Clinical Journal of Gastroenterology (2019) 12:20–24
https://doi.org/10.1007/s12328-018-0895-8

CASE REPORT

Successful treatment of esophageal bleeding due to rupture of major aortopulmonary collateral arteries by transcatheter arterial embolization

Takanori Ito[1] · Masatoshi Ishigami[1] · Yoji Ishizu[1] · Teiji Kuzuya[1] · Takashi Honda[1] · Masaya Matsushima[2] · Taichi Kato[3] · Yoshiki Hirooka[1]

Received: 30 July 2018 / Accepted: 16 August 2018 / Published online: 21 August 2018

Abstract

Major aortopulmonary collateral arteries (MAPCAs) are unique vessels associated with hypoxia induced by congenital heart disease (CHD). Although MAPCAs are essential to supply blood to the lungs, their development and proliferation can induce life-threatening complications, such as rupture into the lung. Here, we describe a rare case of esophageal bleeding from MAPCAs in a CHD patient, which was successfully treated by transcatheter arterial embolization (TAE). A 16-year-old male with CHD experienced a hematemesis and melena after the Bentall procedure to treat valvular heart disease. Emergent esophagogastroduodenoscopy revealed spurting bleeding from the middle esophageal vessels; accordingly, endoscopic variceal ligation (EVL) was performed. However, he had a hematemesis again after 2 weeks of EVL. The arterial phase of dynamic computed tomography indicated that a MAPCA associated with CHD was the origin of bleeding. Hence, TAE of this MAPCA with a mixture of *n*-butyl-2-cyanoacrylate and ethiodized oil was performed to prevent re-bleeding. Color Doppler mode in endoscopic ultrasonography via the esophagus revealed mosaic-like signals in MAPCAs located in the esophageal wall. This finding was consistent with tortuous MAPCAs accompanied by turbulent blood flow. When clinicians encounter CHD patients with unexpected massive esophageal bleeding, bleeding related to MAPCAs should be considered.

Keywords Major aortopulmonary collateral artery · Congenital heart disease · Esophageal bleeding · Transcatheter arterial embolization

Abbreviations

MAPCA	Major aortopulmonary collateral artery
CHD	Congenital heart disease
TAE	Transcatheter arterial embolization
EVL	Endoscopic variceal ligation
CT	Computed tomography
NBCA	*n*-Butyl-2-cyanoacrylate
EGD	Esophagogastroduodenoscopy
EUS	Endoscopic ultrasonography
GI	Gastrointestinal
CA	Celiac artery
SMA	Superior mesenteric artery

✉ Masatoshi Ishigami
masaishi@med.nagoya-u.ac.jp

[1] Department of Gastroenterology and Hepatology, Nagoya University Graduate School of Medicine, 65 Tsurumai-cho, Showa-ku, Nagoya 466-8550, Japan

[2] Department of Radiology, Nagoya University Graduate School of Medicine, Nagoya, Japan

[3] Department of Pediatrics, Nagoya University Graduate School of Medicine, Nagoya, Japan

Background

In congenital heart disease (CHD) patients with cyanosis, unique arteries called major aortopulmonary collateral arteries (MAPCAs) develop to supply blood to the lungs [1–4]. MAPCAs are large systemic collateral arteries usually arising from the descending aorta, branch of the aortic arch, and ascending aorta, and less commonly from other systemic arteries such as the abdominal, carotid, and coronary arteries [2, 5]. In rare cases, expanded and proliferated MAPCAs rupture into the respiratory tract or mediastinum [6]. These cases require an interventional radiology procedure, transcatheter arterial embolization (TAE) to prevent or stop hemorrhage. To date, however, there have been no reports that MAPCAs can induce bleeding into the digestive

tract. Here, we describe a case in which TAE was used to successfully treat MAPCA rupture into the esophagus in a CHD patient with heterotaxy, asplenia syndrome, and single ventricle.

