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Diagnosis of Congenital Obstructive Aortic Arch Anomalies in Chinese Children by Contrast-Enhanced Magnetic Resonance Angiography

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ABSTRACT

***Objective*: The purpose of this study was to evaluate the accuracy of contrast-enhanced magnetic resonance angiography for the diagnosis of congenital obstructive aortic arch anomalies in children and compare it with transthoracic echocardiography and other MR imaging techniques (ECG gated T1-weighted spin-echo imaging and gradient-echo cine imaging). *Materials and methods*: Contrast-enhanced magnetic resonance angiography, ECG gated T1-weighted spin-echo imaging, and gradient-echo cine imaging were performed for the diagnosis of congenital obstructive aortic arch anomalies in 416 patients from April 1999 to March 2005 (age range, 3 days to 12 years; mean age, 2.4 years) using a GE 1.5T MR scanner. Transthoracic echocardiography was performed in all patients prior to MR examination. Surgery and/or conventional X-ray angiocardiography were done in all patients to determine the final diagnosis. *Results*: The population consisted of 416 patients. Congenital obstructive aortic arch anomalies were diagnosed in 213 patients and ruled out in 203 patients by operation and/or conventional X-ray angiocardiography. Among the 213 patients with anomalies, coarctation of aorta was diagnosed in 174, interruption of aortic arch was diagnosed in 35, and persistent fifth aortic arch with fourth aortic arch interruption was diagnosed in 4 patients. Among the 35 patients with interruption of aortic arch, 21 were of type A, and 14 were of type B. The diagnostic sensitivity, specificity and accuracy of contrast-enhanced magnetic resonance angiography for congenital obstructive aortic arch anomalies were 98% (208/213), 99% (201/203) and 98% (409/416), respectively. The diagnostic sensitivity, specificity and accuracy of transthoracic echocardiography were 88% (187/213), 92% (186/203) and 90% (373/416), respectively. The diagnostic sensitivity, specificity and accuracy of other MR imaging techniques (ECG gated T1-weighted spin-echo imaging and gradient-echo cine imaging) were 89% (189/213), 84% (170/203) and 86% (359/416), respectively. *Conclusion*: Contrast-enhanced magnetic resonance angiography is a reliable, noninvasive imaging technique for the diagnosis of congenital obstructive aortic arch anomalies in children. Occasionally, even more information can be obtained from this technique than from conventional X-ray angiocardiography. Contrast-enhanced magnetic resonance angiography is superior to transthoracic echocardiography and other MR imaging techniques (ECG gated T1-weighted spin-echo imaging and gradient-echo cine imaging) for diagnosis of congenital obstructive aortic arch anomalies in children.**

Keywords: Cardiac Defect, Congenital, Magnetic Resonance Imaging, Aortic Arch, Coarctation of Aorta.
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Congenital aortic arch anomalies can be classified into two groups: obstructive and non-obstructive congenital abnormalities. The obstructive form is common in children. For many years, conventional X-ray angiocardiography has been considered the gold standard for diagnosis of congenital obstructive aortic arch anomalies. However, the invasive nature of the procedure and the use of ionizing radiation are of concern to many professionals, which hinders its usage. Echocardiography, on the other hand, is noninvasive and can usually provide adequate images for the diagnosis of congenital heart disease. However, it is limited in its ability to visualize the aortic arch. Contrast-enhanced magnetic resonance angiography (CE-MRA) is a fast, non-invasive imaging technique that does not require radiation, making it a safe and useful tool for diagnosis of congenital aortic arch anomalies (1–4). However, application of CE-MRA in large groups of young children has been limited. To our knowledge, until now, application of CE-MRA in large groups of Chinese children has not been reported. The objective of this study was to evaluate the utility of CE-MRA in the diagnosis of obstructive congenital aortic arch anomalies in young pediatric patients and to compare it with transthoracic echocardiography (TTE) and other MR imaging techniques (ECG gated T1-weighted spin-echo imaging and gradient-echo cine imaging).

