Large Coronary Artery Fistula in an Adult: Rarity, Complexity, and Uncertainty

Rose Tompkins, MD, FACC and Evan M. Zahn, MD, FACC, FSCAI

Introduction

Coronary Artery Fistula (CAF) is a rare connection between a coronary artery and a chamber of the heart or major thoracic blood vessel without an intervening capillary bed. Most CAFs are congenital, can involve one or more coronary arteries, and have a similar prevalence in males and females1. The true incidence of CAFs is difficult to determine as many are small and likely never diagnosed or only discovered incidentally on imaging studies obtained for other reasons. CAFs have been seen in 0.3% of patients with Congenital Heart Disease2, 0.06% of children undergoing echocardiography3, 0.09-0.22% of adults undergoing coronary angiography1, 4-6, and 0.09-0.90% of adults undergoing Coronary Computed Tomography Angiography7-10. There is considerable anatomic and hemodynamic variability observed among CAFs. A majority of CAFs seen in adults are smaller and clinically silent. However, a smaller subset will present with larger, symptomatic fistulas attributed to a significant arteriovenous shunt or myocardial ischemia from a coronary steal phenomenon11. We present the case of a large CAF as an unexpected etiology of heart failure in an older adult and review the unique management challenges encountered in these rare cases.
For over 25 years, B. Braun Interventional Systems has been and continues to be a trusted industry leader in providing high quality valvuloplasty brands. Z-MED and Z-MED II are proven brands with excellent product features for dependable performance and procedural efficiency.

- Rapid inflation/deflation times maximize reperfusion and minimize procedure time
- Short balloon tapers and distal tip for optimal positioning within the valve
- Low profile design provides consistent deliverability and retractability

Z-MED™ and Z-MED II™
Balloon Aortic and Pulmonic Valvuloplasty Catheters

Distributed by:
B. Braun Interventional Systems Inc. | Part of the B. Braun Group of Companies
Bethlehem, PA | USA | 877-836-2228 | www.bisusa.org

Refer to the Instructions for Use for complete indications, relevant warnings, precautions, complications, and contraindications.
Z-MED and Z-MED II are trademarks of NuMED, Inc. Rx only  CV-9119 10/19 ©2019 B. Braun Interventional Systems Inc.
TABLE OF CONTENTS

Large Coronary Artery Fistula in an Adult: Rarity, Complexity and Uncertainty
Rose Tompkins, MD, FACC and Evan M. Zahn, MD, FACC, FSCAI

Pediatric Interventional Cardiology Coding Work Group Introduction
Sergio Bartakian, MD, FSCAI, FAAP; Sarosh Batlivala, MD, MSCI; Dawn Gray, SCAI Staff; Gurumurthy Hiremath, MD, FACC, FSCAI; Mark H. Hoyer, MD, FSCAI; Frank Ing, MD, FACC, MSCAI

2020 SCAI Scientific Sessions
Lee Benson, MD, MSCAI; Daniel Gruenstein, MD, FSCAI; Julie A. Vincent, MD, FSCAI, FACC, FAAP

Medical News
- Children's Hospitals and Pediatric Congenital Heart Association Call for Transparency in Cardiac Surgical Outcomes
- Structural Heart Devices Market Promises Healthy Growth with a Robust 7.5% CAGR to Reach $13 Bn in 2027

@CCardiology

DIGISONICS
Mastering the art of interpretation

Vendor neutral solution with seamless integration to hemodynamics systems, imaging modalities, PACS & EMR
- Extensive library of modifiable Mullins diagrams with 300+ exclusive to Digisonics
- Trend plots with pediatric and fetal z-scores
- Quick report capability for bringing forward data from previous reports
- Robust data mining and business analytics package
- Interoperability with QLAB, EchoPAC & TomTec

Enterprise Pediatric Reporting & Imaging Solutions for Cath, Echo and MR
A 58-year-old woman presented with symptoms of palpitations and shortness of breath. Her history was remarkable for a life-long cardiac murmur that was never fully investigated. She developed Paroxysmal Atrial Fibrillation (pAF) in her mid-40s that was managed with beta-blocker therapy. Systemic anticoagulation was attempted but not tolerated due to conjunctival bleeding and epistaxis, with further diagnostic evaluation revealing Von Willebrand Disease (VWD). In the two years preceding her presentation, she developed an increasing frequency of pAF episodes, as well as, worsening dyspnea on exertion, reduced exercise tolerance, substernal chest pressure, and new onset lower extremity edema. Her exam was notable for a widened pulse pressure, continuous murmur along the left sternal border and trace bilateral pitting edema.

