

● *Original Contribution*

A NEW DIAGNOSTIC APPROACH TO VASCULAR RINGS AND PULMONARY SLINGS: THE ROLE OF MRI

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The conventional diagnostic work-up of a patient suspected of having a vascular cause for stridor, or dysphagia, includes esophagography and bronchoscopy to delineate the abnormal structure without imaging the structure itself. Cine-angiography is regarded as the golden standard, but is not routinely performed. Magnetic resonance imaging (MRI) is non-invasive and has the important advantage over cine-angiography of depicting all structures in the field of view. Color Doppler echocardiography depicts the great vessels, but not the esophagus and trachea. In 14 patients with obstructive symptoms and in four patients without obstructive symptoms MRI successfully imaged the abnormal structure, as was the case in two symptomatic patients using computer tomography. In this series, the findings were confirmed at surgery or by cine-angiography. Conclusion: we suggest that in patients suspected of having a vascular cause for stridor or dysphagia, MRI should be performed. If there is need for a screening procedure, color Doppler echocardiography should be used and if that is equivocal or non-conclusive, esophagography and bronchoscopy should be used. If MRI is difficult to interpret, it should be augmented by magnetic resonance angiography before considering cine-angiography. © 1998 Elsevier Science Inc.

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INTRODUCTION

Findings suggestive of a vascular ring or pulmonary sling will be based on clinical grounds and on routine chest X-ray. The conventional diagnostic work-up includes an esophagogram and bronchoscopy in which indentations in the esophagus and/or trachea and bronchi are correlated with abnormal vascular structures.¹⁻⁷ The need for delineating the actual abnormal structure by cine-angiography is determined by the surgeon's preference. This varies from performing cine-angiography in all cases, to performing cine-angiography only in cases where there is uncertainty with regard to choosing the site where the thoracotomy is to be performed.⁸ In Nikaidoh's series of 68 infants in 1972,⁴ angiography was performed in seven patients, while in Horváth's series of 69 patients in 1992,² cine-angiography combined with

tracheobronchography was routinely done to confirm the diagnosis. If with cine-angiography, the relationship between the abnormal vascular structure and the trachea or bronchi is not apparent, it must be combined with tracheobronchography. Magnetic resonance imaging (MRI), with its multiplanar capabilities, is the most promising of the newer imaging techniques and has the important advantage over cine-angiography of depicting all structures in the field of view. For this reason it has been found to be efficacious in imaging vascular rings and pulmonary slings.^{2,6,9-16} Helical computer tomography (CT) and electron beam CT allow acquisition of a complete volumetric dataset within 30 s, and have a high diagnostic yield.¹⁷⁻¹⁹

Several publications attest to the efficacy of trans-thoracic color Doppler echocardiography (CDE) in imaging these abnormal vascular structures.^{2,20-26} How-

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Table 1. MRI/CT Diagnosis: double aortic arch nine cases

No.	Age (yrs)	Symptoms	Assoc chd	Findings	ANG	BRO	ES	SUR	Corrective surgery
1	1.2	Nil	PA VSD	RDA L>R	+	—	—	+	—
2	0.5	Stridor	—	RDA R>L	—	—	—	+	Division Left Arch
3	0.1	Stridor	—	RDA R>L	—	—	—	+	Division Left Arch
4	0.8	Dysphagia	—	RDA R>L	+	+	+	+	Division Left Arch
5	10.9	Dysphagia	VSD	RDA R>L DK	—	—	—	+	Division Left Arch
		Stridor							
6	21.9	Asthma	—	RDA R>L	—	—	—	+	Division Left Arch
		Stridor							
7	20.4	Dysphagia	—	RDA R>L	—	—	+	+	Division Left Arch
		Stridor							
8	9.0	Dysphagia	—	RDA R>L	—	—	+	+	Division Left Arch
		Stridor							
9	0.3	Resp Insuf	—	RDA R>L	—	+	—	+	Division Left Arch

AGE = age at MRI/CT in years, ASSOC CHD = associated congenital heart disease, ANG = angiography, BRO = bronchoscopy, ES = esophagogram, SUR = surgery, PA VSD = pulmonary atresia and VSD, DK = diverticulum of Kommerell, RDA = right descending aorta, L>R = left arch greater than right arch, R>L = right arch greater than left arch, + = performed, — = not performed, RESP INSUF = respiratory insufficiency.

ever, insonation of ultrasound is limited by bone and air, especially when depicting these often posteriorly running vessels. In addition, the trachea and bronchi cannot be imaged. In two separate studies, success rates of CDE varied between one out of six patients¹⁵ to all of 20 patients²² when using CDE to diagnose a vascular ring or pulmonary sling.