MATERIALS AND METHODS

The ethics commission of Shanghai Second Medical University approved this study. All examinations were performed between April 1999 and March 2005. CE-MRA, TTE, ECG gated T1-weighted spin-echo imaging, and gradient-echo cine imaging were performed for diagnosis of congenital obstructive aortic arch anomalies in 416 Chinese children (age range, from 3 days to 12 years; mean age 2.4 years). Surgery and/or conventional X-ray angiocardiography were done in all patients to determine final diagnosis.

Of the 416 patients, 213 patients had congenital obstructive aortic arch anomalies. Of the 213 patients, 136 were male, and 77 were female (age range, 3 days to 12 years; mean age 2.3 years). Twenty-two patients were younger than 28 days. Postoperative patients were not included in this study.

All MR examinations were performed with a 1.5T MR unit (Signa Echospeed Excite, General Electricity Medical Systems, Milwaukee, WI, USA). CE-MRA, ECG gated T1-weighted spin-echo sequences and gradient-echo cine sequences were performed on each patient. Phase-contrast cine sequences were performed only in some cases. A body coil, an eight-channel cardiac phased-array coil, or a head coil was used depending on the size of the patient. The scan plane usually was sagittal for congenital obstructive aortic arch anomalies. CE-MRA was performed using a 3-D fast spoiled gradient refocused echo sequence in each case, with the following parameters: minimal repetition time and echo time, matrix of 256 × 192, slice thickness of 2 to 3 mm, field of view of 18 to 42 cm, and bandwidth of 31.25 KHZ. Acquisition time for each sequence ranged from 16 to 25 s. Gadopentate dimeglumine (Magnevist, Schering, Berlin, Germany) and Gadodiamide (Omniscan, Nycomed, Oslo, Norway) were used as contrast medium, with a dose of 0.2 mmol/kg body weight followed by a saline bolus injection. The contrast medium was injected manually. The CE-MRA sequence was initiated about 5 s after the start of the injection of contrast medium. The MR source data set was processed on a General Electricity AW 4.0 workstation with maximum intensity projection algorithm (MIP).

MR examinations were performed with sedation in patients younger than seven years old. Chloral hydrate or Phenobarbital (Luminal) was used for sedation. Approximately 15% of the patients failed the MR examination with sedation and were then given general anesthesia.

TTE examinations were performed in all cases with Philips 5500 or Philips 7500 imaging systems (Philips, Best, The Netherlands). Four cardiologists, (three senior cardiologists and one junior cardiologist) performed all of the examinations. X-ray angiocardiography was performed in all cases with a General Electricity LC-LP biplane digital subtraction angiography machine. Forty-two patients underwent CT angiography with a sixteen-row Multi-detector CT (Lightspeed 16, General Electricity Medical Systems).

RESULTS

Of the 416 patients, 213 patients were diagnosed with congenital obstructive aortic arch anomalies, and 203 patients were ruled out for these conditions by surgery and/or conventional X-ray angiocardiography. These 203 patients served as a control group.

Of the 213 patients with a congenital obstructive aortic arch anomaly, 174 were diagnosed with coarctation of aortic (CoA), 35 were diagnosed with an interruption of aortic arch (IAA), and 4 were diagnosed with a persistent fifth aortic arch with interruption of fourth aortic arch.

Of the 174 patients with CoA, 30 had CoA with an isolated lesion (Fig. 1A, B), 29 had CoA with patent ductus arteriosus (PDA) (Fig. 2), 62 had CoA with ventricular septal defect (VSD), 25 had CoA with VSD and PDA (Fig. 3), 6 had CoA with complex congenital heart diseases (Fig. 4), 11 had CoA with complex congenital heart diseases and PDA, 2 had CoA with simple congenital heart disease, and 9 had CoA with left heart obstructive diseases (4 of the 9 cases also with PDA) (Fig. 5) (Table 1).

Of the 35 patients with IAA, 21 had type A IAA (Fig. 6A, B), and 14 had type B IAA (Fig. 7). Twenty-seven of the patients with IAA also had a VSD and a PDA. Eight patients with IAA also had complex congenital heart diseases including 2 with an aorticpulmonary window (Fig. 8), 2 with a double outlet right ventricle, 2 with a truncus arteriosus (Fig. 9), 1 with a single ventricle and 1 with a complete transposition of great arteries 1 (Table 2).

