The utility of multiple imaging modalities to diagnose acute aortic dissection

Katrina F. Hurley, MD;* James Ducharme, MD†

ABSTRACT

A 21-year-old man with Marfan syndrome and known aortic root aneurysm presented to our emergency department with symptoms suggestive of acute aortic dissection. The patient was hemodynamically stable and bilateral upper extremity blood pressures were similar. There was no mediastinal widening on portable chest radiograph. Both contrast CT and retrograde angiography of the aorta failed to identify dissection. Subsequent transesophageal echocardiography demonstrated a Stanford classification type A dissection. This case demonstrates the utility of multiple imaging modalities for identifying aortic dissection in high-risk patients.

Key words: aortic dissection, angiography, transesophageal echocardiogram, computed tomography, magnetic resonance imaging, Marfan syndrome

RÉSUMÉ

Un jeune homme de 21 ans atteint du syndrome de Marfan avec présence connue d'un anévrisme de la racine aortique s'est présenté à l'urgence avec des symptômes évoquant une dissection aiguë de l'aorte. Le patient était stable sur le plan hémodynamique et les pressions artérielles bilatérales des membres supérieurs étaient similaires. Les résultats de la radiographie pulmonaire réalisée sur un appareil portatif n'indiquaient pas d'élargissement du médiastin. Ni la tomodensitométrie avec agent de contraste ni l'angiographie rétrograde de l'aorte n'ont permis de déceler la dissection. Une échocardiographie transœsophagienne subséquente a montré qu'il s'agissait d'une dissection de type A selon la classification de Stanford. Ce cas met en évidence l'utilité d'avoir recours à de multiples techniques d'imagerie médicale pour établir un diagnostic de dissection aortique chez les patients à haut risque.

Introduction

Aortic dissection is reported to be one of the "most undiagnosed serious conditions."[1] In one study, nearly 30% of cases were not diagnosed until the post-mortem examination.[2] Given a mortality rate of 1% per hour in patients with untreated thoracic aortic dissection, diagnosis must be prompt.[3,4] Unfortunately, the diagnostic imaging modalities available have limited sensitivity for identifying aortic dissection. Patients with connective tissue disorders, such as Marfan syndrome account for more than 5% of all patients with aortic dissection[1,4] and many of the cases involving younger patients.[5] We describe a case of acute aortic dissection in a young adult with Marfan syndrome for whom contrast CT and retrograde angiography of the aorta were nondiagnostic, while transesophageal echocardiography (TEE) demonstrated a Stanford classification type A aortic dissection.

*Emergency Medicine Resident, Department of Emergency Medicine, Dalhousie University, Halifax, NS, †Clinical Professor of Medicine, Division of Emergency Medicine, McMaster University, Hamilton, Ont.

Received: Mar. 1, 2007; revisions received: July 12, 2007; accepted: Aug. 3, 2007

This article has been peer reviewed.

CJEM 2008;10(1):75-80

Case report

A 21-year-old man presented to our emergency department (ED) after developing severe chest pain during a game of ball hockey. He described his pain as severe retrosternal chest heaviness (graded 8–9 out of 10). His review of systems included shortness of breath, but he denied nausea, vomiting or palpitations. Past medical history was significant for Marfan syndrome diagnosed at age 5 years and related problems, including an aortic root aneurysm, mitral valve prolapse and bilateral lens dislocations. He had a documented prior episode of ventricular tachycardia and was being treated with sotalol 80 mg daily and atenolol 50 mg twice daily.

Initial examination revealed a regular pulse of 72 beats/minute (bpm), a respiratory rate of 20 breaths/minute, and blood pressures of 128/88 mm Hg in the right arm and 118/78 mm Hg in the left arm. Breath sounds were normal bilaterally and cardiac auscultation revealed a normal S_1, a distant S_2 and a grade III/VI pansystolic murmur that was loudest at the apex and radiated to the left axilla. The patient showed physical features consistent with Marfan syndrome, including a marked pectus excavatum. The remainder of the examination was unremarkable.