Her resting electrocardiogram was normal. Transthoracic echocardiography showed normal biventricular systolic function with four-chamber dilation, no significant valvular disease, and a massively dilated left main coronary artery and coronary sinus. Color and spectral Doppler showed continuous flow from the coronary sinus into the right atrium which was suspicious for a CAF from the left coronary system to the coronary sinus given the significant dilation of the left main coronary artery. (Figure 1). Coronary Computed Tomography Angiogram (CCTA) confirmed a large CAF originating from the distal left circumflex coronary artery and draining into the coronary sinus with a remarkably tortuous course (Figure 2). A pharmacologic nuclear stress test was negative for myocardial ischemia. Cardiac catheterization demonstrated a large left-to-right intracardiac shunt (Qp:Qs 2.5:1.0), elevated pulmonary artery pressure (50/21 mmHg, mean 32 mmHg), capillary wedge pressure of 16 mmHg, and a calculated pulmonary vascular resistance of 1.6 WU or 2.5 WU/m2. The left anterior descending and proximal obtuse marginal branches appeared more normal in caliber.

Given evidence of a hemodynamically significant left-to-right intracardiac shunt in the setting of heart failure symptomatology with elevated pulmonary pressures and an increasing arrhythmia burden, CAF closure was advised. Both surgical and catheter-based options were presented to the patient, but the patient had a strong preference for catheter-based intervention. However, there was concern regarding patient’s ability to tolerate systemic anticoagulation post-procedure due to her VWD. Systemic anticoagulation would be indicated given the unknown but, presumably, high risk for post-closure myocardial infarction related to potential for thrombus formation from stagnant flow in the residually dilated left circumflex coronary artery. Additionally, the anatomical complexity of the fistula also presented possible technical challenges-e.g., ensuring that the fistula was obstructed at an appropriate level to not cause compromise of the coronary flow into the distal left circumflex territory nor affect egress of flow from the coronary sinus. After consultation with hematology and having the impression that VWD appeared acquired presumably from shearing forces associated with the high flow through the CAF into the coronary sinus that was thought to potentially resolve with CAF closure, the patient was ultimately referred for transcatheter CAF closure after risks and benefits were thoroughly reviewed.

Under general anesthesia, ultrasound guidance was used to access the right femoral artery (8f), right femoral vein (8f) and the right internal jugular vein (RIJ) (6f). Systemic heparin (to maintain ACT 250-300s) and prophylactic antibiotics were given. After performing a number of selective left coronary angiograms in a variety of axial projections through a 6Fr EBU guide catheter (Medtronic, MN, USA), supported by an 8Fr Guideliner (Teleflex, Morrisville, NC, USA), the large CAF was navigated with a soft coronary guide wire (0.014” Cougar, Medtronic, MN, USA) housed within a microcatheter (2.6 Fr Corsair, Asahi Intec, Tustin, CA, USA) (Figure 3). Because of uncertainty regarding the precise transition from fistula to true dilated coronary artery, we chose to occlude this CAF at its most distal exit point into the coronary sinus. Therefore, the microcatheter-wire assembly was directed out the exit of the fistula into the coronary sinus, right atrium and ultimately the superior vena cava. A 20 mm gooseneck snare was advanced from the RIJ and used to snare the microcatheter which was exteriorized forming an arteriovenous loop through the CAF. The soft Cougar wire was then replaced with a stiffer Grand Slam guide wire and from the RIJ a 6F Flexor sheath advanced over the guide wire into the CAF. As the exit point into the coronary sinus measured 10 mm, a 12 mm AVPII (Abbott Medical, Minneapolis, MN) was chosen and delivered slowly via the 6Fr RIJ sheath, using a combination of serial angiograms from a coronary guide catheter and tranesophageal imaging to direct proper placement. Quite soon after...
the device was placed, angiography demonstrated complete occlusion of the CAF exit point, stasis of contrast within the body of the occluded CAF and visible contrast for the first time within what was believed to possibly be the true circumflex artery (Figure 3 and 4). At the end of the procedure, repeat pulmonary artery pressure had fallen to 24/10, mean 14 mmHg.

Three months post-CAF closure, the patient was doing well with resolution of symptoms including: no further pAF episodes, normalization of her pulmonary artery pressures, and improvement in her cardiac four-chamber dilation with no residual fistulous flow. She was tolerating systemic anticoagulation with Warfarin and showed resolution of her VVd. Unfortunately, four months post-CAF closure, she suffered a non-ST elevation myocardial infarction associated with dynamic ischemic ECG changes, peak troponin-I level of 30 ng/mL, new inferior wall hypokinesis, and mildly reduced left ventricular systolic function. Coronary angiography showed no acute coronary thrombus nor residual CAF flow and a non-dominant right coronary artery. However, the left main and left circumflex coronary arteries remained severely dilated with sluggish flow. The ischemic event was thought to be thromboembolic into the distal left circumflex territory. Dual anti-platelet therapy with aspirin and clopidogrel was initiated, and Coumadin maintained with an increased INR goal from 2.5 to 3.0. Additionally, she was placed on atorvastatin and lisinopril and continued on metoprolol. Now more than six months post-procedure, patient is asymptomatic and has returned to an active lifestyle. Her left ventricular function has improved on medical therapy.

