

Echocardiographic Features and Management of Neonatal Ductal Aneurysm

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Abstract

Objectives: To determine the case incidence of ductus arteriosus aneurysm among our neonates, and to report on our experience in the presentation, echocardiographic features, management and outcome of this condition. **Methods:** Retrospective review of cases diagnosed within a 1-year period from 1 July 1998 to 30 June 1999. **Results:** Eight cases of neonatal ductus arteriosus aneurysm (DAA) were diagnosed from 1 July 1998 to 30 June 1999. There were 998 new neonatal echocardiograms done during this period, giving us a case incidence of 0.8 per 100 echocardiograms. Only 1 patient was diagnosed antenatally, all others were detected incidentally on echocardiography done for other indications. None had symptoms related directly to the DAA and there was no suggestive mediastinal mass on chest X-ray. Majority were term infants (88%) and there was a predominance of male infants (75%). Three were associated with patent ductus arteriosus (PDA), while in the others, the ductus arteriosus were non-patent at first echocardiography. In the 3 infants whose ductus arteriosus was patent, the PDA was inserted into the pulmonary artery from an unusually superior direction. Close serial echocardiography on some of our patients suggested that resolution of the aneurysm is by thrombosis, manifesting as an echogenic area, followed by regression. In our series, 7 had total resolution, while 1 patient had a persistent echogenic area which became progressively smaller. **Conclusions:** We found ductus arteriosus aneurysms in 0.8% of our neonatal echocardiograms. An unusually superior insertion of PDA into the pulmonary artery is a marker for its presence. Asymptomatic aneurysms resolve spontaneously and should be managed expectantly. Thrombosis plays a part in the resolution of ductal aneurysm.

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Key words: Congenital heart disease, Ductus arteriosus, Echocardiography, PDA, Thrombosis

Introduction

Previously thought to be a rare condition, aneurysms of the ductus arteriosus have been increasingly reported in the medical literature over the past few years. It has been noted as an incidental finding in autopsy, echocardiography and angiography, but can present with symptoms of cough, dyspnoea, hoarseness of voice or as an asymptomatic mediastinal mass on chest X-ray (CXR). Increasingly, ductus arteriosus aneurysms (DAA) are being diagnosed in asymptomatic population due to increased utilisation and sensitivity of echocardiography.

The pathogenesis and natural history of DAA is still unclear, and experience in the management and outcome of such cases are limited. This is especially so in the neonatal age group. The largest case series of neonatal DAA was described by Acherman et al,¹ in which they reported their experience with 11 neonatal DAA over a period of 4 years. In this case series, we report on our experience with

neonatal DAA over a 1-year period from July 1998 to July 1999. This is the largest number of neonatal ductal aneurysms reported within a period of 1 year.

Case Reports

We report on 8 cases of ductus arteriosus aneurysm that were picked up by two-dimensional echocardiography during a 1-year period from July 1998 to July 1999. Their clinical characteristics are summarised in Table I. We define aneurysm of the ductus arteriosus as a segmental dilatation of at least 1.5 times that of adjacent segments in a patent ductus arteriosus, or in cases in which the ductus was not patent, as a remnant ductus manifesting as a fusiform or tubular out-pouching from the aortic end. There were 998 new neonatal echocardiograms done during this period, giving us a case incidence of 0.8 cases per 100 neonatal echocardiograms. The main indication for echocardiography was for assessment of cardiac murmur

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TABLE I: CLINICAL CHARACTERISTICS OF THE 8 PATIENTS WITH DUCTUS ARTERIOSUS ANEURYSM

Case	Sex	Gestational age (weeks)	Birth weight (g)	Apgar (1 min, 5 min)	Antenatal diagnosis	Maternal problem	Indication for 2D-echocardiogram	Patency of ductus arteriosus	Ductal aneurysm (mm)
1	M	38	2550	9, 9	No	PIH	Respiratory distress	Yes	18.0 x 7.0
2	M	38	3160	9, 9	Yes	Nil	Antenatal ultrasound	Yes	17.0 x 6.9
3	F	33	2430	9, 9	No	IDM	Prematurity	Yes	7.0 x 7.0
4	M	37	4340	9, 9	No	Nil	Family history of CHD	No	8.0 x 6.5
5	M	39	3635	9, 9	No	Nil	Family history of CHD	No	17.0 x 9.0
6	F	37	2235	9, 9	No	Nil	Cardiac murmur	No	5.9 x 3.8
7	M	38	3100	9, 9	No	Nil	Cardiac murmur	No	7.6 x 4.6
8	M	39	3530	9, 9	No	Nil	Atrial ectopics	No	7.0 x 4.5

