Obstructive Nephropathy in Children – A Review

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Abstract
Obstructive nephropathy 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. Causes of urinary tract obstruction can be congenital or acquired. Congenital causes are pelvi-ureteric junction obstructions, posterior urethral valves (PUV), urethral atresia, phimosis and vesicoureteric reflux. The acquired causes are calculus, post-inflammation and post-inflammatory strictures and neovascularization. There are some manifestations like prune-belly syndrome, hydronephrosis and renal failure. Diagnostic investigations include ultrasound, intravenous urography, cystography and renography. Advanced technologies have impacted on the treatment of different lesions such as uroto vesico-amniotic shunt and endoscopic valve ablation for PUV and minimally invasive techniques for urolithiasis. Nephrectomy may be indicated in a unilateral damaged kidney. Sometimes treatment may fail because of pretreatment irreversible renal damage. Such as end-stage renal failure is an indication for renal transplantation. So, proper treatment is essential to prevent end-stage renal failure.

Key words: Obstructive Nephropathy, Renal damage, Renal failure

INTRODUCTION
Obstructive nephropathy is a hindrance of normal urinary flow, that leads to renal dysfunction. The kidney is an important organ for maintaining proper homeostasis. The urinary system plays an important role in hormogenesis, metabolism, detoxification & excretion of urine that bare injurious to the body. The kidneys excrete unwanted products of metabolism that depend on adequate flow of urine through urinary tract. Obstructive nephropathy initiates a complex sequence of events resulting in impaired renal function & it is the major cause of renal impairment in infants & children. An indication for paediatric renal transplantation.

Etiology:
Urinary tract obstruction can be caused from congenital (anatomic) lesion or trauma, neoplasia, calculi, inflammation or surgical procedures, although most childhood obstructive lesions are congenital. The obstructive lesion can occur from the calyces to the urinary bladder & it is major cause of renal impairment in infants & children. An indication for paediatric renal transplantation.

Pathophysiology:
urethral obstruction resulting in dilatation of the proximal urethra. Bladder attempt to overcome the obstruction suffers hypertrophy showed by thickening of bladder wall, trabeculation, sacculation & hypertrophy of bladder neck. The increased intravesical...
pressure resulting in decreased emptying of ureter into bladder & reflux of urine into the ureter. Ultimately causing dilatation of ureter, which also undergoes hypertrophy. The intraureteric pressure increases & back pressure bring about hydronephrosis & thinning of renal parenchyma. Stasis of urine dilated urinary tract causes recurrent infections. The combined stasis, repeated infections & increased intraluminal pressure causes renal parenchymal scarring & progressive deterioration in renal function. The impairment of renal function is harmful to normal growth & development.

In acute obstruction (e.g. from a calculus) glomerular filtration stops & tubular transport is markedly decreased. If obstruction becomes prolonged, renal fibrosis & perpetual damage follows. In intrauterine period obstruction is more serious & leads to renal dysplasia. Renal damage is irreversible even if the obstruction is relieved & may lead to ESRD at an early age.

**CLINICAL MANIFESTATIONS**

Most of the clinical features are due to consequences of the obstruction. Urinary tract obstruction causes hydronephrosis, which is typically asymptomatic in its early phase. An obstructed kidney due to a ureteropelvic junction or ureterovesical junction apparent as mass or cause upper abdominal or flank pain on affected side. Pyelonephritis can occur because of urinary stasis. Upper urinary tract stone causes abdominal or flank pain & hematuria with bladder neck obstruction, urinary stream may be poor & dribbling of urine. There may be straining, incontinence & incomplete voiding. Acute obstruction results in flank pain, nausea, vomiting. Chronic obstruction may be silent or can cause vague abdominal pain. In young infant pyelonephritis may cause sepsis. Renal insufficiency can evident itself by failure to thrive, vomiting, diarrhoea or other non-specific features.

