CONGENITAL CARDIOLOGY TODAY
Timely News & Information for Congenital/Structural Cardiologists & Cardiothoracic Surgeons Worldwide
International Edition
Vol. 21 - Issue 9
September 2020

Table of Contents

Surgical Placement of Adjustable PDA Band Facilitates Ductal Stenting in Stage 1 Hybrid Palliation Procedure

A Novel Technique to Help Facilitate Proper PDA Stent Placement

Sharib Gaffar, MD; Joanne P. Starr, MD; Richard N. Gates, MD; Michael R. Recto, MD

Indexing Keywords: Congenital Heart Disease, pediatric intervention, stenting technique
Disclosure: The authors have no conflicts of interest to disclose.

Abstract

Background: The Hybrid Stage I procedure is an accepted alternative for high-risk patients with Hypoplastic Left Heart Syndrome (HLHS) who face increased mortality with the Norwood procedure. The procedure involves bilateral pulmonary artery banding, atrial septostomy, and stenting of the Patent Ductus Arteriosus (PDA). Prior to the procedure, prostaglandin infusion (PGE) is required to maintain ductal patency. Most interventionalists will discontinue PGE prior to the procedure to try to modulate the caliber of the PDA. Although effective in maintaining ductal patency, dose adjustment of PGE cannot accurately predict ductal diameter which is crucial in selection of appropriate stent size. ***Case:*** We describe six patients who were maintained on PGE until just prior to placement of the adjustable PDA band. The adjustable PDA band was placed and appropriately sized after bilateral pulmonary artery banding and prior to PDA stenting. When required, the PDA band was adjusted to the desired diameter after review of the PDA angiogram prior to PDA stent implantation. ***Results:*** All patients underwent successful PDA stenting with either balloon expandable or self-expanding stents. ***Conclusion:*** Adjustable PDA banding allows for more controlled PGE infusion, decreases the likelihood of stent embolization or migration, serves as a landmark for appropriate stent implantation, and allows for creation of an ideal ductal diameter prior to stent implantation.

Introduction

The Hybrid Stage I procedure is a combined surgical and interventional catheterization procedure for high risk hypoplastic left heart (HLHS) patients where a cardiac surgeon will first place bilateral pulmonary artery bands, after which a pediatric interventional cardiologist will stent the Patent Ductus Arteriosus.[1] Depending on the size of the patent foramen ovale, balloon atrioseptostomy with or without atrial stenting is performed either during the initial procedure or at a later date.[2] Because Hypoplastic Left Heart Syndrome

SEPTEMBER 2020

International Edition Vol. 18 - Issue 9

TABLE OF CONTENTS

A HIGH STANDARD FOR DURABILITY **PROVEN BY A DECADE OF DATA**

Melody TPV was the first transcatheter valve implanted in a human anywhere in the world and is the longest-studied transcatheter valve with the largest body of clinical evidence.

That unparalleled body of clinical evidence has recently expanded, with positive 10-year durability, safety, and efficacy data from the U.S. Investigational Device Exemption (IDE) Trial.

Melody™
Transcatheter Pulmonary Valve (TPV) Therapy

Melody™ Transcatheter Pulmonary Valve | Ensemble™ II Transcatheter Valve Delivery System

Important Labeling Information for the United States

Indications: The Melody TPV is indicated for use in the management of pediatric and adult patients who have a clinical indication for intervention on a dysfunctional right ventricular outflow tract (RVOT) conduit or surgical bioprosthetic pulmonary valve that has ≥ moderate regurgitation, and/or a mean RVOT gradient ≥ 35 mm Hg. **Contraindications:** None known. **Warnings/Precautions/Side Effects:** 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.

For additional information, please refer to the Instructions for Use provided with the product or available on manuals.medtronic.com. **CAUTION:** Federal law (USA) restricts this device to sale by or on the order of a physician.

UC202104730 EN 07/2020

Medtronic
Further, Together

is a ductal-dependent lesion, prostaglandin (PGE) is required to maintain ductal patency. We have found that despite discontinuing PGE up to 12 hours prior to the hybrid procedure, the PDA remains large at the time of the operation (**Figure 1**). For this reason, surgeons at our institution have resorted to placing a segment of umbilical tape effectively mildly constricting, or "banding," around the PDA prior to PDA stenting. This PDA banding procedure helps mildly constrict the ductus temporarily, and facilitates the PDA stenting procedure by serving as a landmark for proper PDA stent implantation.

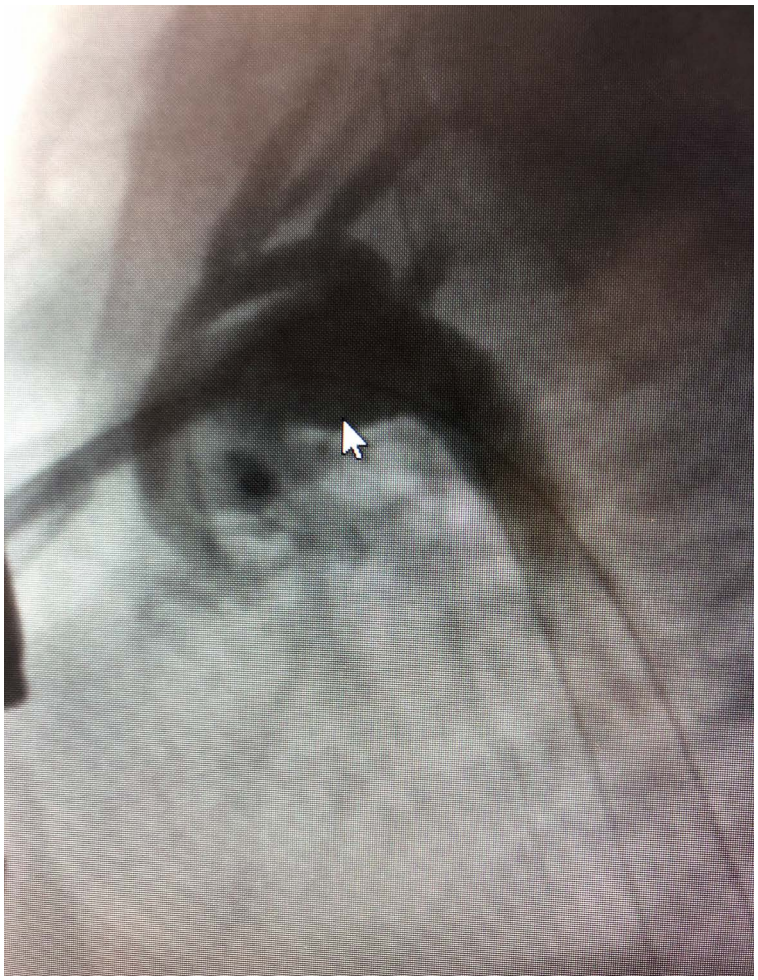


FIGURE 1 *This PDA remained large despite cessation of PGE 12 hours prior to procedure (white arrow).*

Case Series

Six patients (four males, two females), five with HLHS and one with Shone's complex and left ventricular hypoplasia, mean age eight days (range 3-16 days), mean weight 2.72kg (range 2-3.2 kg) underwent Hybrid Stage 1 palliation due to anatomic or weight concerns about the viability of the Norwood procedure. In five cases, banding of the right and left pulmonary arteries and stenting of the PDA were completed as a one-step procedure; in one case, a patient had bilateral pulmonary artery bands completed initially, followed by PDA stenting 11 days later. Umbilical tape was initially wrapped around a 6 mm Hagar and cut to size. It was then wrapped around the PDA and clipped into position utilizing two surgical clips. Following PDA banding, a 7F sheath (Cook) was positioned in the main pulmonary artery (MPA) above the pulmonary sinuses of Valsalva. An angiogram was then performed through the sheath and the PDA with ductal band in place was measured. Ideally the band is positioned in the mid-portion of the PDA with mild constriction (**Figure 2A**). In this particular case the band was too tight and required removal of one of the clips (**Figure 2B**). Conversely, if the ductus remains large, one or more additional clips can be added to mildly constrict the PDA. A follow-up angiogram is then performed. Once the ideal ductal diameter is obtained, either a Cordis Genesis (Cardinal Health, Dublin OH, USA) premounted balloon-expandable stent, or EV3 Everflex (Medtronic, Minneapolis MN USA) self-expandable stent is utilized for ductal stenting with the surgical clips holding the PDA band in place serving as a landmark (**Figure 3A**). Angiography post PDA stenting is performed to confirm proper placement of the stent (**Figure 3B**). The sheath and wire are then removed, and MPA puncture site repaired. All six patients underwent successful PDA stenting without complication. Postoperative course was uneventful.