CHD: congenital heart disease; IDM: insulin-dependent diabetes mellitus; PIH: pregnancy-induced hypertension

detected during routine neonatal screening. Although all of these neonates had routine antenatal ultrasounds done, only 1 had an antenatal diagnosis of abnormal patent ductus arteriosus. One of the neonates was premature at 33 weeks gestation while the others were term babies. The male to female ratio was 3:1. In 3 of the cases, the ductus arteriosus were patent at diagnosis, whereas in the other 5 cases, the ductus arteriosus was non-patent. All of them were asymptomatic with regards to the DAA and none had any of the complications that were described in previous reports. The majority of our patients (7 out of 8) had complete resolution of the ductus arteriosus aneurysm by 3 months of age.

Case 1

The baby presented at birth with respiratory distress and was ventilated for 2 days. The mother had mild hypertension antenatally and antenatal ultrasounds were normal. Chest X-ray (CXR) showed homogeneous haziness of both lung fields consistent with congenital pneumonia; there was no mediastinal mass seen. Echocardiogram, which was done on day 1 of life to exclude cyanotic congenital heart disease, revealed a structurally normal heart and a patent ductus arteriosus with superior insertion into the pulmonary artery (Fig. 1). There was a large fusiform aneurysm of the ductus arteriosus measuring 18 x 7 mm, located anterior to the proximal descending aorta and posterior to the main pulmonary artery (Figs. 2 and 3). The PDA flow was left to right. In view of large ductal flow, he was given a course of indomethacin (0.2 mg/kg/dose 12 hourly x 3 doses). Repeat echocardiography at day 3 of life documented closure of the pulmonary end of the duct with thrombus (seen as an echogenic area) partially filling up the aneurysm (Fig. 4); the aortic end of the duct leading into the aneurysm remained open. The child subsequently made good recovery and had no symptoms related directly to the DAA. Serial echocardiograms showed complete thrombosis and total occlusion of the DAA; echocardiogram done

at 6 weeks of life showed complete resolution of the echogenic area.

Case 2

This is a term baby who was clinically asymptomatic. Antenatal ultrasound done at 34 weeks gestation reported as “ductal arteriosus larger than usual” and post-natal 2D-echocardiogram done at day 4 of life showed an aneurysm of a patent ductus arteriosus measuring 17 x 6.9 mm. The duct was patent, with continuous left to right flow, the aortic end measured 4 mm while the pulmonary end measured 2 mm. From the usual high parasternal “ductal” view, the ductus arteriosus was visualised to be inserted into the pulmonary artery from an unusually superior direction. Serial 2D-echocardiogram showed closure of the pulmonary end of the duct followed by obliteration of the aneurysm by thrombosis; slow regression of this echogenic area was documented on serial echocardiograms. At review at 6 months of age, there was only a small remnant echogenic area left.

Case 3

Born premature at 33 weeks gestation to a 35-year-old mother who had insulin-dependent diabetes mellitus, the baby had mild respiratory distress syndrome and required continuous positive airway pressure (CPAP) support for the first 4 days of her life. Echocardiography at day 9 of life showed a small atrial septal defect and an aneurysm of a patent ductus arteriosus. The duct measured 1.4 mm at the pulmonary end with a fusiform dilatation of 7 mm diameter along the course of the ductus. The flow across the PDA was left to right. Repeat echocardiography at day 22 of life showed complete resolution of the DAA.

Cases 4 and 5

Both were term infants delivered via emergency LSCS for cephalopelvic disproportion. Both were asymptomatic and had normal cardiovascular examination clinically.

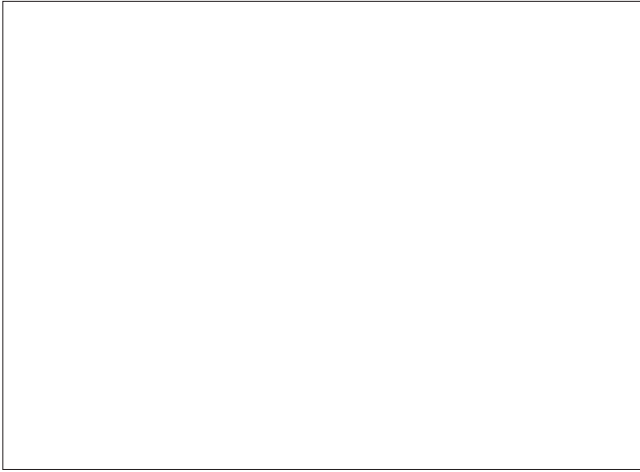


Fig. 1. Superior insertion of the patent arteriosus into the pulmonary artery (high parasternal "ductus" view). LPA: left pulmonary artery; PDA: patent ductus arteriosus; RPA: right pulmonary artery

