Review Article

Testicular Prosthesis in Paediatric Urology: Current Concepts and Available Alternatives

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Abstract: Prosthesis is an artificial material used as a replacement for its natural counterpart. Use of testicular prosthesis in paediatric urology is limited and indications are well defined. In this review we tried to find out and summarize the current indications and available options in paediatric urology for these prostheses.

Keywords: Anorchia, Orchidometer, Testicular prosthesis

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Introduction

Prosthesis is an artificial material used as a replacement for its natural counterpart. Use of testicular prosthesis in paediatric urology is limited and indications are well defined. In this review we tried to find out and summarize the current indications and available options in paediatric urology for these prostheses. An extensive PubMed, Medline and Google scholar search was done to see the available literature and current practice. For the purpose of simplicity the subsequent discussion is under following heads:

- Indications
- Assessment of size required
- Timing
- Procedure
- Complications
- Evolution and currently available options
- Experimental prostheses which are promising

A). Indications of testicular prosthesis placement: Testicular prosthesis is required in following conditions.

1). Congenital anorchia (vanishing testis or pure gonadal atresia)
2). Acquired anorchia due to

a). Trauma
b). Tumor
c). Torsion
d). Dysplasia
e). Dysgenesis
f). Intersex disorders requiring male genitoplasty

B). Assessment of the size of prosthesis required:

This entirely depends upon the age at placement of the prosthesis and the scrotal development. The assessment of the volume of testis is done using an instrument called as Orchidometer/ Orchiometer. The orchidometer was introduced for the first time by Swiss paediatric endocrinologist Prof. Andrea Prader[1] of university of Zurich in 1966. It consists of a string of twelve numbered wooden or plastic beads (some time referred as *Prader's balls, medical worry beads or endocrine rosary*) of increasing size from one to twenty-five milliliters (Fig 1).

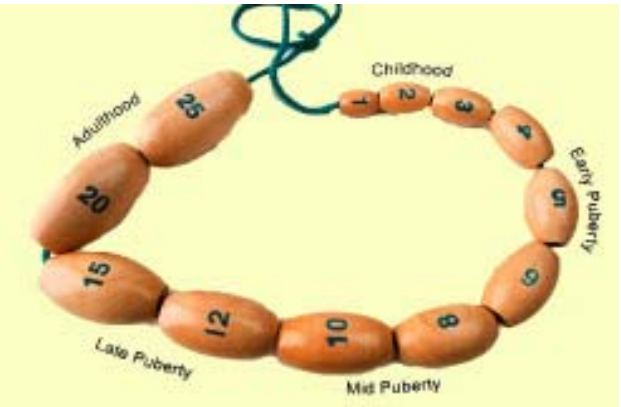

Fig. 1. Prader's orchiometer

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The beads are compared with the testicles of the patient, and the volume is read off the bead which matches most closely in size. Prepubertal sizes are 1-3 ml, pubertal sizes are considered 4 ml and up and adult sizes are 12-25 ml. A number of other orchidometers are also available and marketed commercially for the assessment of testicular volume[2] but still prader's orchidometer is the most commonly used orchidometer in clinical practice.

C). Timing of placement of testicular prosthesis:

The timing of insertion of a testicular prosthesis in a child is not straightforward. The psychological impact of an absent testicle in a child or adolescent is a good reason to consider prosthesis placement at the time of the initial surgery for a cryptorchid testis. As the size is the limitation in this case it may necessitate further surgery to insert a larger prosthesis when the child gets older. However, if a child is satisfied with the size of the original prosthesis, further surgery can be avoided. An alternative strategy is to delay the placement of the definitive prosthesis until the child reaches adolescence. Prosthesis placement at a later age has a limitation that due to the prolonged cryptorchid state scrotal hypoplasia renders further prosthesis placement difficult ultimately leading to the placement of prosthesis of smaller size. This is apart from the psychological trauma of prolonged state of anorchia. Methods for increasing scrotal space for the placement of a testicular prosthesis include the use of tissue expanders such as a silicone balloon attached to a filling port[3] or a Foley catheter balloon.[4] It is thus accepted that a testicular prosthesis should be placed at the time of initial surgery and changed to larger size at a later age if required.

D). Procedure of placement of testicular prosthesis:

Of historical interest, intracapsular insertion of a testicular prosthesis following subscapsular orchidectomy using a scrotal incision in patients with advanced prostate cancer was first described by Tolson in 1944 and endorsed as recently as 1984.[5] In 1972, Abbassian[6] described the insertion of a testicular prosthesis in a subcuticular pouch which was said to be useful in patients with extensive atrophy and scarring of the scrotal area. A skin incision was made in the opposite hemi-scrotum ensuring not to cross the midline raphe. Through this incision, a subcuticular pouch was created for the prosthesis in the empty hemi-scrotum. However, this procedure was associated with a high incidence of prosthesis extrusion. To minimise the risk of extrusion of the prosthesis, Latimmer[7] advocated a high scrotal or low inguinal incision, anchoring the prosthesis to the bottom of the scrotum and narrowing the upper scrotum with additional sutures. This technique was difficult to perform in the presence of a contracted or scarred hemiscrotum. In such circumstances, an appropriate space may be created using a sponge-holding forceps or by using the balloon of a Foley catheter.[4] Fortunately such situations are less seen in paediatric urology. Currently, most paediatric surgeons use a low groin incision whenever possible to implant a testicular prosthesis. This is associated with a lower risk of infection and extrusion.[8,9] A finger is then placed into the scrotal sac and the potential space created. The most dependant part of the scrotum is subsequently inverted and the prosthesis secured with a PDS suture placed through its suture loop. During transfixation of the dartos, particular care must be taken to avoid skin penetration and, thereby, promote infection and possible extrusion of the prosthesis. All prostheses are generally placed surgically on an outpatient basis. Prior to placement all patients should receive perioperative intravenous antibiotics (surgeon choice) and thorough skin preparation with an iodine or chlorhexideine based scrub. The prosthetic device is bathed in antibiotic solution and filled through the self-sealing injection port with normal saline (0.9%) with displacement of all air until the softest possible fluid consistency is achieved without dimpling of the prosthetic wall.

E). Complication of testicular prosthesis insertion:

Marshall[10] reviewed the records of over 2500 testicular prosthetic implantations to establish a list of postoperative complications and their incidence (Table 1). Prosthesis extrusion, the commonest complication, mainly occurred in patients following orchidectomy for epididymo-orchitis, especially if a scrotal incision had been used to implant the device. Marshall also noted that previous scrotal surgery and a long lag time between orchidectomy and the insertion of the prosthesis increased the risk of developing complications. There has been a case report of spontaneous rupture of a silicone testicular prosthesis 11 years after its insertion.[11] The spread of silicone to inguinal lymph nodes is also documented in a case report[12] but, as mentioned previously, there is no evidence of autoimmune disease or malignancy developing following testicular prosthesis implantation. Turek *at el*[13] also reviewed their series of testicular prosthesis for complication and noted a complication rate as shown in table1. In current practice, the most common postoperative complaints concern body image, namely that

Table 1. Observed complications with testicular prosthesis

Complication	Marshal *et al*[10]	Turek *et al*[13]
Extrusion	3-8%	2%
Scrotal Contraction	3-5%	2%
Pain	1-3%	9%
Hematoma	0.3-3%	1%
Infection	0.6-2%	Nil

the device is incorrectly sized/shaped or that it is too high in the hemi-scrotum.[14]

F). Ideal testicular prosthesis:

Ideal testicular prosthesis is one which is chemically inert and does not elicit any inflammatory or hypersensitivity reaction. The material should also resist mechanical strains, take and hold the desired form, be amenable to sterilization and be a proven non-carcinogen.

G). Evolution and currently available options:

Multiple materials have been used for testicular prosthesis. The first testicular prosthetic device was implanted in 1939 by Bowers using the metal alloy vitalium. Vitalium was an alloy composed of cobalt, chromium and molybdenum.[15] In 1943; testicular prostheses made of Lucite were available in a range of sizes.[16] During the 1950s, numerous other materials, including glass marbles, were used.[17] Gelfoam was also used by specifically injecting it into the tunica albuginea following intracapsular orchidectomy performed on patients with metastatic prostate cancer.[18] Plexiglass, Dacron and polyethylene prostheses have also been used without much success. Subsequently in an effort to develop a more cosmetically acceptable prosthetic various materials have been used, including polymerized methyl ethacrylate, methacrylate and polyurethane foam. A dramatic improvement in device consistency was achieved by Lattimer *et al* in 1973 with a silicone gel filled, silicone rubber prosthesis.[19] Used widely until 1995, manufacture of this device was discontinued because of emerging concerns over the association of silicone implants with connective tissue disease.[20,21,22] As a result, silastic and solid silicone rubber prostheses were developed for use in the 1960s.[23] Currently firmer, silicone-coated saline filled product has become the standard prosthesis since 1988.

H). Currently used prosthesis:

There are four companies (Nagor Ltd, Douglas, Isle of Man, UK; Mentor Medical Systems Ltd, Wantage, Oxon UK; Osteotec Plastic Surgery, Dorset, UK; and Silimed, Dieburg, Germany) that supply the majority of testicular prostheses. Osteotec Plastic Surgery supplies the Perthese prosthesis. Nagor prostheses are produced as silicone-gel filled and elastomer versions whereas the Silimed implant is only available in the elastomer version, which has a more solid consistency. The Perthese implant is produced in the gel-filled version; however, Mentor Medical Systems provides a re-inforced silicone elastomer version called the Soft-Solid Testicular Prosthesis (SSTP). They also provide a saline-filled prosthesis which has recently received FDA approval and is the only licensed testicular prosthesis available for common usage in the US. The weight, shape and texture of the Mentor SSTP is designed to approximate the normal testicle and is only licensed for investigational purposes in North America. The company is currently conducting a clinical study to evaluate the safety of its SSTP. A brief description of these prostheses is as under.[24]

a). The Coloplast (formally Mentor) Saline Filled Testicular Prosthesis: This device is about the same weight, shape and softness of a normal testicle. It comes in four sizes-extra-small, small, medium and large. The implant is made of a molded silicone elastomer shell that is approximately 0.035 inches thick. It is not visible on X-ray. The device is filled with saline at the time of surgery and just prior to implantation. It includes a self-sealing injection site at one end that allows for filling with a sterile saline solution. On the opposite end of the implant is a silicone elastomer tab that enables suturing and securing the implant into a set position, if this is desired. The average cost of one prosthesis ranges from $ 3000- 3500.

b). The Coloplast (formally Mentor) Soft-Solid Testicular Prosthesis. This is not US FDA approved, thus is only available in certain countries outside the US. The SSTP is made in five sizes: Extra-small, Small, Medium, Large, Extra-large. The device consists of a molded silicone elastomer shell, ranging from 0.012-0.018 inches thick, filled with cured silicone elastomer. A silicone elastomer Dacron reinforced patch for suturing the prosthesis in position is located at one end of the device shell. This is indicated for cosmetic testicular replacement when the natural testicle has been removed. The weight, shape, and texture of Mentor soft-solid testicular implants is designed to approximate normal testicles, providing patients with a more natural looking and feeling scrotum. They are intended to aid in the restoration of a normal physical appearance for male patients of all ages with one or more missing testicles.

c). Sientra's (formally Silimed) Silicone Elastomer Implant aka Oval Carving Block: The Sientra's oval carving block is composed of an envelope made of chemically and mechanically resistant silicone elastomer which is thin, soft, smooth of surface and contains a certain amount of elastomer whose shape, density and overall consistency have been chosen to make it as similar as possible to the shape and feeling of the human testis it replaces. All materials used are medical grade and proven to be biocompatible (is safe and tolerated well by the body). The silicone envelope membrane is made of a compound of dimethyl polysiloxane and dimethyl fluoro silicone copolymer. The silicone envelope is filled with an elastomer mixture of reinforced dimethyl methylvinyl siloxanes with reinforced dimethyl methylhydrogen siloxanes. Applied Silicone Corporation manufactures the material. Silimed's oval carving block Silicone Implants are available in 5 sizes. Sientra's oval carving block implant is pending approval by the FDA.

d). Custom made testicular prostheses: These are cheaper alternative to the commercially available prostheses. These are made of Teflon which is custom carved into the desired shape, autoclaved and implanted. The disadvantage is the hard feel which is non-physiological.

I). Experimental promising alternatives:

a). Tissue engineered testicular prosthesis with internal support:[25] In an experimental study conducted by Zhongguo Xiu *et al*[25] the chondrocytes were isolated from the swine articular. The PGA scaffold was incorporated with medpor, of which semi diameters were 6 mm and 4 mm respectively. Then, the chondrocytes (5 x 10 7 /ml) were seeded onto Medpor-PGA scaffold and cultured for 2 weeks. The cell-scaffold construct was implanted into subcutaneous pockets on the back of nude mice. Mice were sacrificed to harvest the newly formed cartilage prosthesis after 8 weeks. Macroscopy, histology and immunohistochemistry observations were made. The gross observation showed that on changes were in shape and at size, the color and elasticity were similar to that of normal cartilage and that the cartilage integrated with Medpor in the experimental group. The newly formed complex of Medpor-PGA and cells was very similar to testicle in gross view and to normal cartilage in histology. This pilot technique of creating testicular prosthesis by incorporating tissue-engineered cartilage with Medpor demonstrated success. This however requires a long journey to be practically applicable.

b). Tissue engineered testicular prostheses with prolonged testosterone release:[26] In an experimental study by Raya-Rivera *et al*[26] chondrocytes, harvested from bovine articular cartilage, were seeded on testicular shaped polymer scaffolds at a concentration of 100 x 106 per ml. The scaffolds were maintained in a bioreactor for 4 weeks to form cartilage tissue. Subsequently, testosterone enanthate (100 microgram) was injected into the central hollow space of each testicular prosthesis, and maintained for 40 weeks in culture. A sample of the medium was collected every 2 days for testosterone assays. Another group of ex vivo engineered testicular prostheses was implanted into the scrotal space of castrated athymic mice. Intratesticular injection of testosterone enanthate was made into each prosthesis at a concentration of 100 microgram. Control groups consisted of animals with castration only and sham operations. Testosterone levels were measured prior and 2 weeks after castration, 1 day after testosterone administration, and weekly up to 14 weeks. The engineered testicular prostheses were retrieved at sacrifice for histomorphological and immunocytochemical analyses. They found that ex vivo prostheses showed an initial burst effect of testosterone followed by a broad plateau for 16 weeks (>500 ng/dl) and a decreased level of testosterone until 40 weeks. The testosterone levels were physiologic throughout 40 weeks and the entire testosterone released was calculated as 60% of the injected volume. The circulating testosterone levels in the prostheses implanted animals demonstrated a maximum peak on day 1 and a continued physiologic range during the entire study period. Histologically, the retrieved testicular implants showed mature chondrocytes with a hollow center in each prosthesis. This study demonstrates that engineered cartilage testis can be created in bioreactors, can be implanted in vivo, and can release testosterone for a prolonged period. Furthermore, the levels of testosterone release can be maintained within the physiologic range. Periodic reinjection may potentially provide permanent physiologic hormonal replacement. This novel technology may be beneficial for patients who require testicular prostheses and chronic hormone supplementation. However, this needs to be tried in humans.

Conclusions

A number of available options are available. There are only few which are FDA approved but are still being used. Most of them have a limitation in developing countries due to their high cost thus making custom made Teflon prosthesis as an available alternative. There are many promising experimental studies being conducted that may change the management protocol in near future.

References

1. Prader A. "Testicular size: Assessment and clinical importance". Triangle 1966;7:240-3.
2. Takihara H, Sakatoku J, Fujii M, Nasu T, Cosentino MJ, Cockett AT. Significance of testicular size measurement in andrology. I. A new orchiometer and its clinical application. Fertil Steril 1983;39:836-40.
3. Lattimer JK, Stalnecker MC. Tissue expansion of underdeveloped scrotum to accommodate large testicular prosthesis. A technique. Urology 1989;33:6-9.
4. Chadha A, Saraiya H. Scrotal reconstruction using Foley catheters as tissue expanders. Ann Plast Surg 1991;26:291-2.
5. Short KL, Howerton LW, Holt H, Amin M. Subcapsular orchiectomy with intracapsular testicular prosthesis for metastatic prostate carcinoma. Urology 1984;24:38-40.

6. Abbassian A. A new surgical technique for testicular implantation. J Urol 1972;107:618.
7. Lattimer JK, Vakili BF, Smith AM, Morishima A. A natural-feeling testicular prosthesis. J Urol 1973;110:81-3.
8. Lawrentschuk N, Webb DR. Inserting testicular prostheses: a new surgical technique for difficult cases. BJU Int 2005;95:1111-4.
9. Simms MS, Huq S, Mellon JK. Testicular prostheses: a new technique for insertion. BJU Int 2004;93:179.
10. Marshall S. Potential problems with testicular prostheses. Urology 1986;28:388-90.
11. John TT, Fordham MV. Spontaneous rupture of testicular prosthesis with external leakage of silicone - a rare event. J Urol 2003;170:1306.
12. Doherty AP, Mannion EM, Moss J, Ockrim JL, Christmas TJ. Spread of silicone to inguinal lymph nodes from a leaking testicular prosthesis: a cause for chronic fatigue? BJU Int 2000;86:1090.
13. Turek PJ, Master VA. Safety and effectiveness of a new saline filled Testicular prosthesis. The journal of urology 2004;172:1427-30.
14. Adshead J, Khoubehi B, Wood J, Rustin G. Testicular implants and patient satisfaction: a questionnaire-based study of men after orchidectomy for testicular cancer. BJU Int 2001;88:559-62.
15. Girdansky J, Newman HF. Use of a vitallum testicular implant. Am J Surg 1941;53:514.
16. Rea CE. The use of testicular prosthesis made of lucite with a note concerning the size of the testis at different ages. J Urol 1943;49.
17. Hazzard CT. The development of a new testicular prosthesis. J Urol 1953;70:959-60.
18. Baumrucker GO. Testicular prosthesis for an intracapsular orchiectomy. J Urol 1957;77:756-8.
19. Lattimer JK. Vakili BF. Smith AM, Morishima A. A natural-feeling testicular prosthesis. J Urol 1973;110:81-3.
20. Van Nunen SA, Gatenby PA, Basten A. Postmammoplasty connective tissue disease. Arthritis Rheum 1982;25:694-7.
21. Henderson J, Culkin D, Mata J, Wilson M, Venable D. Analysis of immunological alterations associated with testicular prostheses. J Urol 1995;154:1748-51.
22. Pidutti R, Morales A. Silicone gel-filled testicular prosthesis and systemic disease. Urology 1993; 42:155-7.
23. Lattimer JK, Vakili BF, Smith AM, Morishima A. A natural-feeling testicular prosthesis. J Urol 1973;110:81-3
24. Bukowski TP. Testicular Prostheses. In: Urologic Prostheses: The Complete Practical Guide to Devices, Their Implantation, and Patient Follow up. Carson III CC. (ed), Humana Press, Totowa, New Jersey, 2002:141-54.
25. Wu Y, Jiang H, Zhou G. Experimental study on tissue engineered testicular prosthesis with internal support. Zhongguo Xiu Fu Chong Jian Wai Ke Za Zhi. 2007;21:1243-6.
26. Raya-Rivera AM, Baez C, Atala A, Yoo JJ. Tissue engineered testicular prostheses with prolonged testosterone release. World J Urol 2008;26:351-8.

Review Article

Prune Belly Syndrome: Errors in Management and Complications of Treatment

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Abstract: Prune Belly Syndrome is a condition in which the abdominal wall, urinary tract and testes are affected resulting in a severely curtailed life span. With proper management these patients could have a normal, productive life.

Key words: Abdominal muscular dystrophy, Prune Belly Syndrome, Urinary tract anomalies

Abbreviations: CIC-Clean intermittent bladder catheterization, IVP-Intravenous pyelogram, PBS-Prune Belly Syndrome, MAG3-Tc99m-mercaptoglycine, PUV-Posterior urethral valves, UTI-Urinary tract infection, VCUG-Voiding cystourethrogram, VUR-Vesicoureteric reflux

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Introduction

Prune Belly Syndrome (PBS) is a congenital anomaly that, by definition, occurs only in males. These boys have abdominal muscular dystrophy (prune belly), urinary tract anomalies and intraabdominal testes. Although the basic combination of anomalies is well defined, their severity varies considerably from patient to patient. The abdominal wall defect may range anywhere from a mild "pot belly" to an apparent total lack of abdominal wall musculature with intervening presentations that may involve either the entire abdominal wall or parts of it. The urinary tract may be obstructed, there may be vesicoureteric reflux (VUR) or only urinary stasis. Similarly, the severity of involvement of the kidneys, ureters, bladder and urethra demonstrate a very wide spectrum. In addition, since these patients have a "bell-shaped" chest with a flattened diaphragm they have an ineffective cough thus making them susceptible to respiratory infections. Their intestines are suspended in the abdomen from a universal mesentery, which may make them susceptible to volvulus. Anorectal anomalies, spinal defects, compression defects of the lower extremities are other known associations. Finally, there are patients with an incomplete form of the syndrome, also known as "Pseudoprunes". This group consists of:

1. Females with typical abdominal wall and urinary tract abnormalities.
2. Males or females with abdominal muscular dystrophy without urinary tract involvement.
3. Males or females with typical urinary tract findings and a normal abdominal wall.

It is thus apparent from all these variations that there is considerable scope for misdiagnosis or incorrect intervention.

Our experience in managing 37 children with PBS has provided us with some insight into their management.

