Journal of Cardiology Cases (2012) 5, e51—e54

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Case Report

Supravalvular thrombus after pulmonary artery banding and fontan procedure evaluated by multidetector-row computed tomography

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Received 25 May 2011; received in revised form 21 August 2011; accepted 6 September 2011

KEYWORDS
Fontan procedure;
Multidetector-row computed tomography;
Thrombosis;
Anticoagulation therapy;
Congenital heart disease

Summary The mechanisms responsible for thromboembolic events in children with congenital heart disease have not yet been fully elucidated. Furthermore, establishment of long-term anticoagulation therapy in Fontan patients remains controversial. Here, we report the case of a 9-year-old boy who presented with hemiparesis due to a thromboembolic stroke; the boy had previously undergone staged pulmonary artery banding and Fontan procedure. Cardiac multidetector-row computed tomography (MDCT) clearly showed the supravalvular thrombus at the roofed (blind) pulmonary valve and circulatory stasis, which could be considered a possible source of the thrombus. Follow-up CT examination showed that the thrombus disappeared, but the circulatory stasis remained. Therefore, because the risk of thrombus formation was not eliminated, anticoagulation therapy was continued for the patient. Our case indicates the possible application of cardiac MDCT for providing insight into the hemodynamic mechanisms responsible for the thromboembolic events in children with congenital heart disease.

Introduction

Thromboembolic complication is one of the major factors contributing to late morbidity and prognosis in children with congenital heart disease after Fontan procedure [1—4]. Several researchers have investigated the mechanism of thrombus formation [5], but it is difficult to identify the

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doi:10.1016/j.jccase.2011.09.004

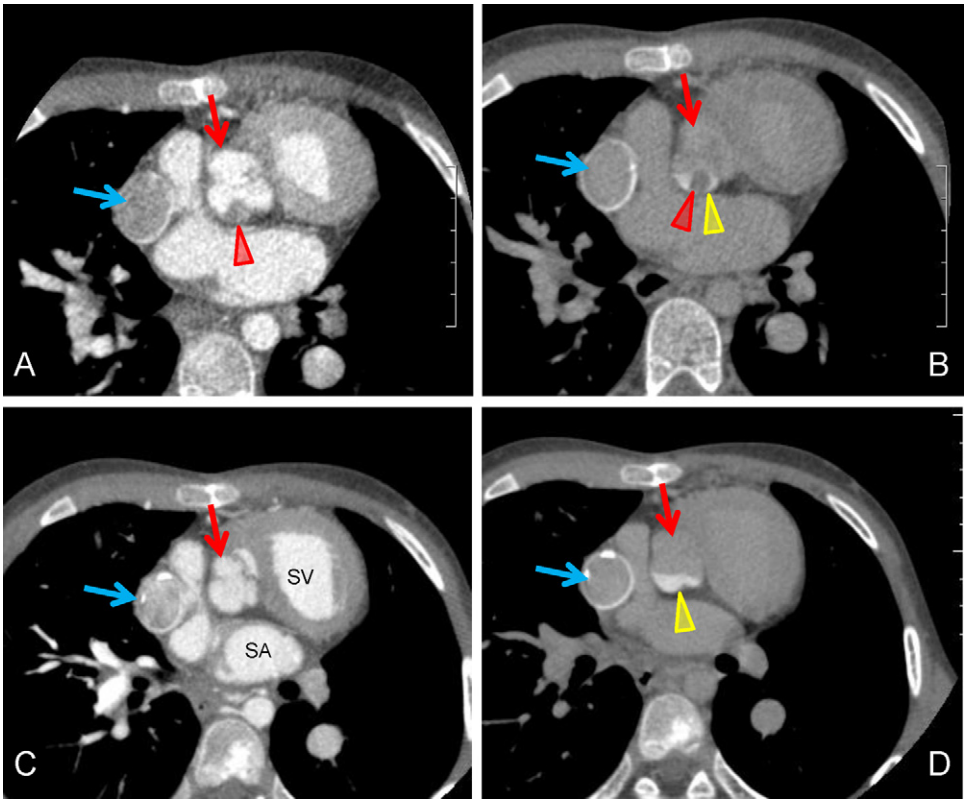

Figure 1 Cardiac multidetector-row computed tomography axial images show serial changes in the thrombus at the roofed (blind) pulmonary valve (red arrow) and the total cavo-pulmonary connection (blue arrow) of the Fontan procedure in the initial and follow-up examinations. The upper panels (A and B) clearly describe the thrombus (red arrowhead) at the roofed (blind) pulmonary valve. The lower panels (C and D) show the thrombus, which remains as a negative contrast in the iodine contrast medium (the fluid—fluid level, yellow arrowhead), implying possible circulatory stasis. SA, single atrium; SV, single ventricle. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

thromboembolic source because of the complex cardiac anatomy.

