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Outcome of Prenatally Diagnosed Posterior Urethral Valve Patients— Experience from a Low Income Country

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Abstract

Purpose: To evaluate the outcome of prenatally diagnosed posterior urethral valve (PUV) patients in our setup where fetal interventions are not available. Methods: This prospective study included 24 PUV patients (July 2007 to December 2012), who were diagnosed prenatally but had no prenatal interventions and delivered in our center. Within 24 hours after birth, all of them had urinary ultrasonography (USG). Voiding cystourethrography (VCUG) and renogram were done in the second week and cystoscopy with valve ablation during the same admission period. Patients were followed up regularly after cystoscopic fulguration. Clinical features, treatment, and outcomes were analyzed. Results: Predominant clinical features were renal failure and urinary ascites. Eleven patients (45.8%) had bilateral vesicoureteric reflux (VUR) of different grades. Eight patients (33.3%) had decreased renal cortical thickness (<10 mm). All patients underwent cystoscopic fulguration except three (expired before fulguration). Four patients (16.6%) needed cutaneous vesicostomy along with fulguration. Follow up period ranged from 3 months to 6 years. At last follow-up, 9 (37.5%) boys had renal failure; 7 patients (29%) had normal renal parameters with good urinary stream. Mortality was 7 (29%) including 3 who died before fulguration. Conclusion: Prenatally detected PUV children seem to have both morphologically and functionally compromised urinary system with poor outcome.

Key words: posterior urethral valve, prenatal diagnosis, renal failure, low income country

INTRODUCTION

Posterior urethral valve (PUV) is the commonest cause of bladder outlet obstruction in boys leading to end stage renal disease. The incidence is about 1:2500 to 4000 male births.1 With the widespread use of prenatal ultrasonogram (USG and increasing awareness, more and more cases of PUV are diagnosed prenatally even in developing countries like Bangladesh.² Prenatal diagnosis has undoubtedly altered the presentation, facilitates early intervention, and influences mid-term outcome of PUV. It is early to judge whether this will improve long-term functional outcome and reduce risk of late onset chronic renal failure. According to Thomas' observation, despite prompt valve ablation, careful management and close follow up, boys with prenatally detected PUV are continuing to enter end stage renal failure in their early teens.² More importantly, no form of postnatal intervention can alter the long-term consequences of renal dysplasia, which appears to play a greater role than was previously thought. Our center is a tertiary level referral

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center and patients from all corners of Bangladesh come here. Fetal intervention setup is not yet established in our country. Here, we are presenting our experience of managing prenatally detected PUV patients without fetal intervention.

METHODS

This prospective study was conducted in the Department of Pediatric Surgery, Chittagong Medical College & Hospital, Chittagong, Bangladesh from July 2007 to December 2012. During the study period, 238 patients with PUV were treated, only patients who were diagnosed prenatally by USG but had no fetal interventions were included. Prenatal USG was done at 24 weeks of gestation and repeated at 28 weeks in case of suspected PUV patients. All of them were delivered in our center at term except two who were delivered at 34th and 36th weeks due to associated pre-eclampsia. Clinical features, investigations, interventions, and outcomes were analyzed. At admission, bladder decompression under strict aseptic conditions was done by a transurethral feeding tube (size 5 or 6 Fr). Antibiotic was commenced empirically (amoxicillin) which was adjusted according to culture and sensitivity report of urine. Within 24 hours of birth, every patient had urinary USG. Voiding cystourethrography (VCUG) and diethylenetriaminepentaacetate renogram were done with diuretic clearance in the second week and endoscopic valve ablation was carried out in the same admission by using 7.5 Fr. cystoscope. Antibiotic was advised to all patients according to culture and sensitivity of the urine and patients with significant residual post void urine were also treated by long term catheterization. All patients underwent routine circumcision. Patients were followed up regularly at 15 days, 1 month, 3 months, and every 6 month after cystoscopic fulguration. In each follow up, weight of the baby, fever, feeding habit, urinary stream, and blood pressure were monitored. Urinary USG was done at 3 months interval. VCUG was done 6-12 months after fulguration to evaluate the patients for residual valves, and status of the upper and lower urinary tract. Renogram was done after one year. Clinical features, treatment, and outcome were analyzed. Follow up ranged from 3 months to 6 years.

RESULTS

Total 24 patients were diagnosed in the prenatal period by positive USG findings (hydroureteronephrosis, oligohydromnios, and delayed bladder emptying). They had no prenatal interventions. All of them were delivered in our center. Twenty-one percent of babies had low birth weight

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| Presenting feature Birth weight | Number of patients (%) |
|------------------------------------|------------------------|
| >2500 gm | 19 (79%) |
| <2500 gm | 5 (21%) |
| Urinary ascites | 7 (29%) |
| Renal failure | 9 (37.5%) |
| Vesicoureteric [VUR] reflux | 11 (45.8%) |
| Grade-II, III | 4 (16.8%) |
| Grade-IV, V | 7 (29%) |
| Decreased renal cortical | 8 (33.3%) |
| thickness (<10 mm) | |
| Urosepsis | 8 (33.3%) |

| Table | 2: | Different | surgical | interventions | done |
|-------|----|-----------|----------|---------------|------|
|-------|----|-----------|----------|---------------|------|

| Surgical interventions | Number of patients |
|--|--------------------|
| Fulguration only | 16 |
| Fulguration with cutaneous vesicostomy | 4 |
| Fulguration twice | 1 |
| Ureteric reimplantation | 2 |

