Obstructive uropathy in childhood: A review

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Abstract

Background: Obstructive uropathy is an affection of the urinary tract characterized by impairment of urine flow through the tract and which, if left untreated, will cause progressive renal damage.

Aim: To present an update on obstructive uropathy in children with emphasis on the situation in Nigeria.

Methods: Contemporary information on the management of obstructive uropathy was obtained by searching the Medline and adding information from the authors' experience.

Results: Obstructive uropathy causes renal impairment in all age groups. The causes in children may be congenital or acquired. The congenital causes include pelvi-ureteric junction obstructions, posterior urethral valves (PUV), urethral atresia, phimosis and meatal stenosis. Associated anomalies include imperforate anus and vertebral malformations. Acquired causes include calculi, post-traumatic and post-inflammatory strictures and meatal stenosis. Some specific manifestations are prune-belly syndrome, hydronephrosis and renal failure. Diagnostic investigations include ultrasonography, intravenous urography, cystography and renography. Recent technological advances have impacted on the treatment of the different lesions. These include in utero vesico-amniotic shunt and endoscopic valve ablation for PUV and minimally invasive techniques for urolithiasis. Nephrectomy may be indicated in a unilateral damaged kidney. Not all lesions require treatment. Criteria to select patients for treatment require definition. Occasionally treatment fails because of pretreatment irreversible renal damage. The resulting end-stage renal failure is an indication for renal transplantation.

Conclusion: Obstructive uropathy is an important cause of renal impairment. Contemporary advances in the management are yet to become available in developing countries. Compromise treatment options therefore prevail. Adequate treatment is essential to prevent end-stage renal failure.

Key words: Obstructive uropathy, Children

Introduction

Obstructive uropathy is any affection of the urinary tract characterized by impairment of urine flow through the tract and which, if left untreated will cause progressive renal damage\(^1\). The kidney is an indispensable organ for maintaining homeostasis. Dysfunction of the renal system imposes significant morbidity and mortality in the individual sufferer. The renal system plays diverse roles including hormoneogenesis, metabolism, detoxification and excretion of urine which contains agents that may be injurious to the body. An important part of the excretory function is fluid and electrolyte homeostasis. The ability of the kidneys to excrete unwanted products of metabolism depends on adequate flow through the urinary tract. This tract extends from the collecting tubules through the calyces, the renal pelvis, the ureter, bladder and finally the urethra. Impairment of flow is the effect of obstruction in one form or the other. These include both mechanical and functional abnormalities. Stone or posterior urethral valves (PUV) may cause a mechanical obstruction. A neurogenic bladder or reflux at the vesico-ureteric junction leads to a functional obstruction. In a recent study of 699 children with renal disorders in this centre\(^2\), 17 (2.4%) had obstructive uropathy...
comprising PUV (6), meatal stenosis secondary to circumcision (9), bladder tumour (1) and pelviureteric junction (PUJ) obstruction (1).

This paper is an overview of obstructive uropathy in children with reference to a centre in Nigeria.

Materials and Methods

Contemporary information on the management of obstructive uropathy was obtained by searching the Medline from 1966 to 2005. Articles on pediatric patients aged between 0-15 years who presented with symptoms and signs of lower or upper urinary tract obstruction or whose conditions were recognized as incidental findings were studied. The most current opinions about the aetiology of this condition were highlighted where available. Information was also obtained concerning newer investigative techniques as well as current and emerging treatment options. Some information from the authors’ experience in a developing country was also included in the study and viewed against what is obtainable elsewhere.

Results

Obstructive uropathy affects all age groups. In the studied group all parts of the urinary tract are affected. Most cases are congenital. The identified causes included:

Congenital:
- Posterior urethral valves
- Pelvi-ureteric junction (PUJ) obstruction
- Megaureter due to vesico-ureteric reflux
- Anterior urethral valves
- Urethral atresia
- Congenital meatal stenosis
- Phimosis
- Prune belly syndrome
- Neuropathic bladder (eg in spina bifida)
- Malrotation leading to hydronephrosis
- Ureterocele
- Multi-cystic dysplastic kidneys
- Mullerian duct cyst which arises from failure of Mullerian duct regression in early foetal development

Acquired:
- Calculi
- Meatal stenosis (post-circumcision)
- Urethral injuries from trauma, infection, and infestations (eg schistosomiasis)
- Embryonic rhabdomyosarcoma of the prostate or bladder
- Iatrogenic ureteric injuries.

