

Biatrial drainage of a right-sided superior vena cava

Roland E. Gazaille, III, D.O., Christopher S. Hofelich, D.O., Matthew J. Zimmerman, M.D., William Meyers, D.O., and Deepthi R. Mosali, M.D.

We report a case of chronic hypoxemia in a 62-year-old woman as a result of biatrial drainage of a right-sided superior vena cava. Radionuclide ventilation and perfusion imaging revealed significant increased radiotracer activity in the kidneys, bowel, and thyroid gland suggesting a right-to-left shunt which was confirmed by contrast enhanced CT of the chest. An anatomically correct right-sided SVC drained through two channels, the larger of which emptied into the roof of the left atrium and a smaller atretic portion feeding the right atrium. We were able to find only nine case reports of this rare anomaly in the English literature. All prior cases demonstrated partial anomalous pulmonary venous return which was also demonstrated to be present in this case with the use of cardiac MRI. According to our literature search, this is one of the few cases to be diagnosed with cardiac MRI.

Introduction

Congenital malformations of systemic venous drainage are rare in the absence of other significant cardiac

anomalies. Isolated anomalies of a right superior vena cava (SVC) are an even more rare abnormality. Isolated malformations of systemic venous drainage can be difficult to diagnose, in part because of their unusual occurrence. These abnormalities are more commonly diagnosed at an early age and are more rarely established as a new diagnosis in late adulthood.

Case Report

We report a case of an unusual cause of chronic hypoxemia in a 62-year-old woman. Her main complaint was that of dyspnea which had been progressively worsening over the past year to the point that it had now begun to affect her activities of daily living. Her dyspnea worsened with activity and was relieved by rest. She had no history of associated chest pain or orthopnea. Her family noted that her chronically light blue

Citation: Gazaille RE III, Hofelich CS, Zimmerman MJ, Meyers W, Mosali DR. Biatrial Drainage of a Right-Sided Superior Vena Cava. *Radiology Case Reports*. [Online] 2009;3:233.

Copyright: © 2008 The Authors. This is an open-access article distributed under the terms of the Creative Commons Attribution-NonCommercial-NoDerivs 2.5 License, which permits reproduction and distribution, provided the original work is properly cited. Commercial use and derivative works are not permitted.

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging; SVC, superior vena cava

Roland E. Gazaille, III, D.O. (Email: regune8@yahoo.com), and William Meyers, D.O., are in the Department of Radiology, Grandview Hospital, Dayton, OH, USA.

Christopher S. Hofelich, D.O., and Deepthi R. Mosali, M.D., are in the Department of Cardiology, Grandview Hospital, Dayton, OH, USA.

Matthew J. Zimmerman, M.D., is in the Department of Cardiology, The Lexington Clinic, Lexington, KY, USA.

