Prenatal Diagnosis of Isolated Ductus Arteriosus Aneurysms with Spontaneous Neonatal Closure: Case Series with Systematic Review of the Literature and Video-Echocardiography Demonstration

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ABSTRACT
A two patient case series of ductus arteriosus aneurysm (DAA) is presented with a systematic review of the literature for consensus of management and video echocardiography demonstrating diagnosis in utero and closure postnatally. To our knowledge, this is the first publication of time-lapse video echocardiography demonstrating the gradual, but spontaneous closure of PDA.

Key Words: ductus arteriosus, aneurysm, neonate, echocardiography

INTRODUCTION
Congenital ductus arteriosus aneurysm (DAA) is a localized saccular or tubular dilation of the ductus arteriosus. The etiology and pathogenesis of DAA is still elusive. The impact of DAA on ongoing pregnancy and postnatal outcome is not well understood. Here we report two cases of isolated congenital DAA; both were diagnosed in the third trimester of gestation and both had spontaneous resolution with no complications as neonates. A targeted systematic review of the literature is included for review of the consensus on DAA management.

CASE 1
The patient was a 36 year old white female, gravida 1, para 0 at 35 weeks’ estimated gestational age and she was referred for a fetal echocardiogram to evaluate for coarctation of the aorta. The fetal echocardiogram of the ductal and aortic arches demonstrated an isolated congenital DAA, measured at 9 mm. (Figure 1) There was mild enlargement of the right ventricle with normal biventricular function. A male infant was born at full term by cesarean section due to fetal distress. The infant was observed in the NICU where he was treated for mild pulmonary hypertension; both DAA and PDA were spontaneously closed on postnatal echocardiography. (Figure 2) The
Spontaneous Closure of Ductus Arteriosus Aneurysm

**Figure 1:** A third trimester fetal echocardiogram revealing the ductal and aortic arches at the time of DAA diagnosis. The aortic arch is revealed with head and neck vessels coming off of the arch, with the aneurysm appearing at the twelve-o’clock position like a sack. The “candy cane” and aneurysmal structures are most evident on the 2-D portion of the echo. The ductal arch comes into view midway into the video and, in contrast the aortic arch, it appears tortuous, elongated, is oriented towards the twelve o’clock position and resembles a “hockey stick”. Color flow-mapping demonstrates swirling of blood within the aneurysm. The video is looped to facilitate viewing. [Editor’s Note: movies can only be viewed in the HTML version of this manuscript, please go to http://www.neonatologyresearch.com/?page_id=2134 to view this movie.]

**Figure 2:** A Transthoracic Echocardiogram (TTE) using short axis (or PDA) view revealing the course of spontaneous PDA closure. A large PDA is shown on the far right in a “three finger” view in the middle of the screen, shortly after birth. It includes the left pulmonary artery, the right pulmonary artery and the ductus. This portion of the video demonstrates a smaller PDA with left to right shunt (indicated by the red color). This is followed by a subsequent TTE on the subsequent day of life demonstrating what is now a “two finger” view after ductal closure. This portion of the video is set to the right, where the predominance of blue flow and the loss of the third and right most “finger” indicates loss of the PDA shunt. The video is looped to facilitate viewing. [Editor’s Note: movies can only be viewed in the HTML version of this manuscript, please go to http://www.neonatologyresearch.com/?page_id=2134 to view this movie.]
infant was discharged home and followed in clinic without complication.

**CASE 2**

The patient was a 22 year old black female, gravida 5, para 4, at 36 weeks’ gestation. She was referred for fetal echocardiogram due to possible “right heart enlargement” by obstetric ultrasound. The ductal and aortic arch views of the fetal echocardiogram revealed a saccular dilation, tortuous course and elongation of the DAA, measured at 6.5 mm (not shown). A male infant was born at full term; his postnatal echo revealed a small PDA, which was spontaneously closed on his follow-up visit at 4 weeks of age. There have been no complications on follow-up visit.

**SYSTEMATIC REVIEW OF THE LITERATURE**

A systematic search of PubMed was made using the search words: ductus arteriosus, aneurysm and neonate and limiting the fields to all English studies greater than 10 patients and after 1997 (the post surfactant era). All such manuscripts were reviewed and a consensus of the literature tabulated for immediate postnatal management (not shown). There was essentially no conflict about the immediate postnatal management of DAA in all studies of greater than ten patients in the post surfactant era. Close observation, with expectation of spontaneous closure in the immediate postnatal period is the consensus method of management. Recommendations for long term cardiac follow up varied but were generally more emphasized if a genetic syndrome (such as a connective tissue disorder) was also diagnosed because of the life-long risk of great vessel aneurysm.

**DISCUSSION**

Although it is rare, the incidence of DAA with fetal echocardiography at third trimester ranges from 1.5 to 2.2%. In a prospective study of 548 full-term neonates, 48 infants (8.8%) were found with DAA. Although no standard criteria were given for the diagnosis of DAA, the typical findings demonstrate a tortuous, dilated vascular structure that protrudes leftward of the aortic arch and the “swirling of blood” within the aneurysm. Jan et al included these cases with DAA measured at 6.5 mm to 11.2 mm; Dyamnalahalli reported 24 cases with DAA measured at 8 to 24 mm. Since the vast majority of the affected infants have no symptoms, without echocardiography, occult DAAs may have gone undetected. However, severe complications with DAA were described in the postnatal period, including spontaneous rupture, thromboembolic disorders, compressions to the neighboring tissues and even sudden death. The larger DAA appears more commonly associated with complications. Spontaneous PDA closure occurred in 70.2% of the newborns during the first 72 hours and all had spontaneous DAA resolution by 7 to 35 days of life. It has been proposed, therefore, that the need for surgical resection should be considered only if the DAA remains patent beyond the neonatal period, the DAA is associated with connective tissue disease, the thrombus extends into other vessels, or there is significant compression of adjacent structures.

The DAA in our series of two cases were relatively small and both had spontaneous closure. There was no associated connective tissue disease or complication. It has been postulated that DAA might be a normal variant of the ductal bump and part of the normal process of spontaneous ductal closure in full term neonates. However, given the potential of complications and
association with connective tissue diseases, continued follow-up of affected infants may be warranted.

REFERENCES