**Posterior Urethral Valve:** Most common causes of childhood obstructive uropathy leads to renal failure. The reported incidence ranges from 1 in 3000 to 1 in 8000 boys. About 5-64% of these patient suffer ESRD during childhood or adolescence. Posterior urethral valves (PUV) are 3 types. Type-I is the commonest form, which radiate distally from the verumontanum & merge into each other to form anterior commisure. Type-II is usually undetectable, do not obstruct the flow of urine. Type-III are less common, consisting of a diaphragm with a central hole.

PUV begins to express effects during developing urinary tract early in second trimester, the back pressure results in vesicoureteric reflux (VUR), hydronephrosis, renal dysplasia & impaired renal function. It can be detected during antenatal ultrasound. The classical features are oligohydramnios, bilateral hydronephrosis & hydroureter, a thick-walled bladder & a dilated posterior urethra, renal dysplasia and pulmonary hypoplasia. The good prognostic factors are sodium <100 mEq/l, chloride <90 mEq/l & osmolality below 200 mosm /L. Vesicoamniotic shunt has been described to overcome this to gain time for prolonging the pregnancy & protecting the developing kidneys & lungs. Treatment option is postnatal surgical valve ablation after treating UTI & correction of fluid & electrolyte abnormality. High loop ureterostomy is considered if the child is very sick, gross pyuria & if the trial of bladder drainage has not helped.

**Pelviureteric Junction Obstruction:** Pelviureteric junction (PUJ) obstruction is the functional obstruction of junction between the renal pelvis & ureter. It is the most common cause of hydronephrosis with an incidence of 1 in 2000 children, with a male female ratio 3:1 & bilateral in 20-25% of cases. This may be due to intrinsic abnormality, muscular abnormalities of the ureter, ureteral polyps, ureteral folds, crossing vessels and rarely secondary to VUR. Children usually present with flank mass, upper abdominal or recurrent flank pain, urinary tract infection (UTI), hematuria. Some may appear acutely with renal pain (Dietl’s crisis). Antenatally it is suspected in a fetus with hydronephrosis without ureteric dilatation & with a normal bladder and normal amniotic fluid volume. Postnatally the majority will tend to deteriorate within the first 6 months of life as a result of maturational changes in
GFR. By micturating cystourethrogram (MCU) & using diethylene triamine pentaacitic acid (99mTC DTPA) diuretic renogram with split renal function it can be diagnosed. With grade 1 or 2 hydronephrosis, observation is usually appropriate. If the hydronephrosis is grade 3 or 4, diameter of renal pelvis more than 3cm, poor upper urinary tract drainage or poor differential renal function (35%) after diuretic renogram then surgery, Anderson Hynes pyeloplasty is recommended.

Vesicoureteric Reflux (VUR): VUR refers to the retrograde flow of urine from the bladder to the ureter & kidney. Reflux occurs when the submucosal tunnel between the mucosa & detrusor muscle is short or absent. About 38% of children with antenatal hydronephrosis it is seen. Reflux usually revealed during evaluation for a UTI. Around 80% are female and the mean age at diagnosis is 2-3 yrs. Reflux is present at birth in 25% of children with neuropathic bladder, 50% of boys with PUV, 15% with multicystic dysplastic kidney or renal agenesis, PUJ obstruction.

Reflux leads to pyelonephritis by facilitating the transport of bacteria from the bladder to the upper urinary tract. The inflammatory reaction may result in renal injury or scarring, renal insufficiency, ESRD. Reflux severity is graded using the international reflux study classification is based on the appearance of the urinary tract on a contrast voiding cystourethrogram (VCUG). Grade-I: reflux into a nondilated ureter; Grade-II: reflux into the upper collecting system without dilatation, Grade-III: reflux into dilated ureter & blunting of calyceal fornices ; Grade-IV: reflux in to a grossly dilated ureter ; Grade V : massive reflux with significant ureteral dilatation & tortuosity loss of the papillary impression.

VUR is treated with antibiotic prophylaxis & follow up for grade I, I I and surgical reimplantation & endoscopic correction for grade IV, V.