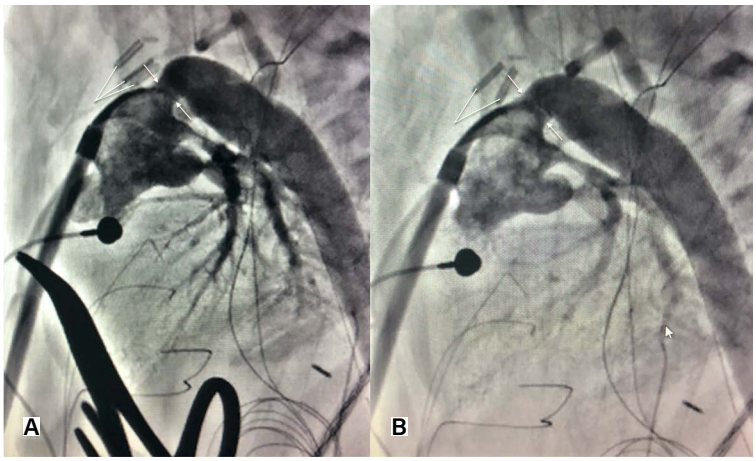


FIGURE 2A *Three hemoclips (white lines) fastening the umbilical tape around the PDA show that the ductal constriction is too tight (long white arrows).*

FIGURE 2B *Less ductal constriction with removal of one hemoclip (white lines).*

Discussion

We describe a series of six infants who underwent ductal stenting as part of Hybrid Stage I palliation. All six patients underwent placement of an adjustable PDA band to mildly and temporarily constrict the large PDA prior to PDA stent implantation. Interventional cardiologists have, for many years, utilized placement of a nasogastric tube to serve as a landmark prior to PDA stent implantation (**Figure 4A, 4B**). The successful placement of a mildly constricting PDA band further helps with PDA stent positioning by serving as an additional landmark and helps decrease the risk of stent embolization in the event that the stent is undersized and embolizes distally (**Figure 5**). This

potential complication can sometimes be rectified by placement of a second proximal stent to help anchor the initial undersized stent that has embolized distally (**Figure 5**).

One of the advantages of having the PDA band fastened with two hemoclips is that the size of the PDA can be easily adjusted by either removal of a clip, thereby loosening the band resulting in increase in PDA diameter, or tightening the band by placement of an additional hemoclip, thereby decreasing ductal diameter. We have found that utilizing a 7 French sheath to deliver the stents allows performance of angiography through the backbleed device to further check stent position prior to stent delivery and deployment. The stent, when expanded to the desired diameter, is then delivered in proper position without evidence of ductal constriction which could impede systemic output.

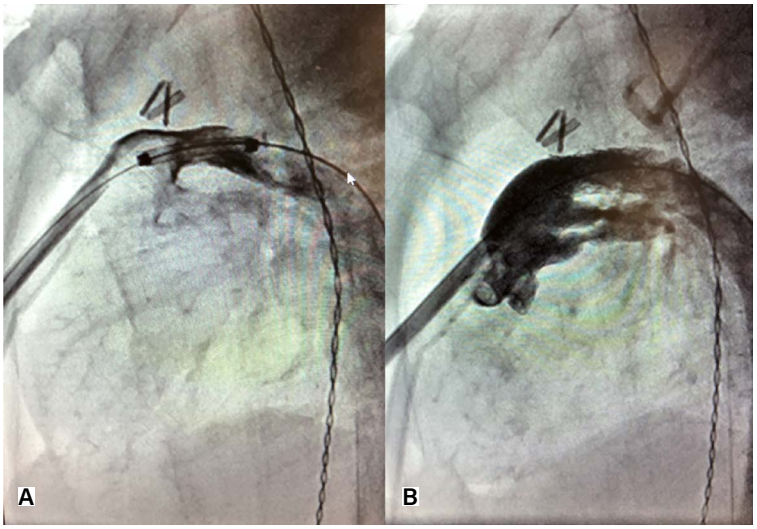


FIGURE 3A *The stent is advanced into place across the PDA.*
FIGURE 3B *Angiography performed following stent expansion into the PDA.*

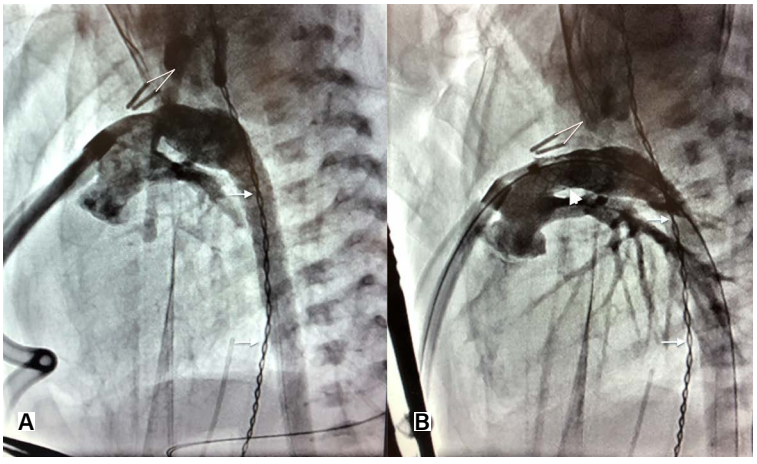


FIGURE 4A & 4B *Both the hemoclips (white lines) and nasogastric tube (white arrows) can be used together as landmarks to facilitate stent placement (large white arrowhead).*

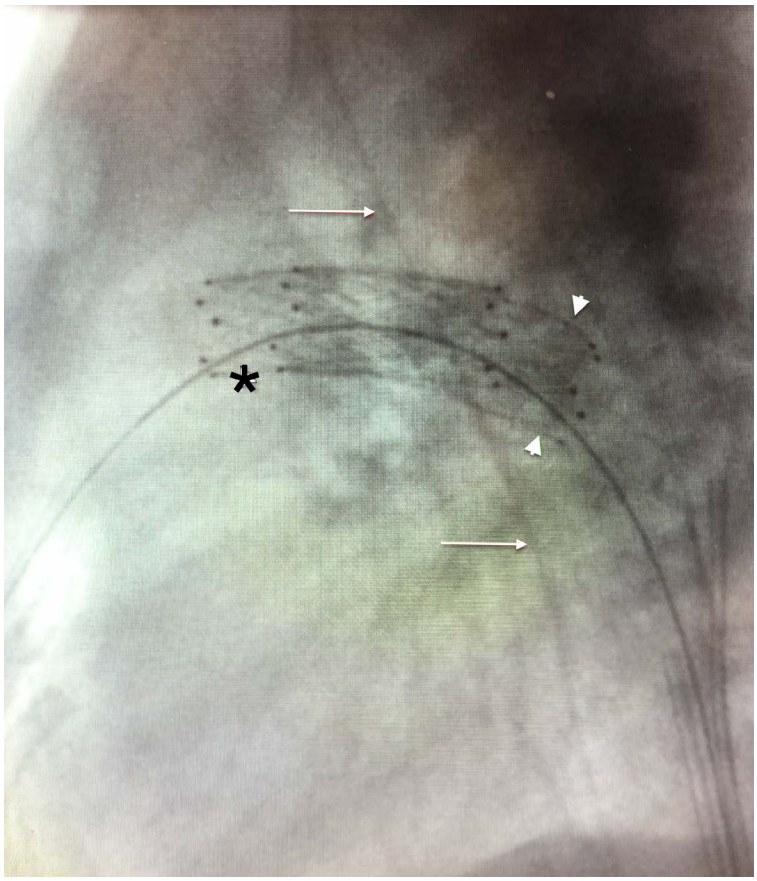


FIGURE 5 *An undersized stent (white arrowheads) embolized distally in this PDA that was not banded prior to stent placement. A second more proximal stent (asterisk) was used to anchor the first stent in place. A temperature probe (long white arrows) serves as sole landmark without PDA banding.*

Conclusion

Adjustable PDA banding with umbilical tape in a Stage 1 Hybrid procedure for HLHS and patients with left-sided obstructive lesions and left ventricular hypoplasia facilitates ductal stenting, allows for PGE infusion until the time of the procedure and decreases the likelihood of stent embolization or migration by creating a mild constriction and serving as a landmark for stent implantation. More importantly, it allows both the surgeon and interventionalist to work together to determine the ideal ductal diameter prior to PDA stent implantation.

