The Management of Fetal Urinary Tract Obstruction

Zvi Appelman, MD, and Mitchell S. Golbus, MD

University of California at San Francisco
San Francisco, California

The diagnosis of fetal urinary tract obstruction can be made quite easily with the help of modern ultrasonography. Correction of an obstruction is not as easy but is possible with several surgical techniques. One of the main problems is the proper selection of patients, because some fetuses, when diagnosed, have suffered irreversible kidney damage. Additionally, when the diagnosis is made after 32 weeks of gestation, it is probably better to deliver early and do ex-utero treatment. Therefore, only a limited number of fetuses require intrauterine intervention.

In this review we will delineate the guidelines we use for selection of fetuses that are going to be treated in utero. We will also delineate our policy regarding the handling of the various types of patients whom we encounter.

A thorough ultrasonographic evaluation is the key factor in the management of fetal urinary obstruction. Identification of fetal urinary tract dilatation is not uncommon, especially before 24 weeks of gestation. However, this dilatation does not by itself reflect an obstruction compromising renal function. It is important to separate these entities. The amount of amniotic fluid is important; if an adequate amount is evident on ultrasound, this signifies reasonable fetal renal function and usually indicates adequate pulmonary development. Oligohydramnios, in contrast, signifies fetal urinary tract obstruction. Further investigation is needed before any intervention. A thorough sonographic evaluation must be performed to rule out any significant anomaly or disease. A fetal karyotype should be done, because chromosomal abnormalities have been associated with urinary tract abnormalities and obstruction.1

Fetal Renal Assessment

Decompression in-utero of fetal hydronephrosis or early delivery for decompression ex-utero is beneficial only for fetuses whose kidneys have not suffered irreversible damage. Appropriate counseling and management, therefore, require accurate assessment of renal function, to allow selection of pregnancies likely to benefit from therapeutic intervention.

Renal dysplasia, defined as abnormal parenchymal development secondary to anomalous differentiation of mesonephric tissue, implies irreversible renal damage.2 The functional capacity of an affected kid-
ney depends on the extent and severity of the dysplasia. Conglomerates of disorganized epithelial structures surrounded by abundant fibrous tissue characterize a dysplastic kidney. Cortical cysts are often, but not necessarily, present. Adzick et al. showed that ureteral ligation in lamb fetuses in early pregnancy causes renal dysplasia. Studies on the morphogenesis of dysplasia indicate that approximately 90% of dysplastic kidneys are associated with urinary tract obstruction during nephrogenesis. The incidence of dysplasia correlates with the severity and site of obstruction. Extensive cortical dysplasia nullifies any beneficial effect of decompression of urinary tract obstruction. Therefore, we turned our attention to developing the ability to recognize the functional status of the fetal obstructed kidney.

**Sonographic Evaluation of Renal Dysplasia**

Dysplastic kidneys are associated with renal cortical cysts and increased echogenicity. In our experience, visible cortical cysts had a sensitivity of 44% and specificity of 100% in predicting renal dysplasia. Increased echogenicity had a sensitivity of 57% and specificity of 89%. The severity of hydronephrosis was least predictive with a sensitivity of 35% and specificity of 78%. The demonstration of renal cortical cysts had the highest predictive value for dysplasia among fetuses with obstructive uropathy. Among the 34 kidneys with dysplasia, all 15 with sonographically visible cortical cysts were also highly echogenic. Ten other kidneys were echogenic without demonstrable cysts, however, and an additional nine dysplastic kidneys had neither sonographically visible cysts nor increased echogenicity. Thus, visualization of a kidney without demonstrable cysts or increased echogenicity does not exclude dysplasia.

