

---

---

## CASE REPORT

---

---

# Evaluation of Congenital Mitral-aortic Intervalvular Fibrosa Aneurysm: Role of Multidetector Computed Tomography in Congenital Heart Disease

YY Kwong, JJK Ip, WWM Lam

Department of Radiology, Queen Mary Hospital, Pokfulam, Hong Kong

### ABSTRACT

*Mitral-aortic intervalvular fibrosa aneurysm is a rare disease entity whose aetiology remains uncertain. Most of the cases reported are believed to be a sequela of infective endocarditis. We present a case of congenital mitral-aortic intervalvular fibrosa aneurysm, representing the first patient to be diagnosed at birth in the literature. This report illustrates the imaging features of mitral-aortic intervalvular fibrosa aneurysm and the role of multidetector computed tomography in the diagnostic process.*

*Key Words:* Aortic aneurysm; Aortic valve; Mitral valve; Tomography, X-ray computed

## 中文摘要

### 先天性二尖瓣－主動脈瓣瓣間纖維膜動脈瘤的評估：多層螺旋CT掃描在診斷先天性心臟病中的作用

鄭彥頤、葉精勤、林慧文

二尖瓣－主動脈瓣瓣間纖維膜動脈瘤是一種罕見疾病，病因至今仍然不明。大多數病例認為是感染性心內膜炎的後遺症。本文報告了一例二尖瓣－主動脈瓣瓣間纖維膜動脈瘤的病例，病人出生時已即診斷此症，是文獻中首個類似病例報告。本報告描述此病例的影像學特徵，以及診斷此症時多層螺旋CT掃描的作用。

### INTRODUCTION

Mitral-aortic intervalvular fibrosa aneurysm (MAIFA) is a rare disease entity whose aetiology remains uncertain. Most of the cases reported are believed to be a sequela of infective endocarditis. We present a case of congenital MAIFA, representing the first patient in the literature to be diagnosed at birth. The imaging features

and differential diagnoses of MAIFA are explored.

### CASE REPORT

A baby girl was born at term by normal spontaneous delivery in January 2011. Fetal echocardiography done at 17 weeks showed pleural effusion, which spontaneously subsided on serial echocardiography.

---

*Correspondence:* Dr Yin-yee Kwong, Department of Radiology, Queen Mary Hospital, Pokfulam, Hong Kong.  
Tel: (852) 6170 6191; Email: char.kwong@gmail.com

Submitted: 10 Mar 2015; Accepted: 28 May 2015.

Disclosure of Conflicts of Interest: All authors have disclosed no conflicts of interest.

Repeated fetal echocardiography at 36 weeks revealed a cystic shadow in the left atrium (LA), which was thought to be an enlarged coronary sinus, and a large right side of the heart.

At birth, she had Apgar scores of 9 and 10 at 1 and 5 minutes, respectively. She was asymptomatic at birth, with good oxygen saturation, and tolerated feeding well. Owing to her antenatal history, she was referred to the paediatric cardiologist for assessment.

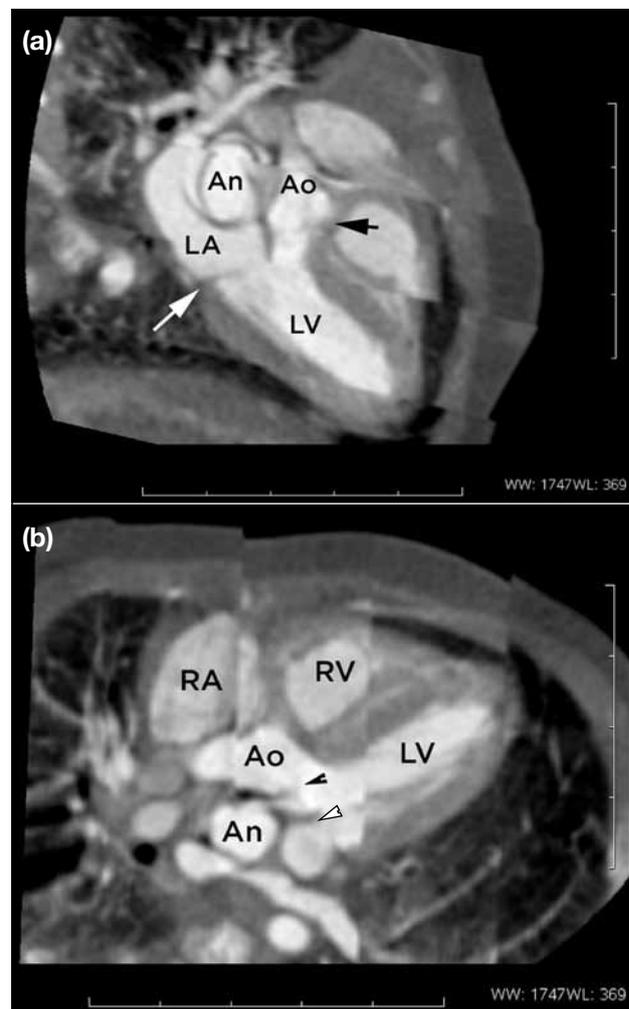
Echocardiogram performed on day 4 of life showed an 8.3 x 7.4 mm aneurysm-like structure in the LA, close to the posterior aspect of the ascending aorta with bi-directional arterial flow. A direct entry point into this structure was difficult to delineate because of the small size of the patient, with limited spatial resolution. A secundum atrial septal defect (ASD) and a small patent ductus arteriosus (PDA) were also noted. No coarctation of the aorta (CoA) was seen. The patient subsequently underwent cardiac catheterisation, confirming a left ventricle (LV) to LA aneurysm.

