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Quality Improvement in Pediatric Cardiac Catheterization: An Update

By Robert Vincent, MD

Introduction

Improving outcomes for patients living with congenital heart disease is a national priority. In 2012, the Centers for Disease Control and Prevention (CDC) tasked experts to identify the major public health gaps related to congenital heart disease and to suggest methods for addressing these gaps.[1] According to the CDC expert consensus, "Simply identifying the key strategies to improve quality will not be enough; these efforts must be widely disseminated, adopted, and tracked."[1]

To date, participation in national data registries, quality initiatives, and accreditation programs related to pediatric congenital heart disease is voluntary. In the future, however, healthcare providers may face various incentives and penalties—financial and otherwise—designed to encourage the implementation of quality improvement initiatives.[1] Therefore, it is vital for all members of the pediatric cardiac catheterization team to understand how quality metrics are defined, how new and emerging initiatives are shaping the standards of quality care in congenital cardiology, and how to become involved in quality improvement efforts.

Quality Benchmarking: The Role of Congenital Heart Disease Registries

Historically, the standardization of care for patients with congenital heart disease has lagged behind that for patients with acquired heart disease, due to a lack of national registries, clinical guidelines, certification of specialty training, and other barriers.[2] The first step toward standardizing care involves establishing baseline values for procedural safety and efficacy. Baseline values are important for comparing quality across facilities and for assessing the impact of quality-improvement efforts.

Several multicenter registries have been collecting procedural data from participating pediatric cardiac catheterization laboratories since the early-1990s, with the goal of establishing benchmarks for quality care. As registry data begin to mature, the congenital cardiology community can begin to examine current performance and set goals for future improvement. In 2014, each of the major congenital heart disease registries made significant contributions to defining the standards of care in congenital cardiology.

Improving Pediatric and Adult Congenital Treatment (IMPACT)

Recognizing the need to evaluate long-term outcomes associated with congenital interventions, the American College of Cardiology (ACC) established the National Cardiovascular Data Registry (NCDR), the first national voluntary data registry for pediatric and adult patients with congenital heart disease. With input from multiple professional organizations such as the American Academy of Pediatrics and the Society for Cardiac Angiography and Interventions (SCAI), the NCDR launched the Improving Pediatric and Adult Congenital Treatment (IMPACT) Registry (https://www.ncdr.com/webncdr/impact/) in 2010 with 2 participating sites. By 2013, the IMPACT Registry grew to include 81 sites with more than 26,000 patient records, exceeding the enrollment of other current and historical congenital catheterization laboratories.[3] As of March 2015, the participation list includes 90 facilities and more than 40,000 records.[4]

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In December 2014, the IMPACT Registry published its first major report based on an analysis of 4,152 catheterization laboratory visits occurring from January 2011 through March 2013. The report described current practice patterns, procedural outcomes, and safety results associated with 6 common congenital interventional procedures: device closure of atrial septal defect (ASD), device closure of patent ductus arteriosus (PDA), pulmonary valvuloplasty, aortic valvuloplasty, coarctation of the aorta (CoA) angioplasty and stenting, and pulmonary artery (PA) stenting.[3] The vast majority of ASD and PDA procedures (>98%) resulted in intended procedural outcomes. By comparison, the success rates for other procedures were lower, with CoA angioplasty being the least successful (51%). Across all procedures, the total adverse event (AE) rates ranged from 5.3% to 24.3%, and the major AE rates ranged from 0% to 3.3%. Although long-term data will be necessary for further insight, these baseline findings provide a solid foundation for developing quality metrics and identifying predictors of procedural success.

In an editorial accompanying the IMPACT Registry report, Michael Landzberg, MD (Director of Boston Children's Heart Center and Boston Adult Congenital Heart Program, Boston) describes the IMPACT Registry as a "game changer" for congenital cardiology.[2] As Dr. Landzberg writes, the initial efforts from the IMPACT Registry, including its preliminary findings, "confirm that the congenital cardiology field has matured and come of age, and can now can now tackle some of its most difficult questions relating to outcomes, quality and worth." Multiple additional reports from the IMPACT Registry, currently in press or under review, are poised to define specific further standards of care in pediatric congenital cardiology.

Congenital Cardiac Catheterization Project on Outcomes (C3PO)

The Congenital Cardiac Catheterization Project on Outcomes (C3PO) was founded in 2006 with the goal of developing outcomes measures for patients undergoing catheterization and other intervention for congenital heart disease.[5] The group's first projects involved analyzing registry data collected from 8 participating institutions to quantify baseline adverse event rates, identify indicators of hemodynamic vulnerability, and define 4 procedure-type risk categories.[6,7] Based on these findings, the C3PO group developed the CHARM (Congenital Heart Disease Adjustment for Risk Method) risk-adjustment model, which provides a mechanism for comparing adverse event rates across institutions performing catheterization for congenital heart disease.[8] In 2012, the National Quality Forum (NQF) endorsed the CHARM adverse event ratio as a pediatric quality measure.[9]

In 2013, the C3PO group launched the C3PO-Quality Improvement (C3PO-QI) initiative and expanded their multicenter collaboration to include 15 institutions. The primary objective of the C3PO-QI project is to effect meaningful change in patient care by reducing radiation exposure.[10] Toward that goal, the C3PO-QI collaborative conducted the first study of procedure-specific radiation doses associated with 6 common catheterization procedures for congenital heart disease (ASD, PDA, pulmonary stenosis, aortic stenosis, CoA, and transcatheter pulmonary valve replacement).[11] In the study of 2,713 procedures conducted in 15 participating catheterization laboratories, investigators found that radiation exposure varied widely by age group and procedure type. Notably, total fluoroscopy time, alone is a poor marker of radiation exposure, indicating the need for alternate tools for radiation safety monitoring. Furthermore, these findings will facilitate the implementation of an ACC-endorsed quality metric that measures the proportion of patients receiving procedure-specific radiation doses in excess of the 95th percentile. Moving forward, the C3PO-QI investigators will conduct biannual assessments of radiation exposure to measure the impact of various quality-improvement initiatives focused on radiation risk reduction.

Congenital Cardiovascular Interventional Study Consortium

The Congenital Cardiovascular Interventional Study Consortium (CCISC) (http://www.ccisc.net/) established the CCISC Risk Registry in 2008. As of 2014, twenty-one participating sites have contributed data on 20,229 patient cases, including information on patient demographics, procedure types, hemodynamic data, radiation

dosage, and complications.[12] The primary goal of the CCISC Risk Registry is to identify opportunities to reduce harm among patients undergoing catheterization for congenital heart disease.

The CCISC Risk Registry is yielding important insights with potential implications for patient care. At the 2014 SCAI annual meeting, Thomas J. Forbes, MD, (Professor of Pediatrics at Wayne State University School of Medicine, Detroit) presented findings from an analysis of risk factors for femoral artery (FA) compromise among patients treated between 2008 and 2013 at pediatric cardiac catheterization laboratories participating in the CCISC Risk Registry.[12]

During a total of 8,958 cardiac catheterization procedures utilizing FA access, the incidence of FA compromise was 1.1%. However, the incidence was significantly higher (5.9%) for infants <12 kg. In a subgroup analysis of infants <12 kg, Dr. Forbes and colleagues identified 2 steps that substantially reduced the risk of FA obstruction for these vulnerable patients by more than two-thirds. First, the use of a size 3 French (1 mm diameter) sheath decreased the risk of FA compromise by 73%, compared with the use of a size 4 French (1.35 mm diameter) sheath (OR, 0.27; p=0.001). Second, the use of a hemostasis patch to seal the arterial puncture site reduced the risk of FA compromise by 68% compared with manual pressure alone (OR, 0.32; p=0.002).

