Absent Right and Persistent Left Superior Vena Cava: Case Report

By Anant Khositseth, MD

Introduction

Absent right superior vena cava (SVC) with persistent left SVC in viscerosubformal situs solitus is a rare congenital anomaly occurring in 0.07-0.13% of congenital heart malformation[1]. It is commonly associated with other congenital heart defects in 46% [2]. Here, we report a case of ostium secundum atrial septal defect (ASD) associated with this anomaly in a patient who underwent Amplatz Septal Occluder (ASO) implantation.

Case Description

An 11-year-old boy presented with presented with frequent tiredness during exertion. Physical examination revealed widely fixed splitting S2 with normal P2 and grade 2/6 systolic ejection murmur along the left upper sternal border (pulmonary valve area). His EKG had an abnormal P axis (-150°), right axis deviation for age (+120°), and pure R in V1 (Figure 1).

Figure 1. A 12-lead electrocardiography demonstrated ectopic atrial pacemaker (upright P wave in I and negative P wave in aVF, bold arrow) and pure R in V1 (dash arrow) indicated right ventricle hypertrophy.
A chest x-ray demonstrated mild cardiomegaly with slightly increased pulmonary vasculature. All of these findings were suggestive of an ASD. Echocardiography demonstrated mild dilatation of the right atrium and ventricle, ostium secundum ASD (8 mm in diameter), and dilated coronary sinus suspected left SVC. He was scheduled for cardiac catheterization for device closure by ASO. Transesophageal echocardiography (TEE) was performed during the catheterization. This demonstrated an ostium secundum ASD with stretched diameter of 14 mm., a dilated coronary sinus, and adequate rims for device closure. A 24-mm balloon sizing was inflated with stretched diameter of 14 mm. by fluoroscopy (Figure 2). A catheter could not be passed from the right atrium into the right SVC, but it could be passed from the right atrium through dilated coronary sinus, left SVC, right innominate vein, and right internal jugular vein. The contrast was injected into the right internal jugular vein and the left SVC, respectively. This injection demonstrated absent right SVC and persistent left SVC draining into the coronary sinus and the right atrium (Figure 3). The pulmonary to systemic blood flow was calculated and equal to 2.8 indicated large left to right shunt. The pulmonary arterial pressure was normal with mean of 20 mm Hg. A 14-mm ASO was selected to close the ASD successfully.

**Figure 2.** A fluoroscopy demonstrated a waist in balloon sizing (bold arrows) indicated a stretched diameter of the atrial septal defect.
The echocardiography performed at 1 day and 3 months after the procedure demonstrated proper position of the device without residual shunt. The ASO device did not obstruct the dilated coronary sinus.

Discussion

Bilateral SVC with persistent left SVC draining into the coronary sinus and then the right atrium which is a normal variation of normal systemic venous return is not uncommon in general, occurring in 0.3% of the general population. This condition is commonly associated with congenital heart defects in 3-34% of patients. However, persistent left SVC with absence of the right SVC is very rare. Bartram et al.[2] reported 121 cases with the absence of the right SVC in visceroatrial situs solitus found that this anomaly is typically characterized by persistent left SVC draining into the right atrium via the coronary sinus and additional cardiac defects could be found in 46% of these cases. Srivastava et al.[3] reported a case with persistent left SVC with absent right SVC associated with ostium secundum ASD which was accidentally detected during the surgical closure of the ASD due to the right internal jugular triple lumen venous catheter placement.

In this case report we successfully closed the ostium secundum ASD by using an ASO device. TEE performed before the procedure demonstrated dilated coronary sinus which was suspected to have the persistent left SVC, but the absent right SVC was not detected. However, the venous catheter course and angiography demonstrated absent right SVC and persistent left SVC.
draining into the right atrium via dilated coronary sinus. We thought that this finding was of concern due to the possibility of the device obstructing the dilated coronary sinus and its orifice. During the device deployment, we selected the size of the device properly using the stretched diameter of the defect by measuring the sizing balloon by TEE and fluoroscopy. We were also aware of the device position related to the large orifice of the coronary sinus by imaging of the TEE. Figure 4 demonstrated the ASO device in the proper position after the deployment. The patient’s EKG had abnormal P axis, which indicated ectopic atrial pacemaker. This finding was reported in 3 of 4 hearts with absent right SVC, and may be the key factor in the development of arrhythmias [4]. The incidence of arrhythmia after device closure may be increased in this patient.

To our knowledge, this was the first case report of Absent Right and Persistent Left Superior Vena Cava in which an associated ASD was closed by the ASO device. Although this is a rare condition, we do recommend all patients with ASD undergoing device closure should be performed complete echocardiography to examine the systemic venous drainage to the heart thoroughly.