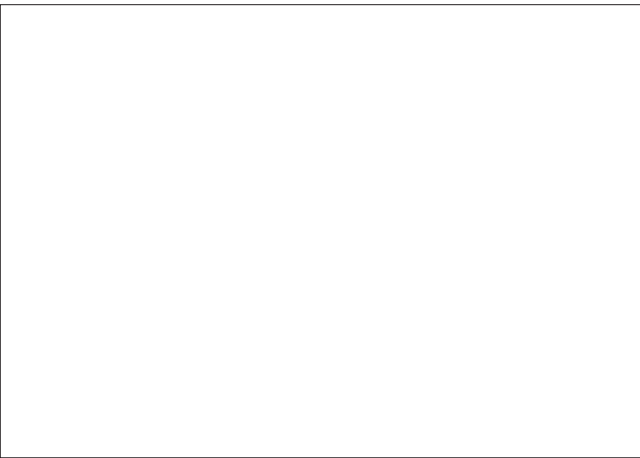


Fig. 3. Aneurysm of the ductus arteriosus (high parasternal view). The descending aorta is in a plane just posterior to this and thus not visualise here. DAA: ductus arteriosus aneurysm; LPA: left pulmonary artery; MPA: main pulmonary artery

Echocardiography was done because of a history of cyanotic congenital heart disease in their previous siblings. The echocardiographic examinations were normal except for the presence of an aneurysm of the non-patent ductus arteriosus, measuring 8.0 x 6.5 mm (tubular) and 17.0 x 9.0 mm (fusiform) (Fig. 5), respectively.

Cases 6 and 7

These 2 term neonates presented with a soft ejection systolic murmur clinically consistent with a functional murmur; they were otherwise well. Echocardiograms done at day 7 of life showed aneurysm of the non-patent ductus arteriosus, measuring 5.9 x 3.8 mm and 7.6 x 4.6 mm, respectively. Both were tubular in morphology.

Case 8

This is a term neonate who had echocardiography done as part of a workup for multiple atrial ectopics. He was

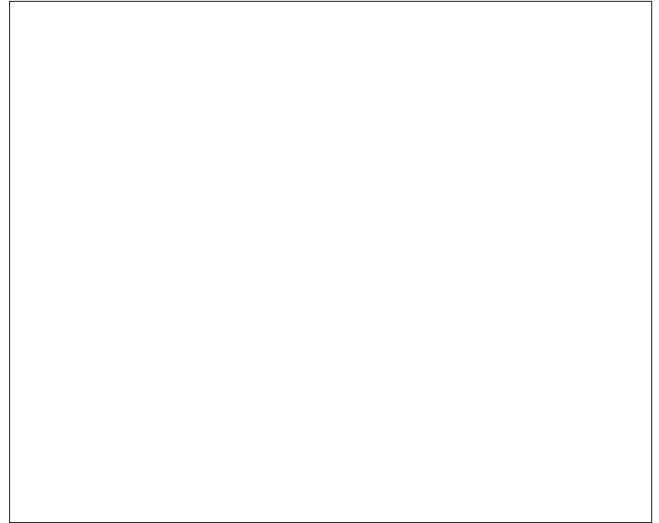


Fig. 2. Aneurysm of the ductus arteriosus (illustration). The ductus arteriosus aneurysm (DAA) is situated posterior to the main pulmonary artery (MPA) and anterior to the aorta (Ao).

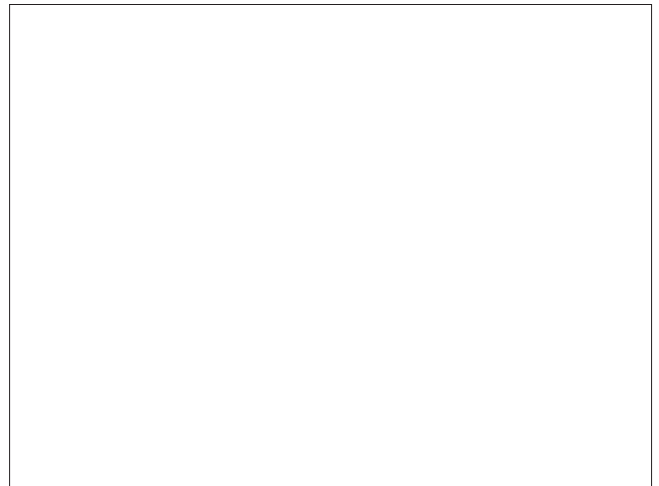


Fig. 4. Echocardiogram showing the arch of the aorta (Ao) and a partially thrombosed aneurysm of the ductus arteriosus (*).

