A RARE CASE OF EARLY URETHRAL OBSTRUCTION SEQUENCE

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Abstract :

Early urethral obstruction is most commonly the consequence of urethral valve formation during the development of prostatic urethra. Less commonly, it is due to urethral atresia, bladder neck obstruction or distal urethral obstruction. A rare case delivered at Pravara Rural Hospital ,Loni with bilateral hydronephrosis with imperforated anus, undescended testis and unilateral Congenital Talipes Equino Varus. Child underwent Exploratory Laparotomy with descending loop colostomy and bilateral nephrostomy.

Key Words : *Urethral obstruction, Prune-Belly, Hydronephrosis.*

Introduction :

Urine formation occurs at 7-8wks of fetal life. With intrauterine urethral obstruction there is progressive back-up of urine flow, leading to the consequences as shown in the flow chart[1],shown in figure 1.

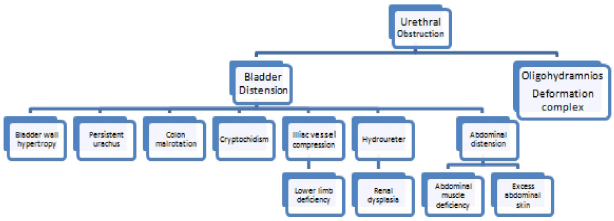

Fig.1:Showing consequences of Urethral Obstruction.

The male to female ratio of 20:1[2] in this disorder is the result of predominant malformations being in the development of penile urethra. Cryptorchidism occurs secondary to the bulk of the distended bladder, preventing full descent of the testes. The bladder pressure usually limits full renal morphogenesis and may result in dilatation of the renal tubules, which by histological section examination may be interpreted as renal "cysts". Similarly the hydrostatic pressure generated by distal obstruction most likely accounts for the hypoplastic prostate, seen in majority of cases. The compressive mass of the bladder may limit full rotation of the colon and may even compress the iliac vessels to the point of causing partial defects or vascular disruption of the lower limb(s). The oligohydramnios will give rise to all the secondary phenomena of the oligohydramnios deformation sequence. Severe early urethral obstruction is often lethal by mid to late fetal life unless the bladder rupture may occur through a patent urachus, an obstructing urethral "valve", or the wall of the bladder or ureter. Following decompression, the fetus will be left with a "prune belly"[3] (**Prune-Belly-Syndrome)**

Case Report:

Full term male child(Fig.2) weighing 3770 grams was born on 20th November 2008 by LSCS under spinal anaesthesia (Indication- previous LSCS with non-progress of labour with congenital hydronephrosis)

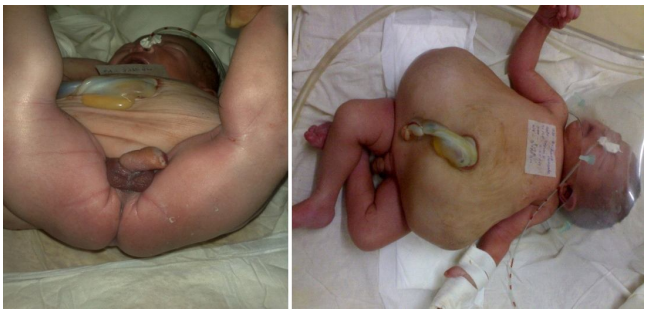

Fig.2:Showing child at birth.

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with APGAR 7, 8, 9 to a $G_3P_1L_1A_1$, 25 years old mother whose present pregnancy on antenatal Ultrasonography showed megaureter with megapelvis with hydronephrosis on left side. Child was born with imperforate anus, undescended testes, bilateral huge palpable kidney with central abdominal wall weakness and hypospadiasis. Urethral Catheterisation was tried to relieve obstruction which failed suggesting Posterior urethral valves. The child was further investigated and reports shown in Table 1.

Investigations	20/1/08 (Cord)	21/11/08	21/11/08	28/11/08
Hb (gm/dl)	12.9	17.2	15.3	
PCV	41.9%	55%	50.1%	
S. urea		18.5	32.3	58
S. creatinine		0.64	0.91	0.98
S. Na		135	131	146
S. K		4.2	5.1	3.3
S. Ca		9.3	8.3	8.9

Table 1:Showing Investigations.

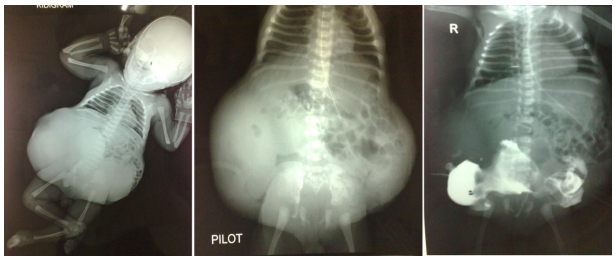

Fig. 3(a-c) Showing Kidigram,X-ray and MCU which showed:1)*Defective anterior abdominal wall musculature and mega extra-renal pelvis bilaterally and mild degree bilateral hydronephrosis with normal size kidneys.2)Non-opacification of ureters and urinary bladder.*

Ultrasonography of abdomen and pelvis revealed bilateral megaureter with megapelvis. Opinion of Pediatric surgeon was taken in view of obstructive uropathy and imperforate anus. Child underwent Descending loop colostomy with bilateral Nephrostomy with drains in-situ under GA on the same day and postoperatively put on IV Fluids and IV Antibiotics. Left nephrostomy tube drain was inadequate. Child was monitored clinically and biochemically for one week. Although there was improvement in child's condition but the renal function gradually deteriorated over period of time.The child was with us for 8 days and then referred to higher centre for fulguration of posterior urethral valve and further management.

Discussion:

Most of the cases of Prune-Belly-Syndrome are stillborn[4]. Urinary tract includes massive dilatation of the ureters and upper tracts and a very large bladder with a patent urachus is present. Most patients have vesico-ureteral reflux. The prostatic urethra is dilated, prostate is hypoplastic. The anterior urethra may be dilated resulting in megalourethra[5]. Kidneys usually show various degree of dyplasia and testis usually is intra-abdominal. Malrotation of bowel is often present cardiac abnormalities occur in 10% and mostly these children have abnormalities of musculoskeletal system, including limb abnormalities and scoliosis. In girls, anomalies of urethra, uterus and vagina are usually present. Most of these children have difficulty in emptying bladder because of defective musculature. The aim of treatment is to prevent Urinary Tract Infection with antibiotic prophylaxis if no obstruction is present. If obstruction is present, temporary drainage procedure such as vesicotomy can be tried. Correction of undescended testis by orchidopexy is difficult as testis is located high up in the abdomen. Reconstruction of abdominal wall offers cosmetic benefits.

Prognosis depends on degree of pulmonary hypoplasia and renal dysplasia[6]. One third[7] of the children with Prune-Belly-Syndrome are stillborn or die in first few months of life because of pulmonary complications. Most of them who survive, develop end-stage[8] renal disease from dysplasia or complications of infection or reflux and finally require renal transplantation, which sometimes offer better results.

In our patient although there was no pulmonary hypoplasia but the renal function was deranged.

Conclusion

Early urethral obstruction is a rare congenital anomaly seen mainly in male infant and can be diagnosed antenatally. The survival and long outcome depends on pulmonary hypoplasia and renal dysplasia.

References

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Medical Quote

Men that look no further than their outsides, think health an appurtenance unto life, and quarrel with their constitutions for being sick; but I that have examined the parts of man, and know upon what tender filaments that fabric hangs, do wonder that we are not always so; and considering the thousand doors that lead to death, do thank my God that we can die but once.

~Thomas Browne