The clinical features in most of the patients were due to consequences of the obstruction. They included urinary retention, calculi, and abdominal distension from hydroureteronephrosis. Haematuria was seen in some cases of calculi, tumours and urinary tract infections. Cases of palpable urethral stones were also seen. Some babies presented with failure to thrive and were subsequently found to have severe renal impairment consequent upon obstructive uropathy. Patients with end-stage renal failure due to bilateral renal involvement presented with such features as generalized oedema and hypertension. Anomalies associated with some congenital causes of obstructive uropathy included imperforate anus as well as vertebral and cardiovascular malformations.

Diagnosis was usually made using ultrasonography, cystography, intravenous urography or renography or combinations of these. Renal function tests, urinalysis and culture as well as plain radiographs are also useful. Radical changes and additions have been made in the treatment of the different lesions due to recent technological advances. For example, in utero vesico-amniotic shunt and endoscopic valve ablation are now being done for PUV. Also, open lithotomies have largely given way to minimally invasive techniques.
Discussion

Urinary tract obstruction may be complete or partial. Complete obstruction results in progressive proximal dilatation and renal damage. Partial or intermittent obstruction is often difficult to diagnose and treat. Although the causes of obstructive uropathy listed above are largely mechanical, immunological factors have been suggested. In chronic upper tract obstruction ureteric dilatation eventually ensues and progresses upwards to involve the pelvis and calyces. The pathological findings are often dependent on the patient's age at onset, being more pronounced in the infant than in the older child. Unrelieved, the obstruction ultimately leads to diminution in the concentrating ability of the kidneys and end-stage renal failure.

Lower tract obstruction, apart from causing pathological changes in the urethra and bladder such as proximal urethral dilatation, trabeculation, sacculcation and diverticula in the bladder, trigonal hypertrophy and vesico-ureteric reflux, also leads to the changes described for upper tract obstruction and these are usually bilateral. Patients may present anytime between birth and late childhood. Some patients remain asymptomatic hence the difficulty in determining the true incidence of congenital anomalies of the urinary tract. In several studies, males are affected more than females. This is partly due to the fact that some conditions such as the prune belly syndrome and posterior urethral valves occur almost exclusively in males. The clinical features relate to the consequences of the obstruction.

Antenatal ultrasound scan (USS) screening in the second trimester is helpful to detect hydronephrosis and PUV. Ultrasound scan should be repeated to distinguish obstructive from non-obstructive dilatation. It is also recommended at 2-6 weeks after birth to confirm the persistence or otherwise of an antenatally diagnosed hydronephrosis. Renal function tests especially urea and creatinine and urine bacteriology are recommended. Plain radiographs may reveal urinary tract stones. Intravenous urography (IVU) is essential in the management of most patients with obstructive uropathy. In unilateral lesions it enables confirmation of the integrity of the unaffected kidney. This is important when considering nephrectomy. It may show dilated tracts in the absence of mechanical obstruction. Other useful radiological investigations include cystography and renography. The latter is useful in the diagnosis, treatment and follow up of patients with hydronephrosis.

Routine prenatal USS is not yet universally available in developing countries. Although antenatal identification of hydronephrosis by USS does not always imply urinary tract obstruction, its use to screen for foetal anomalies should be made mandatory universally in this millennium. In the neonate, there may be ambiguous sex characterization or gender identification. Chromosome studies may therefore, be indicated in some circumstances.

Treatment is tailored to a cause. Ultrasound scan evidence of severe obstructive uropathy is a medical indication for termination of pregnancy. The treatment of the lesions that lead to obstructive uropathy has in the last decade witnessed major advances. However, criteria for selecting patients for treatment require to be properly defined since not all lesions require treatment. In utero surgical treatment for PUV includes placement of vesico-amniotic shunt, endoscopic valve ablation of posterior urethral valves, cutaneous ureterostomies and foetal bladder marsupialization. Post natal surgical treatments of hydronephrosis and posterior urethral valves have long been established. Valve ablation by laser is a new treatment for PUV. Augmentation cystoplasty may be indicated in PUV and prune belly syndrome. Appendicovesicostomy can provide easy access for intermittent bladder catheterization. Technological advances have been made in the treatment of paediatric urolithiasis using extracorporeal shock wave lithotripsy (ESWL), endoscopic ureterolithotomy and percutaneous nephrolithotomy to augment the time-honoured open nephrolithotomy.