Published: July 16, 2009

DOI: 10.2484/rcr.v4i3.233

Bilateral Drainage of a Right-Sided Superior Vena Cava

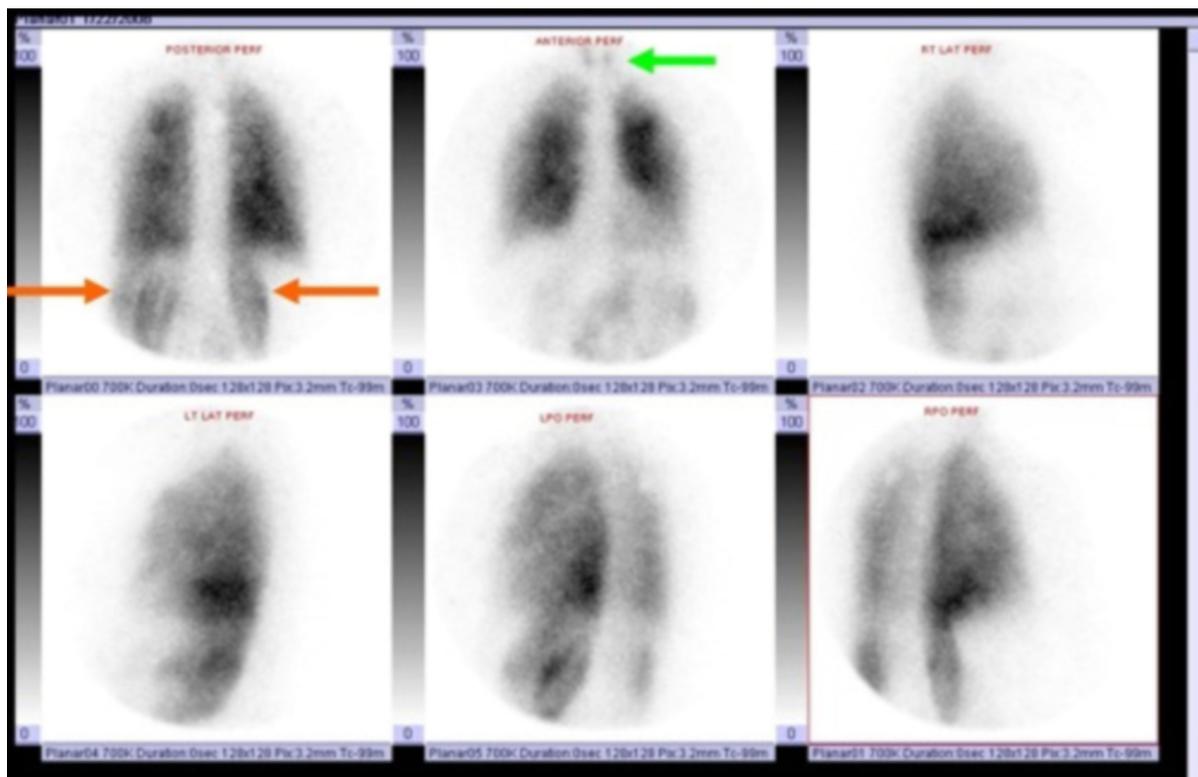


Figure 1. 62-year-old woman with chronic hypoxemia. Perfusion images reveal significant extrapulmonary radiotracer activity in kidneys (orange), bowel, and thyroid gland (green). Radionuclide ventilation images (not shown) were unremarkable.

hands appeared to have darkened in hue over the last few months. She reported difficulty climbing a flight of stairs and her daughter stated that her mother recently had been dyspneic during conversation.

While undergoing a recent colonoscopy, she became hypoxic following the administration of procedural sedation. This prompted a stat blood gas which revealed a pH of 7.45, a pO₂ of 52.5, a PCO₂ of 35.9, and an HCO₃ of 24.4. Her oxygen saturation on room air was 88% which did not improve with the administration of high-flow oxygen. During her resultant hospitalization she underwent a work-up to elucidate the etiology of her hypoxemia.

Her perinatal and childhood histories were significant for cyanosis. Her parents had taken her to the doctor multiple times throughout her early childhood and she was presumptively diagnosed with congenital heart disease. The family considered corrective surgery but declined due to the risks of pediatric cardiac surgery

at that time. During her adolescence she underwent a tonsillectomy without incident. From that point on, she was lost to follow-up and had a medically uneventful life. She did not play competitive sports. She tolerated a single pregnancy and gave birth without incident.

Her past medical history was also significant for uterine carcinoma, colon polyps, diverticulosis, and osteoporosis. She had episodes of syncope in the past for which she was hospitalized in 2005. A cardiac workup at that time was essentially unremarkable. She had no known drug allergies. Her family history was positive for congenital heart disease. She had a 25 pack-year history of smoking and had quit in 1981. She consumed alcohol on a social basis and denied the use of illicit drugs. She recently retired from an office job. Review of systems was negative for fevers, chills, night sweats, cough, sputum production, hemoptysis, orthopnea, lower extremity edema, or unexplained weight changes.

