive a TPV.
- If a stent fracture is detected, continued monitoring of the stent should be performed in conjunction with clinically appropriate hemodynamic assessment. In patients with stent fracture and significant associated RVOT obstruction or regurgitation, reintervention should be considered in accordance with usual clinical practice.

Potential procedural complications that may result from implantation of the Melody device include the following: rupture of the RVOT conduit, compression of a coronary artery, perforation of a major blood vessel, embolization or migration of the device, perforation of a heart chamber, arrhythmias, allergic reaction to contrast media, cerebrovascular events (TIA, CVA), infection/sepsis, fever, hematoma, radiation-induced erythema, blistering, or peeling of skin, pain, swelling, or bruising at the catheterization site. Potential device-related adverse events that may occur following device implantation include the following: stent fracture,* stent fracture resulting in recurrent obstruction, endocarditis, embolization or migration of the device, valvular dysfunction (stenosis or regurgitation), paravalvular leak, valvular thrombosis, pulmonary thromboembolism, hemolysis.

*The term "stent fracture" refers to the fracturing of the Melody TPV. However, in subjects with multiple stents in the RVOT it is difficult to definitively attribute stent fractures to the Melody frame versus another stent.

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Indications: The Melody™ TPV is indicated for use in patients with the following clinical conditions:

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Contraindications

- Venous anatomy unable to accommodate a 22 Fr size introducer sheath
- Implantation of the TPV in the left heart
- RVOT unfavorable for good stent anchorage
- Severe RVOT obstruction, which cannot be dilated by balloon
- Obstruction of the central veins
- Clinical or biological signs of infection
- Active endocarditis
- Known allergy to aspirin or heparin
- Pregnancy

Potential Complications/Adverse Events: Potential procedural complications that may result from implantation of the Melody device include the following: rupture of the RVOT conduit, compression of a coronary artery, perforation of a major blood vessel, embolization or migration of the device, perforation of a heart chamber, arrhythmias, allergic reaction to contrast media, cerebrovascular events (TIA, CVA), infection/sepsis, fever, hematoma, radiation-induced erythema, pain, swelling or bruising at the catheterization site. Potential device-related adverse events that may occur following device implantation include the following: stent fracture.* stent fracture resulting in recurrent obstruction, endocarditis, embolization or migration of the device, valvular dysfunction (stenosis or regurgitation), paravalvular leak, valvular thrombosis, pulmonary thromboembolism, hemolysis.

*The term "stent fracture" refers to the fracturing of the Melody TPV. However, in subjects with multiple stents in the RVOT it is difficult to definitively attribute stent fractures to the Melody frame versus another stent.

For additional information, please refer to the Instructions for Use provided with the product or available on http://manuals.medtronic.com.

The Melody Transcatheter Pulmonary Valve and Ensemble II Transcatheter Delivery System has received CE Mark approval and is available for distribution in Europe.

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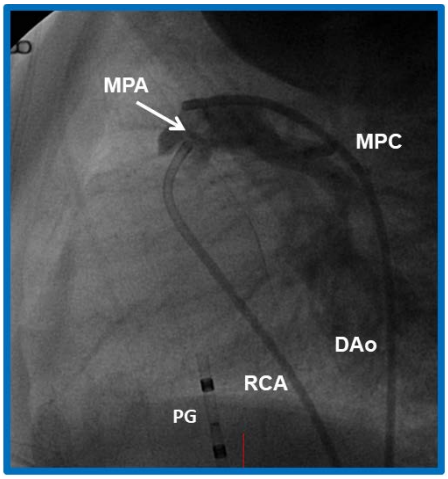

FIGURE 5
Cine-angiographic image of the main pulmonary artery (MPA) in a lateral view; the contrast was injected through a multipurpose catheter (MPC) placed into the MPA from the descending aorta (DAo) via a patent ductus arteriosus (not marked) in a neonate with pulmonary atresia with intact ventricular septum. This cine frame demonstrates the location of the tip of the right coronary artery (RCA) catheter in apposition to the atretic pulmonary valve. PG, pigtail catheter used for calibration purposes. Modified from Reference 36.

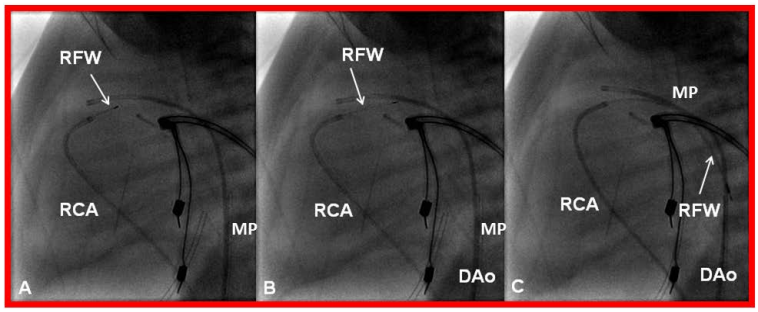

FIGURE 6 A & B) *Cine-radiographic images in straight lateral projections displaying the radiofrequency wire (RFW) (arrows) as it is gradually advanced from the right coronary artery (RCA) catheter into the main pulmonary artery and then into the descending aorta (DAo) via the ductus arteriosus (not labelled).* **C)** *A multipurpose catheter (MP) from the DAo is seen to be positioned in the pulmonary artery via the ductus arteriosus (not labelled) (A, B and C) and served as a guide during the procedure. Unlabeled artifacts are seen and are related to radiofrequency equipment placed beside the baby. Modified from Reference 36.*

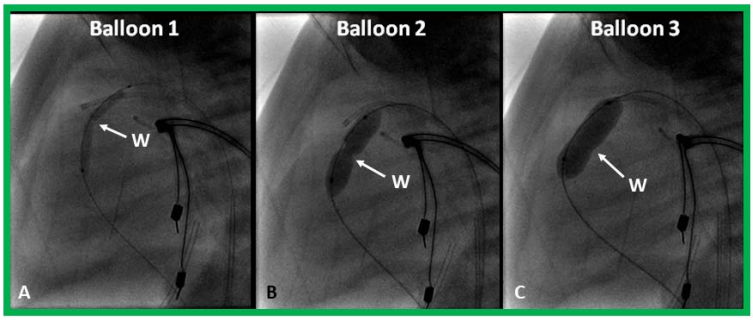

FIGURE 7 *Cine-radiographic frames in straight lateral projections displaying images of balloon dilatation catheters placed across the pulmonary valve following radiofrequency perforation of the atretic pulmonary valve. Progressively larger sized balloons are used and waisting (W) of the balloon (arrows in A, B & C) are seen during the early phases of inflation of the balloons. The waists disappeared after complete inflation of the balloons (not shown). Unlabeled artifacts are seen and are related to radiofrequency equipment placed beside the baby. Modified from Reference 36.*

wires[29,30] in crossing the atretic pulmonary valve have been described in the respective publications[4,5,26-30] and will not be reviewed. Now that radiofrequency guidewires are approved for clinical use by the Food and Drug Administration (FDA) and are commercially available (Baylis Medical Corporation (BMC), Montreal, Canada), we will describe the technique that we use in perforating the atretic pulmonary valve with radiofrequency guidewires.

