

## Case Report

## An Unusual Combination of Congenital Anomalies in an Adult Patient: Patent *Ductus Arteriosus*, Kommerell's Diverticulum with Aberrant Right Subclavian Artery, and Heterotaxy Syndrome

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The heterotaxy syndrome is a rare and sporadic disorder. This syndrome presents with *situs ambiguus*, splenic malformations such as asplenia or polysplenia, and congenital heart disease. Congenital heart diseases associated with this syndrome include a broad variety of manifestations. Patent *ductus arteriosus* is one of them and percutaneous transcatheter closure can be challenging in the setting of this syndrome. Kommerell's diverticulum is a saccular aneurysmal dilation at the origin of an aberrant subclavian artery, and can be related with other congenital anomalies. However, there is no previous report of Kommerell's diverticulum being found together with patent *ductus arteriosus* and heterotaxy syndrome.

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**P**atent ductus arteriosus (PDA) is usually diagnosed and treated during childhood, but it can be seen in adults in developing countries.<sup>1</sup> Percutaneous transcatheter closure is a safe and alternative treatment modality to surgical closure in patients with PDA. The procedure is usually carried out using an antegrade approach via the right femoral vein under local anaesthesia.

The clinical entity characterised by *situs ambiguus*, congenital heart disease and splenic malformations (asplenia or polysplenia) is known as heterotaxy syndrome and is extremely rare in adults.<sup>2</sup> Percutaneous transcatheter closure with an antegrade wire-guided technique via the femoral vein can be challenging in the setting of heterotaxy syndrome, and a retrograde technique via the femoral artery may be used in these patients.

Kommerell's diverticulum is a rare (0.5-2%) saccular aneurysmal dilation of

the descending aorta at the origin of an aberrant right or left subclavian artery.<sup>3</sup> It is usually asymptomatic and most often an incidental finding. In symptomatic patients, the symptoms usually arise as a result of compression of the trachea and oesophagus.<sup>4</sup>

We report a case of PDA, Kommerell's diverticulum with an aberrant right subclavian artery, and heterotaxy syndrome consisting of a left-sided liver and right-sided three-piece spleen, where successful transcatheter closure of the PDA was achieved with a retrograde wire-guided technique using an Amplatzer Duct Occluder device.

### Case presentation

A 38-year-old man with a history of chronic obstructive lung disease was admitted to our department because of worsening of his symptoms, including dizziness,

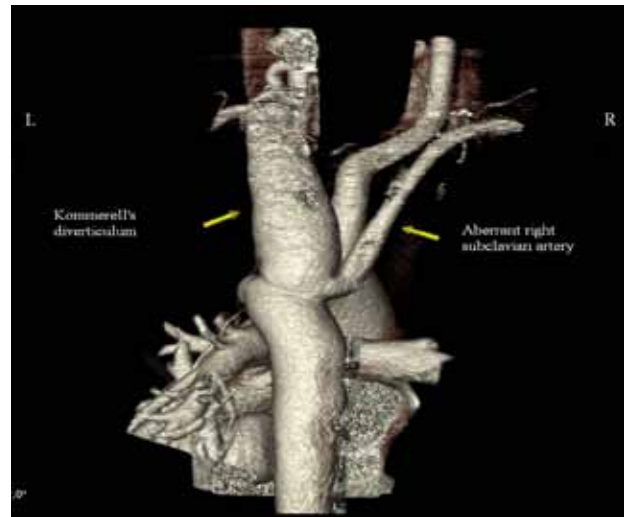
dyspnoea and dysphagia. Physical examination and resting 12-lead electrocardiogram (ECG) were performed at admission. His heart rate was 37 beats/min with a regular pulse, blood pressure was 100/70 mmHg and oxygen saturation was 89%. The electrocardiogram revealed a nodal rhythm with a rate of 37 beats/min. Two-dimensional echocardiography showed a PDA, mitral valve prolapse, degenerative mitral valve insufficiency (moderate), biatrial dilatation and a left ventricular ejection fraction of 66%.

For the evaluation of his dyspnoea and dysphagia, a chest computerised tomography (CT) examination was performed. The CT scan revealed a Kommerell's diverticulum with an aberrant right subclavian artery and a PDA (Figures 1 & 2).

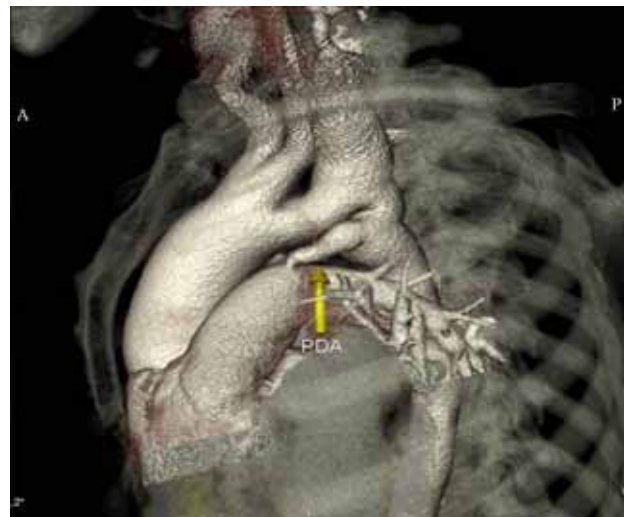
Percutaneous transcatheter closure of the PDA was performed using an antegrade wire-guided technique. The patient was taken to the catheterisation laboratory. The procedure was initiated by puncturing the right femoral vein under local anaesthesia. After several attempts, we were able to advance the 0.0035" guidewire through the inferior *vena cava* (IVC) into the right atrium. However, we were unable to advance the 6 F right Amplatz II diagnostic catheter over the guidewire.