References

1. Chen Q, Parry AJ. The current role of hybrid procedures in the stage 1 palliation of patients with hypoplastic left heart syndrome. Eur J Cardiothorac Surg 2009 Jul;36(1):77-83.
2. Pedra CA, Neves JR, Pedra SR, Ferreiro CR, Jatene I, Cortez TM, et al. New transcatheter techniques for creation or enlargement of atrial septal defects in infants with complex congenital heart disease. Catheter Cardiovasc Interv 2007 Nov 1;70(5):731-739.

SHARIB GAFFAR, MD

Pediatrics Resident, PGY-3
CHOC Children's Hospital of Orange County,
Orange, CA, USA
University of California Irvine
Irvine, CA, USA
sharibgaffar@gmail.com

JOANNE P. STARR, MD

Medical Director, Extracorporeal Life Support Program
CHOC Children's Hospital of Orange County
Orange, CA, USA

RICHARD N. GATES, MD

Surgeon-in-Chief
Co-Medical Director, Heart Institute
Medical Director, Cardiothoracic Surgery
CHOC Children's Hospital of Orange County
Orange, CA, USA

MICHAEL R. RECTO, MD

Medical Director, Cardiology
Medical Director, Cardiac Catheterization Lab
CHOC Children's Hospital of Orange County
Orange, CA, USA

OCTOBER

01

Glasgow Paediatric Echocardiography Course
Glasgow, United Kingdom
https://paediatricecho.co.uk/

05-09

30th Annual International Symposium on Adult Congenital Heart Disease
Virtual
https://www.cincyhearteducationseries.org/achdsymposium

08-09

Pulmonary Vein Stenosis Symposium 2020
Atlanta, GA
https://web.cvent.com/event/c0f8acec-3077-4fad-9329-2681bd833e65/summary

10

10th Annual Fetal Echocardiography Symposium at UCLA: State-of-the-Art 2020
Virtual
https://www.cme.ucla.edu/courses/event-description?registration_id=533268&client_view_p=f

16-17

The Unpartitioned Atrioventricular Connection: From Simple to Complex: 5th Annual Advances in Congenital Heart Disease Virtual Summit
Virtual
http://www.clevelandclinicmeded.com/live/courses/pediatricheart/

27-30

NeoHeart 2020: Cardiovascular Management of the Neonate
Virtual
https://collectedmed.com/index.php/article/article/course_preview/category/12/1025/769

28-29

38th Annual Echocardiography and Structural Heart Symposium
Virtual
http://cme.baptisthealth.net/echo/pages/index.aspx

NOVEMBER

19-20

Plenareno Webinar 2020: Heart Care and Lifestyle
Virtual
https://heart.plenareno.com/

The Fetal Care Center Offers Hope to Families of Infants with Congenital Differences.

The Fetal Care Center's 10-bed special delivery unit, which opened in 2019, is a high risk infant special delivery unit for healthy mothers whose infants will require immediate access to pediatric intensive and critical care after birth. Mother and newborn benefit from staying together in the same hospital, where the infant will have immediate access to our team of renowned specialists. The special delivery unit includes five labor and delivery rooms, five antepartum rooms, and two operating rooms.

Our fetal care nurse navigators are available 24 hours a day to serve as points of contact for our community physicians, pediatricians, perinatologists and neonatologists, and coordinate subspecialty consultations. Nicklaus Children's Hospital offers an array of diagnostic services, including fetal ultrasound, fetal MRI and fetal echocardiography, with the goal to support families in obtaining a definitive diagnosis and plan of care during a single visit.
Nicklaus Children's Hospital. For Health. For Life.

Nicklaus Children's Hospital

Fetal Care Center

Our number one priority is the safety and well-being of our patients and their families. We have implemented additional safety precautions, such as arrival screenings and physical distancing measures throughout the hospital, for everyone's safety. For more information on these enhancements, please visit nicklauschildrens.org/COVID19safety

3100 SW 62nd Ave, Miami, Florida 33155 | **1-844-55FETAL** | **nicklauschildrens.org/FetalCare**

Telemedicine Home Video Visits in Pediatric Cardiology – A Necessary Tool During the COVID-19 Pandemic, A Valuable Resource Moving Forward

Bianca Castellanos, MD & Jonathan Dayan, MD

Background

Various forms of telehealth have been utilized and reimbursed in Pediatric Cardiology worldwide since the 1980s.[1-4] While single ventricle post-Norwood Telemedicine Home Monitoring Programs have suggested a grossly positive impact on patient and provider satisfaction,[5-6] little data exists on Telemedicine Home Video Visits outside this sub-set of patients within Pediatric Cardiology.

Introduction

Telemedicine in the ambulatory pediatric cardiology population at our facility had not previously been established. In March of 2019, patients began participating in our pilot Telemedicine program, referred to in this manuscript as the Telemedicine Home Video Visit Program (THVVP) hereafter. From March 2019, until February of 2020, relevant data on these visits was analyzed in preparation for a large-scale shift in volume in response to the COVID-19 pandemic.

Objective

The purpose of this study is to present diagnosis, purpose of the visit, ages, and real-time data collected on these patients in order to better understand the needs of the patients and to establish ideal candidates for such visits during the COVID-19 pandemic.

Methods

All THVVP encounters between March 2019 and February of 2020 were included. Chart review was conducted. Diagnosis, reason for visit, age at time of visit, saved travel distance, ancillary support staff and real-time data collected is presented for review.

Results

Twenty-six encounters were reviewed. ***Diagnoses:*** Atrial Septal Defect, Ventricular Septal Defect, Patent Ductus Arteriosus, Marfan Syndrome, Ehlers-Danlos, Duchenne muscular dystrophy-associated cardiomyopathy, systemic hypertension, hypercholesterolemia, palpitations, frequent premature ventricular contractions, idiopathic ventricular tachycardia, coronary artery fistula, hypertrophic cardiomyopathy status post AICD, truncus arteriosus with complete heart block status post pacemaker, Shone's complex status post Ross-Konno, and Total Anomalous Pulmonary Venous Return (TAPVR) with stenosis and single ventricle physiology (**Chart 1**). ***Reason for visit:*** test result counseling (38.4%), medication reconciliation (38.4%), feeding problems / poor weight gain (23%), procedural consent (19.2%). A single pilot visit for a wound check was performed. Age groups: Birth to 12 months (30.8%), average five months. One year of age to 17 years of age (46%). Eighteen years of age and older (23%). Average age overall was 14 years old (**Chart 2**). Saved travel distance: Average miles saved: 98 miles. ***Total miles saved:*** 2,344. One patient saved 560.2 miles (**Chart 3**). Notable findings: The most common test result reviewed was a Holter monitor (5/26, 19.2% of total visits). Several encounters were completed in a multi-disciplinary fashion with nutrition, nursing (advanced practice and registered), hospice, and social work in attendance (8/26, 30.7%).