His electrocardiogram showed a ventricular rate of 68 bpm, downsloping ST-segments in the inferior leads, and evidence of left ventricular hypertrophy and left atrial enlargement. A portable chest radiograph revealed a left heart shift, clear lung fields and scoliosis but no mediastinal widening (Fig. 1). Initial laboratory investigations included a hemoglobin level of 139 g/L, a platelet count of 124×10^9/L and an international normalized ration (INR) of 1.35.

A presumptive diagnosis of acute aortic dissection was made. Non-helical CT with contrast and retrograde aortography failed to demonstrate aortic dissection (Fig. 2). Because clinical suspicion remained high, TEE was undertaken with procedural sedation and analgesia in the ED. This revealed an intimal flap in the ascending aorta proximal to the right coronary artery ostium, extending to the mid–ascending aorta (Fig. 3). Trivial aortic insufficiency and severe mitral insufficiency were also noted. All diagnostic tests were performed within 6 hours. The patient remained hemodynamically stable throughout his ED stay.

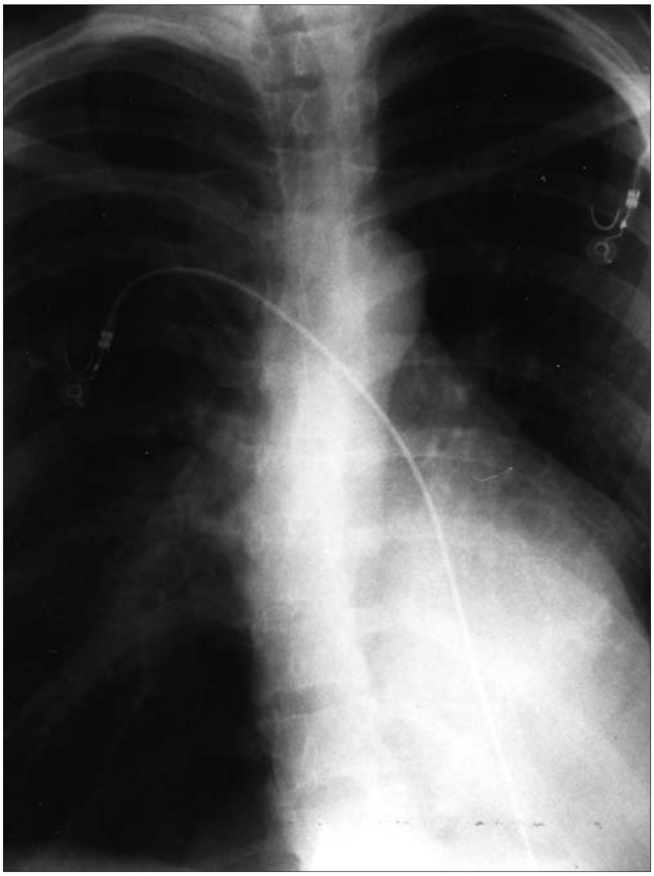

Fig. 1. Portable chest radiograph showing a left heart shift, clear lung fields and scoliosis.

Fig. 2. Retrograde angiography of the aorta showing a grossly dilated aortic root. No dissecting lesion was visible.

https://doi.org/10.1017/S1481803500010046 Published online by Cambridge University Press

The patient was admitted to the intensive care unit and underwent repair with the Bentall procedure.[6] Postoperatively, he developed anterior compartment syndrome in the right leg, necessitating fasciotomy. He recovered well and was discharged home in stable condition.

