

ORIGINAL ARTICLE

ROLE OF PULMONARY ANGIOGRAPHY IN CONGENITAL HEART DISEASES AMONG CHILDREN

Kavita U Vaishnav¹, Chhaya J Bhatt¹, Soujanya D Bolla¹, Dharita S Shah¹**Authors' Affiliation:** ¹NHL Municipal Medical College, AMC Medical Education Trust, VS hospital, Ahmedabad**Correspondence:** Dr. Kavita U Vaishnav, Email: kavitaradio@gmail.com

ABSTRACT

Objective: The aim of this study is to demonstrate MDCT (multi detector computerized scan) angiographic findings of various congenital vascular anomalies in pediatric patients.**Material & Method:** All the pulmonary angiography studies done in our department during the period of March 2012-February 2013 were evaluated to detect congenital heart diseases. Written informed consent was obtained. Routine pre procedure workup was done as per the protocol of the CT angiography study. Plain CT thorax, early arterial phase and venous phase were taken.**Results:** Out of 20 patients, no gender discrimination was seen. Patients with age group of twenty days to seventeen years were included in the study. Six patients suffered from cyanotic heart disease. Eleven patients had associated cardiomegaly. Six patients had involvement of pulmonary veins. The findings of CT angiography were compared with Echo. CT pulmonary angiography was superior to Echo in visualization of the morphology of the congenital anomaly and its associated findings. However angiography was not useful for detecting valvular diseases of the heart. The intraoperative findings were consistent with the pulmonary angiographic findings.**Key Words:** CT scan, angiography, congenital heart diseases (CHD).

INTRODUCTION:

Echocardiography and catheter cardio angiography are the primary cardiac imaging modalities, but both have its own limitations. New generation Multidetector Computed tomography (CT) and magnetic resonance (MR) imaging have important roles in overcoming these limitations. CT angiography is fast method to evaluate vascular structural anomalies by fast image acquisition times and their capacity to obtain volumetric data. For the complete evaluation of CHD high and spatial resolution is necessary which is possible with CT and MR imaging. In this study, we will discuss and illustrate the CT findings (extra cardiac abnormalities, cardiac abnormalities, connection problems) in patients with CHD, and the advantages and disadvantages of CT in this setting.

AIMS

The purpose of this study is to describe the role of CT in the evaluation of congenital cardiovascular disease in children and anatomical anomalies in pulmonary vasculature. Clinical indications, imaging techniques, and illustrations of relevant conditions are presented.

MATERIAL AND METHODS:

The study included CT pulmonary angiography done in our department during the period of March 2012-

February 2013 for various congenital heart diseases. No gender bias was followed. CT scan was done on Multidetector scanner from Siemens or Philips. Written informed consent was obtained. Routine pre procedure was done as per the protocol of the CT angiography study. Breath holding technique was explained to the patient who was able to follow breath holding order.

1. Plain CT scans of thorax followed by
2. Early arterial phases: - was obtained after negative test dose of the non-ionic contrast by injecting 12 to 40 ml of intravenous contrast media (Iopamidol 350mg) through cubital vein by pressure injector at a rate of 1.5 ml/sec. Scan was obtained 6 sec after starting of the intravenous contrast. Scan parameters were as follows:- Volumetric data was obtained from the vessels in axial plane and was reconstructed in saggital, coronal plane. Slice thickness- 1mm.Collimation:- 0.6 mm. Pitch: - 1.5, MAS=200, Kvp120.

RESULT

Results are as shown in tables.

Table 1: Age distribution

Age of presentation	Patient (%)
0-5	9 (45)
6-10	7 (35)
11-15	1 (5)
15-17	3 (15)

Majority of the patients presented between zero days to 5yrs.

Table 2: Association with cardiomegaly:

Association with cardiomegaly	Patient (%)
Cardiomegaly	11 (55)
Without cardiomegaly	9 (45)

11 children had cardiomegaly while 9 did not, showing a slight association with cardiomegaly

Table 3: Association with cyanosis

Association with cynosis	Patient (%)
Cynosis	6 (30)
Acynosis	14 (70)

14 children had acyanotic heart disease and 6 had cyanotic heart disease. There is a higher incidence of acyanotic heart diseases. There were 10 males and 10 female patients. Only 2 out of the 20 children had dextrocardia.