The CCISC Risk Registry will continue to establish safety benchmarks in future database analyses, with the goal of identifying opportunities to reduce harm in the pediatric cardiac catheterization laboratory. To encourage cardiologists and other providers to contribute procedural data, the American Board of Pediatrics grants maintenance of certification (MOC) credit for participation in the CCISC Risk Registry.[9]

Implementing Quality: Pediatric SCAI-Quality Improvement Toolkit

In 2014, SCAI launched the Pediatric SCAI-Quality Improvement Toolkit (SCAI-QIT) (http://www.scai.org/pedqit/default.aspx) to facilitate the adoption of quality and safety standards in pediatric cardiac catheterization laboratories. The Pediatric SCAI-QIT complements the adult SCAI-QIT for catheterization laboratories, but with a focus on the specific clinical, safety, and quality issues surrounding pediatric congenital heart disease.

"Just as pediatric care requires a dedicated approach, pediatric quality improvement requires specific tools and resources tailored to the unique medical needs of children," explains Henri Justino, MD, Chair of the Pediatric SCAI-QIT, Director of the CE Mullins Cardiac Catheterization Laboratories at Texas Children's Hospital, and Associate Professor of Pediatrics at Baylor College of Medicine in Houston, Texas.[13]

The Pediatric SCAI-QIT provides guidance for physicians and cath lab staff on continuous quality improvement, accreditation, external peer review, and public reporting. The Pediatric SCAI-QIT includes four modules on key topics that are critical for delivering quality care in the pediatric cardiac catheterization laboratory:

Catheterization Conferences: This module provides an overview of the types of regular, inclusive, and non-punitive conferences that catheterization laboratories should conduct to facilitate continuous quality improvement. Although facilities can tailor the number and frequency of conferences, SCAI recommends the following key conferences:

1. Invasive Cardiology Morbidity and Mortality (cath lab M&M) Conference, which is separate from the clinical cardiology M&M and focuses on cath lab complications and in-hospital events following invasive procedures;
2. Invasive Case Review Conferences, which include an open review of a random sample of diagnostic and interventional cases; and
3. Catheterization Laboratory Educational Conferences, which are regular, frequent, and formal events that focus on cath lab practices, issues, and the management of complex patients.

Procedural Checklists: This module reviews the rationale for incorporating procedural checklists into every catheterization procedures, and summarizes key components of procedural checklists, and reviews real-world examples of checklists utilized at various pediatric cardiac catheterization facilities.

Procedural Quality: This module discusses the rationale and overriding goals of quality assessment and improvement in the

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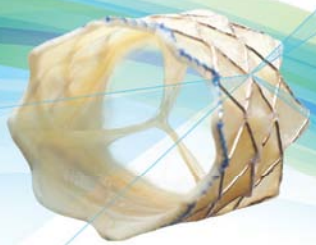

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pediatric cardiac catheterization lab. The module also provides guidance for facilities that wish to establish and run a 10-step procedural quality program.

Radiation Safety: This module describes the rationale and strategies for reducing radiation exposure to patients and staff in the pediatric cardiac catheterization lab. In addition, the module describes strategies for monitoring radiation exposure of staff, improving staff compliance with radiation monitoring and safety procedures, documenting lifetime cumulative radiation exposure, and conducting a follow-up program for the early identification of radiation injury.

The Pediatric SCAI-QIT modules also provide information on and access to the major congenital heart disease registries, as well as important benchmarks from registry data and other evidence-based resources. Physician authors will continuously update the Pediatric SCAI-QIT modules to ensure that the latest quality standards are reflected, and develop additional modules as new quality standards emerge.

According to Dr. Justino, the Pediatric SCAI-QIT modules "should be implemented on a voluntary basis by cardiac cath labs across the country and around the world with the goal of creating a culture of safety and quality." The full interview with Dr. Justino is available on the SCAI website: http://www.scai.org/Press/detail.aspx?cid=72528bf9-b7b0-4423-830e-ee8730a37ab0#.VTKcV8712Os.

Pediatric Congenital Heart Disease Catheterization Laboratory Accreditation

In 2014, ACE partnered with SCAI and ACC to evaluate the need for an accreditation program for pediatric congenital heart disease catheterization laboratories. At present, the preliminary quality standards for pediatric congenital heart disease catheterization remain under review. Once finalized, the consensus quality standards will form the foundation of pediatric congenital heart disease catheterization laboratory accreditation and quality-improvement initiatives. Accreditation is a rigorous process designed to ensure that facilities are meeting the highest standards of patient care. The Accreditation for Cardiovascular Excellence (ACE) (http://www.cvexcel.org/) is an independent, physician-led organization sponsored by SCAI and ACC that provides accreditation, external peer review, and other customized services exclusively for the cardiac catheterization laboratory.[14] With years of leadership in the adult interventional cardiology setting, ACE has established accreditation programs for diagnostic cardiac catheterization, percutaneous coronary intervention (PCI), carotid artery stenting, and peripheral vascular intervention services.

Summary

Continuous quality improvement describes the ongoing cycle of measuring performance data to identify, develop, and implement strategies for improved care. With the identification of new performance benchmarks through registry studies and other initiatives, the quality standards in pediatric interventional cardiology are rapidly evolving. Consensus quality standards are currently under review, and may form the basis of the first accreditation program for pediatric congenital heart disease catheterization laboratories. Cardiologists, surgeons, and other healthcare providers now have access to tools and resources to ensure the delivery of the highest quality care to patients with congenital heart disease. In addition, pediatric cardiac catheterization laboratories have several options for getting involved with quality improvement efforts, including enrolling in the IMPACT Registry or other national data registries, implementing tools and recommendations in the Pediatric SCAI-QIT, and achieving accreditation for pediatric congenital heart disease services.

Biographical Sketch

Robert Vincent, MD, CM, FACC, FSCAI, is a Professor of Pediatrics at Emory University School of Medicine and a Pediatric Cardiologist at Children's Healthcare of Atlanta in Atlanta, Georgia. He currently serves as the President of the Georgia Chapter of the American College of Cardiology, and is Past Chairman of the Congenital Heart Disease Committee of the Society of Cardiac Angiography and Intervention.