Of the 4 cases with persistent fifth aortic arch (Fig. 10A, B), all had interruption of forth aortic arch and coarctation of fifth aortic arch.

Surgery and/or conventional X-ray angiocardiography were done in all 213 patients with congenital obstructive aortic arch

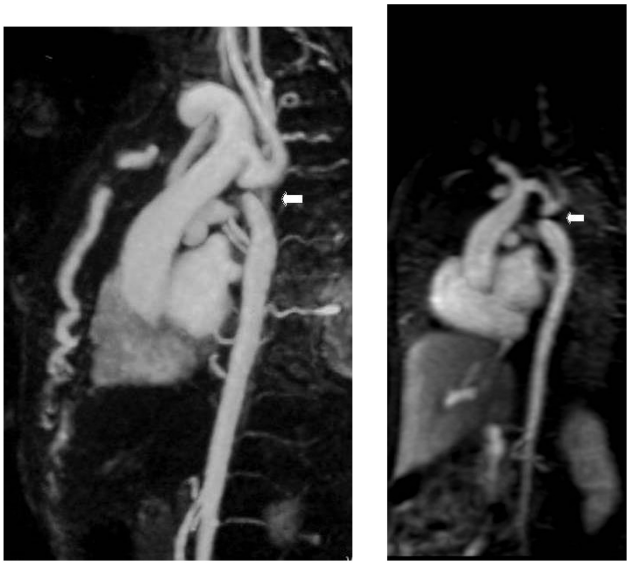


Figure 1. A) Seven-year-old boy with simple CoA. CE-MRA MIP oblique sagittal view image shows the narrowing of aorta (arrow) and collateral vessels. B) Three-month-old boy with simple CoA. CE-MRA MIP oblique sagittal view image shows the narrowing of the aorta (arrow).

anomalies. Of these, 157 underwent both surgery and X-ray angiocardiography, 45 underwent only surgery, and 11 underwent only conventional X-ray angiocardiography. The diagnostic sensitivity, specificity and accuracy of CE-MRA for congenital obstructive aortic arch anomalies were 98% (208/213), 99% (201/203) and 98% (409/416), respectively. The diagnostic sensitivity, specificity and accuracy of TTE were 88% (187/213), 92% (186/203) and 90% (373/416), respectively. Two radiologists reviewed the only other MR imaging data set (ECG gated T1-weighted spin-echo imaging and gradient-echo cine imaging). The diagnostic sensitivity, specificity and accuracy of other MR imaging were 89% (189/213), 84% (170/203) and 86% (359/416), respectively (Table 3).

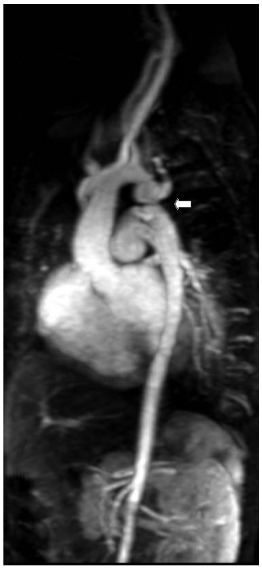


Figure 2. One-year-old boy, CoA with PDA, CE-MRA MIP oblique sagittal image shows the narrowing of the aorta (arrow) and the PDA.

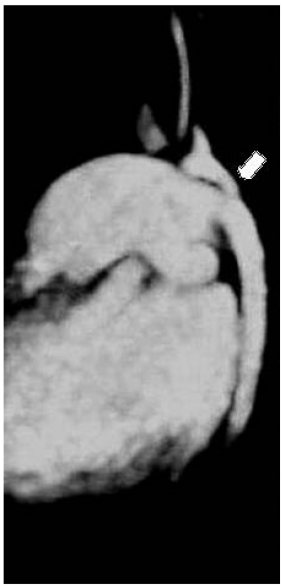


Figure 3. One-year-old boy, CoA with VSD and PDA, CE-MRA MIP oblique sagittal image shows the hypoplastic aortic arch (arrow), dilated pulmonary artery, and the PDA.