The procedure was stopped, and a CT examination of the abdomen and pelvis was performed for the evaluation of the technical difficulties involved in advancing the catheter into the right heart via the femoral vein. The CT abdominal scan revealed a left-sided liver and right-sided spleen in three pieces (Figure 3). In light of these findings, we diagnosed a heterotaxy syndrome. Although there was no evidence for interruption of the IVC with an azygous continuation, we were not able to perform percutaneous closure of the PDA using an antegrade technique. We thus decided to perform a retrograde wire-guided technique via the femoral artery. An Amplatzer Duct Occluder device (ADO II, AGA Medical Corp., Golden Valley MN, USA) was used in this retrograde wire-guided technique. The distal disc of the ADO II device was opened in the pulmonary artery, then the system was withdrawn slowly and the proximal disc opened in the aorta. The device was then released after a check that it had been placed correctly (Figure 4). The closure of the PDA via an arterial approach in this patient was achieved without any difficulty.

The patient also had symptomatic bradycardia and his symptoms persisted. Therefore, a permanent pacemaker programmed in DDD-R mode was implanted via the right subclavian vein and the patient's dizziness was relieved. There was no abnormality in



**Figure 1.** The three-dimensional computed tomography scan shows a Kommerell's diverticulum with aberrant right subclavian artery.

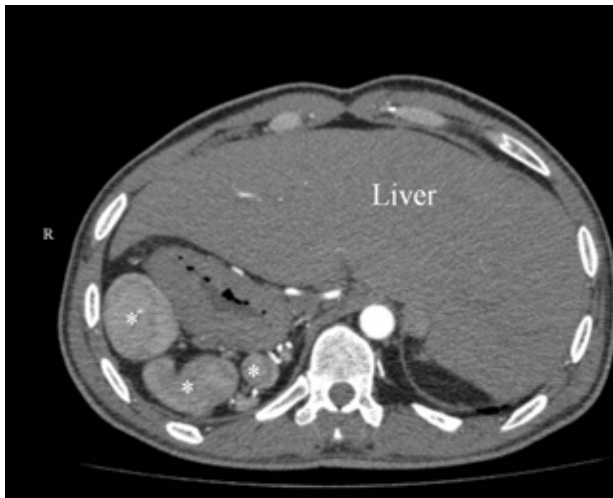


**Figure 2.** The three-dimensional computed tomography scan shows a patent *ductus arteriosus* (PDA).

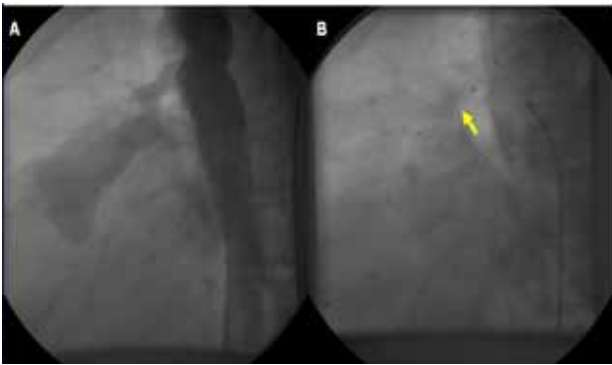
the following days and the patient was discharged in very good condition.

## Discussion

The incidence of PDA is very low in adults, since it is usually diagnosed and treated during childhood.<sup>1</sup> The mortality of untreated PDA in adults is estimated to be 1.8% per year.<sup>5</sup> The first successful transcatheter occlusion was reported by Porstmann et al<sup>6</sup> and this treatment modality has continued to evolve. However, there are various complications and challenges related to this procedure.



**Figure 3.** Computed tomography of the abdomen in horizontal section shows the left-sided liver and right-sided three-piece spleen (parts denoted by \*).



**Figure 4.** The descending aortography demonstrates a patent ductus arteriosus and a severe left-to-right shunt. A. Transcatheter closure of the patent ductus arteriosus with a retrograde guidewire technique using an Amplatzer Duct Occluder (ADO II) device. B. The yellow arrow denotes the double-disc ADO II device with platinum markers.