A Protrach™ Micro catheter (BMC) is preloaded with a radiofrequency perforation wire (BMC) and is positioned beneath the atretic pulmonary valve. After confirming the position of the wire **(Figures 4 & 5)**, a low power (5 watts) radiofrequency energy is delivered for one to two seconds with BMC radiofrequency perforation generator (BMC). This typically produces perforation of the atretic pulmonary valve. The wire is then advanced either into a branch pulmonary artery or into the descending aorta **(Figure 6)**; our preference is descending aorta since such a position provides greater stability of the wire and facilitates successful progression of the procedure. The outer microcatheter is then pushed forward over the radiofrequency wire across the perforated pulmonary valve and placed into either branch pulmonary arteries or into the descending aorta via the ductus **(Figure 6C)**. At this juncture, the radiofrequency wire is swapped with a 0.014" coronary guide wire and the microcatheter removed.

After the tip of the guidewire is stabilized in the pulmonary artery or descending aorta, balloon pulmonary valvuloplasty is undertaken, usually with the use of increasingly larger balloon-diameter catheters **(Figure 7)**. Low profile balloon dilatation catheters such as Tyshak-II or Mini-Tyshak (Braun) of 6 to 8 mm balloon diameter are positioned over the wire, the balloon is inflated with diluted (1 in 4) contrast material, and the pulmonary valve dilated. In most patients, pre-dilation with 3- or 4-mm coronary balloon dilatation catheter may be needed **(Figure 7A)**. The final balloon size should be 1.2 to 1.25 times the diameter of the pulmonary valve annulus. In patients in whom it is not feasible to advance the balloon catheter across the pulmonary valve, snaring the guidewire in the descending aorta and holding it tight may help advance the balloon catheter; we have occasionally used this technique to accomplish the procedure. After completion of the procedure, pressure pullback tracing across the pulmonary valve and RV outflow tract to document residual gradients are made and RV angiography performed **(Figures 2B)**.

While radiofrequency wire perforation of the atretic pulmonary valve is the most frequent method used currently, perforation of the atretic pulmonary valve with the stiff ends of coronary wires,[4,5,26] laser wires[27,28] and coronary recanalization wires[29,30] has been shown to be similarly effective in achieving the objective of crossing the atretic pulmonary valve.

Surgical Relief of Pulmonary Outflow Obstruction

Trans-ventricular[15,16] and trans-pulmonary arterial[14,17] closed pulmonary valvotomy procedures without using cardiopulmonary bypass were used in the past, but are no longer used routinely. At the present time, pulmonary valvotomy is performed via an incision in the pulmonary artery under cardiopulmonary bypass along with mild hypothermia as deemed appropriate. If the pulmonary valve annulus is small, it is enlarged using a pericardial patch or other prosthetic materials. If RV infundibular obstruction coexists, it is also enlarged with a RV outflow tract patch.[14,18,38]

Hybrid Procedure

Some centers have used a hybrid approach if transcatheter intervention did not achieve an appropriate catheter position suitable to perforate the pulmonary valve safely or de novo. The heart is exposed by a sternotomy by the surgeon and the cardiologist introduces a needle through the RV outflow tract and perforates the atretic pulmonary valve. Then sequential balloon dilatations were performed (as describe above) by the balloon valvuloplasty catheters introduced through the RV

outflow tract. Such procedures may be undertaken in a regular surgical theater using transesophageal echo guidance and nominal fluoroscopy, or in a hybrid catheterization-surgical suite.[19-21]

Comments

The concept that the hypoplastic RV will grow in size following opening of the pulmonary valve was introduced in mid 1970s[2] and reiterated subsequently.[3-6] Other investigators[39-41] confirmed these observations in that they demonstrated growth of the RV following relief of RV outflow obstruction, although improvement in tricuspid annular size did not seem to occur. Consequently, opening of the atretic pulmonary valve either by surgical or transcatheter methodology should be pursued in PA with IVS patients provided there is no RV-DCC.

In spite of complete opening of the pulmonary valve by any of the above-described methods and establishing forward flow across the pulmonary valve, a significant number of patients remain hypoxemic, related to right to left shunt across the patent foramen ovale/atrial septal defect (PFO/ASD) which is largely secondary to decreased RV compliance. Such arterial desaturation is addressed by re-initiation of PGE_1 administration, creation of a BT shunt or stenting of the ductus arteriosus. The latter two procedures were initially performed after unsuccessful attempts to wean off of PGE_1. However, some cardiologists/cardiac surgeons are advocating that these procedures are undertaken at the time as the opening of the pulmonary valve.[41,42]

Blalock-Taussig Shunt

A number of procedures have been used in the past to augment pulmonary blood flow, as reviewed elsewhere.[9] Among these, BT shunt has become an attractive surgical procedure to provide pulmonary blood flow. This procedure was originally described by Blalock and Taussig in which the subclavian artery is anastomosed to the ipsilateral pulmonary artery[22] and is now called classic BT shunt. The procedure was modified by de Leval and his colleagues[23] in which a Gore-Tex tube graft is inserted between the right or left subclavian artery and the branch pulmonary artery on the same side. This procedure is usually performed via a lateral thoracotomy. Some surgeons perform the procedure via mid-sternotomy.

The indications for this procedure are severely hypoplastic, unipartite or bipartite RV,[4,12,13] failure to keep satisfactory arterial oxygen saturation (> 70%) following successful transcatheter opening of the atretic pulmonary valve[4,12,13] and patients with RV-DCC.[37] Because of uncertain outcome following surgical opening of the pulmonary valve, some institutions advocate performing BT shunt at the time initial surgery.[41]

Implantation of Stent Within the Ductus Arteriosus

The technique of ductal stenting in patients with PA with IVS was reviewed in the past[4,5,36,37] and will be described here briefly.

Indications and Contraindications

The indications for ductal stenting are similar to those mentioned for BT shunt placement. Because of uncertain outcome following transcatheter opening of the pulmonary valve, some institutions advocate implanting ductal stents at the time of initial procedure.[42] Contraindications are markedly tortuous ductus arteriosus and nonavailability of all materials necessary for ductal stenting. Left pulmonary artery stenosis is another contraindication,[25] although such an abnormality is more likely to be seen in patients with PA with ventricular septal defect.