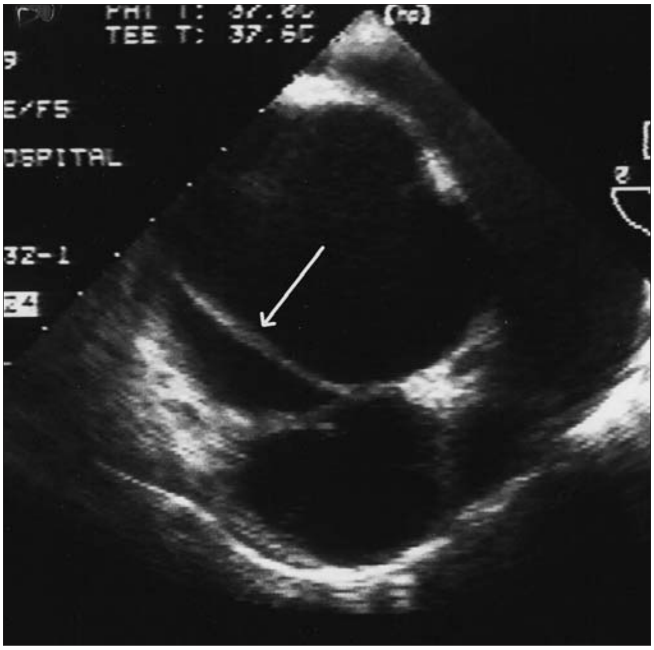

Fig. 3. Transesophageal echocardiography demonstrating dilated aortic root and dissection flap (arrow).

Discussion

Aortic dissection occurs when an intimal tear develops, allowing blood to penetrate the aortic wall, dissect longitudinally through the media and form a false lumen.[1] Conditions associated with medial degeneration, such as connective tissue disorders (e.g., Marfan syndrome) and hypertension, increase the risk for dissection. There are 4 main imaging modalities used to diagnose aortic dissection: retrograde angiography; ultrasound, including transesophageal TEE; CT scanning; and magnetic resonance imaging (MRI).

Retrograde angiography is the historical criterion standard for diagnosing aortic dissection. However, retrograde angiography is invasive and may extend the dissection (Table 1). The patient is also exposed to contrast media and radiation. Retrograde angiography may appear inappropriately normal if the false tract has thrombosed. Recent studies suggest that less invasive studies such as TEE, CT scanning and MRI provide excellent diagnostic accuracy and mitigate some of the risks inherent to retrograde angiography (Table 2).[7–19] Unfortunately, the methodology of the research studies supporting the use of these alternative imaging modalities may make their results inappropriate for populations with a relatively low prevalence of aortic dissection.

Growing familiarity with ultrasound technology and the

Table 1. Comparison of the advantages and disadvantages of TEE, CT, MRI and retrograde angiography of the aorta in diagnosis of acute aortic dissection

Advantages and disadvantages	TEE	CT	MRI	Angiography
Advantages				
Performed at bedside	x			
No radiation	x		x	
Expeditious	x	x		
Assesses entire aorta and branches		x	x	x
Potentially identifies alternative diagnoses		x	x	
Specific advantages	Evaluation of cardiac function and flow in true/false lumens	Most readily available		
Disadvantages				
Invasive				x
Requires experienced operator/interpreter	x		x	
Expensive			x	x
Requires contrast		x		x
Specific risks	Aspiration; exacerbation of hypertension		Contraindicated in patients with some types of aneurysm clips or claustrophobia	Retrograde extension of dissection; cholesterol embolization syndrome; falsely negative when false lumen thrombosed

TEE = transesophageal echocardiography; MRI = magnetic resonance imaging.

https://doi.org/10.1017/S1481803500010046 Published online by Cambridge University Press

use of multi-planar (M-mode) echocardiography rather than 2-dimensional (biplanar) scans have led to higher diagnostic sensitivity and specificity — approaching 100% — for aortic dissection.[7,10,13,14,16,17,19,20] In particular, TEE is reliable for localizing intimal tears and has the added potential benefit of assessing valve function and flow in false lumens.[17,21,22] In addition, TEE may be performed at the bedside in the ED, yielding advantages for hemodynamically unstable patients (Table 2). However, TEE requires esophageal intubation, which may increase systolic blood pressure in awake or inadequately sedated patients, increasing the risk of extension of the dissection or aortic rupture.[23]

CT is now the most frequently ordered diagnostic imaging modality for the initial evaluation of patients with suspected aortic dissection.[24] Multi-detector row CT is the most rapid diagnostic test for aortic dissection, with data acquisition accomplished in less than 30 seconds.[18] Reported sensitivities and specificities range from 79% to 100% for CT, but early studies should be interpreted with caution as this technology is evolving rapidly.[7,11,12,15,16,18] Combining colour Doppler transthoracic echocardiography (TTE) with CT increases diagnostic accuracy. However, TTE may be limited in patients with chest deformities such as those with Marfan syndrome and pectus excavatum.[16]