Biatrial Drainage of a Right-Sided Superior Vena Cava

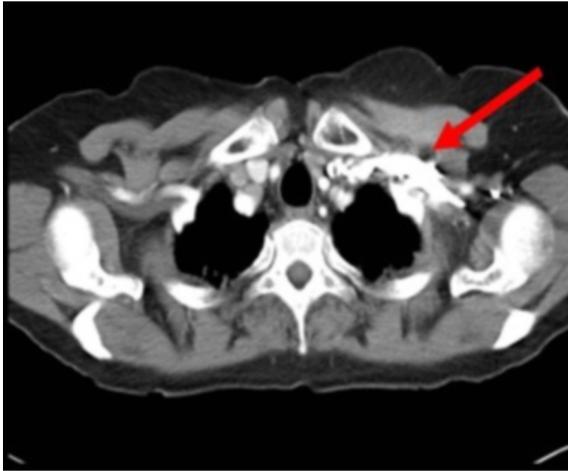


Figure 2. 62-year-old woman with chronic hypoxemia. A large right-to-left shunt was confirmed on contrast-enhanced CT of the chest utilizing a left upper-extremity venous injection (red).

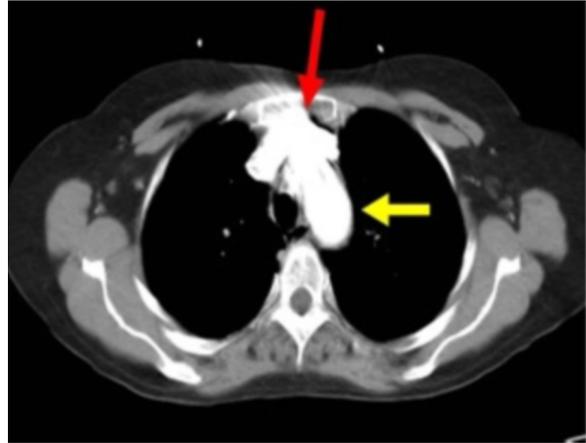


Figure 3. 62-year-old woman with chronic hypoxemia. Immediate and equivalent opacification of the aortic arch (gold) is noted when compared to the left brachiocephalic vein (red).

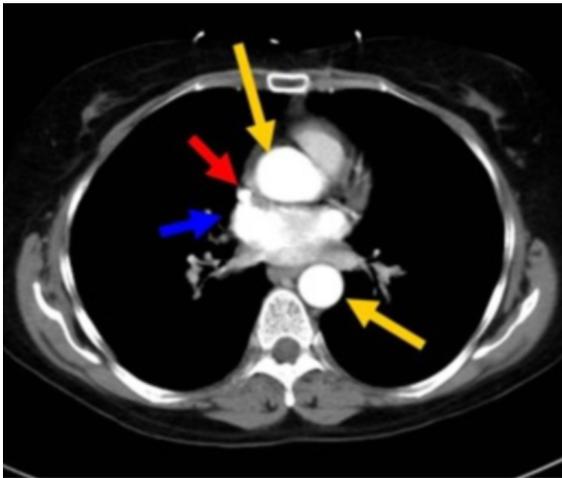


Figure 4. 62-year-old woman with chronic hypoxemia. An anatomically correct right-sided SVC drains through two channels, the larger of which empties into the left atrium (blue) with a smaller atretic portion (red) feeding the right atrium.

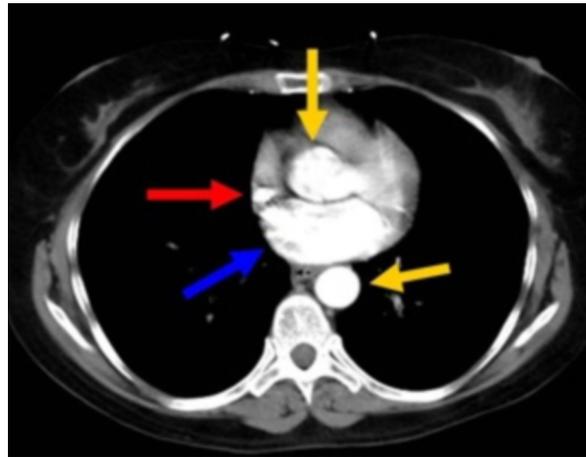


Figure 5. 62-year-old woman with chronic hypoxemia. An anatomically correct right-sided SVC drains through two channels, the larger of which empties into the left atrium (blue) with a smaller atretic portion (red) feeding the right atrium. The ascending and descending thoracic aorta are again noted to be of equivalent opacity as compared to the right-sided venous structures on this immediate post-injection image.

