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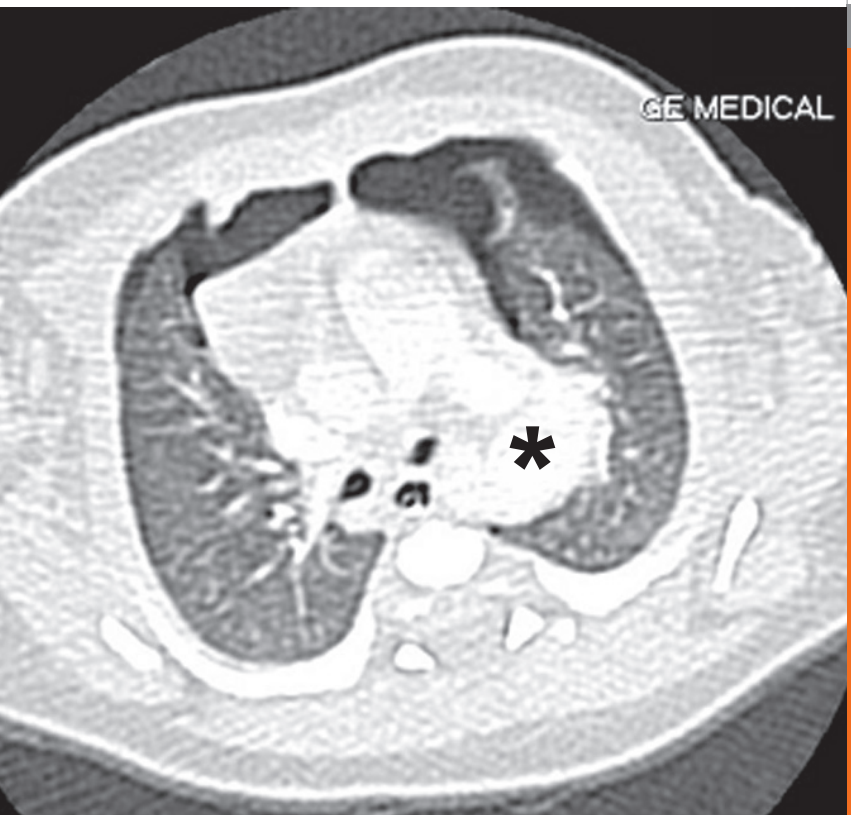
Congenital ductus arteriosus aneurysm: serious or common?



AUGUST 2011

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We report the case of a female infant born from a healthy 40-year-old G1/P1. Pregnancy was remarkable for a well-controlled pre-gestational insulin-dependent diabetes. At 33 weeks of gestation, an aneurysm of the ductus arteriosus was discovered by prenatal ultrasound and echocardiography. It had a 10 mm diameter and was associated with slight ventricular asymmetry. The diameters of the pulmonary artery and aorta were 11 and 5 mm, respectively, with normal flow (Fig. 1). Fetal growth was normal.

Because of these cardiac abnormalities, labor was induced at 38 6/7 weeks of gestation, and when late decelerations occurred, an emergency caesarean section was performed. The girl was born with Apgar scores of 5, 7 and 8 at 1, 5 and 10 minutes, respectively. The umbilical cord pH values were 6.98 and 7.06. Because of non-efficient breathing and respiratory distress, bag mask ventilation with an FiO2 of 0.5 was necessary during the first 3 minutes of life. Respiratory distress disappeared at 10 minutes without any ventilatory support. Capillary pH after 10 minutes showed a mixed acidosis (pH 6.9, pCO2 85 mmHg, lactate 13 mmol/l). Respiratory symptoms and blood gases normalized quickly after 90 minutes of life.

