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Transcatheter Management of Coronary Sinus Atresia in an Infant with Severe Cardiac Dysfunction Following Superior Cavopulmonary Anastomosis for Univentricular Palliation

Sok-Leng Kang, MBBS & Lee N. Benson, MD

Abstract

Coronary sinus ostial atresia (CSOA) associated with coronary venous drainage through a persistent left superior vena cava is rare. However, preoperative recognition of this anomaly is vital in infants undergoing cardiac surgery that might inadvertently lead to impairment of coronary venous return. We report transcatheter treatment of CSOA in an infant with severe cardiac dysfunction following superior bidirectional cavopulmonary anastomosis. Successful decompression of the atretic coronary sinus with radiofrequency perforation and successive balloon dilation resulted in improved ventricular function. Transcatheter management of CSOA is feasible and an attractive alternative to surgery, particularly in patients with significant single ventricle dysfunction.

Keywords: Coronary sinus atresia, functional single ventricle, percutaneous, perforation

Introduction

Coronary sinus ostial atresia (CSOA) is rare and typically associated with anomalous coronary venous drainage through a persistent left superior vena cava (LSVC) and/or intra-cardiac Thebesian veins into the atrium. Although an intrinsically benign anomaly, CSOA associated with a LSVC has serious implication in children with complex congenital heart defects undergoing cardiac surgery.[1] Specifically, creation of a superior bidirectional cavopulmonary connection (BCPC) during second stage univentricular palliation with or without disconnection of the LSVC, can obstruct coronary venous egress resulting in myocardial ischaemia. Surgical strategies to secure coronary venous drainage under such circumstances are well described with good outcomes.[2-5] Here, we describe percutaneous management of CSOA in an infant with single ventricle physiology and severe cardiac dysfunction following a superior BCPC.

Case Report

A 5-month-old infant with double outlet right ventricle, mitral atresia, and bilateral superior vena cava with a bridging innominate vein underwent bidirectional BCPC and Damus-Kaye-Stansel procedure. The right superior vena cava (SVC) was anastomosed to the right pulmonary artery and a very small LSVC was left intact due to intraoperative suspicion of coronary sinus (CS) ostial stenosis. Her previous procedures included pulmonary artery (PA) banding and coarctation repair at four days old,

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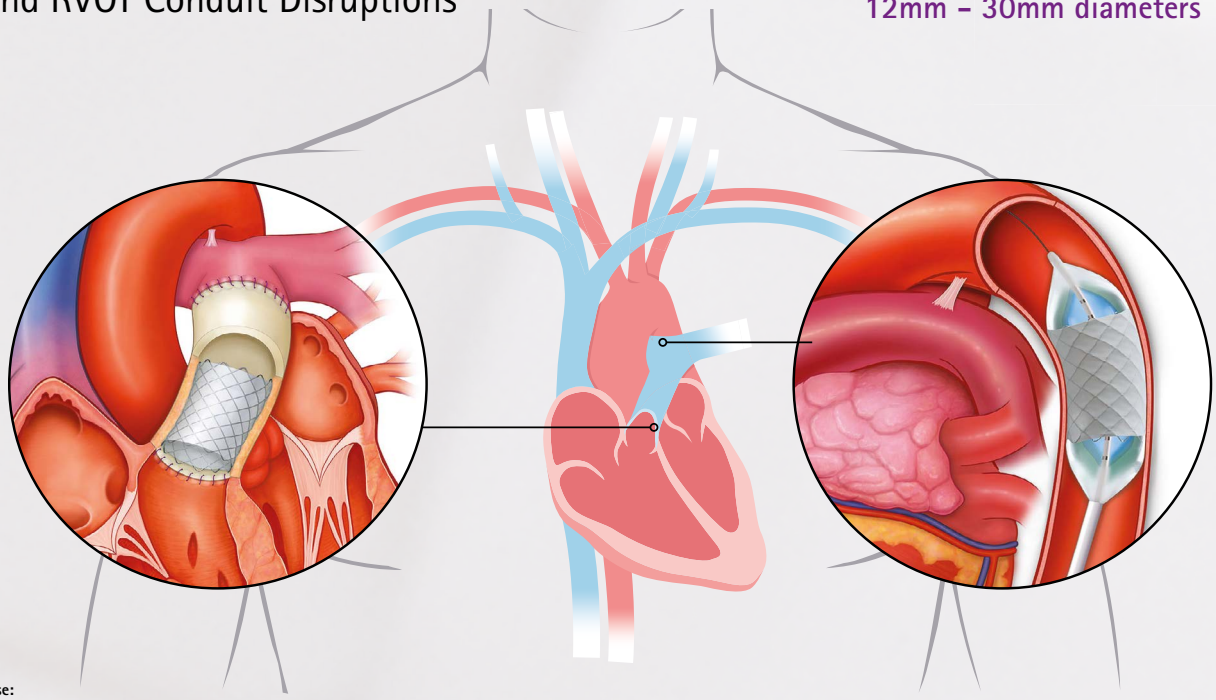