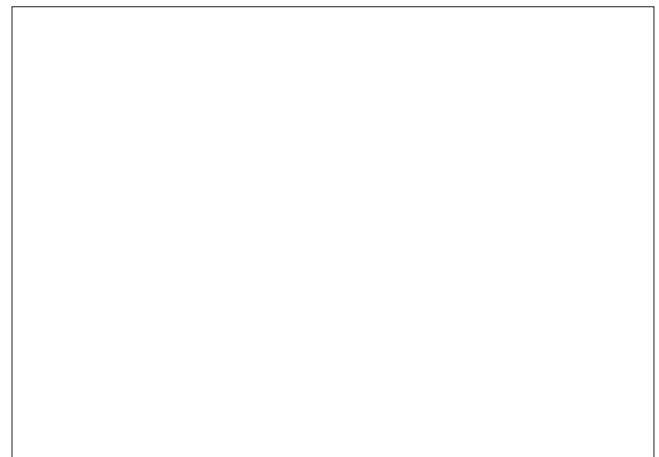


Fig. 5. Aneurysm of the non-patent ductus arteriosus. Suprasternal view showing the descending aorta (DAo) and the aneurysm (DAA) communicating with the aorta. The tissue bar in the aneurysm seen (*) is a cut through a fold of the wall of the aneurysm.

otherwise asymptomatic. Echocardiogram showed structurally normal heart and a tubular aneurysm of the non-patent ductus arteriosus measuring 7 x 4.5 mm.

In cases 4 to 8, they each had an echocardiogram done at diagnosis; they were then reviewed in the outpatient clinic and had a repeat echocardiogram done at 3 months of life, which documented complete resolution of the DAA in each of them.

Discussion

Aneurysms of the ductus arteriosus are described in two groups of patients: in young infants <2 months old, and in adults. Until recently, there are only sporadic case reports, and these are on cases that presented with complications arising from the aneurysm. The prevalence and incidence of DAA in neonates are unclear and are probably underestimated. Two previous neonatal autopsy reports indicated a case incidence of 0.5% to 1%.^{2,3} In adults, frequency of asymptomatic DAA as high as 9% of 103 consecutive aortograms had been reported.⁴ In this study, we found the occurrence of DAA in 0.8% of 998 consecutive new neonatal echocardiograms; this is similar to results of the 2 previous autopsy studies.^{2,3} So far, there has not been any echocardiographic case incidence reported in the literature. We do not perform echocardiography on all newborns delivered in the hospital. Thus, we were unable to give the incidence of ductal aneurysm in terms of per 100 or per 1000 newborns. With improvement in diagnostic methods, DAA will probably be recognised as being more common than previously thought in the general population. Knowledge of such an entity is important in older children and adults as it can be confused with aortic rupture in angiogram done for investigation of thoracic trauma;^{4,5} traumatic aortic rupture can also occur in patients with previously undetected DAA.^{6,7}

The exact pathogenesis of DAA is still debated. It has been reported in association with various conditions in case reports: maternal gestational diabetes, neonatal hypoxia, connective tissue diseases such as Marfan syndrome and Ehlers-Danlos syndrome, Larsen syndrome, interrupted aortic arch and bicuspid aortic valve with dilatation of ascending aorta.⁸⁻¹⁶ Use of medications such as dopamine, dobutamine and prostaglandin E1 has also been suggested to contribute to the development of DAA.^{17,18} In our case series, all except 2 mothers were antenatally well: 1 had insulin-dependent diabetes mellitus and the other had mild pregnancy-induced hypertension; there is no evidence for causal relationship to DAA in this data. Although association of diabetes mellitus with ductal aneurysm has been reported,^{1,8,9} we feel that this relation is casual rather than causal, since diabetes mellitus is a common antenatal problem. None of the babies in our series had inotropic

support or PGE₁ infusion.