Case report

A 16-year-old male with heterotaxy, asplenia syndrome, functional single ventricle, pulmonary stenosis, total anomalous pulmonary venous return, and MAPCAs underwent the Bentall procedure and tricuspid valve replacement to improve cardiac function, which had deteriorated due to aortic and tricuspid regurgitation. Due to the paucity of central pulmonary arteries, the patient was not considered a candidate for the Glenn or Fontan procedures, and no surgical interventions had been performed.

One week after this operation, the patient experienced hematemesis and melena, and soon developed hypovolemic shock. Esophagogastroduodenoscopy (EGD) revealed active bleeding from a vessel at the middle of the esophagus. Accordingly, the patient immediately underwent endoscopic clipping. However, the bleeding was uncontrollable. The patient was intubated and a Sengstaken–Blakemore tube was inserted, followed by transfer to our hospital for further treatment with the transfusion of blood and albumin.

On arrival, his vital signs were follows: blood pressure, 62/30 mmHg; heart rate, 120 beats/min; body temperature, 37 °C; and oxygen saturation in room air, 90% with a ventilator. A physical examination revealed pale palpebral conjunctiva, and the abdomen was soft and flat. Laboratory data on arrival were as follows: white blood cell count, 11,600/μL; hemoglobin, 6.8 g/dL; hematocrit, 20.5%; platelet count, 105×10^3/μL; PT-INR, 1.58 (under warfarinization); fibrinogen, 100 mg/dL; D-dimer, 5.58 μg/mL; antithrombin-·, 38.9%; fibrin/fibrinogen degradation products, 11.9 μg/mL; and brain natriuretic peptide, 1331 pg/mL (Table 1). He underwent emergent EGD and rapid blood transfusion simultaneously in the ICU. EGD revealed spurting bleeding from the proximal side of a vessel with a hemoclip that had been placed at the previous hospital (Fig. 1a). We suspected rupture of esophageal varix, and performed endoscopic variceal ligation (EVL). After one ligation of the bleeding point, hemostasis was achieved. EGD after 1 week revealed tortuous esophageal vessels covering about 70% of the lumen, and the clip remained at the distal side of the post-EVL ulcer (Fig. 1b). Because these vessels were not large and had no red signs, we considered the risk of re-bleeding without additional treatment to be low, and food intake was resumed. However, the patient experienced hematemesis and melena again after 2 weeks of EVL. Emergent EGD revealed spurting bleeding from the same esophageal vessel at the distal side of the post-EVL ulcer, and bleeding was again stopped by EVL. Dynamic computed tomography (CT) with contrast medium was performed to assess the hemodynamics of esophageal varices and the presence of portal thrombus. However, there were no findings of liver cirrhosis, congestive liver, or portal hypertension such as development of esophageal or gastric varices. On the other hand, CT revealed multiple vessels that were enhanced in the early arterial phase around and within the esophageal wall. These vessels were branched from the descending aorta, celiac artery (CA), and superior mesenteric artery (SMA) and flowed into the pulmonary artery (Fig. 2a). Furthermore, the endoscopic clip was attached to one of these tortuous vessels in the middle esophagus (Fig. 2b). Based on these findings, we determined that the esophageal bleeding was caused by rupture of MAPCAs into the esophagus. We believed that MAPCA rupture would not be controllable by EVL alone, and posed the risk of massive, life-threatening bleeding in the event that these vessels ruptured again. Hence, we attempted to perform angiography and TAE of MAPCAs.