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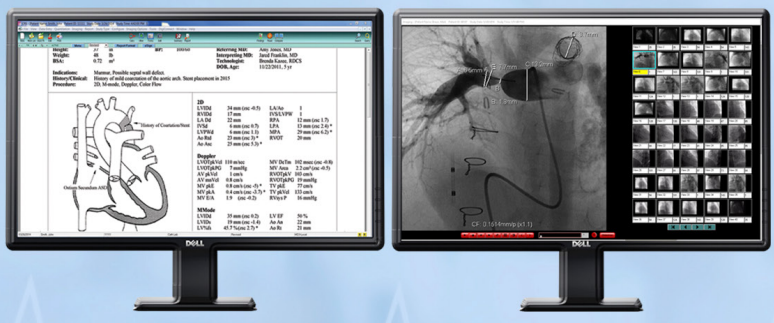

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followed by repeated balloon dilations of the PA band due to progressive cyanosis when she was four months old. On the first post-operative day, the infant had a cardiac arrest shortly after extubation with profound ventricular dysfunction and was cannulated emergently to extracorporeal membrane oxygenation (ECMO). In the absence of cardiac recovery and increasing concerns over adequacy of pulmonary blood flow, the child was taken to the cardiac catheterization laboratory on Day 7 of ECMO support for further evaluation. Initial angiography via the left internal jugular access demonstrated stenosis of the LSVC with decompressing channels to the vertebral plexus and inferior vena cava. The CS was atretic with extensive retrograde filling of the cardiac veins (**Figure 1**). We cannulated the CS with a 5F JR2.5 catheter (Cordis, Santa Clara, CA, USA) and the atretic CS was perforated with a Nykanen wire (Baylis Medical, Montreal, Canada) into the right atrium (RA) with radiofrequency energy of 3W applied for one second. The Nykanen wire was then replaced with a Grand Slam coronary wire, over which sequential dilations of the perforation were performed with 3mm and 5 mm Trek balloons (Abbott Vascular, Illinois, USA). Following decompression of the CS, there was a reduction of CS mean pressure from 13mmHg to 7mmHg and LSVC pressure from 17mmHg to 7mmHg. The right SVC pressure remained at 15mmHg. Repeat angiography in the CS showed rapid contrast entry to the RA and no extravasation of contrast (**Figure 2**). Echocardiography showed gradual recovery of ventricular function in the ensuing 48 hours. Despite improved cardiac function, persistent desaturation made weaning from ECMO support difficult. On Day 12 of ECMO, bedside surgical ligation of the LSVC was performed. On Day 14, a further cardiac catheterization demonstrated an unobstructed BCPC but mildly hypoplastic proximal branch PAs. Mean right SVC pressure was 8mmHg. She underwent balloon dilation of bilateral branch pulmonary arteries and embolization of venovenous collaterals. Subsequent ECMO course was complicated by a massive cerebral infarction and significant coagulopathy, and life-sustaining therapy was ultimately withdrawn on Day 17 of ECMO.