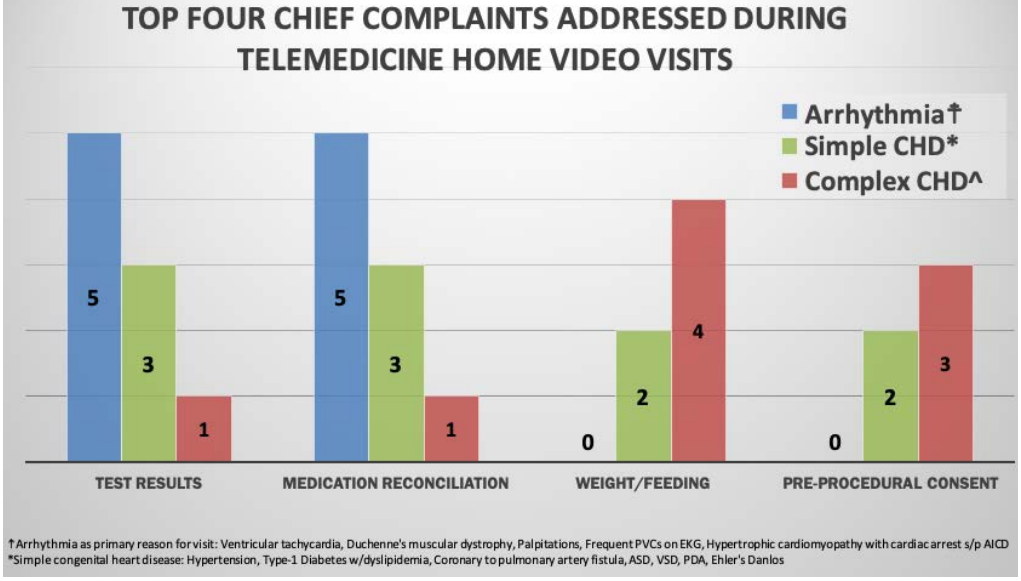


CHART 1 *Patient diagnosis were arranged in three broad categories including arrhythmias, Simple Congenital Heart Disease and Complex Congenital Heart Disease and described in greater detail in the legend at the base. The four most common reasons for the visit were included.*

Real-time data collection: Procedural consent, vital signs, and medical photos were done for at least seven of the encounters (9/26, 34.6%). ***Visit interface:*** Interface offered to providers was an iPad, designated to our division. Providers also had the option to use any secure mobile iOS device, or any secure desktop with video and audio capability and Wi-Fi. The patients used a variety of devices including their personal cell phone, desktops, and tablets, so long as they were logged into their secure medical chart application and had Wi-Fi. At our institution, our electronic medical record software is Epic. Notable complications included technical problems with video feed quality and trouble logging into the protected patient application, which was not common and was isolated to the adolescent population. See **Table 1** for a comprehensive review of patients by diagnosis.

Conclusion

In our experience, the THVVP in Pediatric Cardiology can be used safely with various diagnoses and ages. The majority of encounters were to review test results. The most common test reviewed was an ambulatory Holter monitor. Limitations include a small cohort and short-

TABLE 1 *Patients seen at our facility via the Telemedicine Home Video Visit Program (THVVP) between March 2019 until February 2020.*

Diagnosis[1,^]	Chief Complaint[2]	Age	Miles Saved[3]	Multidisciplinary Staff	Real-time Data	Comments
Palpitations (2)	Test result counseling (2)	12 years (2)	12.7, 25.1			Holter (2). One patient had a family emergency but were able to complete the encounter safely from their mobile phone.
Frequent PVCs (2)	Test result counseling, medication reconciliation	11 years, 16 years	23, 16.8			Holter (2). One patient required sub-specialty referral
Idiopathic VT	Medication reconciliation	17 years	210			
HCM status-post AICD	Wound check	15 years	25.3		Photo of wound	Sub-optimal image quality
Duchenne's[+] (3)	Test result counseling (2), medication reconciliation	21 years, 21 years, 27 years	20.7, 8.7, 29.1			Holter (2). For one encounter, the provider used home computer while out of the office on vacation.
Dyslipidemia	Test result counseling	19 years	53		Patient education material	Patient obtained screenshot of dietary recommendations
Ehlers-Danlos	Medication reconciliation	20 years	395.7			Away at college
Essential hypertension	Blood pressure check	19 years	58.4	Dietician	Blood pressure, weight	Home blood pressure monitor and scale
Coronary artery to pulmonary artery fistula	Test result counseling	10 years	7.2			Patient caregiver completed the encounter on mobile while running errands
ASD	Test result counseling	6 years	560.2			Patient lived out of state
PDA	Pre-procedural consent and counseling	16 months	0.5	Advanced nurse practitioner	Procedural consent	
VSD (3)	Feeding problems / poor weight gain (2), pre-procedural consent and counseling	6 weeks, 4 months, 10 years	13.5, 12.2, 304			Feeds adjusted (2), medications adjusted
Truncus arteriosus status post pacemaker (3)	Feeding problems / poor weight gain (2), test result counseling	6 months, 7 months, 8 months	450.6	Dietician (2)	Weight, pulse oximetry, chest x-ray results	
Shone's Complex status-sost Ross-Konno (2)	Pre-procedural consent and counseling (2)	8 years (2)	533	Social worker (2), advanced practice nurse (2)	Procedural consent (2)	Social worker in their office with patient and caregiver, advanced practice nurse in clinic with provider
Marfan Syndrome, infantile	Feeding problems / poor weight gain	7 months	14.9	Dietician		
TAPVR w/ stenosis and single ventricle physiology (2)	Medication reconciliation, pre-procedural consent and counseling	3 months, 4 months	83.8	Dietician, Hospice	Photo of patient	Home hospice

[1](n) indicates number of encounters per diagnosis, ^diagnosis separated into three broad categories with the first five entries related to arrhythmias, the following seven described as Simple Congenital Heart Disease, and the last four described as Complex Congenital Heart Disease. [2](n) indicates number of encounters per chief complaint. [3]Value combined if from the same household. PVCs = Premature Ventricular Contractions, VT = Ventricular Tachycardia, HCM = Hypertrophic Cardiomyopathy, AICD = Automatic Implantable Cardioverter-Defibrillator, ASD = Atrial Septal Defect, PDA = Patent Ductus Arteriosus, VSD = Ventricular Septal Defect, TAPVR = Total Anomalous Pulmonary Venous Return.

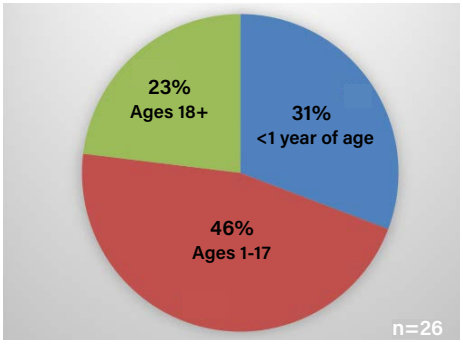


CHART 2 ***Patients participating in the THVVP by age group.*** *Graphic depiction of age distribution as a percentage of the entire cohort.*