Physical examination showed an AGA term infant. There was a heart murmur with dorsal irradiation and slight hepatomegaly extending 3 cm below the costal margin. There was a paracardiac opacity corresponding

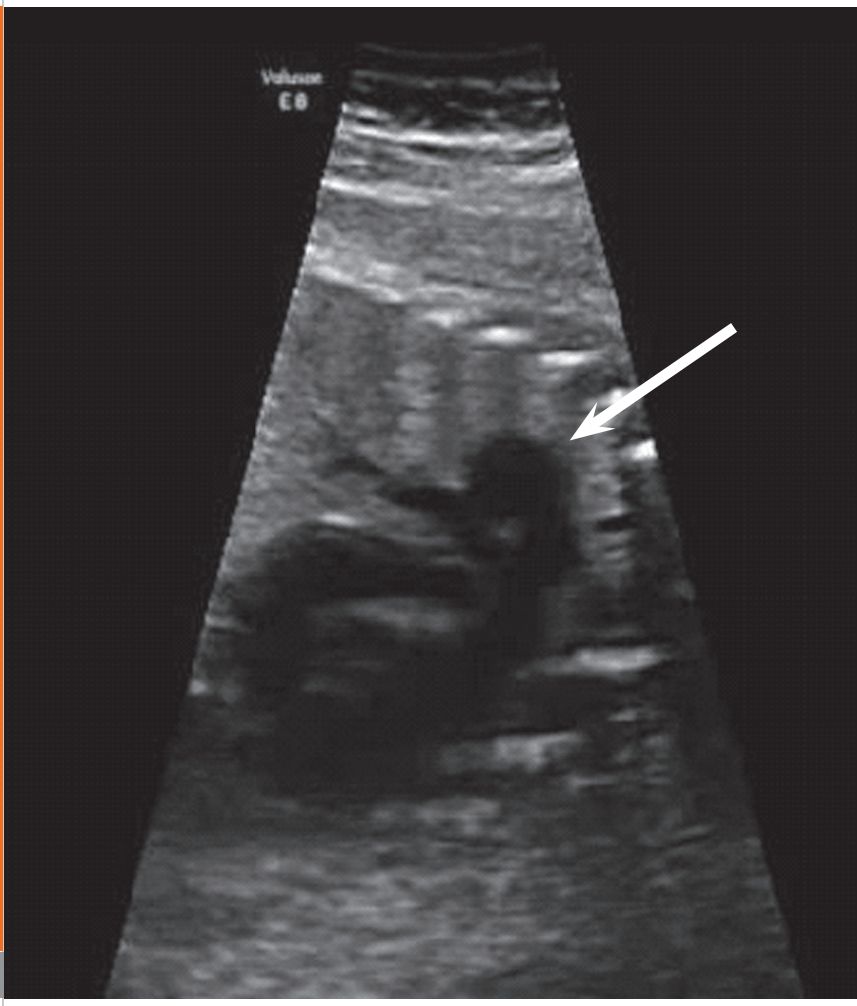

Fig. 1

Fetal ultrasound at 33 1/7 weeks of gestation showing the ductal aneurysm (white arrow).

to a ductal bump on chest X-ray on the first day of life which became progressively smaller over time (Fig. 2). Echocardiography confirmed the presence of a structure measuring 10 mm in diameter consistent with a ductus arteriosus aneurysm (DAA) without any other structural cardiac abnormalities. To better define the structure, a cardiac CT scan was performed and confirmed the presence of a 13 mm DAA without any other cardiac abnormalities (Fig. 3, 4). The CT scan also revealed unexpected bilateral asymptomatic pneumothoraces which resolved spontaneously. The child had no clinical symptoms related to the aneurysm and echocardiographic follow-up 6 months of age documented closure of the ductus arteriosus.