In experienced hands, MRI is highly sensitive and specific for aortic dissection.[8,9,11,15,16] However, MRIs are not universally available and image acquisition time is up to 30 minutes.[18] This relatively prolonged time away from the ED places hemodynamically tenuous patients at risk for delayed aggressive stabilization should they decompensate during the study. Decreasing the duration of MRI studies would, at least partially, mitigate this concern. Two small studies have described faster MRI techniques

Table 2. Studies assessing the characteristics of diagnostic imaging in aortic dissection

Study	Period	Prevalence, %	Criterion standard	*n*	No. of confirmed cases*	Sensitivity, %	Specificity, %
Erbel et al[7]	1983–87	50	OR findings, autopsy or agreement on 2 of 3 imaging studies	164	47†	TEE 99 CT 83 Angio 88	TEE 98 CT 100 Angio 94
Nienaber et al[8]	1988–91	58	OR findings, autopsy or angiography	53	31	TEE 100 MRI 100	TEE 68 MRI 100
Nienaber et al[9]		74	OR findings, autopsy or angiography	35	21	TEE 100 MRI 100	TEE 78 MRI 100
Chirillo et al[10]	1990–93	57	OR findings, autopsy or 6 mo of follow-up	70		TEE 98 Angio 88	TEE 97 Angio 97
Sommer et al[11]		67	OR findings, autopsy, angiography or at least 3 mo of follow-up	49	28	TEE 100 CT 100 MRI 100	TEE 94 CT 100 MRI 94
Small et al[12]	1990–95	32	OR findings, autopsy, other imaging or follow-up	81	17	CT 96	CT 96
Keren et al[13]	1991–94	43	OR findings, autopsy or other imaging	112	60	TEE 95	TEE 98
Evangelista et al[14]		49	OR findings, autopsy, CT/MRI	132	29	TEE 97	TEE 100
von Kodolitsch et al[15]		64	OR findings, autopsy or angiography	120	105	TEE 100 CT 83 MRI 100	TEE 88 CT 90 MRI 96
Losi et al[19]		65	OR findings or angiography	46	45	TEE 97	TEE 100
Kodolitsch et al[16]	1984–94	45	OR findings, autopsy or angiography	86	59	TEE 100 CT 79 MRI 100	TEE 96 CT 87 MRI 96
Pepi et al[17]	1995–98	62	OR findings, autopsy or angiography	86	59	TEE 100	TEE 100
Hayter et al[18]‡	2002–03	18	OR findings, autopsy, DC Dx, follow-up or other imaging	373	37	CT 99	CT 100

OR = operating room; TEE = transesophageal echocardiography; angio = angiography; MRI = magnetic resonance imaging; DC Dx = discharge diagnosis.
*No. of anatomically confirmed cases of aortic dissection (e.g., OR findings or autopsy).
†Registry study.
‡Study included disorders other than aortic dissection.

https://doi.org/10.1017/S1481803500010046 Published online by Cambridge University Press

that do not require either electrocardiographic gating or breath-holding.[25,26] If these techniques, which can be performed in less than a minute, prove accurate, MRI may become the imaging modality of choice for evaluating patients for aortic dissection. Of note, MRI is an observer-dependant technology with reported sensitivities ranging from 52% to 100% depending on the experience of the radiologist.[21,27]

A recent systematic review compared the diagnostic accuracy of TEE, CT scanning and MRI. The authors concluded that these studies are equally reliable for diagnosing or ruling out thoracic aortic dissection.[28] The 95% confidence intervals for sensitivity were 95%–99% for TEE, 96%–100% for CT scanning and 95%–99% for MRI. One interpretation of these findings is that up to 5% of thoracic aortic dissections can be missed when only a single imaging modality is used to diagnose aortic dissection.[28] The 95% confidence intervals for specificity were 92%–97% for TEE, 87%–99% for CT scanning and 95%–100% for MRI. The authors found significant heterogeneity in the study populations. Unfortunately, pooling the results from heterogeneous populations may yield invalid results.[29] In addition, the authors of this systematic review do not discuss the impact of one negative test result on the test characteristics of subsequent tests. Further, the studies included in this systematic review did not adequately take into account pretest probability.