Neurogenic Bladder: Neurogenic bladder means malfunction of the urinary bladder due to disease of the central nervous system or peripheral nerves that controls micturition. The causes includes dysraphism, open and closed spina bifida, sacral agenesis, spinal cord tumor, trauma, transverse myelitis and autonomic neuropathy. Features lead to suspect are poor or impaired urinary stream, straining to pass urine, impaired / lack of bladder sensation, small urinary volumes, continual dribbling, infrequent micturition, impaired bladder emptying, recurrent UTI, abnormality of the spine, abnormality of lower limb & associated constipation.

For diagnosis USG of abdomen, MCU for VUR and dimercaptosuccinic acid (99mTC DMSA) scintigraphy for renal scars can be made. Management done with bladder expression (the crede manoeuvre), bladder straining, clean intermittent catheterization, and anticholinergic drugs. Surgery, like vesicotomy, urinary diversion, sphincterotomy, reimplantation of ureters, endoscopic reflux correction is also recommended.

Urolithiasis: Approximately 7% of urinary calculi occur in children less than 16 years of age. Renal stones are composed of calcium salts (70%), uric acid, magnesium ammonium phosphate or cystine. Metabolic abnormalities seen with these children like hypercalciuria, hyperoxaleuria, hyperuricosuria, UTI, cysteinuria & urinary tract anomalies. Medical management such as thiazides for hypercalciuria, alkali administration in renal tubular acidosis, alopurinol in hyperuricosuria. Surgical measure like extracorporeal shock wave lithotripsy, percutaneous nephrolithotomy or open surgery also done.

Urethral Stricture: Urethral stricture are common in boys. It can be congenital or acquired. Congenital stricture urethra is not common. Acquired stricture occured due to infection, trauma. Child may manifest with dribbling of urine or poor urinary stream associated with straining, palpable bladder and occasionally palpable kidneys. The diagnosis can be made by a intravenous urography (IVU) or retrograde urethrography, ultrasonography. But endoscopy is confirmatory. Definitive treatment is surgery, which includes urethrotomy, urethral dilatation, urothrolaplasty.

Phimosis: Phimosis is inability to retract the prepuce. It is physiologic at birth. But with time the adhesions between

Figure:
the prepuce, glans lyse & the distal phimotic ring loosens.\textsuperscript{29}
At the end of the first year of life retraction of foreskin behind the glandular sulcus is possible in only about 50% of boys, this rises to approximately 90% by the age of 3 years.\textsuperscript{1,35} In true phimosis dispose to smegma collection which causes recurrent balanoprostitis.\textsuperscript{1,30} If physiological phimosis persist with dysuria corticosteroid cream is given.\textsuperscript{1,31} Circumcision is needed in true phimosis.

**Prune Belly Syndrome:** Prune belly syndrome also known as triad syndrome or Eagle Barrett syndrome\textsuperscript{7} occurs in approximately 1 in 29000 to 40000 male births.\textsuperscript{32} Urinary tract abnormalities, abdominal muscle deficiency and undescended testis are the components.\textsuperscript{33} Cardiac (ASD, VSD, TOF), pulmonary hypoplasia, gastrointestinal tract abnormality (Malrotation of Gut, Volvulus, Omphalocele, Imperforate Anus), musculoskeletal anomaly (limb anomaly, scoliosis, hip dislocation, pectus excavatum, talipes equinovarus) are often associate.\textsuperscript{34}

Temporary bladder drainage procedures such as vesicotomy, orchidopexy & abdominoplasty with antibiotic prophylaxis are the treatment options. Renal transplantation also brings good results.\textsuperscript{5,12}

**Hydronephrosis:** Hydronephrosis can be defined as dilatation of the renal pelvis and or calices. It is detected by routine antenatal USG with an incidence of 0.5 to 1.\textsuperscript{36} Commonly detected between 18 and 20 weeks of gestation at the routine anomaly scan.\textsuperscript{15} More than 5mm diameter is considered abormal.\textsuperscript{15,38} It is usually unilateral but may be bilateral in 15-30% cases.\textsuperscript{42}

Society of fetal urology grading of hydronephrosis\textsuperscript{5}.