Historical Errors

In the 1950s many authors, incorrectly equated urinary tract dilation with obstruction and, advocated early, intubated urinary diversion.[1,2] Use of tubes for urinary diversion invariably leads to the expected problems of dislodgment or obstruction of the tube and urinary infections due to this indwelling foreign body. The belief that PBS urinary tracts were obstructed, resulted in the next step in management, in the 1970s, when early complete reconstruction of the urinary tracts of all PBS patients was advocated.[3,4] At the same time others were coming to the exact opposite conclusion that minimal or no interference was the way to go.[5,6]

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Welch and Kearney[7] made the first attempt at classifying these patients into therapeutic groups. A better classification was subsequently proposed by Berdon *et al*.[8] In our hands, the latter classification, with a minor modification of our own, provides the best guideline for managing these patients.

Classification of Berdon *et al*:[8]

They placed patients into three groups:

Group I: Potter's syndrome (oligohydramnios)

Group II: Severe neonatal and infantile urinary tract involvement

Group III: Mild involvement

Group I patients have severe pulmonary hypoplasia and renal dysplasia at birth. Their limbs often demonstrate the effects of intrauterine compression. They die at birth, usually of respiratory failure. Group II patients also have severe respiratory and urinary tract problems, however, both systems are not so severely affected that the child succumbs at birth. Group III patients typically present because of the abnormal appearance of the abdominal wall or for undescended testes. Imaging demonstrates a typically abnormal urinary tract.

Berdon's classification[8] suggests that all patients in Group II require surgical correction. That is not our experience. We further subdivide Group II into three subgroups depending upon whether they have urinary obstruction, VUR or stasis. Patients with urinary obstruction invariably require urinary tract reconstruction, those with VUR require aggressive medical management and often surgical reconstruction is eventually required, whereas those with stasis either do not require surgical intervention or, at most, their urinary drainage is improved by bladder domectomy (to improve detrusor function) and abdominoplasty (to enable a more effective Valsalva maneuver). In our view the difference between Group II stasis and Group III patients lies in the fact that the latter never presents with urinary tract infections or deteriorating renal functions.

Errors in Antenatal Diagnosis and Therapy

Since antenatal ultrasonography became universally available in the 1980s PBS has often been diagnosed and also misdiagnosed as posterior urethral valves (PUV). Initially unnecessary antenatal interventions occurred in many PBS babies since ultrasonographers were unable to distinguish them from PUV. Hydronephrosis, megaureters and a thick-walled bladder are seen in both conditions, however, there are differences. In PBS the abdominal wall tends to be lax and the urinary bladder is massive, whereas in posterior urethral valves the abdominal wall may be distended but it is not lax and the thick walled bladder is small. Finally, the posterior urethra in PBS is dilated, elongated and "heart-shaped" while in PUV the bladder and urethra have a "key-hole" or "pear-shaped" appearance. Subsequently, even though PBS was diagnosed in utero vesicoamniotic shunts were placed because of the erroneous impression that the urinary tract was obstructed. Shunting did not improve their status.[9]

Errors in Initial Evaluation and Management

When a baby is born with PBS, the initial focus should be in determining his respiratory capacity and administering respiratory support as required. A Group I patient will probably succumb despite treatment whereas a Group III patient will require minimal, if any, supportive care. Group II patients will respond to intensive respiratory care. The appearance of the abdominal wall does not correlate at all with the severity of urinary tract abnormalities (Fig. 1 and Fig. 2). Potter's facies,

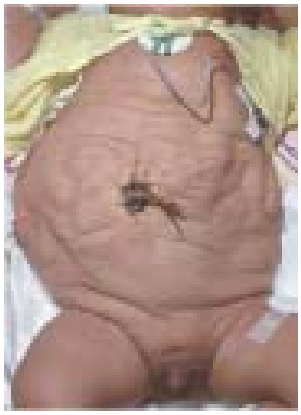

Fig. 1. Evaluation of this child with severe lack of abdominal musculature revealed only urinary stasis. All he required was a bladder domectomy along with orchidopexy and abdominoplasty. Note the bell-shaped chest

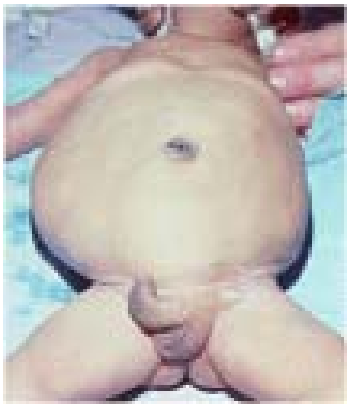

Fig. 2. This patient with very mild abdominal wall involvement had severe bilateral ureteric obstruction and borderline renal functions. Early cutaneous pyelostomies were carried out followed by urinary tract reconstruction. By 9 years of age he required a renal transplant. It is to be noted that he has a bell-shaped chest and a megalourethra. There is a known association between PBS and megalourethra

a bell-shaped chest and compression deformities of the lower extremities all indicate maternal oligohydramnios due to poor renal function or urinary outflow obstruction. Urinary ascites and/or a patent urachus occur due to lower urinary tract obstruction. These obstructed patients require urinary drainage in addition to respiratory support. Urinary ascites is aspirated to permit improvement in respiratory functions. Fortunately, leak from the collecting systems which causes urinary ascites helps maintain low intra-renal pressures, thus protecting the kidneys from further damage. These children improve with adequate drainage of the urinary tract. If patent, the urachus is a convenient site to insert a large catheter for drainage. In this initial stage evaluation of the urinary tract should be confined to blood tests and ultrasonography. In all other patients, unless it is necessary to measure urine output, it is best to avoid catheterizing the neonate for fear of precipitating a urinary tract infection (UTI). Radiographic and nuclear studies should be delayed until the child is stable and they must be evaluated with care since they always demonstrate a dilated urinary tract that retains contrast for long periods of time. Similarly, renal function studies also do not give the true picture. Furthermore, during follow-up, changes in contrast studies and blood tests may lag behind true functional deterioration. In our experience the child's clinical status with regard to appetite, general well-being, somatic growth and development and control of UTIs are much more sensitive indicators of deteriorating renal function than radiographic or chemical tests.

At this early stage we feel that all PBS patients should be placed on urinary suppressive antibiotics as we do not know what their eventual status would be. Initially we like to use an antibiotic from the penicillin group. After the baby is 6 weeks old a sulfa drug or nitrofurantoin may be used. Nitrofurantoin is an excellent drug, however, children often do not like its taste.

Errors in Management After the Neonatal Period

Severity of involvement in these children becomes apparent after the first few weeks. By then Group I patients have succumbed to respiratory failure. All other patients tend to stabilize. The urinary tract should now be evaluated with the child on antibiotics to prevent urinary infection after instrumentation. We prefer a Tc99m-mercaptotriglycine (MAG3) renal scan since in a single test we obtain information regarding the amount of functional renal tissue and outflow obstruction. The test must always be done with a draining bladder catheter in place to minimize confusion caused by urinary stasis and/or VUR. An intravenous pyelogram (IVP), also with a draining bladder catheter, would also give the same information regarding renal concentrating ability and drainage but one is better able to quantify the results of a renal scan (Fig. 3). It is essential to remember that the renal scan and IVP are guidelines for management and no decision should be made based solely upon the tests. Invariably, a borderline obstructive pattern will be apparent in all PBS patients even after administration of diuretics since the ureters and bladder have such a great capacity. A radiographic voiding cystourethrogram (VCUG) is valuable in determining the presence of vesicoureteral reflux (VUR) (Fig. 4), the shape and emptying capacity of the bladder, the appearance of the urethra and the presence of urethral obstruction. Approximately 70% of PBS patients have VUR and they should stay on suppressive antibiotics. The bladder often has an hourglass shape with a urachal cap, which traps urine when the detrusor contracts (Fig. 5). The posterior urethra is "heart-shaped" and there is an area at its junction with the bulbous urethra where it narrows suddenly. This narrowing is usually insignificant, however, it could form a urethral ring that requires division or one may find Type IV valves.[10]

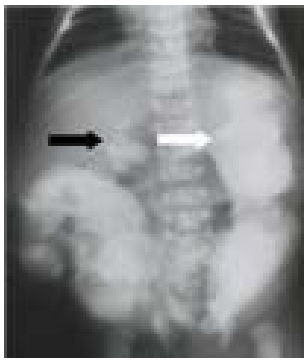

Fig. 3. The massively dilated ureters visible on this intravenous pyelogram were indeed obstructed at the lower end. However, such an appearance is also possible when only stasis is present. In this case the left kidney is extremely hydronephrotic and dysmorphic (white arrow) while the right kidney (black arrow) is only minimally involved. The ureters often absorb back-pressure in an obstructed system, thus preserving renal function. This patient required complete urinary reconstruction

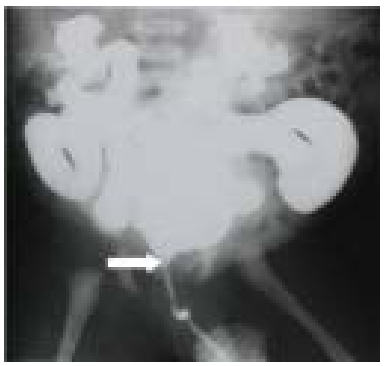

Fig. 4. Massive bilateral VUR is demonstrated in this VCUG. This child had complete urinary reconstruction at 6 weeks of age. White arrow points to the narrow junction between the posterior and the bulbous urethra

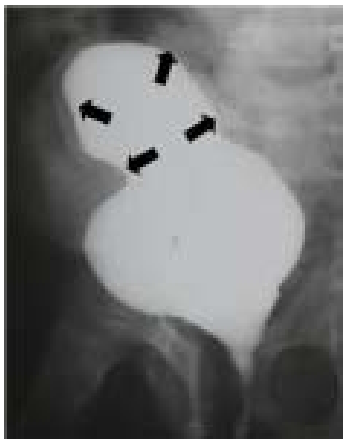

Fig. 5. VCUG demonstrating an hourglass shaped bladder due to a large urachal diverticulum (black arrows)

Once the urinary tract has been evaluated, Group II VUR should be left on suppressive antibiotics. Their follow-up consists of monthly urinalyses and urine cultures to maintain sterile urine. In addition, renal ultrasonography every 3 months or so is valuable in evaluating dilation of the urinary tract, as it demonstrates efficacy of drainage. Patients with stasis alone are typically placed on suppressive antibiotics initially. Antibiotics are discontinued if random monthly urine cultures on three occasions are sterile. They are then followed clinically and if they continue to thrive, they will be placed in Group III. If urinary infection recurs, the child is considered to be Group II stasis and consideration is given to determining whether bladder drainage is adequate. On occasion, clean intermittent bladder catheterization (CIC) is helpful in this group. Group II patients with urinary obstruction have the worst prognosis. They go into renal failure early and they are also particularly susceptible to UTIs. In our experience, keeping a close watch on their general well-being is the best way to catch changes early. If the obstruction is worsening, the child will not thrive and the ultrasound will demonstrate further dilation of the urinary tract. Renal scans and blood chemistries are carried out but they are slow in identifying deterioration. Group II obstructed patients invariably require surgical reconstruction.

Recurrent UTIs are common in PBS patients. Infections can be difficult to clear since the tortuous ureters form pockets of pus. We failed to diagnose a urinary infection in one of our Group II patients. He died of overwhelming sepsis, from resistant E.Coli, 48 hours after total urinary reconstruction. Another Group II patient presented semicomatose with high serum ammonia levels suggestive of Reyes syndrome. On further evaluation it became apparent that the culprit was an ammonia-producing organism in the urinary tract. Finally, one has to maintain a careful balance because clearing pathogens from a urinary tract in which stasis is always present often results in overgrowth of yeast. On occasion the patient may go back and forth between growth of urinary pathogens and yeast.

Errors in Decisions Pertaining to Urinary Diversion

Intubated nephrostomy or pyelostomy: We have already pointed out that the earlier view that every PBS patient is obstructed and requires early urinary diversion is clearly faulty. In addition a nephrostomy or pyelostomy tube tends to get dislodged, kinked, plugged with debris or it becomes the source of infection. Intubated diversions are only indicated as an emergency and for the short-term to resolve an acute problem. They are best performed percutaneously under radiological guidance. The smallest pigtail catheter that would adequately drain the kidney should be placed in a lower pole calyx. If possible, the catheter should be inserted through a part of renal cortex that has the least parenchyma.

Ureterostomies: Ureterostomies are mentioned to be condemned. First, stasis is not avoided since urine still has to pass through these dilated aperistaltic proximal ureters. Secondly, urine has to drain against gravity when ureterostomies are brought out on the abdominal wall (Fig. 6) particularly in infants who spend most of the time on their backs. Thirdly, ureterostomy stomal stenosis is very common. Finally, the longitudinal blood supply of the ureter is placed in jeopardy and the distal ureter may be devascularized when a ureterostomy is taken down.

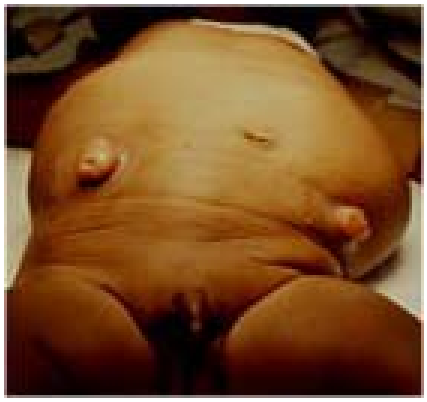

Fig. 6. Bilateral loop cutaneous ureterostomies did not improve drainage in the dilated aperistaltic ureters in this boy because the location of the ureterostomies required anti-gravity drainage of urine. He responded well to total urinary reconstruction

Internal Stents: Internal stents are only of value for obstruction at the ureteropelvic junction or upper ureter and that is rarely, if ever, the case in PBS. Furthermore, stents create free reflux from the bladder, thus compounding stasis and since they are foreign bodies they increase the chance of UTI.

Vesicostomy: A vesicostomy also serves no purpose since urine still has to traverse the full length of the atonic ureters. It does permit continuous drainage from the bladder, but high intravesical pressure is not an issue in these children. In addition, these large, flabby bladders tend to prolapse through the stoma even if a Blocksom vesicostomy[11] is carried out. A suprapubic cystostomy is even worse since the tube is prone to dislodgment and obstruction in addition to being a source of infection.

Perineal urethrostomy: A perineal urethrostomy is never indicated, as it does not bypass any part of the urinary tract.

Indications & Techniques of Urinary Diversion

The only indication for emergent urinary diversion in PBS is the child who develops a severe urinary infection that is not responding to treatment. In such a situation a percutaneous nephrostomy placed under radiographic guidance is the least invasive technique and is easily reversed upon removal of the catheter. While the catheter is in place it carries the already mentioned risks of infection, dislodgment, and obstruction by debris.

Diversion for an extended period is indicated if the kidney has very limited function and one is trying to delay its eventual failure. One or both kidneys can be diverted. A high diversion under such conditions also permits accurate evaluation of individual renal functions.

We prefer cutaneous pyelostomy (pyelocutaneous anastomosis) for diversion. Exteriorization of the renal pelvis provides the shortest and most direct route for egress of urine, and it does not affect the blood supply of the distal ureter. A cutaneous pyelostomy is not without complications. The dysplastic kidney with thin parenchyma can turn inside out and prolapse through the stoma. The stoma could also be the route for an ascending infection and it is difficult to adequately place a device to catch the urine so close to the XII rib and spine. Parents have to place a second diaper around the waist to catch the urine. If bilateral diversions are carried out in the face of deteriorating renal functions the patient is undiverted prior to renal transplantation.

The cutaneous pyelostomy is carried out with the patient prone or in the lateral position. A 1cm transverse incision is made at the angle of the XII rib with the sacrospinalis muscle. The incision is deepened through all muscle layers. The highly mobile kidney can be manually pushed toward the incision and the posterior aspect of the renal pelvis is grasped and delivered out of the wound. A 1cm transverse incision is made on the posterior surface of the renal pelvis and the cut edges of the pelvis are sutured with interrupted absorbable sutures to the full thickness of the skin. It is worth repeating that the incisions in the abdominal wall and the renal pelvis must both be no more than 1cm in length to avoid renal prolapse.

Total Urinary Tract Reconstruction

Indications: Past errors in management have already been discussed. In our experience, all Group II patients with obstruction and a majority of those with VUR require complete urinary tract reconstruction. Patients with an obstructed urinary tract typically become candidates for reconstruction within the first year of life. Occasionally, if their renal function is borderline, a preliminary cutaneous pyelostomy in preparation for reconstruction may be of value. Patients with VUR should initially be managed with suppressive antibiotics, awaiting spontaneous resolution of VUR by two to three years of age. However, if recurrent or persistent UTIs occur or VUR does not resolve, reconstruction is indicated.

Surgical Technique: If the urinary tract is reconstructed, simultaneous abdominoplasty and orchidopexy are carried out but they will be discussed separately. Fig. 7 illustrates the typical appearance of the urinary tract in PBS. The ureters are most tortuous and atonic in their lower halves hence, they are disconnected from the bladder. Any extra length is excised at the lower end and, if necessary, the upper ureters are straightened out. The ureters are reimplanted in the bladder after tapering. During ureteral mobilization it is essential that the peritoneum and periureteric adventitia on its medial aspect be pushed towards the ureter to maintain the blood supply. If a ureterostomy is being taken down, one must take extra pains to avoid damaging the longitudinal blood supply on the medial aspect of the ureter. The ureters are tapered by excising tissue on the lateral aspect and

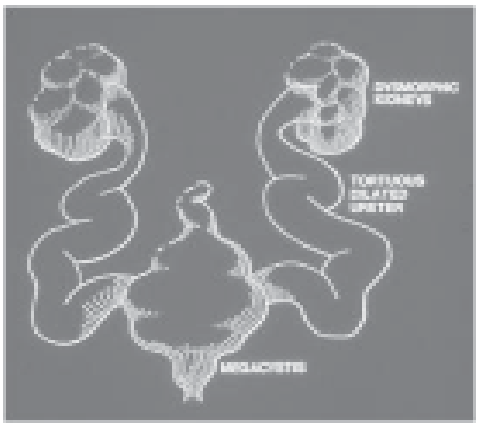

Fig. 7. Typical appearance of the urinary tract in PBS. The kidneys are dysmorphic and hydronephrotic. Renal dysplasia of varying degrees is evident on histologic examination. The ureters are large, elongated and tortuous, particularly at their lower ends. The bladder is large and thick walled with a wide bladder neck, and a urachal diverticulum. The prostatic urethra is "heart-shaped"

retubularization over a 10Fr. catheter. They are reimplanted in a 3-4cm cross-trigone submucosal tunnel. In these large low-pressure bladders reimplantation is relatively simple and results are excellent.

The bladder often has a urachal cap, which collects urine when the detrusor contracts. This cap is best removed by making a "fish-mouth" incision that runs in the anteroposterior direction. Once the cap is excised a spherical bladder with good detrusor function is obtained. Bladder domectomy is carried out at the beginning of the reconstruction and ureteral mobilization and reimplantation are carried out through this opening. It is important to remember that these children often produce large volumes of urine and it is crucial that an adequate bladder capacity be maintained (Fig. 8 and Fig. 9).

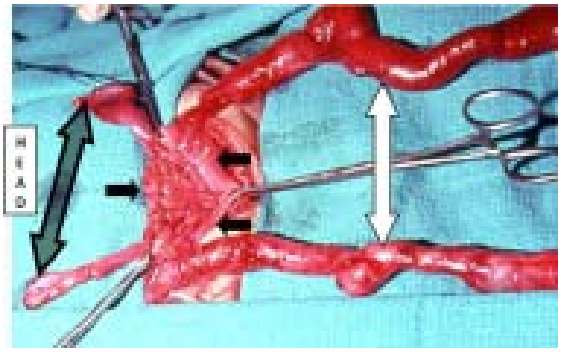

Fig. 8. Total urinary reconstruction. The patient's head is to the left. The megaureters have been dissected out and straightened (white arrow). The testes have been mobilized on the vessels of the vas deferens (shaded gray arrow). The dome of the bladder has been removed (small black arrows). The ureters will be shortened, tapered and reimplanted into the bladder which will be reconstructed so as to have a spherical shape. The testes will be brought down into the scrotum

In those rare instances when urethral outflow obstruction coexists, it is dealt with at the time of reconstruction. Valves should not be resected in a diverted or "dry" urinary tract lest a urethral stricture develops.

The ureters and bladder are stented for a two-week period after which the stents are removed if contrast studies demonstrate no leak from suture lines.