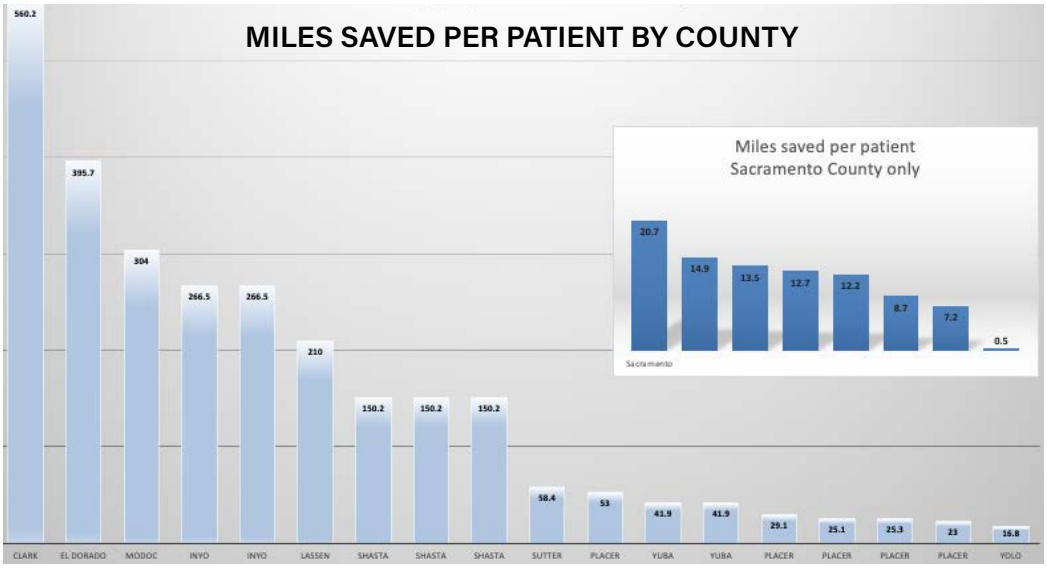


CHART 3 *The miles saved by patients by virtual encounters. Presented as total miles from their home address to the Sacramento, California main campus. Larger graph represents miles saved for counties outside of Sacramento County. Smaller, embedded graph represents only Sacramento County.*

term observation period. THVVP visits for wound checks are deemed uncertain at this time due to poor resolution of image between devices; visits of this nature will likely continue depending on family reliability and type of device used. The results of our observations suggest future studies should include special attention to arrhythmia patients. Existing data suggests expanding telemedicine care to include application-based diagnostic tools available on smart phones, such as electrocardiographic tracings, are accurate and may be a useful consideration moving forward. Indeed, the SPEAR Trial, which reported the accuracy of smart phone electrocardiogram and ambulatory rhythm monitoring as determined by 238 tracings, of which 96% of the tracings with sinus rhythm, sinus tachycardia, supraventricular tachycardia and atrial fibrillation, were deemed of diagnostic quality.[7] Importantly, family surveys to assess value of mileage saved, quality of visit and level of concerns addressed in this format compared to traditional visits should be included in future studies.

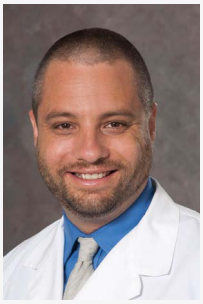
Lastly, a mention of the COVID-19 pandemic and its impact on the need for remote medical care is warranted. Our center was fortunate enough to set some groundwork in preparation for the COVID-19 pandemic. The dramatic increase in the THVVP use, however, was not anticipated previously. Pending publication within our center cites the Pediatric Ambulatory THVVP use increase from approximately 1.2% during the period between March 2019 until the March 2020 shelter in place order for the state of California, to 41.8% in the three months following. This sharp increase in use, and need, for the THVVP has shed some light on the broad applications of this tool in the comprehensive care of our cardiac patients, which was made possible for patients far and near while keeping them safely and comfortably in their homes.

References

1. Phillips A, Sable CA, Waggaman C, Harahsheh AS. 2019. Abstract 10150: Direct-to Consumer Cardiology Telemedicine: A Single Large Academic Pediatric Center Experience. Circulation. 140:A10150.
2. Casey F, Brown D, Corrigan N, Craig BG, Quinn M, McCord B, Rogers J, Mulholland HC. 1998. Value of a low-cost telemedicine link in the remote echocardiographic diagnosis of congenital heart defects. J Telemed Telecare. 4 Suppl 1:46-8.
3. Castela E, Ramalheiro G, Pires A, Carreira LM, Santos I, Costa H, Mota A, Ribeiro L. 2005. Five years of teleconsultation: experience of the Cardiology Department of Coimbra Pediatric Hospital. Rev Port Cardiol. 24(6):835-40.
4. Cloutier A. 2000. Distance diagnosis in pediatric cardiology: a model for telemedicine implementation. Telemed Today. 8(3):20-1.
5. Harahsheh AS, Horn LA, Clauss SB, Cross RR, Curtis AR, Steury RD, Mitchell SJ, Martin GR. 2016. The Impact of a Designated Cardiology Team Involving Telemedicine Home Monitoring on the Care of Children with Single-Ventricle Physiology After Norwood Palliation. Pediatr Cardiol. 37(5):899-912.
6. Morgan GJ, Craig B, Grant B, Sands A, Doherty N, Casey F. 2008. Home videoconferencing for patients with severe congenital heart disease following discharge. Congenit Heart Dis. 3(5):317-24.
7. Nguyen HH, Van Hare GF, Rudokas M, Bowman T, Silva JNA. 2015. SPEAR Trial: Smartphone Pediatric ElectrocARdiogram Trial. PLoS One. 10(8):e0136256.

BIANCA CASTELLANOS, MD

Associate Physician
The University of California – Davis
Children's Hospital, The Pediatric Heart Center
Sacramento, CA, USA
416.813.1500
Bianca.castellanos@sickkids.ca



JONATHAN DAYAN, MD

Assistant Professor
The University of California – Davis,
Children's Hospital, The Pediatric Heart Center
Sacramento, CA, USA

Interventional Systems
B|BRAUN
SHARING EXPERTISE

NuMED

Nano4Imaging
YOUR HEALTH - OUR VISION

pfmmedical

MAKING A DIFFERENCE

NuDEL®
CP Stent® Delivery System

EmeryGlide™
MR Conditional Guidewire

Nit-Occlud® PDA
Coil System For PDA Closure

NuDEL™ Indications for Use:
The NuDEL is indicated for use in the treatment of native and/or recurrent coarctation of the aorta involving the aortic isthmus or first segment of the descending aorta where there is adequate size and patency of at least one femoral artery associated with one or more of the following: acute or chronic wall injury; nearly atretic descending aorta of 3 mm or less in diameter; a non-compliant stenotic aortic segment found on pre-stent balloon dilation; a genetic or congenital syndrome associated with aortic wall weakening or ascending aortic aneurysm.

The NuDEL is indicated for use in the treatment of right ventricle to pulmonary artery (right ventricular outflow tract) conduit disruptions that are identified during conduit pre-dilatation procedures performed in preparation for transcatheter pulmonary valve replacement.

Caution: Federal (USA) Law restricts this device to sale by or on the order of a physician. **Contraindications:** Clinical or biological signs of infection. Active endocarditis. Pregnancy. **Contraindications (CoA only):** Patients too small to allow safe delivery of the stent without compromise to the systemic artery used for delivery. Unfavorable aortic anatomy that does not dilate with high pressure balloon angioplasty. Curved vasculature. Occlusion or obstruction of systemic artery precluding delivery or the stent. Known allergy to aspirin, other antiplatelet agents, or heparin. **Contraindications (RVOT only):** Patients too small to allow safe delivery of the stent without injury to a systemic vein or to the right side of the heart. **Warnings / Precautions:** Administer appropriate anticoagulation therapy to reduce potential thrombosis. If the patient is not appropriately anticoagulated, thrombus formation may occur. The sheath must be flushed with heparinized saline via the proximal side port prior to introducing the delivery system into the body. The inflated diameter of the stent should at least equal the diameter of the intended implant site. Excessive handling and manipulation of the covering while crimping the stent may cause the covering to tear off of the stent. Retracting the covered stent back into the sheath may cause the covering to catch and tear off of the stent. Do not exceed the RBP. An inflation device with pressure gauge is recommended to monitor pressure. Pressure in excess of the RBP can cause balloon rupture and potential inability to withdraw the catheter into the sheath. Confirm that the distal end of the introducer sheath is at least 2.5cm back from the most proximal image band before inflating the outer balloon. Failure to do so may stretch the outer tubing and severely hinder balloon deflation. Exercise caution when handling the stent to prevent breakage. The NuDEL system, especially at the stent, is rigid and may make negotiation through vessels difficult. The inflation diameter of the balloon used during stent delivery should approximate the diameter of the obstructive vessel and the intended implant site. If resistance is encountered upon removal, the whole system (balloon, guidewire and sheath) should be removed as a single unit, particularly if balloon rupture or leakage is known or suspected. **Warnings / Precautions (CoA only):** Coarctation of the aorta involving the aortic isthmus or first segment of the descending aorta should be confirmed by diagnostic imaging. The NuMED CP Stent has not been evaluated in patients weighing less than 20kg. The platinum/iridium stent may migrate from the site of the implant. As with any type of implant, infection secondary to contamination of the stent may lead to aortitis, or abscess. Over-stretching of the artery may result in rupture or aneurysm formation. **Warnings / Precautions (RVOT only):** During the Premarket Approval study the Medtronic Melody valve was used for valve restoration. The safety and effectiveness of the Covered CP Stent for pre-stenting of the right ventricular outflow tract (RVOT) landing zone (i.e. prophylaxis or prevention of either RVOT conduit rupture or TPVR fracture; use as a primary RVOT conduit) in preparation of a transcatheter pulmonary valve replacement (TPVR) has not been evaluated. As with any type of implant, infection secondary to contamination of the stent might lead to endocarditis, or abscess formation. The Covered Stent can migrate from the site of implant potentially causing obstruction to pulmonary artery flow. Over-stretching of the RVOT may result in rupture or aneurysm of the RV-PA conduit or the native pulmonary artery.

