Spontaneous Closure of a Large Ductus Arteriosus Aneurysm

To the Editor,

Spontaneous ductus arteriosus aneurysm (DAA) is a sack-like or spindle shaped dilatation due to an intrinsic anomaly of intraductal tissue related to elastin. It is usually diagnosed within the first 2 months of life,¹ but can be detected at any age, occasionally after surgical closure of a patent ductus.² This is a rare potentially severe heart condition; however, prenatal screening has shown a greater prevalence than expected and a more benign natural evolution during infancy.²

We present the case of a child with a large DAA diagnosed at birth by echocardiography and computed tomography angiography (CT-angio). A see and wait attitude was adopted with series of echocardiograms and complete resolution was seen. Infant of 49 days of age, born at term, weighed 4.330 kg at birth, with a normal perinatal period, without maternal diabetes, brought for observation and treatment to our service with a diagnosis of DAA. At birth, an echocardiographic study showed a round image on the left of the pulmonary trunk was seen with marked autocontrast in its interior, indicating a ducal aneurysm, subsequently confirmed by CT-angio: partially thrombosed aneurysmatic formation of 20 mm in diameter with a narrow opening (6 mm) on the anterior face of the descendent aorta and with no communication with the pulmonary artery.

No symptoms were found on physical exam on admission, no murmurs were auscultated, peripheral pulses were preserved and there was no Marfanoid habitus. On x-ray it was possible to see a convex image between the first and second left costal arches. A transthoracic echocardiogram confirmed a large DAA adjacent to the left pulmonary branch, with thrombotic material in its interior and with no evidence of a shunt, which correlated with the CT-angio findings (Figure).

Given the lack of symptoms and the evidence
of thrombosis within the aneurysm a conservative strategy was maintained. Echocardiographic control at 3 months was completely normal, with no image of an aneurysm of the permeable ductus.

Spontaneous or congenital DAA is usually not detected and, in many cases, is an incidental finding. In adult patients the pulmonary end is frequently found closed and this is confused with a thoracic aorta aneurysm, which means that its incidence could be underestimated. Predisposing risk factors are considered to be maternal diabetes mellitus, high weight at birth and connective tissue disease. Current incidence is estimated at 1.5%-8.8%. Diagnosis can be reliably performed with echocardiography, CT-angio and magnetic resonance.

Management of spontaneous DAA must be careful without ignoring the possibility of potential complications. Spontaneous rupture with fatal consequences has been described in patients with Marfan syndrome. Furthermore, there have been reports of thrombi extension to the pulmonary arteries or aorta and even thromboembolism, including association with bacterial infections. Large ones may compress or cause erosion of neighbouring structures, vascular, airways or the recurrent laryngeal nerve. The true incidence rate of these complications is unknown, although it is known to be high in older children and adults. In some revisions surgical treatment is indicated in patients of more than 2 months of age if they have connective tissue disease, infectious syndrome or if there is a aortopulmonary shunt with clinical repercussion. It is advisable to closely monitor the patient with these characteristics up to 2-3 months of age to confirm spontaneous remission. Previously aneurysms of more than 10 mm were operated, although there is no evidence that size is related to a greater complication rate. In our case, the DAA was large, but the absence of symptoms and aortopulmonary communication, as also the age of the patient, led us to adopt an attitude of “wait and see.” Echocardiographic control at 3 months showed total resolution of the condition with no events; however we continue with regular check-ups to rule out connective tissue diseases of late appearance.

Iria A González, Carmen González, Mario Cazzaniga, and Luis Fernández

Servicio de Cardiología, Hospital Clínico Universitario, Valladolid, Spain
Unidad de Cardiopatías Congénitas y Cardiología Pediátrica y Congénita, Hospital Ramón y Cajal, Madrid, Spain

REFERENCES


To the Editor,

Patent foramen ovale (PFO) is an incomplete closure of the interatrial septum. This is a frequent finding in healthy individuals. Under conditions in which there is an increase of pressure in the right chambers of the heart, a right-left shunt may arise, with variable degrees of secondary hypoxaemia.

A 26-year-old male was admitted for craniocephalic trauma, which evolved without complications, and chest trauma, with pneumothorax and lung contusion. The patient was treated with bed rest, mechanical ventilation (MV) and insertion of a pleural drainage. The patient was kept in controlled-assisted ventilation with a FiO$_2$ of 0.4 and without PEEP. On the second day he developed hypoxaemic respiratory failure secondary to lung infection, which required a FiO$_2$ of 1. PEEP was increased, but no significant changes were seen in oxygenation or ventilation.

On day 10 it was necessary to start noradrenaline administration. A transthoracic echocardiogram (TTE) was performed that did not detect any alteration.

On day 12 PEEP had reached 20 cmH$_2$O and respiratory failure persisted. A chest computed tomography (CT) was taken, this showed lung condensation, with no other complications, and a cranial CT was taken, that showed an acute ischaemic lesion. A transesophageal echocardiogram (TEE) was performed that showed an aneurysm in the interatrial septum and a patent foramen ovale (PFO) (Figures 1 and 2). The administration of shaken saline fluid as bubble contrast confirmed the shunt. PEEP was reduced from 20 to 5 cmH$_2$O and SaO$_2$ improved immediately from 75% to 92%. Simultaneously, thrombosis of the left jugulosubclavianaxillary axis was detected by catheterisation. The stroke was attributed to paradoxical embolism through the persistent foramen ovale (PFO).

As the patient continued to require a FiO$_2$ of 1, the percutaneous closure of the PFO was planned. Twenty-four hours before placement of the device the patient presented pneumothorax. Drainage was performed, but respiratory failure persisted and the patient died.

A PFO is an incomplete closure of the interatrial septum. This condition is present in 25%-30% of healthy individuals, with similar prevalence in both sexes. Its cause is unknown, but it is possible there may be a genetic component. It can be associated with aneurysm of the interatrial septum or a Chiari network or a more prominent Eustachian valve. PFO is diagnosed by TEE; TTE, or transcranial Doppler. Transesophageal echocardiogram (TEE) is the most sensitive and specific of these techniques and makes it possible to study other associated structures. Transthoracic echocardiogram (TTE) has a high specificity, but less sensitivity and is more operator-dependent and acoustic window-dependent, which makes it of more limited use in ventilated patients with positive pressure.

Most individuals with PFO are asymptomatic. Its importance has been classically associated with ischaemic stroke caused by paradoxical embolism. For it to take place the pressure gradient between the atria must become inverted. PFO has also been implicated in paradoxical embolisms of other kinds: fatty, tumour and air (in decompression accidents) and is associated with migraine, vascular headache and platypnea-orthodeoxia syndrome.