Orchidopexy

Invariably the testes are located intraabdominally, at the pelvic brim (Fig. 10). In Group II patients, orchidopexy is best carried out in conjunction with urinary reconstruction. In most instances the spermatic vessels are too short for the testes to reach the bottom of the scrotum. The spermatic vessels are divided close to the kidneys and mobilized with the blood

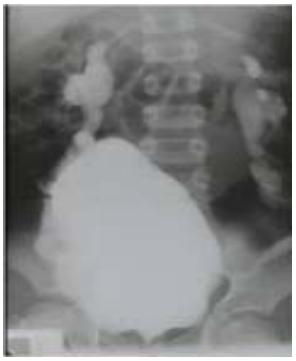

Fig. 9a

Fig. 9b

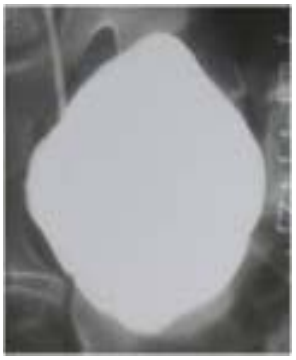

Fig. 9c

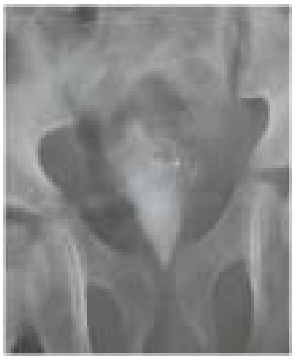

Fig. 9d

Fig. 9a. Intravenous pyelogram demonstrating a large bladder
Fig. 9b. Huge postvoid residue in the large bladder
Fig. 9c. Cystogram after bladder domectomy
Fig. 9d. Minimal postvoid residue after domectomy

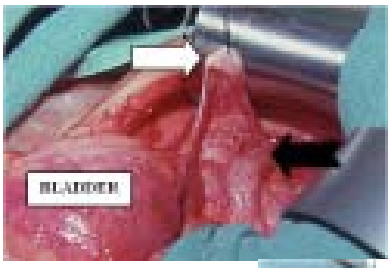

Fig. 10a

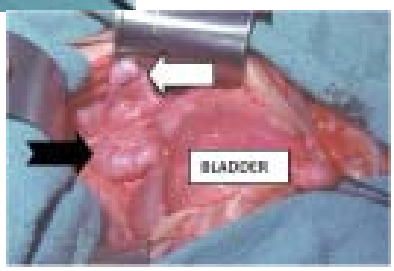

Fig. 10b

Fig. 10. Location of the intraabdominal testes at the pelvic brim. In both figures the bladder has been displaced downwards. The solid white arrows point to the testes. The notched black arrows demonstrate the megaureters
Fig. 10a. Right testis
Fig. 10b. Left testis

supply to the ureter. The testes are then brought down into 1the scrotum on the collateral blood supply from the vas deferens.[12] On occasion, in the infant, it may be possible to bring the testes down into the scrotum with the spermatic vessels intact.[4] Since there is no inguinal canal, an opening is made in the anterior abdominal wall over the pubic tubercle to develop the shortest possible route to the scrotum. The large bladder tends to get in the way and the opening has to be just lateral to it. Group III patients may require orchidopexy alone or in association with abdominoplasty. If abdominoplasty is planned, the orchidopexy is carried out through the same incision. If only orchidopexy is planned, the PBS bladder, which occupies the area of the lower abdomen behind the recti upto the umbilicus, has to be avoided (Fig. 11). We prefer bilateral transverse 3cm incisions extending anteriorly from the anterior superior iliac spines. Such incisions avoid the bladder, and place us directly above the testes. With this approach also, the spermatic vessels are divided and the testes are brought into the scrotum on the vessels from the vas, by the shortest route possible.

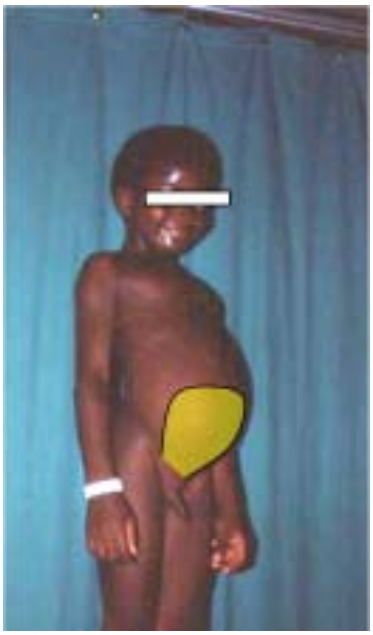

Fig. 11. category III patient with a large pot belly. The yellow structure outlined corresponds to the large bladder. The incisions extend anteriorly for 3 cm from the anterior superior iliac spine on either side

Abdominoplasty

Abdominoplasty is of great value to the majority of these patients. If the abdominal wall is tightened, respiratory functions, particularly coughing, and the ability to Valsalva to effectively evacuate feces and urine are improved. In addition we believe patients are better able to sit up from a supine position after abdominoplasty. Only a few patients in Group III, with a pot-belly, do not require abdominoplasty. Various operations have been described for abdominoplasty.[13-16] It is true that the infraumbilical rectus abdominis muscles demonstrate the greatest laxity, however, the deficiency may be worse on one or the other side, and it may extend to the supraumbilical recti and even the whole abdominal wall. If the defect is symmetrically located in the infraumbilical recti one obtains an excellent result from an abdominoplasty carried out through an infraumbilical "smile" incision (Fig. 12). On the other hand, the patient in figure 13 required a "reverse D" incision as his defect involved the supra- and infraumbilical portions of the right side of the abdomen while the left side was almost normal (Fig. 13). Another patient with an eccentric infraabdominal defect in which the "smile" incision was widened on the more affected side (Fig. 14). We believe that no single incision is adequate for all cases and it must be tailored to the defect. In our

Fig. 12a

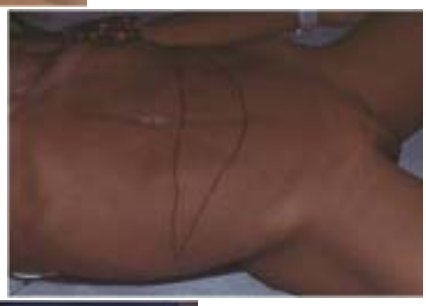

Fig. 12b

Fig. 12c

Fig. 12a. The patient's defect was essentially infraumbilical and symmetrical on both sides
Fig. 12b. The incision has been marked
Fig. 12c. The appearance of the abdominal wall one week after surgery

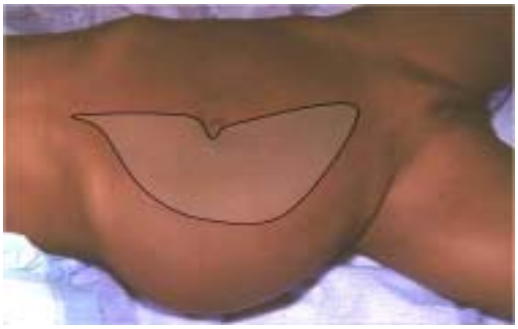

Fig. 13. This patient's major problem was in the upper and lower abdominal muscles on the right side. The reverse D shaped incision marked on the abdomen corrected the defect

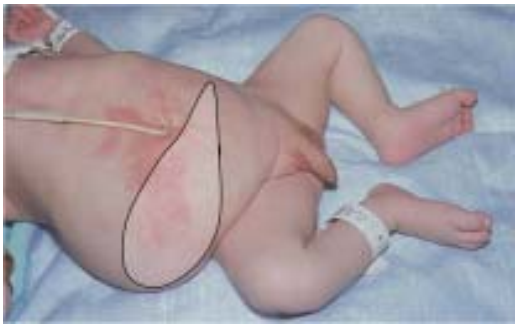

Fig. 14. This patient had an eccentric infraumbilical defect involving more of the right side hence the "smile" incision was widened in the right infraumbilical area as marked in the photograph

experience careful physical examination in which the patient is made to contract his abdominal muscles is all that is required to delineate the defect. Electromyography is unnecessary, expensive and it scares the child.[17]

Perioperative Management

It is essential that the entire team taking care of these children understand that that great care is required in their management. The patient's respiratory functions should be at their best prior to the operation and even the slightest respiratory infection should be grounds for postponement. The urine must also be sterile.

The immediate postoperative period is a critical time in which pulmonary infection and respiratory failure result in deaths. Vigorous pulmonary toilet and ventilator support are crucial at this time. Adequate postoperative pain control is essential for the patient to move, take deep breaths and cough. An epidural catheter maintained for 3-4 days is invaluable.

Conclusion

Prune belly syndrome is a complex previously lethal congenital anomaly. Numerous errors were made in the management of these patients in the past due to improper and inadequate understanding of its pathology. With careful and committed care these patients can go on to function very well in society.

References

1. Lattimer JK. Congenital deficiency of the abdominal musculature and associated genitourinary anomalies: a report of 22 cases. J Urol 1958;79:343-52.
2. McGovern JH, Marshall VF. Congenital deficiency of the abdominal musculature and obstructive uropathy. Surg Gynec Obstet 1959;108:289-300.
3. Randolph JG. Total surgical reconstruction for patients with abdominal muscular deficiency ("Prune-Belly") syndrome. J Pediatr Surg 1977;12;1033-43.
4. Woodard JR, Parrott TS. Reconstruction of the urinary tract in prune belly uropathy. J Urol 1978;119:824-8.
5. King LR. Idiopathic dilatation of the posterior urethra in boys without bladder outlet obstruction. J Urol 1969;102:783-7.
6. Snyder HM, Harrison NW, Whitfield HN, Williams INNES. Urodynamics in the prune belly syndrome. Brit J Urol 1976;48:663-70.
7. Welch KJ, Kearney GP. Abdominal musculature deficiency syndrome: prune belly. J Urol 1974;111:693-700.
8. Berdon WE, Baker DH, Wigger HJ, Blanc WA. The radiologic and pathologic spectrum of the prune belly syndrome. The importance of urethral obstruction in prognosis. Radiol Clin North Amer 1977;15:83-92.
9. Woodard JR. The impact of fetal diagnosis on the management of obstructive uropathy. Postgrad Med J 1990;66:37-43.
10. Stephens FD. Type IV valves in prune belly syndrome. Congenital malformations of the urinary tract. Chapter 5. Philadelphia PA, Praeger Press September 1983.
11. Blocksom BH Jr. Bladder pouch for prolonged tubeless cystostomy. J Urol 1957;78:398-401.
12. Fowler R Jr, Stephens FD. The role of testicular vascular anatomy in the salvage of high undescended testes. Austr NZ J Surg1959; 29:92-106.

13. Randolph J, Cavett C, Eng G. Abdominal wall reconstruction in the prune belly syndrome. J Pediatr Surg 1981;16;960-4.

14. Monfort G, Guys JM, Bocciardi A. A novel technique for reconstruction of the abdominal wall in prune belly syndrome. J Urol 1991;146;639-40.

15. Ehrlich RM, Lesavoy MA. Umbilicus preservation with total abdominal wall reconstruction in prune belly syndrome. Urology 1993;41:231-2.

16. Furness PD III, Cheng EY, Franco I, Firlit CF. The Prune-belly syndrome: A new and simplified technique of abdominal wall reconstruction. J Urol 1998;160:1195-7.

Review Article

Modern Management of Neurogenic Bladder: Making the Child the Focus of Therapeutic Efforts

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Abstract: The management of children with neurogenic bladder (NB) has remained a source of great dilemma for paediatric urologists since time immemorial. Several paths have been approached to reach the ultimate goal of upper tract preservation as well as to grant a socially acceptable life to these children and their families. Over the years, the 'proactive' approach of universal institution of therapy from the neonatal period onwards has gradually taken precedence over a 'reactive' approach, based on urodynamic parameters or the radiological sequelae. In this review we have summarized the various available options in the managment of neurogenic bladder.

Key words: Detrusor instability, Neurogenic bladder, Urodynamic parameters

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Introduction

The management of children with neurogenic bladder (NB) has remained a source of great dilemma for paediatric urologists since time immemorial. Several paths have been approached to reach the ultimate goal of upper tract preservation as well as to grant a socially acceptable life to these children and their families. Over the years, the 'proactive' approach of universal institution of therapy from the neonatal period onwards has gradually taken precedence over a 'reactive' approach, based on urodynamic parameters or the radiological sequelae.

Of the neuropathic and non-neuropathic causes of NB, spinal dysraphism is the most common aetiology, with myelomeningocele (MMC) leading with 80% incidence.[1] Other causes are uncommon in children including acquired cases occurring as a consequence of traumatic, oncologic or vascular pathology of the spinal cord, supraspinal and peripheral lesions and the non-neuropathic lesions.

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Conventionally, the etiology of the neuropathic bladder is divided into the following categories: central, peripheral or mixed. But this classification has very little role to play in therapeutic decision-making. Management is dictated by the basic state of the bladder after the neurological event:

- Detrusor overactivity with sphincter overactivity
- Detrusor inactivity with sphincter overactivity
- Detrusor overactivity with sphincter inactivity
- Detrusor inactivity with sphincter inactivity

A particular child tends to stick to the same category over the course of time owing to the unchanged primary pathology. Of the four categories, 'detrusor underactivity and sphincter underactivity' is inherently 'safe' in that, if untreated, the upper tracts remain preserved whereas 'detrusor overactivity with sphincter overactivity' which, when diagnosed, should sound alarm bells immediately lest the upper tracts get affected.[2]

Given the 20% incidence of death due to renal damage during the first year of life and the invariably dismal outcome of the disease, management should start as soon as a diagnosis is made.[3,4] For the individual patient, therapy is a lifelong requisite. Management starts with explaining the family about the cause, prognosis and ultimate goals of treatment so as to sensitise them adequately to get out of the 'shame and blame' game and accept the child and the problem.

Time is of essence regardless of the strategic approach instituted. The urologic sequelae of this condition are quite morbid. The apparent discrepancy between the treatment end-

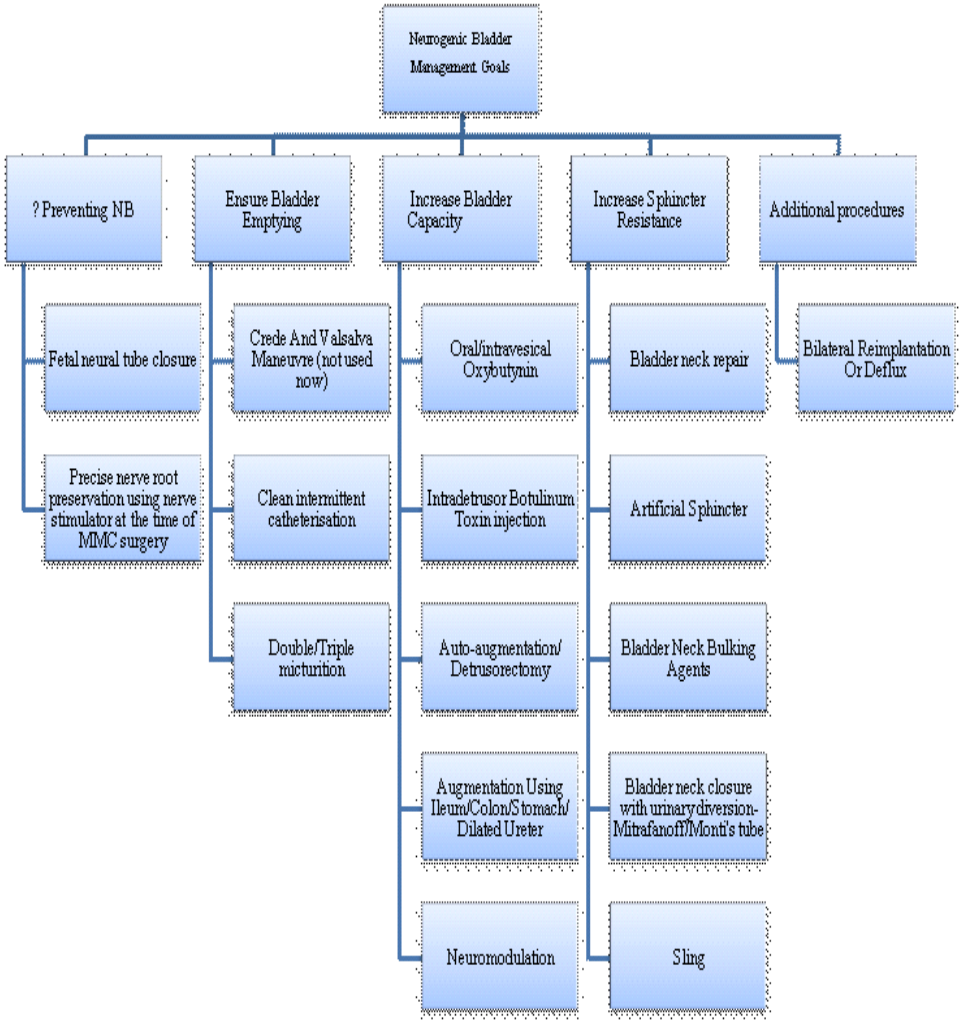
points of a doctor and a parent must be clarified before initiating treatment. The care-givers should realise that the doctor prioritizes the preservation of the upper tracts, while the doctor should realise that the social stigma of an incontinent child is nerve-wracking for any parent.

Approach to A Child with Neurogenic Bladder

Accurate evaluation and characterization of the neurological status of the lower urinary tract form the bedrock of treatment. This begins with a careful history of bladder and bowel habits. This includes questions to assess LUTS (lower urinary tract symptomatology) such as urinary frequency, urgency, intermittency, and incontinence or any history of UTIs. It is also necessary to assess whether the bladder evacuates completely after each void or if the child is simply overflowing urine from a chronically full bladder. A careful history of bowel habits is equally important and must address the frequency of defecation and the character of stool. The correlation of bladder and bowel dysfunction remains an enigma with some suggesting a local phenomenon due to rectal distension and others postulating a role of the interaction between the locus ceruleus and the pontine micturition centre.[5]

The physical exam should include a careful abdominal examination, palpating for any masses, such as due to fecoliths or retained urine. A careful genitourinary examination should confirm normal anatomy, as well as check for the presence of ammoniacal dermatitis. The back is checked for any anomalies of the spine as well as any midline skin lesions, such as a hemangioma, nevus, isolated tuft of hair, or dimples. One should also note the presence and symmetry of the gluteal cleft. The anus is examined for sphincter tone and the integrity of the local reflex arcs.

Table 1. Observed complications with testicular prosthesis



The assessment continues with a radiologic evaluation and urodynamic study. The initial assessment must include a renal bladder ultrasound (with a note of the amount of postvoid residual urine), voiding cystourethrogram (VCUG) to assess bladder capacity, shape and presence of vesicoureteric reflux (VUR), and an urodynamic study. NB should be suspected in any child with a long history of micturition disturbances or during diagnostic work-up for other symptoms *e.g.* severe constipation or psychological disorders with encopresis. It is very important not to label every child with voiding issues as a patient with a "neurogenic" bladder to avoid over-diagnosis and stigmatization. An algorithm for the basic tenets of management are summarised as a flow chart:-

Intrauterine Intervention

Fetal surgery for antenatally diagnosed neural tube defects has been pondered over and even attempted several times but no definite conclusions have been drawn. Clayton *et al* in 2011 studied the long term outcomes of fetal MMC surgery and demonstrated a range of urological dysfunctions in this subset including decreased bladder capacity, detrusor overactivity, and increased detrusor pressure with no significant difference compared with individuals who underwent traditional postnatal repair.[6]

MOMS trial (management of MMC study)[7] was a prospective, multicentre, randomised controlled trial established by the National Institutes of Health in 2003 to evaluate the results of intrauterine surgery in the United States. The end-points evaluated were:

1. Fetal and infant mortality
2. Need for a ventriculoperitoneal shunt at the age of 1 year
3. Mental and motor development at 30 months age

The rates for shunt placement were 40% in the prenatal-surgery group and 82% in the postnatal-surgery group. The prenatal surgery group had better motor function compared with the postnatal surgery group. The trial was prematurely terminated because the overall data limited efficacy of prenatal surgery. A retrospective analysis of the data from this trial will evaluate the urologic outcomes.[7]

Fetal surgery is still nascent and thus, an attempt must be made to prevent damage at the time of the primary repair. Neurophysiological monitoring may be used to assist the surgeon with intraoperative planning and prevent iatrogenic worsening of urological function.[8] Tarcan reported on 56 patients with secondary tethering after repair of MMC and evaluated outcomes after a detethering procedure. They evaluated the outcomes according to the initial grade of urinary tract dilation. Individuals with Grade 1-2 dilation had an improvement in grade in 40% of the cases and complete resolution of the dilation in 33.3%. Those with Grade 3-4 dilation had an improvement in 50% of the cases and resolution in none.

The surgeon should be aware of the possibility of re-tethering during follow-up and consider early detethering to give such bladders another chance to improve.

Non-surgical Modalities

Before surgery is considered, conservative protocols have to be maximized because two-thirds of patients can become continent by clean intermittent catheterization (CIC) and medication alone.[9] Urodynamic studies have a major role to play in diagnosing and following these children since they forebode the urologist of the deleterious effects of a high pressure system rather than looking at the effects on the upper and lower tracts per se radiologically. McGuire and colleagues noted that 80% of patients with intravesical pressures greater than 40cm H20 at urinary leakage (detrusor leak point pressure [DLPP]) had reflux and/or hydronephrosis.[10] Beyond this pressure, upper tract damage occurs in 70% of the children.[11]

Pharmacological Agents

The aim of pharmacotherapy in neurogenic bladders is to bring about one or more of the following effects-decrease detrusor overactivity, increase bladder capacity, and/or increase bladder outlet resistance.

