BDR NEWS

The official newsletter of The Birth Defects Registry, Chennai
(Unit of Fetal Care Research Foundation)

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Proceedings of the meeting of Chennai Birth Defects Registry held on the 25th July 2002

The registry meeting was held on the 25th of July 2002 at Fetal Care Research Foundation, Chennai. Chennai BDR has now become the first functional unit of the Birth Defects Registry of India (BDRI). Similar registries at selected towns such us Erode, Thanjavur, Theni, Chidambaram and Pondicherry are in the process of getting initiated shortly.

Dr. Sujatha Jagadeesh welcomed the members and introduced Dr. Sripathy, Pediatric Urologist, who was invited as a guest speaker to deliver a talk on congenital obstructive uropathy. She observed that acquiring knowledge on correctable disorders is essential and it is a part of the goals set by the registry. This was followed by an excellent presentation of the guest speaker on different obstructive renal anomalies and their management in children. Given below is the synopsis of the talk.

Dr. S.Suresh proposed vote of thanks and called for the involvement of the pediatricians in registering cases up to one year of age in the registry, as this information would throw light on the supportive measures that could be adopted for the correctable congenital problems.

Approach to diagnosis and treatment of obstructive uropathy in children

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Introduction:

The term 'obstructive uropathy' commonly conjures up visions of two anomalies, namely hydronephrosis due to pelvi-ureteric junction obstruction (PUJ) and posterior urethral valve disease (PUV). But in reality there are a number of other conditions, which can cause severe damage to the growing kidneys if not diagnosed and treated appropriately. Broadly speaking, obstructive lesions of the urinary tract in children can be classified as anatomical obstructions and functional obstructions. Anatomical obstructions can be intrinsic or extrinsic.

Intrinsic anatomical obstructions of the urinary tract include:

- Hydronephrosis due to PUJ obstruction
- PUV disease
- Primary Obstructive Megaureter
- Obstructive Refluxing Megaureter
- Ectopic ureteroceles obstructing the bladder outlet
- Anterior urethral valves
- Cowpers duct cysts
- Severe phimosis caused by Balanitis Xerotica obliterans

Extrinsic anatomical obstructions of the bladder outlet include:

- Utricular cysts in males (with severe hypospadias)
- Prolapsing bladder diverticulae
- Urethral or vaginal ectopic ureters
- Rhabdomyosarcoma of the prostate or the vagina

Apart from these causes the bladder outlet can be pressed by

- Presacral tumors
- Loaded rectum

Functional obstructions of the urinary tract are insidious and often present only with recurrent urinary infections or dribbling. These diseases are poorly categorized and lumped under the term 'neurogenic bladder disease'. All of them are characterized by high pressure within the urinary bladder. The unique ability of the bladder to store and expel urine under low pressure is lost. When pressures in the bladder exceed 20 CM of water, severe backpressure is exerted on the renal tubules and the

glomeruli resulting in gradual loss of renal substance and onset of renal failure.

Children with neurogenic bladders often have myelodysplasia. But children with normal spinal cords can also develop neurogenic bladders. This is now classified as 'dyselimination Syndrome' meaning thereby that the problem lies in the urinary and rectal sphincters and their control. These children have to be taught to pass urine in a coordinated manner. This re-training process may take many months and involve biofeedback with transcutaneous stimulation and even 'hypnotherapy' at times. The diagnosis and management of hydronephrosis and PUV disease will now be discussed in detail including recent advances in management.

Antenatally diagnosed PUJ Obstruction:

When antenatal ultrasound was widely introduced, it was found that a number of fetuses had hydronephrotic kidneys. This raised the problem of deciding which of these kidneys were truly obstructed and which of the kidneys were exhibiting transient dilatation. Unfortunately to this day, this controversy has not been fully sorted out and the management of an antenatally diagnosed hydronephrotic kidney is still based on the beliefs of a given individual. However, a balanced approach based on current evidence will be presented here as a guideline to safe practice.

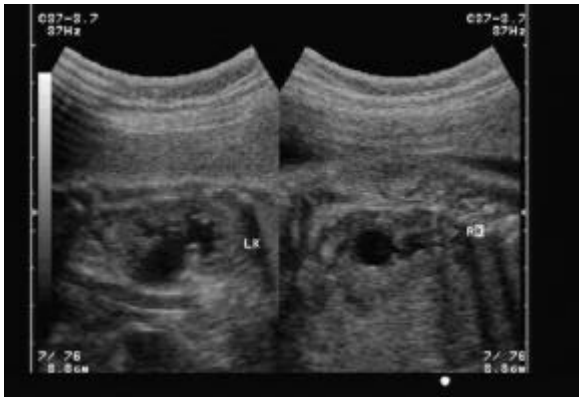

Fig.1: Antenatal ultrasound shows pelviureteric junction obstruction (Picture courtesy: Mediscan Systems)

Antenatal pointers to the need for surgery after birth:

- If third trimester scan shows the antero-posterior (AP) diameter of the renal pelvis to be greater than 20 MM with calyceal dilatation then surgery will in all probability be needed postnatally.
- If third trimester scan shows the AP diameter of the renal pelvis to be between 20-30 MM then careful follow up is needed. Thirty one percent of these children will ultimately need surgery.
- If third trimester scan shows a renal pelvis less than 20 MM in AP diameter then surgery will probably not be needed.