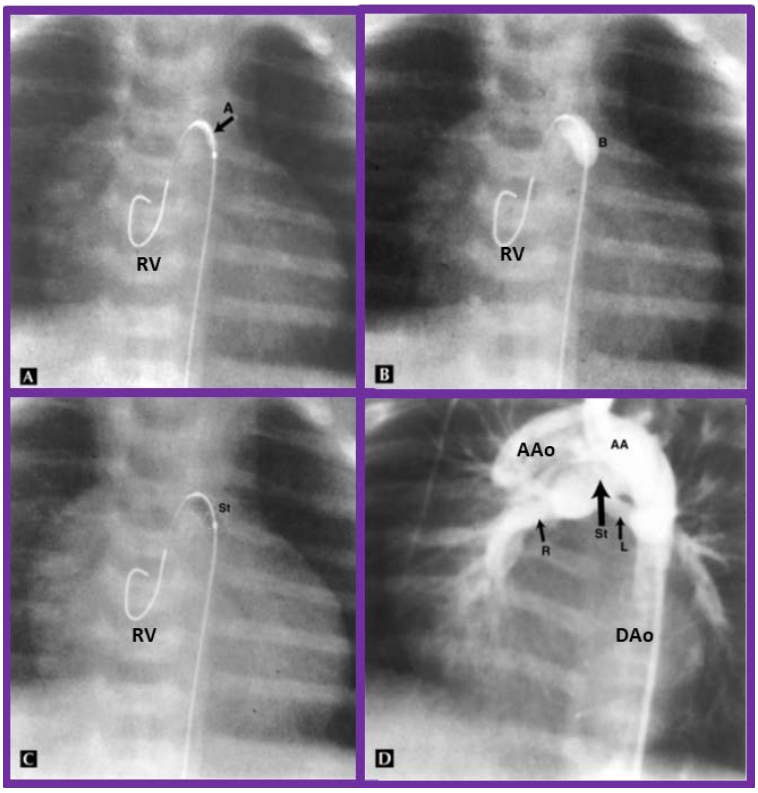

FIGURE 8 **A)** *Cine-radiographic image illustrating the position of the distal end of the guidewire placed in the right ventricle (RV) through the ductus arteriosus and main pulmonary artery (not marked). The uninflated stent is located within the ductus and the arrow (A) identifies the articulation within the stent.* **B)** *This cine image demonstrates inflated balloon (B).* **C)** *The balloon is deflated and removed leaving the stent (St) within the ductus.* **D)** *Cine-angiographic picture of aortic arch (AA) in left anterior oblique view showing visualization of the St (large arrow) within the ductus and the right (R) and left (L) pulmonary arteries (small arrows). AAo, ascending aorta; DAo, descending aorta. Modified from Reference 5.*

Procedure

At first, cine-aortography in straight lateral and sitting-up (15^0 left-anterior oblique and 35^0 cranial) or 30^0 right anterior oblique projections is performed to demonstrate the morphology of the ductus arteriosus. The ductal length and minimal ductal diameter are measured. Frozen frames of the aortograms serve as road maps during the procedure. Once the ductal morphology is assessed, a #4-French right coronary artery, a cut-pigtail catheter, or a similar catheter is placed in the aortic arch and its tip is pointed against the aortic ampulla of the ductus. The tip of a 0.014-in coronary guide wire (Choice PT Extra S'port [Boston Scientific], Hi Torque Spartacore guide wire [Guidant] or a similar wire) is advanced into the ductus and from that position conveyed into either distal left or right pulmonary arteries. If the tip of the guidewire crosses the pulmonary valve and enters the RV **(Figure 8A)**, such a wire position is also accepted. There is a possibility of change in ductal length following its straightening by the introduction of the guidewire; therefore, we recommend re-measuring the ductal length by performing a test angiogram with guidewire across the ductus. At this juncture, the catheter used for angiography is withdrawn and the femoral arterial sheath exchanged with a #4-French (or #5-French) long sheath (Cook, Bloomington, IL). Alternatively, stent implantation may be performed without the long sheath as per the interventionalist's preference. A 3.5 to 4.5 mm diameter pre-mounted coronary stent is selected for deployment; stent diameter choice is based on the measured minimal ductal

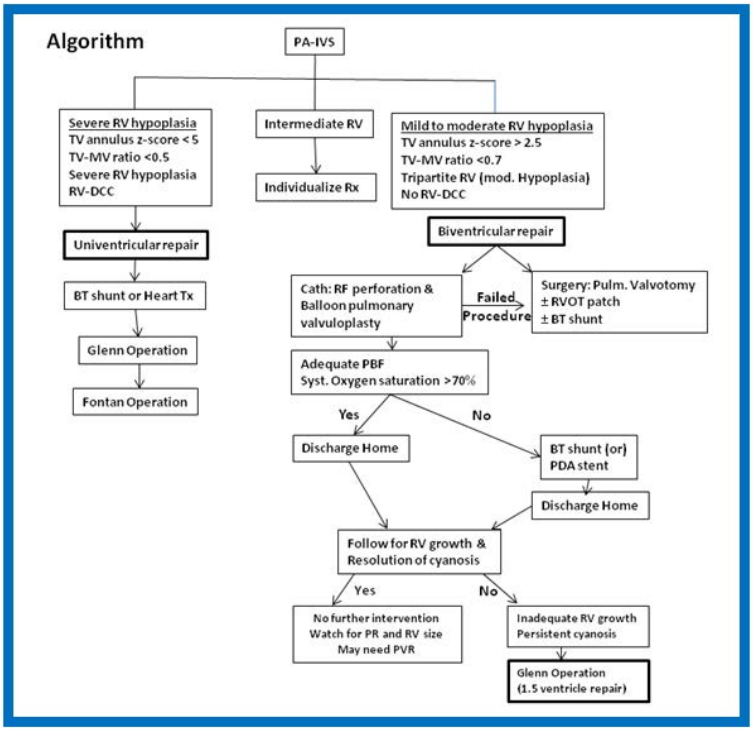

FIGURE 9 *An algorithm for clinical decision-making in pulmonary atresia with intact ventricular septum (PA-IVS). This algorithm is partly based on References 13, 59 and 66. BT, Blalock-Taussig shunt; MV, mitral valve; PBF, pulmonary blood flow; PDA, patent ductus arteriosus; PR, pulmonary regurgitation; PVR, pulmonary valve replacement; RF, radiofrequency; RV, right ventricle; RV-DCC, right ventricular dependent coronary circulation; RVOT, right ventricular outflow tract; Rx, treatment; TV, tricuspid valve; Tx, transplant. Reproduced from Reference 13.*