References

1. Oster ME, Riehle-Colarusso T, Simeone RM, et al. Public health science agenda for congenital heart defects: report from a Centers for Disease Control and Prevention experts meeting. J Am Heart Assoc. 2013;2(5):e000256.
2. Landzberg MJ. The impact of IMPACT: a game changer for congenital cardiology. J Am Coll Cardiol. 2014;64(23):2452-2454.
3. Moore JW, Vincent RN, Beekman RH 3rd, et al. Procedural results and safety of common interventional procedures in congenital heart disease: initial report from the National Cardiovascular Data Registry. J Am Coll Cardiol. 2014;64(23):2439-2451.
4. Improving Pediatric and Adult Congenital Treatment (IMPACT) Registry. Available online at: https://http://www.ncdr.com/webncdr/impact/.
5. Bergersen L, Marshall A, Gauvreau K, et al. Adverse event rates in congenital cardiac catheterization—a multi-center experience. Catheter Cardiovasc Interv. 2010;75(3):389-400.
6. Bergersen L, Gauvreau K, Marshall A, et al. Procedure-type risk categories for pediatric and congenital cardiac catheterization. Circ Cardiovasc Interv. 2011;4(2):188-194.
7. Lin CH, Hegde S, Marshall AC, et al. Incidence and management of life-threatening adverse events during cardiac catheterization for congenital heart disease. Pediatr Cardiol. 2014;35(1):140-148.
8. Bergersen L, Gauvreau K, Foerster SR, et al. Catheterization for Congenital Heart Disease Adjustment for Risk Method (CHARM). JACC Cardiovasc Interv. 2011;4(9):1037-1046.
9. Barach P, Jacobs J, Lipshultz SE, Laussen P, eds. Pediatric and Congenital Cardiac Care: Volume 1: Outcomes Analysis. Springer-Verlag London. 2015.
10. Chaudhry-Waterman N, Coombs S, Porras D, Holzer R, Bergersen L. Developing tools to measure quality in congenital catheterization and interventions: the congenital cardiac catheterization project on outcomes (C3PO). Methodist Debakey Cardiovasc J. 2014;10(2):63-67.
11. Ghelani SJ, Glatz AC, David S, et al. Radiation dose benchmarks during cardiac catheterization for congenital heart disease in the United States. JACC Cardiovasc Interv. 2014;7(9):1060-1069.
12. Forbes T, Kobayashi D, Moore P, Nykanen D. Femoral artery compromise from cardiac catheterization in children–a multicenter study by the CCISC (Congenital Cardiovascular Interventional Study Consortium). Presented at the 2014 SCAI Annual Meeting. May 28-31, 2014; Las Vegas, NV. Abstract O1-002.
13. Society for Cardiovascular Angiography and Interventions (SCAI). Pediatric SCAI-QIT. Available at: http://www.scai.org/PEDQIT/Default.aspx.
14. Accreditation for Cardiovascular Excellence (ACE). Available online at: http://www.cvexcel.org/.

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Total Anomalous Pulmonary Venous Connection: Outcomes of Surgical Correction and Management in a New Saudi Center

By Abdullah Alkhudhayri, MD; Ayed A. Shati, MD; Broering DC, MD; Jamil Azzi, MD; Antonio F. Corno, MD

Abstract

Objective

To see the outcome of surgical correction and management in a newly established centre in the Middle East among the patients with total anomalous pulmonary venous connections.

Introduction & Background

Total Anomalous Pulmonary Venous Connection (TAPVC) consists of an abnormality of blood flow in which all 4 pulmonary veins drain into systemic veins, or the right atrium, with or without pulmonary venous obstruction. How severe this condition is depends on whether the pulmonary veins are blocked or obstructed as they drain. Obstructed TAPVR causes symptoms early in life and can be deadly very quickly if it is not found and corrected with surgery.

Result

There were no early or late deaths in a mean follow-up of 16 months (range 7 to 24 months) in Middle Eastern patients with an anomalous pulmonary venous connection.

Conclusion

In a new unit the patients with TAPVC can be surgically managed with good results, even in the presence of associated complex malformations.

Key Words: Total anomalous pulmonary venous connection, modern centre.

Introduction

Total anomalous pulmonary venous connection consists of an abnormality of blood flow in which all 4 pulmonary veins drain into systemic veins or the right atrium with or without pulmonary venous obstruction. All pulmonary venous return connects to the systemic venous system, right atrial and right ventricular enlargement occurs, and, if significant pulmonary venous obstruction develops, right ventricular hypertrophy occurs. Total anomalous pulmonary venous connection occurs alone in two thirds of patients and occurs as part of a group of heart defects (eg, heterotaxy syndromes) in approximately one third of patients.[1] In some cases, it can be detected prenatally.[2] There are four variants:

1. **Supracardiac** (50%): blood drains to one of the innominate veins (brachiocephalic veins) or the superior vena cava;
2. **Cardiac** (20%), where blood drains into coronary sinus or directly into right atrium;
3. **Infradiaphragmatic** (20%), where blood drains into portal or hepatic veins; and,
4. **A mixed** (10%) variant.

How severe this condition is depends on whether the pulmonary veins are blocked or obstructed as they drain. Obstructed TAPVR causes symptoms early in life and can be deadly very quickly if it is not found and corrected with surgery. [3]

The result of the surgical correction of this anomaly is associated with acceptable morbidity and mortality, depending on early referral and surgery, without progression of the pulmonary vascular hypertension findings.[5]

Surgical repair is used as treatment for total anomalous pulmonary venous connection whenever it best serves the individual patient. Stabilizing the patient prior to surgery as much as possible from a cardiovascular and metabolic standpoint is important. [6,7]

Many infants who have had TAPVR surgical repair will grow and develop normally. However, after TAPVR repair, an infant will need to be followed periodically by a pediatric cardiologist who will make assessments to check for any heart-related problems. As some children grow, the area where the pulmonary arteries were reconnected to the left atrium may become narrowed, preventing blood from moving from the lungs to the left atrium. Treatment may include placement of a wire mesh device called a stent into the narrowed vein(s) to open it, which is done during a cardiac catheterization procedure or surgical enlargement of the narrowed pulmonary vein connection(s).[8-11]

Methods

Type of study; It is a retrospective, cross-sectional type of study.

Duration and Place of the Study: From October 2010 to March 2013 in the Heart Centre at King Fahad Medical City, Riyadh, Saudi Arabia.

Ethical Considerations: The World Medical Association's Ethical Principles for Medical Research Involving Human Subjects has been our standard. We have followed all those ethical principles i.e. protect the life, health, dignity, integrity, right to self-determination, privacy, and confidentiality of personal information of research subjects.

Type of Sampling: Non-probability, purposive sampling.

Inclusion criteria: All the patients with stated condition / diseases during the study duration were included in the study.

Exclusion Criteria: Exposure to any investigations, drug or procedure within 1 month prior to study entry or enrolled in a concurrent study that may confound results of this study.

List of Variables: Age, gender, mortality.

Data Analysis: Statistical Package for Social Sciences (SPSS) ver. 20 was used for entering and analysing the data. Data were coded for entering purpose.

Statistical Analyses: Descriptive statistics i.e. mean, standard deviation, percentages of variables, etc. were calculated. To check the significance differences among the variables statistical tests, i.e. chi square test, correlation test was used.

Level of Significance: We fixed the level of significance at 95%, which means any p-value less than 0.05 considered significant.

Practice points

In the new modern centre, patients with TAPVC can be surgically managed with good results, even in the presence of associated complex malformations.

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Results

- Figure 1 depicts the gender distribution to be: 57.14% male, 42.86% female.
- Table 1 shows the age distribution of the patients, and that the median age was 4-years-old.
- Table 2 shows the comparison between age and mechanical ventilation in hours / days shows the insignificant difference p-value =0.13, the median mechanical ventilation is 12 hours.
- Table 3 shows that 100% of the patients survived after the treatment.

Discussion

In TAPVC, the pulmonary vasculature often has a thickened medial layer; thus, Pulmonary Vascular Resistance (PVR) does not decrease normally after repair and the right ventricle has to pump against an increased after load. The left ventricle is unable to support the circulation probably because it is under filled and underutilized prior to correction. Its function is also hampered by septal displacement. Thus, the postoperative period is complicated by existence of a low output state, persistence of pulmonary hypertension (PHT) and a highly reactive pulmonary vasculature.[12] Our study results are in line with another study conducted by Hyde et al. demonstrated that total anomalous pulmonary venous connection (TAPVC) can be corrected with low mortality and good outcome. If complicated by Pulmonary Vein Stenosis (PVS), either at presentation or secondary to the repair, the long-term outcome is compromised. We have evaluated an institutional experience with TAPVC, with particular regard to the evolving management of PVS.[13]

Another study stated that TAPVC is a very uncommon cyanotic anomaly comprising 1% of all congenital heart disease, since the pulmonary veins drain into the systemic venous circulation.