Theories proposed to explain the pathogenesis of DAA include:

- a) delay in closure of the aortic end of the ductus arteriosus with subsequent exposure to system arterial pressure;⁸
- b) abnormality of the arterial wall promotes formation of DAA;¹⁹
- c) DAA representing post-stenotic dilatation of the ductus due to turbulent flow through a stenotic segment at its pulmonary artery end during fetal life;¹
- d) remnant of partially involuted right dorsal aortic root.²⁰

The second theory would explain the association of DAA with connective tissue diseases. The third and fourth theories would be consistent with the fact that DAA has been diagnosed antenatally^{1,9,21} and that majority of DAA are diagnosed soon after birth. In our patients, 1 had antenatal evidence of ductal abnormality and all of them were diagnosed within the first few days of life (mean 4 days, range 1 to 9 days). In fact, in 2 cases whose antenatal ultrasounds were normal, echocardiograms were performed just a few hours after birth with the documentation of occurrence of DAA. Antenatal ultrasounds are performed at 22 to 24 weeks and again at 32 to 34 weeks of gestation at our centre. This would suggest that neonatal ductal aneurysm is a congenital lesion which develops *in utero*. None of our cases was syndromic clinically and all of them had structurally normal heart (apart from the DAA and PDA, and a small ASD in 1 case), with no echocardiographic evidence of main or branch pulmonary artery stenosis.

Falcone et al²² have proposed an anatomic classification of ductus arteriosus aneurysms into patent and non-patent types, with the latter sub-classified according to their morphology into fusiform, tubular and dissecting variants. In our series, 3 DAAs were patent, 1 was non-patent fusiform type and the other 4 were of the non-patent tubular variety. In our cases in which the ductus arteriosus were patent at diagnosis of DAA, it was interesting to note that the insertion of ductus arteriosus into the pulmonary artery was from a more superior direction compared to the usual insertion of PDA. This unusual insertion of PDA into the pulmonary artery should serve as an indication to search for the presence of a ductal aneurysm. In their case series, Acherman et al also noted a horizontal ductal jet in some of their patients with ductal aneurysm.¹ This observation can be explained by the ductal aneurysm expanding superior to the bifurcation of the pulmonary artery; the pulmonary end of the ductus arteriosus is thus seen to come from a more superior direction than usual (Fig. 2).

The DAAs were incidental findings in all of our cases, as none of them has any symptoms or signs directly related to the aneurysms. Reported complications of spontaneous

DAA include rupture, dissection, thromboembolism, infection, bronchial obstruction, acute lung collapse, recurrent laryngeal and phrenic nerve palsies, and pulmonary artery obstruction.^{1,12,23-31} In their review of literature, Lund et al³² found a complication rate of 31% in spontaneous DAA in infants younger than 2 months of age with an operative mortality of 2 out of 8 cases. This is probably an overestimation due to selection bias, since the cases reported previously were mainly those with symptoms or had presented with complications. In the series by Acherman et al¹ published recently, 8 of 11 aneurysms resolved completely without complication, while the other 3 underwent surgical resection (1 due to failure to regress, 2 due to thrombosis). In our experience, the aneurysms regressed without complication on follow-up. In 2 of our patients who had very close serial echocardiographic observations, thrombi were observed to form in the aneurysms following closure of the pulmonary end of the ductus arteriosus. The thrombi occluded the aneurysms, manifesting as highly echogenic areas at the site of the aneurysm. These areas were then observed to undergo gradual regression in size on serial echocardiography. There was no evidence of embolic phenomenon arising from the thrombi in these 2 cases. This suggests that resolution of DAA is by thrombosis, at least in some cases. The observation of thrombus formation within the ductal aneurysm by itself should not be an indication for surgical intervention.

From our series and that of Acherman et al,¹ we conclude that asymptomatic neonatal DAAs are likely to remain so, and the majority of them resolve spontaneously. We therefore recommend conservative management in asymptomatic DAA, with regular echocardiographic follow-up for resolution. Complete resolution of the majority of ductal aneurysms is expected by 3 months of age.

The fact that most neonatal DAAs regress completely also brings up an interesting question – are the DAAs found in older children and adults those that did not regress in the neonatal period, or do they arise *de novo* via a different pathogenesis?

Conclusion

Aneurysm of the ductus arteriosus is not a rare entity, occurring in 0.8% of our neonatal echocardiograms. A superior or horizontal insertion of PDA into the pulmonary artery from a superior direction may indicate the presence of a ductal aneurysm. Our series of consecutive patients shows that asymptomatic ductus arteriosus aneurysms regress spontaneously and can be managed conservatively. Serial echocardiograms showed that thrombosis plays a part in the resolution of ductus arteriosus aneurysms.

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