*Cardiology in the Young*

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# Successful device closure by Amplatzer vascular plug II of a rare celiacomesenteric trunk-to-right atrium fistula in a repaired giant omphalocele

Yu-Hsuan Chen[1], Kun-Shan Cheng[2] and Wei-Li Hung[1]

[1]Department of Pediatric Cardiology, MacKay Children's Hospital, Taipei, Taiwan and [2]Department of Pediatric Cardiology, MacKay Memorial Hospital, Hsinchu, Taiwan

## Brief Report

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**Author for correspondence:**
Y.-H. Chen, Department of Pediatric Cardiology, MacKay Children's Hospital, No. 92, Sec. 2, Zhongshan N. Rd., Taipei City 10449, Taiwan.
Tel: (886)-2-2543-3535; Fax: (886)-2-2543-3642.
E-mail: j724603@gmail.com

**Abstract**

Aorto-right-atrial fistula is an uncommon condition with an unclear pathogenesis. We present the case of a 3-year-old girl with a giant omphalocele repaired days after birth and incidentally discovered with a celiacomesenteric trunk-to-right atrium fistula. Three-dimensional reconstruction CT unveiled its anatomical pattern, and the fistula was successfully closed using a Amplatzer vascular plug II percutaneously.

A 3-year-old girl who underwent giant omphalocele repair (an omphalocele containing most of the liver) days after birth was referred to our hospital because of a grade 3/6 continuous murmur with epigastric thrills incidentally discovered during a common cold visit. Plain chest radiography showed cardiomegaly with a cardiothoracic ratio of 61.6% and increased pulmonary vascularity.

**Figure 1.** CT with three-dimensional reconstruction of the aorto-right-atrial fistula (white arrow).

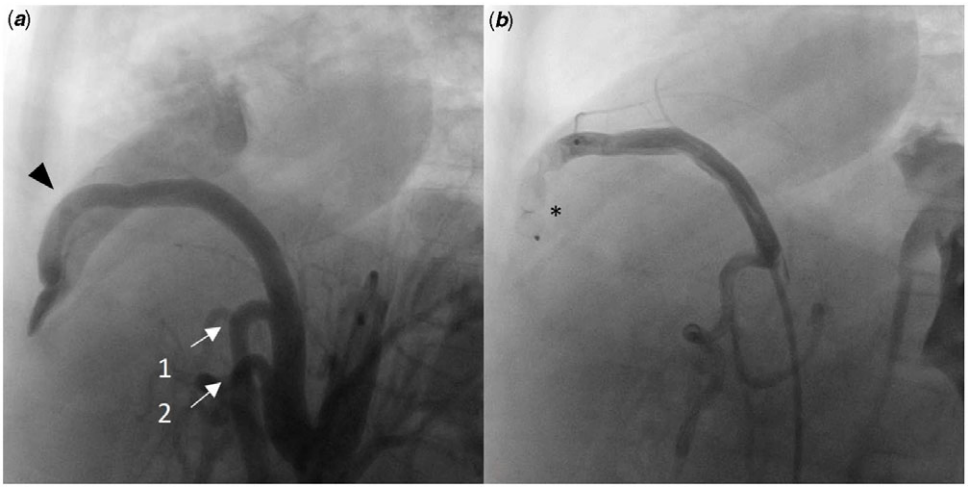


**Figure 2.** Lateral view of abdominal aorta angiograms showing the course of the aorto-right-atrial fistula (black arrowhead), celiac trunk (white arrow 1), and superior mesenteric trunk (white arrow 2) (***a***). After deployment of AMPLATZER™ VASCULAR PLUG II (*), which showed total occlusion of the aorto-right-atrial shunt without a residual shunt (***b***).

Electrocardiography revealed right atrial enlargement and incomplete right bundle branch block. Echocardiography identified a 5-mm fistula originating from the common celiacomesenteric trunk and draining into a pouch just below the right atrium. CT later confirmed that the pouch drained into right atrium. Three-dimensional reconstruction unveiled the complete course of fistula (Fig 1). After discussion with the parents and obtaining their consent, we performed the fistula closure via catheterisation.

During the cardiac catheterisation examination, right and left heart catheterisation were performed. The pulmonary-to-systemic flow ratio was 1.5. An aortogram showed a tubular-shaped fistula (Fig 2a) coming from the common celiacomesenteric trunk and draining into the pouch below the right atrium at a wide angle. No other fistulas were observed in the liver. In addition, the inferior vena cava angiogram showed an unusual vein route that drained anterior to the liver and joined the same pouch below the right atrium. Such inferior vena cava anomalies have been previously reported in giant omphaloceles and were classified as type II.[1]

The fistula diameter was 5.18 mm at the origin, 4.41 mm at its mid-point, and with stenosis, 2.03 mm at the exit. Considering the inadequate length to deploy the device in the narrowest part of the fistula and the possibility of the device to protrude into the pouch, we decided to deploy the device near the exit but closer to the proximal part of the fistula. The diameter was approximately 5 mm at the deployment location. Hence, we chose an 8-mm AMPLATZER™ VASCULAR PLUG II to smoothly occlude the fistula. A fistula angiogram was repeated, revealing the position of the device (Fig 2b). The patient was transferred to the general ward after the procedure and was discharged on the following day without complications. The echocardiography performed 3 months later at follow-up after the procedure showed a turbulent flow originating from the common celiacomesenteric trunk, and the device was seen in the distal end. The patient remained well during the follow-up clinic visits over the next 6 months.

Aorto-right-atrial fistula is an uncommon condition with an unclear pathogenesis. Most cases occur as complications of surgical procedures or result from bacterial endocarditis, paravalvular abscess, ruptured sinus of Valsalva, and aortic dissection. Congenital cases also exist but are extremely rare. Our patient had an omphalocele and underwent primary closure surgery a few days after birth, leaving the aetiology unclear. However, complications of the surgery could not be ruled out, because the echocardiography performed before the omphalocele was repaired and showed no structural heart abnormalities.

The exact incidence of aorto-right-atrial fistulas is unknown; a systemic review reported that 63.2% originate in the ascending aorta and extend to the right atrium.[2] Others arise from the aortic root, and a few originate in the thoracic aorta. Only one such case has been reported by Alkan et al.; in 2017 in Turkey,[3] an abdominal aorta-to-right atrium fistula was found. The patient had a vessel root similar to that in our case, which was successfully closed using AMPLATZER™ VASCULAR PLUG IV via catheterisation.

Patients with aorto-right-atrial fistulas may be asymptomatic; however, once diagnosed, closure is indicated to prevent potential complications such as congestive heart failure or pulmonary hypertension. Surgical or percutaneous closure is a therapeutic option. AMPLATZER™ VASCULAR PLUG II is widely used in arteriovenous fistulas with good efficacy and minor safety concerns.

**Supplementary material.** To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951122003456

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**Conflicts of interest.** None.

**Ethical standards.** This study complies with the Declaration of Helsinki and was approved by the relevant ethics committee.

## References

1. Gorenstein A, Meyer S, Schiller M. Inferior vena cava anomalies associated with giant omphalocele–a proposed classification. Z Kinderchir 1983; 38: 380–382.
2. Jainandunsing JS, Linnemann R, Bouma W, et al. Aorto-atrial fistula formation and closure: a systematic review. J Thorac Dis 2019; 11: 1031–1046.
3. Alkan F, Pabuscu Y, Cetin M, Coskun S. Successful treatment of abdominal aorto-right atrial fistula by vascular plug: a previously unreported cardiac malformation. Anatol J Cardiol 2017; 18: 433–435.