Patent ductus arteriosus in adults

Case report and review illustrating the spectrum of the disease

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Patent ductus arteriosus (PDA) is a congenital heart defect in which the *ductus arteriosus*, a vascular structure between the pulmonary artery and the aorta that normally closes shortly after birth, remains open. We present two cases of adults with PDA. A 28-year-old lady had a small PDA without any symptoms or left heart overload. The PDA was closed for endocarditis prevention using a coil. In a 37-year-old lady with palpitations, collapse, dyspnoea and left heart overload caused by a large PDA, percutaneous closure of the PDA was performed with an Amplatzer device. Transcatheter closure has been established to be the method of choice for treating a PDA in adults. (*Neth Heart J* 2008;16:255-9.)

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Ductus arteriosus is a vascular structure that connects the proximal descending aorta to the roof of the main pulmonary artery near the origin of the left pulmonary artery. It normally closes spontaneously within 24 to 48 hours after birth by the contraction of medial smooth muscle in the vessel wall, due to the increased oxygen tension and reduced prostaglandin E₂ and I₂ levels. Endothelial adhesion followed by replacement of the muscle fibres with connective tissue results in the remaining ligamentum arteriosum within two to three weeks. If this does not happen, there is a patent or persistent ductus arteriosus (PDA).

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The incidence of PDA is approximately 1 in 2000 in full-term infants and consists 5 to 10% of all congenital heart disease in children.¹ PDA is found twice as often in females than in males.¹ In adulthood a PDA is not often encountered since it is usually discovered and treated during childhood. The mortality of untreated PDA in adults (without correction for the size of the PDA) is estimated to be 1.8% per year.² We present two cases illustrating the spectrum of patent ductus arteriosus at adult age and its management.

Case report

Patient 1

A 28-year-old sport instructress with no cardiac history came to the outpatient clinic for cardiac screening for congenital heart disease because her brother had a bicuspid aortic valve.

She had no symptoms and an excellent exercise capacity. On physical examination we saw a healthy young lady with a height of 173 cm and a weight of 68 kg. Her heart rate was 60 beats/min and her blood pressure 110/60 mmHg. First and second heart sounds were normal; a continuous murmur grade 2/6 left

Figure 1. Preclosure transthoracic echocardiography (suprasternal view) with colour Doppler flow from the proximal descending aorta (desc. Ao) through the PDA to the pulmonary artery (PA) of patient 1.

Figure 2. A. Preclosure angiography of the PDA in patient 1. Desc. Ao=proximal descending aorta, PA=pulmonary artery. B. Postclosure angiography of the PDA in patient 1. C. Picture of a coil.

subclavicular was heard. The breathing sounds were normal. There were no signs of venous congestion.

The ECG and chest X-ray showed no abnormalities. During echocardiographic examination, a small shunt from the proximal descending aorta to the main pulmonary artery was seen (figure 1). The diagnosis of patent ductus arteriosus (PDA) was made. There were no signs of left ventricle volume overload and no signs of increased pulmonary artery pressure. There was no evidence of calcification of the ductus.

The patient underwent successful percutaneous closure of her PDA with a four-loop 5 mm coil (Flipper[®] Cook Medical, Ireland), which was introduced from the aortic side (figure 2).

Patient 2

A 37-year-old cleaning lady, with a documented PDA, visited the emergency room because of palpitations, collapse and dyspnoea. An attempt to percutaneously close the PDA was made in 1993, which was unsuccessful because the shunt was too large using the available devices at that time. She did not return to the outpatient clinic until she developed the symptoms described above.

In addition, she had progressive limitations of her daily activities and had to stop working. At physical examination, she was 155 cm with a weight of 57.5 kg. The heart rate was 83 beats/min and blood pressure 155/70 mmHg. There were normal first and second heart sounds and also a third heart sound was heard with a continuous murmur in the second left intercostal space. The breathing sounds were normal. There were no signs of venous congestion.

The ECG showed normal sinus rhythm with signs of left atrial dilatation, left ventricle hypertrophy or dilatation. The heart rhythm monitor showed paroxysmal atrial fibrillation (PAF). The chest X-ray demonstrated cardiomegaly and increased pulmonary vascular markings.

Echocardiography demonstrated a continuous wide flow from the aorta through the ductus arteriosus to the pulmonary artery (figure 3A and B). Maximum flow velocity was 4.5 m/sec, indicating an elevated pulmonary artery pressure. The left ventricle was dilated with an end-diastolic diameter of 82 mm and end-systolic diameter of 65 mm (figure 3C). The systolic function was moderately impaired.

Cardiac catheterisation revealed systolic pulmonary pressure of 70 mmHg and a Qp:Qs of 3:1. The duct diameter was 11 mm. Because of these findings (pulmonary hypertension and left ventricle volume overload) the PDA was successfully closed with an Amplatzer ductus occluder 16-14 mm (ADO, AGA Medical Corporation, Minnesota, USA) (figure 4).