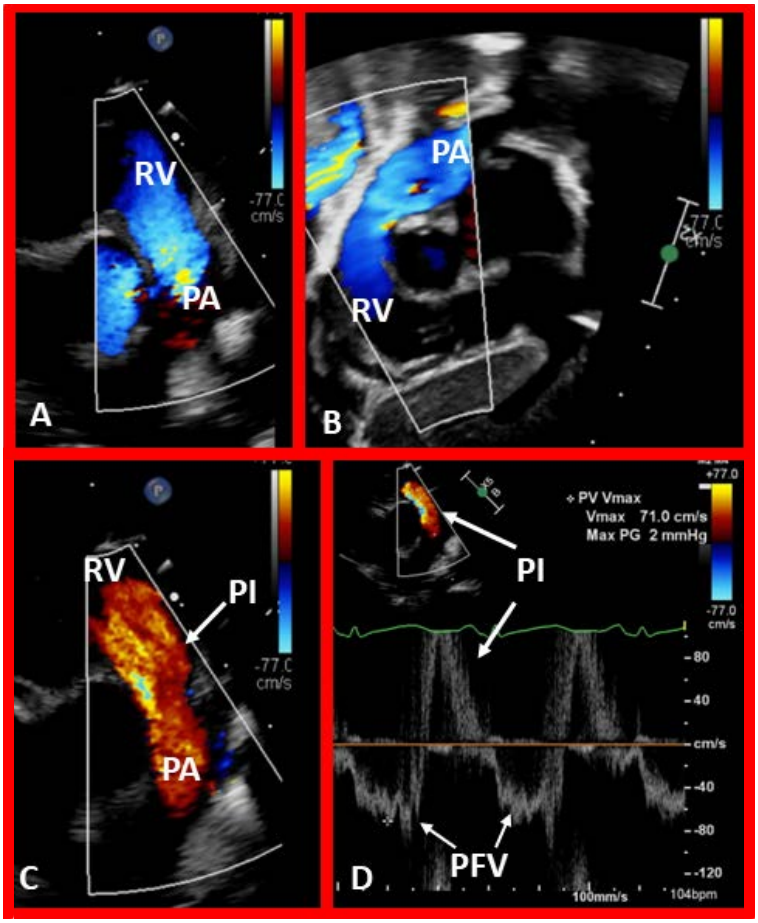

FIGURE 10 *Selected echo images from parasternal (A, C & D) and subcostal (B) views demonstrating laminar flow across the pulmonary valve (A, B & D) and pulmonary insufficiency (PI) (C & D) in a nine-year-old child who had transcatheter opening of the atretic pulmonary valve as a neonate. Note low pulmonary flow velocity (PFV) indicating no residual gradient (D). PA, pulmonary artery; RV, right ventricle.*

diameter and weight of the infant (<3.0 kg - 3.5 mm diameter stent; 3.0 to 4.0 kg - 4.0 mm; 4.0 to 5 kg - 4.5 mm[43,44]). The length of the stent should be 1 to 2 mm longer than the measured length of the ductus. The selected stent is advanced through the arterial sheath, but over the guidewire already in position and deployed across the ductus arteriosus by inflating the balloon **(Figures 8B and 8C)**. If a long sheath was used for stent deployment, the sheath is withdrawn into the descending aorta before balloon inflation. Several types of stents are available for use[36] and selection of the type of stent is mostly based on the availability at that particular time at a given institution. Following successful stent deployment, the deflated balloon is cautiously withdrawn without dislodging the stent. Similarly, the guidewire is also taken out. It is critical to ensure that the entire length of the ductus is covered by the stent in an attempt to prevent constriction of the unstented ductus. A repeat aortogram is performed to demonstrate patency of the stent and visualization of the branch pulmonary arteries **(Figure 8D)**. PGE_1 is discontinued shortly before inflating the balloon. Measurement of systemic arterial oxygen saturation and PO_2 is done before removal of the catheters and sheaths used during the procedure.

Comparison of Blalock-Taussig Shunts and Ductal Stents

When relative merits of these two procedures in maintaining adequate pulmonary flow were examined,[45] procedural success (~93% for both) and major complication rates were similar between the two cohorts. But the hospital stay was shorter for the ductal stent group (10 days vs. 23 days). Need for acute re-intervention was more frequent for BT shunt group (31% vs. 8%) than for the ductal stent group while post-discharge re-interventions occurred more often in the ductal stent group (58% vs. 14%). The authors concluded that ductal stents may offer safer and more effective alternative strategy to BT shunts in patients with PA with IVS.[45] Another study in which these two procedures were compared included patients with several types of heart defects (not just PA with IVS), but concluded that both procedures had similar need for reintervention to sustain acceptable pulmonary blood flow.[46]

3. Provide Satisfactory Egress of Blood Flow From the Right Atrium

If successful opening of the pulmonary valve by any of the methods described in the preceding section can be performed, the egress of the RA blood into the RV will occur naturally. But, in cases with very hypoplastic RVs or those with RV-DCC are usually treated with a BT shunt for providing pulmonary blood flow. In these patients, balloon atrial septostomy[36,47,48] should be performed to provide unrestricted right to left shunting, as has been demonstrated to be beneficial in a prior study.[49]

4. Separate Right and Left Heart Circuits

All babies, irrespective of type of intervention to open the atretic pulmonary valve during the neonatal period, should be re-evaluated between 6 and 12 months of age (and time-to-time thereafter) to assess the adequacy of relief of RV outflow tract obstruction and to

appraise the RV growth. Patients with residual RV outflow tract obstruction should be treated with transcatheter (balloon valvuloplasty) or surgical (resection of obstructive lesion and/or outflow patch) intervention to relieve the RV outflow obstruction, as deemed appropriate. In patients with adequate-sized RVs, closure of the atrial defect by transcatheter methodology[50,51] should be performed, preferably following test occlusion[50] of the atrial defect.

Other Treatment Modalities

A number of other procedures are performed to treat PA with IVS patients and these are bidirectional Glenn,[52] one and one-half ventricle repair,[53] staged Fontan surgery,[54-58] Starnes procedure[59] for tricuspid valve leaflet dysplasia with or without Ebstein's type of abnormality, cardiac transplantation particularly for patients with RV-DCC,[60,61] and surgical[62-64] or device closure[50,51,65,66] of PFO/ASD. Because of limitations of space, these treatment modalities will not be detailed.