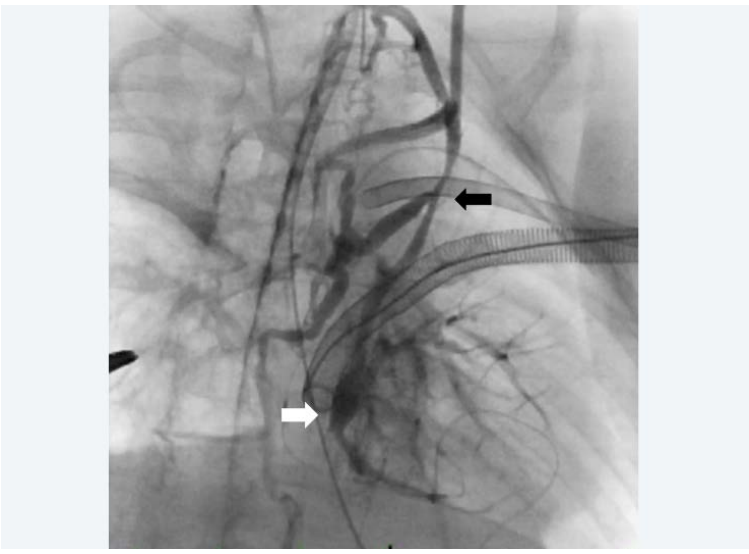

FIGURE 1 *Angiogram via the left internal jugular access demonstrating stenosis of the left superior vena cava (black arrow) with decompressing channels to the vertebral plexus and inferior vena cava. The coronary sinus was atretic (white arrow) with extensive retrograde filling of the cardiac veins.*

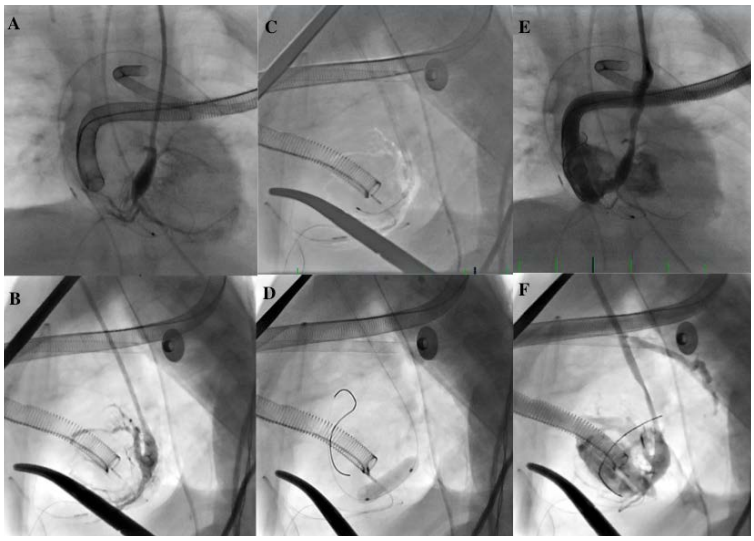

FIGURE 2 *(**A,B**) The atretic coronary sinus (**CS**) was cannulated with a 5F JR2.5 catheter and the position of the venous ECMO cannula in the right atrium was used as a target for a (**C**) radiofrequency perforation of the CS with a Nykanen wire. (**D**) The perforation was dilated sequentially with 3mm and 5mm coronary balloons. (**E,F**) Repeat angiography showed widely patent CS with rapid contrast entry to the RA.*

Discussion

Recognition of CSOA and persistent LSVC is crucial prior to a superior BCPC in staged univentricular repair. In the majority of children with bilateral SVCs, both caval veins are divided and anastomosed to the pulmonary arteries. If the LSVC was considerably smaller, particularly in the presence of a bridging innominate vein, the LSVC is simply ligated to prevent venous runoff to the right atrium. When the LSVC is the only outlet draining an atretic CS, disconnection of the LSVC results in interruption of coronary venous egress, acute coronary venous hypertension and myocardial ischaemia. Even when the LSVC is preserved, the higher-pressure in the caval system after a superior BCPC can adversely affect the coronary arterio-venous gradient and myocardial perfusion.[6] Our patient had normal ventricular function preoperatively; thus, in retrospect, the cardiac arrest was likely precipitated by progressive coronary venous obstruction due to the stenotic LSVC and elevated venous pressures post BCPC. Further, recovery of cardiac function was observed following decompression of the atretic CS.

When CSOA is diagnosed pre- or intra-operatively in the absence of ventricular dysfunction, surgical treatment has yielded good outcomes. Various surgical techniques of CS unroofing or rerouting of LSVC to the atrium are well reported.[1-5] Transcatheter management of CSOA offers a less invasive approach and may reduce operative risks particularly in the setting of significant ventricular dysfunction. To the best of our knowledge, percutaneous treatment of CSOA associated with single ventricle physiology has only been reported twice in the literature. El-Said et al described successful radiofrequency perforation of an atretic CS and subsequent stent implantation in a 3-month-old infant with progressive ventricular dysfunction following Norwood Stage 1 procedure.[7] Petit et al described a similar technique in a young adult who presented with symptomatic heart failure after Fontan completion.[8] Both showed good

