Lower Urinary Tract Obstruction in the Fetus and Neonate

Douglass B. Clayton, MD*, John W. Brock III, MD

KEYWORDS

• Urethral obstruction • Prenatal diagnosis • Prune-belly syndrome
• Prenatal ultrasonography • Posterior urethral valves • Fetal intervention

KEY POINTS

- Congenital lower urinary tract obstruction in the fetus and neonate is rare but is increasingly identified in the era of prenatal sonography and can result in substantial perinatal mortality with lifelong morbidity.
- Posterior urethral valves, urethral atresia, and prune-belly syndrome are the frequently reported causes of lower urinary tract obstruction, with posterior urethral valves seen most often.
- The management of these diseases in the newborn period requires appropriate urinary tract decompression, subspecialty support, and definitive diagnosis with endoscopy and radiography.
- Despite numerous human series and the recent publication of a randomized trial, the survival benefit in lower urinary tract obstruction afforded by fetal intervention with vesicoamniotic shunt placement remains unclear.

INTRODUCTION

Routine prenatal sonography has made prenatal anomaly detection a reality. Congenital anomalies affect up to 2% of all pregnancies,[1,2] and ultrasound technology has high sensitivity for urologic anomalies, particularly diagnoses involving obstruction. Urologic diagnoses account for 20% of all prenatally identified congenital anomalies.[3] Fetal hydronephrosis is one of the most commonly detected findings, but alone it does not necessarily portend a persistent postnatal obstruction.[1,2,4] Prenatal care providers must rely on additional findings to increase the degree of suspicion for urinary tract obstruction.

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Division of Pediatric Urologic Surgery, Department of Urologic Surgery, Monroe Carrel Jr. Children's Hospital, 2200 Children's Way, 4102 DOT, Nashville, TN 37232, USA
* Corresponding author.
E-mail address: douglass.b.clayton@vanderbilt.edu

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Several genitourinary anomalies with a high propensity for morbidity and mortality are causes of lower urinary tract obstruction (LUTO).[5] **Box 1** lists the causes of LUTO that are discussed in this article. The article focuses on:

- Review of the current literature regarding LUTO in the fetus and newborn
- Clinical aspects of each diagnosis and management strategies
- Prenatal sonographic appearance suggesting LUTO
- Fetal intervention for LUTO

EPIDEMIOLOGY AND INTRODUCTION OF LUTO

Congenital LUTO is a rare phenomenon primarily affecting the male fetus.[6] Epidemiologic data suggest that the birth prevalence of LUTO is stable. In a large population-based study from the United Kingdom, the birth prevalence of postnatally confirmed LUTO was 3.34 per 10,000 live births, with no significant change in birth prevalence occurring between 1995 and 2007.[7] **Table 1** shows the birth prevalence of LUTO diagnoses. A United States report using the kids inpatient database also found no significant changes in the prevalence of prune-belly syndrome (PBS) or posterior urethral valves (PUV) between 1997 and 2009.[8]

PUV

The most common cause of LUTO is PUV, with a birth prevalence of 1 to 2 per 10,000 live male births.[8,9] The male urethra consists of the posterior urethra, which comprises the prostatic and membranous sections, and the anterior urethra, which is distal to the membranous urethra and traverses the entire penis.[10] A posterior urethral valve is an obstructing membrane or mucosal fold within the posterior urethra leading to complete or partial bladder outlet obstruction.[11] Hugh Hampton Young[12] originally described 3 types of PUV in the early 1900s. Type I valves are most common and appear as leaflets that extend in an inferior and anterior direction originating posteriorly from the verumontanum. Type II valves are not obstructive and represent mucosal folds extending superiorly from the verumontanum to the bladder neck. Type III valves

Box 1
Congenital LUTO diagnoses

1. Most common
   a. Posterior urethral valves
   b. Urethral atresia
   c. Prune-belly syndrome
2. Less common
   a. Anterior urethral valves/anterior urethral diverticulum
   b. Congenital megalourethra
   c. Obstructing ureterocele
3. Rare mimics
   a. Isolated megacystis
   b. MMIHS
   c. Megacystis-megaureter association