Trans-esophageal CDE can image these posteriorly running vessels, and the technical state of art is such that infants of 2.5 kg can be examined with a biplane probe.²⁷ Disadvantages of trans-esophageal CDE are that it is invasive, and that, particularly in small infants, an already compromised trachea may become more compromised due to the presence of the probe in the esophagus. A third disadvantage of trans-esophageal CDE is that the delineation of the anterior connections of the abnormal vascular structure may not be optimal.

PATIENTS AND METHODS

In this retrospective study we selected patients suspected of having a vascular cause for stridor who had undergone MRI or CT. Patients with a vascular ring or pulmonary sling without obstructive symptoms were also included. For MRI the spin-echo technique with standard and angulated projections was used. The CT images were contrast-enhanced. One observer (RPB) first examined the MRI and CT examinations before confirmation was gained by examining the angiographic or operative findings. The bronchoscopic and esophagographic findings were recorded if these examinations had been performed, but were not separately assessed.

RESULTS

Eighteen patients underwent 19 MRI examinations, one patient being examined twice after a recurrence of

Table 2. MRI/CT Diagnosis: aberrant subclavian artery four cases

No.	Age (yrs)	Symptoms	Assoc chd	Findings	ANG	BRO	ES	SUR	Corrective Surgery
10	0.03	Nil	TOF	Right SA	+	—	+	+	—
11	1.5	Dysphagia	—	Right SA	—	—	+	+	Reimplantation
									Anterior
12	0.2	Stridor	—	SEE TEXT	+	+	+	+	Aortopexy
13	9.7	Nil	PA VSD	Right SA	+	—	—	+	—

AGE = age at MRI/CT in years, ASSOC CHD = associated congenital heart disease, SA = subclavian artery, ANG = angiography, BRO = bronchoscopy, ES = esophagogram, SUR = surgery, TOF = Tetralogy of Fallot, PA VSD = pulmonary atresia and VSD, + = performed, — = not performed.

Case 12 has a left aortic arch, without a right innominate artery. An aberrant vessel arises from the left common carotid artery and runs horizontally across the trachea, causing compression and giving rise to the right common carotid and subclavian arteries.

Table 3. MRI/CT Diagnosis: pulmonary sling 3 cases

No.	Age	Symptoms	Assoc CHD	ANG	BRO	ES	SUR	Corrective Surgery
14	12.4	Nil	COARC	+	-	-	+	-
15	0.3	Stridor RESP INSUF	Nil	-	+	-	+	Reimplantation LPA
16	0.3	Stridor RESP INSUF	Nil	-	+	+	+	Trachea Resection and Anastomosis

AGE = age at MRI/CT in years, ASSOC CHD = associated congenital heart disease, ANG = angiography, BRO = bronchoscopy, ES = esophagogram, SUR = surgery, COARC = Coarctation of the Aorta, LPA = left Pulmonary artery, + = performed, - = not performed, RESP INSUF = respiratory insufficiency.

symptoms (case 6 and case 21 refer to one patient). Cases 9 and 16 underwent CT. In all cases the MRI or CT diagnosis was confirmed by either angiography or by surgical findings. Cases with a double aortic arch are summarized in Table 1. In case 1 the double aortic arch was an incidental finding in a patient with pulmonary atresia and VSD. The double aortic arch was not divided because there was no evidence of obstruction.