Nit-Occlud® Indications for Use:
The Nit-Occlud® PDA coil is a permanently implanted prosthesis indicated for percutaneous, transcatheter closure of small to moderate size patent ductus arteriosus with a minimum angiographic diameter less than 4mm.

Nit-Occlud® Brief Statement:
Do not implant the Nit-Occlud PDA into patients who have endocarditis, endarteritis, active infection, pulmonary hypertension (calculated PVR greater than 5 Wood Units), thrombus in a blood vessel through which access to the PDA must be obtained, thrombus in the vicinity of the implantation site at the time of the implantation or patients with a body weight < 11 lbs. (5kg). An angiogram must be performed prior to implantation for measuring length and diameter of the PDA. Only the pfm medical implantation delivery catheter should be used to implant the device. Administration of 50 units of heparin per kg bodyweight should be injected after femoral sheaths are placed. Antibiotics should be given before (1 dose) and after implantation (2 doses) to prevent infection during the implant procedure. Do not implant the Nit-Occlud PDA in an MR environment. Do not pull the Nit-Occlud coil through heart valves or ventricular chambers. Contrast media should not be injected through the implantation catheter. The catheter must not be connected to high pressure injectors. Patients may have an allergic response to this device due to small amounts of nickel that has been shown to be released from the device in very small amounts. If the patient experiences allergic symptoms, such as difficulty in breathing or swelling of the face or throat, he/she should be instructed to seek medical assistance immediately. Antibiotic prophylaxis should be performed to prevent infective endocarditis during first 6 months after coil implantation. Potential Adverse Events: Air embolism, Allergic reaction to drug/contrast, Apnea, Arrhythmia requiring medical treatment or pacing, Arteriovenous fistula, Bacterial endocarditis, Blood loss requiring transfusion, Chest pain, Damage to the tricuspid or pulmonary valves, Death, Embolization of the occluder, requiring percutaneous or surgical intervention, Endarteritis, False aneurysm of the femoral artery, Fever, Headache/ Migraine, Heart failure, Hemolysis after implantation of the occluder, Hypertension, Hypotension or shock, Infection, Myocardial infarction, Occluder fracture or damage, Perforation of the heart or blood vessels, Stenosis of the left pulmonary artery or descending thoracic aorta, Stroke/TIA, Thromboembolism (cerebral or pulmonary), Valvular Regurgitation, Vessel damage at the site of groin puncture (loss of pulse, hematoma etc.)

Refer to the IFUs for a complete listing of indications, contraindications, warnings and precautions. www.bisusa.org

Rx only CV-9112 Rev. 8/19

Distributed by:
B. Braun Interventional Systems Inc. | Part of the B. Braun Group of Companies
824 Twelfth Avenue | Bethlehem, PA 18018 | USA
Tel 877-836-2228 | Fax 610-849-1334 | www.bisusa.org

Excellence in pediatric care, delivered with heart

Nationally ranked specialties. World-renowned team. Collaborative approach.

BEST CHILDREN'S HOSPITALS U.S.News & WORLD REPORT CARDIOLOGY & HEART SURGERY 2020-21

Nicklaus Children's pediatric specialty programs have time and again been ranked among the best in the nation according to *U.S.News & World Report*. Established in January 2015 as Pediatric Specialists of America and rebranded in January 2020, **Nicklaus Children's Pediatric Specialists** is the multispecialty medical group practice of Nicklaus Children's Health System with a regional, national and international presence in providing pediatric-centric care through a collaborative team approach, excellence in clinical care, education and research.

The Heart Program at Nicklaus Children's serves as a beacon to families confronting the reality of a child or newborn with a heart condition. The program offers the most innovative, least invasive approaches to the treatment of congenital heart disease, including many first-in-the-world procedures that were pioneered by our own internationally renowned team.

Nationally ranked specialties. World-renowned team. That's Nicklaus Children's Pediatric Specialists.

Nicklaus Children's Pediatric Specialists

(Formerly Pediatric Specialists of America)

nicklauschildrens.org/Heart

Our number one priority is the safety and well-being of our patients and their families. Because of this, we have implemented additional safety precautions, such as arrival screenings and physical distancing measures in waiting rooms. For parents who wish to make arrangements for their child to see their specialty physician virtually, telehealth options may also be available. For more information on our safety enhancements visit nicklauschildrens.org/covid19safety

An Expandable Heart Valve May Reduce Number of Pediatric Open-Heart Surgeries

Someday, this Prosthetic Heart Valve might be the Only One a Child Needs

More than 330,000 children worldwide are born with a heart valve defect, and millions of others develop rheumatic heart disease requiring early valve replacement. Current prosthetic heart valves are fixed in size, so typically need to be replaced every few years as a child grows. For children receiving their first replacement before age two, that means as many as five high-risk open-heart operations before they reach adulthood.

But a surprising new design created at Boston Children's Hospital could allow children to keep the same prosthetic valve until adulthood. The research team, led by Pedro J. del Nido, MD, **http://www.childrenshospital.org/directory/physicians/d/pedro-delnido**, Chairman of Cardiovascular Surgery at Boston Children's Hospital, envision patients having the valve expanded through a minimally invasive balloon catheter procedure as needed.

As described in Science Translational Medicine, **https://stm.sciencemag.org/content/12/531/eaay4006**, the valve underwent extensive benchtop studies, computer simulations, and testing in large animal models. All showed that the new design enables the valve to work across a wide range of sizes, retaining its functionality.

"We hope to bring this new device into clinical testing fairly rapidly," says del Nido, the paper's senior author. "If our preclinical results hold up in human testing, this could transform the field."

Less is More: A Bileaflet Heart Valve

Commercially available prosthetic heart valves have three leaflets, tiny flaps that provide a one-way inlet or outlet for blood to keep it flowing in the right direction. The new design was inspired by human venous valves, found in the deep veins of the leg. Unlike our hearts' native outflow valves, venous valves have just two leaflets. Nature has optimized their geometry to maintain closure and one-way blood flow even when the veins expand in diameter.