The patient was referred to the Department of Radiology for multidetector (MD) computed tomography (CT) evaluation to confirm the diagnosis and to better delineate the cardiac anatomy. MDCT was done at the age of 3 weeks in an inpatient setting. After a 24-gauge intravenous cannula was inserted into the right arm, midazolam (Dormicum; Roche, Vienna, Austria) 0.1 ml/kg was injected for sedation.

No oral or intravenous beta-blocker was given. The scan was done with a 64-detector row CT scanner (LightSpeed VCT; General Electric, Fairfield [CT], USA) with the following protocol: 64 x 0.625 mm collimation, 100 kVp tube voltage, 199 mA tube current, rotation time of 0.35 seconds, and pitch of 0.24 with retrospective electrocardiogram gating. Contrast material (Iopamiro 370; Bracco Sine Pharmaceutical Corp Ltd, Shanghai, China) 10 ml was injected followed by 10 ml of saline flush. The flow rate was 1.8 ml/second. The scan was performed from the carina to the base of the heart. Bolus tracking was used, with the region of interest in the LV cavity. After enhancement had reached 150 HU, a 5-second post-threshold delay was applied before the scan. The average heart rate during the scan was 141 beats/min (range, 138-147 beats/min). Four equally spaced phases of images were reconstructed from 40% to 80% of the RR intervals with a slice thickness of 0.625 mm. A senior paediatric

radiologist interpreted the scan on a dedicated MDCT workstation with three-dimensional reformation.

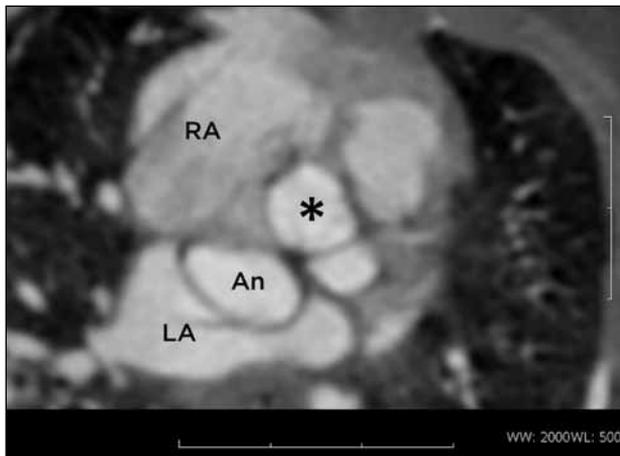
Motion artefacts and step artefacts were present due to fast heart rate and inadequate sedation. Regardless, MDCT demonstrated a 1.3 x 1.1 cm aneurysm at the mitral valve-aortic valve junction, communicating with the LV and indenting into the LA (Figure 1). The aortic valve was intact (Figure 1a) and there was no