Cases with an aberrant subclavian artery are summarized in Table 2. Two of the four patients had obstructive symptoms and were operated on. Case 12 has a left aortic arch, without a right innominate artery. The left common carotid artery gives rise to an aberrant vessel running horizontally across the trachea, causing compression and giving rise to the right common carotid and subclavian arteries. With regard to the imaging of this unusual anomaly, the MRI findings were correctly interpreted after the angiographic images were studied. The three cases with a

pulmonary sling are summarized in Table 3. One asymptomatic patient with coarctation of the aorta was found to have a pulmonary sling as an incidental finding. Case 16 had additional severe tracheal pathology. Eight tracheal rings were circular without a membranous part and had to be resected.

The remaining miscellaneous group of patients is summarized in Table 4. Cases 17 and 19 had previously been operated on for a double aortic arch. Case 17 still had a vascular ring due to a left arterial ligament. In case 19 a dense connective tissue band which appeared not to be an arterial ligament formed a vascular ring between the pulmonary artery and left aortic arch between the left common carotid and subclavian arteries. In case 18 the vascular ring was formed by a left arterial ligament arising out of a retro-esophageal diverticulum from the right descending aorta, causing compression of the esophagus and trachea. An aberrant left subclavian artery also arises from this diverticulum. Case 20 is interesting as she is

Table 4. MRI/CT Diagnosis: miscellaneous group 6 cases

No.	Age (yrs)	Symptoms	Assoc CHD	Findings	ANG	BRO	ES	SUR	Corrective Surgery
17	10.6	Stridor Dysphagia	Nil	Left ART LIG.	+	+	+	+	Division Ligament
18	8.5	REC CHEST INF	Nil	Left ART LIG	-	+	+	+	Division Ligament
19	6.5	Stridor	Nil	Scar Tissue	+	+	+	+	Resection Scar Tissue
20	4.7	Stridor	TOF	B-T Shunt	+	-	-	-	-
21	24.9	Stridor	Nil	Comp. Right Bronchus	-	+	-	+	Dacron Interposition Descending Aorta

AGE = age at MRI/CT in years, ASSOC CHD = associated congenital heart disease, ANG = angiography, BRO = bronchoscopy, ES = esophagogram, SUR = surgery, ART LIG = arterial ligament, REC CHEST INF = recurrent chest infections, TOF = Tetralogy of Fallot, B-T = Blalock-Taussig, COMP = compression, + = performed, - = not performed.

Case 17: Previously operated double aortic arch in which the smaller right dorsal aorta was divided, compression due to a remaining left arterial ligament.

Case 18: Ring formed by a left arterial ligament arising out of a retro-esophageal diverticulum in the right descending aorta causing compression of the oesophagus and trachea. An aberrant left subclavian artery also arises from this diverticulum.

Case 19: Previously operated double aortic arch in which the smaller left arch and arterial duct were divided. Compression was caused by a connective tissue band arising from the left arch between the left common carotid and left subclavian arteries, connecting to the main pulmonary artery.

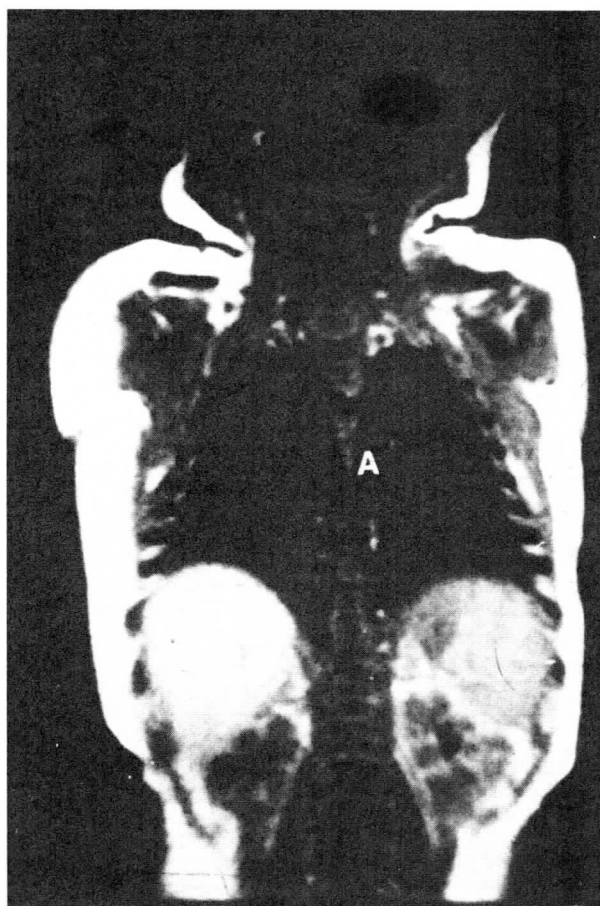
Case 20: Compression on the right lateral wall of the trachea (Figs. 6a, 6b) resulting from scar tissue from previously ligated left and right Blalock-Taussig shunts.