Biatrial Drainage of a Right-Sided Superior Vena Cava

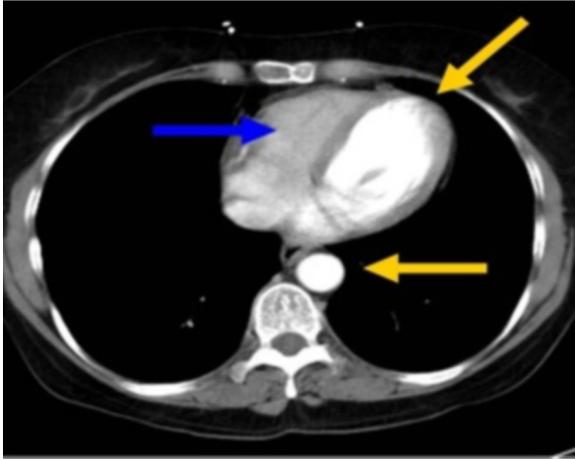


Figure 6. 62-year-old woman with chronic hypoxemia. A paucity of contrast is noted in the RV (blue) as compared to LV and aorta (gold), which is an abnormal finding on this early venous phase image.

Physical examination revealed a 62 year-old caucasian female in no acute distress. Her vitals signs were within normal limits with the exception of a low oxygen saturation of 88% on room air. The cardiac exam was remarkable for a soft systolic murmur. Mild clubbing of the digits along with cyanosis were also present. Pulses were of equal quality and palpable in all four extremities.

Pulmonary function testing revealed mild obstructive mechanics with minimal improvement following the administration of bronchodilators. Her echocardiogram revealed mild to moderate mitral insufficiency, trace pulmonic and tricuspid insufficiency, as well as Grade I diastolic dysfunction. The presence of a mildly dilated left atrium and a prominent coronary sinus were noted. Her left and right ventricular systolic function was considered normal. A markedly abnormal Qp/Qs ratio of 0.511 was consistent with a large right-to-left shunt. The remainder of the non-invasive cardiac workup was negative and following a short stay in the hospital. She was discharged with instructions to follow up with the consulting cardiologist.

Radionuclide ventilation and perfusion imaging performed on an outpatient basis revealed significant increased radiotracer activity in the kidneys, bowel, and thyroid gland along with unremarkable ventilation images (Figure 1). Images of the brain were not obtained.

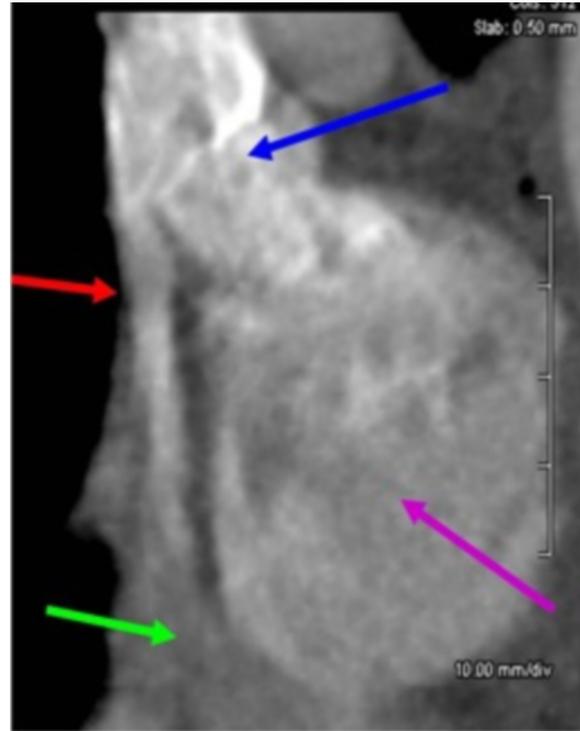


Figure 7. 62-year-old woman with chronic hypoxemia. Coronal 3-D reconstructed image reveals the small atretic segment of the SVC (red) feeding the right atrium (green), and the larger segment (blue) feeding the left atrium (purple).