Prenatal intervention is placing a vesicocamniotic shunt\textsuperscript{37,39}. Postnatally with hydronephrosis, evaluation of

<table>
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<th>Grade</th>
<th>Central renal complex intact</th>
<th>Renal Parenchymal Thickness Normal</th>
<th>Ultra sound scan</th>
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<td>I</td>
<td>Intake</td>
<td>Normal</td>
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</tr>
<tr>
<td>II</td>
<td>Slight splitting of pelvis</td>
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<td><img src="image2" alt="Image" /></td>
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<tr>
<td>IV</td>
<td>Wide splitting of pelvis and calices</td>
<td>Normal</td>
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<td>V</td>
<td>Further splitting of pelvis and calices</td>
<td>Normal</td>
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VUR & antibiotic prophylaxis recommended. At 4-6 weeks of age MCU, diuretic renography could be done. Surgery is required in 28% cases. PUV account for 1-4%, where definitive treatment is surgery.\textsuperscript{37}

**INVESTIGATION AND IMAGING MODALITIES FOR EVALUATION**

Urine R/M/E, Culture & sensitivity test, Complete blood count, Serum creatinine, electrolyte, bicarbonate, calcium, phosphorous, parathyroid hormone.

Urinary tract dilatation, renal cortical thickness, calyx size, diameter of pelvis, ureter, thickness of bladder, tumor and calculi can be obtained by renal USG. Doppler USG needed for evaluation of vascular anomalies.\textsuperscript{1,40} Plain x-ray KUB for urinary tract calculi.\textsuperscript{1}

Radionuclide studies includes $^{99m}$Tc DTPA, $^{99m}$Tc DMSA, $^{99m}$Tc MAG3 (Mercaptoacetyl triglycine), $^{99m}$Tc GHA (Glucoheptonate) for renal scarring and VUR.\textsuperscript{1}

Radiodiagnostic studies includes IVU for hydronephrosis, calculi, ureterocele; MCU for bladder and urethral obstruction, VUR; retrograde urethrography for urethral
obstruction.1
Endoscopy for direct visualization of lesion e.g PUV.
Magnetic resonance urography may be carried out for
determination of cause, level and degree of obstruction in
poorly functioning kidneys.41

COMPlications
Congenital obstructive nephropathy is the most common
cause of chronic kidney disease (CKD) and ESRD in
children.1 Symptomatic and asymptomatic UTI are both
common. Hypertension is common in scarred kidney with
proteinuria. Fluid and electrolyte abnormalities, systemic
acidosis (distal RTA) occurs due to insufficient urinary
acidification.16 Growth retardation due to bony
abnormalities and failure to thrive are also common.1,42

TREATMENT
After diagnosing obstructive nephropathy therapy
emphasizes on the rapid restoration of normal urine flow
either by medical or surgical interventions.

There are some newer surgical interventions for VUR as
conventional, open ureteric re-implantation increasingly
replaced by alternative techniques:

*Sub-ureteric transurethral injection (STING) of
dextranomer/hyaluronic acid co-polymer (Dx/HA) or
De/flux procedure.

*Laparoscopic procedure
*Robot assisted procedure

Bladder drainage, vesicostomy, valve ablation and high
diversion surgery and VUJ stents are indicated in PUV.42,43
Initially patient may need acute renal replacement therapy
to remove toxins and maintain fluid, electrolytes & acid
base balance.43 Some patients may experience enough
recovery of renal function and compensatory function
dialysis once established.12

In patients with CKD hyperkalemia and hyper-
phosphatemia can be compensate with low potassium, low
phosphorus diets with phosphate binders.12 Anemia is
corrected with iron supplementation or human
recombinant erythropoietin. Production of 1,25-dihydroxy
vitamin D is reduced, requiring supplementation.
Nutritional supplementation and recombinant growth
hormone is necessary to maintain normal growth.44

PROGNOSIS

16.3% of pediatric obstructive nephropathy patients goes
into ESRD requiring renal transplantsations.44 Long term
renal function is variable depends on underlying pathology
and associated complications. Prognosis is worse if UTI
remains untreated.22 Earlier diagnosis and skilled
intervention by pediatrician and pediatric urologists can
give a better outcome.

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