Case 21: This is the same patient as case no. 6. Compression was caused on the right main bronchus by the ascending and descending Aorta (Fig. 7b)

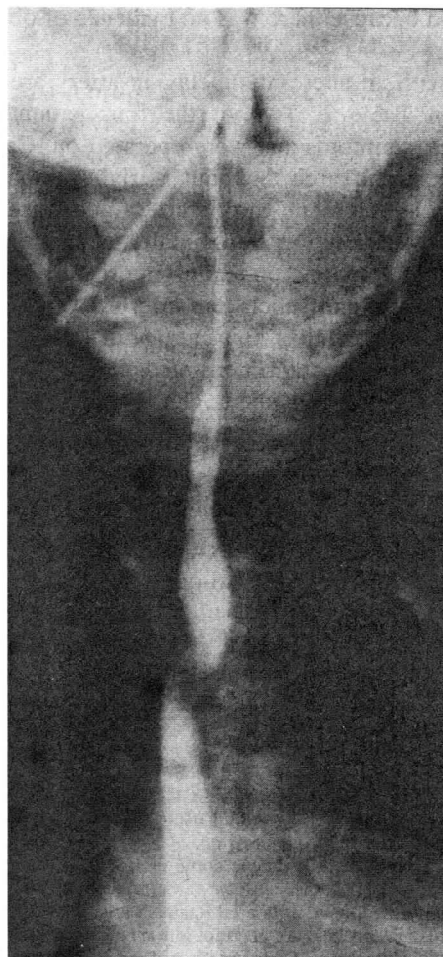


Fig. 1. Case 3. MRI axial section with double aortic arch. The arrowhead points to the smaller left aortic arch. A = ascending aorta, D = descending aorta. The trachea and esophagus can be seen between both arches

the only patient in this series with an acquired vascular ring. This patient with Tetralogy of Fallot was given a left modified Blalock-Taussig shunt at 3 months. Two years later the parents who were Jehovah's witnesses refused total correction. Partial obstruction of this Blalock-Taussig shunt necessitated a right Blalock-Taussig shunt. Mild stridor was noted for the first time thereafter. Total correction of Tetralogy of Fallot with division of both Blalock-Taussig shunts was performed at 3 years and 4 months in another center. Although the compression on the trachea is clearly visible on Fig. 6a, she has mild stridor, and her parents have not opted for surgery as yet. Case 21, previously operated on as case 6 in this series, was diagnosed by MRI and bronchoscopy to have compression of the right main bronchus between the ascending and de-

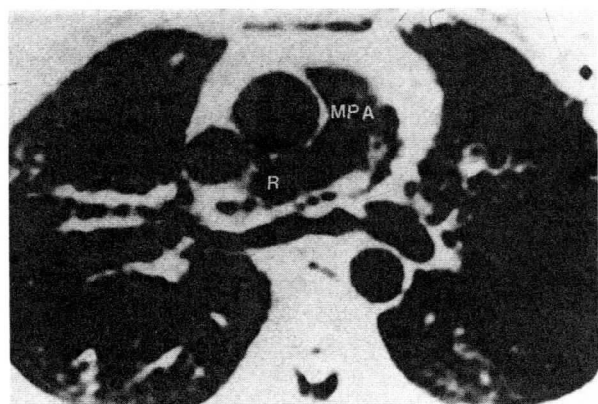


(a)