recovery of cardiac function and symptomatic improvement at short-term follow-up.[7,8] From a technical perspective, the trajectory of CS perforation can be guided by placing a snare or curved tip catheter at the right atrial end of CS; or in our case, using the existing venous ECMO cannula in the RA as a target. Additionally, multimodality imaging such as transoesophageal echocardiography, pre-procedural computed tomography (CT) or magnetic resonance image (MRI) fusion with live fluoroscopy may be useful for procedural guidance. Once unobstructed coronary sinus flow to the RA is established, the LSVC can be occluded percutaneously or ligated surgically at the time of BCPC or Fontan.[7,8]

The diagnosis of CSOA is challenging, with the majority of reported cases found incidentally during coronary angiography, intra-operatively or at autopsies.[2,3,9] In infants with complex cardiac anomalies undergoing single or biventricular repair; a thorough knowledge of the anatomy of systemic veins and patency of coronary sinus is necessary to inform surgical planning. The presence of left SVC on preoperative echocardiogram, with retrograde flow and/or dilation of coronary sinus should prompt suspicion of coronary sinus atresia and further evaluation with CT or MRI.[9] Similarly, when a persistent LSVC is identified during cardiac catheterization, the direction of flow should be scrutinized. If retrograde flow is suspected, selective LSVC angiography is indicated to determine the patency of the coronary sinus and presence of anomalous pulmonary venous drainage.[10]

Conclusion

Atresia of the coronary sinus associated with persistent LSVC is rare. However in the setting of univentricular repair, one should always remain cognizant of the possibility of CSOA to avoid the catastrophic consequences of interrupted coronary venous drainage. Our case supports the feasibility and safety of percutaneous decompression of CSOA. The transcatheter approach is less invasive and an attractive alternative to surgery in patients with single ventricle dysfunction.

Declarations

Funding: None
Conflicts of interest: The authors declare that they have no conflict of interest.

References

1. Fulton JO, Mas C, Brizard CP, Karl TR (1998) The surgical importance of coronary sinus orifice atresia. Ann Thorac Surg 66:2112-2114.
2. Tadokoro N, Hoashi T, Kagisaki K, Shimada M, Kurosaki K, Shiraishi I, Ichikawa H (2016) Clinical Features and Surgical Outcomes of Coronary Sinus Orifice Atresia. Pediatr Cardiol 37:387-391.
3. Santoscoy R, Walters HL, 3rd, Ross RD, Lyons JM, Hakimi M (1996) Coronary sinus ostial atresia with persistent left superior vena cava. Ann Thorac Surg 61:879-882.
4. Kang SR, Park WK, Kwon BS, Ko JK, Goo HW, Park JJ (2018) Management of Coronary Sinus Ostial Atresia during a Staged Operation of a Functional Single Ventricle. Korean J Thorac Cardiovasc Surg 51:130-132.
5. Yokoyama S, Yonekura T, Nishiwaki N, Taniguchi S (2017) Coronary Sinus Orifice Atresia After the Fontan Completion: Unique Repair Technique. Ann Thorac Surg 104:e151-e153.
6. Ilbawi MN, Idriss FS, Muster AJ, DeLeon SY, Berry TE, Duffy CE, Paul MH (1986) Effects of elevated coronary sinus pressure on left ventricular function after the Fontan operation. An experimental and clinical correlation. J Thorac Cardiovasc Surg 92:231-237.
7. El-Said H, Hegde S, Moore J (2014) First report of atretic coronary sinus stenting in a 5-kg infant resulting in dramatic improvement of ventricular function in functional single ventricle. Congenit Heart Dis 9:E175-178.
8. Petit CJ, Webb GD, Rome JJ (2007) Creation of a coronary sinus to atrial communication in coronary sinus ostial atresia improves cardiac function after Fontan. Catheter Cardiovasc Interv 70:897-899.
9. Kim C, Goo HW, Yu JJ, Yun TJ (2012) Coronary sinus ostial atresia with persistent left superior vena cava demonstrated on cardiac CT in an infant with a functional single ventricle. Pediatr Radiol 42:761-763.
10. Prasad D, Strainic JP, Pandya K, Kouretas PC, Ashwath RC (2016) Venous Myocardial Infarction in an Infant with Obstructed Totally Anomalous Pulmonary Venous Drainage and Coronary Sinus Ostial Atresia. Tex Heart Inst J 43:430-432.

