Prenatal Sonographic Diagnosis of Congenital Ductus Arteriosus Aneurysm: A Case Report

Fuanglada Tongprasert, MD*,
Theera Tongsong, MD*, Rekwan Sittiwangkul, MD**

** Department of Obstetrics and Gynecology, Faculty of Medicine, Chiang Mai University, Chiang Mai*
*** Department of Pediatrics, Faculty of Medicine, Chiang Mai University, Chiang Mai*

The objective of this report was to demonstrate prenatal sonographic features of congenital ductus arteriosus aneurysm (DAA), a rare, but possibly fatal abnormality. It is characterized by a saccular or fusiform dilatation of the ductus arteriosus. The majority of affected neonates are clinically asymptomatic and tend to progressively diminish in size of DAA and spontaneous closure. However, serious complications can occur during waiting periods, including spontaneous rupture or thromboembolism. Case: A 35 year-old Thai woman, G2P1, underwent ultrasound examination at 34 weeks' gestation. Fetal echocardiography revealed markedly enlarged and tortuous ductus arteriosus with saccular dilation at the distal end, just before joining the descending aorta. The cross-sectional diameter of the saccular portion of the ductus arteriosus was 10 mm. The active female baby was vaginally delivered at 34 weeks' gestation, weighing 1050 grams. Postnatal echocardiography confirmed the prenatal findings. The DAA was gradually decreased in size and finally spontaneously closed without medical or surgical intervention.

Keywords: *Ductus arteriosus aneurysm, Echocardiography, Ultrasound*

J Med Assoc Thai 2005; 88(4): 541-4
Full text. e-Journal: *http://www.medassocthai.org/journal*

Congenital ductus arteriosus aneurysm (DAA) is a rare, but possibly fatal abnormality. It is characterized by a saccular or fusiform dilatation and elongation of ductus arteriosus[1]. Congenital DAA may be identified in infants, children and adults, and recently it has been recognized in the fetus by echocardiography[2,3]. Although the true incidence of congenital DAA is still unknown, recent reports suggest it may be much more common among fetuses and neonates than previously thought[4]. DAA likely develops in the third trimester rather than in the first or second trimester perhaps due to abnormal intimal cushion formation or elastin expression[2]. A late onset of prenatal development, perhaps due to an altered circulation, weakening of the wall of the ductus arteriosus or a combination of both. It is during the third trimester that the arterial blood pressure and cardiac output are greatest. In addition, it may be more commonly observed in patients with connective tissue disease. Infants with large for their gestational age, poorly controlled diabetic mothers and mothers with blood group A also have a high risk of DAA[4]. The majority of affected fetuses are asymptomatic at birth, however some reports suggest that symptomatic congenital DAA may be associated with serious complications[2]. The objective of the present report was to demonstrate prenatal features of congenital ductus arteriosus aneurysm detected by fetal echocardiography.

Correspondence to : Tongprasert F, Department of Obstetrics and Gynecology, Faculty of Medicine, Chiang Mai University, Chiang Mai 50200, Thailand.

Case Report

A 35 year-old Thai woman, G2P1, attended the antenatal care clinic at 34 weeks' gestation. Her previous pregnancy had been uneventful. The past surgical or medical history was unremarkable except for well-controlled chronic hypertension. Pregnancy-aggravated hypertension was clinically diagnosed. Ultrasound examination was requested due to small-for-date uterine size and for anomaly screening. Fetal biometry suggested growth restriction. Fetal echocardiography revealed normal cardiac structures on