Table 1 Laboratory data on arrival

AST	37 U/L	WBC	11,600/μL
ALT	30 U/L	RBC	2.14×10^6/μL
LDH	258 U/L	Hemoglobin	6.8 g/dL
ALP	96 U/L	Hematocrit	20.5%
γ-GTP	13 U/L	Platelet count	105×10^3/μL
Total bilirubin	0.7 mg/dL	PT-INR	1.58
Total protein	6.0 g/dL	APTT	62.6 s
Albumin	4.9 g/dL	Fibrinogen	100 mg/dL
UN	21.6 mg/dL	D-dimer	5.58 μg/mL
Creatinine	0.36 mg/dL	AT-·	38.9%
BNP	1331 pg/mL	FDP	11.9 μg/mL

AST aspartate aminotransferase, *ALT* alanine aminotransferase, *LDH* lactate dehydrogenase, *ALP* alkaline phosphatase, ·-*GTP* ·-glutamyl transpeptidase, *T.bil* total bilirubin, *UN* urea nitrogen, *BNP* brain natriuretic peptide, *WBC* white blood cell count, *RBC* red blood cell count, *PT-INR* prothrombin time-international normalized ratio, *APTT* activated partial thromboplastin time, *sec* second, *AT-·* antithrombin-·, *FDP* fibrin/fibrinogen degradation products

On angiogram, we observed several MAPCAs branched from the aorta, CA, and SMA feeding the lung, one of which arose from the CA and ran close to the hemoclip. A microcatheter was advanced near the hemoclip, and embolization was performed with a 2:3 mixture of *n*-butyl-2-cyanoacrylate (NBCA) and ethiodized oil (Lipiodol) after confirming rupture of the MAPCA with the clip by digital subtraction angiography and enhanced CT during angiography (Fig. 2c–e). CT after embolization revealed that Lipiodol was deposited at the artery around clip, and the patient experienced no abdominal pain or oxygen desaturation. Oral

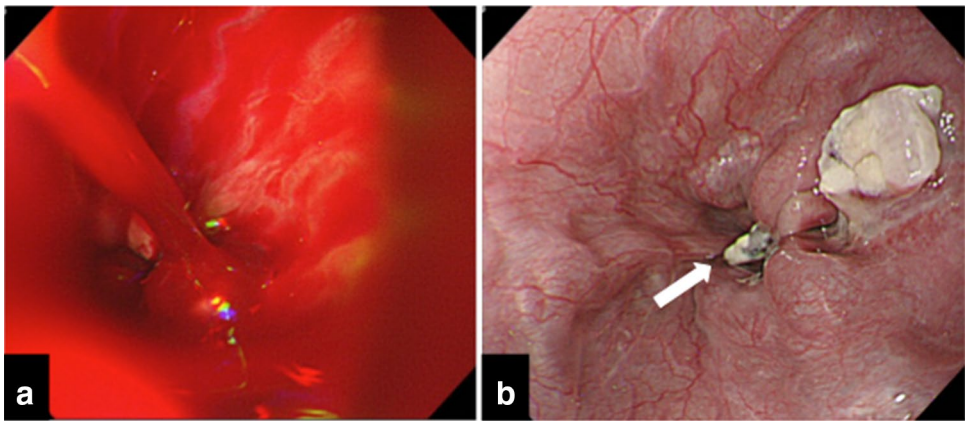

Fig. 1 **a** Emergent esophagogastroduodenoscopy (EGD) showed spurting bleeding of the middle esophageal vessels. **b** EGD showing tortuous esophageal vessels after EVL and the hemoclip attached at the distal side of the post-EVL ulcer (white arrow)