**Discussion**

Due to the rarity of the diagnosis and heterogeneity of the clinical presentation, our knowledge of the natural history, indications for intervention, and long-term outcomes following closure of large, hemodynamically significant CAF remain limited. Prior 2008 American College of Cardiology (ACC)/American Heart Association (AHA) guidelines recommended closure of large CAF regardless of symptomatology, and closure of smaller CAF in setting of symptoms clearly attributable to the fistula including: myocardial ischemia, arrhythmia, ventricular dilation or dysfunction, or endarteritis. However, the updated 2018 ACC/AHA guidelines no longer provide clear recommendations regarding indication nor method for CAF closure (surgical vs. transcatheter), but rather advise consultation with an expert team. In general, expert consensus would advocate for CAF closure in an adult when a large left-to-right shunt is present resulting in heart failure and/or increased pulmonary pressures secondary to over-circulation and/or clear association with a patient’s symptomology not explained by another etiology. Surgical or transcatheter closure remains the mainstay of treatment for large and/or symptomatic CAF, with transcatheter approach being preferred in patients with favorable anatomy and no indications for concomitant surgery. However, anatomic and hemodynamic variables must be taken into account regarding risks of closure, technical considerations regarding preferred method of closure, and post-closure complications and management strategies.

Small single-center studies suggest good procedural success with transcatheter CAF closure and relatively low morbidity among select patients. Procedural complications include: transient ST-T wave changes, atrial arrhythmia, device embolization, and coronary dissection. Late post-closure complications, including Myocardial Infarction (MI), are a concern and have been reported in up to 15% of patients, with older age at time of closure, distal origin of the CAF off the coronary artery, and CAF drainage into the coronary sinus having a higher association with post-closure MI. All these characteristics were present in our patient.

El-Sabawi et al. described Mayo Clinic’s transcatheter CAF closure experience over a 21-year period that included a total of 45 adult patients, of which 37 patients had congenital CAF and 14 of these had termination of the CAF into the coronary sinus. This represents one of the larger retrospective series for transcatheter CAF closure published to date in an adult-only cohort. Procedural success was reported in 89% of the cases, with a trend of increasing success rate and higher use of larger vascular plug devices over embolization coils in the more contemporary period. Over a median follow-up of approximately one year, two patients experienced post-procedural MI related to residual stagnation of flow in the originating coronary artery, both were distal right coronary artery to coronary sinus fistulas. One MI occurred one day post-closure and the other occurred one year post-closure despite
The Heart Institute at the UPMC Children’s Hospital of Pittsburgh is EXPANDING!

With a strategic plan for growth and expansion, the Division of Cardiology within the Heart Institute of the UPMC Children’s Hospital of Pittsburgh / University of Pittsburgh School of Medicine is recruiting additional faculty positions.

**Director of Cardiology Clinical Services**

The Heart Institute is seeking an exceptional individual to lead the Clinical Services within the Division of Cardiology, actively participating with the Division Chief and Heart Institute Leadership in the supervision and development of clinical services, strategic planning, program coordination and expansion. The applicant should have demonstrated evidence of strong leadership skills and recognized expertise as academic physician. A commitment to excellence, integrity, collegiality and professionalism is a must. Applicants should be at the Associate Professor level (or above), and Board Certified in Pediatric Cardiology.

**Director of Pediatric Non-Invasive Imaging (Echocardiography Laboratory)**

For this leadership level position, we are seeking an outstanding board-certified pediatric cardiologist with strong expertise in non-invasive imaging including all forms of echocardiography and/or cardiac MRI & cardiac CT. Applicants should be at the Associate Professor level (or above). In addition, evidence of solid leadership skills to take the Director role and help build up the Non-Invasive Imaging Program, working closely with division chief and hospital leadership. Candidates must have completed a 4th year pediatric imaging advanced fellowship and demonstrated an academic commitment in the field of imaging, with dedication to teaching, research and quality improvement. Candidates must be Board Certified in Pediatric Cardiology.

**Imaging Faculty With Expertise In Echocardiography, Including Fetal**

We are recruiting for a full-time Board Eligible/Certified non-invasive imaging faculty with expertise on TTE, TEE and FETAL echocardiography. Completion of a 4th year imaging fellowship plus skill and independence in transesophageal echocardiography is a requirement.

Imaging faculty will join an outstanding team: Including eleven echocardiographers, 16 pediatric sonographers in a highly productive echo lab – with over 20,000 echocardiograms, including over 1600 fetal echo’s and 550 TEE’s.

Echocardiography program covers Children’s Hospital, Magee Women’s hospital and multiple outreach sites and a robust tele-echo program. Further collaboration with the adult cardiology program for ACHD cMR program is anticipated, as well as with Radiology enhancing the cardiac MRI program, and MFM colleagues to expand the Fetal Cardiac Program. Candidates must be board-eligible/certified in pediatric cardiology.

**Adult Congenital Heart Disease Faculty**

The Division of Cardiology at UPMC Children’s Hospital of Pittsburgh / University of Pittsburgh School of Medicine is recruiting for additional faculty to join the Adult Congenital Heart Disease (ACHD) program. The well-established ACHD program is currently supported by 2 ACHD physicians (including one ACDH Director), 2 advanced practice providers, a dedicated RN, research coordinator and social worker. The applicant should have expertise in the management of adult congenital heart disease with prominent clinical, teaching and research skills. He or she will be working closely with division chief, ACHD Director and hospital leadership to support program expansion. Candidates must be Board-Eligible/ Certified in Pediatric Cardiology or Adult Cardiovascular Diseases and in Adult Congenital Heart Disease.