The small residual shunt, still seen with echocardiography one day after intervention, disappeared during follow-up (figure 5). After 18 months of follow-up, the left ventricular dimensions diminished to an end-

Figure 3. A. Preclosure transthoracic echocardiography (supra-sternal view) with colour Doppler flow from the proximal descending aorta (desc. Ao) through the PDA to the pulmonary artery (PA) of patient 2. B. Preclosure pulsed wave transthoracic echocardiography in the PDA of patient 2 showed continuous flow. C. Transthoracic echocardiography of patient 2 showing a dilated left ventricle (LV). LA=left atrium, RV=right ventricle, RA=right atrium

Figure 4. A. Preclosure angiography of the PDA in patient 2 with the catheter from the pulmonary artery (PA) through the PDA to the proximal descending aorta (desc. Ao). B. Postclosure angiography of the PDA in patient 2. Amplatzer closure device in situ. C. Picture of an Amplatzer closure device.

Figure 5. Postclosure transthoracic echocardiography (suprasternal view) with and without colour Doppler flow through the PDA of patient 2 showed no rest shunt. Desc. Ao=proximal descending aorta, PA=pulmonary artery.

diastolic diameter of 64 mm and end-systolic diameter of 57 mm. The left ventricle systolic function had not significantly improved. Despite this, the patient felt much better after treatment. She had no symptoms and was able to perform her job normally.

Both of these patients were recommended to take endocarditis prophylaxis until six months after treatment.

Discussion

The PDA in adults is usually a coincidental finding during physical examination or echocardiography screening. Clinically, a PDA has a typical continuous murmur which can be heard at the higher left sternal edge. It may be associated with a wide pulse pressure due to the runoff to the pulmonary circulation.

The ECG of patients with a large PDA (left-right shunt) may show left atrium dilatation and left ventricle strain (patient 2). In patients with a small shunt, the ECG is expected to be normal. Echocardiography is

important to establish the diagnosis of PDA, recognise volume load, estimate pulmonary artery systolic pressure with flow velocity through the duct and identify associated cardiac pathology. Even an extremely tiny patent ductus can be detected by a colour flow signal entering the pulmonary artery.

The clinical severity grading of the PDA in adults is depicted in table 1.³

Cardiac catheterisation is used to assess the haemodynamic situation before closure of the PDA, and is particularly important in adults to evaluate the pulmonary vascular resistance and the degree of shunting. Temporary test occlusion with balloon catheter may provide important information regarding advisability of closure.⁴

Patient 1 had a tiny PDA without haemodynamic significance; therefore, diagnostic cardiac catheterisation for decision-making or preparation of closure of the PDA was not necessary. In this patient the risk of endocarditis was the indication for closure.

Patients with a significant left heart volume overload caused by a moderate to large PDA, such as patient 2, have a risk of congestive heart failure and irreversible pulmonary hypertension.¹

In patient 2 at longer term follow-up, the left ventricular dimensions had decreased but the ventricular function was still impaired. This is conform the findings of Young et al. They showed that the left ventricular dimensions had significantly decreased after PDA closure and preclosure left ventricular ejection fraction (LVEF) was the only independent predictor of late normal postclosure LVEF. They saw no improvement of LVEF late after closure of the PDA compared with the preclosure state in adults and suggested that closure should be performed before LVEF decreases.⁵

Patients with a small duct, such as patient 1, often remain asymptomatic during infancy and childhood and may never develop symptoms. Regardless of the size, complications may arise; most importantly, endarteritis (endocarditis of the artery). The incidence of

Table 1. The clinical severity grading of the PDA in adults.

Type of the PDA	Murmur	Wide pulse pressure	Dilated left ventricle	Pulmonary hypertension
Silent	-	-	-	-
Small	Continuous	-	-	-
Moderate	Continuous	+	+	+
Large	Systolic + Diastolic ±	±	++	++
Eisenmenger	Ejection murmur	-	++	+++

endarteritis in adults with a PDA has been reported to be over 0.45% per year.² However no recent data on the incidence are available. Endocarditis is responsible for almost half of the deaths in patients with untreated PDA.⁶ Although patients with a small PDA have a normal life expectancy, the Dutch guidelines advise closing the PDA when a murmur is present to prevent endarteritis. Vegetations usually occur on the pulmonary side of the ductus, and embolic events occur in the lungs rather than in the systemic circulation. The risk of endarteritis, and therefore associated complications, is abolished by closure of the PDA, which carries very little morbidity and almost no mortality.¹ This is the reason why patient 1 was advised to undergo an interventional closure of the PDA.

The definitive treatment of PDA is by closing it either by a transcatheter approach or by surgery. Transcatheter closure has been established to be the method of choice for treating a PDA in adults with very good outcome. However, surgical closure is still the method of choice for treating very large PDAs not amendable for catheter intervention.¹

At present, thrombogenic coils are used to treat a small ductus and an Amplatzer occluder for a ductus larger than 3 mm. Transcatheter techniques are effective and safe with complete closure rates of 90 to 95% in most studies.^{1,7} With the present techniques residual shunting is usually resolved within 24 hours; however, it may take several weeks to months when large occluders are used. Complications occurring after closure are rare but may include delayed migration of the device, flow disturbance in the left pulmonary artery

or descending aorta from a protruding device, haemolysis from high velocity residual shunting, distal embolisation, thrombosis of the vascular access, and infection.^{1,7,8}

Conclusion

Transcatheter closure is the primary choice for treating a PDA in adults. Next to volume overload, prevention of endocarditis is an indication for treatment, also in small ducts. In a small PDA a coil can be used while in moderate to large PDA it is recommended to use an Amplatzer device. ■

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