Algorithms of Management

How to approach a given PA with IVS patient is largely dependent upon the morphology of the RV. Feasibility of achieving biventricular circulation, going with univentricular palliation or in-between (one and one-half ventricle) should be assessed at the time of initial presentation and time to time thereafter. There is no universally accepted algorithm of management, and the algorithm that we use **(Figure 9)** is developed on the basis of our experience and that of others.[3-5,12,13,60,67]

Adequate-sized RV

If the RV is bipartite or tripartite with mild to moderate hypoplasia, the tricuspid valve annulus diameter Z-score is larger than –2.5 and the tricuspid/mitral valve annulus ratio is greater than 0.7, the RV is likely to grow to support the pulmonary circulation[12,13] and biventricular option should be selected. Transcatheter (preferable) or surgical opening of the pulmonary valve should be undertaken as described in the preceding sections. Of course, absence of RV-DCC should be assured prior to opening of the pulmonary valve. Then, percutaneous closure of the atrial septal defect (if present) should be performed[50,51,65,66] to maintain normal oxygen saturations and to prevent paradoxical embolism. While opening up the pulmonary valve reduces/abolishes pulmonary valve gradient **(Figure 10)** with laminar flow across the pulmonary valve **(Figure 10 A and B)**, pulmonary insufficiency **(Figure 10 C and D)** is present in most patients. Some of these patients may require replacement of the pulmonary valve in their adolescence or adulthood.

Small and Hypoplastic RV

If the RV is unipartite or bipartite with an absent sinus portion, tricuspid valve annulus diameter Z-score is smaller than –5 and the tricuspid/mitral valve annulus ratio is less than 0.5, the RV is unlikely to grow sufficiently to support the pulmonary circulation.[12,13,68] Consequently, these patients are candidates for single ventricle palliation by Fontan procedure (staged total cavo-pulmonary connection) which would involve BT shunt (or ductal stent) during the neonatal period (along with balloon septostomy or surgical atrial septostomy), bidirectional Glenn[52] around six months of age and inferior vena caval blood flow diversion into the pulmonary arteries, usually via an extra-cardiac conduit and a fenestration[55-58] a year afterwards.

Marginal-sized RV

If the RV morphology falls in between the above two scenarios, transcatheter or surgical opening of the atretic pulmonary valve may not achieve a RV size large enough to support pulmonary circulation. These patients are addressed with one and one-half ventricle repair[53] in which bidirectional Glenn procedure[52] is performed, thus reducing volume that needs to be handled by the RV. The atrial defect should also be closed.

Other Scenarios

RV-DCC

Since PA with IVS patients with RV-DCC do not tolerate opening of the pulmonary valve, they are either addressed with cardiac transplantation, Starnes procedure or by Fontan, largely based institutional preference.

Tricuspid Atresia

In some patients the tricuspid valve is extremely small[69] and these patients are addressed similar to tricuspid atresia patients[70-72] by Fontan procedure in stages as alluded to above.

Tricuspid Valve Regurgitation

In PA with IVS patients with severe tricuspid regurgitation due to valve dysplasia with or without Ebstein's are probably best address by Starnes procedure[59] during the neonatal period and then staged Fontan.

Summary and Conclusions

A comprehensive program of medical, transcatheter, and surgical treatment strategy is essential to improve the long-term prognosis of these babies. Algorithms of treatment plans based on the size and morphology of the right ventricle, tricuspid annular diameter Z-scores and the presence of RV-DCC should be implemented. In babies with tripartite or bipartite RVs, transcatheter radiofrequency perforation is the first-line therapy at most institutions at the present time. Improving pulmonary blood flow by infusion of PGE_1, stenting the ductus, or a modified Blalock-Taussig shunt may be necessary in some of these babies. These babies will eventually achieve biventricular status. Subjects with a unipartite or very hypoplastic RV or RV-DCC require staged Fontan approach. Cardiac transplantation is used for RV-DCC patients at some institutions. Patients with anatomy between above two scenarios may be addressed with one and one-half ventricle repair. It may be concluded that PA with IVS can be successfully assessed with the presently available diagnostic methods and these patients can effectively be managed with the currently existing medical, catheter interventional, and surgical therapeutic practices.

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Work-Life Balance: The Expectations and Perspectives as a Female Interventional Cardiologist

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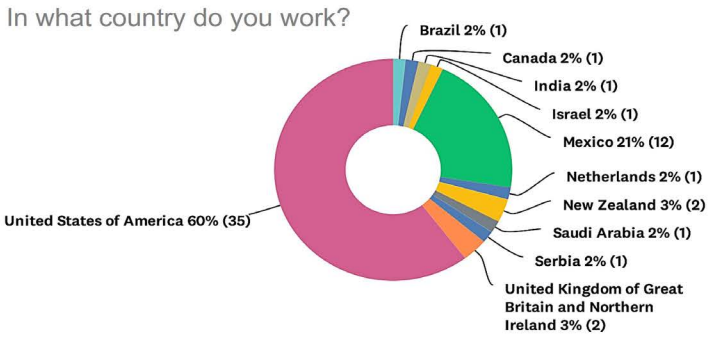
Introduction

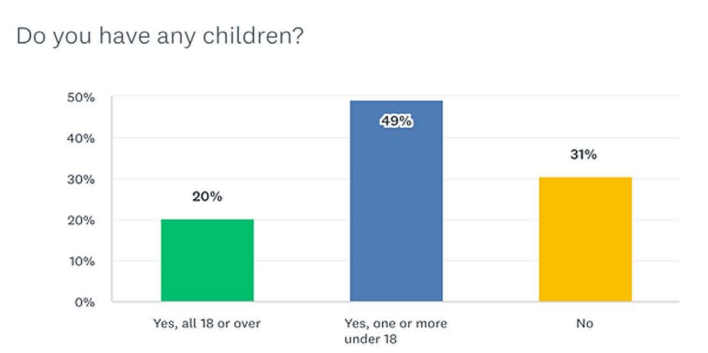
Work-life balance (WLB) refers to the harmony and interference that occurs between work and personal life. WLB can also simply be defined as the equal time of priority to personal and professional activities. This has become a primary concern to all of us wishing to have a good quality of life. A more complex term, Work-life integration (WLI) has become known in more recent years; and refers to the integration of life and work creating synergy between them. WLI works only if it is combined by "balance" in both areas, otherwise there is the concern that it would be difficult to maintain work-life boundaries.

Being women interventional cardiologists seeking career advancement we manage multifaceted roles and responsibilities, and promotion into leadership positions has been marked with increased pressure and frustrations regarding women's roles and our desire to balance our work and non-work lives.