Heterotaxy syndromes are characterised by complex abnormalities of abdominal organs, such as *situs ambiguus*, asplenia or polysplenia.<sup>7</sup> Congenital heart diseases are frequently associated with heterotaxy syndromes,<sup>7,8</sup> and the prognosis mainly depends on the degree of malformation. Atrial and ventricular septal defects, PDA, absent coronary sinus, abnormal location of the cardiac apex, common atrioventricular canal, pulmonic stenosis, pulmonary atresia, anomalous pulmonary venous connection, anomalous systemic venous return (e.g. bilateral superior *vena cava*, interruption of IVC with azygous continuation), and malposition or transposition of the great arteries, are the congenital heart defects that have been reported in association with this syndrome.

Heterotaxy syndromes are less frequent in adults because of the early death of children with severe cardiac abnormalities. However, 5-10% of patients have no major cardiovascular defects and are not diagnosed until adulthood.<sup>9</sup> Our patient was not diagnosed with heterotaxy syndrome until he was 38 years old, because of the absence of severe structural or functional cardiac defects.

Interruption of the IVC is defined as lack of development of the IVC below the level of the hepatic vein, with a well developed azygous or hemiazygous collateral continuation, and can occur in the setting of heterotaxy syndrome. A congenital anomalous or interrupted IVC is one of the challenges in interventional cardiology, as regards the traditional antegrade approach via the femoral vein. In our case, the CT examination did not reveal an interruption of the IVC. However, advancing the catheter into the right heart via the right femoral vein proved to be so difficult that we decided to perform a retrograde wire-guided technique via the femoral artery using an ADO II device. The procedure was completed successfully.

The saccular aneurysmal dilation of the descending aorta at the origin of the right or left subclavian artery is called Kommerell's diverticulum. There are no published data showing the presence of Kommerell's diverticulum in patients with heterotaxy syndrome. Curiously, Kommerell's diverticulum with an anomalous right subclavian artery was detected incidentally during a CT scan in our case. Although most patients with Kommerell's diverticulum are asymptomatic, dilatation of Kommerell's diverticulum and a retro-oesophageal course of the aberrant subclavian artery can compress the surrounding structures, such as the oesophagus and the trachea. As a result, the patient may experience symptoms such as dysphagia, dyspnoea, stridor, wheezing, cough or chest pain.<sup>10</sup> In our patient the CT examination did not reveal compression of the trachea or oesophagus and we decided that no operation for Kommerell's diverticulum was necessary.

In conclusion, heterotaxy syndromes are frequently associated with various congenital heart diseases and are rare in adults. These uncommon entities will be diagnosed more frequently by the use of CT imaging techniques. To the best of our knowledge, this is the first case report concerning the association between PDA, Kommerell's diverticulum and heterotaxy syndrome. Furthermore, the traditional antegrade wire-guided percutaneous transcatheter closure of PDA, which seems to be a simple proce-

dure, can be challenging in the setting of heterotaxy syndrome, especially in the presence of interruption of the IVC. A retrograde wire-guided approach using an ADO II device should be chosen for percutaneous transcatheter closure of PDA in these patients.

## References

1. Wiyono SA, Witsenburg M, de Jaegere PPT, Roos-Hesselink JW. Patent ductus arteriosus in adults: Case report and review illustrating the spectrum of the disease. *Neth Heart J*. 2008; 16: 255-259.
2. Ivemark BI. Implications of agenesis of the spleen on the pathogenesis of conotruncus anomalies in childhood; an analysis of the heart malformations in the splenic agenesis syndrome, with fourteen new cases. *Acta Paediatr Suppl*. 1955; 44: 7-110.
3. Backer CL, Hillman N, Mavroudis C, Holinger LD. Resection of Kommerell's diverticulum and left subclavian artery transfer for recurrent symptoms after vascular ring division. *Eur J Cardiothorac Surg*. 2002; 22: 64-69.
4. van Son JAM, Konstantinov IE, Burckhard F, Kommerell and Kommerell's diverticulum. *Tex Heart Inst J*. 2002; 29: 109-112.
5. Campbell M. Natural history of persistent ductus arteriosus. *Br Heart J*. 1968; 30: 4-13.
6. Porstmann W, Wierny L, Warnke H. Closure of persistent ductus arteriosus without thoracotomy. *Ger Med Mon*. 1967; 12: 259-261.
7. Phoon CK, Neill CA. Asplenia syndrome: insight into embryology through an analysis of cardiac and extracardiac anomalies. *Am J Cardiol*. 1994; 73: 581-587.
8. Winer-Muram HT. Adult presentation of heterotaxic syndromes and related complexes. *J Thorac Imaging*. 1995; 10: 43-57.
9. Winer-Muram HT, Tonkin IL, Gold RE. Polysplenia syndrome in the asymptomatic adult: computed tomography evaluation. *J Thorac Imaging*. 1991; 6: 69-71.
10. Patiniotis TC, Mohajeri M, Hill DG. Right aortic arch with aberrant left subclavian artery: aneurysmal dilatation causing symptomatic compression of the right main bronchus in an adult. *Aust N Z J Surg*. 1995; 65: 690-692.