The drug oxybutynin hydrochloride has brought a paradigm shift in the outlook of patients with NB and is the most studied.[12,13] The anti-cholinergic action against the M3 muscarinic receptors on the wall of detrusor muscle relaxes it and thereby decreases intravesical pressures and uninhibited contractions and also indirectly increases bladder capacity. It also has an anti-spasmodic, local anaesthetic and calcium channel blocking properties which augment its effect on the overactive detrusor and "pharmacologically convert" it into an inactive reservoir.[14] It is administered orally in the dose of 0.2-0.4 mg/kg/day in 2-3 divided doses. The dose has to be limited sometimes in view of the side effects of altered thermoregulation and constipation. Alternatives also exist, such as transdermal patches and intravesical instillation (tablets are dissolved in water and instilled directly into the bladder by catheterization). It was demonstrated that a reduced first-pass metabolism of oxybutynin after intravesical instillation, resulting in a reduced generation of the N-desethyl metabolite, may explain the clinically relevant reduction of systemic side effects that characterizes intravesical compared with oral oxybutynin therapy.[15]

Other bladder relaxant drugs include propiverine (0.8mg/kg/day), trospium, and tolterodine. The current experience with compounds other than oxybutynin is still limited in children with NB. Propiverine hydrochloride, with its anti-muscarinic

and calcium-channel modulating properties, has been of proven efficacy in adults with neurogenic detrusor overactivity.[16,17] Madersbacher *et al*[18] demonstrated the superior tolerability of propiverine over oxybutynin with comparable efficacy in children. They followed up 17 children over an average of 3.6 years and achieved a maximum cystometric capacity within the normal range in 11 patients. They concluded that long-term efficacy and tolerability of propiverine in children is promising with clinically relevant improvements in key urodynamic outcomes.[19]

Another study evaluated long-term efficacy and safety of tolterodine in 30 children with neurogenic detrusor overactivity. Efficacy was evaluated urodynamically and using parents completed 3-day bladder diaries. Functional bladder capacity (volume at first leakage, first sensation of bladder fullness or 40cm H2O pressure) increased by month 12 with no change in detrusor leak point pressure. Hence they concluded that tolterodine was effective and well tolerated in children with neurogenic detrusor overactivity.[20]

One should ensure maximal medical therapy including dose escalation and the addition of a second anticholinergic prior to embarking on surgical management in children with NB. For children who void spontaneously, but experience urinary intermittency or incomplete bladder emptying related to detrusor-sphincter dyssynergia, one may try alpha-blocker therapy such as tamsulosin, prazosin or doxazosin. These have achieved success, are low risk to try, and can be followed for effectiveness by symptomatic improvement, as well as with a noninvasive uroflow study with postvoid residue.[8] Though useful, the studies which recommended the paediatric usage of these drugs lacked controls and long term follow up data.[2]

Another facet of pharmacotherapy is the use of prophylactic antibiotic cover in these children. However, the downside of daily antibiotics is the risk of developing antibiotic-resistant organisms.[21] Low dose, low efficacy antibiotics such as co-trimoxazole and nitrofurantoin should be used in an alternative fashion for prophylaxis[22] and replaced by high-efficacy full dose drugs for an active infection.

New drug development is taking place at a rapid pace. Bladder specific calcium channel openers, intravesical vanilloid (resiniferatoxin) treatment, tachykinin antagonists and calcium mobilizing and calcium signal modulating agents are all being studied as potential pharmacotherapeutic agents in NB.

Clean Intermittent Catheterization (CIC)

Lapides *et al* in the 1970s introduced CIC as a simple, effective way of keeping the bladder empty and thereby preventing the reflux of urine in case of high intravesical pressures.[23] Assuming fewer than 10% of children with congenital NB will develop satisfactory bladder control without need for CIC, all parents are initially counselled and reminded at periodic follow-ups to expect this intervention by the age of toilet training if urodynamic evaluation does not indicate earlier management.[24]

Some authors prefer early institution of CIC in all infants with NB, given the fact that by the age of 3 years, CIC will be required in all for achieving continence, and given the difficulties of starting CIC in toddlers.[25] Such early institution of CIC seems to improve the compliance of caregivers and their ability to assist the child in coping with his/her condition.[26] CIC can be done by parents till the child achieves sufficient dexterity to do it on his own, usually by around 7-8 years of age.[27] CIC done at regular intervals has several advantages:[28]

- Empties the bladder adequately without leaving any residual urine and hence no risk of infection
- Keeps the upper tracts safe of reflux prior to high pressure voiding
- Valuable tool to keep the child dry

Efforts to perform a sterile technique each time have not shown a significant difference in the incidence of UTI, although long-term randomized studies are limited.[29-30] Starting CIC will frequently result in chronic colonization of the bladder by bacteria. These bacteria are generally of low clinical consequence to urinary tract health if they are evacuated on a regular basis. Cloudy or foul-smelling urine is a common phenomenon of the chronically colonized bladder, but does not necessitate antibiotic treatment. Antibiotic use should be reserved only for a symptomatic episode of UTI. Overuse of antibiotics will contribute to formation of antibiotic-resistant organisms. The urine culture should be checked periodically for the presence of certain urease-positive organisms, such as Pseudomonas and Proteus, which need treatment to minimize formation of urinary tract stones or biofilms.[31]

A useful adjunct to a program of intermittent catheterization is a watch that is scheduled to alarm on a regular basis every 3-4 hours to help remind the child to catheterize on a timed schedule and not based on their urge to urinate. A major problem with doing CIC from an artificially created stoma is the risk of urine being left and consequently leading to infection, hence the tube inserted should be left to drain into a bowl kept at a lower level (on the floor) for complete evacuation. Spontaneous reflux resolution was recorded in 43% to 58% of cases with standard conservative treatment with a combination of CIC, anticholinergics and prophylactic antibiotics.[32]

Intradetrusor Botulinum Toxin (BOTOX) Injection

Persistent high pressure or uninhibited contractions (UIC)

are treated by increasing oral oxybutynin to tolerance and/or adding intravesical instillation of 5 mg two to three times daily as needed. In unusual cases with continued UIC, Botox injection is recommended. BOTOX results in decreased muscle activity by blocking the release of acetylcholine from the neuron by preventing the vesicle where the acetylcholine is stored from binding to the membrane where the neurotransmitter can be released. This effectively weakens the muscle for a period of three to four months. This ultimately projects as an increase in bladder capacity, decreased pressures, decreased incidence of reflux and improves continence.

A recent systematic review (six studies) encompassing 108 children (mean age 9.8 years) with neurogenic detrusor overactivity managed with CIC who were treated with BOTOX injection were evaluated from 2002 to 2006.[33-38] Follow-up was reported at 4 to 12 week intervals up to 26 weeks. BOTOX injection resulted in a mean 40-80% reduction from baseline, and 65-87% of children became completely continent between catheterizations. All six studies showed a significant reduction in maximum detrusor pressure and corresponded with an increase in maximum cystometric capacity (percentage increase from baseline ranging from 35-80%). Clinical efficacy was noted within 2 weeks from injection and these benefits were noted to persist for 3 to 6 months.

Bowel Management

One cannot effectively manage this disorder without careful attention to neurogenic bowel management. The degree of fecal incontinence is unpredictable and depends on the extent of the neuropathy and the effectiveness of the anal sphincter action. The large volume of hard stool retained in the colon occupies space in the pelvis and places pressure on the bladder that compromises the ability to store and evacuate urine effectively. It is useful to consider strategies of scheduled evacuation. This includes healthy dietary fiber intake and a daily stool softener, coupled with a method of daily evacuation to achieve effective bowel management. Biofeedback programmes have not been shown to be more effective in achieving fecal continence than a bowel management programme.[39]

Surgical Modalities

To Increase1 Bladder Storage Capacity

The ideal state of the bladder with minimum long term effects on the upper and lower tracts is the presence of low pressure storage and physiologic storage pressure at an adequate volume. The volume at which pressure increase occurs is defined as the "reflex volume".[40] It is intuitive that a high reflex volume will aid in the protection of the upper tracts. A number of surgical options are available should a combination of pharmacotherapy and CIC prove inadequate in halting the progression of urinary tract damage. It is imperative that one must have an unscarred bladder to work with and a mechanism of satisfactory bladder emptying.

As mentioned previously, a more conservative approach to improving the storage capacity of the detrusor is by the intramural Botox injection. Failing this, one may think of "partial autoaugmentation" with the use of detrusor myomectomy.[41] The urothelium is left intact and post-procedure urinary decompression by regular CIC is mandatory while the anatomic and physiologic reconfiguration takes place (usually in the next 1-2 years). The use of additional tissue to cover the muscular defect does not contribute significantly to outcomes.[42,43]

Bladder augmentation is usually kept low on the list of surgical options due to the complexity and inherent long-term morbidity of these procedures. Enterocystoplasty and clam cystoplasty are good alternatives when the surgeon is faced with a small, scarred bladder. Clam cystoplasty involves the insertion of a patch of intestine on the longitudinally opened posterior wall of the bladder.[44] Despite the simple elegance of autoaugmentation, it has not proved as useful as the use of intestinal segments to increase storage capacity.[39] The choice of surgery leans towards urinary diversion in the form of continent or incontinent urinary diversion and bladder reconstruction/replacement in the event of intractable incontinence, diminishing renal functions and hypertension following long-term renal damage.

Neuromodulation

Neuromodulation therapy aims to treat the abnormal innervation of the bladder, trying to "re-train" the nerve-muscle interaction to attain more normal bladder function. The available treatments include nonsurgical therapies, such as transurethral electrical bladder stimulation; minimally invasive procedures, such as implantation of a sacral neuromodulation pacemaker device; and operative procedures that reconfigure sacral nerve root anatomy.

Sacral neuromodulation by a reversible implantable device (InterStim, Medtronic, Minneapolis, MN, USA), is thought to improve bladder function either by consistent stimulation of the efferent fibers of the sacral nerve roots or by providing rhythmic contractions of the pelvic floor.[45] Hagerty, Richards and Kaplan were the main proponent of the technique of transurethral electrical bladder stimulation in children and reported a series of 372 patientss with a mean age of 5.5 years and mean follow-up 6.6 years. In 76.9% of these patients, the increase in bladder capacity was 20% or greater.[46]

Another method of counteracting a small, hyperactive bladder that is available for patients with high and complete cord lesions is the intradural transection of the S2-S4 nerve roots which in effect achieves deafferentation of the urinary bladder.

A necessary pre-requisite is that the bladder should be compliant and the person must possess enough manual dexterity and means to achieve regular intermittent self-catheterization. As most of the paitents in whom this modality is chosen have a high cord lesion, the latter criterion may not be fulfilled. In such cases, sacral electrostimulation (*e.g.* Brindley stimulator) can be utilized to initiate voluntary voiding, defaecation and even penile erections.

Sauerwein *et al*[47] and Kutzenberger *et al*[48] noted a less than satisfactory effect of these implants in erectile dysfunction in this group of patients. Addressing this along with the myriad psychosexual issues that develop as a natural consequence is important as eventually our paediatric patients grow into adolescence and adulthood due to the longer survival times. Guys *et al*[49] described the results of sacral nerve stimulation (SNS) therapy in children with neurogenic bladder dysfunction. They studied the results in 42 children with neurogenic bladder dysfunction, mainly due to spina bifida. Twenty-one children were randomized between the conservative and interventional arms. This group did not find significantly better results in the SNS group and postulated that it could be due to a small study group or very severe bladder function.

Surgery for the Incompetent Bladder Outlet

Bladder Neck Bulking Agents

In case of sphincter underactivity/pelvic floor paresis, bladder neck procedures are the way to achieve continence. Agents that have been employed for urethral injection include autologous fat, polytetrafluoroethylene, bovine collagen, and pyrolytic carbon-coated zirconium oxide beads (Durasphere). Bulking agents also include silicon grains in povidone (MACROPLASTIQUE) injected by suprapubic puncture under transurethral endoscopic vision control. In the current era, Deflux is being used as a bulking agent in the region of bladder neck.

In a series of patients with mixed causes of incontinence and mean follow-up of 28 months, bladder neck injection using dextranomer/hyaluronic acid polymer (Deflux) resulted in a dry interval of 4 hours in 48% of 27 children with neuropathic bladders, 4 of whom underwent failed treatment with slings.[50] Similarly, polydimethylsiloxane injection ended pad use in 34% of 44 children with NBs, 24 of whom underwent prior bladder neck procedures, at median follow-up of 28 months.[51] The main disadvantage with this approach is the difficulty in CIC which follows.

Neel *et al*[52] have introduced the concept of total endoscopic management (TEM) in children with noncompliant NB to address continence and treatment of VUR. Ten children with NB (mean age 5.9 years) who did not respond to maximal anticholinergic dosing and CIC were treated with cystoscopic injection of botulinum-A toxin in the detrusor and subureteric injection of Deflux. At 1 month, a significant increase in bladder capacity (79±49 to 155±55 ml), increase in bladder compliance (from 1.4 to 4.3 ml/cmH2O), and decrease in maximum detrusor pressure (from 55±16 to 37±11cmH2O) were noted. These improvements were maintained at 6-months follow-up. VUR was successfully treated in 15 of 16 ureters (94%). Five of six incontinent patients achieved complete dryness. This study provides an interesting approach to avoid bladder augmentation in this population.

Bladder Neck Slings and Bladder Neck Reconstruction

One of the more enduring questions when choosing these techniques is whether to carry out concomitant bladder augmentation. Male children have a potential for prostatic growth and thus may be served by bladder augmentation alone despite low outlet resistance.[53]

Kreder *et al*[54] advocated that supporting the bladder neck with a sling may be enough to abolish leakage if the preoperative urodynamic assessment, performed with some form of bladder outlet occlusion, shows a stable bladder with sufficient capacity and normal compliance. Whatever the choice based on pre-operative radiological studies and urodynamics, the durability of the sling and longevity of the procedure must be foremost in the surgeon's mind when applied to the paediatric population.

Artificial Urinary Sphincter (AUS)

American Medical Systems currently produced the AMS 800 model which is the only AUS currently available in the market. The three components include a cuff which fits around the urethra or bladder neck; a balloon fluid reservoir which is implanted in the abdomen; and a pump which is implanted in the scrotum or labia to control activation. The device can be activated a few weeks after surgery by squeezing the bulb of the pump which forces fluid into the hollow cuff which occludes the urethra. Prior to voiding, the bulb is pumped to push fluid back into the reservoir. Care must be taken during cystourethroscopic visualization.

There is a controversy regarding the ideal time for implantation of the AUS in the paediatric age group. Both Kryger *et al*[55] and Levesque *et al*[56] found no increase in the rate of AUS re-insertion with growth. Kryger *et al* also found no difference in the continence rate, revision rate, augmentation rate, or number of complications in patients who received an AUS before age 11 compared to those placed later in life. AUS insertion, in fact, may be easier in prepubertal patients secondary to the shallower pelvis and lesser degree of periurethral venous plexus engorgement. Revisions for retraction of the pump in the scrotum were uncommon, occurring only in 1 of their 25 patients.[55]

Conclusion

What should and can be achieved is a more or less adequate, low-pressure, functional capacity of the bladder that is emptied as completely as possible. Newer techniques are being pioneered at an astonishing rate and the modern surgeon has a smorgasbord of techniques to choose from and can "custom-fit" the therapy to his/her patient. Tissue engineering will play an increasing role in the future with the first reports of the use of this ground-breaking technique reported by Atala *et al* in 2006.[57] It is an alternative to the free grafts that are conventionally used and is grown by seeding autologous cells harvested from the bladder onto a scaffold of "extracellular matrix".

The ultimate goals though are to provide continence, to prevent bladder-wall destruction, and to preserve upper tract function. It is not always possible to reach these objectives. Therefore, one should keep in mind some basic rules. First, renal function preservation is more important than continence. Second, noninvasive measures take priority over surgery, as the former are reversible but the latter is not. Ultimately, the treating physician has to realise the primacy of the individual in the holistic view of management. The 'child' is the focus-not the 'disease'.

References

1. Bauer SB. Neurogenic bladder: etiology and assessment. Pediatr Nephrol 2008;23:541-51.
2. Verpoorten C, Buyse GM. The neurogenic bladder: medical treatment. Pediatr Nephrol 2008;23:717-25.
3. de Jong TP, Chrzan R, Klijn AJ, Dik P. Treatment of the neurogenic bladder in spina bifida. Pediatr Nephrol 2008;23:889-96.
4. Spindel MR, Bauer SB, Dyro FM, Krarup C, Khoshbin S, Winston KR *et al*. The changing neurourologic lesion in myelodysplasia. JAMA. 1987;258:1630-3.
5. Rouzade-Dominguez ML, Miselis R, Valentino RJ *et al*. Central representation of bladder and colon revealed by dual transsynaptic tracing in the rat: substrates for pelvic visceral coordination. Eur J Neurosci 2003;18:3311-24.
6. Clayton DB, Tanaka ST, Trusler L, Thomas JC, Pope JC 4th, Adams MC *et al*. Long-term urological impact of fetal myelomeningocele closure. J Urol 2011;186:1581-5.
7. Adzick NS, Thom EA, Spong CY, Brock JW 3rd, Burrows PK, Johnson MP *et al*. A randomized trial of prenatal versus postnatal repair of myelomeningocele. N Engl J Med 2011;364:993-1004.
8. Pouratian N, Elias WJ, Jane JA Jr, Phillips LH 2nd, Jane JA Sr. Electrophysiologically guided untethering of secondary tethered spinal cord syndrome. Neurosurg Focus 2010;29:E3
9. Frimberger D, Cheng E, Kropp BP. The current management of the neurogenic bladder in children with spina bifida. Pediatr Clin North Am 2012;59:757-67.
10. McGuire EJ, Woodside JR, Borden TA, Weiss RM. Prognostic value of urodynamic testing in myelodysplastic patients. J Urol 1981;126:205-9.
11. Wan J, McGuire EJ, Bloom DA, Ritchey ML. Stress leak point pressure: a diagnostic tool for incontinent children. J Urol 1993;150:700-2.
12. Youdim K, Kogan BA. Preliminary study of the safety and efficacy of extended-release oxybutynin in children. Urology 2002;59:428-32.
13. Goessl C, Sauter T, Michael T, Bergé B, Staehler M, Miller K. Efficacy and tolerability of tolterodine in children with detrusor hyperreflexia. Urology 2000;55:414-8.
14. Andersson KE, Chapple CR. Oxybutynin and the overactive bladder. World J Urol 2001;19:319-23.
15. Buyse G, Waldeck K, Verpoorten C, Björk H, Casaer P, Andersson KE. Intravesical oxybutynin for neurogenic bladder dysfunction: less systemic side effects due to reduced first pass metabolism. J Urol 1998;160:892-6.
16. Madersbacher H, Mürtz G. Efficacy, tolerability and safety profile of propiverine in the treatment of the overactive bladder (non-neurogenic and neurogenic). World J Urol 2001;19:324-35.
17. Stöhrer M, Mürtz G, Kramer G, Schnabel F, Arnold EP, Wyndaele JJ. Propiverine compared to oxybutynin in neurogenic detrusor overactivity--results of a randomized, double-blind, multicenter clinical study. Eur Urol 2007;51:235-42.
18. Madersbacher H, Mürtz G, Alloussi S, Domurath B, Henne T, Körner I *et al*. Propiverine vs oxybutynin for treating neurogenic detrusor overactivity in children and adolescents: results of a multicentre observational cohort study. BJU Int 2009;103:776-81.
19. Schulte-Baukloh H, Mürtz G, Heine G, Austin P, Miller K, Michael T *et al*. Urodynamic effects of propiverine in children and adolescents with neurogenic bladder: results of a prospective long-term study. J Pediatr Urol 2012;8:386-92.
20. Reddy PP, Borgstein NG, Nijman RJ, Ellsworth PI. Long-term efficacy and safety of tolterodine in children with neurogenic detrusor overactivity. J Pediatr Urol 2008;4:428-33.

21. Schlager TA, Anderson S, Trudell J, Hendley JO. Nitrofurantoin prophylaxis for bacteriuria and urinary tract infection in children with neurogenic bladder on intermittent catheterization. J Pediatr 1998;132:704-8.

22. Salomon J, Denys P, Merle C, Chartier-Kastler E, Perronne C, Gaillard JL *et al.* Prevention of urinary tract infection in spinal cord-injured patients: safety and efficacy of a weekly oral cyclic antibiotic (WOCA) programme with a 2 year follow-up--an observational prospective study. J Antimicrob Chemother 2006;57:784-8.

23. Lapides J, Diokno AC, Silber SJ, Lowe BS. Clean, intermittent self-catheterization in the treatment of urinary tract disease. J Urol 1972;107:458-61.

24. Snodgrass WT, Gargollo PC. Urologic care of the neurogenic bladder in children. Urol Clin North Am 2010;37:207-14.

25. van Gool JD, Dik P, de Jong TP. Bladder-sphincter dysfunction in myelomeningocele. Eur J Pediatr 2001;160:414-20.

26. Kessler TM, Lackner J, Kiss G, Rehder P, Madersbacher H. Early proactive management improves upper urinary tract function and reduces the need for surgery in patients with myelomeningocele. Neurourol Urodyn 2006;25:758-62.

27. de Jong TPVM. Neurogenic bladder, modern treatment. J Paediatr Surg Special 2007;1:55-9.

28. Edelstein RA, Bauer SB, Kelly MD, Darbey MM, Peters CA, Atala A *et al*. The long-term urological response of neonates with myelodysplasia treated proactively with intermittent catheterization and anticholinergic therapy. J Urol 1995;154:1500-4.

29. Getliffe K, Fader M, Allen C, Pinar K, Moore KN. Current evidence on intermittent catheterization: sterile single-use catheters or clean reused catheters and the incidence of UTI. J Wound Ostomy Continence Nurs 2007;34:289-96.

30. Moore KN, Burt J, Voaklander DC. Intermittent catheterization in the rehabilitation setting: a comparison of clean and sterile technique. Clin Rehabil 2006;20:461-8.

31. Hung EW, Darouiche RO, Trautner BW. Proteus bacteriuria is associated with significant morbidity in spinal cord injury. Spinal Cord 2007;45:616-20.

32. Engel JD, Palmer LS, Cheng EY, Kaplan WE. Surgical versus endoscopic correction of vesicoureteral reflux in children with neurogenic bladder dysfunction. J Urol 1997;157:2291-4.

33. Altaweel W, Jednack R, Bilodeau C, Corcos J. Repeated intradetrusor botulinum toxin type A in children with neurogenic bladder due to myelomeningocele. J Urol 2006;175:1102-5.

34. Kajbafzadeh AM, Moosavi S, Tajik P, Arshadi H, Payabvash S, Salmasi AH *et al*. Intravesical injection of botulinum toxin type A: management of neuropathic bladder and bowel dysfunction in children with myelomeningocele. Urology 2006; 68:1091-6.

35. Riccabona M, Koen M, Schindler M, Goedele B, Pycha A, Lusuardi L *et al.* Botulinum-A toxin injection into the detrusor: a safe alternative in the treatment of children with myelomeningocele with detrusor hyperreflexia. J Urol 2004;171:845-8.