The exceptions to this rule are severe caliceal clubbing and a small intra-renal pelvis. Such children will need early corrective surgery according to HK Dhillon.

Postnatal management of a PUJ obstruction:

Confirmatory ultrasound is done a week after birth. There is little to be gained from doing a scan within the first 48 hours after birth. These children are relatively dehydrated and even a significant obstruction may appear misleadingly innocuous. It has been my personal practice to ask for a DTPA renogram at 60 days of age. This study should be done in a well-hydrated child following injection of a diuretic. The decision to operate is based on a careful study of the following:

- Intraparenchymal transit time
- Washout of isotope following injection of diuretic (called T½)
- Excretion pattern of the kidney as seen on a graph

Commonly a kidney with function of less than 40%, with a T½ of greater than 20 minutes and a climbing curve is a candidate for surgery. There are a group of surgeons who will not operate unless the renal function dips below 30%. Unfortunately this approach needs a close follow-up and renal recovery following surgery cannot be guaranteed according to SA Koff.

Newer isotopes like MAG 3 are promising better delineation of renal function due to better extraction by the kidney. More recently Gadolinium enhanced DTPA MRI is being used as a tool to further choose those obstructions, which will truly benefit from surgery, postulated by CK Yeung.

There are a number of investigators who believe that the diuretic renogram is not a very accurate study and are trying to evolve more accurate tests to define PUJ obstruction. A test called upper tract urodynamics used by AE Khoury et al, which involves perfusion of the renal pelvis and measurement of the pressure differential across the PUJ.

As a rule of thumb in clinical practice, however, a palpable kidney warrants surgery. The results of dismembered pyeloplasty are excellent and in large series success rates of 98% are routinely reported. Follow-up is generally maintained for two years from the date of surgery.

Posterior urethral valve disease:

Antenatal diagnosis is of crucial importance in this condition, as treatment needs to be instituted soon after birth for a good outcome. The antenatal finding of bilateral ureterohydrnephroses with a distended bladder and a dilated posterior urethra ('key-hole' sign) is pathognomonic of PUV.

Antenatal pointers to a poor outcome:

The following indicate that the child will have a poor outcome after birth:

- Renal pelvis > 10 mm in AP diameter

- Consistently imaged fetal bladder at less than 28 weeks gestational age
- Increased renal parenchymal echogenicity
- Renal cortical cysts

Eighty nine percent of PUV children with these antenatal findings were either dead or in chronic renal failure in infancy as per the study done by Hutton et al in 1997.

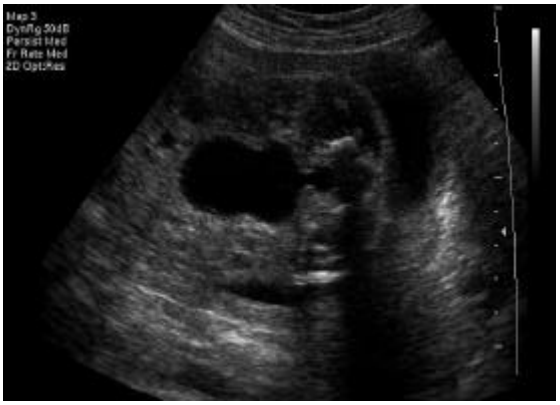

Fig.2: Antenatal ultrasound shows posterior urethral valve disease (Picture courtesy: Mediscan Systems)

Management of antenatally diagnosed PUV:

A renal ultrasound is taken to confirm the presence of bilateral ureterohydronephroses, dilated thickened bladder and a distended posterior urethra. In the nursery the bladder is punctured supra-pubically and dye instilled. A compression cystogram is done to confirm PUV disease. This contrast X-ray also serves to assess adequacy of valve fulguration at a later date. Under GA, valve fulguration is done with an 8 Fr cystoscope and a Bugbee electrode. With this instrument children 2.5 KG and above can be safely subjected to primary fulguration.