Studies over the years demonstrate the differences in WLB in physicians and especially female physicians. In serial studies by Shanafelt et. al, with the latest one in 2020 and included over 7,500 physicians, the results showed that physicians had a lower rate of satisfaction with WLI than the general US working population (43.6% vs 62.5%; OR, 0.46; 95% CI, 0.421 to 0.512; P<.001). On multivariable analysis of the 2020 data, being female and working more hours per week were independently associated with higher rates of burnout and lower degrees of satisfaction with WLI.[1,2]

FIGURE 1 *Demographics*





Interventional Women Cardiologists in 2023 and WLB: Objective Data

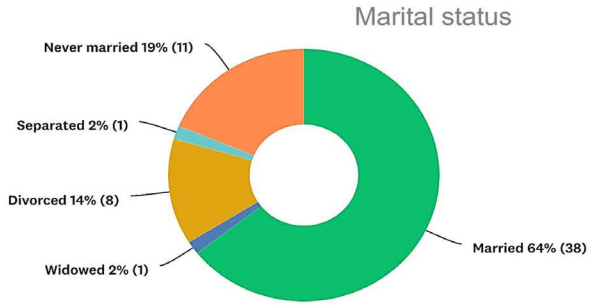
We decided to understand the current rate of WLB among female interventional cardiologists and some life and work factors that may affect our perception of WLB.

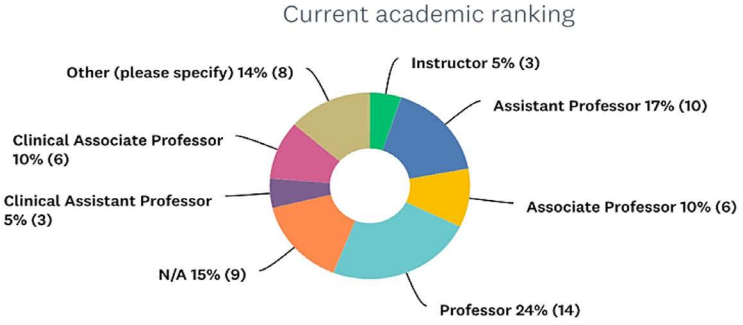
We sent a survey to female congenital interventional cardiologists around the world, participation was anonymous and voluntary. The survey was sent on March 16th, 2023, and 59 responses were collected by March 24th, 2023.

Fifty-nine female interventional cardiologists from 11 different countries completed the survey, 60% of them work in the United States. Seventy-six percent are working full time and in academic centers, 64% percent of them are married and 69% have one or more children (49% with children younger than 18 years of age, **Figure 1**).

Only 44% of the interventional cardiologists that answered the survey are satisfied with their work life balance. In contrast, for 78% of them, WLB is extremely or very important. Fifty-nine percent of the women interventionists are somewhat happy or very happy with the opportunities for advancement at their organization, when multivariate analysis was performed, the more unhappy with the advancement opportunities at their organization, the more likely to be unsatisfied with the WLB **(Figure 2)**.

Eighty percent of the surveyed answered that they think that they have to work harder than their male colleagues to accomplish the





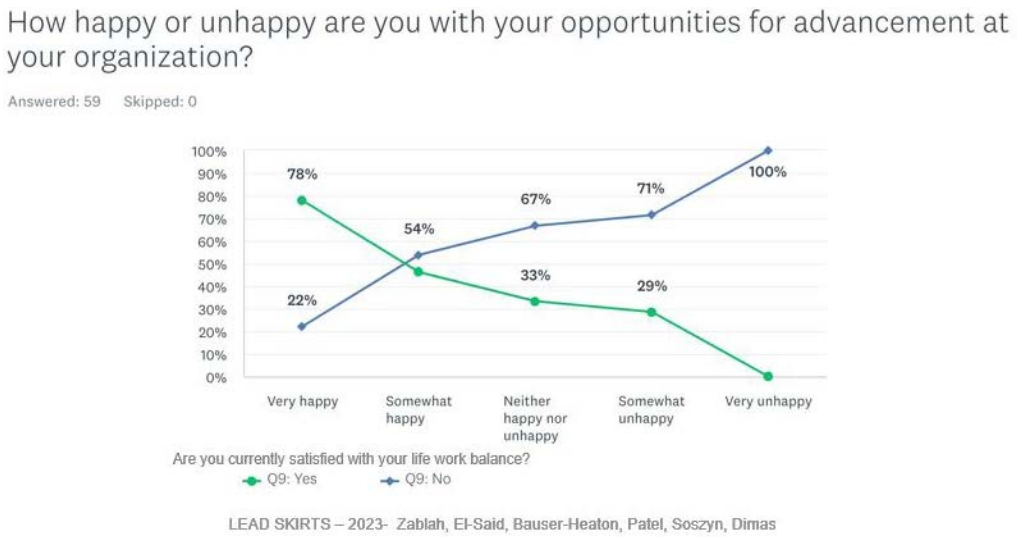


FIGURE 2

same work **(Figure 3)**. When multivariate analysis was performed, having kids younger than 18 years old, work in academic practice, and being unhappy with the opportunities for advancement at work were associated with less likely to be satisfied with the WLB. Sixty-four percent of the surveyed said that decreasing the number of hours at work would allow them to accomplish WLB, while 52% said that learning to say "no" was critical to accomplishing this goal. All of the surveyed have hobbies that include sports, music, gardening, and art.

– Jenny E. Zablah, MD

When and How to Say "No"

Cultivating a proper work-life balance is essential to success in both facets of your day, in that you are better able to devote yourself to each respective component when you understand and implement boundaries. A vital aspect of distinguishing your work and personal lives is being able to say no; when you recognize your own limits and make the sometimes-difficult decision to prioritize yourself when faced with favors, you can better commit yourself to the things you are already doing. No one wants to seem unfriendly or unwilling to help those around them, but you cannot properly contribute to society when you stretch yourself too thin.

Simply put, the quality of your work will deteriorate for the sake of quantity. Instead, it is important to realize what is feasibly possible for you to accomplish at a level you would be proud of, and to avoid mistreating yourself by crossing into the realm of the impossible. In fact, saying no can be as simple as–quite literally–saying no, apologizing, and explaining your rationale. Offering a less demanding alternative for yourself can also convey your intentions, characterizing you as responsible and thoughtful while still allowing you to maintain your work-life balance. Emphasizing that the timing of the request is difficult, rather than the nature of the question or the individual posing it, is key. In this manner, saying no can facilitate the preservation of an even proportion of professional and personal tasks in your day-to-day life.

– Priti M. Patel, MD

Keeping Our Hobbies and Our Work/ Family…is There Enough Hours in the Day?

When recently interviewed about my directorship in a specialty program, I was asked "what are your hobbies?" I had to stop and truly think – what do I do as a hobby? Certainly, childrearing doesn't seem quite hobby-like. For me, and true for many, my health had taken a backseat to nearly every activity. Patient calls? Covering for a colleague? Mystery reader at school? Balloon arch for a birthday party? All prioritized. Even if this meant staying up until late into the night to sacrifice sleep to do so. What I realized, however, is no one would make me get back to things I once loved, but rather this was my responsibility. Hobbies for me now include things that have dual meaning and purpose. To take on running and yoga as a hobby enables me to maintain my health and mental balance. Continuing to be a co-leader in Girl Scouts allows me to spend time with my eldest while camping or hiking (once two of my favorite hobbies) and sewing costumes for my children allows to me to feed the creative that once was vibrant. The musician in me is enveloped in listening to music while writing cath reports. As our children age, time for hobbies will change but at stages where their social lives are more active than a college freshman, time is precious. That being said, time for self-care in many forms is vital to being a good physician and good mother. Without allowing some freedom for wandering thoughts or physical activity to take priority, we lose that which made us unique and without carving out that protected time to do so leaves us wishing. For me, this can only be accomplished by literally scheduling time for myself on my calendar.