four-chamber view but abnormal great vessels on the long-axis view. The pulmonary trunk was markedly greater than the ascending aorta. Ductus arteriosus aneurysm (DAA) was incidentally visualized, appearing as a cystic mass at the left upper chest, which proved to be vascular on color flow mapping. The ductus arteriosus was markedly dilated and tortuous with a saccular dilation at the distal end before joining the descending aorta (Fig. 1-4). The DAA was protruded leftward and cephalic to the aortic arch. The cross-sectional diameter of the saccular portion of the ductus arteriosus was 10 mm. Color flow mapping showed dilated and tortuous ductus arteriosus (DA) and reversed flow in aortic arch during diastole (Fig. 4). In addition, minimal pericardial effusion and mild cardiomegaly are also demonstrated. After stabilization with $MgSO_4$, induction of labor was performed, resulting in vaginal delivery. The female infant was growth-restricted, weighing 1050 grams. Apgar scores were 8 and 8 at 1 and 5 minutes, respectively. The immediate postnatal period was unremarkable. Echocardiogram performed by a pediatric cardiologist several hours after birth confirmed the prenatal findings. However, DAA was spontaneously decreased in size due to hemodynamic change of normal postnatal circulation. The moderate size of patent ductus arteriosus aneurysm was gradually decreased on subsequent serial echocardiographic examinations. Finally, spontaneous regression and closure of DAA without complications were observed without any medical or surgical intervention.

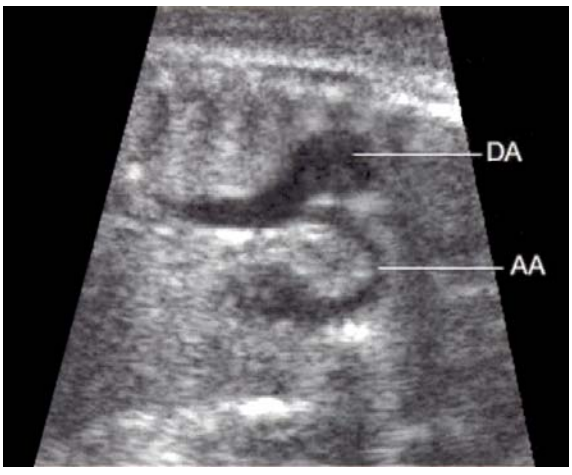

Fig. 1 Sagittal scan of the fetal thorax shows smaller aortic arch (AA) and saccular portion of ductus arteriosus (DA), close to the junction with aortic arch becoming descending aorta

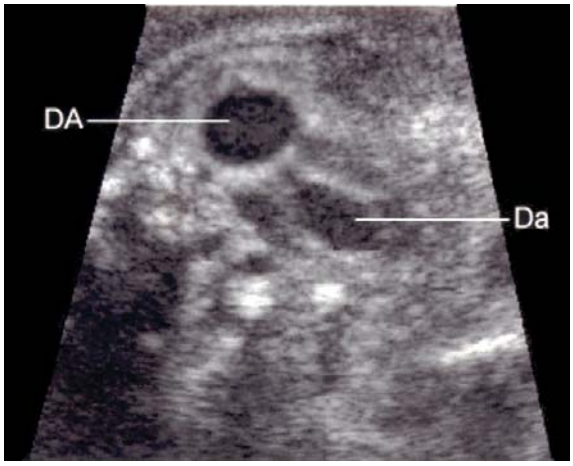

Fig. 2 Cross-sectional scan at the upper thorax shows the greatest dimension of the ductus aneurysm (DA), (Da; ductal arch)

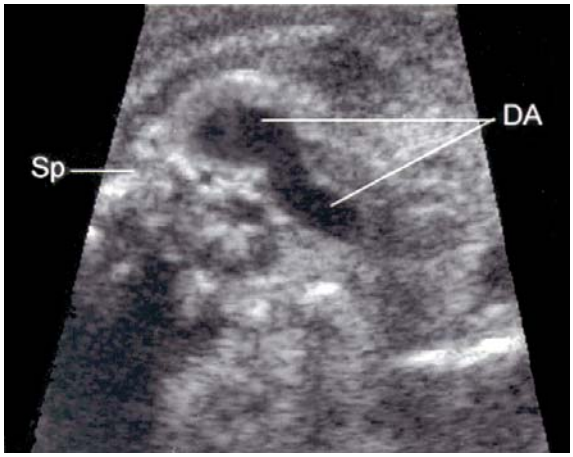

Fig. 3 Long-axis view of the great arteries shows dilated and tortuous ductal arch (DA), (Sp; spine)