**Biochemical/Physiologic Evaluation**

Because sonography cannot identify all dysplastic kidneys and, therefore, cannot identify accurately which fetuses with bilateral urinary tract obstruction can benefit from decompression, we considered biochemical studies. Fetal urine is produced by the 13th gestational week and is an ultrafiltrate of fetal serum made hypotonic by selective tubular absorption of sodium and chloride. Fetal urine composition remains constant throughout gestation. In our retrospective experience, fetuses that had had hypotonic urine were later found to have good renal function, while fetuses with isotonic urine were found to have poor renal function. Figure 1 (top) shows the correlation between sodium and chloride levels and renal function. There is clear separation of the fetuses with good functioning kidneys from those with irreversibly damaged, dysplastic kidneys.

Figure 1 (bottom) shows a similar predictive value of the urine osmolarity. Urine output (Fig. 2) usually was significantly different in fetuses defined as having good function compared with those with poor function. In contrast, the differences in fetal iohalamate excretion and fetal urine potassium and creatinine concentrations were not predictive (Fig. 3). On the basis of these results, prognostic criteria to identify the fetus with good function and poor function were generated (Table 1).

The most difficult problem in the management of the fetus with urinary tract obstruction has been selection of the fetus that might benefit from intrauterine treatment. Study of the natural history of untreated obstruction has shown that the fetus with unilateral obstruction and the fetus with mild bilateral obstruction and normal amniotic fluid volume require no intrauterine therapy. Also, the fetus with severe oligohydramnios and severe dysplastic kidney changes seen by sonography is unlikely to benefit from intrauterine therapy. However, between these extremes is a grey zone where potentially fatal renal and pulmonary damage may be averted by intervention. Assessment of the functional potential of the obstructed fetal
FIG. 1. Top. Urinary electrolytes in the fetus with bilateral urinary tract obstruction. Fetal urine sodium and chloride concentrations were significantly different in the good and poor renal function groups. (---) indicates a value at least two standard deviations from the mean of the normal-function group. Bottom. Urinary osmolality in the fetus with bilateral urinary tract obstruction. Hypotonic fetal urine correlates with good function after birth.

<table>
<thead>
<tr>
<th>Predicted Function</th>
<th>Sonographic Appearance of Kidneys</th>
<th>Initial Amniotic Fluid Status</th>
<th>Fetal Urine</th>
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<tbody>
<tr>
<td></td>
<td>Echogenic/cystic</td>
<td>Moderate to severely decreased</td>
<td>Sodium (mEq/ml)</td>
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<tr>
<td>Poor</td>
<td></td>
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<td>Chloride (mEq/ml)</td>
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<tr>
<td>Good</td>
<td>Normal/echogenic</td>
<td>Normal to moderately decreased</td>
<td>Osmolarity (mosm)</td>
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FIG. 2. Urine output in the fetus with bilateral urinary tract obstruction. Fetal urine output correlates fairly well with renal function. (---) indicates a value at least two standard deviations below the mean of the good-function group.

FIG. 3. Fetal urine potassium and creatinine concentrations in the fetus with bilateral urinary tract obstruction do not correlate with fetal renal function.

urinary tract has proved difficult. Indirect methods, such as sonographic determination of bladder filling and emptying or Lasix (Hoechst-Roussel Pharmaceuticals Inc., Somerville, NJ) stimulation of urine production have proved unreliable in our experience. The development of the previously mentioned prognostic criteria that predict the potential for recovery has greatly simplified counseling of families and selection of appropriate management.
Management of Fetal Urinary Tract Obstruction

Our current approach to the management of fetal urinary tract obstruction is outlined in Figure 4. Patients referred for evaluation undergo a thorough static and real-time ultrasound examination. If other significant or life-threatening abnormalities are identified, we counsel the parents about these findings and their significance. Most couples, at this time, elect to terminate the pregnancy.