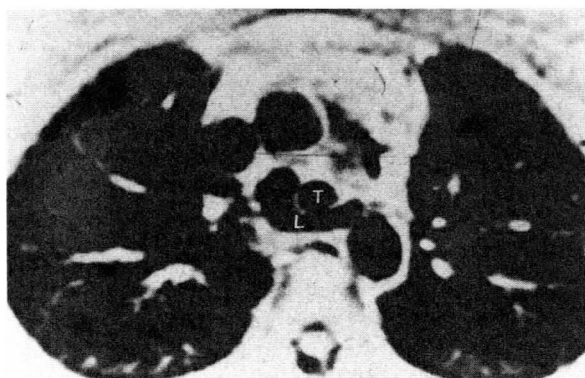


(b)

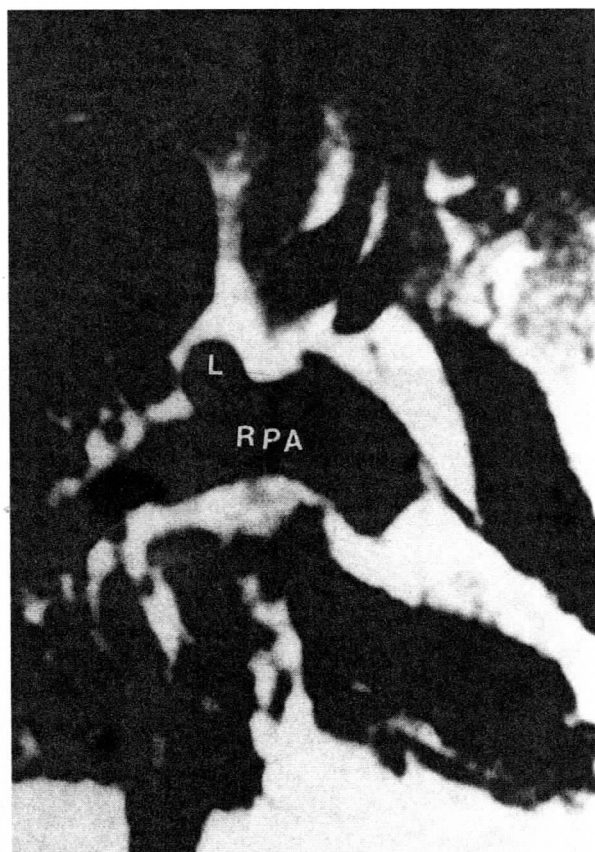
Fig. 2. Case 10. MRI coronal image depicting an aberrant right subclavian artery coming off the descending aorta (A) (a). Case 10. Esophagogram depicting the impression made by an aberrant right subclavian artery (b).



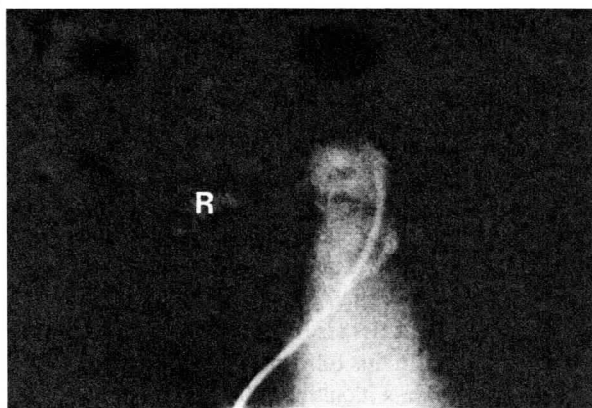
(a)



(b)



(c)



(d)

Fig. 3. Case 14. MRI axial section showing the main pulmonary artery (MPA) and the right (R) pulmonary artery of a patient with a pulmonary sling. A normal origin of the left pulmonary artery cannot be seen (a). Case 14. MRI axial section below Fig. 3a. The left (L) pulmonary artery is seen to pass between the trachea (T) anteriorly and the esophagus posteriorly (b). Case 14. MRI oblique sagittal section depicting the right pulmonary artery (RPA) coming off the main pulmonary artery and giving rise to the aberrant left (L) pulmonary artery (c). Case 14. Angiographic view of the central pulmonary arteries. The aberrant origin of the left pulmonary artery comes from the right (R) pulmonary artery (d).

scending aorta as can be seen on Fig. 7b. To create room for the right main bronchus, the right aortic arch was mobilized to the right by placing a tubular dacron graft in the descending aorta.