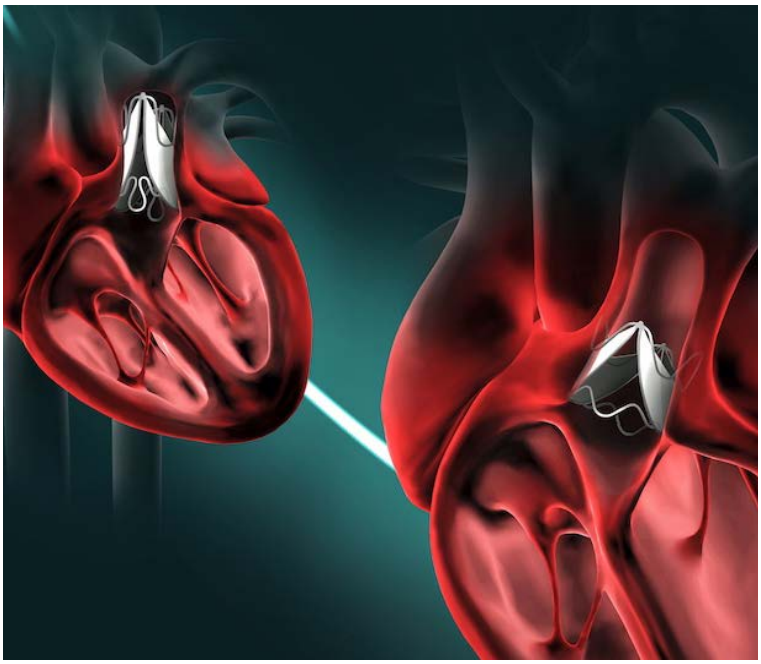
"Veins carry approximately 70 percent of our blood volume, and their dimensions can change dramatically depending on body position," explains Sophie Hofferberth, MD, a surgical resident at Brigham and Women's Hospital who led the research in del Nido's Boston Children's lab. "We mimicked the geometric profile of the human venous valve to design a bileaflet heart valve of programmed dimensions that is adaptable to growth without loss of one-way flow control."

In multiple rounds of testing, valve prototypes were able to expand to accommodate growth and structural asymmetries within the heart. They remained fully functional across a range of dimensions, pressures, and flow rates.

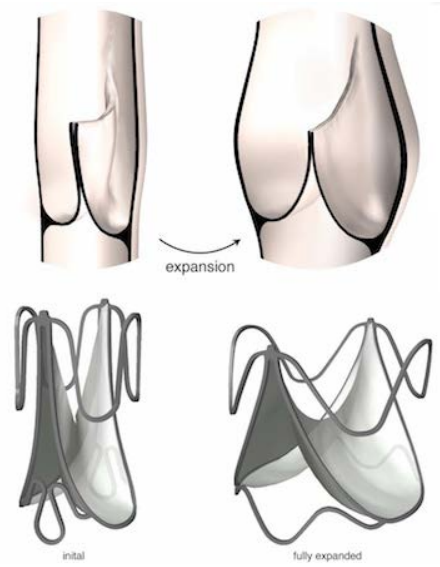
Because the valve can expand without requiring the frame and leaflet to stretch or enlarge, it is compatible with a range of off-the-shelf materials, the researchers say. The researchers successfully and safely expanded the device at multiple timepoints in a growing sheep model, using a balloon catheter.

Potential for Fewer Blood Clots

The researchers also observed that the bileaflet heart valve design encourages good blood flow through the valve. This could potentially reduce the risk for blood clots, a complication often seen with existing prosthetic valves. The team saw no evidence of blood clot formation in the growing sheep model over 10 weeks of observation, even without the use of blood-thinning medication typically given to prosthetic valve recipients.



A breakthrough design could spare children from repeated operations to replace outgrown valves and could also benefit adults with valve defects. At left, the valve in its unexpanded state; at right, its expanded configuration in a larger heart. (Sophie Hofferberth, Boston Children's Hospital; Lara Tomholt and James Weaver, Wyss Institute for Biologically Inspired Engineering)



At top, human venous valve geometry at rest and during expansion. At bottom, an expandable bileaflet heart valve inspired by the venous valve. The two-leaflet design enables the heart valve to adapt to growth, unlike existing three-leaflet valve prostheses. (Sophie Hofferberth, Boston Children's; Lara Tomholt, Wyss Institute; and Matheus C. Fernandes, John A. Paulson School of Engineering and Applied Sciences, Harvard University)

"Flow disruptions that lead to blood clot formation and early valve deterioration are a shortcoming of many existing devices," says Hofferberth, who is first author on the paper. "Our design achieves a favorable flow profile that seems to facilitate effective valve washout and minimize flow stagnation, which is likely to be an important determinant of long-term device durability."

The research team believes their data support initiation of a clinical study within one to two years.

Mossab Saeed, Christopher Payne, Karl Price, and Peter Hammer of Boston Children's Department of Cardiac Surgery were coauthors on the paper, together with Lara Tomholt, Matheus Fernandes, and James Weaver of the Wyss Institute for Biologically Inspired Engineering; Gerald Marx, Jesse Esch, and David Brown of Boston Children's Department of Cardiology; Jonathan Brown and Elazer Edelman of MIT; and Richard W. Bianco of the University of Minnesota. The study was supported by a NIH-NRSA postdoctoral fellowship grant (1F32HL138993-01), an Early Career Award from the Thrasher Research Fund, and the Oakwood Foundation. A provisional patent has been filed naming several of the authors.

American Academy of Pediatrics Releases First Policy Statement on Organ Transplants for Children with Intellectual, Developmental Disabilities

(COLUMBUS, Ohio) – *The American Academy of Pediatrics (AAP)* has released its first policy statement addressing children with intellectual and developmental disabilities as organ transplantation recipients.

The policy statement, published online in *Pediatrics*, recommends that transplant teams consider both the cognitive and adaptive skills of a patient when determining if a transplant could be of benefit, and recognize that children without disabilities are no more deserving of organ transplants than children with disabilities. The policy states that it is unethical to deny organ transplants to individuals with disabilities and may constitute illegal and unjustified discrimination.

Often, children with intellectual and developmental disabilities are not considered for organ transplants. A perceived lower quality of life compared to peers without disabilities is frequently cited as a reason for denial. Because children with disabilities can be organ donors, the policy states it would be unfair to exclude that patient population as recipients of organ transplants.

"An individual's quality of life is unique and multifaceted, and it is known that those with disabilities rate their own quality of life similarly to their peers without disabilities," said Garey H. Noritz, MD, Section and Division Chief of the Complex Care Program at Nationwide Children's Hospital and co-author of the policy statement. "This also applies to patients post-transplant: recipients with disabilities have reported improved quality of life, despite the potential challenges that surgery, immunosuppression and other therapies pose."

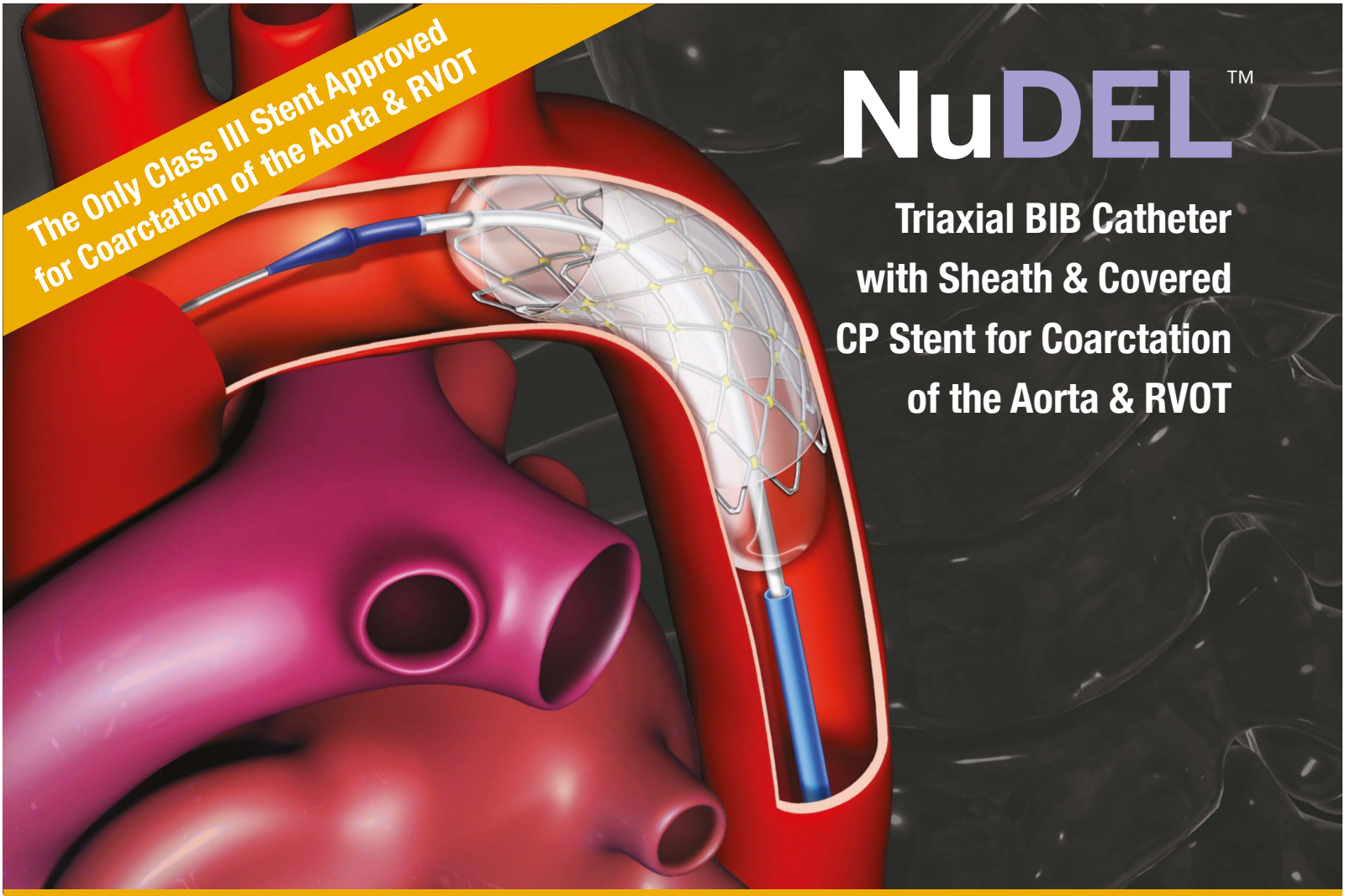
According to the policy statement, *AAP* recommendations include:

