June

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Editorial Board
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ASSESSMENT PROGRESS:
Total Questions: 10
Questions Answered: 8
Correct Answers: 3

Question 8

A local pediatrician asks you to "check out a baby" in the newborn nursery. He has been called by
the nurses because the infant had a single umbilical artery. The infant was delivered at 38 weeks'
gestation at a birthweight of 3,200 g. The woman's prenatal course and family history were
normal. Gestational age was confirmed on ultrasonography at 10 weeks and repeat
ultrasonography findings at 24 weeks' were normal. No abnormalities are detected on physical
examination. You ask the resident to consider the association of congenital anomalies with a single
umbilical artery and value of added testing.

Of the following, isolated single umbilical artery HAS BEEN shown to be associated with:

- **A.** Afro-Caribbean heritage
- **B.** cardiovascular anomalies
- **C.** macrosomy
- **D.** occult renal anomalies
- **E.** post-term birth

**Incorrect:**

Correct Answer: D

Single umbilical artery (SUA) often has been described in association with adverse perinatal
outcomes, with morbidity or mortality in individual patients attributed to chromosomal or
structural abnormalities. SUA occurs with persistence of the original artery of the embryonic
body stalk, atrophy of a previously normal umbilical artery, or agenesis of one of the
umbilical arteries.

The incidence of SUA is higher in infants of white women than in infants of African-American,
Afro-Caribbean, or Japanese heritage. Infants of multiple gestations have
higher risks also. Maternal diabetes also increases the risk. When a SUA is
discovered prenatally, significant fetal anomalies are reported in about 20%
of cases, and include cardiovascular, gastrointestinal, renal anomalies—
alone or in combination. Chromosomal abnormalities affect 20% of cases,
with trisomy 18 being the most frequent. Perinatal mortality ranges from
5% to 20%, with two thirds of the deaths occurring in utero. Neonatal
mortality is highly correlated with the presence of multiple anomalies. Because of these
aforementioned factors, the finding of SUA in a fetus is a call to action, which includes a
detailed anatomic survey; fetal echocardiography; fetal karyotyping; and serial evaluation of fetal growth, even in the absence of structural or chromosomal abnormalities.

Of what significance is the finding of an isolated single umbilical artery in an otherwise normal-appearing infant? Of the factors listed, only occult renal anomalies is associated with isolated SUA.

In surveys of records of infants having isolated SUA, significant associations include the following:

- gestational age is lower (39 vs 40 weeks; \( P < .001 \)),
- mean birthweight is lower (3,160 vs 3402 g; \( P < .001 \)),
- prematurity is more common (15% vs 7.4%) as is extreme prematurity (3.9% vs 1.2%; \( P = .019 \))
- intrauterine growth restriction is more prevalent (5.4% vs 1.9%; \( P < .001 \))

When infants having chromosomal or multisystem anomalies are excluded, occult multisystem anomalies have not been reported in association with SUA.

Recent surveys of the literature and case studies have addressed the relationship between isolated SUA and occult or significant renal anomalies. The existing data are confusing and a bit conflicting. In a metaanalysis of 37 studies of SUA over a 40-year period, only seven studies contained data regarding isolated SUA. The investigators found significant renal or urinary abnormalities in 8% of cases, with the most common abnormality, vesicoureteral reflux (VUR) of grade 2 or higher, in 2.9% of cases. Although controls were not available in the studies in the metaanalysis, investigators made estimates from the reported incidence of renal anomalies in the general population and concluded that silent renal anomalies of any severity may be sixfold higher and severe malformations up to be threefold higher among infants with isolated SUA. Because VUR comprised the majority of the severe cases, and because VUR and associated urinary tract infection may precede reflux nephropathy, the authors of the metaanalysis recommended that infants having isolated SUA undergo renal ultrasonography and a micturating cystourethrogram. Renal ultrasonography alone has been associated with a positive predictive value of only 32.5% for suggesting VUR. Follow-up recommendations include having a low threshold for diagnosis of urinary tract infection in these patients.

In another analysis, isolated SUA was found in 129 of 33,067 sequential newborns at a single institution, an incidence of about four cases per 1,000 live births. The data confirmed the association with prematurity and fetal growth restriction. Of the 122 infants who underwent renal ultrasonography, two had clinically significant renal anomalies (absent kidney; unilateral hypoplastic kidney with grade 2 VUR) and three had minor, transient abnormalities. The authors of the study also compared their data with data from the general population. Their data also found that occult renal anomalies occurred more often among infants with isolated SUA (4.1% vs 0.9%; \( P = .005 \)), but no significant differences were found in the prevalence of clinically significant renal anomalies (1.6% vs 0.4%, \( P = .74 \)), albeit the trend suggests the need for more data. The authors concluded that although the overall incidence of renal abnormality may be higher in association with SUA, the risk for significant abnormality is no greater than that in the general population. They believe that the incidence of significant renal anomalies in infants with isolated SUA does not warrant routine postnatal renal imaging. Their conclusion was supported in a second study of 52 cases of isolated SUA over 8 years at a single hospital in Europe. Although 10% of the infants were found to have abnormalities on renal ultrasonography, none was clinically significant and the authors do not recommend routine screening.

Prenatal ultrasonography may play a role in decision making about renal studies in cases of SUA. If the mother had undergone sophisticated ultrasonography, including examination of fetal anatomy, especially if the SUA had been detected and the renal system was well visualized; further imaging may be avoided, as suggested in the vignette. Of note, if screening is desired, delay until the second month after birth may allow transient findings to resolve.

Although there is no consensus regarding screening for renal anomalies in infants having isolated SUA, screening has regularly revealed an increased prevalence of minor renal anomalies. Prenatal screening may play a role in making individual case decisions.

References:
Beali M, Ross M. Umbilical cord complications. Available at: 


Related readings from Neoreviews.org

American Board of Pediatrics Content Specification(s):

07_Water_Salt_Renal: Know how to diagnose specific anatomic abnormalities of the kidneys and urinary tract in infants

13_Skin: Know how to evaluate and manage disorders of the umbilical cord, including granulomas, persistent omphalomesenteric duct remnant, and patent urachus