TAPVC is incompatible with life unless a communication between the right and left sides of the heart exists; usually via a patent foramen ovale or atrial septal defectale or atrial septal defect.[12]

As in our study, some cases are also comes in the neonatal age in line with other study that in the neonatal period, the surgical mortality of palliation is extremely high for asplenia syndrome complicated by single ventricle combined with TAPVC.[14]

The overall survival rate of our study is almost in line with Karamlou et al, who recently reported an operative 5-year survival of 97% for patients undergoing repair at the Hospital for Sick Children in Toronto since 2000. The 3-year operative survival reported in the present study was seemingly poorer at 85%; this may reflect the population basis rather than institutional basis for the present study, which comprised the full range of morphology and/or surgical and medical management.[15-18]

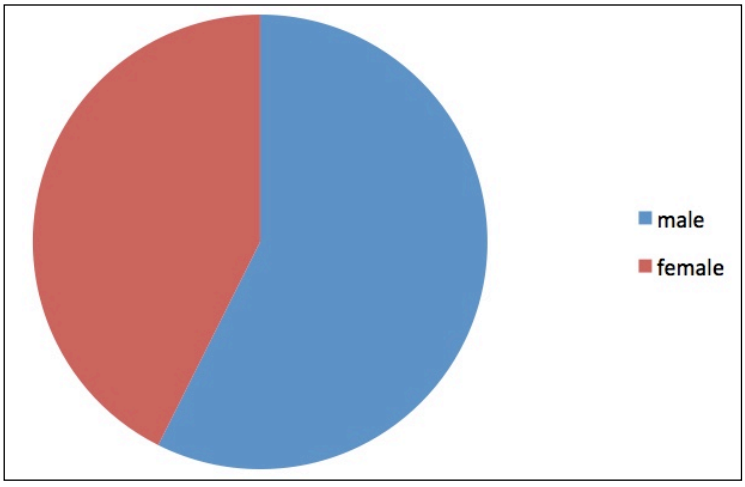

Figure 1: Gender Distribution: 57.14% of the patients were male while 42.86 were female.

Table 1. Age Distribution of the Patients and the Median Age was 9.70

S. No	Age distribution
1	4
2	12
3	1.6
4	4
5	0.3
6	0.1
7	27
Mean Age	7
Standard deviation	9.70
Median Age	4

Table 2: Comparison Between Age and Mechanical Ventilation in Hours / Days - Insignificant Difference P-value = 0.13, Median Mechanical Ventilation is 12 Hours

S. No	Age	Mechanical Ventilation in Hours / Days
1	4	0.1
2	12	16.5
3	1.6	360
4	4	7
5	0.3	6
6	0.1	48
7	7	12
Median	4	12
p-value = 0.130		

Table 3 : Survival Percentage - 100% Patients Survived after the Treatment

Survival upon follow-up (up to 1.8 years)		
	Frequency	% age
Survived	7	100%
Not survived	0	0%
Total	7	100%
Median	4	12

Declaration of Interest

No declarations of interest. We have not disclosed any affiliation or financial involvement with organization or entities with a direct financial interest in the subject matter or materials discussed in the manuscript.

Acknowledgment

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References

1. http://emedicine.medscape.com/article/899491-overview.
2. Raisher BD, Grant JW, Martin TC, et al. Complete repair of TAPVC in infancy. J Thoracic Cardiovasc Surg. 1992;104:443–448 [PubMed].
3. Webb GD, Smallhorn JF, Therrien J, Redington AN. Congenital heart disease. In: Bonow RO, Mann DL, Zipes DP, Libby P, eds. Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine. 9th ed. Philadelphia, Pa: Saunders Elsevier; 2011:chap 65.
4. Antonio F. Corno, Sami A. Alahda and Karuna Moy Das , Systemic venous anomalies in the Middle East , published: 26 February 2013 doi: 10.3389/fped.2013.00001.
5. Atik FA1, Irun PE, Barbero-Marcial M, Atik E Total anomalous drainage of the pulmonary veins-- surgical therapy for the infradiaphragmatic and mixed anatomical types. [PubMed].
6. http://emedicine.medscape.com/article/899491-treatment.
7. Morray JP, Lynn AM, Mansfied PB, et al. Effect of pH and PaCO2 on pulmonary and systemic hemodyanamics after surgery in children with congenital heart disease and pulmonary hypertension. J Pediatr. 1998;113:474–479. [PubMed].
8. Braunwald's Heart Disease—A Textbook of Cardiovascular Medicine. Bonow, Robert O., MD. 2012, 9th edition, pp. 1411–1467.
9. Nelson Textbook of Pediatrics. Kliegman, Robert M., MD. 2011, 19th edition, pp. 1549–1551.
10. Konstadt S. Nitric oxide: has it progressed from molecular year to wonder drug of decade? J Thorac Cardiovasc Surg. 1995;9:625–626. [PubMed].
11. Bando K, Tumentine MW, Sharp TG, et al. Pulmonary hypertension after operation for congenital heart disease: analysis of risk factors and management. J Thorac Cardiovasc Surg. 1996;112:1600–1607. [PubMed].
12. Sandeep Khanna, Minati Choudhury, and Usha Kiran Total Anomalous Pulmonary Venous Connection: Post Operative Problems and Management, Indian J Anaesth. 2009 Feb; 53(1): 71–74.
13. Middlebrooks E, Wymer D, Chandran A. Caseofpersistent left superior vena cava draining via the left atrium with a normal coronary sinus utilizing cardiac CT. J Cardiovascular Comput Tomogr. (2012) 6:289–91.
14. J A Hyde, O Stümper, M J Barth, J G Wright, J V de Giovanni, W J Brawn , B sethia ;Total anomalous pulmonary venous connection: outcome of surgical correction and management of recurrent venous obstruction. European Journal of Cardio-Thoracic Surgery (Impact Factor: 2.81). 07/1999; 15(6):735-40; discussion 740-1. DOI: 10.1016/S1010-7940(99)00104-9.
15. Takashi Higaki, Eiichi Yamamoto, Takeshi Nakano, Masaaki Ohta, Hidemi, Takata Kikuko, Murao, Toshiyuki Chisaka Tomozo Moritani, Mitsugi Nagashima, Fumiaki Shikata, Eiichi Ishii; Successful stenting of the ductus venosus in 2 neonates with asplenia syndrome complicated by infracardiac type total anomalous pulmonary venous connection; Journal of Cardiology Cases Volume 1, Issue 3, June 2010, Pages e129–e132.
16. Karamlou T, Gurofsky R, Al Sukhni E, Coles JG, Williams WG, Caldarone CA, Van Arsdell GS, McCrindle BW. Factors associated with mortality and reoperation in 377 children with total anomalous pulmonary venous connection. Circulation. 2007;115:1591–1598.
17. Koudieh M, McKenzie ED, Fraser CD. Outcome of Glennanasto-mosisforheterotaxy syndrome withsingle ventricle. Asian Cardiovasc Thorac Ann. (2006) 14:235–8.
18. Kula S, Cevik A, SanliC, Pektas A, TunagluFS, OguzAD,etal. Persistent left superior venacava: experience of tertiary health- care centre. Pediatr Int. (2011) 53:1066–9.
19. Curtil A,Tronc F,Champsaur G, Bozio A,Sassolas F,Carret JP, et al.Thelef tretro-aorticbra chiochephalicve in:morphologic data and diagnostic ultrasound in 27 cases Surg RadiolAnat. (1999)21:251–4.