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Congenital Interventional Cardiology Coding Work Group Part Four: Project Timelines

Sergio Bartakian, MD, FSCAI, FAAP; Sarosh Batlivala, MD, MSCI; Gurumurthy Hiremath, MD, FACC, FSCAI; Frank Ing, MD, FACC, MSCAI

AMA American Medical Association
CCCHD Cardiac Catheterization for Congenital Heart Disease
CMS Centers for Medicare and Medicaid Services
CPT® Current Procedural Terminology
PDA Patent Ductus Arteriosus
CICCW Congenital Interventional Cardiology Coding Workgroup
RUC Relative value scale Update Committee
RVU Relative Value Units
SCAI Society for Cardiovascular Angiography and Interventions

In the initial three articles of this series, we focused primarily on the components of the physicians' description of work and briefly touched on the RUC survey. These were important as part of the effort to educate the Congenital Cardiology community on the importance of CPT® coding and the RUC valuation for their work. The majority of the work of the CICCW, however, has been focused on the creation of a comprehensive congenital cardiac catheterization code set. Previously, a simplistic set of diagnostic codes had been created with the bulk of the work performed being captured by codes borrowed from other specialties: namely, non-congenital cardiac catheterization and interventional radiology. This left many procedures without an appropriate code resulting in the lack of any reimbursement for many complex interventions. Whereas these borrowed codes may have served their purpose early on in the era of predominantly diagnostic catheterization, they fail miserably in capturing the complexity of patients in a specialty that has seen dramatic changes over the past 2-3 decades. In fact, the field continues to transform every year, with ongoing advancements in imaging technology, catheterization equipment, and interventional devices.

The process of creating of a new code is not straightforward. The timeline is quite lengthy and even in the absence of any delays, will require at least 20 months from start to finish (**Figure 1**).

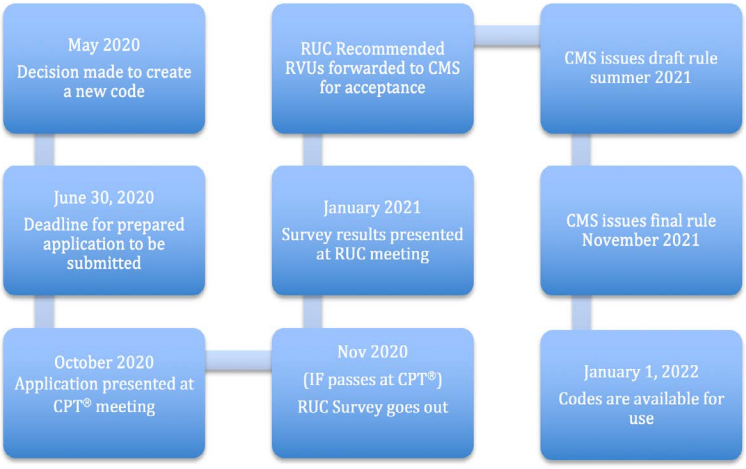

FIGURE 1 *Timeline for a new CPT® code*

Creating a new code for each and every item for which we have a need is, therefore, a significantly time-consuming and ineffective plan. Alternatively, there are other pathways through which resolution could be achieved for some problems (**Figure 2**).

Editorial revisions are also processed through the same CPT® pathway and are essentially suggestions for corrections or changes to an existing item in the CPT® book. These can often be expedited (somewhat), as they typically do not need to go through the RUC process. The CICCW used this pathway to correct an error regarding the ability to report a pulmonary angiogram (93568) at the time of a PDA device closure procedure (93582).

CPT® Assistant Articles are a paid-for service used by coders/billers and offer another possible solution to some problems. They are essentially a collection of question and answer scenarios used to describe difficult/confusing coding scenarios and how to appropriately report them. These articles are typically developed based on questions from providers, which are ultimately forwarded to the respective specialty society and coding experts for guidance regarding the proper coding. This guidance is then released in the form of a CPT® Assistant Article to explain the proper coding.

Finally, the National Correct Coding Initiative (NCCI) is an entity developed by the CMS. The work performed by NCCI is contracted, by the CMS, to a private firm to provide coding guidance and control improper coding leading to inappropriate payment in Part B claims.[1] The SCAI, along with our partners at the ACC, has submitted several letters to NCCI requesting corrections to flawed prior directives which improperly conflated congenital and non-congenital procedures.