Cardiac multidetector-row computed tomography (MDCT) can provide a three-dimensional view of the cardiac anatomy with high spatial resolution. Recently, clinical application of cardiac MDCT has been expanded from adult cardiology to pediatric cardiology.

Case report

A 9-year-old boy was referred to a local hospital because of sudden onset of left hemiparesis. He was diagnosed with thromboembolic stroke by means of magnetic resonance imaging and admitted to an intensive care unit. Four days after the thromboembolic event, his neurological deficits improved, and he was transferred to our hospital. His medical history included the following: diagnosis of a single atrium and single ventricle and total anomaly of pulmonary venous drainage (TAPVD) at birth, followed by a series of complex cardiac surgeries; pulmonary artery banding (at 3 months); bidirectional Glenn procedure combined with ligation of the vertical vein and common pulmonary vein to atrium anastomosis (at 2 years); and total cavo-pulmonary connection (TCPC) of Fontan procedure (at 3 years).

For systemic workup of his complex cardiac anatomy, cardiac MDCT (Brilliance iCT, Philips Healthcare, Cleveland, OH, USA) was performed. Early images (retrospective ECG-triggering, 80 kV, 585 mAs; effective dose: 6.1 mSv) clearly revealed a supravalvular thrombus at the roofed (blind) pulmonary valve and the TCPC conduit as a contrast defect (Fig. 1A). Late images (prospective electrocardiogram-triggering, 80 kV, 180 mAs; effective dose: 1.8 mSv) also depicted the negative contrast-enhanced thrombus surrounding the iodine contrast described as the fluid—fluid level and the well-enhanced TCPC conduit without mural thrombus (Fig. 1B). Considering the possibility that the source of the thromboembolic event was not eliminated, anticoagulation therapy was continued. Six months later, a follow-up cardiac MDCT was performed, which showed that the thrombus disappeared, but the congestion of the iodine contrast, i.e. circulatory stasis, remained (Fig. 1C and D).

Discussion

Previous studies have shown that thromboembolic events after Fontan procedure are not rare and range from 3% to 16% [1—4]. Jacobs and Pourmoghadam reported that thromboembolism after Fontan procedure is attributed to various

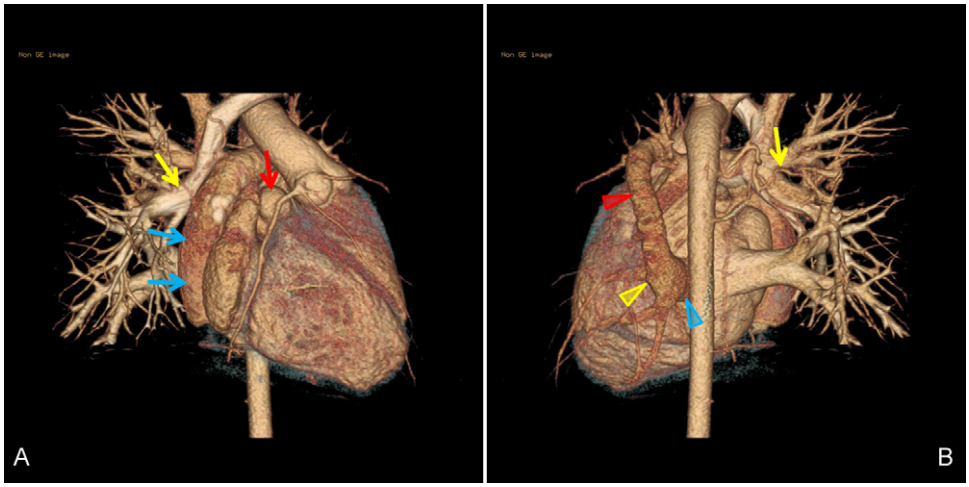

Figure 2 Cardiac multidetector-row computed tomography volume-rendering images in the anterior (A) and posterior (B) views of a child with congenital heart disease after complex cardiac surgery. Anterior view shows the roofed pulmonary valve (red arrow) after pulmonary artery banding, the connection between the superior vena cava and the right pulmonary artery (yellow arrow) by bidirectional Glenn procedure, and total cavo-pulmonary connection (blue arrow) of the Fontan procedure. Posterior view shows the vertical vein (red arrowhead) and common pulmonary (yellow arrowhead) vein to atrium anastomosis (blue arrowhead). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

conditions such as low-flow states, stasis in the venous pathways, right-to-left shunts, blind cul-de-sac, prosthetic materials, atrial arrhythmias, and hypercoagulation states [1]. Previous studies have shown an association of a history of pulmonary artery banding with an increased risk of stroke after Glenn or Fontan procedure [5,6]. Recently, surgical procedures have been chosen to prevent thromboembolism of the pulmonary artery stump origin. These procedures typically involve completely closing the pulmonary artery stump, including the pulmonary valve, or removing all leaflets followed by oversewing the pulmonary artery stump at the time of a bidirectional Glenn or Fontan procedure.