There was strong correlation between CT pulmonary angiographic and echo findings. Valvular pathologies were not evaluated by angiography. However patent ductus arteriosus and persistent truncus arteriosus were diagnosed exclusively by angiography only. Detailed morphology of coarctation of aorta was better depicted by angiography. Only 4 out of the 20 children had changes in lung fields in form of ground glass opacities.

Six patients had anomalies in the pulmonary veins of which two patients had partial anomalous pulmonary venous return and one had total anomalous pulmonary venous return. 11 patients had anomalies of the pulmonary artery of which 5 patients had dilated main pulmonary artery suggestive of pulmonary arterial hypertension and 5 patients had features pulmonary arterial hypoplasia in which the pulmonary artery was narrowed or not visualized.

Intra operative findings were consistent with the pulmonary angiographic findings in patients who had undergone surgery while other received supportive treatment only.

DISCUSSION

CT pulmonary angiography can be considered the first-line imaging technique in a few clinical cases for example evaluation of suspected vascular ring or sling, suspected aortopulmonary collateral circulation in patients with severe right ventricular outflow tract obstruction, and in individuals with implanted pacemakers and metal surgical hardware in whom MRI is contraindicated. [1] The role of CT in the evaluation of intracardiac anomalies has been more promising, owing to advances in MDCT scanners and ECG-gating techniques. In a small study with a 40-MDCT scanner

(without ECG gating), Lee et al. [2] detected 53 of 54 intracardiac and extracardiac anomalies in neonates with complex congenital heart disease. CT scan is also helpful in the assessment of extracardiac systemic and pulmonary arterial and venous structures. For a radiologist, it is important to have precise knowledge of cardiovascular anatomy, physiology, and surgical techniques. [3]

Compared with old generation CT scanners, 16 slice MDCT and higher scanners yield images with better temporal and spatial resolution, greater anatomic coverage per rotation, more better enhancement with a lesser volume of intravascular contrast media, and higher-quality of multiplanar reconstruction and 3D reconstruction owing to acquisition of an isotropic data set. [4, 5] Rapid imaging with these CT scanners requires less patient sedation or short breath holding time than older-generation CT, MRI, and conventional angiography. [1, 4]

Total anomalous pulmonary circulation (TAPVC) is the abnormal diversion of oxygenated blood into the systemic venous circulation, and mixed blood flows through an interatrial septal defect or a patent foramen ovale to systemic organs. [6] TAPVC is seen in nearly 1.5% of all patients with cardiovascular malformation and in 6.8 per 100,000 live births. [6] The anomalous venous communication can be cardiac, supracardiac, infracardiac, or mixed,

depending on the sites of connection with the supracardiac communication being the commonest. [7]

Aortic arch: Aorta is nicely seen on contrast enhanced CT scan.

- **Left Aortic Arch with an Aberrant Right Subclavian Artery (ARSA):** Here, right common carotid artery arises directly from the aortic arch as the first branch. The second branch is the left carotid artery followed by left subclavian arteries and aberrant right subclavian artery.

- **Double Aortic Arch:** In this anomaly, ascending aorta splits into two arches, the right and the left. The left or anterior arch has a similar course as that of a normal left aortic arch, and the right arch which lies posterior to the left arch courses towards the left side posterior to the esophagus to join the left arch.

- **Interrupted Aortic Arch:** An Interrupted aortic arch is rare anomaly and is characterized by the discontinuity between descending and ascending aorta, without any structural connection between them and thus can be differentiated from severe aortic coarctation or aortic atresia. [8]

- **Right Aortic Arch (RAA):** The RAA is a relatively common anomaly, seen in approximately 0.05% of the population. [9]

Atrial septal defect (ASD): The failure of the septum primum to fuse with the endocardial cushions causes the formation of a primum ASD. Failure of the septum

secundum to grow and cover the ostium primum results in development of a secundum ASD. The septum secundum ASDs are the most common type. [10] [figure 3a]

Ventricular septal defect (VSD): Ventricular septal defects (VSD) are the most common shunt defects in infants. The majority of these are small muscular defects that gradually close. The interventricular septal defect can be divided into four main types-inlet septum, outlet septum, membranous septum and muscular VSD. [10] [figure 7 and 8]