A large right-to-left shunt was confirmed on a contrast enhanced CT of the chest. However, to our surprise, the shunt was noted to be pre-cardiac as opposed to intracardiac (Figures 2-6). An anatomically correct right-sided SVC was demonstrated to drain through two channels, the larger of which emptied into the roof of the left atrium with a smaller atretic portion feeding the right atrium (Figure 7). Evidence of partial anomalous pulmonary venous return was demonstrated on the cardiac MRI/MRA with a right upper lobe pulmonary vein draining to the superior vena cava (Figure 8).

Biatrial Drainage of a Right-Sided Superior Vena Cava

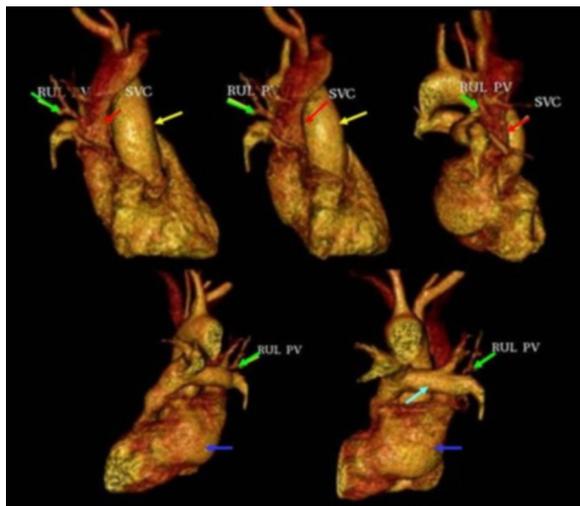


Figure 8. 62-year-old woman with chronic hypoxemia. Volume rendered images from a cardiac MRA demonstrate partial anomalous pulmonary venous return (green) to the SVC (red) which was not appreciated on the contrast-enhanced CT of the chest. Blue – Left atrium, Yellow – Aorta, Light blue – Right pulmonary vein

Discussion

Venous anomalies of the superior vena cava are the result of variations in persistence and/or regression of the anterior and common cardinal veins [1]. During normal embryologic development, the left anterior cardinal vein regresses and the remaining right anterior and common cardinal veins form a right-sided SVC. Persistence of the left anterior cardinal vein results in a duplicated SVC, the most common anomaly of the superior vena caval system. A solitary left-sided SVC is due to persistence of the left anterior cardinal vein accompanied by regression of the right anterior cardinal vein. Isolated anomalies of a right-sided SVC are rare. Variations include a low insertion into the right atrium, complete drainage into the left atrium, or congenital dilatation which can mimic a superior mediastinal mass on standard chest radiography [2].

Extrapulmonary accumulation of ^{99m}Tc -MAA can be seen in a variety of situations. These scenarios may include right-to-left cardiac or pulmonary shunts, shunting to the portal vein before reaching the right atrium and ventricle of the heart, free pertechnetate due

to poor preparation of the radiopharmaceutical, and with degradation of the tagged radiopharmaceutical to submicron-particulate size allowing passage through the pulmonary capillary bed [3]. Therefore, with the exception of situations involving a problem with the radiopharmaceutical, visualization of organs other than the lungs or demonstration of extrapulmonary hotspots is most often suggestive of abnormal hemodynamics with a physiologic right to left shunt. Given the clinical symptomatology in this patient, the radionuclide findings were felt to be highly suspicious for a right-to-left shunt. These findings were then confirmed with the contrast enhanced CT of the chest as well as the cardiac MRI.

The most comprehensive review of the rare anomalous biatrial or left atrial drainage of a right superior vena cava was completed in 2003 [4]. Following review of three newly reported pediatric cases, the authors highlighted some of the trends noted in the nine (9) biatrial and nineteen (19) left atrial drainage patterns of the right superior vena cava cases (3 new and 25 old) reported in the English literature.

Van Praagh and colleagues believe these anomalies result from an anomalous venous confluence that may more accurately be termed a cavopulmonary venous defect. Its anatomic variations include the sinus venosus defect of the superior vena cava (or high type), the biatrial, and the left atrial drainage of the right superior vena cava. All three variations exhibited “unroofing” of the right upper or upper and middle pulmonary veins into the normally positioned right superior vena cava. They noted that surgical treatment of left atrial drainage of the right superior vena cava has been successfully accomplished. They also noted that concomitant congenital cardiac malformations are very rare in the presence of anomalous venous drainage of the right superior vena cava.