Two studies did take into account sequential diagnostic imaging modalities and pretest probability. A decision analysis by Sarasin and colleagues concluded that, in patients with low pretest probability ($< 15\%$), a single negative TEE, CT, MRI or aortic angiogram is sufficient to rule out dissection, while in patients with higher pretest probability, additional imaging is required.[30] Unfortunately, this model has not been validated and is limited by the quality of published data and the ability of clinicians to determine pretest probability. An analysis by Barbant and colleagues also discusses the importance of pretest probability and the application of Bayes' theorem.[31] These authors reported that when the prevalence of disease was varied from intermediate (10%) to high (50%), the negative predictive value of CT decreased from 98% to 85%, reflecting the need for a second diagnostic test in high-risk populations.[31] In our very high–risk patient, 3 imaging modalities were needed to convincingly diagnose aortic dissection.

Conclusion

Our case demonstrates that for high-risk patients multiple imaging modalities may be needed to adequately diagnose aortic dissection. Given the associated mortality, reliance on a single imaging modality to rule out aortic dissection in high-risk patients is unwise.

Acknowledgements: We would like to thank Dr. Doug Baggs for his help with the images in this case.

Competing interests: None declared.

References

1. Robicsek F, Thubrikar MJ. Hemodynamic considerations regarding the mechanism and prevention of aortic dissection. Ann Thorac Surg 1994;58:1247-53.
2. Spittell PC, Spittell JA Jr, Joyce JW, et al. Clinical features and differential diagnosis of aortic dissection: experience with 236 cases (1980 through 1990). Mayo Clin Proc 1993;68:642-51.
3. Hirst AE Jr, Johns VJ Jr, Kime SW Jr. Dissecting aneurysm of the aorta: a review of 505 cases. Medicine (Baltimore) 1958;37: 217-79.
4. Hagan PG, Nienaber CA, Isselbacher EM, et al. The International Registry of Acute Aortic Dissection (IRAD): new insights into an old disease. JAMA 2000;283:897-903.
5. Januzzi JL, Marayati F, Mehta RH, et al. Comparison of aortic dissection in patients with and without Marfan's syndrome (results from the International Registry of Aortic Dissection). Am J Cardiol 2004;94:400-2.
6. Bentall HH. Operation for ascending aortic aneurysm and aortic regurgitation — pathological influence on survival. Jpn J Surg 1987;17:425-30.
7. Erbel R, Engberding R, Daniel W, et al. Echocardiography in diagnosis of aortic dissection. Lancet 1989;1:457-61.
8. Nienaber CA, Spielmann RP, von Kodolitsch Y, et al. Diagnosis of thoracic aortic dissection. Magnetic resonance imaging versus transesophageal echocardiography. Circulation 1992;85:434-47.
9. Nienaber CA, von Kodolitsch Y, Brockhoff CJ, et al. Comparison of conventional and transesophageal echocardiography with magnetic resonance imaging for anatomical mapping of thoracic aortic dissection. A dual noninvasive imaging study with anatomical and/or angiographic validation. Int J Card Imaging 1994;10:1-14.
10. Chirillo F, Cavallini C, Longhini C, et al. Comparative diagnostic value of transesophageal echocardiography and retrograde aortography in the evaluation of thoracic aortic dissection. Am J Cardiol 1994;74:590-5.
11. Sommer T, Fehske W, Holzknecht N, et al. Aortic dissection: a comparative study of diagnosis with spiral CT, multiplanar transesophageal echocardiography and MR imaging. Radiology 1996;199:347-52.
12. Small JH, Dixon AK, Coulden RA, et al. Fast CT for aortic dissection. Br J Radiol 1996;69:900-5.

13. Keren A, Kim CB, Hu BS, et al. Accuracy of biplane and multiplane transesophageal echocardiography in diagnosis of typical acute aortic dissection and intramural hematoma. J Am Coll Cardiol 1996;28:627-36.