Patent ductus arteriosus: The ductus arteriosus is a communication between the distal aortic arch and the proximal left pulmonary artery that is essential in fetal life for shunting of the mother's oxygenated blood to the systemic fetal circulation. In patients with pulmonary atresia or hypoplastic left heart syndrome, the ductus arteriosus should remain patent for survival. Urgent surgical intervention is required to prevent this communication from constricting and closing within 7–10 days after birth. When the ductus fails to close it results in patent ductus arteriosus (PDA). [10]

Coarctation of aorta: Coarctation of the aorta was first described by Morgagni in 1760. The narrowing is most commonly seen beyond the origin of the left subclavian artery, just distal to it. Interrupted aortic arch is the severest form. The internal mammary arteries serve as collateral channels and are often dilated and easily visible on CTA. CTA can be a useful tool in patient selection and for planning of surgical procedures. Moreover, complications such as restenosis or dissection can be evaluated on CTA. [10]

Dextrocardia: Dextrocardia is defined as cardiac positional anomaly where the heart is located in the right hemithorax with its apex is directed to the right side and caudad. [figure 5,6].

- **Pulmonary Arteries:** CT can be used to assess numerous pulmonary arterial congenital anomalies as well as to assess the postoperative appearance of the pulmonary arterial anatomy. [figure 3b]. Echocardiography is limited in the evaluation of branch pulmonary arteries because of the presence of surrounding air-containing lung. [11, 12] CT is useful in such circumstances as the presence, confluence, patency, or caliber of the pulmonary arteries are clearly seen [13, 14] because these findings can be useful in surgical decision making. CT is useful for noninvasive assessment of pulmonary arterial growth and stenosis, after surgery and shunt placements. [12, 15]
- CT can depict the pulmonary arteries in obstructing lesions of the right ventricular outflow tract, such as fallot's tetralogy and atresia of the pulmonary artery. [12, 16] More than 10% of patients with tetralogy of Fallot have either central or peripheral pulmonary arterial stenosis. [17] [fig4a] In the most severe forms of tetralogy of Fallot, CT can be used to accurately visualize patent and atretic pulmonary arteries. [13, 18]

- **Pulmonary Veins:** CT can be useful to evaluate congenital unilateral pulmonary venous atresia [19] and congenital and acquired pulmonary venous stenosis. [20, 21] CT findings of pulmonary venous atresia are a small ipsilateral hemithorax and pulmonary artery, interlobar septal thickening due to interstitial edema and engorged lymphatic channels along with areas of ground-glass attenuation. [19] Ou et al. [21] found that CT is more sensitive than echocardiography in the detection of pulmonary venous stenosis and is a non invasive alternative to invasive pulmonary venography.

CT findings can be used to diagnose partial and total anomalous pulmonary venous return in pediatric patients. [22, 23, 20, 24, 25] In these conditions, the pulmonary veins drain to another location other than the left atrium. CT is a valuable noninvasive add on to echocardiography in evaluating pulmonary venous structures. [26] [fig 1a and 1b] and [figure 2].

CT is also useful in the follow-up of operated cases of CHD. CTA can be used to confirm stent patency and used to assess various complications, like stent occlusion, stent fracture, stent separation from the vessel wall, residual vessel narrowing, and pseudoaneurysms. [27]

CONCLUSION

CT is a useful imaging modality for the morphologic evaluation of CHD. CT is useful in identifying different types of congenital anomalies of the cardiovascular system in children. It is essentially useful in the evaluation of vascular anomalies located outside the heart and in the assessment of complications that occur post operatively.



Figure 1a and 1b: 6yr old girl who presented with breathlessness and easy fatigability, on CT pulmonary angiography showed right superior and inferior pulmonary veins draining into the right atrium.

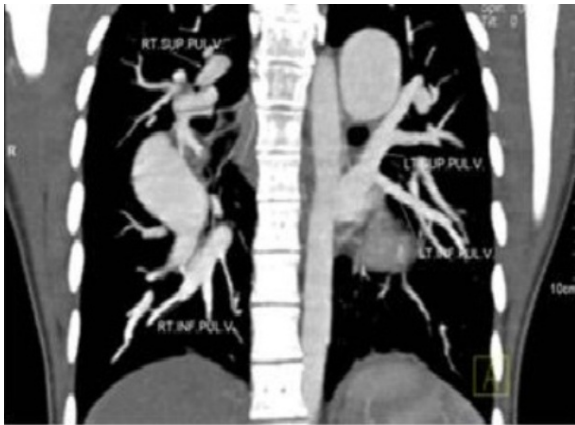


Figure 2: A 6yr old girl who presented with breathlessness and easy fatigability, on CT pulmonary angiography showed left superior and inferior pulmonary veins draining into the left atrium. The figures 1a and 1b of the same patient show the right superior and inferior pulmonary veins draining into the right atrium, suggestive of partial anomalous pulmonary venous drainage.