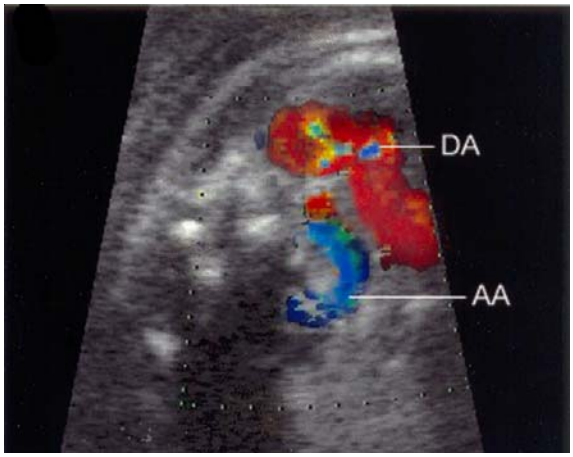

Fig. 4 Color flow mapping shows dilated and tortuous ductus arteriosus (DA) and reversed flow in aortic arch (AA)

Discussion

Congenital ductus arteriosus aneurysm has been considered a rare cardiovascular lesion. It can be diagnosed either before or after birth. Although the incidence of neonatal DAA was previously reported to be 0.8% upon neonatal autopsies, the true incidence is unclear and varied, depending on the criteria of diagnosis. For example, it was observed in 1.5% by fetal ultrasound imaging at the fetal age of more than 30 weeks, and interestingly, possibly as high as 8.8% of full-term neonate[2,4]. However, the rather large aneurysm like in the case presented here is probably rare. DAA has been uniformally identified in the third trimester and has not been detected earlier in gestation, in spite of widespread use of routine ultrasound screening. This suggests a late onset of prenatal development perhaps may be the result of abnormal intimal cushion formation or defective elastin in ductus arteriosus[2]. DAA may be observed in the patients with connective tissue diseases such as Marfan, Ehler-Danlos and Larsen syndromes. Previous reports showed the association of trisomy 21, 13 or Smith-Lemli-Opitz syndrome with DAA[2]. High risk factors associated with DAA include newborns with large for gestational age, maternal DM and mothers with blood group A[4]. However, there does not appear to be any typical association of DAA with extracardiac malformations.

The mean DAA diameter measured in a previous report was 12.6 ± 4.1 mm. with a range of 8 to 24 mm[1]. The natural history of the smaller DAA was benign in the majority of cases, with 70% demonstrating progressive regression in size of DAA followed by spontaneous closure[4].

Although the majority of affected neonates are clinically asymptomatic, several reports demonstrated several potential serious complications including spontaneous rupture, thromboembolism, erosion into airways, infection, and compression of surrounding vessels, airways and nerves (particularly the recurrent laryngeal nerve)[2]. It may also be associated with chromosomal anomalies and, more importantly, connective tissue disorders which may be progressive and place the infant at higher risk of spontaneous aneurysm rupture[5]. To avoid such lethal complications, surgical intervention may be considered if 1) the ductus arteriosus with DAA remains patent beyond the neonatal period, 2) the DAA is associated with connective tissue disease, 3) there is evidence of thrombus extension into other vessels or thromboembolism or 4) there is significant compression of adjacent structures[1,2,4]. Although it does not appear to warrant intervention in the majority of cases, it would be wise to document complete closure of the aneurysmal duct prior to neonatal discharge. Like most cases, the case presented here was asymptomatic, closed spontaneously and did not required treatment.

In addition to surgical considerations, close follow-up of the affected neonate is important because the majority of cases may spontaneously deminute after a period of time[4]. The use of indomethacin has been reported to close DAA successfully in one case report, however the definitive effect of it on DAA is uncertain[6].

In summary, increased use of fetal echocardiography has detected more cases of congenital DAA. The majority of affected neonates are clinically asymptomatic and tend to progressively diminish in size of DAA and spontaneous closure, especially in cases of smaller DAA. However, serious complications can occur during waiting periods indicating surgical intervention, especially in cases of persisting DAA beyond the neonatal period or associated connective tissue diseases. Finally, indomethacin treatment may be used to successfully close DAA in premature infants even though its definitive effect on DAA is still uncertain.