References


Dr. Mullins retired at the end of 2006 and this series started with a lecture by Dr. Philip Bonhoeffer, entitled, “The Pulmonary Valve Stent” in January, 2007. The second lecture of this series was provided by Dr. Zahid Amin, entitled “Perventricular Occlusion of Muscular VSD’s.”

The event started with an informal dinner held the evening before, attended by current staff and some of Dr. Mullins’ former trainees working in other pediatric cardiac centers. Following the lecture, which was attended by approximately 50 people, a “Charles E. Mullins” display case was unveiled at the Texas Children’s Hospital heart center library and learning center. The display case was intended to show off some of Chuck’s collection of publications, catheters, antiques and other historically important artifacts pertaining to pediatric cardiology interventions.

“Texas Children’s Hospital held its 2nd Annual, “Charles E. Mullins Interventional Lecture Series,” in honor of the lifetime contribution of Dr. Mullins.”

Overall, it was a great time for a mixture of academic discussions and renewal of old friendships. If the reader has any “old artifacts” of Chuck’s, and would like to donate them to the “Mullins” display case, please contact the author.

The date for the third lecture will be sometime in the Spring of 2009. Those interested in possibly attending, are encouraged to send an email to the author, and information will be sent when available.
Highlights of PICS-AICS 2008

By Ziyad M. Hijazi, MD

With over 700 attendees from 65 countries, PICS (Pediatric and Adult Interventional Therapies for Congenital and Valvular Heart Disease) was a huge success. Fifty percent of the attendees were from the US. A faculty of ninety-five members participated, the largest of its kind in the world.

Similar to last year, we combined the pediatric and adult sessions for interventional therapy of congenital and structural heart disease. Live cases from twelve sites were transmitted via satellites to the venue at the Bellagio Hotel. The quality of live cases this year was outstanding.

PICS started Sunday July 20th at 8:30 AM with an industry-sponsored workshop (Cook Medical). The workshop was well-attended due to its excellent faculty and subject matter. Among the speakers in this workshop was Dr. Robert White from Yale, who is the world’s expert on pulmonary AVMs. Other speakers included: Drs. Lee Benson, Shakeel Qureshi, Jeff Feinstein, Jo De Giovanni and Omar Galal. Feedback from attendees on this workshop was overwhelmingly excellent.

In the afternoon, we had an excellent workshop on the atrial septum and imaging. There were fourteen talks discussing various aspects of imaging and devices for the septum. In that workshop, there were two excellent debates: one on ICE vs TEE and another on surgery for ASD vs device closure. The last debate between Dr. Fu and Dr. Ilbawi was particularly lively.

The day ended with “Meet the Expert” sessions and oral abstract presentations. At the end of the day, all attendees were invited to the welcome reception in the exhibit hall. This year, we had 29 exhibitors representing the wide range of manufacturers in our field.

The following day, Monday, July 21st, 2008, was full of action. Live cases were transmitted from Danta Pazzanesse Instituto de Cardiologia in Sao Paulo, Brazil, where Dr. Carlos Pedra and his team performed four excellent cases; from Rush University Medical Center in Chicago, where Dr. Hijazi and his team performed two cases; from Miami Children’s Hospital, where Dr. Evan Zahn and his team performed two cases, and from Seattle Childrens’ Hospital where Dr. Tom Jones performed three cases. Between the live cases, there were seventeen excellent talks discussing various aspects of interventional therapies in children and adults. Topics included were the following: Covered Stents, Medical Simulation, Percutaneous Aortic Valve Therapies, the Mitral Valve (Imaging and Therapies), and Percutaneous Pulmonary Valve Implantation. At the end of the day, there were two excellent sessions: one, a debate between Drs. Schranz and Bacha about management of AS in infants <3 months of age, and the other session was about catheterizing critically ill babies. Monday included the ever-popular “My Nightmare Case in the Cath Lab” session!

At the end of the day, we had our traditional “PICS Achievement Award.” This year’s recipient was Dr. Carlos Ruiz. Dr. Bill Hellenbrand reviewed Dr. Ruiz’s many accomplishments. Everyone was very impressed with Carlos’ background, and what he has achieved in our field.

Tuesday July 22nd, 2008 was the third full day of the meeting. Again, we had many excellent live cases transmitted from Atlanta, where Dr. Bob Vincent and his team performed two excellent cases; from Columbus Nationwide Children’s Hospital, where Drs. John Cheatham and Mark Galantowicz performed two excellent cases; from Detroit, where Dr. Tom Forbes and his team performed three very good cases, and from San Diego, where Dr. John Moore and his team performed three very good cases. Also on Tuesday, we had twenty-five talks discussing various aspects of interventional therapies in children and adults. The topics included the following: a debate about whether to angioplasty native coarctation, ventricular septal defects, the PFO, the LAA, heart failure monitoring devices, and the PDA.