**The Heart Institute** provides comprehensive pediatric and adult congenital cardiovascular services to the tri-state region and consists of 27 pediatric cardiologists, 5 pediatric cardiothoracic surgeons, 8 pediatric cardiac intensivists and 9 cardiology fellows along with 19 physician extenders and a staff of over 100. We are honored to be ranked #3 nationally and #1 in Pennsylvania for pediatric cardiology and heart surgery by U.S. News and World Report. Our Cardiac surgical program is one of the top in the country, with a 3-star rating from Society of Thoracic Surgery (STS).

UPMC Children’s Hospital of Pittsburgh has been named one of the top U.S. News & World Report’s Best Children’s Hospitals. Consistently voted one of America’s most livable cities, Pittsburgh is a great place for young adults and families alike.

The position comes with a competitive salary and faculty appointment commensurate with experience and qualifications at the University of Pittsburgh School of Medicine. The University of Pittsburgh is an Equal Opportunity/Affirmative Action Employer. Interested individuals should forward letter of intent, curriculum vitae and three (3) letters of references. Informal inquiries are also encouraged.

**For more information, please contact:**

**Jacqueline Kreutzer, MD, FSCAI, FACC**
Chief, Division of Pediatric Cardiology
UPMC Children’s Hospital of Pittsburgh, 4401 Penn Avenue, Pittsburgh, PA 15224
Jacqueline.Kreutzer@chp.edu or 412.692.3216

The University of Pittsburgh is an Affirmative Action/ Equal Opportunity Employer and values equality of opportunity, human dignity and diversity. EOE, including disability/vets
the use of anticoagulation and aspirin. The former was managed conservatively without future complications, but the latter underwent coronary artery bypass grafting. Late fistula recanalization occurred infrequently and was more likely to occur among those patients that had some residual flow at time of closure.

In summary, this case highlights a very rare diagnosis of a symptomatic large CAF causing heart failure in an older adult, the complexities involved in the technical approach to closure, and the risk of post-closure thrombosis despite systemic anti-coagulation. Specific anatomic criteria and optimal timing of CAF closure remain poorly understood as many incidentally diagnosed CAFs are likely to remain small and clinically inconsequential. However, earlier closure of large CAFs, regardless of symptoms, seems reasonable given increased symptoms and higher complication rate post-closure with advancing age as demonstrated by our case.

References

**FIGURE 4** Transesophageal Intra-procedural Echocardiography
A, B) Mid-esophageal views with color Doppler comparison showing drainage of the coronary artery fistula (yellow arrow) into the coronary sinus with flow then into the right atrium.
C, D) Mid-esophageal views with color Doppler comparison showing presence of a vascular plug (red arrow) occluding the communication of the coronary artery fistula into the coronary sinus with no evidence of residual flow and no obstruction to normal egress of coronary sinus flow into the right atrium. CS, coronary sinus; RA, right atrium.
The only transcatheter valve designed specifically for RVOT conduits and bioprosthetic valves. Thin leaflets from naturally derived tissue open and close under minimal pressure. The flexible delivery system is designed for the right side of the heart and offers controlled, stepwise deployment of the valve with balloon-in-balloon technology.

*Melody Transcatheter Pulmonary Valve Study: Post Approval Study of the Original IDE Cohort
©2019 Medtronic. All rights reserved.
UC2018090308 EN 07/2019

Not intended to constitute medical advice or in any way replace the independent medical judgment of a trained and licensed physician with respect to patient needs or circumstances. Melody TPV is not suitable for all patients and ease of use, outcomes, and performance may vary. See the Instructions for Use for indications, contraindications, precautions, warnings, and adverse events.
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 to severe 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, 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.

CAUTION: Federal law (USA) restricts this device to sale by or on the order of a physician.

Important Labeling Information for Geographies Outside of the United States

Indications: The Melody TPV is indicated for use in patients with the following clinical conditions:
- Patients with regurgitant prosthetic/conduit RVOT conduits or bioprostheses with a clinical indication for invasive or surgical intervention. OR
- Patients with stenotic prosthetic RVOT conduits or bioprostheses where the risk of worsening regurgitation is a relative contraindication to balloon dilatation or stenting.

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

medtronic.com
710 Medtronic Parkway
Minneapolis, MN 55432-5604
USA
Tel: (763) 514-4000
Fax: (763) 514-4879
Toll-free: (800) 328-2518

LifeLine
CardioVascular Technical Support
Tel: (877) 526-7890
Fax: (763) 526-7888
rs.cstechsupport@medtronic.com

©2019 Medtronic. All rights reserved. Medtronic, Medtronic logo, and Further, Together are trademarks of Medtronic. All other brands are trademarks of a Medtronic company.