(less than 2.5 kg). Five babies were delivered by caesarian section. Predominant clinical features were renal failure and urinary ascites. Presenting features of the patients are summarized in Table 1. Three patients expired before intervention. All the remaining patients (21) underwent cystoscopic fulguration. Four patients (16.6%) needed additional cutaneous vesicostomy along with fulguration due to gross upper tract dilatation (high grade vesicoureteric reflux ([VUR]) and urosepsis. Table 2 shows the different surgical interventions done. During follow up, renal failure persisted in 5 patients (20.8%) and another 4 patients (16.6%) progressed to renal failure, VUR resolved in 4 (16.6%), downgraded in 4 (16.6%), and persisted in 3 (12.5%) patients. Renal cortical thickness improved in 5 (20.8%) patients. One of the patients needed fulguration twice due to residual valve. At last follow up, 10 patients (41.6%) gained significant weight, 9 patients (37.5%) were in renal failure, and 7 patients (29%) had normal renal parameters with good urinary stream. A total of 7 (29%) patients died during study period (3 before and 4 after fulguration) due to renal failure and urosepsis. Five of them had urinary ascites also. Findings of
 Table 3: Findings of the patients before and after fulguration

| Findings of the patients | Before fulguration | After fulguration |
|---|-----------------------|----------------------|
| Renal failure | 9 | 5 |
| Vesicoureteral reflux | 11 | 7 |
| Decreased renal cortical thickness (<10 mm) | 8 | 3 |
| Urosepsis | 8 | 3 |

the patients before and after fulguration are summarized in Table 3.

DISCUSSION

With the widespread use of antenatal USG and increasing awareness, the vast majority of patients with PUV are being diagnosed in utero. The incidence of prenatally diagnosed PUV patients are however limited to 10% in developing countries.3 Thomas stated that more than 80% cases of PUV are now detected prenatally, but Sudarsanan et al. showed that 30% cases of PUV were diagnosed prenatally in their study.^{2, 4} In our center, 10% cases of PUV are detected prenatally which is gradually increasing. Prenatal diagnosis helps serial monitoring of urinary tract dilatation and amount of amniotic fluid for early intervention in well selected patients. It also gives the opportunity to counsel parents regarding their babies' potential for normal or near normal life style, long-term follow up, and the possibility of long-term renal sequelae.5 Prior to prenatal diagnosis, 50-60% of boys with PUV presented with infection that was often of life threatening severity and associated with septicemia, hypovolemic shock, and acidosis. Prenatal diagnosis gives us an opportunity to commence antibiotic prophylaxis and decompress the obstructed system within the first few hours or days after birth. Cases that are not detected prenatally tend to be at the milder end of the spectrum and probably carry a lower risk of overwhelming sepsis.² Primary transurethral fulguration of PUV has become the most widely accepted technique of valve ablation worldwide.6,7 Although controversial, prenatal intervention is technically difficult and the ethical dilemma associated with it precludes its use in many surgical centers worldwide.8 No prospective randomized trials or uniform written guidelines exist that support the therapeutic benefit of prenatal intervention in altering the natural history of obstructive uropathy. It also carries a considerable riskto the fetus with a fetal mortality rate of 43%, even when performed at a developed tertiary care center.^{9,10} Review of the literature demonstrated a complication rate for antenatal intervention of 21-59%.¹⁰⁻¹² No evidence exists that demonstrates the benefit of antenatal intervention in terms of renal function and only in a select number of cases will this benefit pulmonary function.¹³ In low income countries like Bangladesh, there are no facilities available for prenatal intervention of PUV patients. We advise our obstetrician colleagues to deliver these babies in our center so that we can follow and manage them postnatally. Several instruments and approaches have been used for primary valve ablation especially in neonate and small infant.¹⁴⁻¹⁶ We use 3 Fr. ureteric catheter through cystoscope for fulguration of valves. Controversy continues to revolve round the use of temporary high diversion for PUV. Even in most severe cases, many pediatric urologists still do not accept the efficacy of this treatment. They believe that high diversion prevents bladder cycling resulting in noncompliant bladder and does not change the outcome of kidney function in the long run.¹⁷⁻²⁰ We prefer cutaneous vesicostomy for gross upper tract dilatation and urosepsis; because this is easy to perform and manage, and acceptable to parents. Also, renal injury may be exacerbated by febrile urinary tract infections, residual bladder dysfunction, and poorly compliant valve bladder after PUV ablation.^{21,22} Mechanisms that relieve bladder pressure may have a protective effect on renal function. Large bladder diverticula, bladder rupture with urinary ascites, and renal urinary extravasation with urinoma formation are thought to be a pop-off mechanism that protect renal function. In our study, a total of 7 patients (29%) presented with urinary ascites. Unfortunately 5 of them expired who had ascitic fluid more than 500 ml. Proper bladder management was to be taken to reduce morbidity related to valve bladder syndrome. Regular follow up with detailed urinary history is essential.

CONCLUSION

Prenatally detected PUV children seem to have both morphologically and functionally compromised urinary system with poor outcome.

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