Patients should not be excluded from consideration for solid organ transplant solely based on an intellectual or developmental disability. Delaying transplantation to individuals with disabilities on the basis of a perceived lower quality of life may constitute discrimination.

Transplantation programs should standardize the definition and assessment of intellectual disability so transplant decisions can be individualized, equitable and transparent. Transplant teams should consider both the cognitive and adaptive skills of the individual.

Transplant evaluations are collaborative, should occur in person, and should include caregivers such as therapists and developmental specialists who can describe the patient's degree of function. Evaluations for transplantation to an individual with a disability should include professionals with expertise in the evaluation and management of individuals with disabilities.

"This statement is extremely important, as exclusion of these children from transplant consideration needs to end," said Dr. Noritz. "Individuals with disabilities are just as deserving of any life-saving treatment as those without disabilities, and organ transplants are no exception."



NuDEL™

Triaxial BIB Catheter with Sheath & Covered CP Stent for Coarctation of the Aorta & RVOT

The All-In-One Stent System

The NuDEL™ Stent Delivery System is designed for the efficient and effective treatment of Coarctation of the Aorta and RVOT Conduit Disruption.

The NuDEL™ includes a triaxial balloon in balloon designed catheter with a pre-mounted Covered CP Stent™, which is then covered by a sheath as an all-in-one system. Combining the proven technologies of the NuMED BIB® balloon catheter and the Covered CP Stent™, the NuDEL™ System employs both a compact delivery method and the "zig" pattern stent design.

The NuDEL™ System is available worldwide. Contact us or your local distributor to place an order.

NuMED
For Children
World Leader in Pediatric Cardiology

This device is subject to individual country regulations regarding the importation and sale of this product.

NuMED | 2880 Main Street | Hopkinton, NY 12965 USA
Tel: 315.328.4491 | Fax: 315.328.4941 | www.numedforchildren.com

CE 1639

Minneapolis Heart Institute Foundation® Welcomes Dr. Vinayak Bapat to the Valve Science Center Research Team

The Minneapolis Heart Institute Foundation® (MHIF) announced today that Vinayak (Vinnie) Bapat, MD, is joining the Valve Science Center team, bringing his extensive expertise as a cardiothoracic surgeon and innovator in treatment of heart valve disease. He will also be working directly with patients as a cardiothoracic surgeon at the Minneapolis Heart Institute®. Most recently, Dr. Bapat was a cardiac surgeon at New York Presbyterian Hospital and professor of cardiothoracic surgery at Columbia University Medical Center, both in New York. Dr. Bapat also previously served as professor of cardiothoracic surgery at Guys and St. Thomas' Hospital, London and continues to hold that position as visiting professor.

"Our team is excited to welcome a leader and innovator in the world of heart valve disease to our team at the Valve Science Center," said Paul Sorajja, MD, Roger L. and Lynn C. Headrick Family Chair for Valve Science Research. "Dr. Bapat brings a depth of experience and passion for helping patients overcome life-threatening valvular disease. We look forward to engaging his leadership and drive for innovation as we continue to pursue important research and advances on behalf of our patients and the hope they can treat their valve disease and return to full life."

"I look forward to joining the MHIF Valve Science Center team and contributing to new research and advances for heart valve disease patients, knowing there is still much opportunity to understand and improve the technologies and treatment approaches," said Dr. Bapat. "I am joining a talented research team with the expertise to remain at the forefront of research for valvular disease. My vision is to work collaboratively across imaging and cardiology research to bring new solutions that can, ultimately, improve life for even more patients with access to the right technologies."

Dr. Bapat grew up in Mumbai, India and learned his passion for medicine at a young age after taking his father to the cardiology clinic for heart failure management each week. His education and training began at King Edward Memorial Hospital and University of Bombay where he earned Bachelor of Medicine and Bachelor of Surgeon degrees, followed by a Master of Surgery degree and eventually a Master of Cardiothoracic Surgery degree. He completed his Fellowship of the Royal Colleges of Surgeons (FRCS) in cardiothoracic surgery at the Royal College of Surgeons in Edinburgh, United Kingdom. He also completed an internship, three residencies and two fellowships at various institutions, including King Edward Memorial Hospital and University of Mumbai, Toronto General Hospital in Canada, and as part of rotations at institutions in the United Kingdom, including Guys and St. Thomas' Hospital NHS Foundation Trust, Kings College Hospital NHS Foundation Trust, and Royal County Sussex Hospital, Brighton.

In addition to his clinical accomplishments, Dr. Bapat has been a collaborator among his peers in the world of transcatheter valves through the creation of two apps that are downloadable on mobile devices – Valve-in-Valve (VIV) Mitral and VIV Aortic. New transcatheter valve systems are the latest, minimally invasive technologies that have changed the way heart valve disease is treated. The apps provide imaging and clinical information, such as dimensions and characteristics of the various valve repair technologies and have been downloaded in 132 countries. Through these apps, physicians access data and clinical insights

Minneapolis Heart Institute Foundation®

Creating a world without heart and vascular disease

to determine the technology options based on their individual patient needs, including guidance and insights on the implant procedure.

"In medicine, there are unknowns and new challenges and not everything works for every patient," said Dr. Bapat. "When I think about my experience, if I decide to use one treatment and it doesn't work, I always have a plan B or plan C. It is part of my passion to continue the research that is needed to make sure we always have the additional options we need to treat more patients successfully."

"We are proud to welcome Dr. Bapat, a recognized leader in valvular disease, to the MHIF team," said Scott Sharkey, MHIF chief medical officer. "The Valve Science Center, under the leadership of Dr. Sorajja, has distinguished itself with internationally acclaimed breakthroughs in heart valve disease research, including first-in-human and first-in-world procedures to repair or replace damaged heart valves. With the arrival of Dr. Bapat, we look forward to further acceleration of this research as we strengthen our commitment to creating a world without heart and vascular disease."

CONGENITAL CARDIOLOGY TODAY

CORPORATE OFFICE

11500 Elk Horn Drive
Clarksburg, MD 20871 USA

CORPORATE TEAM

EDITORIAL BOARD

OFFICIAL PUBLICATION OF
THE CHiP NETWORK

ISSN 1554-7787 print. ISSN 1554-0499 electronic.
Published monthly.