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Medical News, Products & Information

Compiled and Reviewed by Tony Carlson, Senior Editor

Children's Hospital of Pittsburgh of UPMC - Congenital Cardiac Morphology Program Announcement

Children's Hospital of Pittsburgh of UPMC's 2015 Master Class to Focus on Univentricular Heart

Understanding the functionally univentricular heart is the topic of the *2015 Master Class in Congenital Cardiac Morphology* to be held at Children's Hospital of Pittsburgh of UPMC, Sep. 30th to Oct. 2nd.

"Single ventricles are the most complex congenital heart defects and the ones that we find to be the most challenging to diagnose accurately in our pediatric patients," said Vivek Allada, MD, Co-Director of Children's Heart Institute, one of the event sponsors.

Robert Anderson, MD, FRCPath, professorial fellow at Newcastle University's Institute of Genetic Medicine and visiting professor of pediatrics, Medical University of South Carolina (MUSC)

Now in its eighth year, the annual event draws an international audience ranging from cardiologists, surgeons, and cardiac interventionists to nurse practitioners, medical students, device makers, and medical sonographers. Held at the John G. Rangos Sr. Conference Center, located on Children's campus, the event is co-sponsored with Children's Division of Pediatric Pathology and the University of Pittsburgh School of Medicine's Center for Continuing Education in the Health Sciences.

Featuring multiple lecturers, the event will be again moderated by Robert Anderson, MD, FRCPath, professorial fellow at Newcastle University's Institute of Genetic Medicine and visiting professor of pediatrics, Medical University of South Carolina (MUSC). This year's sessions will spotlight lesions such as pulmonary atresia-intact ventricular septum, Hypoplastic Left Heart Syndrome (HLHS), double inlet ventricle, atrioventricular valve atresia, and straddling atrioventricular valves, as well as isomerism in the setting of the functionally univentricular heart.

Through the program, participants will receive a multimodal look at all aspects of the developing heart using the latest imaging technologies (3D echocardiography and CT angiography) and hands-on demonstrations of pathologic specimens of each lesion. In addition, developmental embryology, clinical treatment for both cardiac catheterization and cardio thoracic surgery will be covered in depth.

"The strength of the course is that it gives a sound fundamental basis for the understanding of congenital heart disease," said Allada, adding, "The survival for congenital heart disease has vastly improved over the past decade, and much of that can be

PEDIATRIC ECHOCARDIOGRAPHY

OCHSNER HEALTH SYSTEM in **NEW ORLEANS** is searching for a **BC/BE PEDIATRIC CARDIOLOGIST** to join the Section of Pediatric Cardiology. The section is recruiting a **Pediatric Cardiologist specializing in non-invasive imaging with expertise in fetal echo, TEE, MRI, and/or Cardiac CT** to join 3 other imaging physicians in a group of 9 pediatric cardiologists. Completion of an advanced non-invasive imaging fellowship is preferred. Salary with be commensurate with training and experience.

The Department of Pediatrics and our pediatric subspecialists (collectively, Ochsner for Children™) represent over 110 physicians. Our pediatric subspecialties include: Allergy, Cardiology, CV Surgery, Critical Care Medicine, Dermatology, ENT, Endocrinology, Emergency Medicine, Gastroenterology, Hematology/Oncology, Hospital Medicine, Infectious Diseases, Medical Genetics, Neonatology, Nephrology/Hypertension, Neurology, Neurosurgery, Ophthalmology/Optometry, Orthopedics, Physiatry/Sports Medicine, Plastic and Reconstructive Surgery, Psychiatry, Pulmonology, General Surgery, Transplant, and Urology. We also have a Pediatric Anesthesiology Team, Pediatric CV Surgery Anesthesia Team and Pediatric Radiology support. We support a pediatric heart failure program, including left ventricular assist devises and transplantation. We also have a pediatric transcatheter valve program. We utilize a 54 bed Level III-C NICU and 14 bed PICU as well as a dedicated, full-time, region-wide in-house pediatric transport team.

Ochsner is the region's leading integrated provider of multi-specialty care for infants, children and adolescents. Our physicians practice in multiple sites across Louisiana, including a large, state-of-the-art dedicated pediatric ambulatory facility on our main campus. We have an outstanding primary care pediatric network across the region with over 40 general pediatricians, in addition to a large outside referral base. Ochsner pediatric physicians care for over 55,000 unique pediatric patients annually, and Ochsner hospitals have more than 4,000 deliveries per year.

Ochsner Health System is Louisiana's largest non-profit, academic, healthcare system. Driven by a mission to Serve, Heal, Lead, Educate and Innovate, coordinated clinical and hospital patient care is provided across the region by Ochsner's 19 owned, managed and affiliated hospitals and more than 50 health centers. Ochsner is the only Louisiana hospital recognized by U.S. News & World Report as a "Best Hospital" across nine specialty categories caring for patients from all 50 states and more than 90 countries worldwide each year. Ochsner employs more than 16,000 employees, nearly 1,000 physicians in over 90 medical specialties and subspecialties and conducts over 750 clinical research studies. Ochsner Health System is proud to be a tobacco-free environment. For more information, please visit www.ochsner.org and follow us on Twitter and Facebook.

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The Department of Pediatrics and pediatric subspecialists (collectively, Ochsner for Children™) is comprised of 110 physicians with specialties including: General Pediatrics, Allergy, Cardiology, CV Surgery, Critical Care Medicine, Dermatology, ENT, Endocrinology, Emergency Medicine, Gastroenterology, Hematology/Oncology, Hospital Medicine, Infectious Diseases, Medical Genetics, Neonatology, Nephrology/Hypertension, Neurology, Neurosurgery, Ophthalmology/Optometry, Orthopedics, Physiatry/Sports Medicine, Plastic and Reconstructive Surgery, Psychiatry, Pulmonology, General Surgery, Transplant, and Urology.

Congenital Cardiovascular Program support includes a pediatric CV Surgery Anesthesia Team, perfusion and pediatric radiology. We support comprehensive catheter intervention and EP services including a pediatric transcatheter valve program and a pediatric heart failure program that includes percutaneous and surgical circulatory assist devices and transplantation. We utilize a 54 bed Level III-C NICU and 14 bed PICU as well as a dedicated, full-time, region-wide in-house pediatric transport team.

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attributed to the work of people like Professor Anderson giving us a greater understanding of how the heart is malformed and how we can best treat it."

Registration details will be provided online at www.chp.edu/masterclass in June, with enrollment limited to 75. The program has been approved for AMA PRA Category 1 continuing education credits. For more information, contact Lynda Cocco, 412-692-3216, at the Heart Institute at Children's Hospital of Pittsburgh of UPMC.

First Patient Enrolled in GORE® TAG® Thoracic Branch Endoprosthesis Early Feasibility Study - First Early Feasibility Study of a Gore Device Will Assess Treatment of Aortic Arch Aneurysms

W. L. Gore & Associates (Gore) has announced that Michel Makaroun, MD, Co-Director of the University of Pittsburgh Medical Center (UPMC) Heart and Vascular Institute and the Chair and Professor of Surgery in the division of Vascular Surgery, enrolled the first brachiocephalic treated patient in the GORE® TAG® Thoracic Branch Endoprosthesis Early Feasibility Study. The US-based study will assess the treatment of aortic arch aneurysms in which perfusion of the brachiocephalic or left common carotid artery is maintained through a branch device.

"Working with Gore on this early feasibility study gives us the opportunity to explore the GORE TAG Thoracic Branch Endoprosthesis as a potentially new less invasive treatment option that will greatly benefit this patient population."

Principal Investigator of the study, Michael Dake, MD, Thelma and Henry Doelger Professor of Cardiovascular Surgery at the Stanford School of Medicine, will present on the study this week at the Charing Cross (CX) International Symposium in London. The study represents Gore's first use of a recent Food and Drug Administration (FDA) early feasibility study guidance to help patients gain access to breakthrough medical devices in order to maintain innovation of new technologies in the US.