DISCUSSION

MRI successfully delineated the anatomy and anomalies of the great vessels in all these patients. This also

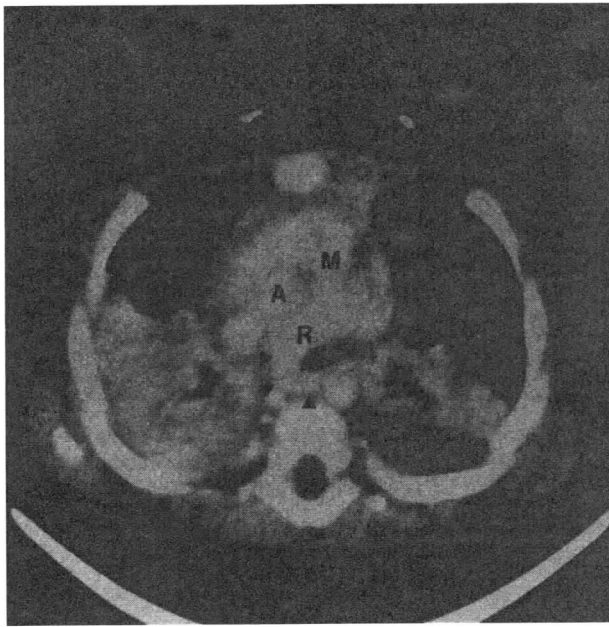


Fig. 4. Case 16. Axial CT section of a patient with a pulmonary sling. A = aorta, M = main pulmonary artery, R = right pulmonary artery. The black arrowheads point to the left pulmonary artery coming off the right pulmonary artery running between the trachea and esophagus before reaching the left lung

applies to the two patients who underwent CT. In all but one of the cases the MRI diagnosis which was evaluated first, was correct. The angiographic and surgical findings were then evaluated and confirmed the MRI and CT findings. The unusual anatomy in case 12 was missed at the initial MRI evaluation, but was seen in retrospect after angiographic images had been studied. Therefore MRI is a very reliable diagnostic procedure to confirm the diagnosis and to give the surgeon the anatomic information of the abnormal vascular structure and its effect on the adjacent tissues, once a vascular cause for stridor is suspected. In this retrospective series there is no discernable pattern in which the different diagnostic modalities were used. Personal preference is probably the single most important factor in choosing the diagnostic tool.

In setting out a diagnostic path in a patient suspected of having a vascular cause for stridor or dysphagia because of a suggestive history and/or chest x-ray, MRI is the next definitive step and further diagnostic steps can then be dispensed with. Only in cases where there is uncertainty with regard to the cause of stridor, is there a role for esophagography, bronchoscopy and CDE. However many patients will have undergone a variety of imaging examinations prior to being referred to a congenital cardiologist.

Whereas, there will be no false negatives with esophagography, other than anterior compression of the trachea caused by the distal origin of the right innominate artery,² which is similar to case 12 in this series, it cannot rightly be regarded as a diagnostic procedure because the abnormal vascular structures are not imaged, but inferred. Esophagography is therefore useful as a screening procedure. Similarly CDE has the inherent problem of not being able to image the trachea and oesophagus. Like esophagography, CDE should strictly speaking be seen as a screening tool. Even cine-angiography has the inherent deficiency of not depicting the esophagus and trachea but does depict the vascular structure without the restriction of a tomographic acquisition. This issue of the relative advantages of a tomographic technique in which all structures are seen in that two-dimensional acquisition against an angiographic technique in which a three-dimensional acquisition of only vessels is presented in two-dimensions, was raised by Singh in a case report in which, as was seen in our case 12, the appreciation of the vascular abnormality occurred only after studying the angiogram.²⁸ This argument also applies to MRI and magnetic resonance angiography