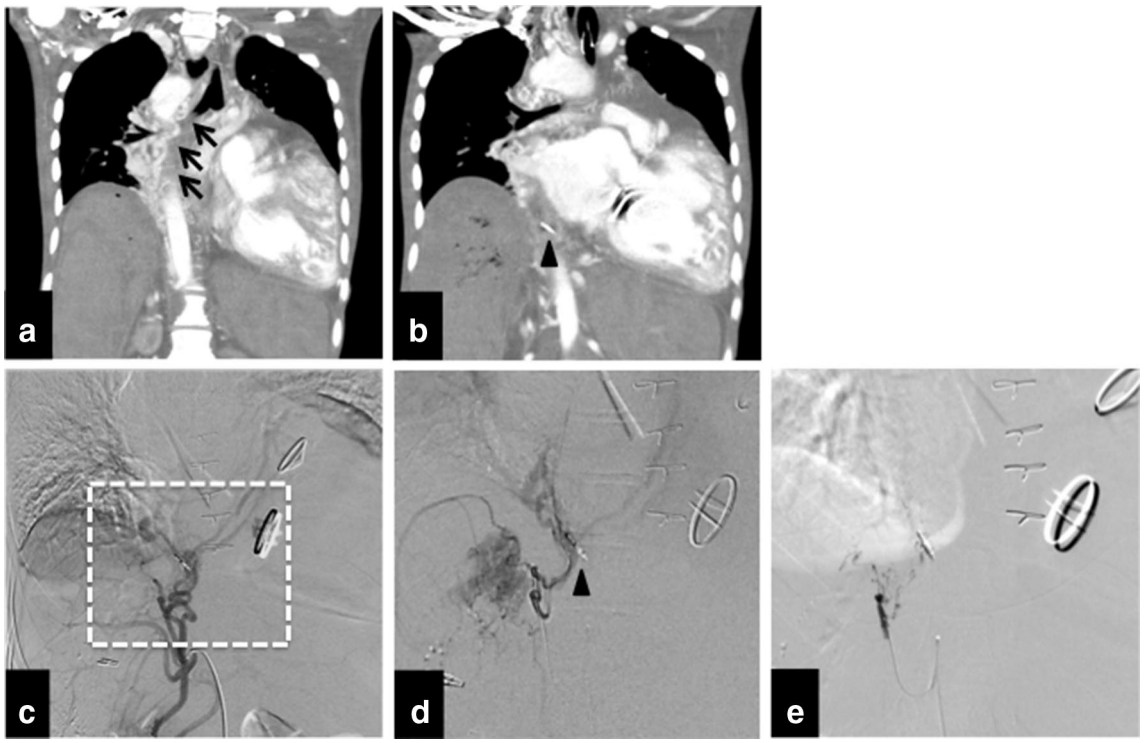

Fig. 2 **a** Dynamic computed tomography (CT) image with contrast medium revealed many vessels contrasted in the early phase around the esophagus (coronal plane). These vessels branched from the descending aorta, celiac artery, and superior mesenteric artery and flowed into the lung (arrow). **b** The hemoclip was attached to one of these tortuous vessels in the middle esophagus (arrow head). **c, d** Angiography of the branch of celiac artery revealed the culprit vessel with the clip (arrow head). **e** Transcatheter arterial embolization (TAE) was performed using a 2:3 mixture of *n*-butyl-2-cyanoacrylate (NBCA) and ethiodized oil

intake was resumed, the patient was transferred to the previous hospital without hematemesis or melena.

Ten months after discharge, a follow-up EGD revealed that the ruptured site had scarred completely, and the size of vessel had decreased (Fig. 3a). CT without contrast medium revealed a few Lipiodol remnants in the same vessel (Fig. 3b). At the same time, we performed endoscopic ultrasonography (EUS) to confirm that these vessels were MAPCAs. EUS via esophagus using a GF-UE260-AL5 (Olympus, Tokyo, Japan) with balloon and dual scanning frequencies of 7.5 and 12 MHz revealed many vessels with low echoic area under and inside the esophageal wall, some of which were near the surface of the esophageal wall. Color Doppler mode in EUS revealed mosaic-like signals in these vessels (Fig. 3c, d). This finding indicated rapid blood flow with turbulence, accompanied by tortuous MAPCAs. No further bleeding was noted over 20 months of follow-up after the TAE procedure.