UC201809495a EN 07/2019
**Pediatric Interventional Cardiology Coding Work Group Introduction**

Sergio Bartakian, MD, FSCAI, FAAP; Sarosh Batlivala, MD, MSCI; Dawn Gray, SCAI Staff; Gurumurthy Hiremath, MD, FACC, FSCAI; Mark H. Hoyer, MD, FSCAI; Frank Ing, MD, FACC, MSCAI

**Abbreviations**

AMA - American Medical Association  
CCCHD - Cardiac Catheterization for Congenital Heart Disease  
CMS - Centers for Medicare and Medicaid Services  
CPT* - Current Procedural Terminology  
NCCI - National Correct Coding Initiative  
PICCW - Pediatric Interventional Cardiology Coding Workgroup  
RUC - Relative value scale Update Committee  
RVU - Relative Value Units  
SCAI - Society of Cardiovascular Angiography and Interventions

This article represents the first in a series of papers introducing the Pediatric Interventional Cardiology Coding Workgroup (PICCW) to the pediatric cardiology community, and provides updates and education regarding the process of Current Procedural Terminology (CPT* ) code creation.

In 1966, the American Medical Association (AMA) first published a set of standard terms and descriptors to document medical procedures, known as CPT. Over the next 30+ years, numerous revisions took place with each progressively more detailed, in line with the increasing complexity of the health care system. In 2000, after a thorough review by the AMA, CPT became the national coding standard for reporting medical services and procedures.

The CPT Editorial Panel is responsible for maintaining the CPT code set and is supported by the larger body of specialty society advisors, the CPT Advisory Committee. The members of this committee are primarily physicians nominated by the national medical specialty societies. Currently there are 156 members serving in this capacity. The importance of adequate representation on this committee cannot be overstated. The number of representatives per specialty is variable; some specialties are represented by a dozen or more members due to representation by multiple societies while others, such as the National Athletic Trainers Association and the American Massage Therapy Association have two advisors each. Unfortunately, the pediatric cardiology community never sought to be formally involved in this process until 2017 with the nomination and appointment of the author through the Society of Cardiovascular Angiography and Interventions (SCAI).

Whereas the CPT Panel oversees the process of creating new codes, it is the task of the Relative Value Scale Update Committee (RUC) to recommend a value to the Centers for Medicare and Medicaid Services (CMS). Overall, the process of creating a new code involves initial presentation by the Specialty Society Advisor to the CPT panel. At these sessions, the code is assigned a 5-digit number, and the description of the work performed is outlined. Once approved by CPT, the proposed code is surveyed among the stakeholder society members, according to stringent rules, regarding the time and intensity needed to perform the respective procedure. A society representative then presents the society’s recommended value for the code based on the RUC survey, to the RUC panel for valuation. The results of the RUC meeting and their recommended Relative Value Units (RVU) are then forwarded to CMS for formal acceptance.

Until recently, the use of RVUs for determining physician productivity was mostly relegated to adult medicine, while pediatricians were relatively protected. This was primarily due to utilization data being available only for Medicare patients, leaving no accurate means to track physician productivity in pediatric medicine. Additionally, the pediatric cardiology community mistakenly perceived CPT merely as a means by which to be paid for services. Since the majority of pediatric interventional cardiologists were (are) hospital salary-based practitioners, this simply did not matter. Unfortunately, over the years, this lack of representation led to increasing gaps in coding due to the ever-increasing complexity of Cardiac Catheterization for Congenital Heart Disease (CCCHD). Whereas once, CCCHD was primarily a diagnostic tool, the field has significantly evolved into one of predominantly therapeutic intervention.

Beyond the fallacy that RVUs are merely used to justify a physician’s salary, there is a greater understanding required of the implication of having an inadequate coding structure for one’s specialty. The CPT code not only provides a means to assign value for physician work, but also includes built-in metrics to capture liability and facility and non-facility practice expenses. Lack of appropriate reimbursement is typically perceived by administrations as the service you provide being unworthy of investment. This will result in budgetary shortages that can be used to limit the hiring of necessary additional personnel and necessary replacement of outdated lab equipment. All of these deficiencies result in slower turnaround times, less efficiency, and increased complications, thus further diminishing your value to your institution.

As of early 2016, despite the dozens of CPT codes used during CCCHD procedures, only 12 were created specifically for the congenital patient, and all but one of those were 13+ years old (Table 1), and nearly obsolete. An internal audit conducted by the author in 2016 while at the University of Texas, identified that only about 50% of all work performed for CCCHD was being captured by the available coding system. This rate has since been further validated by the author to be consistent at other institutions, and is further diminished by any coding errors made by the operators and coding specialists, as well as inappropriate denials by payers.
In early 2017, SCAI members began to voice concerns regarding the increasing gaps in appropriate coding. The problem was compounded by the growing trend of denials from payers for clearly erroneous reasons being provided by the so-called experts in coding/billing. In response, SCAI formed the PICCW, and established primary goals to 1) identify gaps in CPT coding specific to CCCHD procedures, 2) identify and address sources of coding misinformation, and 3) develop a structured approach to address the needs through various resolution pathways. These pathways, among others, would include revision of long-standing codes in need of updates, creation of entirely new CPT codes, issuing formal coding guidance to the coding/billing community and payers, and educating providers on proper coding practices. This latter issue quickly proved to be one of the most daunting issues SCAI was to face as we moved forward. It became readily apparent that the lack of education not only involved the providers, but also the coding community.