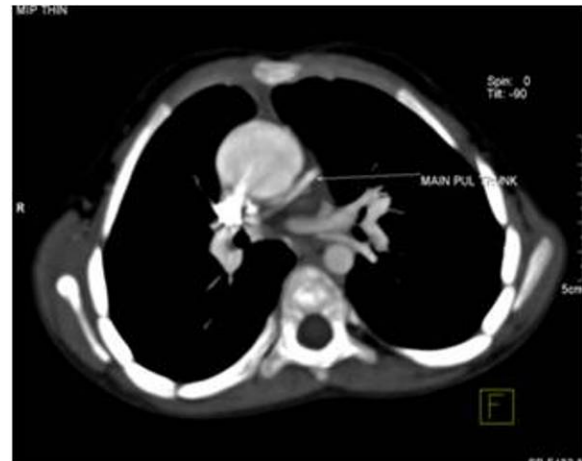


Figure 4a : 6 year boy, a case of hypo plastic pulmonary arteries showing severely hypoplastic main pulmonary artery that measures 4mm.

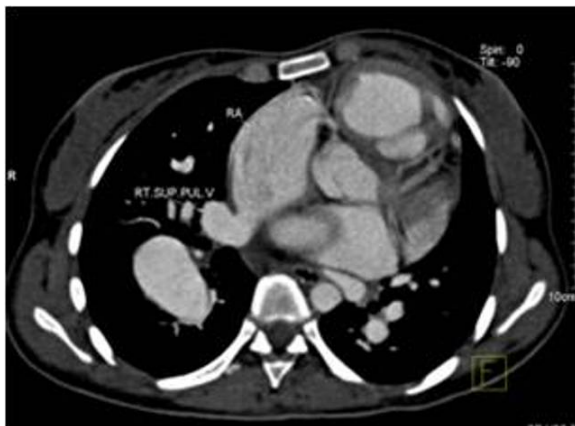


Figure 3a: A 17 yr old girl who presented with palpitations and breathlessness, CT pulmonary angio showed, cardiomegaly with dilated right atrium and right ventricle with an atrial septal defect.

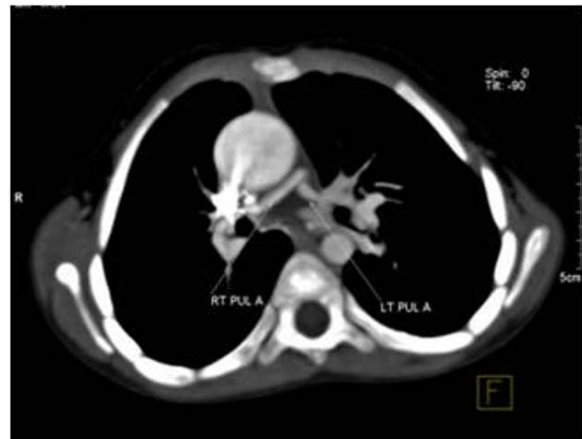


Figure 4b: 6year boy, a case of hypo plastic pulmonary arteries showing severely hypo plastic right and left main pulmonary arteries that measure 4mm and 4.9mm respectively.

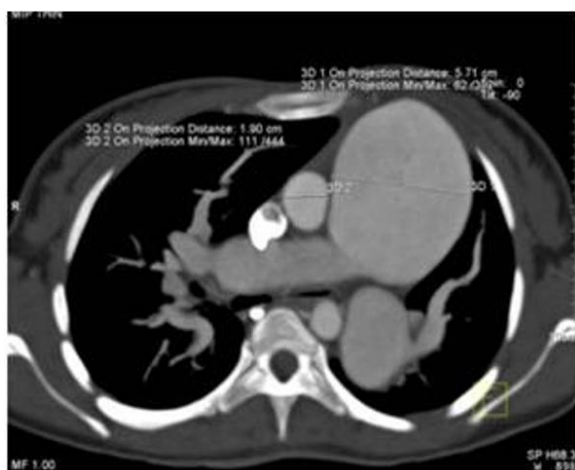


Figure 3b: The same patient as in the previous figure also showed dilated pulmonary artery and its branches, with the main pulmonary artery measuring 57mm, right main pulmonary artery measuring 34mm and left main pulmonary artery measuring 28mm.ascending aorta measures 19mm, suggestive of pulmonary arterial hypertension