– Holly D. Bauser-Heaton, MD

Looking to the Future as a Woman Interventionist in Training - My Expectations of Work-Life Balance

As physicians and women in the field of interventional cardiology, we aim for excellence by trying to be "able, affable, and available" – with available often being the easiest to achieve.

We need to have the ability to juggle everyone's expectations – often leaving our own self-care to last on the checklist. There is also an unspoken expectation that as a woman you must be a "diamond-in-the-rough" able to outlast the training and come out the other side with your personality and relationships intact. So - where does that leave work-life balance? Work-life balance means something different for everyone. For me, it means meeting life and work expectations and being satisfied with the division of your time - without feeling you could or should do more. There has been a societal pivot towards awareness that maintaining work-life balance means less burn out and more satisfaction. It is important; therefore, we actively seek to maintain our external identities. Finding purpose in life and work and how best to harmonize the two is an individual process but my expectation is that it can be achieved with support and a mutual acceptance that the choice of where to draw the line between work

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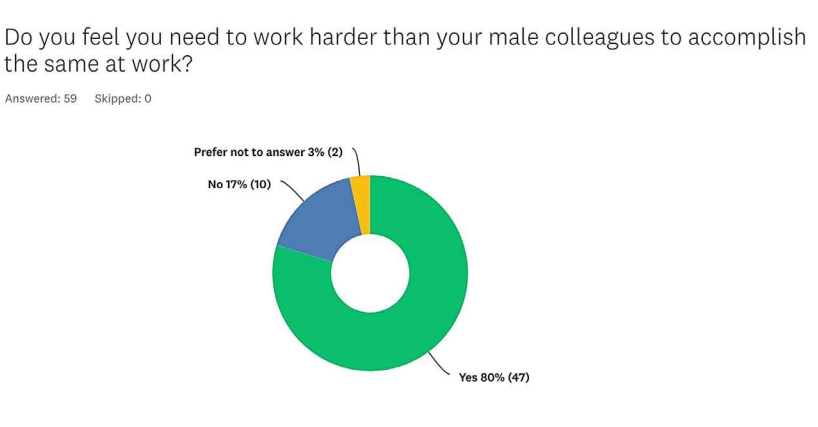

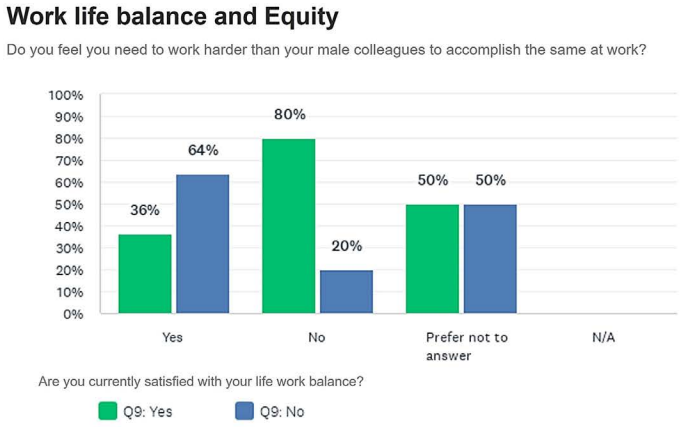

FIGURE 3

and life commitments should be that of the individual. As I am about make the transition from fellow to attending, it seems to me with more female interventionalists and more awareness that this balance matters, things are changing. It may never be perfect but enough to ensure that as women interventionalists we can not only enjoy what we do, but thrive in all aspects of our lives.

– Natalie Soszyn, MBBS

Words of Wisdom

Don't let anyone tell you can't do it all! Because you absolutely can!!! In almost every culture, women have been the cornerstone of the family. Men went out to hunt and women did EVERYTHING else! Recent research has shown that women are better multitaskers than men.[3] So, believe in yourself and believe you can do it all! As a mother of three and the director of the Cath lab, this has not been easy. But at the end of the day when all is said and done, I am happy to report that I would do it all again. I will share a moment that put it all together for me. One day I was very upset about something that happened at home and was sobbing and saying maybe if I had spent more time at home this wouldn't have happened. My son who was only 11 years old at that time came to my rescue and said: "Mom, you are a great Mom! Do you think that all the stay-at-home Mom's spend all the time with their kids, no they don't. When you are home, you're home and you show us how much you care in every way you can. So, get up and be proud!" Your kids will learn from your actions. They will grow up to be strong, motivated, witty, kind, independent and accountable and you will grow up fulfilled with your achievements both at home and at work.

– Howaida El-Said, MD

Dear Younger Self,

You will be happy to know that in the end, you were ultimately able to find career satisfaction, equality in pay and raise two beautiful children while continuing to work in a demanding male dominated field. This certainly came at a price personally and professionally, with lessons learned and mistakes made all along the way. You will be told that women "can't do it all," that if we choose to become mothers we will have to sacrifice either family or professional growth. Yes, you will have to make decisions and hard choices, but remember that the only person who has to be happy with the road you take is you. You will need help raising your children, but this will not mean that you will love them any less or that they will somehow feel neglected if you are not there every hour of every day. Remember that no one can actually have it all… not even men. For they will also sacrifice family time for work, but somehow society will accept that more. Stay at home moms will criticize you for not being home, but just being home is not a guarantee that you will always be present. If you are happy and fulfilled, you will be able to help others become happy and fulfilled, including your family. When you are at work, focus on work and when you are home, focus on home. You must create boundaries and protect them fiercely. You will find this challenging.

It will be a roller coaster until they get to school and then illnesses, school pick-ups and after school activities will cause every day to feel like a mad but glorious dash. You will need to exercise patience and grace for you, your partner, and your children. Accepting that you will not make it to everything, but that your family will ALWAYS come first. Your children and spouse will know that your love is constant even if your presence is not. As your children age, you will develop amazing relationships with them and demonstrate to them that it is, in fact, possible to work and have a family. They will tell you they understand you needed help and how much they loved their nanny. You will find your people. You need to find your people. Those men, and especially women, support you, think what you do is amazing and are willing to help in times of need. You may in return, gladly provide free medical advice and endless carpools on the weekends when you are free. Always remember that home is the one place where you cannot be replaced. Remind yourself to enjoy the crazy struggle and to be present whatever you are doing.