With respect to the providers, our observations and internal survey results suggest this problem stems from the simple fact that the majority of pediatric interventional fellows receive inadequate training on coding and billing during their formal training year. Those who claimed to have received some training, usually referred to the long-standing paper billing sheet with boxes to check at procedures’ end; the “this is how we have always done it” method of education. Beyond the task of checking boxes, there was no education regarding: what documentation was necessary, the CPT, RUC, taking a RUC survey, the actual work included in the CPT codes they were checking boxes for, and very importantly, understanding the times associated with performance of work. Many of the problems with the valuation of existing pediatric interventional codes are due to this lack of education regarding procedure times.

As an example, a diagnostic right and left heart catheterization through existing septal communication (CPT 93533, created in 1998) includes 120 minutes of intra-procedure time. Just five years later, when the community created the bundled code for an Atrial Septal Defect device closure (CPT 93580, created in 2003), they claimed the same 120 minutes of intra-procedure work was needed; despite the fact that this bundled code included all of the diagnostic work of the aforementioned 93533 code, with the addition of the work of placing the closure device. In a recent survey for a new bundled code describing placement of an intra-cardiac stent (such as for a Fontan fenestration, restrictive atrial septum, or right ventricular outflow tract, etc.) that also included a complete right and left diagnostic study, the median time those surveyed claimed was a mere 92 minutes. In fact, one respondent stated this highly complex procedure, including all of the diagnostic work, with higher than typical intensity, only required a total of 40 minutes.

The concept of time perception and its many fallacies and pitfalls have long been studied and described in the literature. We know from decades of research on this topic that individuals have a very poor ability to gauge the amount of time it takes to do something, particularly in stressful situations. Unfortunately, this is the most important factor used in the RUC valuation system. The other factor, being the intensity of the procedure, is a purely subjective determination. In the next article in this series, we will dive deeper into what work constitutes each of the three segments in a RUC assessment of total procedural time: the pre, intra, and post procedure times.

### Table 1

<table>
<thead>
<tr>
<th>Year Created</th>
<th>CPT Codes</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1992</td>
<td>92992, 92993</td>
<td>Atrial Septostomy</td>
</tr>
<tr>
<td>1992</td>
<td>92990</td>
<td>Pulmonary Valvuloplasty</td>
</tr>
<tr>
<td>1998</td>
<td>92997, 92998</td>
<td>Pulmonary Angioplasty</td>
</tr>
<tr>
<td>1998</td>
<td>93530, 93531, 93532, 935533</td>
<td>Base Congenital Cath Codes</td>
</tr>
<tr>
<td>2003</td>
<td>93580, 93581</td>
<td>ASD and VSD Device Closure</td>
</tr>
<tr>
<td>2014</td>
<td>93582</td>
<td>PDA Device Closure</td>
</tr>
</tbody>
</table>

In early 2017, SCAI members began to voice concerns regarding the increasing gaps in appropriate coding. The problem was compounded by the growing trend of denials from payers for clearly erroneous reasons being provided by the so-called experts in coding/billing. In response, SCAI formed the PICCW, and established primary goals to 1) identify gaps in CPT coding specific to CCCHD procedures, 2) identify and address sources of coding misinformation, and 3) develop a structured approach to address the needs through various resolution pathways. These pathways, among others, would include revision of long-standing codes in need of updates, creation of entirely new CPT codes, issuing formal coding guidance to the coding/billing community and payers, and educating providers on proper coding practices. This latter issue quickly proved to be one of the most daunting issues SCAI was to face as we moved forward. It became readily apparent that the lack of education not only involved the providers, but also the coding community.

With respect to the providers, our observations and internal survey results suggest this problem stems from the simple fact that the majority of pediatric interventional fellows receive inadequate training on coding and billing during their formal training year. Those who claimed to have received some training, usually referred to the long-standing paper billing sheet with boxes to check at procedures’ end; the “this is how we have always done it” method of education. Beyond the task of checking boxes, there was no education regarding: what documentation was necessary, the CPT, RUC, taking a RUC survey, the actual work included in the CPT codes they were checking boxes for, and very importantly, understanding the times associated with performance of work. Many of the problems with the valuation of existing pediatric interventional codes are due to this lack of education regarding procedure times.

As an example, a diagnostic right and left heart catheterization through existing septal communication (CPT 93533, created in 1998) includes 120 minutes of intra-procedure time. Just five years later, when the community created the bundled code for an Atrial Septal Defect device closure (CPT 93580, created in 2003), they claimed the same 120 minutes of intra-procedure work was needed; despite the fact that this bundled code included all of the diagnostic work of the aforementioned 93533 code, with the addition of the work of placing the closure device. In a recent survey for a new bundled code describing placement of an intra-cardiac stent (such as for a Fontan fenestration, restrictive atrial septum, or right ventricular outflow tract, etc.) that also included a complete right and left diagnostic study, the median time those surveyed claimed was a mere 92 minutes. In fact, one respondent stated this highly complex procedure, including all of the diagnostic work, with higher than typical intensity, only required a total of 40 minutes.