Nephrectomy may be indicated in a unilateral damaged kidney as in vesico-ureteric reflux (VUR) and nephrolithiasis. When all else have failed, end-stage renal failure results. The treatments include
renal dialysis in the short-term. Renal transplantation is the final option where available and when affordable.

Technological advances in the management of patients with obstructive uropathy are lacking in most developing countries. Paediatric endoscopy is not established in these countries such as Nigeria. Improvisations such as balloon ablation, transvesical valve ablation and bouginage to dilate the urethra are practised to relieve the obstruction of posterior urethral valves. Here in Port Harcourt there were initial successful results with dilatation in PUV, but many of the patients were soon lost to follow up. Thus long-term results of these treatments in our environment remain inadequately documented. However, from the literature, it is uncertain whether antenatal diagnosis or treatment (e.g. with vesicoamniotic shunts) improves the long-term outcome.

The technological tools currently employed in the management of urolithiasis are lacking in the developing countries and open methods of lithotomy are therefore prevalent in these countries. The new methods of treatment including the placement of the Rodeck vesicoamniotic shunt are yet to be assessed. Although one study suggests that antenatal intervention such as the vesicoamniotic shunt placement may help those fetuses with most severe forms of obstruction, another report concluded that shunt placement is beneficial if done before 20 weeks of gestation. There are doubts as to the long-term benefits of early treatment of posterior urethral valves by valve ablation. In about 15% of patients, the creatinine fails to normalize after bladder decompression. This has been attributed to intrinsic renal disease, including irreversible renal damage prior to treatment, poor bladder development and function. Upper tract obstruction e.g. at the vesicoureteric junction was thought to account for persistently high serum creatinine and deteriorating renal function after bladder decompression. Decompression of the upper tract by nephrostomy or high loop ureterostomy has not confirmed this hypothesis. A recent study in India concluded that persistent distal renal tubular acidosis can predict the development of overt nephropathy after surgery in children with PUV in spite of a normal preoperative serum creatinine.

The effectiveness of relief of hydronephrosis in restoring and preserving renal function is uncertain. Attempts are being made to develop criteria to select patients for operative or nonoperative treatment using serial USS and renographic studies. Worsening deterioration of renal function or hydronephrosis is an indication for surgery. Caution needs to be exercised in this circumstance of nephrectomy for VUR as VUR in a kidney may be protective to the other kidney just as urinoma from an obstructively distended renal pelvis protects its kidney from back pressure effects of the obstruction. Unfortunately both the short-term and the long-term options of renal dialysis and transplantation respectively elude us in these parts.

It has been claimed that there has been a fall in infant mortality from obstructive uropathy. This fall in mortality has been attributed to prenatal diagnosis with USS together with surgical and medical care. It is possible that the fall in mortality may be contributed to by termination of pregnancies in fetuses found to be severely obstructed. However, a recent study in the University of Port Harcourt, Nigeria, where prenatal diagnostic facilities are not available, showed a mortality rate of 35% in 20 patients over a period of six years.

Some specific entities

Phimosis

Phimosis is the inability to retract the foreskin. It may be physiologic in which case intermittent penile erections may gradually separate the foreskin from the glans, thus relieving the phimosis. Secondary phimosis may follow forceful retraction of the foreskin leading to re-epithelialization of the foreskin and glans and consequent recurrent adhesions. It may also follow balanitis or balanoposthitis. Physiologic phimosis in the absence of urinary
obstruction is better left alone as most of them are relieved by 3 years of age$^3$. The secondary ones may respond to topical corticosteroid cream$^4$. Circumcision is curative in all cases. The authors have had to insert a suprapubic catheter in a cachectic 5-day old boy with voiding difficulties and a distended bladder. The working diagnosis was a posterior urethral valve. Circumcision was done to save the parents the additional costs and inconveniences at a later date or another place. The catheter fell off after 2 days. While waiting to reinsert it, the child was observed to void urine with a good stream. Phimosis was probably the cause of his inability to void after birth.