If ultrasonography reveals no other evidence of significant anomalies, the amount of amniotic fluid present becomes the deciding factor in our management scheme. There are instances of hydramnios and dilated urinary tract systems, most often associated with unilateral obstruction. If the amniotic fluid volume is normal, the nature of the obstruction becomes paramount. Uncomplicated, unilateral obstruction with normal fluid volume does not warrant the risk of invasive interventional decompression. In a report by Harrison et al.,6 eight fetuses with unilateral obstruction and normal amniotic fluid volumes were managed expectantly and delivered at or near term when pulmonary maturity was assured. All eight fetuses continue to do well, with six fetuses having undergone postdelivery pyeloplasties and two fetuses requiring a unilateral nephrectomy. As we gain more experience and expertise, thereby significantly decreasing the risk of intrauterine shunting, one might make an argument for unilateral decompression to prevent the need for postnatal nephrectomy, which occurred in 25% of these cases. However, the current yet ill-defined risks of intrauterine shunting need to be substantially lessened in order to justify this rationale.

For bilateral urinary tract obstruction with normal amniotic fluid volumes, we normally do not recommend invasive intervention. However, if the hydronephrosis

FIG. 4. Evaluation and management of the fetus with bilateral urinary tract obstruction.
is sufficiently severe, we attempt acceleration of pulmonary maturity with betamethasone and proceed with delivery between 32 and 34 weeks gestation by the vaginal route, unless cesarean section is dictated by other obstetric indications. Of some concern, and in need of further evaluation, are recent, pooled data presented at an international meeting on fetal therapy that suggested that as many as one-third of fetuses who died of pulmonary hypoplasia with associated urinary tract obstruction were reported to have "adequate" amniotic fluid volumes. We have not seen this in our experience and, in fact, have reported cases in which the obstruction actually resolved with expectant management, and term deliveries resulted with normal neonatal urinary and pulmonary function.9

An important question is whether restoration of normal amniotic fluid to the oligohydramniotic fetus and decompression of the fetal bladder before birth will result in normal pulmonary development and prevent pulmonary hypoplasia. This question was studied in the lamb model of urinary tract obstruction4; fetal urinary tract decompression before birth resulted in close to normal pulmonary development and prevented pulmonary hypoplasia and respiratory insufficiency.

As shown in Figure 4, our approach to invasive assessment and therapy for urinary tract obstruction is reserved primarily for cases that demonstrate isolated bilateral urinary tract obstruction with oligohydramnios. We start with needle aspiration of the fetal bladder to determine the osmolarity of the urine and its sodium and chloride concentrations, thus separating dysplastic from nondysplastic kidneys. We do not measure urine output any longer because it involves temporary fetal bladder catheterization, which can be avoided by using the biochemical parameters.

Patients whose fetuses have severe oligohydramnios and severely compromised renal function are offered the options of early termination of the pregnancy or non-intervention. The outcome of these pregnancies, in our experience, has been very poor, with no survivors.9

In the presence of good renal function, if the gestation is beyond 32 weeks we recommend early delivery and extraterine decompression. However, in the future some of these pregnancies may become candidates for in-utero shunting to provide more time for pulmonary development after restoration of normal amniotic fluid volume. Finally, if the gestational age is less than 32 weeks and adequate renal function is present, then we offer intrauterine shunting. Usually we use a double-pigtailed fetal bladder catheter. This allows shunting of the fetal urine into the amniotic cavity, bypassing the obstruction and preventing further renal damage while restoring the normal amniotic volume. The renal decompression, additionally, prevents pulmonary hypoplasia.

The drawback of this technique is, in our experience, that those catheters are often displaced or blocked and recatheterizations are often necessary, complicating management and making it riskier and unsatisfactory. Therefore, we offer patients before 28 weeks' gestation another option, that of an open procedure done under general anesthesia. In this technique, the uterus is opened in an area chosen by sonographic evaluation, the lower abdominal wall of the fetus is exposed, and marsupialization of the fetal bladder is done. Our experience is limited, but the approach appears promising.

Conclusions

It now appears that with a combination of the sonographic and biochemical/physiologic criteria described in this article, we can select those fetuses with bilateral urinary tract obstruction that will be most likely to be aided by therapeutic inter-
vention. This is probably the most significant step we have taken in this endeavor. The struggle to provide adequate in-utero therapy will be helped immensely by our ability to choose the appropriate patient for these efforts.

References

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