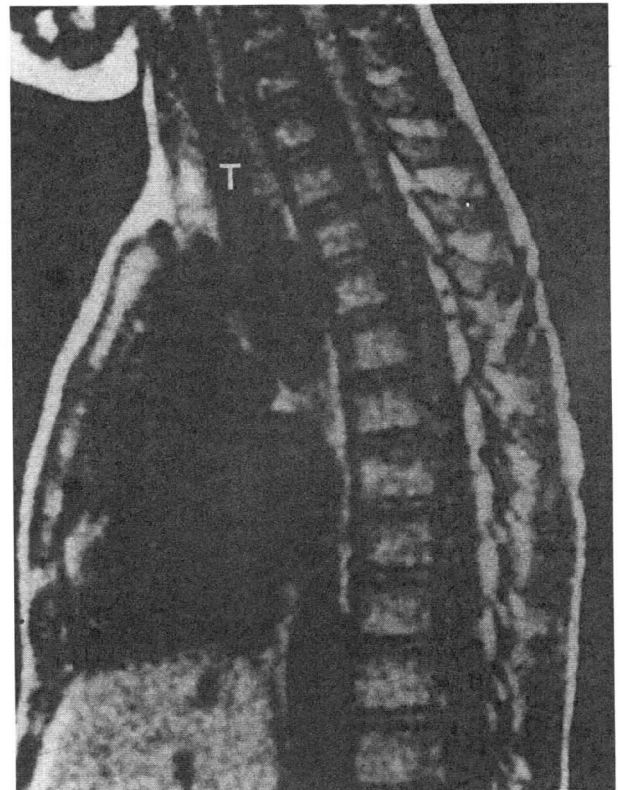
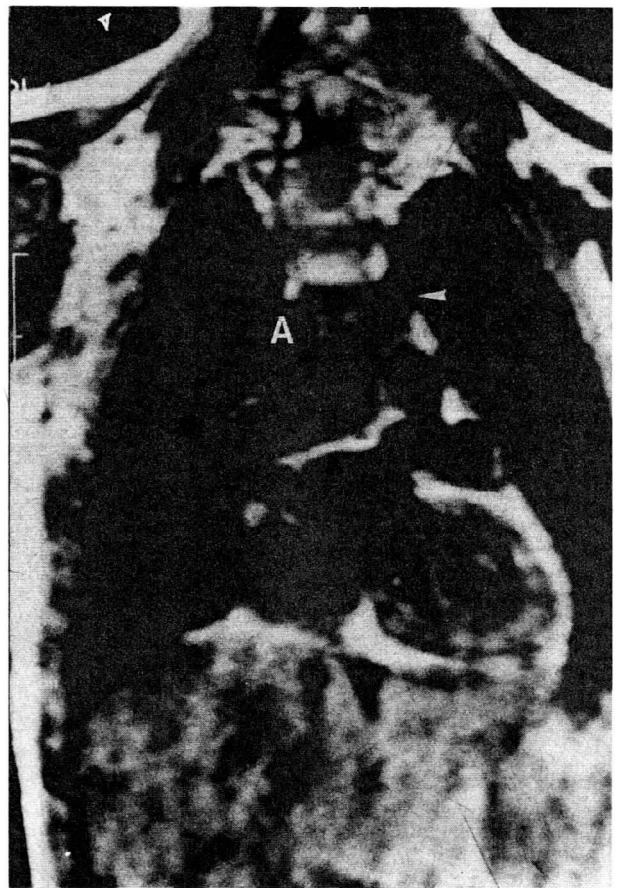


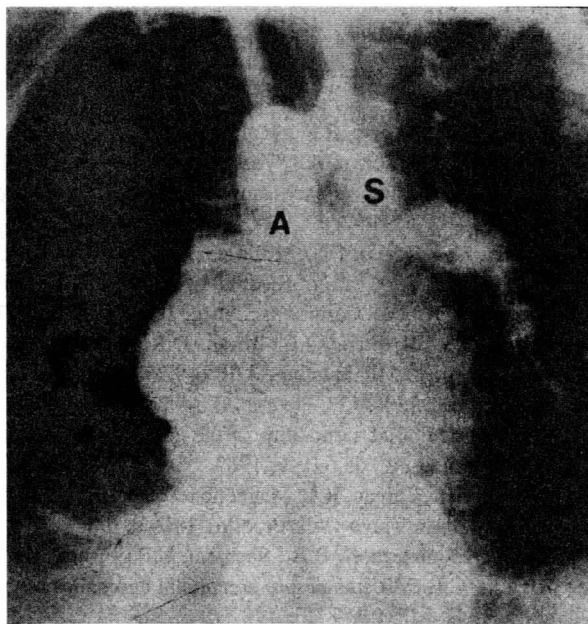
Fig. 5. Case 18. MRI sagittal section depicting posterior compression into the trachea (T) by a diverticulum arising from the right descending aorta



