Images in Pediatric Cardiology

An Aberrant Umbilical Vein Draining into the Coronary Sinus

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Case Report

A 31-year-old G1P0 at 20 weeks of gestation presented for evaluation of an abnormal ultrasound. Fetal echocardiogram revealed a large abnormal umbilical vein lateral to the aorta that entered what was believed to be the coronary sinus (Figs. 1 and 2). The right atrium was mildly enlarged, but the remainder of the study was normal. Repeat echocardiograms revealed persistence of the aberrant umbilical vein. Delivery of a full-term female occurred after induction of labor. Initial physical exam was notable for an enlarged umbilicus left of midline in the midsagittal line (Fig. 3B). The child was otherwise well with normal oxygen saturation and discharged home at 48 hours of life. Echocardiogram at birth revealed a structurally normal heart with a very large coronary sinus (Fig. 3A).

Discussion

Umbilical vein anomalies are a rare group of congenital malformations. Mende [2] described an aberrant umbilical vein on autopsy in 1826 that passed externally to the liver and terminated in the right atrium. Aberrant umbilical venous drainage into the inferior vena cava, iliac vein, and superior vena cava via subcutaneous anastomoses (caput medusae) and persistence of the right umbilical vein have also been described [1]. Umbilical vein anomalies can be associated with gastrointestinal, cardiovascular, musculoskeletal, and genitourinary anomalies [1].

Proposed theories of umbilical vein anomalies include derangement in embryology, folic acid deficiency, and teratogens. Paired umbilical veins appear in week 2 of gestation. They initially drain into the sinus venosus along with the paired common cardinal veins and the vitelline veins. As the liver bud develops, the cranial portions of the vitelline veins are incorporated into hepatic sinusoids. At the same time, “critical anastomosis” between the umbilical veins and hepatic sinusoids are formed, terminating the

Fig. 1. Fetal echocardiogram at 6 months of gestation showing drainage of anomalous umbilical vein (asterisks) into coronary sinus in black-and-white (A) and color Doppler (B) images. Arrows show termination of umbilical vein into very enlarged coronary sinus. CS, coronary sinus; LV, left ventricle; RA, right atrium; RV, right ventricle.

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connection between the umbilical veins and sinus venosus. When connections between the umbilical vein and hepatic sinusoids are not successfully formed, blood can be diverted and form an aberrant course [2]. In 1957, Monie [3] reported an association between folic acid deficiency in pregnant Long–Evans rats from days 9 to 11 of gestation and persistence of the right umbilical vein. The right umbilical vein then terminated in the inferior vena cava. Monie and Khemmani [4, 5] reported similar findings after administration of retinoic acid.

This is the first published report of an umbilical vein draining into coronary sinus. The ectopic enlarged umbilicus may be explained by the aberrant course of the umbilical vein that prevented blood from the placenta from being divided between the liver and the heart. With an absent ductus venosus, there is no modulation of blood flow by the constricting sphincter mechanism at the origin of the ductus. Thus, there is an increased volume of blood and greater pressure in the aberrant vein that could possibly account for the increased umbilical size. Fortunately, this anom-

Fig. 2. Coronal color Doppler images of fetal thorax at 6 months of gestation showing entrance (A) and course (B) of anomalous umbilical vein. Thick arrow shows the entrance of umbilical cord to the left of the normal insertion site (double asterisks). Thin arrows show the course of the umbilical vein. Ant., anterior; L, left; Post., posterior; R, right; S, Spine.

Fig. 3. (A) Parasternal long-axis echocardiographic image at 8 hours of life shows a very large coronary sinus. The left atrium and left ventricle are normal size. The pulmonary and systemic venous drainage were otherwise normal and the coronary sinus was not unroofed. (B) Photograph of the patient at the same time as the echocardiogram was obtained shows the large, laterally displaced umbilicus. CS, coronary sinus; LA, left atrium; LV, left ventricle; RV, right ventricle.