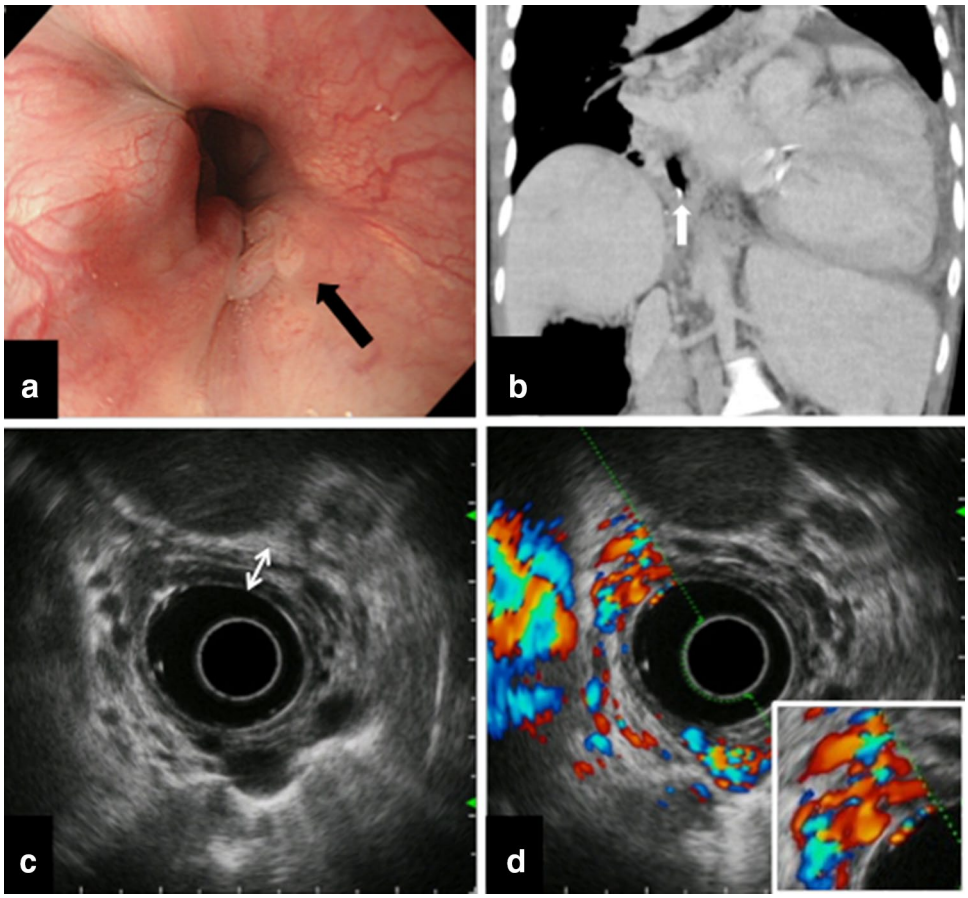

Fig. 3 Images at 10 months after TAE. **a** EGD revealed that the ruptured site had healed completely, and the size of vessel had decreased relative to its state prior to TAE. **b** Coronal image of CT without contrast medium revealed a few deposits of ethiodized oil at the vessel in the middle esophagus. **c** Endoscopic ultrasound (EUS) revealed many vessels with low echoic area under and within the esophageal wall (two-way arrow, esophageal wall). **d** Vessels exhibiting mosaic signals in Color Doppler mode of EUS

Discussion

Herein, we report a rare case of MAPCA rupture into the esophagus in a patient with asplenia, heterotaxia, and single ventricle. In rare cases, MAPCAs induce pulmonary bleeding by rupture into the lung [6]. This is the first report of MAPCA rupture into the esophagus in a patient with CHD. We successfully treated refractory bleeding caused by MAPCA rupture by TAE, which was not accompanied by adverse events.