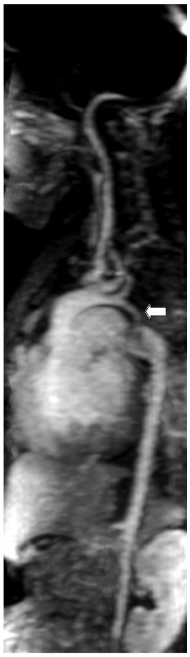


Figure 4. Forty-five-day-old boy, double outlet right ventricle with CoA, CE-MRA MIP oblique sagittal image shows the hypoplastic aortic arch (arrow), dilated pulmonary artery and the aorta arising from the right ventricle.

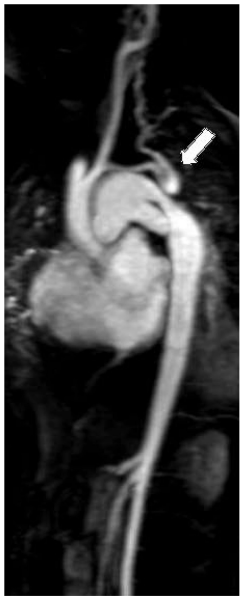
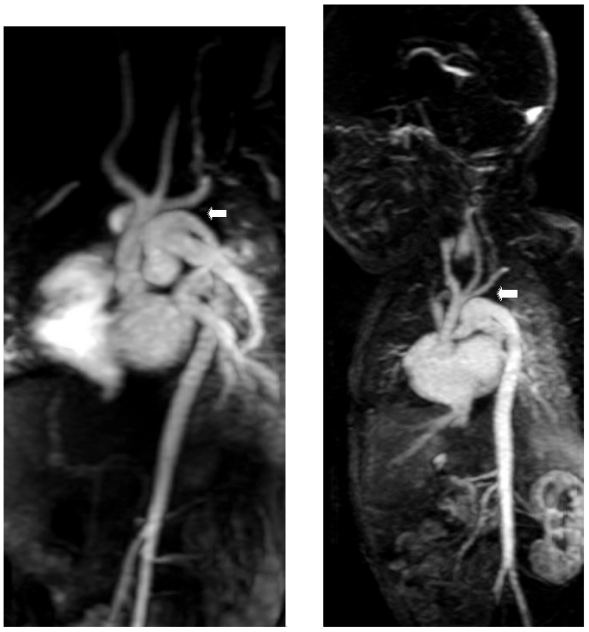


Figure 5. Two-year-old boy, CoA with subvalvular aortic stenosis and PDA. CE-MRA MIP oblique sagittal image shows hypoplastic aortic arch (arrow) and PDA.



(A) (B)

Figure 6. A) Fourteen-month-old boy, type A IAA, CE-MRA MIP oblique sagittal image shows the interruption position (arrow). B) Three-day-old boy, type A IAA, CE-MRA MIP sagittal image shows interruption position (arrow).

Four patients with CoA and 1 with IAA in this study were not accurately diagnosed by CE-MRA. Two patients with Takayasu disease and narrowing of the aortic isthmus (Fig. 11) were incorrectly diagnosed as having a CoA by CE-MRA. However, no patient who was misdiagnosed by CE-MRA was correctly diagnosed by ECG gated T1-weighted spin-echo sequence and gradient-echo cine sequence.

DISCUSSION

Management of patients with congenital obstructive aortic arch anomalies relies predominately on imaging. Routine imaging modalities, such as echocardiography and X-ray angiocardiography, have been successfully complemented by cardiac MR, which provides the capability of imaging in any plane. MR imaging has quickly evolved and 3D contrast-enhanced MR angiography has proved to be particularly valuable in diagnosing some congenital obstructive aortic anomalies (4, 5). It is an accurate technique that allows assessment of congenital malformation of the aorta, pulmonary arteries, and veins.