"Aortic arch aneurysms present physicians with a very difficult challenge, as no other approved endovascular method exists to treat this region," said Dr. Makaroun. "Working with Gore on this early feasibility study gives us the opportunity to explore the GORE TAG Thoracic Branch Endoprosthesis as a potentially new less invasive treatment option that will greatly benefit this patient population."

Currently, treatment of aortic arch aneurysms is performed either through complex open surgical repair or through hybrid procedures involving the combination of devices whose use in this application has yet to be approved by the FDA and still requires a thoracotomy or incision of the sternum. In contrast to these alternatives, the GORE TAG Thoracic Branch Endoprosthesis offers a less invasive treatment option specifically designed for these challenging cases.

"Over the course of my career, I have witnessed a sustained focus on the development of less invasive approaches to aortic arch aneurysms,"

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said Dr. Dake. “This clinical trial represents a culmination of years of research and development, beginning with the custom fabrication of a device to meet a specific patient's anatomy and now reaching a more practical off-the-shelf solution, which I am confident will improve outcomes for patients.”

Designed for long-term durability, the GORE TAG Thoracic Branch Endoprosthesis is an off-the-shelf device, which includes aortic and branch components constructed specifically for use in the arch. The device allows for femoral-only access over a pre-positioned branch guidewire designed to minimize the risk of branch vessel coverage and improve ease of implantation. The branch device also features the CBAS® Heparin Surface. This technology, used in many of Gore’s peripheral products, is intended to provide improved thromboresistance within the branch component of the device.

“This feasibility study is a testament to our longstanding commitment to innovation in new products and pathways, and to bringing products to market quickly,” said Ryan Takeuchi, Gore Aortic Business Leader. “We are the first to investigate this indication with an off-the-shelf endovascular device. We are meeting this challenge using our aortic branch portfolio to address this new and rather difficult clinical application.”

Gore has met several other key milestones in its commitment to pursuing new clinical applications with its aortic branch and thoracic portfolio. In 2014, Gore initiated a clinical study evaluating the use of the GORE TAG Thoracic Branch Endoprosthesis for the treatment of thoracic aortic aneurysms that require coverage of the left subclavian artery (LSA). Gore also enrolled its first patient in the Gore Thoracoabdominal Aortic Aneurysm Clinical Study, which will assess the durable repair of aortic aneurysms encroaching on or involving visceral branch vessels using the GORE® EXCLUDER® Thoracoabdominal Branch Endoprosthesis. Enrollment in the Gore® EXCLUDER® Iliac Branch Clinical Study was completed in February 2015. Additionally, the FDA approved the Conformable GORE® TAG® Thoracic Endoprosthesis for the treatment of acute and chronic dissections in September 2013, making it the first stent-graft approved to treat aneurysms, traumatic transections, and Type B dissections of the thoracic aorta.

Visit the Gore aortic portal (www.goremedical.com/aortic/#innovation) for additional information.

For more information on Gore Medical devices, visit www.goremedical.com.

Identify 'Missing Culprit' in Heart Failure

Newswise - Working with lab animals and human heart cells, scientists from Johns Hopkins and other institutions have identified what they describe as “the long-sought culprit” in the mystery behind a cell-signaling breakdown that triggers heart failure. The condition, which affects nearly 6 million Americans and 23 million people worldwide, is marked by progressive weakening and stiffening of the heart muscle and the organ’s gradual loss of blood-pumping ability.

The research results, described in the March 18th journal *Nature*, reveal that an enzyme called PDE-9 interferes with the body's natural "braking" system needed to neutralize stress on the heart. The experiments demonstrate that the enzyme wreaks mischief by gobbling up a signaling molecule, cGMP, which normally stimulates the production of a heart-protective protein called PKG, known to shield the heart muscle from the ravages of disease-causing stress, such as long-standing high blood pressure.

Naturally found in the gut, kidneys and brain, PDE-9 is already a prime suspect in neurodegenerative conditions such as Alzheimer's, the researchers say. But the new study shows the enzyme's footprints are also present in heart cells and markedly elevated in patients with heart failure — evidence that PDE-9 is a multitasking "offender" and a key instigator of heart muscle demise, the researchers say.

To understand the enzyme's role, the scientists exploited the knowledge that heart muscle health is safeguarded by two separate mechanisms, or signaling pathways. Activated by two different chemicals — nitric oxide and natriuretic peptide — each pathway produces cGMP, which in turn stimulates the all-important heart muscle protector PKG. Most cases of heart failure, the researchers say, are fueled by breakdowns in both.

"The existence of two separate pathways with overlapping but distinct functions is nature's insurance policy, a fail-safe redundancy to ensure that should one pathway falter, the other one can compensate and maintain heart muscle function," says senior investigator David Kass, MD, Professor of Medicine at the Johns Hopkins University School of Medicine and its Heart and Vascular Institute.

Nearly a decade ago, a team led by Kass identified the culprit responsible for breakdown in one of the signaling pathways, an enzyme called PDE-5 — also known to cause erectile dysfunction — and ever since then scientists have searched for the second "offender" that causes glitches in the other pathway. The discovery of PDE-9 provides that long-sought "break in the case," the team says.

"Like a play with multiple characters, heart muscle function is the result of a complex but perfectly synchronized interaction of several proteins, enzymes and hormones," says lead investigator Dong Lee, MS, PhD, a cardiology research associate at the Johns Hopkins University School of Medicine. "Our findings reveal that, like two subplots that converge in the end of the play, PDE-5 and PDE-9 are independent rogue operators, each leading to heart muscle damage but doing so through different means."

Kass' earlier studies showed that PDE-5, like its newly identified accomplice PDE-9, inflicts damage by feeding on heart-protective cGMP and PKG. But the new findings reveal an important difference — PDE-9 has an appetite for the form of cGMP stimulated by the second signaling pathway.

In other words, Kass says, too much PDE-9 can interfere with the second of the two heart-protective systems by speeding up the breakdown of cGMP, which in turn reduces levels of PKG, rendering heart cells prone to malfunction and the heart muscle vulnerable to scarring and damage.

The research team also notes that heart failure treatments blocking the activity of PDE-9 may be right around the corner, with drugs that inhibit PDE-9 already being tested for use in people with Alzheimer's disease.

In the current study, such PDE-9 blockers not only stopped heart muscle enlargement and scarring in mice with heart failure, but they nearly reversed the effects of the disease.

"We believe the identification of PDE-9 puts us on the cusp of creating precision therapies that target the second pathway or developing combined therapies that avert glitches in both pathways," Kass says.

The new findings could be even more relevant for the nearly one-half of all heart failure patients with an especially recalcitrant form of the disease known as

heart failure with preserved ejection fraction — a treatment-defying condition in which the heart appears to pump normally but is, in fact, scarred and hardened. Heart cells obtained from people with this form of the disease showed PDE-9 levels six times higher than normal hearts, suggesting that people with that form of the disease have far less heart-protective PKG, the researchers note.

In one set of experiments, the researchers chemically stimulated human heart cells and cells from newborn mice to mimic the effects of heart failure. The cells ballooned beyond their normal size, but when treated with a PDE-9 antidote, the cells returned to near normal.

In another test, the scientists created mice genetically incapable of making PDE-9 and compared their heart cells and overall organ function to mice with intact PDE-9 genes. The heart cells of mice lacking PDE-9 had higher cGMP levels than the cells of mice with intact PDE-9. Next, mice in both groups underwent surgical narrowing of the aorta, the body's main blood vessel, a procedure intended to create extra workload for the heart and induce heart failure. Mice lacking the PDE-9 gene fared much better. Their hearts had far less scarring, muscle thickening and dilation than the hearts of mice with intact PDE-9.