**Figure 1.** Multiplanar multidetector computed tomography images of a mitral-aortic intervalvular fibrosa aneurysm in a neonate. (a) Three-chamber view shows an aneurysm at the mitral-aortic intervalvular fibrosa, which is the junction between mitral valve (white arrow) and aortic valve (black arrow). The aneurysm indents into the left atrium. The aortic valve is intact, which allows exclusion of a diagnosis of sinus of Valsalva aneurysm. Step artefacts degrade the image quality. (b) Two-chamber view demonstrates the neck of the aneurysm arising from the mitral-aortic intervalvular fibrosa between the mitral valve (white arrowhead) and aortic valve (black arrowhead). Abbreviations: An = aneurysm; Ao = aorta; LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

dilatation of the sinus of Valsalva (Figure 2), so the diagnosis of sinus of Valsalva aneurysm was excluded. The interventricular septum was intact so the diagnosis of septal aneurysm was excluded (Figure 3). The ASD previously noted on echocardiogram was again noted (Figure 3). Furthermore, focal narrowing was observed at the distal aortic arch (narrowest 3.3 mm) [Figure 4], thus an additional diagnosis of CoA was made.

Since the MAIFA was present at birth, before any intervention, acquired causes such as infection or iatrogenic cause could be confidently excluded.



**Figure 2.** Multiplanar multidetector computed tomography image of the sinus of Valsalva (asterisk) shows that it is of normal size. Abbreviations: An = aneurysm; LA = left atrium; RA = right atrium.

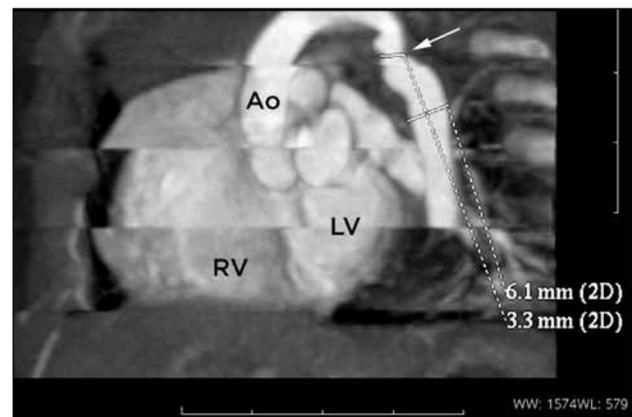


**Figure 3.** Multiplanar multidetector computed tomography in four-chamber view clearly shows the atrial septal defect (black arrow). The interventricular septum is intact, which excludes a diagnosis of perimembranous septal aneurysm. Abbreviations: LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

The patient underwent surgical repair for CoA and ligation of PDA. The paediatric cardiologist decided to observe the MAIFA, which showed no significant enlargement on serial echocardiography (Figures 5 and 6). At the time of writing this report, the child was 4 years old, and remained well and asymptomatic to date.

## DISCUSSION

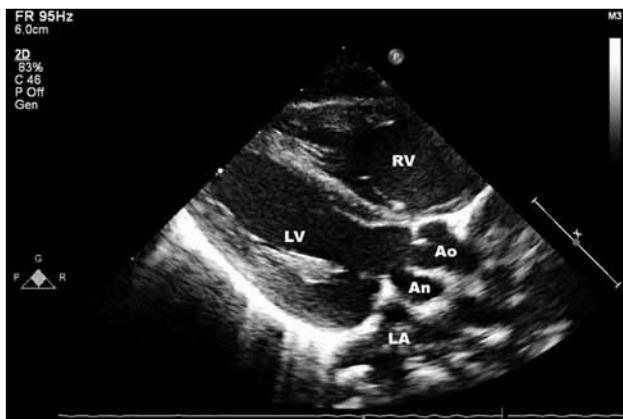
The mitral-aortic intervalvular fibrosa is the junctional tissue between the elements of the aortic and mitral valves.<sup>1</sup> Perforation or rupture of this area is usually secondary to blunt chest trauma, a complication of cardiac surgery, or as a result of infection.<sup>1,2</sup>



**Figure 4.** Multiplanar multidetector computed tomography maximum intensity projection image of the aorta. Focal stenosis at the distal aortic arch is indicated by the white arrow. Abbreviations: Ao = aorta; LV = left ventricle; RV = right ventricle.



**Figure 5.** Echocardiogram short-axis view at the aortic valve level shows an aneurysm posterior to the aortic root at the level of the aortic valve (white arrow). Abbreviation: An = aneurysm.



**Figure 6.** Echocardiogram parasternal long-axis view shows an aneurysm posterior to the aortic root and anterior to the left atrium.

Abbreviations: An = aneurysm; Ao = aorta; LA = left atrium; LV = left ventricle; RV = right ventricle.