References

1. Hornberger LK. Congenital ductus arteriosus aneurysm. J Am Coll Cardiol 2002; 39: 348-50.
2. Dyamenahalli U, Smallhorn JF, Geva T, Fouron JC, Cairns P, Jutras L, et al. Isolated ductus arteriosus aneurysm in the fetus and infant: a multi-institutional experience. J Am Coll Cardiol 2000; 36: 262-9.
3. Puder KS, Sherer DM, Ross RD, Silva ML, King ME, Treadwell MC, et al. Prenatal ultrasonographic diagnosis of ductus arteriosus aneurysm with spontaneous neonatal closure. Ultrasound Obstet Gynecol 1995; 5: 342-5.
4. Jan SL, Hwang B, Fu YC, Chai JW, Chi CS. Isolated neonatal ductus arteriosus aneurysm. J Am Coll Cardiol 2002; 39: 342-7.
5. Leal SD, Cavalle-Garrido T, Ryan G, Farine D, Heilbut M, Smallhorn JF. Isolated ductal closure in utero diagnosed by fetal echocardiography. Am J Perinatol 1997; 14: 205-10.
6. Lee WJ, Chen SJ, Wu MH, Li YW. Regression of ductus arteriosus aneurysm in a neonate demonstrated by three-dimensional computed tomography. Int J Cardiol 1999; 68: 231-4.

การวินิจฉัยก่อนคลอดด้วยภาพคลื่นเสียงความถี่สูงของภาวะเส้นเลือด ductus arteriosus โป่งพอง

เฟื่องลดา ทองประเสริฐ, ธีระ ทองสง, แรกขวัญ สิทธิวางค์กูล

วัตถุประสงค์ของรายงานนี้เพื่อจะแสดงลักษณะทางคลื่นเสียงความถี่สูงก่อนคลอดของภาวะเส้นเลือด ductus arteriosus โป่งพอง ซึ่งเป็นภาวะที่พบได้น้อย แต่อาจมีอันตรายรุนแรงได้ ในภาวะนี้เส้นเลือด ductus arteriosus จะขยายเป็นถุง หรือ เป็นท่อคดเคี้ยว ส่วนมากทารกแรกคลอดจะไม่มีอาการ แต่มีแนวโน้มลดขนาดลง และปิดไปได้เองในที่สุด แต่อย่างไรก็ตามบางรายอาจมีภาวะแทรกซ้อนรุนแรง เช่น แตก หรือ มีก้อนเลือดอุดตัน

***กรณีศึกษา**: หญิงไทยคู่ อายุ 35 ปี ครรภ์ที่สอง ได้รับการตรวจคลื่นเสียงความถี่สูงก่อนคลอดขณะอายุครรภ์ 34 สัปดาห์ คลื่นเสียงความถี่สูงของหัวใจพบว่าเส้นเลือด ductus arteriosus ขยายขนาดใหญ่และคดเคี้ยว และมีการเปลี่ยนแปลงเป็นถุงน้ำขนาดใหญ่ มีเส้นผ่าศูนย์กลางประมาณ 10 มม. แต่ไม่พบมีความผิดปกติของโครงสร้างอื่น ๆ ทารกคลอดขณะอายุครรภ์ 34 สัปดาห์ คลอดปกติทางช่องคลอด ทารกเพศหญิง น้ำหนัก 1,050 กรัม แข็งแรงดี การตรวจคลื่นเสียงความถี่สูงของหัวใจ พบลักษณะเช่นเดียวกับที่ตรวจพบก่อนคลอด แต่ขนาดเล็กลง ซึ่งเมื่อตรวจซ้ำเป็นระยะ ๆ พบว่ามีขนาดเล็กลงเรื่อย ๆ และปิดได้เองในที่สุด โดยไม่ต้องผ่าตัดหรือให้ยารักษา*