MAPCAs are unique collateral arteries associated with hypoxia induced by CHD, e.g., pulmonary atresia with ventricular septal defect, asplenia syndrome, or single ventricle. MAPCAs are congenital and systemic-to-pulmonary collateral arteries, representing remnants of the embryonic ventral splanchnic arteries. They normally regress concomitant with the formation of normal pulmonary arteries during gestation. However, in cases of early maldevelopment of the pulmonary valve or the central pulmonary arterial system, MAPCAs can persist to supply blood to the pulmonary arteries [3, 7]. MAPCAs run near the esophagus on the way to the pulmonary arteries. Richard et al. reported that 67% of the patients undergoing primary surgical procedures for pulmonary atresia/ventricular septal defect/MAPCAs had retroesophageal MAPCAs, and that 46% of retroesophageal MAPCAs coursed within the muscular fibers of the esophagus (intraesophageal) [8], as was the case in our patient. Although MAPCAs that penetrate the esophageal wall have the potential to induce esophageal bleeding, to date there have been no reports of MAPCA rupture into the esophagus.

Meanwhile, it was very difficult to distinguish abnormal collateral arteries with developed para-esophageal vessels. In this case, EUS demonstrated that some vessels existed near the surface of the esophageal wall. MAPCAs might contain parts of developed para-esophageal vessels, therefore, it was impossible to identify whether the bleeding was due to the rupture of perforator from developed para-esophageal vessels communicated with MAPCAs or direct rupture of MAPCAs.

In this case, at first we tried to stop bleeding by EVL alone, but the patient experienced vomiting bleeding again; therefore, we performed TAE with NBCA and Lipiodol. Bleeding of common esophageal varices induced by portal hypertension can be treated by EVL. However, the blood flow in a MAPCA, which is an abnormal artery rather than a vein, is more rapid than that in common

varices; consequently, it is difficult to control MAPCA-induced bleeding by EVL alone. In pre-postoperative management of CHD patients, TAE for MAPCAs is often performed to unifocalize the blood flow toward the lungs [9]. However, almost no previous reports described the use of TAE to treat MAPCAs for preventing bleeding. Although MAPCA rupture is very rare, it can lead to severe hemorrhage and sudden death [5, 10]. Sharma et al. reported a case in which a ruptured aneurysm of a large MAPCA in a patient with CHD was treated by TAE with endovascular deployment of a vascular plug [6]. In our case, the previous doctor attached an endoscopic hemoclip to the culprit vessel. Therefore, we could identify the culprit artery from among several MAPCAs by confirming the clip during angiography.

We selected NBCA, rather than coil, as the embolus material for TAE under selective cannulation of the microcatheter. NBCA is a low-viscosity liquid adhesive mainly used in the field of intravascular radiology for the treatment of arteriovenous fistulas/malformations and pseudo-aneurysm of the brain [11, 12]. In addition, NBCA is considered to be useful as a secondary embolization material in TAE conducted emergently to control massive gastrointestinal (GI) bleeding [13]. In particular, NBCA is suitable for tortuous or narrowed vessels in which it is difficult to deeply insert a microcatheter, or in cases with coagulopathy [14]. Because the culprit vessel of this case was extremely tortuous, we anticipated that it would be difficult to perform TAE with coils under deep cannulation of the microcatheter. Additionally, this case had a number of collateral vessels near the bleeding point. Coiling is likely to induce the embolization of only proximal vessels, and blood flow from the collateral vessels to the bleeding point can occur. Hence, NBCA was the better choice for embolizing the entire culprit vessel in this case. Ultimately, TAE with NBCA was performed successfully and safely without complications such as severe pain, cerebral infarction, pulmonary embolism, or GI ischemia and stenosis.

In conclusion, we encountered a patient with severe esophageal bleeding induced by a CHD-associated MAPCA, and treated him successfully with TAE. Recent reports have shown that the medical and surgical treatment of CHD has advanced remarkably, and over 90% of CHD patients survive until adulthood [15, 16]. Therefore, we may have the opportunity to treat MAPCA-induced hemorrhage in CHD patients, as in this case. When we encounter unexpected severe esophageal bleeding resembling variceal rupture in patients with CHD, we should consider the possibility of rupture from a MAPCA into the esophagus, and add dynamic CT examination to obtain detailed information. For treatment of MAPCA bleeding, TAE with NBCA and Lipiodol may contribute to hemostasis under the marking of clip.