36. Schulte-Baukloh H, Knispel HH. Repeated botulinum-A toxin injections in treatment of children with neurogenic detrusor overactivity. Urology 2005;66:865-70.

37. Schulte-Baukloh H, Michael T, Schobert J, Stolze T, Knispel HH. Efficacy of botulinum-a toxin in children with detrusor hyperreflexia due to myelomeningocele: preliminary results. Urology 2002;59:325-7.

38. Schulte-Baukloh H, Michael T, Stürzebecher B, Knispel HH. Botulinum-a toxin detrusor injection as a novel approach in the treatment of bladder spasticity in children with neurogenic bladder. Eur Urol 2003;44:139-43.

39. Tekgül S, Riedmiller H, Gerharz E, Hoebeke P, Kocvara R, Nijman R *et al*. Management of neurogenic bladder in children. In: Guidelines on paediatric urology. European Association of Urology, European Society for Paediatric Urology; Arnhem, The Netherlands: 2009 Mar. p. 31-41.

40. Stöhrer M, Goepel M, Kondo A, Kramer G, Madersbacher H, Millard R *et al*. The standardization of terminology in neurogenic lower urinary tract dysfunction with suggestions for diagnostic procedures. Neurourol Urodyn 1999;18:139-58.

41. Cartwright PC, Snow BW. Bladder auto-augmentation: early clinical experience. J Urol 1989;142:505-8.

42. Oge O, Tekgul S, Ergen A, Kendi S. Urothelium-preserving augmentation cystoplasty covered with a peritoneal flap. BJU Int 2000;85:802-5.

43. Perovic SV, Djordjevic ML, Kekic ZK, Vukadinovic VM. Bladder autoaugmentation with rectus muscle backing. J Urol 2002;168:1877-80.

44. Mast P, Hoebeke P, Wyndaele JJ, Oosterlinck W, Everaert K. Experience with augmentation cystoplasty. A review. Paraplegia 1995;33:560-4.

45. Das AK, White MD, Longhurst PA. Sacral nerve stimulation for the management of voiding dysfunction. Rev Urol 2000;2:43-60.

46. Hagerty JA, Richards I, Kaplan WE. Intravesical electrotherapy for neurogenic bladder dysfunction: a 22-year experience. J Urol 2007;178:1680-3.

47. Sauerwein HD. The use of nerve deafferentation and stimulation in the paraplegic female patient. In: Female Urology. Raz S (ed). Philadelphia: WB Saunders, 1996: 656-64.

48. Kutzenberger J, Pannek J, Stohrer M. Neurourology: Current developments and therapeutic strategies. Urologe A 2006;45:158-66.

49. Guys JM, Haddad M, Planche D, Torre M, Louis-Borrione C, Breaud J. Sacral neuromodulation for neurogenic bladder dysfunction in children. J Urol 2004;172:1673- 6.

50. Lottmann H, Traxer O, Aigrain Y, Melin Y. Posterior approach to the bladder for implantation of the 800 AMS artificial sphincter in children and adolescents: techniques and results in eight patients. Ann Urol (Paris) 1999;33:357-63.

51. Guys JM, Breaud J, Hery G, Camerlo A, Le Hors H, De Lagausie P. Endoscopic injection with polydimethylsiloxane for the treatment of paediatric urinary incontinence in the neurogenic bladder: long-term results. J Urol 2006;175:1106-10.

52. Neel KF, Salem M, Soliman S. Total endoscopic management (TEM approach) of children with non-compliant neuropathic bladder: a preliminary report. J Pediatr Urol 2008;4:124-6.

53. Raz S, McGuire EJ, Ehrlich RM, Zeidman EJ, Wang SC, Alarcon A *et al*. Fascial sling to correct male neurogenic sphincter incompetence: the McGuire/Raz approach. J Urol 1988;139:528-31.

54. Kreder KJ, Webster G. Management of the bladder outlet in patients requiring enterocystoplasty. J Urol 1992;147:38-41.

55. Kryger JV, Lerverson G, Gonzalez R. Long-term results of artificial urinary sphincters in children are independent of age at implantation. J Urol 2001;165:2377-9.

56. Levesque PE, Bauer SB, Atala A, Zurakowski D, Colodny A, Peters C *et al*. Ten year experience with the artificial urinary sphincter in children. J Urol 1996;156:625-8.

57. Atala A, Bauer SB, Soker S, Yoo JJ, Retik AB. Tissue-engineered autologous bladders for patients needing cystoplasty. Lancet 2006;367:1241-6.

Original Article

Pelviureteric Junction Obstruction Associated with Vesico-ureteric reflux and Vesico-ureteric Junction Obstruction: Challenges and Management Protocols

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Abstract. ***Objective:*** To assess the incidence and outcome of cases of pelviureteric junction obstruction (PUJO) with associated vesicoureteric reflux (VUR) and/or vesicoureteric junction obstruction (VUJO). ***Methods:*** This retrospective analysis included cases of PUJO with associated VUJ anomalies managed between January 2000 and April 2013. All the cases were operated first for PUJO by Anderson-Hynes pyeloplasty via lumbotomy approach. Follow up was done by ultrasound and renal dynamic scan (RDS) at 6 weeks after surgery. Second surgery done was Cohen's ureteric reimplantation after 6 weeks or later. They were subsequently followed by using standard follow up protocol. ***Results:*** Out of a total of 29 cases, four cases had an associated VUJO while remaining 25 cases had an associated VUR. VUJO cases presented with poorly functioning kidneys with average split renal function (SRF) of 7%. These cases underwent PCN as an initial management. Follow up RDS were suggestive of VUJO in two cases and were then confirmed with an antegrade dye study. Two cases were found to have VUJO on intraoperative saline test. These cases subsequently underwent Cohen's ureteric reimplantation. Mean SRF in patients with VUR was 22%. Mean number of surgeries per patient was 2. Two cases with consequent VUR didn't show improvement in SRF in last follow up. Rest of the patients showed a significant improvement in SRF after surgery. ***Conclusion:*** Concurrent presence of PUJO and VUJ anomalies is a diagnostic challenge. These cases can be managed nicely if a protocol based approach is followed.

Keywords: Pelviureteric junction obstruction, Vesicoureteric reflux, Vesicoureteric junction obstruction, Vesicoureteric junction anomalies

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Introduction

Congenital pelviureteric junction obstruction (PUJO) is one of the most common causes of hydronephrosis in children. A child may present to a clinician with complaint of dull aching pain on the affected side or with an obvious palpable lump. This condition can be diagnosed by ultrasonography and confirmed by a renal dynamic scan. The management protocols are based on the presence of symptoms. On the contrary when the condition is asymptomatic, the split renal function of the affected kidney determines the line of further management. There has been a lot of debate regarding when to operate and when to follow up these patients. Accepted consensus is to operate the obstructed side when the split renal function is < 35 % with an obstructed pattern of clearance on renal dynamic scan.[1] PUJO is associated with a number of conditions and the management of these conditions largely varies. Association of vesicoureteric junction anomalies is not very commonly noticed. Vesicoureteric reflux (VUR) is among the most common vesicoureteric junction anomaly associated with PUJO and infact PUJO is considered secondary to the associated VUR. Few uncommon cases are associated with a vesicoureteric junction obstruction (VUJO). In this article we have tried to address these issues.

Objective

To find out the incidence of associated VUR and/ or VUJO in operated cases of PUJO and assess their outcome.

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Materials and Methods

This was a retrospective analysis of cases of PUJ obstructions managed in Wednesday Paediatric Urology Clinic (WPUC), AIIMS, between January 2000 and April 2013 who presented with an associated vesicoureteric junction anomalies. Case sheets of all these patients were analyzed and data regarding clinical presentation, age at presentation, management offered and investigations done were obtained and analyzed. Those with incomplete data or those who were lost to follow up were excluded. Those with neurogenic bladder, urethral valves, ureterocele, bladder diverticula and duplex system were also excluded. VUR was defined as the presence of reflux documented in micturating cystourethrogram (MCU). PUJO was defined as an obstructive pattern of drainage in renal dynamic scan using LLEC. VUJO was defined as an evident vesicoureteric junction obstruction on antegrade dye study (Fig. 1), intraoperative diagnosis based on saline test or an evident hydroureteronephrosis in a well-tempered renogram with a bladder catheter in situ showing obstructive pattern in vesicoureteric junction.

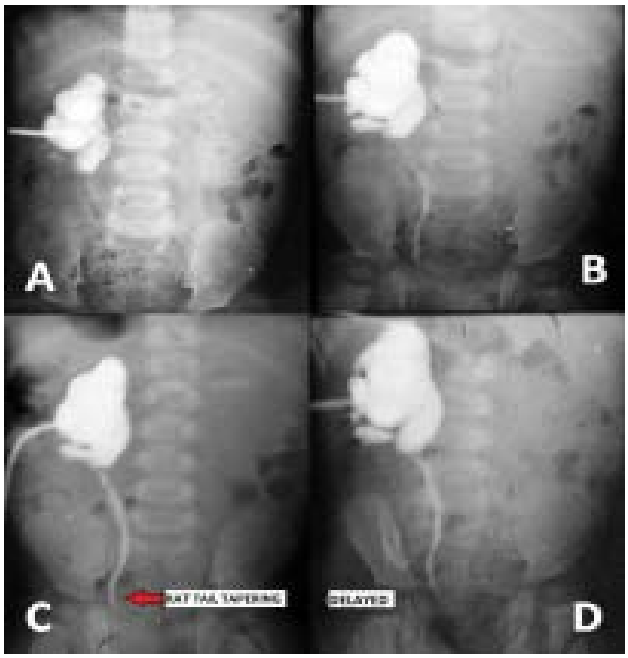

Fig. 1. Antegrade dye study in operated PUJO patient showing rat tail tapering suggestive of associated VUJ obstruction (A): Initial (B) After 15 sec (C) After 2 min (D) After 45 min

All the cases were operated first for a PUJO using Anderson-Hynes pyeloplasty via lumbotomy approach. Follow up was done using an ultrasound and a renal dynamic scan at 6 weeks after surgery using follow up protocol as shown in Fig. 2.

Second surgery done was Cohen's ureteric reimplantation after 6 weeks or later from pyeloplasty. They were than

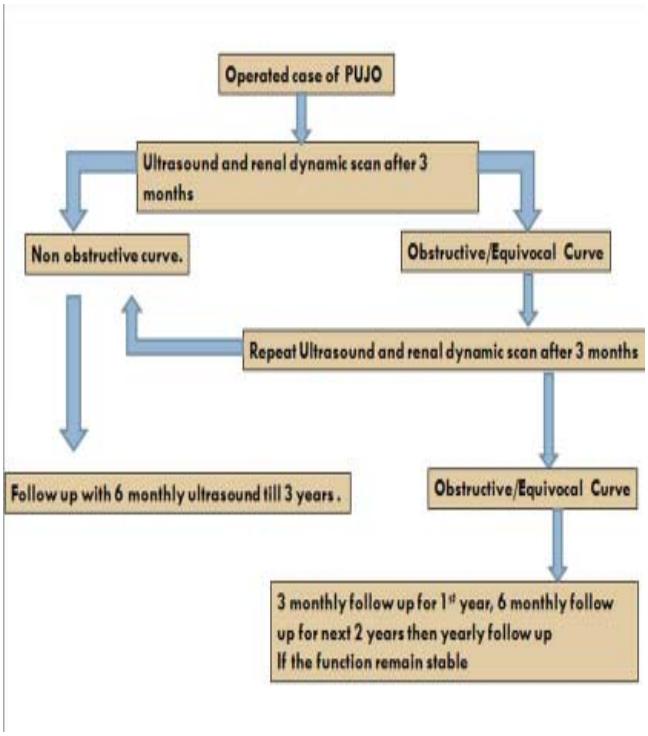

Fig. 2. Follow up protocol after pyeloplasty

subsequently followed by doing an ultrasound scan at 3monthly interval, MCU and RDS after 6 months of second surgery and subsequently by ultrasounds. Success was defined as no sign of reflux on MCU, resolution or significant decrease in hydroureteronephrosis, complete clinical improvement, resolution of preoperative symptoms, improvement in drainage curve on diuretic renogram, regression in pelvic diameter on serial ultrasonography, and improvement or maintenance of renal function on the renal dynamic scan using 99mTc-LLEC scan. The result obtained was analyzed.

Results

There were a total of 293 cases of pelviureteric junction obstruction managed in the clinic during the same period. A total of 29 cases that had either VUJO or VUR formed the study group. The mean age at presentation was 37.26 months (range: 3-148 months). There were 3 females and 26 males with the male to female ratio of 9:1. Four cases had an associated VUJO while remaining 25 cases had an associated VUR. There were 7 cases with right sided pathology, 19 with left sided pathology while 3 had bilateral pathology. VUJO cases presented with poorly functioning kidneys with average split renal function of 7% (range 0-10%). These cases underwent percutaneous nephrostomy (PCN) as an initial management. Follow up renal dynamic scans were suggestive of VUJO in two cases and were then confirmed with an

antegrade dye study. Two cases were found to have VUJO on intraoperative saline test. These cases subsequently underwent Cohen's ureteric reimplantation. Mean post-operative follow up was 24 months (range 8-48 months). Of the 25 cases with VUR, 20 cases had higher grades of VUR (grade 4 and 5) while 3 cases had grade 3 VUR and 2 cases had lower grades of documented VUR. Those with VUR below grade 3 were followed for VUR and not operated. Mean Split renal function in patients with VUR was 22% (Range 13- 38%). Mean number of surgeries per patient was 2 (range 1-4). Two cases with consequent VUR didn't show improvement in the split renal function in last follow up though they had a non-obstructive clearance and are in follow up. Rest of the patients showed a significant improvement in the split renal function after surgery.

Discussion

The coexistence of obstructions at the proximal and the distal ends of the ureter is rare. There are very few published series in English literature showing such coexistence. Most often the distal obstruction is masked either due to the poor function of the concerned kidney or due to the focus of interest on the proximal obstruction. Most of the cases who presented to us with dual obstruction had poorly functioning kidneys and VUJ obstruction was picked up intraoperatively in two while others were diagnosed in follow up imaging. Pak *et al* first reported a case of coexistence of PUJ obstruction with VUJ obstruction in a horseshoe kidney.[2] Pfister and Hendren reported 150 primary mega-ureters (predominantly in children) and described five patients (3%) who had coexisting PUJ obstruction.[3] Pitts reviewed 80 patients (about one half were children) with primary megaureter and noted that 20 (25%) had simultaneous PUJ obstruction.[4] Subsequently McGrath at el reported 14 cases of double obstruction after reviewing about 225 cases of PUJ obstruction and 115 cases of VUJ obstruction diagnosed over an 11year period.[5] The ages at presentation ranged from birth (prenatal diagnosis) to 11 years. In 11 cases the left ureter was affected; in 2 of these it was draining the child's only kidney. The diagnosis was made preoperatively in only 3 patients. In 3 the second obstruction was initially suspected at surgery; in the other 8, the second obstruction was diagnosed during follow-up radiologic studies performed 2 weeks to 2.5 years after surgery.

In most of the cases the concerned side with dual obstruction has very poor function. Thus the diagnosis remains obscure and most of the time PCN is done to see the pattern of improvement. After 6-8 weeks the renal dynamic scan is done to see the improvement in pattern. Most of the time, as there is an associated PUJ obstruction, we can't get an idea about the lower obstruction. Most of these cases are thus diagnosed in the follow up investigations. A fraction of cases are diagnosed intraoperatively on saline injection test or while inserting a DJ stent. Many surgeons still contest in the issue and argue that both the obstruction can be relieved at a time while others propose a staged management with upper obstruction to be addressed first. As of now there is no established series comparing the results of these two approaches so as a dictum proximal obstruction is relieved first. The second surgery which is the reimplantation of the obstructed ureter is done after 6-8 weeks of the first surgery, till then the child is maintained on a nephrostomy. After reimplantation the nephrostomy is clamped and subsequently removed.

The presence of PUJO with an associated VUR is rather commonly noted. There is contest regarding the pathogenesis of the obstruction secondary to VUR. The obstruction is mostly congenital or primary; however, it can occur secondary to severe VUR.[6] The coexistence of VUR and PUJO has been reported.[7] Approximately 10% of patients with PUJO are found to have concomitant VUR coincidently, whereas less than 1% of patients with the diagnosis of VUR have simultaneous PUJO.[8] The etiology of the coexistence is not well established.[9] However, ureteral kinking and inflammation resulting from reflux, the presence of a single developmental abnormality, or a random event have been proposed as possible etiologies.[9] Management of these patients can be complicated, and no straightforward approach has been described in the literature. The traditional management of children with VUR and PUJO is pyeloplasty as the initial step, and if the high-grade reflux remains symptomatic after pyeloplasty, surgical correction of the reflux is the next step.[10] It was postulated that the nearby obstruction to the renal parenchyma might cause more renal damage, so it should be managed sooner rather than later. Moreover, in a few paediatric urology centers, this concept has ultimately been changed by surgical correction of reflux (ureteral reimplantation) initially, followed by pyeloplasty in persistent PUJO. Kim *et al* also favoured of initial pyeloplasty because they hypothesized that edema at the ureterovesical junction after ureteral reimplantation, and not endoscopic injection, leads to acute worsening of the PUJ obstruction.[11] Peters *et al*[12] and Onen *et al*[13] suggested that because conservative management can be applied to low-grade hydronephrosis caused by PUJO, all patients with PUJO, even with coexisting VUR, should not undergo pyeloplasty as the initial intervention. Bomalaski *et al*[14] reported a pyeloplasty rate of 75% in children with simultaneous VUR and PUJO. Kajbafzadeh *et al*[15] approached patients with concomitant VUR and PUJO by endoscopic injection of Deflux using the HABIT technique. They included 2810 cases of VUR and found 143 cases having a concomitant PUJO. Of the 76 (89 ureters) included in the study they showed a self-resolution of PUJO in 76.9% per patient (74.1% per ureter) after the first injection and 84.6% per patient (80.6% per ureter) after the

second. Only 3 patients (11.5%) underwent ureteral reimplantation, and 3 (11.5%) required pyeloplasty. This recent publication has started the debate on the management of this concomitant lesion with the management of VUR first.

We, however, believe in the traditional management option and feel that the upper obstruction should be addressed first and then VUR should be subsequently addressed after 6-8 weeks of the first surgery. Follow up of these cases should be done regularly. The follow up protocol should be as shown in figure 2. An MCU should be repeated after 3 months of reimplantation and repeated after 6 months. If MCU shows no reflux it may not be repeated as such.

Conclusion

Concurrent presence of PUJO and VUJ anomalies is a diagnostic challenge. Presence of poorly functioning kidney on the affected side makes diagnosis further difficult. These cases can be managed nicely if a protocol based approach is followed.

References

1. Chandrasekharam VV, Srinivas M, Bal CS, Gupta AK, Agarwala S, Mitra DK, *et al*. Functional outcome after pyeloplasty for unilateral symptomatic hydronephrosis. Pediatr Surg Int 2001;17:524-7.
2. Pak K, Konishi T. Horseshoe kidney in an infant associated with ureteropelvic junction and ureterovesical junction obstructions. Hinyokika Kiyo 1985;31:457-61.
3. Pfister PC, Hendren WH. Primary megaureter in children and adults: clinical and pathophysiologic features of 150 ureters. Urology 1978;12:160-76.
4. Pitts WR, Muecke EL. Congenital megaloureter: a review of 80 patients. J Urol 1974;111:468-73.
5. McGrath M A, Estroff J, Lebowitz. RL. The Coexistence of Obstruction at the Ureteropelvic and Ureterovesical Junctions. AJR 1987;149:403-6.
6. Carr MC, Casale P. Anomalies and surgery of the ureter in children. In: Wein A, Kavoussi L, Novick A, *et al*., eds. Campbell-Walsh Urology. 9th ed. Philadelphia, PA: Saunders; 2007:3212-3235.
7. Karnak I, Woo LL, Shah SN, Sirajuddin A, Kay R, Ross JH. Prenatally detected ureteropelvic junction obstruction: clinical features and associated urologic abnormalities. Pediatr Surg Int 2008;24:395-402.
8. Byrne LN, Koyle MA. Ureteropelvic junction obstruction unmasked after correction of vesicoureteral reflux using endoscopic treatment with Deflux. J Pediatr Urol 2006;2:205-9.
9. Stauss J, Connolly LP, Connolly SA, Zurakowski D, Treves ST, Peters CA. Dynamic renal scintigraphy in children with vesicoureteral reflux and suspected coexisting ureteropelvic junction obstruction. J Urol 2003;170:1966-70.
10. Maizels M, Smith CK, Firlit CF. The management of children with vesicoureteral reflux and ureteropelvic junction obstruction. J Urol 1984;131:722-7.
11. Kim YS, Do SH, Hong CH, Kim MJ, Choi SK, Han SW. Does every patient with ureteropelvic junction obstruction need voiding cystography? J Urol 2001;165:2305-7.
12. Peters CA. Urinary tract obstruction in children. J Urol 1995;154:1874-83.
13. Onen A, Jayanthi V, Koff S. Long-term followup of prenatally detected severe bilateral newborn hydronephrosis initially managed nonoperatively. J Urol 2002;168:1118-20.
14. Bomalaski MD, Hirschl RB, Bloom DA. Vesicoureteral reflux and ureteropelvic junction obstruction: association, treatment options and outcome. J Urol 1997;157:969-74.
15. Kajbafzadeh AM, Tourchi A, Ebadi M. The Outcome of Initial endoscopic Treatment In the Management of Concomitant Vesicoureteral Reflux and Ureteropelvic Junction Obstruction. Urology 2013;81:1040-6.