Special situations:

- Massive reflux into a defunct kidney: The reflux serves to emphasize that bladder pressures are very high and the ureter therefore is acting as an extension of the bladder. These children should be managed conservatively. Nephrouretrectomy should never be done as the 'pressure pop-off' provided by the dilated ureter will be irretrievably lost.
- Severe sepsis and metabolic acidosis: Children who present in infancy with urosepsis should be subjected to bladder drainage, IV antibiotics and supportive measures. If renal recovery is seen as shown by a rapid drop in serum creatinine then primary fulguration may be safely proceeded with. If renal recovery is not forthcoming or urosepsis is persistent then upper tract diversion (ureterostomies) may be indicated.
- Persistently high Serum Creatinine even after adequate fulguration. In this condition the thick walled high-pressure bladder does not permit adequate urine drainage from the ureters. These children should be subjected to ureterostomies to salvage renal function.

Follow-up after valve fulguration is extremely important. It is now recognized that abnormal bladder function is the eventual cause of renal failure in this condition (SB Bauer). In Bauer's review only 10% of PUV bladders were normal in the long term. Thirty five percent went into myogenic failure, 25% had detrusor hyperreflexia, 25% had small capacity with poor function and 5% exhibited bladder neck hypertrophy.

The greatest advance in the management of PUV in this decade has been the recognition and early treatment of these bladder abnormalities so that renal failure and transplantation is avoided. A study by J Gennaro et al reveals that uroflowmetry and urodynamics are important tests of bladder function which need to done regularly in children following valve fulguration in order to identify various functional disorders.

Indications for urgent urodynamics in PUV:

- Increasing Serum Creatinine
- Increasing Hydronephrosis
- Poor voiding in spite of adequate fulguration
- Persistent wetting
- Poor somatic growth

If bladder dysfunction is left untreated, tubular dysfunction results from backpressure leading to polyuria. Large quantities of poor quality urine result in persistent bladder distension and myogenic detrusor failure.

Recent advances in pharmacotherapy of PUV disease:

It has been recognized that children below three years of age with PUV disease have a high incidence of bladder dysfunction (hyperreflexia and poor compliance). Anticholinergic therapy is routinely being advised in these children till five years of age. This serves to relax the detrusor and reduce bladder pressure. In children with persistent bladder neck hypertrophy, obstructed voiding and wetting, alpha-blockers are being tried to reduce outlet resistance.

Good prognostic factors in PUV:

- Rapid fall of serum creatinine 5-7 days after fulguration
- Serum creatinine <1 mg/dl at 1 year of age
- Absence of reflux with a smooth bladder outline

Bad prognostic factors in PUV:

- Oligohydramnios
- Bilateral reflux
- Ultrasound showing loss of cortico-medullary differentiation and increased echogenicity

Bladders, which are non-responsive to pharmacotherapy, will need to be augmented and drained by clean intermittent cauterization. It should be remembered that even a transplanted kidney can be affected by a poor bladder hence correction of bladder dysfunction is paramount in PUV disease.

Following is the list of Doctors who represented their Hospitals at the sixth BDR meeting on 25th July 2002

Name of the Hospital	Participants	Code
Mediscan Prenatal Diagnosis & Fetal Therapy Centre	Dr. S. Suresh Dr. Indrani Suresh Dr. Sujatha Jagdeesh Dr. Suchitra Dr. Gazala Jabeen Dr.G. Thangavel Dr. Jayalakshmi Mrs. Vijayalakshmi Raja Mrs. Ranjani Pathasarathy,	001
Kanchi Kamakoti Child Trust Hospital	Dr. V. Ganapathy	003
Vijaya Hospitals	Dr. Lalitha Dr. Mona Dr. Vani Pujari Dr. Uma Dr. Sujatha Dr. Noorudhin Dr. Nithya Dr.K. Priya Dr. Shanthi	006
Sri Ramachandra Medical College Hospital	Dr. Usha Viswanath	008
Durgabai Deshmukh General Hospital	Dr. Indira	009
Saidapet Corporation Hospital	Dr. Sheela Gopinath	010
Public Health Centre, West Mambalam	Dr. Prabha Ganapathy Dr. Subbulakshmi	011
Nagamani Hospital	Dr. S. Prema Kumari	014
G G hospital	Dr.Deepa Hariharan	015
Devaraj Manikchand Maternity Hospital	Dr. Revathi Ravikumar	016
Ritherdon Nursing Home	Dr. Zunaidha	017
Lakshmi Maternity Hospital	Dr. Sivasankari	018

Other member hospitals

Name of the Hospital	Code
EV Kalyani Medical Centre	002
Sundaram Medical Foundation	005
Apollo Hospitals	007
CSI Rainy Multi Specialty Hospital	012
CSI Kalyani Hospital	013
Seethapathy Nursing Home	019

If you know of any hospital willing to join the registry, please ask them to contact us. We are available at:

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