In a third experiment, the scientists divided mice with surgically induced heart failure into three groups. One group received treatment with a PDE-9 blocking drug, another was treated with the PDE-5 blocker sildenafil and a third was given a placebo.

After four weeks of therapy, mice treated with a placebo developed full-blown heart failure, while mice treated with either PDE-9 or PDE-5 blockers showed marked improvements in heart muscle function and size, their hearts' pumping ability reverting to near normal.

To compare the effects of PDE-5 and PDE-9 inhibitors on heart function, the researchers then fed half the mice with a chemical that turned off the heart-protective signaling pathway regulated by PDE-5. In that group, treatment with a PDE-5 inhibitor made no difference. However, when given a PDE-9 blocker, the animals' hearts showed notable improvement. This finding, the research team says, illustrates the notion that heart muscle failure is fueled by flaws in two separate signaling cascades, each one regulated by a different enzyme and benefiting from different treatment.

"In practical terms, this affirms that preserving the function in one pathway can avert clinical disease, even if the other one goes bad," Kass says.

The work was supported in part by the National Institutes of Health under grant numbers HL-119012, HL-093432, HL-089297 and HL-07227; Fondation Leducq TransAtlantic Network of Excellence; the Peter Belfer Laboratory Foundation; the Abraham and Virginia Weiss Professorship in Cardiology; the American Heart Association; and the Austrian Academy of Sciences. Drug manufacturer Pfizer provided the PDE-9 inhibitors and the genetically altered mice.

Other Johns Hopkins investigators involved in the study included: Guangshuo Zhu, Manling Zhang, Thomas Danner, Peter P. Rainer, Djahida Bedja, Jonathan A. Kirk, Mark J. Ranek, Jennifer Van Eyk and Eiki Takimoto.

Other organizations participating in the research were: the Cardiovascular Research Institute at VU University Medical Center in Amsterdam, Cedars-Sinai Medical Center in Los Angeles, Kangwon National University College of Medicine in South Korea, the University of Vermont and the Mitsubishi Tanabe Pharma Corporation of Japan.

Letters to the Editor

Congenital Cardiology Today welcomes and encourages Letters to the Editor. If you have comments or topics you would like to address, please send an email to: LTE@CCT.bz, and let us know if you would like your comment published or not.

Case Report: Fontan Patient with Recurrent Cerebral Embolic Phenomenon and Pulmonary Artery Stump

By Tabitha Moe, MD

A 33-year-old female presented to the Emergency Department for evaluation after her home care giver noticed that she was having increasing difficulty speaking, and decreased arm movements. Discussion with the patient's husband, to whom she has been married for eight years, revealed the medical history including an atrial septal defect closure at the age of five years, complicated by a perioperative stroke with residual hemiparesis, and resultant contractures. However, the patient typically functions relatively independently, is able to cook and clean, and communicate verbally. She does not have any medical insurance and has not seen a cardiologist in many years. She takes no medications. An electrocardiogram demonstrated a regular atrial rhythm without ectopy (Image 1).

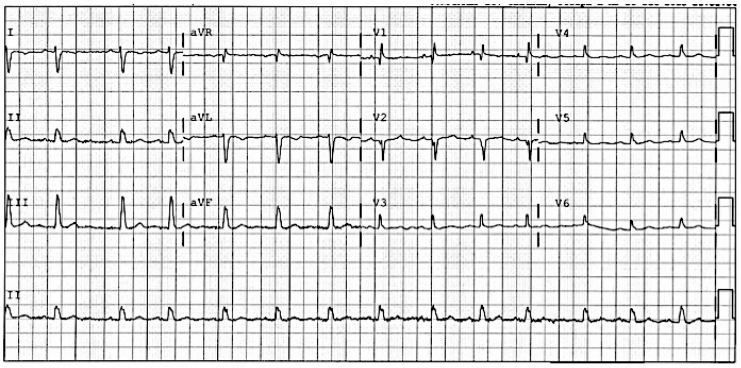


Image 1. Electrocardiogram.

Initial evaluation of the patient was concerning as her peripheral oxygen saturation was 78%; however, she was not in respiratory distress. An emergent chest CT with pulmonary embolus protocol was ordered. The interpretation of this test eluded the radiologist secondary to her complex anatomy, and a congenital cardiology consult was called. It was determined after review of the CT, and a transthoracic echocardiogram that the patient has a single morphologic left ventricle, and a hypoplastic right ventricle. The aorta arises from the morphologic right ventricle remnant. There is no substantial interventricular septum. Her operative repair appeared to have been a single operative palliation with a classic Fontan, right atrial appendage to pulmonary artery conduit. She also has a dilated persistent left superior vena cava to coronary sinus, a dilated bridging innominate vein, and a pulmonary artery stump with swirling of contrast seen on CT. Due to multiple levels of right-to-left shunting the patient is desaturated. She is also at high risk for recurrent embolic stroke in the setting of a pulmonary artery stump. Unfortunately she developed hemorrhagic conversion of her middle-cerebral artery embolic stroke, and therefore full-anticoagulation could not be initiated for four weeks. She was referred for outpatient cardiac catheterization with plans to occlude the bridging innominate vein in attempts to improve saturations by reducing right to left shunting. Successful vascular plug occlusion of two large venous collaterals, one draining from the innominate vein to the coronary sinus to the atrium, and the second draining via the azygous vein to the coronary sinus to the atrium was performed, each using a fourteen-millimeter vascular plug. After occlusion of both collaterals, oxygen saturations improved to 90% without any increase in Fontan circuit pressures.

Discussion

Adult Congenital Heart Disease (ACHD) management is a balancing act. Patients may be lost to follow-up for many years prior to a re-presentation with a new acute issue. An initial diagnosis and interval surgical procedures may be incorrect, or absent altogether, further complicating management. Adult patients may not be aware of their

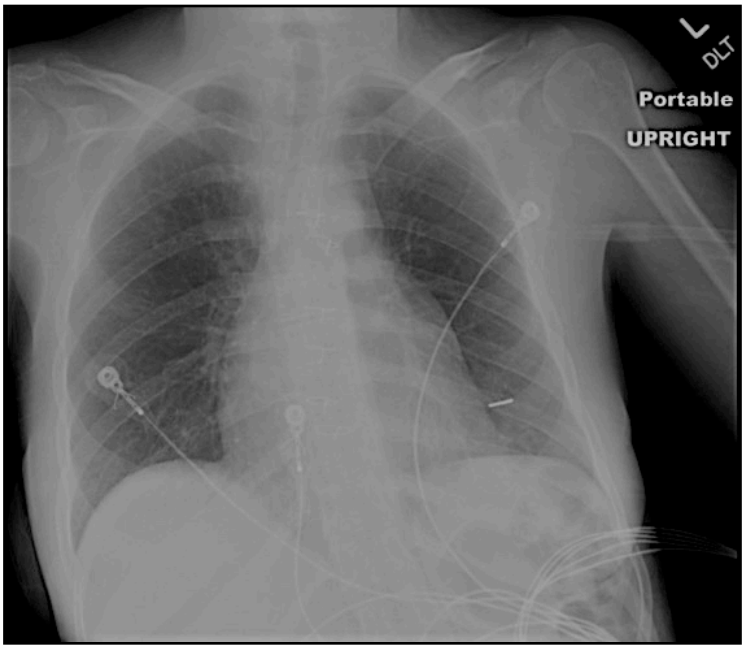


Image 2. Standard Antero-Posterior Chest Xray demonstrates right atrial enlargement, and cardiomegaly.