Table 1. The associated cardiac diseases of CoA

	CoA		
Associated cardiac defect	Without PDA	With PDA	Total
Normal heart	30	29	59
VSD	62	25	87
Complex heart diseases	6	11	17
Simple heart diseases	2		2
Left heart obstructive diseases	5	4	9
Total	105	69	174

The data in this study were acquired from one medical center. The patients in this study were relative young and most of the examinations were performed without patient cooperation (ie, breath holding). The diagnostic accuracy of CE-MRA was

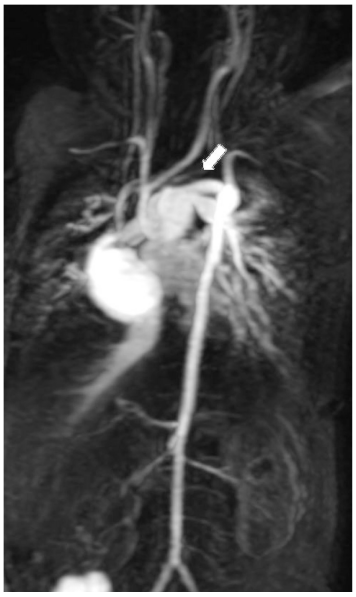


Figure 7. Six-month-old boy, truncus arteriosus with type B IAA, CE-MRA MIP oblique sagittal image shows interruption position (arrow).

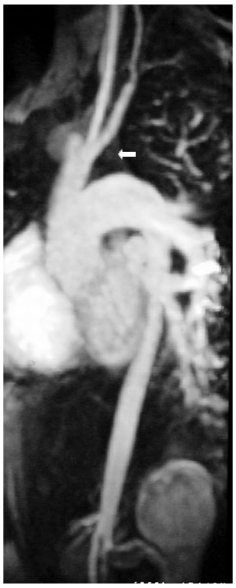


Figure 8. One-year-old girl, aortopulmonary window with type A IAA, CE-MRA image shows the interruption position (arrow).

98%, which was superior to that of TTE (90%) and the other MR imaging techniques (86%). This study demonstrates that CE-MRA is a reliable, noninvasive imaging diagnostic tool for congenital obstructive aortic arch anomalies in children.

TTE is currently the main initial imaging modality for diagnosis of congenital obstructive aortic arch anomalies. In this study, all patients were initially examined by TTE. The cardiologists sent the patient for MR scanning if they could not make a definitive diagnosis or confidently rule out congenital obstructive aortic arch anomalies by TTE. Our result showed the diagnostic accuracy of TTE was 90%. TTE could be complemented by CE-MRA to diagnose congenital obstructive aortic arch anomalies in patients with tortuous or severe narrowing of the aortic arch, exceedingly dilated pulmonary artery or PDA, or older age and complex lesions such as persistent fifth aortic arch (6, 7). In addition, the anatomy and involvement of branch vessels and collateral pathways are better demonstrated by CE-MRA. Moreover, CE-MRA imaging can provide a large field of view, allowing for identification of descending aorta anomalies such as Takayasu disease, which has symptoms that are similar to CoA. CE-MRA can clearly show the abdominal aorta, which is very helpful in differentiating the abdominal aortic narrowing of Takayasu disease from CoA (8).

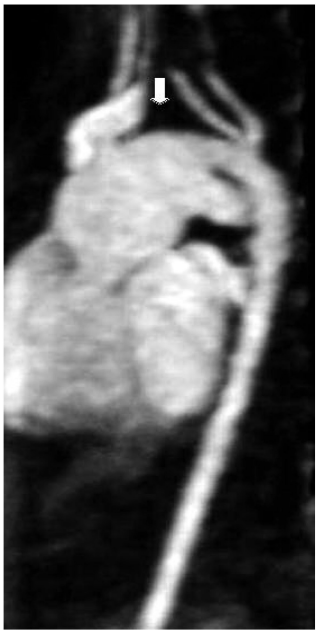


Figure 9. Five-year-old girl, truncus arteriosus with type B IAA and aberrant right subclavian artery, CE-MRA image shows interruption position (arrow) and aberrant right subclavian artery.