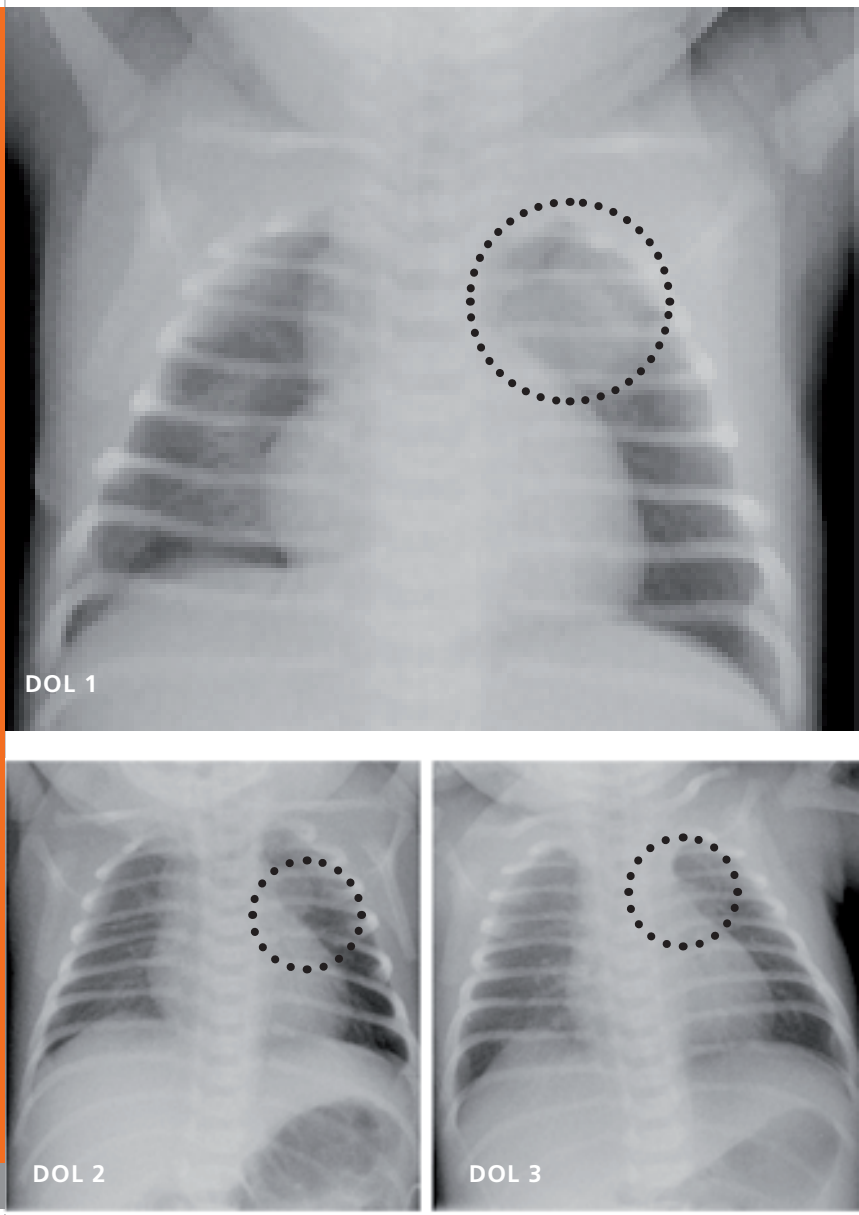

Fig. 2

Chest X-rays performed on day of life (DOL) 1, 3 and 5 illustrate progressive disappearance of the ductal bump (circles).

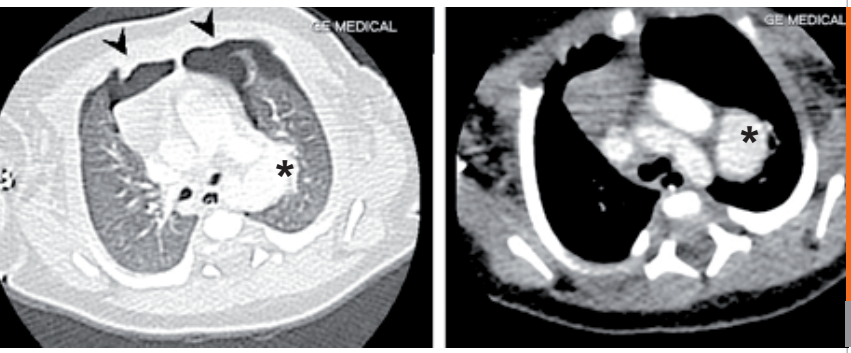

Fig. 3

Cardiac CT scan (DOL 2) showing the ductal aneurysm measuring 13 mm in diameter (asterisk); in addition, there are bilateral anterior pneumothoraces (arrow heads).