Congenital MAIFA is a rare entity and only a few case reports are available in the literature. Therefore, its congenital aetiology has long been a subject of debate.

The youngest patient reported in the literature without a history of infection was 6 months old.<sup>3</sup> This patient showed a LA cystic shadow as early as 36 weeks of gestation on fetal echocardiography and was diagnosed with echocardiography and MDCT soon after birth, before any surgery or infective episode, which further supports the hypothesis that the condition could be congenital in origin. This information is also helpful to understand the timing of development of this disease.

The natural history and outcome of congenital MAIFA are not well understood. Previous reports suggested that these lesions could be asymptomatic and carry a better prognosis than their pseudo-aneurysm counterparts.<sup>3,4</sup> The clinical course of this patient is in line with this hypothesis. The patient did not have an operation on the aneurysm and has been well at follow-up (aged 4 years at the time of writing this report). Serial echocardiography did not show significant enlargement of the aneurysm or presence of thrombus.

In this patient, MDCT was shown to be extremely helpful in the management of congenital heart disease. The major advantage of MDCT is its excellent spatial resolution, which is beneficial in evaluating tiny intracardiac structures.<sup>5</sup> In this patient, the aneurysm opening could not be identified on echocardiography, even

by an experienced paediatric cardiologist, due to its limited spatial resolution. Moreover, congenital MAIFA may mimic other intracardiac anomalies, for example, sinus of Valsalva aneurysm or interventricular septal aneurysm.<sup>3</sup> MDCT was able to demonstrate the intact aortic valve and the intact interventricular septum to exclude these diagnoses.

This patient also illustrates the usefulness of MDCT to detect extracardiac anomalies in complex congenital heart disease. Previous studies have shown that MDCT can detect additional extracardiac anomalies in up to 29% of patients, when compared to echocardiography.<sup>5</sup> In this patient, CoA could not be detected on echocardiography or cardiac catheterisation. MDCT could therefore be a ‘one-stop shop’ for preoperative planning.

A major disadvantage of MDCT, especially in children, is the exposure to ionising radiation. Children are at much greater risk than adults from a given dose of radiation, both because they are inherently more radiosensitive and because they have more remaining years of life during which a radiation-induced cancer could develop.<sup>6</sup> To minimise these risks to ‘as low as reasonably achievable’ levels, it is recommended that MDCT be used only when other modalities such as echocardiography or cardiac magnetic resonance (CMR) is inconclusive, unavailable, or contraindicated.<sup>6</sup> However, in clinical practice, neonates often need general anaesthesia for CMR and the extended scan time could be a challenge for unstable neonates with cardiopulmonary anomalies. At Queen Mary Hospital, MDCT remains the first-line investigation for neonates when echocardiography is inconclusive.

Another challenge for MDCT when imaging neonates with congenital heart disease is the fast heart rate. In this patient, her heart rate was up to 140 beats/min, thus considerable motion artefacts and step artefacts were present. Newer 320-detector row CT scanners are able to cover the scan range of the entire heart within a single heartbeat, and may minimise these artefacts.

In conclusion, this report presents the first patient to be diagnosed with congenital MAIFA at birth, while the congenital aetiology has long been challenged. MDCT is a valuable tool for demonstrating the intracardiac anatomy and comprehensively assessing extracardiac anomalies.

## REFERENCES

1. Bansal RC, Graham BM, Jutzy KR, Shakudo M, Shah PM. Left ventricular outflow tract to left atrial communication secondary to rupture of mitral-aortic intervalvular fibrosa in infective endocarditis: diagnosis by transesophageal echocardiography and color flow imaging. *J Am Coll Cardiol.* 1990;15:499-504. [cross ref](#)
2. Tak T. Pseudoaneurysm of mitral-aortic intervalvular fibrosa. *Clin Med Res.* 2003;1:49-52. [cross ref](#)
3. 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. [cross ref](#)
4. He D, Sinha P, Olivieri L, Jonas RA. Congenital aneurysm of the aortomitral intervalvular fibrosa. *Ann Thorac Surg.* 2015;99:314-6. [cross ref](#)
5. Lee T, Tsai IC, Fu YC, Jan SL, Wang CC, Chang Y, 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;36:1273-82. [cross ref](#)
6. Prakash A, Powell AJ, Geva T. Multimodality noninvasive imaging for assessment of congenital heart disease. *Circ Cardiovasc Imaging.* 2010;3:112-25. [cross ref](#)