Table 2. The associated cardiac diseases of IAA

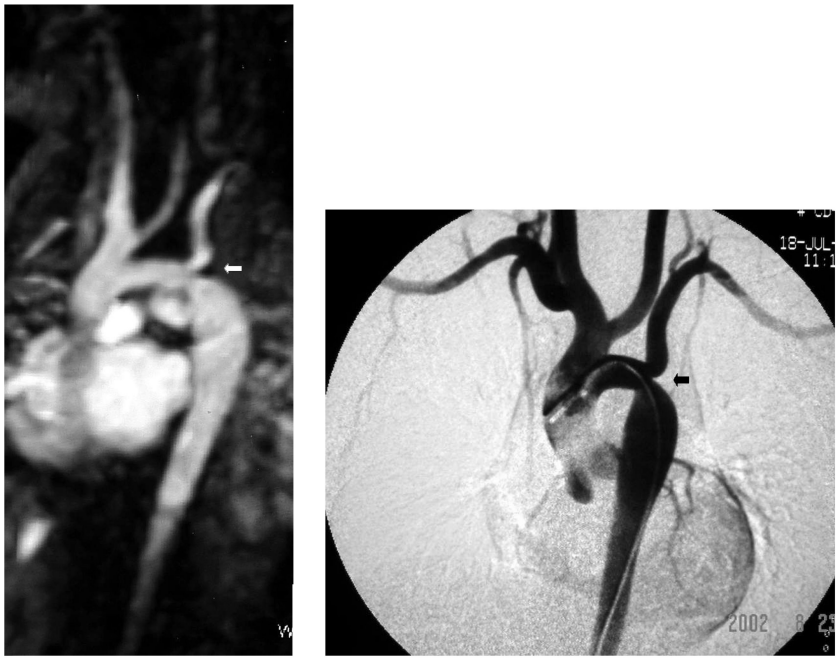
Associated cardiac defect	Interruption of aortic arch Type A	Type B	Total
VSD&PDA	17	10	27
Complex heart diseases	4	4	8
Total	21	14	35

Multi-detector CT (MDCT) angiography is an alternative diagnostic approach for diagnosing of congenital obstructive aortic arch anomalies (9). In this study, 42 patients underwent CT angiography with a sixteen-row MDCT. However, the MDCT imaging quality of these patients was not satisfactory because these patients had required sedation and were not cooperative (ie, unable to hold their breaths); also, the high heart rates of pediatric patients (usually over 120 bpm) interfered with the image quality. Unlike MDCT 2-D axial imaging, CE-MRA was performed using a 3-D sagittal acquisition technique. CE-MRA was able to provide reasonable image quality in sedated patients and in patients with high heart rates. With no ionizing radiation exposure and no known adverse effects associated with the contrast medium (gadolinium-DTPA), CE-MRA is a safer and more suitable imaging technique for small children.

Conventional MR techniques for evaluation of aortic arch anomalies include ECG gated T1-weighted spin-echo imaging and gradient-echo cine imaging. Although these methods are useful for evaluation of congenital heart diseases, they are not as accurate as CE-MRA for diagnosis of congenital obstructive aortic arch anomalies. An oblique sagittal view should be acquired for conventional MR imaging, due to the oblique

Table 3. The diagnostic sensitivity, specificity and accuracy of different imaging modalities for congenital obstructive aortic arch anomalies

Imaging modalities	Sensitivity %	Specificity %	Accuracy %
CE-MRA	98 (208/213)	99 (201/203)	98 (409/416)
TTE	88 (187/213)	92 (186/203)	90 (373/416)
Other MR imaging	89 (189/213)	84 (170/203)	86 (359/416)



(A) (B)

Figure 10. A) Three-year-old girl, persistent fifth aortic arch with interruption of fourth aortic arch and fifth aortic arch narrowing (arrow), CE-MRA imaging. B) Same patient as Fig. 10 A, X-ray angiocardiography imaging shows persistent fifth aortic arch with interruption of fourth aortic arch and fifth aortic arch narrowing (arrow).

orientation of the aortic arch. The image obtained by the technique can be degraded if the child moves or if there is severe aortic arch narrowing and a tortuous aortic arch.