To date, the CICCW has completed several projects with others in progress and many more in various stages of planning. These are shown in **Table 1**, along with a timeline for anticipated dates for when completed projects will be available, as well as future dates for subsequent projects and meetings. Of note, the septostomy and intracardiac stent code project was completed and received positive RVU valuations. These values are currently pending CMS final approval and will be released in November 2020 for use as of January 1, 2021.

The current project which has recently passed through CPT® and now at the RUC stage is the new set of diagnostic congenital catheterization codes; a monumental task which the CICCW members have been working on for nearly two years. Unlike the older CPT® codes (93530, 93531, 93532, and 93533), these now include two codes for right heart catheterization alone, one code for congenital left heart catheterization alone, and two codes for combined right and left heart catheterization. Whereas the left heart catheterization only code is for all scenarios, the right heart only and combined right and left heart catheterization codes are divided into two codes each, normal vs abnormal connections. As the name implies, the codes for normal connections are designed to be used for straightforward

defects (e.g. ASD, VSD, PDA, etc), where the connections from the vena cavae-right atrium-right ventricle-pulmonary arteries are normal for the right heart, and left atrium-left ventricle-aorta, for the left heart (atrio-ventricular and ventriculo-arterial concordance and normal levocardia). The codes for abnormal connections are designed to capture the patients with a more complex congenital heart anatomy involving any abnormal atrio-ventricular or ventriculo-arterial connections, such as variations of single ventricle anatomy, transposition, dextrocardia, etc. Importantly, post-surgical connections through shunts/conduits are considered abnormal connections (e.g. Blalock Taussig or Sano shunts, Glenn and Fontan patients, and those with homografts). Soon after these codes pass the RUC process and values are assigned, CICCW has planned for an SCAI webinar to explain the codes in detail and answer any questions providers may have. The webinar should take place in late 2020 and will likely be repeated in late 2021 as a refresher prior to the codes being available for use on January 1, 2022. This further delay adds another layer to the examples provided in **Figure 1**, caused by the printing process, as the 2021 CPT® book has already been printed and soon to be released.

Finally, for the current project at the CPT® application stage, we will be introducing new transcatheter repair of aortic coarctation codes this October, just as this edition of *Congenital Cardiology Today* comes out in print. This application includes three new codes, two for coarctation stenting and one for angioplasty alone. The stent codes are divided into two categories: one for involvement of any major side branches (such as transverse arch stenting or stenting of abdominal aorta for mid-aortic syndrome), and the second code for the more typical juxta-ductal coarctation where no head and neck vessels are involved. For consistency, this model was copied from the one used for endovascular graft for aortic aneurysms, designed to rightfully capture the increased complexity of stenting the more complex defects. Finally, a code for balloon angioplasty of aortic coarctation has been designed for all scenarios and with the intent of capturing all of the work performed as compared to the peripheral extremity angioplasty code (CPT® 37246) that we have been relegated to for so many years. Please refer to **Table 1** for a timeline of this project as well as future projects.

The CICCW would like to extend a special thanks to the leadership at SCAI for their continued support of this project. Additionally, our gratitude to Dr. Robert Vincent for his contributions to the work of the CICCW. Dr. Vincent has rotated off the workgroup and is being replaced by Dr. John Rhodes.