The thromboembolic source is usually evaluated by echocardiography; however, this can occasionally be challenging due to the small field of view, insufficient acoustic window, and various echocardiographic artifacts. Magnetic resonance imaging is also an alternative diagnostic imaging technique, which avoids radiation exposure, but has several disadvantages, such as low spatial resolution, long scanning duration, motion artifacts, sedation, and use of metallic devices that hamper its routine clinical use.

Clinical application of cardiac MDCT has been expanded from adult cardiology to pediatric cardiology [7,8]. In this case, the opening of the pulmonary valve could be visualized on the echocardiogram, but the presence or absence of the thrombus could not be evaluated; however, a three-dimensional view of the complex cardiac anatomy and the thrombus could be visualized on MDCT images; hemodynamic information could thus be obtained by assessing the contrast retention in the late image (Fig. 2, Movie 1). Our study also shows that the 256-slice MDCT was feasible for use in children with a high heart rate. The combination of a low-tube voltage technique and a special reconstruction algorithm to reduce the image noise would be promising for decreasing radiation exposure in children and maintaining the quality of image.

A few researchers have been considering long-term anticoagulation therapy in Fontan patients [1—5,9]. MDCT can provide a complete evaluation of the thromboembolic source and the risk of thrombus formation in symptomatic patients. Moreover, the complementary use of MDCT and echocardiography would be effective in children with congenital heart disease after surgery for long-term and/or repeated follow-up examination, based on the extent of radiation exposure and contrast medium.

In conclusion, we presented the case of a thromboembolic stroke in a child with congenital heart disease, after previous pulmonary banding and Fontan procedure. This study shows the usefulness of cardiac MDCT to evaluate thromboembolic risk in patients with complex congenital heart disease after surgery.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.jccase.2011.09.004.

References

[1] Jacobs ML, Pourmoghadam KK. Thromboembolism and the role of anticoagulation in the Fontan patient. Pediatr Cardiol 2007;28:457—64.
[2] Mahnke CB, Boyle GJ, Janosky JE, Siewers RD, Pigula FA. Anticoagulation and incidence of late cerebrovascular accidents following the Fontan procedure. Pediatr Cardiol 2005;26:56—61.
[3] Kaulitz R, Ziemer G, Rauch R, Girisch M, Bertram H, Wessel A, Hofbeck M. Prophylaxis of thromboembolic complications after the Fontan operation (total cavopulmonary anastomosis). J Thorac Cardiovasc Surg 2005;129:569—75.
[4] Monagle P, Karl TR. Thromboembolic problems after the Fontan operation. Semin Thorac Cardiovasc Surg Pediatr Card Surg 2002;5:36—47.

[5] Oski JA, Canter CE, Spray TL, Kan JS, Cameron DE, Murphy AM. Embolic stroke after ligation of the pulmonary artery in patients with functional single ventricle. Am Heart J 1996;132: 836—40.
[6] Chun DS, Schamberger MS, Flaspohler T, Turrentine MW, Brown JW, Farrell AG, Girod DA. Incidence, outcome, and risk factors for stroke after the Fontan procedure. Am J Cardiol 2004;93:117—9.
[7] Lee EY, Kritsaneepaiboon S, Zurakowski D, Arellano CM, Strauss KJ, Boiselle PM. Beyond the pulmonary arteries: alternative diagnoses in children with MDCT pulmonary angiography negative for pulmonary embolism. Am J Roentgenol 2009;193:888—94.
[8] Tsai IC, Fu YC, Lin PC, Lin MC, Jan SL. MDCT evaluation of congenital mitral-aortic intervalvular fibrosa aneurysm: implications for the aetiology and differential diagnosis. Pediatr Radiol 2009;39:80—3.
[9] Cheung YF, Chay GW, Chiu CS, Cheng LC. Long-term anticoagulation therapy and thromboembolic complications after the Fontan procedure. Int J Cardiol 2005;102:509—13.