The concept of time perception and its many fallacies and pitfalls have long been studied and described in the literature. We know from decades of research on this topic that individuals have a very poor ability to gauge the amount of time it takes to do something, particularly in stressful situations. Unfortunately, this is the most important factor used in the RUC valuation system. The other factor, being the intensity of the procedure, is a purely subjective determination. In the next article in this series, we will dive deeper into what work constitutes each of the three segments in a RUC assessment of total procedural time: the pre, intra, and post procedure times.

**SERGIO BARTAKIAN, MD, FSCAI, FAAP**

Children's Hospital of Michigan
AMA CPT Advisor for SCAI
dctrbar@gmail.com
A very special thank you to Ms. Dawn Gray, our dedicated SCAI staff member, who has worked tirelessly in helping the PICCW. Without her immense knowledge, experience, and guidance, we would not have achieved any of our major successes over the past three years.
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.

(Formerly Pediatric Specialists of America)

nicklauschildrens.org/Heart
The 2020 SCAI Scientific Sessions are almost here. As interventional cardiologists or other professionals involved in the care of children or adults with Congenital Heart Disease, we are privileged to participate in one of the great success stories in modern medicine. As we enter the third decade of the 21st century, there is a tsunami of information to assimilate. Attending the 2020 annual meeting will help you in learning what's new, what works, and what doesn't. If you have not registered, please keep reading!

As in past meetings, SCAI 2020 has a dedicated Learning Track devoted to interventional procedures for patients with congenital heart disease. The curriculum for the Congenital Heart Disease Track will be presented by experts and thought leaders in the field, in an environment that encourages a friendly, approachable discourse. We encourage attendee participation through questioning and discussion during the sessions and are informal enough to "corner" the speakers for more extensive individual conversations. In addition to the Congenital Heart Disease (CHD) Track, there are four other Learning Tracks, which will provide comprehensive programs in structural, peripheral, and coronary interventions. Selectively visiting these Tracks will provide opportunities for crossover learning. This year's SCAI meeting is at The Hyatt Regency Atlanta, May 13th through the 16th. This is a wonderful venue in a city which has unlimited cultural, culinary and entertainment options, and you can plan on warm sunny days! Being the home of CNN, this year's SCAI Featured Speaker is Dr. Sanjy Gupta.

Just a few words about the Congenital Heart Disease Track Program.....The organizational theme of this year's CHD Track is to address complications that occur in the interventional management of patients with CHD: awareness of the problems that can occur, how to avoid them through preparation and how to deal with them when they do occur. The sessions begin on Thursday afternoon, with our initial session on what we can learn from the past; registries and determination of who is at risk. Later that day we will examine various lesions and how to stay out of trouble. During the day a new session has been added with presentations of interesting interventional cases, followed by a moderated poster session. Our first full day, Friday, begins with oral CHD abstracts (a prize going to the best of the best; moderated or oral presentations). The PICES session this year, planning for complicated interventions and dealing with unpredictable complications, will be followed by the annual Mullin's Lecture. This year the honor goes to Dr. Shak Qureshi, well-known to all of us as an innovator and teacher. This will be followed by our 'I Blew It' and 'Cath Lab Jeopardy' sessions, both looking at and poking fun at ourselves. On Saturday we pick up the theme of complications, taking a critical look at pulmonary vein stenosis, obstructions and ruptures, and other stuff that doesn't go so well! Later in the day we will have a CHD live case presentation from San Diego and finally an ACHD debate on the advisability of closing that fenestration and what can happen down the road. As always, there will be opportunities to reunite and network with colleagues and old friends while we discuss the most important and the newest techniques and technologies in interventional congenital cardiology as we strive for Semper Ad Meliora! Your organizing committee has tried very hard to put together an interesting, comprehensive program. We hope you will join us in Atlanta! Register and download the advance program at www.scai.org/SCAI2020.
MEETING CALENDAR

2020

May

02

ACHA Michigan Regional Conference
Grand Rapids, MI, USA
https://www.achaheart.org/get-involved/events/?c=1655

13-16

SCAI 2020 Scientific Sessions
Atlanta, GA, USA
http://www.scai.org/SCAI2020

19

AlMed Cardiology 2020
Newport Beach, CA, USA
https://ai-med.io/all-events/clinician-series/cardiology/

June

11-13

30th Annual International Symposium on Adult Congenital Heart Disease
Cincinnati, OH, USA
https://www.cincyhearteducationseries.org/achdsymposium

07-20

TVT2020
Chicago, IL, USA
https://www.crf.org/tvt

19-22

ASE 2020 – The Next Millennium
Denver, CO, USA
https://www.asescientificsessions.org/2020registration/

2020

Atlanta

May 13-16

Scientific Sessions

The Intersection of Innovation & Practice

Registration is open
www.scai.org/SCAI2020

Keynote Speaker
Dr. Sanjay Gupta
The NuDEL™ Stent Delivery System is designed for the efficient and effective treatment of Coarctation of the Aorta.

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

The NuDEL System is available for immediate purchase in the EU. Contact us or your local distributor to place an order.