DISCUSSION

Congenital DAA is in fact a frequent entity, but probably underdiagnosed. Already in 1850, a prevalence of 0.8% of DAA was observed in neonatal autopsies, and in 1974 a retrospective review of chest X-rays of newborn infants with respiratory distress revealed a "ductal bump" in 1% of patients (1, 2). More recently, a retrospective review of 200 fetal ultrasound examinations performed after 30 weeks of gestation, suggested a DAA incidence of 1.5% (3) in the analysed population. An echocardiographic longitudinal study of 509 fetuses from 32 weeks of gestation until term revealed DAAs in 2.2% (4). Interestingly, ultrasound screening of 548 healthy term newborns showed an even higher prevalence of 8.8 % of isolated DAAs (5). Jan et al. suggested that the incidence was previously underestimated because of the absence of symptoms

before complications occur and because most patients do not develop complications. Complications of isolated DAAs include compression of adjacent structures, erosion into airways, infection, thromboembolism and spontaneous rupture (5).

How DAAs develop is not clearly understood and probably multifactorial. Several theories have been presented. The first theory suggests that a delayed closure of the aortic end of the ductus arteriosus exposes the ductal wall to systemic pressure and leads to aneurysm formation (3). However, this would not explain antenatal cases. A second hypothesis is based on the assumption of a congenital weakness of the ductal wall. Under normal conditions, modifications of the intimal cushions of the ductus arteriosus occur during the third trimester in preparation for postnatal closure. These modifications consist of 1) smooth muscle cell proliferation and migration into the subendothelial space and 2) an increased production of glycosaminoglycans, hyaluronan, chondroitin sulphate and fibronectin which are all essential for smooth muscle cell migration. Interestingly, DAAs have never been identified before the third trimester. Dyamenahalli et al. (3) suggested that DAAs may be the result of reduced intimal cushion formation or abnormal deposition of elastin resulting in weakening of the vessel wall. Indeed, several cases of DAA have been described in the literature in patients with connective tissue disorders (Marfan syndrome, Ehler-Danlos syndrome) (3).

Finally, it is known that the intimal thickening process begins at the pulmonary end of the ductus arteriosus. Higher blood flow velocities at the aortic end of a more markedly curved ductus can cause a stress in the ductal wall and contribute to DAA formation (5) (Fig. 5). Hormonal factors such as estrogen and progesterone have also been implicated in DAA development.

Jan et al. (5) describe an increased incidence of DAAs in large for gestational age infants and in infants of diabetic mothers. This could perhaps be explained by circulating factors influencing the development of intimal cushions or extracellular matrix deposition in the ductus arteriosus (6). Further, maternal blood group A seems to be more common among neonates with DAA but the underlying mechanism remains unclear (5, 6).

Diagnosis is mostly done in the prenatal period or as an incidental finding during investigations for other symptoms after birth. Echocardiography is the most important tool for diagnosis and follow-up of DAA and mostly sufficient. Alternative imaging techniques such as CT scan or MRI should be considered if the evaluation is difficult or if there are clinical signs of complications. Chest X-ray can suggest the diagnosis by the presence of "ductal bump" corresponding to the DAA as seen in our patient.