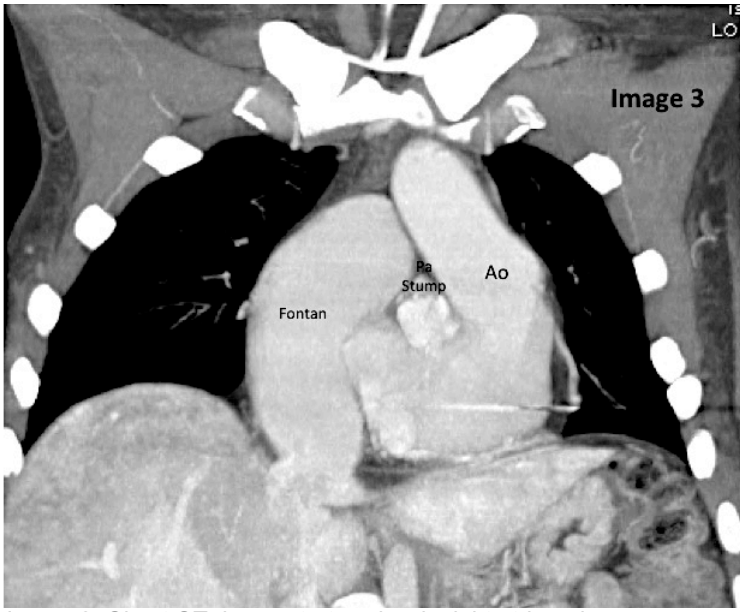
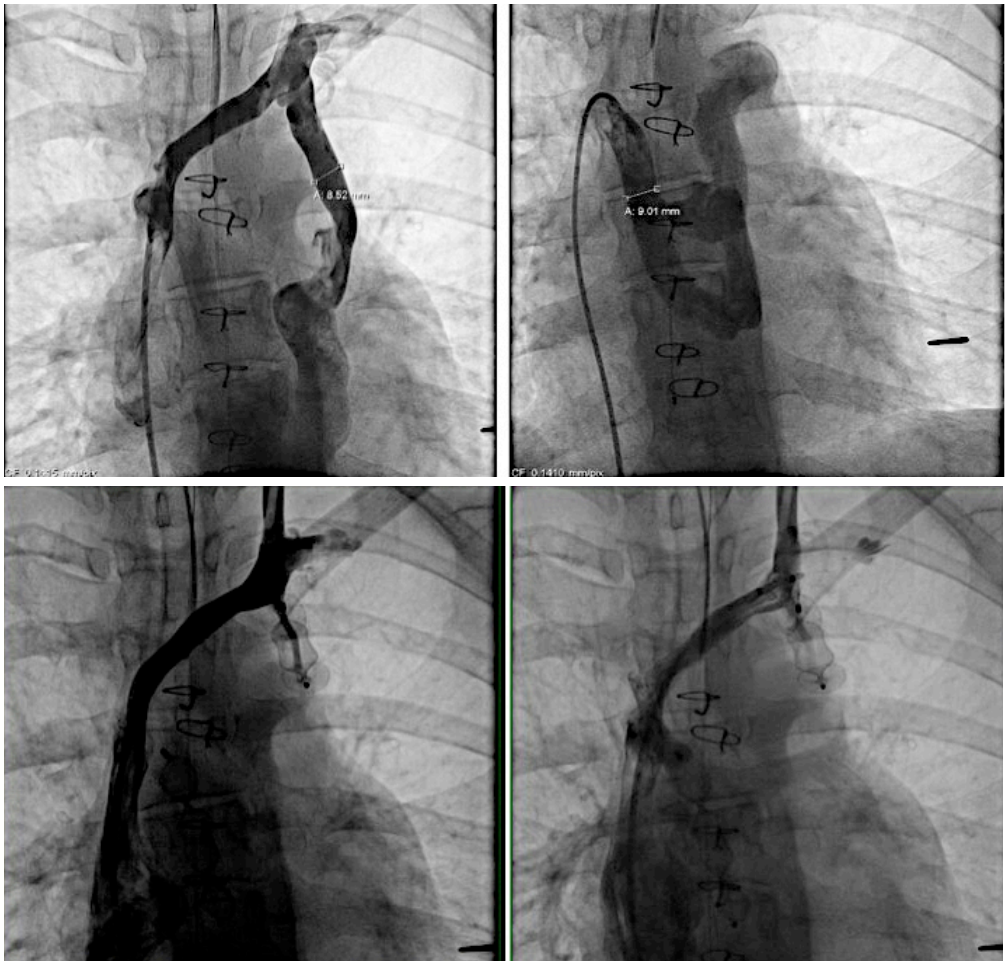


Image 3. Chest CT demonstrates classic right atrio-pulmonary Fontan circuit, and contrast within the pulmonary artery stump.

diagnosis or surgical history, their parent or grandparent may no longer be available and surgical records lost.

Patients who historically underwent a classic or modified intracardiac Fontan, may or may not have had their pulmonary valve oversewn as a part of their palliation. The risk of having a pulmonary valve with some antegrade flow is the risk of nonlinear flow in any fluid dynamics. Nonlinear flows cause eddies in the stream. Loss of linear fluid dynamics in an already distended Fontan circuit increases the risk of thromboembolism, with subsequent distal embolic phenomenon associated with thromboembolism.[1] This variation on anatomic repair in patients with congenital tricuspid atresia has since been recommended for full anticoagulation with warfarin.[2-6] Pharmacodynamics of the novel oral



Images 4 (Top) and 5 (Bottom) Pre- and post-coil embolization of left SVC collateral.

anticoagulants in patients who have undergone cavopulmonary and atriopulmonary anastomoses with resultant sluggish hepatic flow have not been studied. It is not recommended at this time for those patients to receive anticoagulation with the new novel anti-Xa agents, or the direct thrombin inhibitors.[7]

One of the challenges with this patient population, however, is that these patients may be under the impression that their childhood repairs do not require life-long congenital cardiac care, therefore, making following recommendations for interventions, therapeutic or otherwise problematic. Construction of a comprehensive U.S. database for ACHD is one step towards improving quality of care, and quality of life.

References

1. Whitehead KK, Pekkan K, Kitajima HD, Paridon SM, Yoganathan AP, Fogel MA. Nonlinear Power Loss During Exercise in Single-Ventricle Patients After the Fontan. Insights from Computational Fluid Dynamics. Circulation. 2007;116:165-171.
2. Oski JA, Canter CE, Spray TL, Kan JS, Cameron DE, Murphy AM. Embolic Stroke After Ligation of the Pulmonary Artery in Patients With a Functional Single Ventricle. Am Heart J. 1996;132:836-840.
3. Hutto RL, Williams JP, Maertens P, Wilder WM, Williams RS (1991) Cerebellar infarct: late complication of the Fontan procedure? Pediatr Neurol 7:293–295.
4. Mathews K, Bale JF, Clark EB, Marvin WJ, Doty DB (1986) Cerebral infarction complicating Fontan surgery for cyanotic congential heart disease. Pediatr Cardiol 7:161–166.
5. Russo P, Danielson GK, Puga FJ, McGoon DC, Humes R (1988) Modified Fontan procedure for biventricular hearts with complex forms of double-outlet right ventricle. Circulation 78(Suppl III):III20–III25.
6. Sanders SP, Wright GB, Keane JF, Norwood WI, Casteneda AR (1982) Clinical and hemodynamic results of the Fontan operation for tricuspid atresia. Am J Cardiol 49:1733–1740.
7. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol. 2008;52:e143–e263.

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