NuMED
World Leader in Pediatric Cardiology

NuMED, Inc.  I  2880 Main Street  I  Hopkinton, NY 12965 USA
Tel: 315.328.4491  I  Fax: 315.328.4941  I  www.numedforchildren.com
Children’s Hospitals and Pediatric Congenital Heart Association Call for Transparency in Cardiac Surgical Outcomes

PRNewswire - Congenital Heart Disease (CHD) is the most common birth defect in the United States, occurring in approximately one in every 100 babies. However, information about the quality of care available, along with short- and long-term outcomes for patients, is limited and oftentimes difficult to interpret. To address this shortfall Children's Hospital Colorado and others are partnering with the Pediatric Congenital Heart Association (PCHA), https://www.conqueringchd.org/, in an effort to make this information more accessible and understandable to families impacted by CHD. This effort, which comes on the heels of PCHA’s Transparency Summit, hosted by Nationwide Children's Hospital and attended by families, clinicians, researchers, and hospital leaders from around the country, has the goal of improving information sharing, overall outcomes and quality, and patient and family experience.

PCHA is developing a website that will provide patients and families with program-specific pediatric and congenital cardiology data combined with improved explanation and context for families to aid interpreting the data. The Heart Institute at Children's Hospital Colorado, https://www.childrenscolorado.org/doctors-and-departments/departments/heart/, the Herma Heart Institute at Children's Hospital of Wisconsin and The University of Michigan Congenital Heart Center at C.S. Mott Children's Hospital in Ann Arbor were among the first hospitals to share their outcomes data with the PCHA for testing and development of the website.

"Of the 42,000 babies diagnosed with CHD each year in the United States more than 13,000 will require a life-saving intervention in the first weeks or months of life," said David Kasnic, Executive Director of the PCHA. "My daughter was one of those 13,000 and since that time it has been my goal to improve education, awareness, research and advocacy for all those living with CHD. This website will go a long way toward ensuring parents have the information they need to choose the best care for their child."

"Children's Hospital Colorado has long been committed to transparency about our outcomes, as well as the length of time our patients stay post-surgery," said Jim Jaggers, MD, Cardiothoracic Surgeon and Medical Director of the Heart Institute at Children's Hospital Colorado. "We believe open, honest reporting and dialogue is key to improving outcomes for all patients."

The PCHA’s new website is not intended to rank or compare hospitals but instead, it will provide data to support an educated conversation between families and the care teams. The PCHA will continue to add metrics and subsequent updates to the site.

“When a family receives a diagnosis, they are often overwhelmed and don’t know what questions to ask,” said Kasnic. “With the launch of this site, we can help families not only identify questions but also provide a centralized consumer-facing tool for getting some of those questions answered. In the end, we hope this information will lead to more informed and thoughtful conversations between families and their providers.”
Structural Heart Devices Market Promises Healthy Growth with a Robust 7.5% CAGR to Reach $13 Bn in 2027

Shift in procedures for Aortic Stenosis and Mitral Regulation continue to drive robust growth in the structural heart devices market. The technical advancements in healthcare procedures continue to make way for minimally-invasive methods, which are ideal for elderly patients. The tissue-engineered heart valves or biological valves will emerge as the largest segment in the market with an anticipated valuation of US $8 bn by 2027 end. The segment accounted for a 55% market share in 2018.

Key Impediments for Structural Heart Device Market Players
- Stringent government regulations, and large costs associated with hospitalization, and limited reach of private insurance players remain a concern for players in the structural heart devices market.
- Additionally, the lack of awareness about regular medical check-ups for elderly patients also remains a restraint in expansion of the structural heart devices market. However, this is changing slowly. In 2017, the Alliance for Aging Research ran a National Heart Valve Disease Awareness campaign in United States. Collaborations with organizations like these will make way for more opportunities for growth for players in the structural heart devices market.

Structural Heart Device Market: Region-wise Analysis
North America will likely hold the largest share of total revenues in the global structural heart devices market. The region accounted for 42% share of the total revenues in 2018, due to the innovative procedures in the region, and positive adoption of technological advancements, and product launches remain major drivers for growth.

Procedures like interventional catheter technologies remain at the forefront of driving growth globally. These have been adopted globally to support hybrid procedures, and modernization of healthcare systems. The growing collaboration between surgeons, cardiologists, and interventionists will drive major growth for the structural heart devices market in the near future.

Structural heart device market growth in 30+ countries includes: U.S., Canada, Germany, United Kingdom, France, Italy, Russia, Poland, Benelux, Nordic, China, Japan, India, and South Korea. Request a sample of the study, https://www.transparencymarketresearch.com/sample/sample.php?flag=S&rep_id=54915.

Structural Heart Device Market: Competitive Analysis
Main players in the structural heart device market are Medtronic, Abbott Laboratories, Boston Scientific Corporation, Edwards Lifesciences Corporation, LivaNova plc, Lepu Medical. New product innovations to meet patient needs for mitral valve replacement remains a major strategy adopted by key players in the market. Additionally, acquisitions and mergers also remain integral to the approach of key players to expand their footprint in the global structural heart device market.