You will spend your days at work keeping your head down and doing good work, worrying more about taking care of your patients than recognition. Understand that ultimately recognition will come. You will endure bias that you must look within yourself to overcome. Not with emotion, but with sheer facts. Remember this when you are justifying your pay increase to equal that of your male colleagues. It will be shocking to you that discrimination remains present, but you will find colleagues who support you and together you will be successful in getting equal treatment for equal work. But it will take time, patience, and persistence. You will realize you really can do whatever you set your mind to. But, you must, surround yourself with people who share your vision and want to help you get there, and then return the gift by mentoring other women to walk in your same footsteps. Accept that no one, not even you, can do it all alone, but you can still do it fantastically.

Signed,
Your Older and Much Wiser Self

– Vivian Dimas, MD

Acknowledgments

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Illinois Tech Professors' Paper Challenging Classical View of Muscle Contraction Could Lead to New Cardiac Treatments

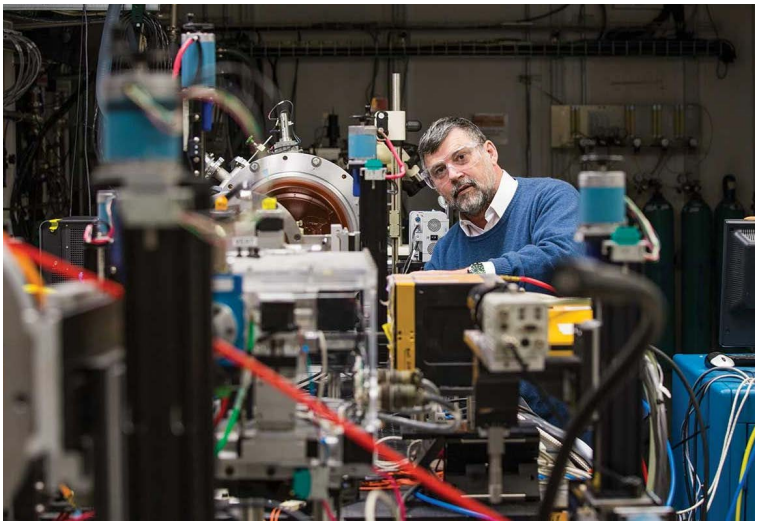
Professor Thomas Irving and Research Assistant Professor Weikang Ma, working with a team from the University of Washington, found that motor proteins have to be activated before muscles can contract

CHICAGO—February 1, 2023—In a newly published paper, a team including two professors from Illinois Institute of Technology (Illinois Tech) have reported a new mechanism for muscle contraction that could play a significant role in developing treatments for inherited cardiac conditions.

Illinois Tech Research Assistant Professor Weikang Ma and Professor of Biology and Physics Thomas Irving, working in collaboration with Professor of Bioengineering Michael Regnier's group at the University of Washington, challenged the conventional understanding of muscle contraction, which held that the relationship between the thin and thick filaments that comprise muscle tissue was relatively straightforward. Rather than myosin proteins that make up thick filaments automatically finding their way to thin filaments when their targets were activated, the researchers discovered that myosin motor proteins have to be turned on before they can generate force and contract the muscle.

By better understanding this relationship between the thick and thin filaments in regards to muscle contraction, genetic conditions that have gone untreated until now may eventually have medical remedies. The diseases can range from dilated cardiomyopathy—where the heart muscle is too weak and ultimately struggles to pump blood throughout the body—to hypertrophic cardiomyopathy, where the muscle works too hard and eventually becomes thickened, potentially reducing blood flow.

Part of what has allowed the team to make its case is the information that it gathered using a state-of-the-art technique called small-angle X-ray diffraction, a technique that Irving has been perfecting as part of the Biophysics Collaborative Access Team (BioCAT) in partnership with groups such as the National Institutes of Health and Argonne National Laboratory since the 1990s. This technique allows researchers to gather data in a way that Irving says is more accurate than other techniques currently being used.



Professor of Biology and Physics, Thomas Irving, in the lab

"In electron microscopy, you can't see motion, it's a static measurement. You can't do a time result," says Irving. "In our case, we can take movies; we can do a physiological experiment and see these molecules moving in real physiological time. That's what's so special about it."

Their paper, "Structural OFF/ON transitions of myosin in relaxed porcine myocardium predict calcium-activated force," was published January 23, 2023, in the Proceedings of the National Academy of Sciences.

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The Berlin Heart Active Driver Trial Update from ACTION and Berlin Heart, Inc.

Cincinnati, OH – March 15, 2023 – A leading pediatric heart failure organization ACTION (the Advanced Cardiac Therapies Improving Outcomes Network in Cincinnati, OH), in conjunction with Berlin Heart, Inc. (The Woodlands, TX), launched the first prospective FDA regulated device trial using a pediatric ventricular assist device (VAD) registry in November 2022.

The Berlin Heart ACTIVE Driver Trial is sponsored by Berlin Heart, Inc. ACTION is serving as the clinical research organization (CRO), with the ACTION registry serving as the repository of the clinical trial data.

The goal of the trial is to gain FDA approval of the new ACTIVE driving unit, which will replace the approved IKUS driving unit that powers the commercially available EXCOR Pediatric Ventricular Assist Device system. Of the 58 Children's Hospitals who are member sites in the ACTION Network, 17 have been chosen to participate in the trial.

Historically, there has been a paucity of pediatric device and drug trials due to cost, patient volume and variability in heart failure etiology. ACTION is collaborating with industry, patients and families and regulatory agencies to develop novel ways to bring innovation to the bedside and improve the treatment and outcomes for the pediatric population with heart disease.

The Berlin Heart ACTIVE Driver study will allow young heart failure patients implanted with a Berlin Heart VAD to rehabilitate and become stronger while awaiting heart transplantation. The currently approved IKUS driver is large, heavy and cumbersome. The new Active Driver is smaller and allows patients much greater mobility and quality of life.

As of February 3, 2023:

- Eleven of 17 sites have been activated to enroll patients.
- Seven of 17 sites have enrolled patients.
- Ten of 40 patients have enrolled in the study.

"We are thrilled to partner with Berlin Heart on this important study," said Angela Lorts, MD, MBA and Co-Executive Director of ACTION. "We are using the ACTION registry data as 'real world data' for both prospective clinical trials and to apply for expanded FDA indications for pediatrics. This study and others will continue our important work in making a difference to improve outcomes for patients with heart failure, especially children."

To learn more about the Berlin Heart ACTIVE Driver Trial, click HERE. To learn more about ACTION and how you could join its efforts, please visit www.actionlearningnetwork.org.

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https://stollery-cardiac.wixsite.com/stollery-cardiac-sym

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JULY

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