At the end of the day, the attendees were treated to a night of fun at the traditional Gala Dinner. The gala was very well-attended, and similar to each year, we had drawings from B. Braun and the PICS Foundation for free registration and hotel for two for next year’s PICS.

The last day of the meeting, Wednesday, July 23rd, 2008 was as enjoyable as the first. Again, four sites transmitted live cases. From Cincinnati, Dr. Russel Hirsch and his team transmitted three very good cases. Dr. David Nykanen and his team transmitted two very good cases from Orlando. From St. Louis, Dr. David Balzer transmitted two very educational cases, and from Vancouver, Dr. John Webb and his team transmitted two excellent cases. For the first time at PICS, we had a trans-apical aortic valve replacement transmitted live from Vancouver. The same day we had twelve excellent talks covering cath lab issues for nurses and techs (inventory
management, sedation in the cath lab, vessel closure devices and surgical procedures in the cath lab); pericardiocentesis; vascular thrombosis and its management; vascular closure devices; hybrid management of HLHS. At the end of the day, five speakers talked about what is coming down the pipe that we should be looking for.

That was our last meeting in Las Vegas, at least for the next two years.

I hope that you all can join us in Cairns, Australia, June 21-23rd, 2009 for PICS at the World Congress of Pediatric Cardiology and Cardiac Surgery. It promises to be educational as usual, but more fun for you and for your family. For more information, go to www.picsymposium.com.

See you in Cairns,

Ziyad M. Hijazi, MD
on behalf of all course directors

CCT
Should We Use Echocardiography to Screen Young Athletes?

Sudden and unexpected deaths in young competitive athletes are uncommon, but highly visible events, which raise concern and ethical issues in both the lay public and medical community. Which is the best strategy to timely identify individuals with cardiac disease responsible for sudden death (primarily, HCM) is largely debated. Namely, the extent to which sophisticated testing, such as echocardiography, is needed is still undefined.

To address this question, we carried out an echocardiographic assessment of the structural cardiac diseases in a population of 4,450 athletes, initially judged free of cardiac disease and eligible for competition on the basis of pre-participation screening with 12-lead ECG.

None of the 4,450 athletes showed evidence of HCM. Other cardiac abnormalities were detected in only 12 athletes, including myocarditis (n=4), mitral valve prolapse (n=3), Marfan’s Syndrome (n=2), aortic regurgitation with bicuspid valve (n=2), and arrhythmogenic right ventricular cardiomyopathy (n=1). In addition, four athletes were identified with borderline LV wall thickness (i.e., 13 mm) in the “gray-zone” between HCM and athlete’s heart. In two of these athletes, subsequent genetic analysis or clinical changes over an average 8-year follow-up resulted, respectively, in a diagnosis of HCM.

In conclusion, the pre-participation screening program including 12-lead ECG appears to be efficient in identifying young athletes with HCM, leading to their timely disqualification from competitive sports. The data also suggest that routine echocardiography is not an obligatory component of large population screening programs designed to identify young athletes with HCM. For more information, European Society of Cardiology - www.escardio.org

Stem Cell Regeneration Repairs Congenital Heart Defect

Mayo Clinic investigators have demonstrated that stem cells can be used to regenerate heart tissue to treat dilated cardiomyopathy, a congenital defect. Publication of the discovery was expedited by the editors of Stem Cells and appeared online in the “express” section of the journal’s web site at http://stemcells.alphamedpress.org/.

The study expands on the use of embryonic stem cells to regenerate tissue and repair damage after heart attacks and demonstrates that stem cells also can repair the inherited causes of heart failure.

“We’ve shown in this transgenic animal model that embryonic stem cells may offer an option in repairing genetic heart problems,” says Satsuki Yamada, MD, PhD, cardiovascular researcher and first author of the study. “Close evaluation of ge-

Cardiac Ultrasound Imaging Goes to Handheld

Cardiac ultrasound imaging, also known as echocardiography, has been recently challenged by several new imaging methods. However, echocardiography has unique characteristics that make it very attractive: it is cheap, can be done bedside and without ionizing radiation. Recently, devices have also become very small.

Actually, in echocardiography there are two diverse and ong-
Hypertrophic cardiomyopathy (HCM) is a genetic disorder that can result in heart failure (HF) with or without obstruction (non-ischemic cardiomyopathy). A new study suggests that stem cell therapy can achieve functional repair in non-ischemic cardiomyopathy.