There are several advantages in using CE-MRA for the diagnosis of congenital heart disease (2). CE-MRA 3-D data acquisition can be finished in a short time period (16–25 s in this study). In addition, CE-MRA has the advantage of providing a 3-D data set that can be processed and analyzed using several post reconstruction techniques, such as surface shield display and maximum intensity projection. This also has the advantage that unsuspected findings or abnormalities will not be missed because imaging in a particular plane was not performed. The other advantage of this technique is that CE-MRA can be performed by a technologist and does not require the presence of a cardiologist or radiologist during the acquisition of images (even in complicated cases). CE-MRA is also an excellent technique for the evaluation of extra-cardiac vascular anatomy. The ease of visualizing all vascular anatomy in various projections is its biggest advantage in comparing to other 2-D imaging techniques. This gives doctors the time to consider and it reconstruct at any desired plane. Because CE-MRA can give more information for evaluation of the aortic arch anomalies, it should be used before multi-plane gradient-echo cine sequence in sedated patients.

Conventional X-ray angiocardiography is the gold standard for diagnoses of congenital aortic arch anomalies. In some diseases such as mild CoA with very small PDA, X-ray angiocardiography is superior to CE-MRA. In CoA and IAA with aberrant subclavian artery, X-ray angiocardiography can show blood flow direction through vertebra arteries, which cannot be demonstrated by CE-MRA. In contrast, in those patients who have a very severe CoA and IAA and in whom a catheter cannot be placed into their ascending aorta, CE-MRA can provide more information than X-ray angiocardiography. Also both the ascending aorta and descending aorta can be simultaneously visualized by CE-MRA; this view, which is important for surgical planning, is limited in X-ray angiocardiography. In CoA and IAA with aberrant subclavian artery, CE-MRA cannot show vertebral artery blood flow direction, but phase-contrast imaging can be used as a complementary tool of CE-MRA to show vertebral artery blood flow direction. MR ECG gated T1-weighted spin-echo sequence can show the relationship between the aberrant artery and trachea.

The practical aspect of performing CE-MRA is complicated. The lack of cardiac synchronization leads to blurring of the image due to cardiac motion, which is particularly problematic when assessing intra-cardiac anatomy. CE-MRA can clearly show extra-cardiac anatomy, but it does not resolve intra-cardiac anatomy adequately. Our study demonstrates the main problem of CE-MRA for diagnosis of extra-cardiac obstructive aortic arch anomalies is that it cannot be used to rule out very tiny PDA. Two Takayasu disease cases with aortic isthmus narrowing were incorrectly diagnosed as CoA by CE-MRA; however, this problem may be overcome by the presence of other findings. For example, brachiocephalic artery narrowing was one of the diagnostic clues of Takayasu disease and can be detected.

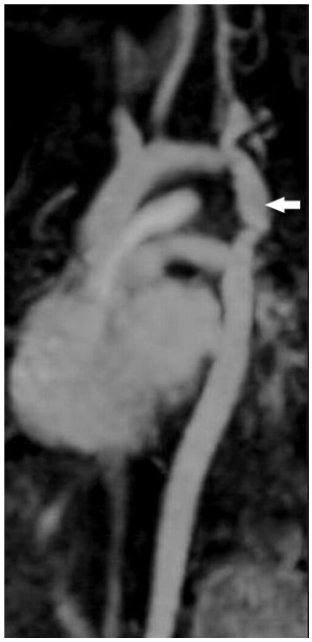


Figure 11. Seven-year-old girl, Takayasu disease with aortic isthmus narrowing (arrow) and brachiocephalic artery narrowing, CE-MRA MIP image shows brachiocephalic artery narrowing.

This study provides information about racial differences in congenital obstructive aortic arch anomalies. In 174 Chinese children with CoA, only 9 patients were CoA with left heart obstructive diseases, which is very low incidence. Type A IAA was more common than type B IAA in these patients, which appears to be different from the incidence seen in children with IAA from western countries (10). However, more children need to be studied to verify this conclusion.

In conclusion, CE-MRA is a reliable, noninvasive imaging technique. CE-MRA is superior to transthoracic echocardiography and other MR imaging techniques for diagnosis of congenital obstructive aortic arch anomalies in children.

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