Since this comprehensive 2003 review, at least seven [7] case reports on this topic have also been reported. In 2004, Drs. Recto and colleagues reported on a four year-old child that presented with cyanosis and polycythemia [5]. SVC angiography demonstrated flow into both the left and right atria. Predominant flow was into the left atrium, and an anomalously draining right upper lobe pulmonary vein was also seen. Also in 2004, Varghese and colleagues reported in the Indian Journal of Thoracic and Cardiovascular Surgery on a patient with anomalous drainage of the right superior vena cava to the left atrium [6]. Arora and colleagues in 2005 reported on a neonate with unexplained cyanosis and normal intra-

Biatrial Drainage of a Right-Sided Superior Vena Cava

cardiac anatomy by echocardiography [7]. The diagnosis of a right superior vena cava that drained into the left atrium was made via cardiac catheterization. In 2006, a case of an anomalous right-sided superior vena cava was reported in *Circulation (Images in Cardiovascular Medicine)* by Sadek and colleagues in which the diagnosis was made, in part, based on magnetic resonance imaging [8]. In 2006, at least three more articles appeared that chronicled cases of a right-sided superior vena cava draining into the left atrium. Aminololama-Shakeri and colleagues from UC Davis reported on a case of a toddler with asymptomatic hypoxemia who was ultimately determined to have a right-sided superior vena cava that drained into his left atrium [9]. Also in 2006, Vassallo and colleagues reported on the first case per our literature review of a prenatal diagnosis of anomalous drainage of a right-sided superior vena cava to the left atrium [10]. In addition, this patient also had anomalous pulmonary venous return and an atrial septal defect. Lastly, Oppido and colleagues published a case report of a case report of a newborn with a right-sided superior vena cava draining into the left atrium in *The Annals of Thoracic Surgery* in 2006 [11].

This particular case certainly raises some interesting questions as to the cause of her physiologic decompensation at the time of her colonoscopy. In addition, there are some important implications regarding the immediate and long-term management of this patient. The most important and imminent risk is that of paradoxical embolus with the potential for stroke and/or development of brain abscesses. These phenomena have been documented as part of the clinical presentation in prior reported cases of both biatrial and left atrial drainage of a right-sided superior vena cava [12]. Due to the risk of thromboemboli to the systemic circulation, systemic anticoagulation and consideration of placement of a superior vena cava filter may be the most appropriate immediate method of preventing the possibility of these devastating sequelae of this rare anomaly. Preferential placement of all future intravenous access lines in the lower extremity vessels is recommended to minimize the risk of systemic thromboembolization.

The matter of whether the patient truly decompensated or whether the findings of hypoxemia during her colonoscopy were coincidental is also up for debate. It should be noted that this workup was initiated upon the fact that the patient had a low oxygen saturation noted on pulse oximetry which did not improve with the use of high-flow oxygen. The nuclear medicine ventilation

and perfusion study and the contrast-enhanced CT of the chest were subsequently ordered in an effort to come up with an explanation as to the patient's persistently low oxygenation status. Systemic uptake following intravenous injection of radiotracer is consistent with a right-to-left shunt as was noted in this particular case. The patient had obviously been chronically compensating for this abnormality given the polycythemia. It is therefore plausible that perhaps the patient may not have "decompensated" at all and that these were merely coincidental findings of the most unusual variety. Potential causes for her decompensation include but are not limited to interval development of mild diastolic heart failure, as noted on her cardiac workup, with her underlying fixed anatomic abnormality leaving her no room for additional physiologic compensation. A second confounding possibility may also include decreased ventilatory drive as a result of the conscious sedation administered at the start of her colonoscopy.