Compliance with ethical standards

Conflict of interest The authors who have taken part in this study declared that they do not have anything to disclose regarding funding or conflict of interest with respect to this manuscript.

Human rights All procedures were performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

Informed consent Informed consent for being included in this report was obtained from family of this patient.

References

1. Boshoff D, Gewillig M. A review of the options for treatment of major aortopulmonary collateral arteries in the setting of tetralogy of Fallot with pulmonary atresia. Cardiol Young. 2006;16:212–20.
2. Tchervenkov CI, Roy N. Congenital heart surgery nomenclature and database project: pulmonary atresia—ventricular septal defect. Ann Thorac Surg. 2000;69:97–105.
3. Reinhartz O, Reddy VM, Petrossian E, et al. Unifocalization of major aortopulmonary collaterals in single-ventricle patients. Ann Thorac Surg. 2006;82:934–8 **(discussion 8–9)**.
4. Cheng J, Fan CM, Tang M, et al. Initial research on postoperative management of tetralogy of fallot with major aortopulmonary collaterals. Cardiology. 2016;134:406–10.
5. Lanjewar C, Shiradkar S, Agrawal A, et al. Aneurysmally dilated major aorto-pulmonary collateral in tetralogy of Fallot. Indian Heart J. 2012;64:196–7.
6. Sharma A, Kumar S, Priya S. Ruptured aneurysm of major aortopulmonary collateral artery: management using amplatzer vascular plug. Cardiovasc Diagn Ther. 2016;6:274–7.
7. Carotti A, Albanese SB, Filippelli S, et al. Determinants of outcome after surgical treatment of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. J Thorac Cardiovasc Surg. 2010;140:1092–103.
8. Mainwaring RD, Patrick WL, Carrillo SA, et al. Prevalence and anatomy of retroesophageal major aortopulmonary collateral arteries. Ann Thorac Surg. 2016;102:877–82.
9. Cheng J, Fan C, Tang M, et al. Initial research on postoperative management of tetralogy of Fallot with major aortopulmonary collaterals. Cardiology. 2016;134:406–10.
10. Miyazaki O, Nosaka S, Hayakawa M, et al. Sudden death due to rupture of major aortopulmonary collateral arteries in a patient with tetralogy of Fallot and pulmonary atresia. Emerg Radiol. 2001;8:293–6.
11. Kish JW, Katz MD, Marx MV, et al. *N*-butyl cyanoacrylate embolization for control of acute arterial hemorrhage. J Vasc Interv Radiol. 2004;15:689–95.
12. Pollak JS, White RI. The use of cyanoacrylate adhesives in peripheral embolization. J Vasc Interv Radiol. 2001;12:907–13.
13. Toyoda H, Nakano S, Kumada T, et al. Estimation of usefulness of *N*-butyl-2-cyanoacrylate-lipiodol mixture in transcatheter arterial embolization for urgent control of life-threatening massive bleeding from gastric or duodenal ulcer. J Gastroenterol Hepatol. 1996;11:252–8.
14. Frodsham A, Berkmen T, Ananian C, et al. Initial experience using *N*-butyl cyanoacrylate for embolization of lower gastrointestinal hemorrhage. J Vasc Interv Radiol. 2009;20:1312–9.
15. Marelli AJ, Mackie AS, Ionescu-Ittu R, et al. Congenital heart disease in the general population: changing prevalence and age distribution. Circulation. 2007;115:163–72.
16. Khairy P, Ionescu-Ittu R, Mackie AS, et al. Changing mortality in congenital heart disease. J Am Coll Cardiol. 2010;56:1149–57.