Figure 5: A case of dextrocardia with aneurysmal dilatation of the morphological left atrium. In this case the inferior and superior vena cava was draining into the morphological right atrium.



Figure 6a and 6b : The same patient as in figure 5 shows dextrocardia with the morphological positions of left and right ventricles.



Figure 6a and 6b : The same patient as in figure 5 shows dextrocardia with the morphological positions of left and right ventricles.



Figure 7: A 20 day old baby who presented with cyanotic spells since birth, on CT pulmonary angiography shows a small subvalvular type of ventricular septal defect. The patient also had persistent truncus arteriosus.

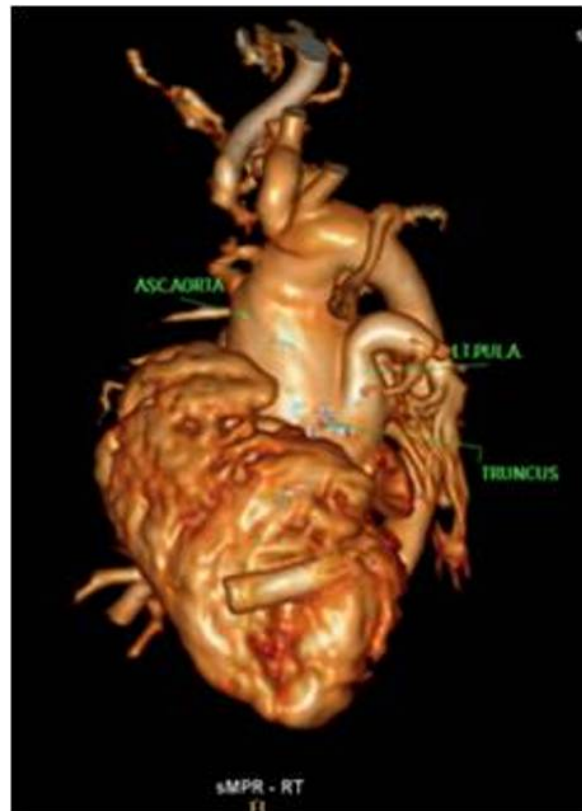


Figure 8 : The same patient as mentioned in figure 8 showing persistent truncus arteriosus, which is a common arterial trunk which has failed to properly divide into the pulmonary trunk and aorta.

REFERENCES:

1. Taylor AM. Cardiac imaging: MR or CT? Which to use when. *Pediatr Radiol* 2008;38 [suppl 3]:S433 -S438
2. Lee T, Tsai IC, Fu YC, et al. Using multidetector-row CT in neonates with complex congenital heart disease to replace diagnostic cardiac catheterization for anatomical investigation: initial experiences in technical and clinical feasibility. *Pediatr Radiol* 2006 ; 36:1273 -1282
3. Gaca AM, Jagers JJ, Dudley LT, Bisset GS 3rd. Repair of congenital heart disease: a primer. Part 1. *Radiology* 2008 ; 247:617 -631
4. Siegel MJ. Multiplanar and three-dimensional multi-detector row CT of thoracic vessels and airways in the pediatric population. *Radiology* 2003;229 : 641-650
5. Spevak PJ, Johnson PT, Fishman EK. Surgically corrected congenital heart disease: utility of 64-MDCT. *AJR* 2008 ; 191:854 -861
6. Correa-Villasenor A, Ferencz C, Boughman JA, Neil CA. Total anomalous pulmonary venous circulation: Familial and environmental factors. *Teratology*. 1991;44:415–28.
7. Chowdhary SK, Bhan A, Sharma R, Airon B, Devagourou V, Saxena A, et al. Total anomalous venous connection: Surgical experience in Indians. *Indian Heart J*. 2001;53:754–60.
8. Aysel Türkvatın, MD, Fatma Gül Büyükbayraktar, MD, Tülay Ölçer, MD, and Turhan Cumhuri, MD Congenital Anomalies of the Aortic Arch: Evaluation with the Use of Multidetector Computed Tomography. *Korean J Radiol* >v.10(2); Mar-Apr 2009 >PMC2651449