Although there have been positive outcomes with surgical intervention of left atrial drainage of a right sided superior vena cava in more recent literature [4], the patient has elected to forego invasive treatment at this time. Given that she is a cyanotic adult survivor who is essentially stable at this time, invasive iatrogenic intervention at this time would risk making her symptoms and current quality of life worse. Reasons for her to consider a complete physiologic repair may include worsening cyanosis, decrease in functional capacity, or the development of symptomatic arrhythmias [13]. In the meantime, the patient will need to be followed closely on an outpatient basis as suggested in a consensus statement from the Canadian Cardiology Society regarding recommendations for the management of adults with congenital heart disease. This includes yearly CBC, ferritin level, clotting profile, renal function and uric acid levels, and monitoring for symptoms of hyperviscosity, systemic complications of cyanosis, change in exercise tolerance or oxygen saturation levels, and development of arrhythmias [13]. In addition to risk modification and surveillance for acquired cardiovascular disease, anticoagulation and serial echocardiograms will be of paramount importance as the enlargement of the left atrium will predispose the patient to the development of mural thrombi.

Biatrial Drainage of a Right-Sided Superior Vena Cava

References

1. Kume N, Suga K, Uchisako H, Matsui M, Shimizu K, Matsunaga N. Abnormal extrapulmonary accumulation of 99m Tc-MAA during lung perfusion scanning. *Ann Nucl Med* 1995; 9: 179-84. [PubMed]
2. Demos TC, Posniak HV, Pierce KL, Olson MC, Muscato M. Venous anomalies of the thorax. *Am J Roentgenol* 2004; 182: 1139-50. [PubMed]
3. Arat N, Sokmen Y, Golbasi Z. Persistent left superior vena cava with absent right superior vena cava and coronary sinus dilatation mimicking a paracardiac mass. *Texas Heart Inst J* 2007 34: 492-3. [PubMed]
4. Van Praagh S, Geva T, Lock JE, Nido PJ, Vance MS, Van Praagh R. Biatrial or left atrial drainage of the right superior vena cava: Anatomic, morphogenetic, and surgical considerations – report of three new cases and literature review. *Pediatr Cardiol* 2003; 24: 350-363. [PubMed]
5. Recto MR, Sobczyk WL, Yeh T Jr. Right superior vena cava draining predominantly into the left atrium causing cyanosis in a young child. *Pediatr Cardiol* 2004; 25: 163-4. [PubMed]
6. Varghese R, et al. Anomalous drainage of right superior vena caval vein to left atrium with intact atrial septum – a case report. *Indian J Thorac Cardiovasc Surg* 2004; 20(3): 135-6.
7. Arora G, Carlson KM, Neish SR. Anomalies of systemic venous return: Right superior vena cava draining to the left atrium. *Pediatr Cardiol* 2005; 26: 115-6. [PubMed]
8. Sadek H, Gilkeson RC, Hoit BD, Brozovich FV. Case of anomalous right superior vena cava. *Circulation* 2006; 114: e532-3. [PubMed]
9. Aminololama-Shakeri S, Wootton-Gorges SL, Pretzlaff RK, Reyes M, Moore EH. Right-sided superior vena cava draining into left atrium: A rare anomaly of systemic venous return. *Pediatr Radiol* 2007; 37(3) 317-20. [PubMed]
10. Vassallo M, Pascotto M, Pisacane C, Santoro G, Paladini D, Russo MG, Calabrò R. Right superior vena cava draining into the left atrium: Prenatal diagnosis and postnatal management. *Ultrasound Obstet Gynecol* 2006; 27(4): 445-8. [PubMed]
11. Oppido G, Napoleone CP, Turci S, Giardini A, Formigari R, Angeli E, Gargiulo G. Right superior vena cava draining in the left atrium: Anatomical, embryological, and surgical considerations. *Ann Thorac Surg* 2006; 81(6): 2313-5. [PubMed]
12. Shapiro EP, Al-Sadir J, Campbell NP, Thilenius OG, Anagnostopoulos CE, Hays P. Drainage of right superior vena cava into both atria: Review of the literature and description of a case presenting with polycythemia and paradoxical embolization. *Circulation* 1981; 63(3): 712-7. [PubMed]
13. Canadian Cardiovascular Society. CCS Consensus Conference 2001 update: Recommendations for the management of adults with congenital heart disease. Patient Management – Section XVI: Management of Cyanotic Patients. Parts III, IV, and IV. http://www.cachnet.org/managing_recomp3.html#Section%20XVI