**Meatal stenosis**
This is the abnormal narrowing of the external urethral meatus. It is more often acquired than congenital. The acquired ones most often result from circumcision. It may follow inflammatory reaction from the procedure or it may be caused by damage to the frenular artery$^5$. It may be a cause of neonatal urinary obstruction but recurrent urinary tract infection is also a common mode of presentation. Meatoplasty using local anaesthetic cream is now being practised$^6$. Meatotomy is also an effective cure. Congenital meatal stenosis is most often seen in babies with hypospadias, particularly the coronal or subcoronal varieties.

**Hydronephrosis**
This is an abnormal dilatation of the renal pelvis including the calyces (hydrocalycosis). There are three main types including:
(i) Chronically obstructed hydronephrosis
(ii) Intermittently obstructed hydronephrosis
(iii) Reflux-induced hydronephrosis.

The cause may be at the PUJ or distally. Associated syndromes include Fanconi anaemia syndrome, foetal alcohol syndrome, pyloric stenosis, Noonan syndrome and infants of diabetic mothers. The investigations include screening USS preferably in the 2nd trimester, diuresis renography, IVU and cystogram if reflux is suspected and retrograde ureterography. Diuresis renography may be misleading in poorly functioning kidneys. In the circumstances, percutaneous pressure flow studies may be required$^7$. The size of the hydronephrosis relates directly to the severity of the predisposing abnormality and to the type of abnormality$^8$. The complications of hydronephrosis include susceptibility to trauma, calculi, infection, hypertension and renal impairment.

**Posterior urethral valves (PUV)**
This is the most common cause of childhood obstructive uropathy leading to renal failure$^9$. The condition may present initially in adulthood or late childhood$^{10,11}$. The embryogenesis of PUV is unknown. However some hypotheses have been propounded. These are:

a. Overdeveloped posterior urethral fold;
b. A remnant of the mesonephric duct;
c. An anomalous opening of the ejaculatory duct.

In addition to obstructive uropathy, PUV may predispose to impaired fertility due to slow ejaculation and retention of the ejaculate in the posterior urethra$^{12}$, resulting in retrograde ejaculation.

**Prune belly syndrome or Urethral obstruction syndrome**
The components of this syndrome include absence of abdominal wall muscles, undescended testis and general maldevelopment of the urinary tract$^{13}$. The initial defect is urethral obstruction (due to urethral agensis or PUV) leading to dilatation of the proximal urinary tract, bladder distension and hydroureter. This may cause abdominal distension, abdominal wall muscle deficiency and excess abdominal wall skin, giving rise to wrinkled appearance over the abdomen. It has been suggested that the abdominal laxity in this syndrome is secondary to abdominal distension and stretching during foetal development$^{14}$. Obstruction of urine flow interferes with normal nephrogenesis, resulting in renal agenesis. Involvement of the kidneys, however, is often asymmetric as severe dysplasia on one side may co-exist with a completely normal contralateral kidney. This is an important determinant of prognosis. In prune-belly syndrome, recurrent infection, rather than obstruction, usually represents the greatest threat to
renal parenchyma. Uretero-pelvic junction obstruction may also be present. Gross dilatation of the ureters is due to reduced number of smooth muscle cells which are replaced by fibrous tissue. Also, effective propulsion of urine is lacking. Vesico-ureteric reflux is seen in most cases. The bladder capacity is increased but the voiding profile is usually compromised. There is dilatation of the prostatic urethra and narrowing at the region of the membranous urethra.

The future
Routine USS screening in pregnancy will enable early detection of obstructive uropathy and other congenital malformations. In spite of the initial costs, the benefits are undoubtable. Governments should decree that this be done in the developing world.

Facilities for early detection and treatment of obstructive uropathy in our environment are urgently required. Training of personnel to man these facilities is also necessary. Prevalent conditions in our environment such as malaria and sickle cell disease, impact on renal diseases although there has been no direct influence in obstructive uropathy. Adequate preventive and therapeutic actions in these circumstances will be beneficial. In Nigeria certain centres have been designated as centres of excellence for renal disease care including dialysis and transplantation. Function in these places should be actualized.

One can only hope that the present gloom in the management of obstructive uropathy in the developing world is only in the short-term, to be followed in the long-term by a bloom.

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