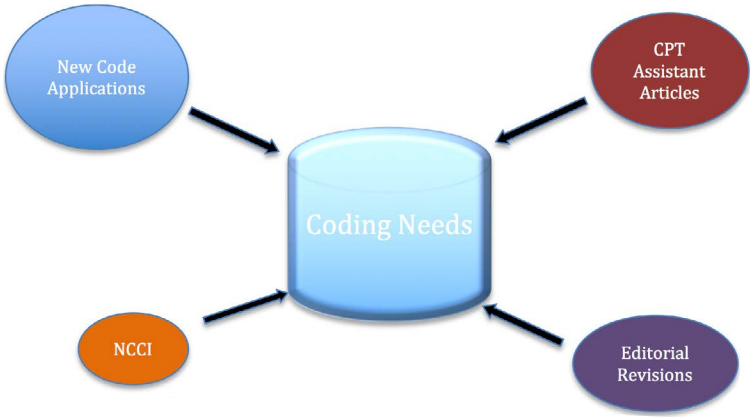

FIGURE 2 *Various pathways for resolution towards individual coding needs*

TABLE 1 *Project Timelines*

Project	CPT Meeting Deadline	CPT Meeting Presented	RUC Survey	RUC Meeting Presented	Status	When Available	New code (value)
Pulmonary artery angiography with device closure bundles		June 2017	n/a	n/a	Passed	Dec 2017	n/a
Directed by CMS to create new pericardiocentesis codes		Sep 2018	Nov 2018	Dec 2018	Passed	1-Jan-2020	33016 (4.4), 33017 (4.62), 33018 (5.4)
Modifier 63 for infants under 4 kg		Sep 2018	n/a	n/a	Passed	Jun 2019	n/a
Atrial septostomy	Nov 2019	Feb 2020	Feb 2020	Apr 2020	Passed	1-Jan-2021	33XX0*
Intracardiac stenting	Nov 2019	Feb 2020	Feb 2020	Apr 2020	Passed	1-Jan-2021	33XX1* and 33XX2*
ICE for all use with all cardiac cath procedures		May 2020	n/a	n/a	Passed	1-Jan-2021	
Congenital diagnostic catheterization base codes	Feb 2020	May 2020	Jun 2020	Oct 2020		1-Jan-2022	
Stent / angioplasty for repair of coarctation of the aorta	Jun 2020	Oct 2020	Oct 2020	Jan 2021		1-Jan-2022	
Pulmonary artery ductal stenting	Feb 2021**	May 2021**	Jun 2021**	Oct 2021**	Drafted		
Intra-cardiac / vascular RF perforation					First draft		
Possible Hybrid procedure with co-surgeon code					Discussions		
Completed	* Final code number to be determined, values approved and soon to be released						
In process / scheduled	** Anticipated dates barring any delays						

References

1. https://www.cms.gov/Medicare/Coding/NationalCorrectCodInitEd/index

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UC Davis School of Medicine
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Co-director, Pediatric Heart Center
UC Davis Children's Hospital

Jorge E. Suarez-Cavelier, MD, FACS is Recognized by Continental Who's Who

PRNewswire/ -- Jorge E. Suarez-Cavelier, MD, FACS, is being recognized by Continental Who's Who as a Top Surgeon for his remarkable contributions in the field of Medicine and acknowledgment of his dedication and commitment as a Cardiothoracic Surgeon with the Advent Health Cardiovascular Institute.

Proudly serving Orlando, Advent Health is committed to delivering quality care to patients from everyday wellness and preventive health care, to life-saving diagnostic services and innovative medical treatments in cancer, and heart failure through the Advent Health Cardiovascular Institute. Patients have direct access to some of the most advanced cardiovascular treatment options available at one of America's most experienced cardiac hospitals.

Board-certified Medical Director and Cardiothoracic Surgeon, Dr. Jorge E. Suarez-Cavelier has garnered a laudable reputation for his vast repertoire of expertise and professional experience in structural heart disease and heart transplants. Fluent in English and Spanish, he offers services in those languages. He has served as Chief of the Department of Cardiothoracic Surgery for over 10 years performing cardiothoracic and vascular surgeries and providing the highest standard of patient-centered care. More specifically, he diagnoses and treats a collection of heart diseases that includes heart failure, coronary artery disease, hypertrophic cardiomyopathy, and congenital heart disease that is obtained through wear and tear, or heart disease that people are born with.

An academic scholar, Dr. Suarez-Cavelier received his Medical Degree and completed his general surgical residency from Javeriana University in Bogota, Columbia. He relocated to the United States for a general surgical residency at Maimonides Medical Center in New York. In light of his academic success he was invited for a Fellowship in vascular and cardiac surgery at Caroline Medical Center, Charlotte, NC, and later invited for a Fellowship in cardiothoracic surgery with Methodist Hospital, Baylor University, Houston, Texas. He has also completed a Fellowship in cardiac surgery under Dr. Albert Starr in Portland, Oregon.

Remaining abreast of the latest developments in his field, he remains a Fellow of the American College of Surgeons and is a member with the Cardiac Surgery Association, Society of Thoracic Surgeons and the Southern Society of Thoracic Surgeons.

Dr. Jorge E. Suarez-Cavelier dedicates his success to his mentors Michael DeBakey, MD, and Francis Robichek, MD.