(a)



(b)



(c)

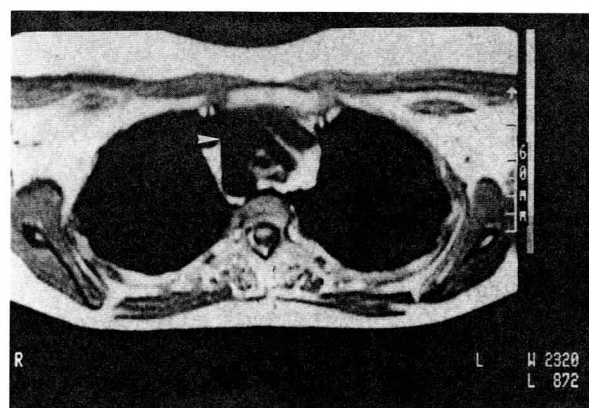
Fig. 6. Case 20. MRI coronal section depicting a right aortic arch (A) compressing the trachea (T). The arrowhead points to the remnant of the left Blalock-Taussig shunt (a). Case 20. MRI coronal section posterior to Fig. 6a (b). Case 20. Cine-angiographic image of right aortic arch (A) and the remnant of the Blalock-Taussig shunt (S) (c).

(MRA), in which case MRA is a non-invasive alternative to cine-angiography which can be performed in the same sitting as MRI. In this series two patients were successfully examined by CT.

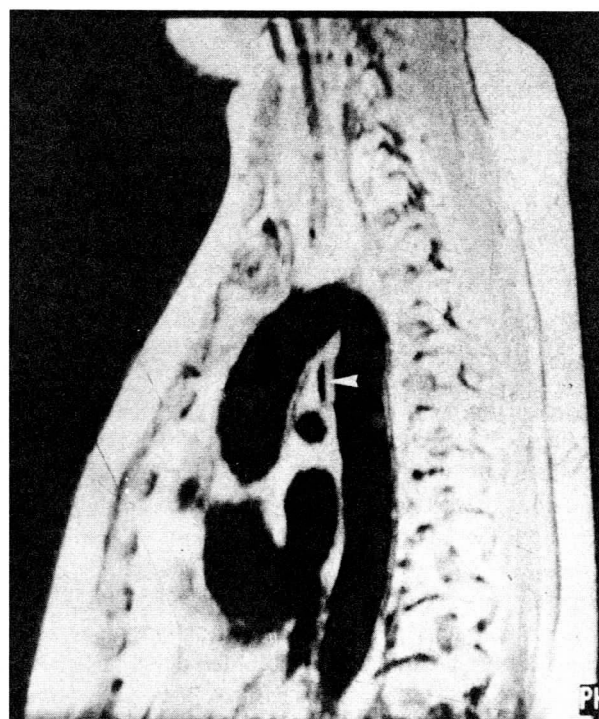
We have no experience with ultra-fast CT in these patients, but in a recent study by Zeidberg *et al.*, helical CT of the upper airway with three dimensional imaging was employed in 10 volunteers and 20 patients with

either intrinsic or extrinsic abnormalities affecting the airway, and was found to significantly assist the understanding of upper airway disease.¹⁹

In conclusion we suggest that in patients suspected of having a vascular cause for stridor or dysphagia, MRI should be performed. If there is need for a screening procedure, CDE and if that is equivocal or non-conclusive, esophagography and bronchoscopy can be used. If



(a)



(b)

Fig. 7. Case 21. MRI axial section in patient who has been operated on for a double aortic arch. The white arrowhead points to the right aortic arch (a). Case 21. MRI sagittal section with white arrowhead pointing to the compression of the right main bronchus (b).

MRI is difficult to interpret, it should be augmented by MRA, before considering cine-angiography.

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