9. Stewart JR, Kincaid OW, Edwards JE. An atlas of vascular rings and related malformations of the aortic arch system. Illinois: Springfield; 1964. pp. 12-131.
10. Matthew J. Budoff and Jerold S. Shinbane Editors Associate Editors Stephan Achenbach, Paolo Raggi, and John A. Rumberger Cardiac CT Imaging Diagnosis of Cardiovascular Disease: Congenital Heart Disease and Computed Tomography Jamil AboulHosn and Ronald J. Oudiz page 227.
11. Spevak PJ, Johnson PT, Fishman EK. Surgically corrected congenital heart disease: utility of 64-MDCT. *AJR* 2008 ; 191:854-861
12. Hernandez RJ, Bank ER, Shaffer EM, Snider AR, Rosenthal A. Comparative evaluation of the pulmonary arteries in patients with right ventricular outflow tract obstructive lesions. *AJR* 1987 ; 148:1189-1194
13. Greil GF, Schoebinger M, Kuettner A, et al. Imaging of aortopulmonary collateral arteries with high-resolution multidetector CT. *Pediatr Radiol* 2006;36 : 502-509
14. Frush DP, Herlong JR. Pediatric thoracic CT angiography. *Pediatr Radiol* 2005;35 : 11-25
15. Ley S, Zaporozhan J, Arnold R, et al. Preoperative assessment and follow-up of congenital abnormalities of the pulmonary arteries using CT and MRI. *Eur Radiol* 2007;17 : 151-162
16. Taneja K, Sharma S, Kumar K, Rajani M. Comparison of computed tomography and cineangiography in the demonstration of central pulmonary arteries in cyanotic congenital heart disease. *Cardiovasc Intervent Radiol* 1996; 19:97-100
17. Dabizzi RP, Teodori G, Barletta GA, Caprioli G, Baldrighi G, Baldrighi V. Associated coronary and cardiac anomalies in the tetralogy of Fallot: an angiographic study. *Eur Heart J* 1990 ; 11:692-704
18. Gaca AM, Jagers JJ, Dudley LT, Bisset GS 3rd. Repair of congenital heart disease: a primer. Part 2 *Radiology* 2008 ; 248:44-60
19. Heyneman LE, Nolan RL, Harrison JK, McAdams HP. Congenital unilateral pulmonary vein atresia: radiologic findings in three adult patients. *AJR* 2001;177 : 681-685
20. Gilkeson RC, Ciancibello L, Zahka K. Multidetector CT evaluation of congenital heart disease in pediatric and adult patients. *AJR* 2003; 180:973-980
21. Ou P, Marini D, Celermajer DS, et al. Non-invasive assessment of congenital pulmonary vein stenosis in children using cardiac-non-gated CT with 64-slice technology. *Eur J Radiol* 2008 Mar 18
22. Tsai IC, Chen MC, Jan SL, et al. Neonatal cardiac multidetector row CT: why and how we do it. *Pediatr Radiol* 2008 ; 38:438-451
23. Leschka S, Oechslin E, Husmann L, et al. Pre- and postoperative evaluation of congenital heart disease in children and adults with 64-section CT. *RadioGraphics* 2007;27 : 829-846
24. Goo HW, Park IS, Ko JK, et al. CT of congenital heart disease: normal anatomy and typical pathologic conditions. *RadioGraphics* 2003;23 : S147-S165
25. Kim TH, Kim YM, Suh CH, et al. Helical CT angiography and three-dimensional reconstruction of total anomalous pulmonary venous connections in neonates and infants. *AJR* 2000 ; 175:1381-1386
26. Lee T, Tsai IC, Fu YC, et al. Using multidetector-row CT in neonates with complex congenital heart disease to replace diagnostic cardiac catheterization for anatomical investigation: initial experiences in technical and clinical feasibility. *Pediatr Radiol* 2006 ; 36:1273-1282
27. Fidler JL, Cheatham JP, Fletcher SE, et al. CT angiography of complications in pediatric patients treated with intravascular stents. *AJR* 2000; 174:355-359.