14. Evangelista A, Garcia-del-Castillo H, Gonzalez-Alujas T, et al. Diagnosis of ascending aortic dissection by transesophageal echocardiography: utility of M-mode in recognizing artifacts. J Am Coll Cardiol 1996;27:102-7.

15. von Kodolitsch Y, Simic O, Nienaber CA. Aneurysms of the ascending aorta: diagnostic features and prognosis in patients with Marfan's syndrome versus hypertension. Clin Cardiol 1998;21: 817-24.

16. Kodolitsch Y, Krause N, Spielmann R, et al. Diagnostic potential of combined transthoracic echocardiography and x-ray computed tomography in suspected aortic dissection. Clin Cardiol 1999;22:345-52.

17. Pepi M, Campodonico J, Galli C, et al. Rapid diagnosis and management of thoracic aortic dissection and intramural haematoma: a prospective study of advantages of multiplane vs. biplane transoesophageal echocardiography. Eur J Echocardiogr 2000;1:72-9.

18. Hayter RG, Rhea JT, Small A, et al. Suspected aortic dissection and other aortic disorders: multi-detector row CT in 373 cases in the emergency setting. Radiology 2006;238:841-52.

19. Losi MA, Betocchi S, Briguori C, et al. Determinants of aortic artifacts during transesophageal echocardiography of the ascending aorta. Am Heart J 1999;137:967-72.

20. Vignon P, Spencer KT, Rambaud G, et al. Differential transesophageal echocardiographic diagnosis between linear artifacts and intraluminal flap of aortic dissection or disruption. Chest 2001;119:1778-90.

21. Wiet SP, Pearce WH, McCarthy WJ, et al. Utility of transesophageal echocardiography in the diagnosis of disease of the thoracic aorta. J Vasc Surg 1994;20:613-20.

22. Mastrogiovanni G, Masiello P, Leone R, et al. Emergency surgical management of acute aortic dissection: role of transesophageal echocardiography. G Ital Cardiol 1999;29:1137-41.

23. Silvey SV, Stoughton TL, Pearl W, et al. Rupture of the outer partition of aortic dissection during transesophageal echocardiography. Am J Cardiol 1991;68:286-7.

24. Moore AG, Eagle KA, Bruckman D, et al. Choice of computed tomography, transesophageal echocardiography, magnetic resonance imaging, and aortography in acute aortic dissection: International Registry of Acute Aortic Dissection (IRAD). Am J Cardiol 2002;89:1235-8.

25. Nitatori T, Yokoyama K, Nakamura A, et al. Clinical application of single shot GRE-EPI as non-enhanced MRA (EPI-MRA) for aortic aneurysm and dissection. Radiat Med 1999;17:393-7.

26. Summers RM, Sostman HD, Spritzer CE, et al. Fast spoiled gradient-recalled MR imaging of thoracic aortic dissection: preliminary clinical experience at 1.5 T. Magn Reson Imaging 1996;14:1-9.

27. Kersting-Sommerhoff BA, Higgins CB, White RD, et al. Aortic dissection: sensitivity and specificity of MR imaging. Radiology 1988;166:651-5.

28. Shiga T, Wajima Z, Apfel CC, et al. Diagnostic accuracy of transesophageal echocardiography, helical computed tomography, and magnetic resonance imaging for suspected thoracic aortic dissection: Systematic review and meta-analysis. Arch Intern Med 2006;166:1350-6.

29. Egger M, Smith GD. Misleading meta-analysis. BMJ 1995;310: 752-4.

30. Sarasin FP, Louis-Simonet M, Gaspoz JM, et al. Detecting acute thoracic aortic dissection in the emergency department: time constraints and choice of the optimal diagnostic test. Ann Emerg Med 1996;28:278-88.

31. Barbant SD, Eisenberg MJ, Schiller NB. The diagnostic value of imaging techniques for aortic dissection. Am Heart J 1992;124: 541-3.

Correspondence to: Dr. Katrina Hurley, Emergency Department, QEII, Halifax Infirmary — Rm 3021, 1796 Summer St., Halifax NS B3H 3A7**;** kfhurley@dal.ca

https://doi.org/10.1017/S1481803500010046 Published online by Cambridge University Press