The research was supported by the National Institutes of Health, the American Heart Association, the Marriott Foundation, the Ralph Wilson Medical Research Foundation, and the Japanese Ministry of Education, Science, Sports, Culture and Technology.

**How They Did It**

The team reproduced prominent features of human malignant heart failure in a series of genetically altered mice. Specifically, the “knockout” of a critical heart-protective protein known as the KATP channel compromised heart contractions and caused ventricular dilation or heart enlargement. The condition, including poor survival, is typical of patients with heritable dilated cardiomyopathy.

Researchers transplanted 200,000 embryonic stem cells into the wall of the left ventricle of the knockout mice. After one month the treatment improved heart performance, synchronized electrical impulses and stopped heart deterioration, ultimately saving the animal’s life. Stem cells had grafted into the heart and formed new cardiac tissue. Additionally, the stem cell transplantation restarted cell cycle activity and halved the fibrosis that had been developing after the initial damage. Stem cell therapy also increased stamina and removed fluid buildup in the body, so characteristic in heart failure.

The researchers say their findings show that stem cells can achieve functional repair in non-ischemic (cases other than blood-flow blockages) genetic cardiomyopathy. Further testing is underway.

Others members of the multidisciplinary team are: Timothy Nelson, MD, PhD; Ruben Crespo-Diaz; Carmen Perez-Terzic, MD, PhD; Xiao-Ke Liu, MD, PhD; and Atta Behfar, MD, PhD, of Mayo Clinic; Takashi Miki, MD, Chiba University, Japan; and Susumu Seino, MD, Kobe University, Japan.

The research was supported by the National Institutes of Health, the American Heart Association, the Marriott Foundation, the Ralph Wilson Medical Research Foundation, and the Japanese Ministry of Education, Science, Sports, Culture and Technology.
tion compared favorably to traditional methods.

"Our findings suggest that Internet-based instruction is an effective way to teach health care professionals," says Dr. Cook. "We now can confirm that, across a wide variety of learners, learning contexts, clinical topics, and learning outcomes, Internet-based instruction appears to be as effective as similar to traditional methods."

Dr. Cook also notes that Internet-based instruction has unique advantages, including flexible scheduling, adaptability of instruction, and readily available content that is easily updated. "As health care workers balance challenging practice demands, the ever-expanding volume of medical knowledge requires us to find more effective, efficient ways to learn," says Dr. Cook. "Internet-based instruction will be an important part of the solution."

He also notes that this research likely applies to training outside of health care, citing studies in the engineering, computer science, and teaching fields that have shown similar results.

"There is more research to be done as we try to find out how to make Internet-based instruction most appropriate," says Dr. Cook. "We are currently conducting research looking at this issue. We also are reviewing other studies to see how to optimize Internet-based instruction."

Other researchers were Denise Dupras, MD, PhD, Patricia Erwin, and Victor Montori, MD, all of Mayo Clinic; and Anthony Levinson, MD, and Sarah Garside, MD, PhD, from McMaster University.

Last Minute Appeal from the International Children's Heart Foundation (ICF)

SANTIAGO–DOMINICAN REPUBLIC

The ICHF is in need of more PICU nurses for their trip to Santiago, Dominican Republic (DR) from Oct. 26-Nov. 8, 2008. The hospital is the Hospital De Ninos Dr. Arturo Guillon - in Santiago Dominican Republic - The ICHF has been assisting this unit since January 2006, and currently makes 4 trips/year there. The surgeons on this trip are Drs. Tom Karl and Joanne Starr, who will each do a week. The total trip duration is two weeks, and preference is for volunteers who can do both weeks, although they will look at people who cannot make the full two weeks, in case they are unable to recruit for the full period. Useful, but not essential is any level of Spanish speaking ability, and prior visits to DR or any ICHF site - please specify if any of these apply.

BEIJING–CHINA

The ICHF is in need of another pediatric cardiac intensivist for their trip to Beijing Children’s Hospital from Oct. 19 - Nov. 1, 2008. The ICHF has been assisting Beijing Children’s Hospital for 4 years now, and the unit now does over 400 cases a year independently. The surgeon on this trip will be Dr. Novick. The total trip duration is two weeks, and their preference is for volunteers who can do both weeks, although they will look at people who cannot make the full two weeks, in case they are unable to recruit for the full period. You will be one of two intensivists. Useful, but not essential is any level of Chinese speaking ability, and prior visits to China or any ICHF site - please specify if any of these apply.

For more information, contact:
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www.ACC09.ACC.org

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