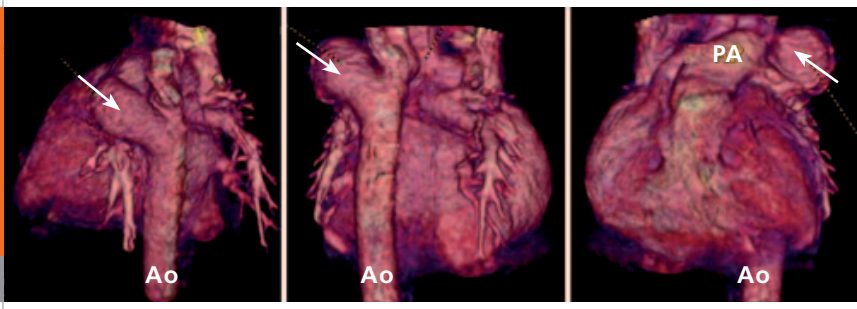

Fig. 4

Three dimensional CT scan reconstruction (DOL 2) showing the ductal aneurysm (arrows) (PA: pulmonary artery; Ao: aorta).

Management consists in regular follow-up and most of the time the outcome is favourable with spontaneous closure of the ductal aneurysm. In all cases described in the literature, spontaneous closure occurred mostly during the first month of life and never later than at two months (3-5).

According to the experience of Jan et al, ductal aneurysm resolution may occur in two ways. In two thirds of the patients, aneurysms become progressively smaller with or without thrombus formation after ductal closure. In remaining one third of patients, progressive thrombus formation in the aneurysm becomes organised and fibrosed (5).

Because of its potential association with connective tissue disorders, it is important to exclude a relevant family history or additional clinical evidence of such

A

B

Fig. 5

Wall stress at the pulmonary end of the ductus arteriosus with a locus minoris resistentiae (A) leads to DAA formation in the curved part of the ductus arteriosus (B).

a disease entity. Without additional evidence, clinical follow-up of neonates with DAA will be sufficient. The presence of both a DAA and bilateral pneumothoraces in our patient raised concerns regarding a connective tissue disorder in our patient. However, we could not find any cases describing neonatal pneumothoraces as an early manifestation of a connective tissue disorder in the literature. In addition, there were no additional clinical signs and the patient's family history was negative.

Ductal aneurysm regression after indomethacin has been described in two case reports, but the real effect of this drug on DAA is uncertain and spontaneous evolution is mostly favorable (5, 7, 8).

Surgical resection of the aneurysm can be considered in some particular situations such as DAA persisting beyond the neonatal period, association with connective tissue disorder, thrombus extension into adjacent vessels, evidence of thromboembolism and functional compression of adjacent structures (3-6). Size of aneurysm does not seem to be relevant to decide if surgical resection is necessary because spontaneous rupture has been described with DAA diameter of only 8 mm (3).

In conclusion, DAA is not uncommon. Most infants with DAA are asymptomatic and spontaneous closure within one to two months after birth is the rule. Because of its potential association with connective tissue

disorders, long-term follow-up of these children is recommended.

REFERENCES

1. Thore J. De l'anévrisme du canal artériel. Arch Gen Med (Paris) 1850;23-30

2. Heikkinen ES, Simila S, Laitinen J, Larmi T. Infantile aneurysm of the ductus arteriosus. Acta Pediatr Scand 1974;63:241-248

3. Dyamenahalli U, Smallhorn J, Geva T et al. Isolated ductus arteriosus aneurysm in the fetus and infant: A multi-institutional experience. JACC 2000;36:262-269

4. Tseng JJ, Jan SL. Fetal echocardiographic diagnosis of isolated ductus arteriosus aneurysm: a longitudinal study from 32 weeks of gestation to term. Ultrasound Obstet Gynecol 2005;26:50-56

5. Jan SL, Hwang B, Fu YC et al. Isolated neonatal ductus arteriosus aneurysm. JACC 2002;39:342-347

6. Hornberger LK. Congenital ductus arteriosus aneurysm. JACC 2002;39:348-350

7. Doege C, Linderkamp O, Gorenflo M et al. Treatment of an aneurismal dilatation of the ductus arteriosus with indomethacin in a premature infant. Pediatr Cardiol 2006;27:166-167

8. Lee WJ, Chen SJ, Wu MH et al. Regression of ductus arteriosus aneurysm in a neonate demonstrated by three-dimensional computed tomography. Int J Cardiol 1999;68:231-234

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