Original Article

Pelviureteric Junction Obstruction in Duplex System: Management and Review of Literature

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Abstract. ***Objective:*** Objective of this study was to review the management and outcome of 17 children with pelvi-ureteric junction obstruction (PUJO) in a duplex kidney. ***Materials and Methods:*** We analyzed our record from January 2000 to December 2011. A total of 258 pyeloplasty in a single system and 17 pyeloplasty in duplex system were performed during this period. Lower and upper pole moiety obstruction was seen in both complete and incomplete duplex systems. The duplex was suspected in ultrsonography (USG), renal scintigraphy and confirmed on magnetic resonance urography (MRU). ***Results:*** Ten children had upper moiety obstruction while seven had a lower moiety obstruction. Upper and lower pole obstructions were seen in both complete and incomplete duplex moiety. Eleven children underwent Anderson-Hynes pyeloplasty, three underwent ureteropyelostomy, and two underwent heminephrectomy while one underwent uretero-ureterostomy ***Conclusion:*** There are a multitude of surgical options to deal with PUJO in duplex system and management should be based on the pathological and functional anatomy and the experience of the surgeon.

Keywords: Duplex system, Lower moiety, Pelviureteric junction obstruction, Upper moiety

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Introduction

Ureteral duplication is one of common anomalies affecting the genitourinary tract. Pelviureteric junction obstruction (PUJO) is the most common site of obstruction in the upper urinary system. PUJO in duplex system is an unusual entity and infrequently reported. Managing PUJO in duplex system is difficult for the treating surgeon. We are reporting our experience in managing PUJO in a duplex system with review of literature.

Objective of this study was to review the management and outcome of cases with pelvi-ureteric junction obstruction (PUJO) in a duplex system.

Materials and Methods

We retrospectively analyzed our record from January 2000 to December 2011. The inclusion criteria were all cases of PUJO in a duplex system and minimum postoperative follow up of 24 months. Exclusion criterias were incomplete data and presence of associated hypertension and other associated genitourinary anomalies.

Diagnostic investigations included renal ultrasonography, renal scintigraphy, diethylenetriamine Penta-acetic acid (DTPA), dimercaptosuccinic acid (DMSA), and magnetic resonance urography (MRU). The duplex was suspected on USG and renal scintigraphy and was confirmed by MRU. We have preferred MRU over DTPA simply because it provided the best functional and anatomical information needed by the surgeon dealing with duplex system. DTPA was used for quantifying differential renal function because of cost and availability as compared to MAG 3. MRU was done using heavily T2-weighted images; contrast enhanced T1-weighted MR sequences and maximum intensity projection (MIP) after proper hydration in 1.5T MR scanner imaging. Patients were sedated with trichlorofos (pedicloryl) in the dose of 50 mg/kg. Intravenous fluids were given as per body weight to maintain proper hydration. Intravenous gadolinium contrast was used for the study for image acquisition using abdominal or body coil, with patient in supine position and coil positioned over upper abdomen and centred on kidneys. After initial localizing images were obtained in the following

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sequences: T2 HASTE single slice, T2 HASTE multislice, 3D GRE T1, TRUFI 2D, FL2D 80 and TSE FS T2.

Post contrast images in T1 sequences were obtained. MR urography as a single investigation gives an excellent anatomical description of the duplex renal moieties even when they are nonfunctioning. The MRU is a noninvasive examination method that does not entail ionizing radiations and does not require iodinated contrast as in intravenous urogram (IVU) or computed tomography urogram (CTU) and safer in children. It is excellent in identifying non-functioning or poorly functioning duplex system. The heavily weighted T2 images best pick up the dilated non functioning moieties using static fluid as hyperintence images This technique does not require contrast. The T1-weighted images pick up non dilated and functioning unit as hyperintence. Various studies have shown almost 99% sensitive and more than ninety five percent specific for the diagnosis of the duplex renal system by MRU.[1-2] The complete duplex system was defined as kidney with 2 pyelocaliceal systems and 2 ureters that drained separately into the urinary bladder. The incomplete duplex system was defined as kidney with 2 pelvicaliceal systems with a single ureter or with a bifid ureter which joined before draining into the urinary bladder. For grading of severity of hydronephrosis, SFU grades were used. All cases were operated by an experienced single surgeon using dorsal lumbotomy approach. Cystoscopy was performed to document ureteric openings and their anomalies. Presenting symptoms and indications for surgery were as shown in table1.

Pyeloplasty was done with double J stent in all cases and stents were removed after 6 weeks following surgery. Postoperatively patients were followed up with USG abdomen and renal dynamic scans done at 3 months, 6 months and one year after surgery. Follow up period ranged from 24 to 111 months. All cases were given chemoprophylaxis till 6 months after surgery.

TABLE 1. Presentation and Indications for surgery in PUJO in duplex system

Sl no.	Presenting symptoms (n), (%)	Indications for surgery
1.	UTI (6) (35.2) Recurrent UTI (3) (17.6) Febrile UTI (1) (5.8) VUR (2) (11.7)	Recurrent UTI, Febrile UTI , and VUR
2.	Lump (4) (23.5)	Lump
3.	Pain (4) (23.5)	Pain and Differential renal function less than 40%
4.	ANH (3) (17.6)	More than 10% fall in differential renal function

Results

A total of 258 pyeloplasties in a single system and 17 pyeloplasties in duplex system were performed during this period. There were 11 males and 6 females. Patient characteristics were as shown in table 2.

Median age at surgery was 40.11 months (range 2-120 months). Complete duplex was seen in 8 (47.0%) cases while incomplete duplex was seen in 9 (52.9%) cases. Six (35.2%) cases presented with UTI which includes 3 cases of recurrent UTI (17.6%), 1 case of febrile UTI (5.8%) and 2 cases of vesicoureteric reflux (VUR) (11.7%) respectively. Antenatal diagnosis was present in 3 (17.6%) cases. Ipsilateral lump and pain was seen in 4 (23.5%) cases each. Mean age of all four cases who presented with lump was 55 months (range 33-91 months). The renal pelvis anterior posterior (AP) diameter was 7.3 cm, 6.9 cm, 6.3 cm and 5.7 cm respectively. Renal dimensions were 14.2 x 8.5 cm , 11.8 x 9.1 cm, 12.7 x 10.2cm and 10.6 x 9.1 cm respectively .The left side was affected in 10 (58.8%) cases while the right side in 7 (41.1%). Upper pole moiety was involved in 10 (58.8%) cases while lower pole moiety was seen in 7 (41.1%). Anderson-Hynes pyeloplasty was performed in 11 (64.7%) cases while in 3 (17.6%) ureteropyelostomy was done. Two (11.7%) cases required hemineprectomy, both cases presented with delayed presentation and poor renal function (<10%) of upper moiety. Ureteroureterostomy was performed in 1 (5.8%) case in which caudal ectopia was present. This child having a caudal ectopia did not have a megaureter. Six cases (35.2%) developed immediate postoperative complication, ipsilateral lump in 3 (17.6%) requiring external drainage in the form of percutaneous nephrostomy, febrile UTI in 2 (11.7%) and UTI in 1 (5.8%) case. Mean Follow up period was 59.5 months ranged from 24-111 months.

Discussion

PUJO is commonly seen with a single system with the girls more affected than boys. These cases are now-a-days diagnosed prenatally.[3,4] The reported incidence of PUJO in a duplex system varies from 2-7%. The PUJO in a duplex system was first described by Freyer and Deming in 1942 in the lower pole.[5] Since 1976; various series have been published in the English literature describing PUJO in a duplex system (Table3).[6-9]

Historically it was considered that PUJO in a duplex system affects the lower moiety but now it has become apparent that the proximal ureter of an upper moiety can also be obstructed.[10-13] Anatomically, the lower segment is the analogue of a single renal system with usually about two-

TABLE 2. Demographic profiles of cases and their mean follow up

Patient	Age at surgery (Months)	Sex	Side	Site	Anatomy	Presentation	Treatment	Follow up (Months)
1.	2	M	L	LP	C	ANH	Pyeloplasty	60
2.	9	M	R	LP	I	UTI	Pyeloplasty	84
3.	15	M	R	LP	C	ANH	Pyeloplasty	120
4.	36	F	L	UP	I	UTI	Pyeloplasty	72
5.	42	M	R	LP	I	LUMP	Pyeloplasty	48
6.	38	M	L	UP	C	PAIN	Ureteropyelostomy	36
7.	24	F	R	UP	C	UTI	Pyeloplasty	92
8.	120	M	L	UP	I	PAIN	Heminephrectomy	36
9.	91	M	L	UP	I	LUMP	Heminephrectomy	29
10.	60	F	R	LP	C	PAIN	Ureteropyelostomy	53
11.	4	F	L	UP	I	ANH	Pyeloplasty	111
12.	11	F	L	UP	C	UTI	Pyeloplasty	73
13.	19	M	L	UP	I	UTI	Pyeloplasty	33
14.	54	M	R	LP	C	LUMP	Ureteropyelostomy	46
15.	16	F	L	UP	C	UTI	Pyeloplasty	59
16.	33	M	L	LP	I	LUMP	Pyeloplasty	36
17.	82	M	R	UP	C	PAIN	Ureteropyelostomy	24

TABLE 3. Various published series in duplex system with PUJO

Authors	Sex M/F	Age	Total patients	Side R/L	Site UP/LP	Anatomy C/I	Presentation	Treatment
Joe et al.[7]	2 / -	Neonates	2	NA/NA	- / 2	2 / -	ANH	NA
Cacciaguerra et al.[8]	- / 1	13 months	1	- / 1	1 / -	1 / -	UTI	Excision of fibrous band
Ho et al.[9]	4 / 3	3 weeks-14 years	7	NA/NA	3 / 4	2 / 5	ANH (4) Pain (1) UTI (1) Mass (1)	Pyeloplasty (4) Heminephrectomy (2) Pyelopyelostomy (1)
Fernbach et al.[10]	11 / 5	Neonates-9 years	17	6 / 11	- / 16	16 / -	NA	Pyeloplasty (10) be Pyelpureterostomy (2) Heminephrectomy (2) No operation (3)
Gonzalez et al.[4]	5 / -	6 weeks-6 years	5	1 / 4	- / 5	4 / 1	ANH (2) Pain (3)	Pyeloplasty (4) Ureteric calycostomy (1)
Horst and Smith[32]	9 / 2	1 month-13 years	11	3 / 8	3 / 8	4 / 7	ANH (9) Febrile UTI (2)	Pyeloplasty (8) Pyeloureterostomy (1) Upper-pole heminephrectomy (1)

thirds of the parenchyma and at least two calyces and a true renal pelvis.[11] That explains the predilection of PUJO for the lower moiety. In our study group obstruction in upper pole moiety was seen in 10 (58.8%) cases while in 7 (41.1%) lower pole moiety was affected. The complete duplex system was found in 8 (47.0%) cases while incomplete system in 9 (52.9%) cases. Our study is in concordance with other reported series.[8-10],[13-22] Fernbach *et al* in 1995 reported higher incidence of complete duplex system with PUJO of lower pole in males as compared to females.[10] The reported incidence of complete duplication is 0.2%.[12] The left sided lesion was more common than right *i.e* 10 (58.8%) versus 7 (41.1%) respectively. The clinical presentation of these patients is similar to that in single-system PUJO. In our study group most common presentation was UTI which was seen in 6 (35.2%) followed by a lump, and pain in 4 (23.5%) cases each. ANH was seen in 3 (17.6%) cases. All cases were investigated with USG, renal scintigraphy followed by MRU to clearly delineate the surgical and functional anatomy as per our routine protocol. In all cases cystoscopy was performed followed by a definitive surgical procedure using lumbotomy approach.[23] DJ stents were used in all cases. Immediate postoperative complications were seen in 6 (35.2%) cases as explained previously. Chemoprophylaxis was given till 6 months post surgery in all cases. Initially follow up were done at 3, 6, and 12 months after surgery with USG and RDS followed by annual surveillance with alternate USG and RDS.

In our study group the overall complication rate was 64.7% which appears to be too high simply because of small sample size. We could not find any series documenting their complication rate in the English literature except Avlan D *et al* who had reported no complications in seven cases of lower moiety PUJO.[24] There is no such series available which has compared complication rate in single system versus duplex system PUJO. If individual series are considered, the complication rate in single system ranges from 10-15%.[25-27] Managing PUJO in duplex system is a challenging task for the treating surgeon. Careful radiological evaluation of an obstructed duplex system is important, especially to decide the best surgical approach. MRU can be used as the primary diagnostic method for assessing a duplex ectopic ureter even when not picked up by conventional radiological investigations.[28-29] Not only the surgical anatomy but also the functional status of the kidney unit dictates the type of procedure required. The use of DMSA for assessment of differential function is of great help in managing cases of PUJO with duplex system. The management of upper urinary tract obstruction should follow similar guidelines to those used for the single-system PUJO, with the intention to improve drainage and to save as much functioning tissue as possible. Various surgical options available are pyeloplasty, pyeloureterostomy, calyceoureterostomy, ureterostomy and heminephrectomy. In the present era of minimal surgery laparoscopic intervention in duplex system has been reported by various authors with comparable results.[30-32] The indication of reconstructive surgery or heminephrectomy depends on the morphology and function of the involved renal moiety, ureter, and possible presence of a lateralised or ectopic ureteric ureteral orifice, or ureteral stenosis. We are of the opinion that preoperative evaluation with USG, renal scintigraphy, and MRU followed with cystoscopy before the definitive surgical procedure adds in the proper diagnosis and treatment allowing the best surgical approach to be planned. The limitations of this study include that the subjects were retrospectively analyzed and the sample size was relatively small. Hence, a prospective study with a larger sample will be required to enhance the validity of this study in the future.

Conclusion

There are a multitude of surgical options to deal with PUJO in duplex system and management should be based on the pathological and functional anatomy and the experience of the surgeon.

References

1. Campbell MF. Embryology and anomalies of the urogenital tract. In: Clinical paediatric urology. Kelalis PP, King LR, Belman AB. (eds). Philadelphia, PA: WB Saunders; 1976. p. 159.
2. Pearlman ES, Rosenfield AT, Wexler JS, Glickman MG. CT urography in the evaluation of urinary tract diseases J Comput Assist Tomogr 1996;20:620-6.
3. Kaplan WE, Nasrallah P, king LR. Reflux in complete duplication in children. J Urol 1978;120:220-2.
4. Parks, Han SS, Choi HK. Duplicated ureters: 2 cases treated with ipsilateral ureteroureterostomy (end to side anastomosis). Korean J Urol 1985;26:339-42.
5. Freyer S, Deming CL. Congenital hydronephrosis in the lower half of a double kidney; report of a case. N Engl J Med 1942;226:220-23.
6. Gonzalez F, Canning DA, Hyun G, Casale P. Lower pole pelvi-ureteric junction Obstruction in duplicate collecting systems. BUJ Int 2006;97:161-5.
7. Jee LD, Rickwood AM, Williams MP, Anderson PA. Experience with duplex system anomalies detected by prenatal ultrasonography. J Urol 1993;149:808-10.
8. Cacciaguerra S, Arena C, Rizzo L, Distefano A, Di Benedetto AG, DiBenedetto A. Pyeloureteral junction obstruction in upper moiety of duplex system. Scand J Urol Nephrol 1995;299:511-5.

9. Ho DS, Jerkins GR, Williams M, Noe HN. Ureteropelvic junction obstruction in upper and lower moiety of duplex renal systems. Urology 1995;45:503-8.
10. Fernbach SK, Zawin JK, Lebowitz RL. Complete duplication of the ureter with ureteropelvic junction obstruction of the lower pole of the kidney. AJR Am J Roentgenol 1995;164:701-4.
11. Fernbach SK, Feinstein KA, Spencer K, Lindstrom CA. Ureteral duplication and its complications. Radiographics 1997;17:109-27.
12. Amis ES, Cronan JJ, Pfister RC. Lower moiety hydronephrosis in duplicate kidneys. Urology 1985;26:82-8.
13. Wah TM, Weston MJ, Irving HC. Lower moiety pelvic-ureteric junction obstruction (PUJO) of the duplex kidney presenting with pyonephrosis in adults. Br J Radiol 2003;76 909-12.
14. Koyanagi T, Takamatsu T, Terashima M, Nonomuri N, Tsuji J. Internal vascular obstruction of superior ureteropelvic junction causing nephralgia in a woman with complete duplex kidney. Int Urol Nephrol 1978;10:267- 74.
15. Jerkins GR, Noe HN. Unusual presentation of ureteropelvic junction obstruction in an incomplete duplex system. Urology 1985;26:402-4.
16. Ossandon F, Androulakakis P, Ransley PG. Surgical problems in pelvicoureteral junction obstruction of the lower moiety in incomplete duplex systems. J Urol 1981;125:871-2.
17. Armor AD. Congenital hydronephrosis of lower segment in duplex kidney. Urology 1976;7:480-5.
18. Joseph BJ, Stuart BB, Colodny AH, Mandell J, Lebowitz RL, Retik AB. Lower pole ureteropelvic junction obstruction and incomplete renal duplication. J Urol 1989;141:896-9.
19. Dewan PA, Penington E, Jeyaseelan D. Upper pole pelviureteric junction obstruction. Pediatr Surg Int 1998;13:290-2.
20. Yamamoto N, Takeuchi T, Hayashi H, Miwa K, Tanoguchi M, Sakai S. Successful management of lower pole ureteropelvic junction obstruction in a partially duplicated collecting system using retrograde endoureteropyelotomy with the Holmium: YAG laser. J Urol 2001;166:2301-2.
21. Shelfo SW, Keller MS, Weiss RM. Ipsilateral pyeloureterostomy for managing lower pole reflux with associated ureteropelvic junction obstruction in duplex systems. J Urol 1997;157:1420-2.
22. Ulchaker J, Ross J, Alexander F, Kay R. The spectrum of ureteropelvic junction obstruction occurring in duplicate collecting systems. J Pediatr Surg 1996;31:1221-4.
23. Bajpai M, Kumar A, Gupta AK, Pawar DK. Lumbotomy approach for upper urological tract surgery in children--an analysis of 68 consecutive lobotomies. Eur J Pediatr Surg 2004;14:163-7.
24. Avlan D, Gündo?du G, Deliba? A, Nayci A. Pyeloureterostomy in the management of the lower pole pelvi-ureteric junction obstruction in incomplete duplicated systems. Urology 2010;76:1468-71.
25. Braga LH, Lorenzo AJ, Farhat WA, Bägli DJ, Khoury AE, Pippi Salle JL. Outcome analysis and cost comparison between externalized pyeloureteral and standard stents in 470 consecutive open pyeloplasties. J Urol. 2008;180:1693-8.
26. Smith KE, Holmes N, Lieb JI, Mandell J, Baskin LS, Kogan BA *et al*. Stented versus nonstented paediatric pyeloplasty: a modern series and review of the literature. J Urol. 2002;168:1127-30.
27. Elmalik K, Chowdhury MM, Capps SN. Ureteric stents in pyeloplasty: a help or a hindrance?. J Pediatr Urol 2008;4:275-9.
28. Avni FE, Nicaise N, Hall M, Janssens F, Collier F, Matos C *et al*. The role of MR imaging for the assessment of complicated duplex kidneys in children: preliminary report. Pediatr Radiol 2001;31: 215-23.
29. Yanagisawa N, Yajima M, Takahara T, Nosaka S, Iwamoto T. Diagnostic magnetic resonance-urography in an infant girl with an ectopic ureter associated with a poorly functioning segment of a duplicated collecting system. Int J Urol 1997;4:314-7.
30. Lowe GJ, Canon SJ, Jayanthi VR. Laparoscopic reconstructions for obstruction in children with duplex renal anomalies. BJU int 2008;101:227-30.
31. Hisanom. Denes FT, Brinto AH , Lucon M, Machado MG, Bruschini H *et al*. Laparoscopic ureteropyeloanastomosis in the treatment of duplex system. Int Braz J Urol 2012;38:235-41.
32. Horst M, Smith GH. Pelvi-ureteric junction obstruction in duplex kidney BJU Int 2008;101:1580-4.

Original Article

Cloacal Exstrophy: A Histomorphological Analysis of the Bladder Plate and Correlation with Bladder Dynamics

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Abstract. ***Objective:*** Physiological outcomes in patients of cloacal exstrophy are governed primarily by neurological innervation and detrusor histomorphology. An evaluation of the bladder plate histomorphology for the presence of neural elements, collagen fibres and smooth muscle components was done and correlation attempted with detrusor physiology. ***Methods:*** From three cases of cloacal exstrophy, full thickness biopsies were taken from the bladder plate at the time of single stage total reconstruction and slides stained with hematoxylin and eosin, masson's trichrome and immunohistochemical staining done with S-100, SMA and CD-117. Bladder biopsy taken at the time of cystolithotomy from a child with vesical calculus served as a control. ***Results:*** Cloacal exstrophy bladders showed squamous metaplasia, submucosal edema, increased collagen in the muscle layer and poor staining with S-100 (neural elements) and CD-117 (interstitial cells). Whereas the control biopsy showed normal transitional epithelium, good muscle layer with presence of both neural elements (S-100) and interstitial cells (CD-117). ***Conclusion:*** Poor muscle layer with increased fibrosis and scarcity of neural elements in the bladder wall may account for the poor compliance of such bladders and explain the incontinence and upper tract deterioration due to high intravesical pressure.