Offering Hope to Families of Infants with Congenital Differences

BEST CHILDREN'S HOSPITALS U.S. News & World Report RANKED IN 5 SPECIALTIES 2020-21

Nicklaus Children's Hospital's Fetal Care Center is dedicated to families expecting an infant who will need medical attention immediately after birth. The program offers comprehensive, coordinated care from prenatal diagnosis to delivery, postnatal care and the transition to infant care.

Nurse navigators are available to optimize care access and serve as points of contact for the expectant family, community physicians, pediatricians, perinatologists and neonatologists, as well as coordinate the subspecialty consultations.

Nicklaus Children's 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's subspecialty programs are routinely ranked by *US. News & World Report* in its annual listing of "*Best Children's Hospitals*" in the nation.

We have implemented additional safety precautions, such as arrival screenings and physical distancing measures. For parents who wish to make arrangements for their child to see his or her specialty physician virtually, telehealth options may also be available. For more information visit **nicklauschildrens.org/covid19safety**

The Fetal Care Center's special delivery unit is a birth center for healthy mothers of high-risk infants who will require immediate access to our team of pediatric intensive and critical care specialists after birth.

- **2** High-technology operating rooms
- **5** Antepartum rooms
- **5** Labor and delivery rooms

Nicklaus Children's Hospital
Fetal Care Center

To reach a Fetal Care Center nurse navigator, available 24 hours a day, call 1-844-553-3825 | Nicklauschildrens.org/FetalCare

Nicklaus Children's Hospital. For Health. For Life.

Two-Year-Old Palestinian "Miracle Boy" Returns Home After More Than Two Months in Critical Condition in Israel

The Israeli medical team of Save a Child's Heart fought for Hamza's life, treating him and functioning as his parents, while his parents had to stay at home in Ramallah, Palestine due to COVID-19 lockdown

PRNewswire - Hamza was born with life-threatening Congenital Heart Disease and had his first surgery as an infant. The Save a Child's Heart (SACH) medical team decided a few months ago that he needed a second surgery to survive.

On February 24, 2020, Hamza underwent open heart surgery at Wolfson Medical Center (WMC) in Holon, Israel, by the SACH volunteer medical team.

His recovery from this surgery was very complicated. "We tried to extubate Hamza a few times after his surgery, but he had a hard time breathing by himself. We had to connect him to an ECMO machine, and it kept him alive for a few weeks," said Dr. Racheli Sion Sarid, a senior physician at the Pediatric Intensive Care Unit at Wolfson.

In addition to the huge challenge of keeping Hamza alive through this period, the medical team had to face another major difficulty: Hamza's parents, who went home to the West Bank city of Ramallah for a short visit to see Hamza's siblings, could not return to Israel due to the COVID-19 lockdown.

"The whole medical team in the Pediatric Cardiology unit became his parents," said Dr. Ahmed Amer, a Pediatric Resident at WMC. Dr. Amer, an Israeli Arab physician who is part of the SACH medical team, became the main contact person for Hamza's parents since he speaks Arabic. "We all became his family. The nurses did shifts to hold him in their arms and play with him. We did not keep him alone for a minute. A child his age and in his condition needs to be hugged and loved in order to recover and get stronger, and that's exactly what we did."

Dr. Amer spoke with Hamza's parents every day, sent them pictures and videos and updated them on his recovery.

As Hamza's condition improved, he was able to communicate with his parents through video calls. "The first few times he saw them on video he began crying and it took us a long time to calm him down," said Dr. Amer, "But he got used to it and we even celebrated his birthday a couple of weeks ago together with his parents on video."

Hamza is now walking, laughing, and playing, and soon he will be ready to return home to his family in Ramallah. The medical team at the hospital are preparing to say goodbye, knowing they will never forget him and his story.

"His story is amazing," says Dr. Sarid. "It is a story about a medical team who fought tirelessly for the life of a critically ill child, a story of a brave little boy who survived against all odds, and a story of people, of human beings, helping each other regardless of their origin and religion. This is the story of Save a Child's Heart."

Palestinian child, Hamza, and one of the nurses taking care of him.

About Save a Child's Heart

SACH is an Israeli-based international non-profit organization that has provided care to more than 5,400 children from 62 countries in Africa, the Middle East, Asia, Europe, and South America and trained more than 120 medical professionals from these countries.

SACH is committed to the health and welfare of all children, regardless nationality, religion, color, gender, or financial situation.

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CONGENITAL HEART INTERNATIONAL PROFESSIONALS

The congenital heart professionals network exists to facilitate communications between congenital heart professionals locally, regionally, and globally.

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