Keywords: Bladder plate, Cloacal exstrophy, Collagen, Histomorphological analysis, Metaplasia

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Introduction

Cloacal exstrophy is a rare congenital paediatric urological problem, the management of which continues to challenge the paediatric surgeons not only for anatomical optimisation but also for improving the long term outcomes and quality of life-social, psychological, physical and physiological. It is the extreme end of a spectrum of defects that can occur in the formation of the ventral abdominal wall and represents one of the most severe anomalies that is compatible with viability. This entity is extremely rare, occurring in 1 in 200,000-400,000 live births.[1-3] The male to female ratio has most recently been reported in a large contemporary study to be equal between the sexes, 1:1.[4]

The goals of surgical reconstruction in these patients are to correct the urogenital and gastrointestinal defects providing a reservoir that is adequate for urinary storage at low pressures with the ability to empty completely without compromising renal function, to create functional and cosmetically acceptable external genitalia, and to maximizing patient quality of life. Urodynamics in such cases is governed primarily by two entities-the neurological innervations and the detrusor histomorphology. Neurospinal abnormalities have been noted in 85-100% of patients with cloacal exstrophy with a distribution of lumbar (80%), thoracic (10%) and sacral defects (10%).[5] The presence of a significant neurologic deficit is associated negatively with the ability to develop continence.[6] Various authors have tried to correlate the detrusor histomorphology with muscle contractility in bladder exstrophy patients and few cases have been similarly reported for cloacal exstrophy.[7-8]

We have, here, attempted to correlate the histomorphological findings with neurological outcomes in terms of increase in capacity and detrusor contractility over time.

Materials and Methods

Three neonates presented to our department over a period of one year from July 2012 - June 2013 with a classical exstrophy of the cloaca. All three underwent single stage total

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reconstruction of the defect after optimal stabilisation. Intra-operatively full thickness biopsies were taken from the bladder plate and fixed in formalin. Thereafter, slides prepared from the tissue specimen were stained with Hematoxylin-Eosin and Masson's trichrome stains and immunohistochemical staining was done with S-100, CD-117 (c-kit) and SMA (smooth muscle antigen).

Full thickness bladder biopsy during a cystolithotomy for a primary vesical calculus was used as a control to compare the findings. The distribution of various parameters in the specimen was subjectively analyzed without any definite measurements. An average analysis of the three cases' histopathological findings was taken.

Results

All three cloacal exstrophy cases were consistent in their histomorphology with marked variation as compared to the control *i.e* apparently healthy bladder. In contrast to the normal-looking transitional epithelium of the control (Fig. 1a and Fig. 1c), the mucosa of the bladder plate in the cloacal exstrophy cases had squamous metaplasia (Fig. 1b and Fig.1d). The submucosa showed severe edema and congestion in the cases vis-a-vis mild congestion in the control. The muscle layer contained only healthy muscle fibres in the control (Fig. 1e) whereas both collagen fibres as well as hypertrophied smooth muscle elements constituted the muscle layer in the cloacal exstrophy specimens (Fig. 1f).

On immunohistochemical analysis, scarcity of neural elements, as denoted by poor expression of S-100, was noted in the cases (Fig. 2d). Also, interstitial cells were markedly reduced in the cases as visualised on staining with CD-117 (c-kit) (Fig. 2b). SMA staining showed marked hypertrophy of the muscle bundles in the cases. All these findings were in contrast with a healthy muscle layer in the control specimen with adequately present neural elements (Fig. 2c) and interstitial cells (Fig. 2a).

Comparison of the histological findings of cloacal exstrophy bladder plate with an apparently healthy bladder tissue is described in table 1.

Discussion

Ultrastructural changes in the bladder plate have been looked into in cases of classical bladder exstrophy many times in literature. Mathews *et al* obtained bladder biopsies from 32 patients of bladder exstrophy at various stages of reconstruction and evaluated them for various histological parameters and found that patients with good parameters had good outcomes in terms of increased bladder capacity with age. They found an increase in the collagen fibres in the muscle layer as well as a decrease in the smooth muscle to collagen ratio stating thereby, that increased collagen is responsible for the decreased compliance of these bladders and their poor ability to 'catch-up' their capacity with increasing age of the child.[7]

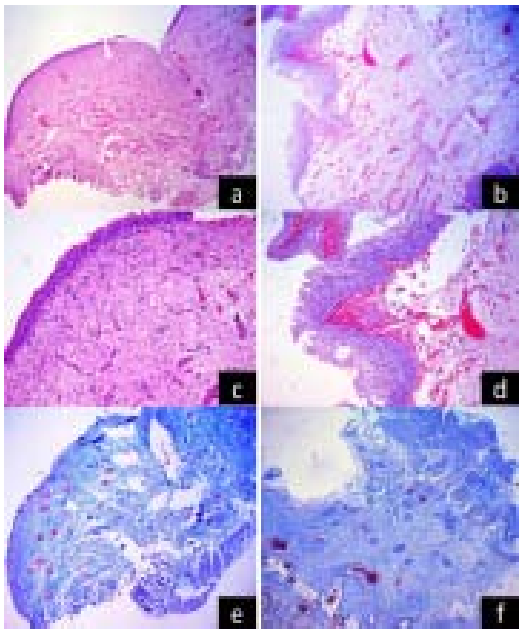

Fig. 1. Fig. 1a. & Fig. 1c. Photomicrograph shows mucosal fragment lined by transitional epithelium [H&E x40, cx100], Fig. 1b. & Fig. 1d. Photomicrograph shows mucosal fragment lined by transitional epithelium with marked squamous metaplasia and submucosal edema [H&Ex40, ex100], Fig. 1e. Masson's trichrome stain shows minimal collagen [cx100], Fig. 1f. Masson's trichrome stain with increased collgen and also fibrosis of muscle layer [fx100]

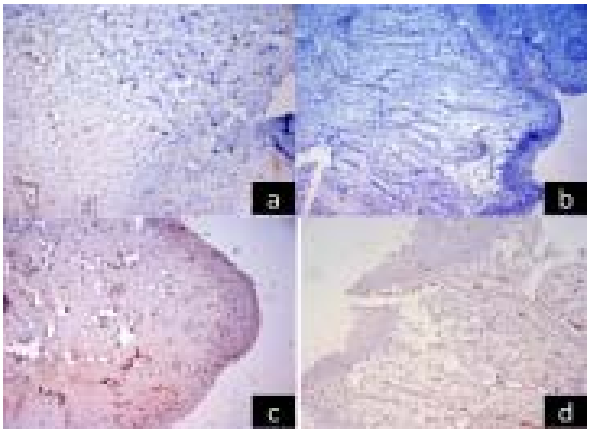

Fig. 2. Fig. 2a. Immunohistochemistry shows few cells positive for CD117 [x100], Fig. 2b. Cloacal exstrophy bladder plate showing absence of CD117 positive cell [x100] Fig. 2c. Presence of nerve bundles positive for S100 [x100] Fig. 2d. Decreased nerve bundles [x100]

Table 1. Comparison of the histological findings of cloacal exstrophy bladder plate with an apparently healthy bladder tissue

	Control bladder biopsy	Cloacal exstrophy bladder biopsy
Mucosa (H-E)	Transitional epithelium normal (Fig. 1a & Fig. 1c)	Squamous metaplasia (Fig. 1b & Fig. 1d)
Submucosa	Mild congestion	Severe edema, congestion
Muscle	Normal muscle fibres	Haphazardly arranged and hypertrophied
Masson's trichrome	Few collagen fibres in submucosa (Fig. 1e)	Collagen fibres both in submucosa and muscle layer (Fig. 1f)
IHC-S-100	Nerve bundles present (Fig. 2c)	Markedly decreased (Fig. 2d)
IHC-CD-117	Interstitial cells present (Fig. 2a)	Markedly decreased (Fig. 2b)
IHC-SMA	Normal muscle	Hypertrophy

In the present report, similarly, an increase in the collagen elements was represented by an increase in the Masson's trichrome staining in the muscle layer of the specimens. It has been reported that kit-positive interstitial cells in the submucosal layer can play a key role in communicating stimulation to afferent nerves in the bladder.[9] Moritoki *et al* evaluated the bladder plate and hindgut biopsy in a cloacal exstrophy patient and tried to correlate the findings with bladder neuropathic dysfunction. Fewer kit-positive interstitial cells were found in the submucosal layer in the cloacal exstrophy tissue than in the normal bladder tissue. Hence, they hypothesized that fewer neurons and interstitial cells in the bladder submucosa of a cloacal exstrophy patient may be responsible for its neuropathic nature.[8]

Likewise, markedly decreased expression of CD-117 (c-kit) and S-100 was noted signifying scarcity of interstitial cells and neural elements respectively in these cases. These important findings could mean poor neural activity in the wall of such bladders which will ultimately present as 'bad bladders' with poor compliance and impaired growth potential. Such 'bad bladders' can in future pave the way for upper tract deterioration as a consequence of a high pressure system. Likewise, continence in such cases is a function of altered bladder dynamics. Anticholinergics are routinely used in bladders with small capacity and high pressures, followed by adjuncts, like clean intermittent catheterization and bladder augmentation in different permutations and combinations, where necessary.

The drug "Oxybutynin hydrochloride" has brought a paradigm shift in the outlook of patients with high pressure, small capacity bladders with a so-called 'normal' wall with a 'normal' muscle layer.[10] The anti-cholinergic action against the M3 muscarinic receptors on the wall of detrusor muscle relaxes it and thereby decreases intravesical pressures and indirectly increases bladder capacity and pharmacologically converts it into an inactive reservoir.[11] However, based upon on our observations on the histomorphology in the present study, we believe, that the same will have questionable application.

With increasing knowledge on the surgical management of these cases, survival is approaching 100%[12] and now the emphasis is shifting on improving the quality of life of these vulnerable children. The above findings lead us to believe that these bladders are, in a way, 'neuropathic' with respect to poor neural innervation and coupled with an inelastic wall constantly pose a threat to the upper tracts.

Conclusion

This is the first reported case series of cloacal exstrophy bladder plate histomorphology. We have tried to correlate these with the bladder dynamics and upper tract changes. We hope to further our observations on more specimens and include newer histomorphological parameters in literature to come.

References

1. Ziegler M, Duckett JW, Howell JG. Cloacal exstrophy. In: Welch J, (ed). Paediatric Surgery, Chicago: Year Book Medical Publishers, 1986.
2. Tank ES, Lindenauer SM. Principles of management of exstrophy of the cloaca. Am J Surg 1970;119:95-8.
3. Hurwitz RS, Manzoni GA, Ransley PG, Stephens FD. Cloacal exstrophy: a report of 34 cases. J Urol 1987;138:1060-4.
4. Schober JM, Carmichael PA, Hines M, Ransley PG. The ultimate challenge of cloacal exstrophy. J Urol 2002;167:300-4.
5. McLaughlin KP, Rink RC, Kalsbeck JE, Keating MA, Adams MC, King SJ *et al*. Cloacal exstrophy: the neurological implications. J Urol 1995;154:782-4.

6. Husmann DA, Vandersteen DR, McLorie GA, Churchill BM. Urinary continence after staged bladder reconstruction for cloacal exstrophy: the effect of coexisting neurological abnormalities on urinary continence. J Urol 1999;161:1598-602.

7. Mathews R, Gosling JA, Gearhart JP. Ultrastructure of the bladder in classic exstrophy: correlation with development of continence. J Urol 2004;172:1446-9.

8. Moritoki Y, Kojima Y, Mizuno K, Kamisawa H, Kohri K, Hayashi Y. Histopathologic analysis of bladder in patient with cloacal exstrophy. Urology 2012;79:1368-71.

9. Johnston L, Woolsey S, Cunningham RM, O'Kane H, Duggan B, Keane P *et al*. Morphological expression of KIT positive interstitial cells of Cajal in human bladder. J Urol 2010;184:370-7.

10. Youdim K, Kogan BA. Preliminary study of the safety and efficacy of extended-release oxybutynin in children. Urology 2002;59:428-32.

11. Andersson KE, Chapple CR. Oxybutynin and the overactive bladder. World J Urol 2001;19:319-23.

12. Diamond DA, Jeffs RD. Cloacal exstrophy: a 22-year experience. J Urol 1985;133:779-82.

Original Article

Early Experience of Lumboscopy in Paediatric Upper Urinary Tract Calculi

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Abstract. ***Objective:*** Objective of this study was to present our early experience with lumboscopy in treatment of paediatric urolithiasis of upper urinary tract. ***Methods:*** Paediatric urolithiasis cases treated by lumboscopy approach from July 2013 to December 2013 were included in this prospective study. Lumboscopy was performed in prone position using three ports to approach kidney and ureter in all patients. The patient characteristics were analysed. Intraoperative and postoperative details were taken. Data was analyzed regarding age, sex, diagnosis, surgical procedure, anaesthetic details, intraoperative problems encountered, postoperative pain, time to oral feed, length of hospitalization, complications and final outcome. ***Results:*** Between July 2013 and December 2013, a total of 6 patients underwent lumboscopic procedures for upper urinary tract calculi. There were 5 males and one female; median age at surgery was 7.5 years (range 2-12 years). Four patients underwent lumboscopic pyelolithotomy, one lumboscopic ureterolithotomy and one lumboscopic bilateral ureterolithotomy and pyelolithotomy. The mean operating time was 120 minute; range 80-200 minute. There were no special anaesthetic requirements and no intraoperative problems were encountered. There was minimal postoperative pain. All the patients were started orally within 24 hours. The mean duration of hospital stay was 5 days; range 3-10 days. There were no incision related or port site complications. All the patients were asymptomatic in the follow up period. ***Conclusion:*** Lumboscopy is an easy, effective, technically safe, low morbidity alternative for management of upper urinary tract calculi in paediatric population.

Keywords: Lumboscopy, Paediatric urolithiasis, Pyelolithotomy, Ureterolithotomy

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Introduction

Paediatric urolithiasis is an uncommon occurrence but the incidence has been increasing over the last decade. It poses a technical challenge to the paediatric urologist. Despite the success of minimally invasive techniques, open surgery is still commonly practiced for treatment of paediatric urolithiasis. The wide spectrum of paediatric upper urinary tract calculi warrants for an approach which focuses on decreasing morbidity, cosmetically acceptable smaller incisions and technically feasible access. The lumboscopy approach fits into the requirements and is an affordable option.

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We are reporting our early experience with lumboscopy in children suffering from urolithiasis of upper urinary tract.

Materials and Methods

This prospective study was conducted from July 2013 to December 2013 and included children presenting with upper urinary tract stones managed via lumboscopic approach. Cases with parents/guardians refusing consent for the procedure, history of any prior surgery were excluded. Diagnostic investigations included X-ray kidney, ureter, bladder (KUB) and noncontrast computed tomogram (NCCT) KUB with 3D reconstruction (Fig. 1). The patient's characteristics were analyzed. Intraoperative and post operative details were noted. Data was collected regarding age, weight, sex, diagnosis, surgical procedure, anaesthetic details, intraoperative problems encountered, postoperative pain, time to oral feed, length of hospitalization and complications. All patients were operated by single surgeon. The data from anaesthetic chart was also analysed regarding difficulty in maintaining the airway in prone position,

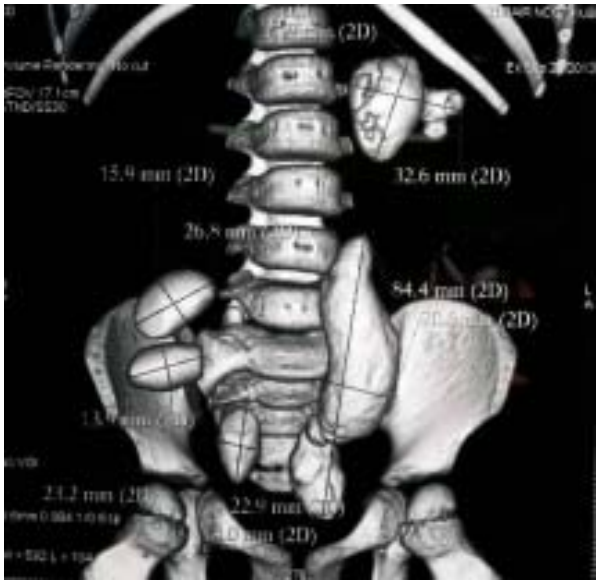

Fig. 1. Non contrast computed tomography of kidney, ureter and bladder (NCCT KUB) with 3D reconstruction showing stones in both ureters and left pelvicalyceal system

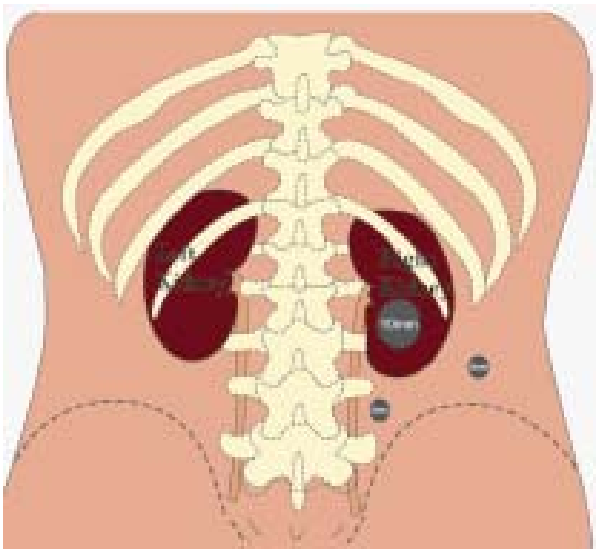

Fig. 2. Port positions used in lumboscopy

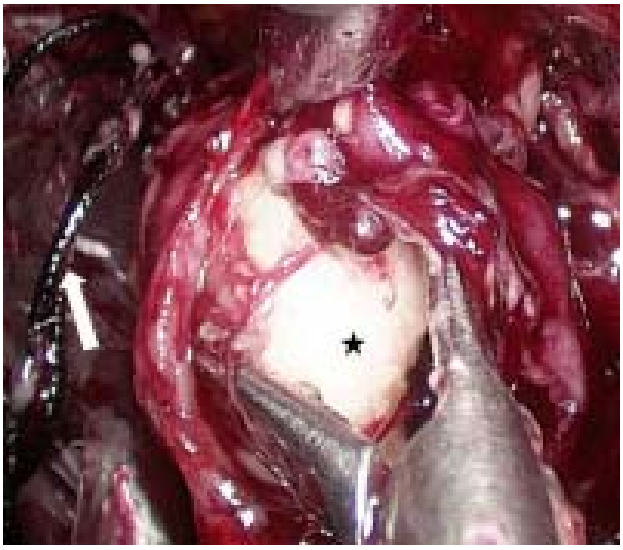

Fig. 3. Lumboscopic view showing a stone being delivered out of ureter (black asterix: stone, white arrow: stone retrieval bag)

intravenous analgesia requirements and any intraoperative hemodynamic event such as bradycardia, fall in blood pressure and electro-cardio-graphic (ECG) changes.

The procedures were performed after informed consent as per hospital protocol. Children were intubated after standard intravenous and inhalational anaesthesia. Prone position was maintained by roll under the pelvis, thick pad under the shoulders and upper chest leaving the thorax and abdomen free for movements. Head was turned to one side for securing the airway. The following landmarks were identified: 12th rib, vertebral column, erector spinae and the iliac crest. A small 12mm incision was given at the renal angle at the lateral margin of paraspinal muscles. Posterior lamina of the dorsolumbar fascia was pierced vertically. A retroperitoneal space was created using blunt and sharp dissection. Gerota's fascia was then opened. Indigenous balloon made using the middle finger of a surgical glove (No 8) fixed at the end of a suction catheter was inserted into the space created and gradually inflated by insufflating 200-300ml of air. After insufflation (which was gradual and over 5-10 min) we waited for 20-25 minutes for sufficient creation of the space. Once the space was created a 12mm port was inserted and 10mm telescope inserted to visualize the retroperitoneum. Standard three port technique was used (Fig. 2). Subsequent 5mm ports were then inserted under vision and fixed. Urinary tract (ureter or pelvis) was identified and opened with laparoscopic scalpel and stone was removed intact using indigenously designed stone retrieval bag (Fig. 3). Double J stent was placed and urinary tract closed with 4-0 absorbable sutures. Penrose drain was placed in all the cases. Operating time, intraoperative complications, postoperative pain, time to oral feeds, duration of hospital stay and final outcome were analyzed.

Results

A total of 6 children underwent lumboscopic procedures for upper urinary tract calculi from July 2013 to December 2013. Five were males while one was a female. The median age at surgery was 7.5 years; range 2 to 12 years. Four had renal calculi, one had ureteric calculi of one side and one had renal and bilateral ureteric calculi. Children with renal calculi

underwent lumboscopic pyelolithotomy, while in cases of ureteric calculi lumboscopic ureterolithotomy was performed. One case underwent lumboscopic bilateral ureterolithotomy and pyelolithotomy, each side approached separately. The mean operating time was 120 minutes with a range of 80-200 minutes. There were no special anaesthetic requirements and no intraoperative problems were encountered. None of our patients experienced any significant anaesthetic problem. All of the patients were maintained on intravenous and inhalational anesthetic agents. Intraoperative analgesia was in form of intravenous fentanyl (one microgram/kg) and all patients were extubated smoothly. There was minimal postoperative pain reported in all cases which was managed in form of intravenous paracetamol (PCM) on first day and oral PCM on the day after. Intravenous fluids were stopped by next morning and oral feeds were encouraged. All the patients were started orally within 24 hours of surgery. Penrose drain was removed on post-operative day 2 or 3. The mean duration of hospital stay was 5 days; range 3-10 days. There were no incision related or port site complications. None of our patients had wound disruption or infection. All of them were asymptomatic in the follow up period.

Discussion

With the adoption of evidence based medicine in past couple of decades the practice of surgery has been more or less standardized. The newer concepts and approaches are frequently questioned and tested in relation to the established practices. The surgeon by his own nature keeps innovating and practicing such techniques which he finds more suitable and convenient. Such techniques often evolve in the process of improvisation and meeting needs of the time. Surgeries involving the retroperitoneal structures like kidney are quite common in paediatric age group. The wide spectrum of urological diseases in these age groups warrants for an approach which focuses on decreasing morbidity, cosmetically acceptable smaller incisions and technically feasible access.

Paediatric urolithiasis though uncommon as opposed to adults, has shown an increase in incidence in recent years. Use of minimally invasive techniques has been a major breakthrough in paediatric urology in the past decade. Despite this, the management of paediatric urolithiasis remains a surgical challenge. More and more such cases are now being managed laparoscopically with the improved expertise of the operating surgeon and availability of paediatric laparoscopic instruments. Laparoscopy can be performed by transperitoneal or retroperitoneal approach. Transperitoneal route has the obvious disadvantage of peritoneal breach and resultant complications which can be avoided in retroperitoneal approach.

Lumboscopy with gas insufflation was described in an animal model by Roberts and in humans by Wikham in 1976, who first performed retroperitoneal endoscopic ureterolithotomy using a standard laparoscope in 1979.[1] Laparoscopic procedures are still not widely performed in paediatric urology because of operating time and costs as well as the lack of indications and, thus, the lack of sufficient surgeon experience.[2]

Retroperitoneal approach provides a laparoscopic technique comparable to conventional renal surgery.[2] By avoiding the peritoneal cavity the risk of visceral and vascular injury may be reduced. Also, the risk of adhesive obstruction is eliminated. It is a more direct approach to the kidney and other retroperitoneal structures. In 1996, Valla *et al* reported their experience with 18 cases of retroperitoneal laparoscopic nephrectomies in children aged 3 months to 14 years. The median operating time was 106 minutes. There were no complications and only one conversion.[3] In a systematic review of 689 paediatric nephrectomies by Kim C *et al* in 2009, it was reported that retroperitoneal approach was associated with a shorter operative time, comparable hospital stay and overall complication rate compared to transperitoneal approach.[4]

The lumboscopy approach fits into the requirements and is an affordable option. A surgeon who is familiar with lumbotomy approach can perform lumboscopy and identify structures more easily and confidently than a real beginner for this approach. The data on lumboscopic treatment of paediatric urolithiasis is limited. In 2013, Agarwal *et al* reported their experience of 22 cases of children with urinary tract stones who were treated by transperitoneal laparoscopy. Twelve underwent pyelolithotomies while in 10 ureterolithotomy was performed. Complete removal of calculi was achieved in 21 (95.45%) and one required conversion to open surgery. One patient developed a small localized urinoma after laparoscopic pyelolithotomy while one had omental prolapsed on drain removal which was managed by repositioning and repair of fascia. Mean hospital stay was 3 days. None had recurrence or residual stones, in a mean follow up of 11 months.[5]

Senior author who did lumboscopy in all cases had vast experience of lumbotomy approach for upper urological diseases.[6-7] So it became easy and technically safe for the surgeon and his team members to do the procedure like lumboscopy for upper urinary tract stones. We used the three port technique with patient in prone position. Our mean operating time was 120 minutes and it improved as the number of cases increased. Postoperative recovery was very quick in these patients, early feeding was initiated and there was minimal postoperative pain, most were discharged without any complication. With increasing availability of small sized

laparoscopic instruments, lumboscopy holds great promise for treatment of paediatric urolithiasis.

Conclusion

Lumboscopy is an effective procedure for management of ureteric and renal calculi in paediatric population with low morbidity. We continue to use and advocate this approach as primary option for removal of all upper urinary tract calculi.

References

1. Wickham JEA. The surgical treatment of renal lithiasis. In: Wickham JEA, (ed). Urinary calculus disease. New York, Churchill Living; 1979:145-98.
2. El Ghoneimi A, Valla JS, Steyaert H, Aigrain Y. Laparoscopic renal surgery via a retroperitoneal approach in children. J Urol 1998;160:1138-41.
3. Valla JS, Guilloneau B, Montupet P, Geiss S, Steyaert H, El Ghoneimi A *et al*. Retroperitoneal laparoscopic nephrectomy in children. Preliminary report of 18 cases. Eur Urol 1996;30:490-3.
4. Kim C, McKay K, Docimo SG. Laparoscopic nephrectomy in children: systematic review of transperitoneal and retroperitoneal approaches. Urology 2009;73:280-4.
5. Agarwal V, Bajaj J, Acharya H, Chanchalani R, Raina VK, Sharma D. Laparoscopic management of paediatric renal and ureteric stones. J Paediatr Urol 2013;9:230-3.
6. Bajpai M, Kumar A, Gupta AK, Pawar DK. Lumbotomy approach for upper urological tract surgery in children--an analysis of 68 consecutive lumbotomies. Eur J Pediatr Surg. 2004;14:163-7.
7. Bajpai M, Kumar A, Tripathi M, Bal CS. Dorsal lumbotomy incision in paediatric pyeloplasty. ANZ J Surg. 2004;74:491-4.

Case Report

Cloacal Exstrophy with Intravesical Phallus: An Intra-operative Revelation in a case of OEIS Complex

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Abstract. Cloacal exstrophy is a complex congenital malformation and presents more often than not as a spectrum of anomalies. Gender assignment is difficult in these babies because of the ambiguity of the genitalia and the major surgical undertaking which may make the salvage of certain delicate structures an arduous task. Awareness of the variety of ways in which the phallus may be positioned with respect to the exstrophy patch aids surgeons in further reconstruction. We describe a neonate with an extremely rare variant of an intravesical phallus (eight published cases of this entity were found in literature) and discuss the implications of the same.

Keywords: Cloacal exstrophy, Intravesical phallus, Staged management

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Introduction

The many synonyms of cloacal exstrophy including "vesicointestinal fissure" and the descriptive term, "OIES complex", introduced by Carey, Greenbaum and Hall in 1978,[1-2] denote the same anatomic defect. The multitude of variations around the central theme of Omphalocele, Exstrophy, Imperforate anus and Spinal defects makes reconstruction a daunting task. Gender assignment is challenging because of the innate ambiguity of the genitalia which accompanies this most severe form of the exstrophy-epispadias defect. The entity of the intravesical phallus (revealed more often than not during surgical exploration) aids surgeons in taking the correct decision for the child entrusted in their care. We describe a neonate with this extremely rare variant and discuss the implications of the same.

Case Report

A 1.4 kg neonate presented to us on the first day of life with cloacal exstrophy, omphalocele, sacral agenesis and bilateral congenital talipes equinovarus. After initial stabilization, a thorough physical examination revealed an omphalocele with a large exstrophy patch which contained a prolapsed ileal segment (the "elephant trunk" deformity) (Fig. 1) and a caecal patch bounded on both sides by the hemibladders. A single ureteric opening was visible within the edematous mucosa. The genitalia were ambiguous and a possibility of widely separated bifid clitoris and labio/scrotal folds was kept in the absence of palpable testes. Aphallia was evident.

An ultrasound of the abdomen revealed orthotopic location of radiologically normal kidneys. An infantogram showed diastasis pubis and sacral agenesis which correlated with a 5

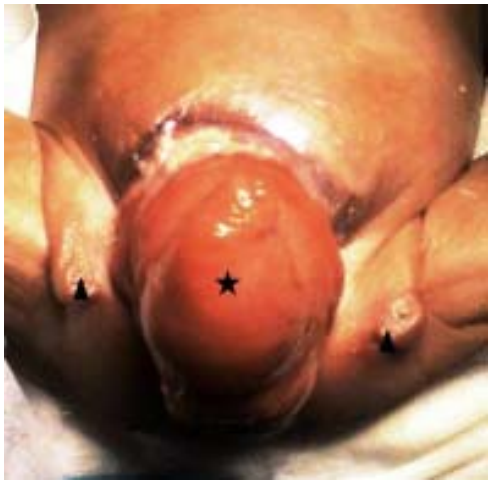

Fig. 1. Pre-operative photograph of the cloacal exstrophy with prolapsed ileal segment (the "elephant trunk" deformity) (asterix) and widely separated labio/scrotal folds (arrow head)

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x 4cm, soft, globular lump located in the midline just above the shallow natal cleft, possibly a lipoma. The baby was taken up for surgery after routine haematological and blood chemistry panels. The dilated ileum was found to open into the right side of the caecal patch. As in the classical description, two appendiceal openings and approximately 10 cm of distal atretic colon were found towards the left. The hemibladders flanked the intestinal structures and there was no evidence of gross cystitis cystica. Both ureteric orifices could be comfortably cannulated with 5 Fr infant feeding tubes. A 2cm phallus-like structure was noted to be ensconced within the vesical area just deep to the symphysis pubis and adherent to the caecum by a fibrotic band of tissue (Fig. 2). The left testis was seen in the inguinal canal, no attempt was made to dissect out the right testis.

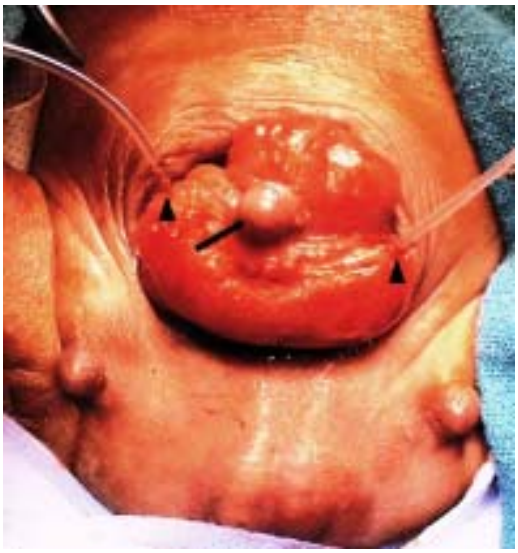

Fig. 2. Per-operative findings after prolapsed ileal segment was reduced inside the abdomen showed intravesical phallus (arrow) and bilateral ureteric openings catheterised by no 5 infant feeding tube

Our team was unprepared for this surgical surprise of an intravesical penis. As a lack of accurate anatomic information would have made single stage surgical reconstruction potentially dangerous, a decision was taken to separate the urogenital and intestinal tracts and tackle this unusual entity after further growth of the child. The dilated terminal ileocaecal region and the proximal 2cm of the distal colon were resected and an end to end ileocolic anastomosis was achieved with 5-0 PDS suture. The blind ending colon was pulled through the pelvic floor into the site of the future neoanus and the bowel hung for 2cm (Fig. 3) after resecting the distal 1cm. This hanging bowel will be trimmed at a later date after auto-anastomosis of the bowel to the pelvic floor has taken place. Thus a colostomy was avoided and the twin aims of adequate bowel decompression and diverting the faecal stream from the perineal wound were achieved. The bladder patch was reconstructed and sutured to the abdominal wall thus creating a classical bladder exstrophy (Fig. 3).

The child continues to do well in the post-operative period and is thriving.

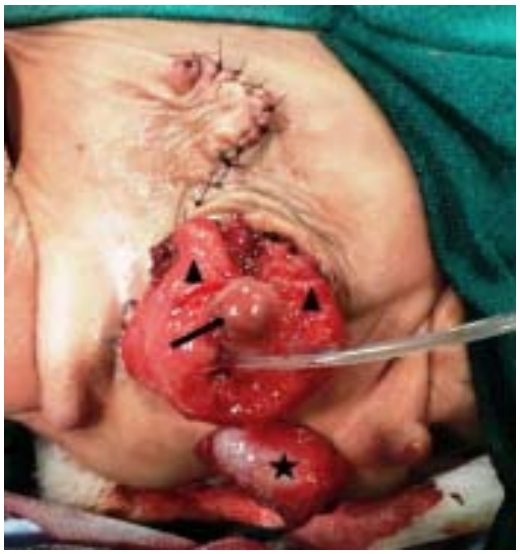

Fig. 3. Operative photograph after separation of the bowel plate from bladder plate showing intravesical phallus (arrow), reconstructed bladder patch to classical bladder exstrophy (arrow head) and pulled through hanging bowel from neoanus (asterix)

Discussion

This uncommon variant of phallic morphology in cloacal exstrophy is the ninth case of its kind.[3] Apart from this, the reported continuum of phallic abnormalities include, widely separated hemicorpora, epispadias, diphallia, bifid penis and aphallia. We are aware of eight cases in world literature and each description is unique which only adds to the fact that this is a spectrum of malformations. Arunachalam *et al* described a 46 XY child with an entrapped and buried penis.[4] Although the child developed multiple post-operative complications and died, a biopsy from the tip of the phallic structure was confirmatory.

A review of the prevailing literature was presented in the light of gender assignment by Tomaszewski *et al*.[5] All the cases described had anatomic similarities in terms of fused bladder plates. This differs from our experience as the bladder plate was cleaved completely by the bowel patch. The penile morphology in the case studies described previously consisted of partially or fully fused corpora[3-6] or a solitary corporal body[7] covered completely by urothelium. Our baby had fused corpora and the crura could be seen disappearing under the widely separated pubic bones. Though the penis

was hidden within mucosal folds, it was not covered by urothelium and had adequate skin cover.

Though antenatal diagnosis was possible in two cases,[7-8] this entity is usually discovered at the time of bladder closure. The embryology of cloacal exstrophy is the subject of much controversy and even the origin of the intra-exstrophic phallus is disputed. Lakshmanan *et al*[7] theorized that the abnormal anterior abdominal wall mechanically approximates the exstrophic bladder tissue and thus traps the phallus within it. In the present case, the band connecting the base of the phallus to the caecal patch may have had a role in halting the natural evolution of the penile tissue and the fusion of the bladder plates. The paucity of reported cases could be due to a case of mistaken identity. The urothelial covering coupled with the mucosal edema in a clinically diagnosed aphallic patient can lead the surgeon to excise it as a bladder hamartoma. Pathological examination in such cases has shown a hamartoma of squamous mucosa surrounding erectile tissue.[8]

The true incidence of cloacal exstrophy is unknown but it does account for 10% of the exstrophic defects.[9] An improvement in overall survival and continence rates in the modern era makes it imperative that we strive to give these babies a wholesome and productive life. A greater degree of knowledge about such variants could influence reconstruction and the choice of gender in children presumed to be aphallic. Genetic males need not be assigned a female sex which may go against testosterone imprinting and consign them to a life beset by psychosexual issues along with the other long-term complications of this anomaly.

References

1. Carey JC, Greenbaum B, Hall BD. The OEIS complex (omphalocele, exstrophy, imperforate anus, spinal defects). Birth Defects Orig Artic Ser 1978;14:253-63.
2. Carey JC. Exstrophy of the cloaca and the OIES complex: one and the same. Am J Med Genet 2001;99:270.
3. Fox JA, Banihani O, Schneck FX. Is it really a hamartoma? Bringing awareness to the possibility of an intravesical phallus in the aphallic 46 XY cloacal exstrophy patient. J Ped Urol 2013;9:1237-8.
4. Arunachalam P, Pillai SB, Sridhar DC. Classical cloacal exstrophy with intravesical phallus. J Pediatr Surg 2012;47:5-8.
5. Tomaszewski JJ, Smaldone MC, Cannon GM, Schneck FX, Hackam DJ, Docimo SG. Cloacal exstrophy variant with intravesical phallus: further implications for anatomy and gender reassignment. J Pediatr Urol 2012;8:426-30.
6. Shaw MB, West K, Gitlin J, Casale AJ, Rink RC. Intravesical phallus in cloacal exstrophy. Urology 2003;61:644.
7. Lakshmanan Y, Bellin PB, Gilroy AM, Fung LC. Antenatally diagnosed cloacal exstrophy variant with intravesical phallus in a twin pregnancy. Urology 2001;57:1178.
8. Carr MC, Benacerraf BR, Mandell J. Prenatal diagnosis of an XY fetus with aphallia and cloacal exstrophy variant. J Ultrasound Med 1994;13:323.
9. Ziegler MM, Duckett JW, Howell CG. Cloacal Exstrophy. In: Welch KJ, Randolph JG, Ravitch MM, O'Neil JA, Rowe MI (eds) Paediatric Surgery. Year Book Medical Publishers, Chicago, 1986, pp 764-771.

Case Report

Laparoscopic Nephroureterectomy in Wilms' tumor

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Abstract. Open surgery has remained the standard surgical technique for Wilms' tumor. But laparoscopic nephrectomy in Wilms' tumor is a feasible and a safe option in selected subset of patients. It reproduces all steps similar to an open surgical approach with the advantage of less postoperative pain, shorter hospital stay, a more acceptable cosmetic scar and early initiation of adjuvant chemotherapy. Here we describe a 13-month-old patient of Wilms' tumor successfully treated using a minimally invasive approach.

Keywords: Indications of laparoscopy, Laparoscopic nephroureterectomy, Wilms' tumor

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Introduction

Wilms' tumor is the most common malignant renal tumor of childhood affecting approximately one child per 10,000 younger than fifteen years. Surgical staging and tumor resection remain the central components of tumor therapy. Treatment protocols recommend nephroureterectomy with neoadjuvant/adjuvant chemotherapy. The surgeon must remove the tumor, determine the intra abdominal stage by lymph node sampling and carefully examine the liver and the contralateral kidney. The surgical guidelines for the management of Wilms' tumor have been clearly outlined. A large trans-abdominal, trans-peritoneal incision is recommended for adequate exposure. Recently, the advancement in laparoscopic techniques and increased laparoscopic nephroureterectomies for benign renal lesions has encouraged surgeons to apply this potentially beneficial approach to treat malignant renal lesion such as Wilms' tumor. But standard selection criteria for patients undergoing laparoscopic nephroureterectomy in Wilms' tumor is lacking and the question of safety of laparoscopic nephroureterectomy in children with Wilms' tumor still lingers in the mind of most paediatric surgeons. Herein is described a case of Wilms' tumor successfully treated using a minimally invasive approach. Also recommendations for selection criteria for laparoscopic nephroureterectomy are discussed.

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Case Report

A 13-month-old boy presented to us with fever and anorexia for 2 months and lump in the right flank for the last 20 days. The child had no complaints of hematuria, pyuria, graveluria, bowel irregularities, lethargy, loss of appetite and weight loss. Clinically the child had pallor and a firm, ballotable lump, measuring 5 x 4cm palpable in the right lumbar and hypochondrium region.

The contrast enhanced computed tomogram (CECT) of the chest and abdomen revealed a large heterogenous enhancing mass lesion 7.1 x 7.1 x 6.6cm arising from the anterior surface of the right kidney with multiple necrotic areas with no calcification. Renal vein, inferior vena cava and the liver were normal. There was no lung metastasis. Fine needle aspiration cytology (FNAC) was suggestive of biphasic Wilms' tumor with predominant blastemal components. A diagnosis of stage two right Wilms' tumor was made and the child received six weeks of DD4A neoadjuvant chemotherapy consisting of vincristine, actinomycin-D and doxorubicin. Post chemotherapy CECT (Fig. 1) showed a partial response and an exophytic tumor of 4 x 4 x 3cm localized to the mid-polar region of the right kidney.

Thereafter, a three port laparoscopic nephrectomy with lymph node sampling was planned for this patient. After induction of general anaesthesia, an orogastric tube and a urinary catheter were inserted. The patient was placed supine and was secured to the table with straps. The operating surgeon, first assistant and the scrub nurse were positioned on the left

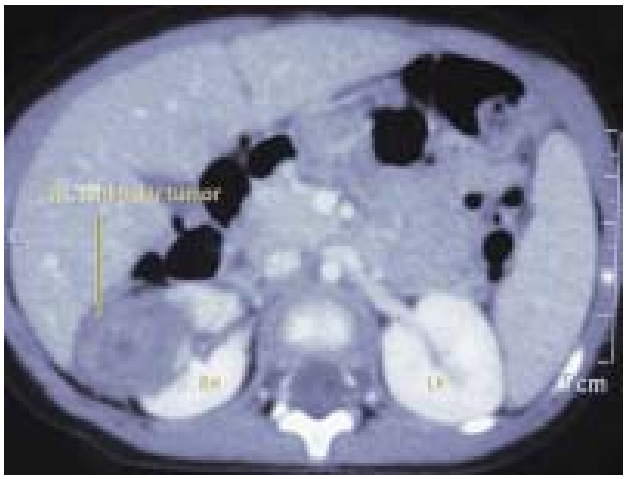

Fig. 1. CECT abdomen after six cycles of neoadjuvant chemotherapy showing the right mid-polar renal tumor

of the patient facing the primary video monitor and the second assistant on the right. The first 12mm port was placed in the umbilicus for the camera. Then two 5mm ports were placed under vision in the epigastrium and the right hypochondrium in the mid clavicular line (Fig. 2). The head end was raised to about 45 degrees and right side was raised. The dissection was started along the line of Toldt, releasing the ascending colon, the hepatic flexure and the transverse colon. The mobilized colon was reflected medially and the retroperitoneal fat was dissected to identify the gonadal vein and the right ureter. The ureter was traced proximally to reach the renal hilum (Fig. 3a). The lower pole of the right kidney was mobilized using harmonic scalpel. Thereafter the upper pole was mobilized by releasing it from undersurface of the liver and the diaphragm, after retracting the liver. The hilar dissection was carried out meticulously. The renal artery and the renal vein were clipped and cut (Fig. 3b). The posterior adhesions of the kidney with the psoas muscle were released using sharp and blunt dissection. After complete mobilization of right kidney (Fig. 4a), ureter was traced caudally, clipped and cut just proximal to the bladder. Thereafter the supra-hilar, infra-hilar and the para-caval lymph nodes were sampled using electrocautery. Titanium clips were applied to mark the limits of the tumor extension. The specimen was delivered from the right inguinal skin crease incision (Fig. 4b) and the specimens were sent for histo-pathological examination. The child was extubated and the total duration of the surgery was one hour and thirty minutes.

He was started on enteral feeds on day two and discharged on postoperative day three. The duration of hospital stay was six days. The child received week seven chemotherapy on postoperative day seven. The histo-pathological report showed a 4 x 3 x 3cm homogenous tumor in the mid-portion of

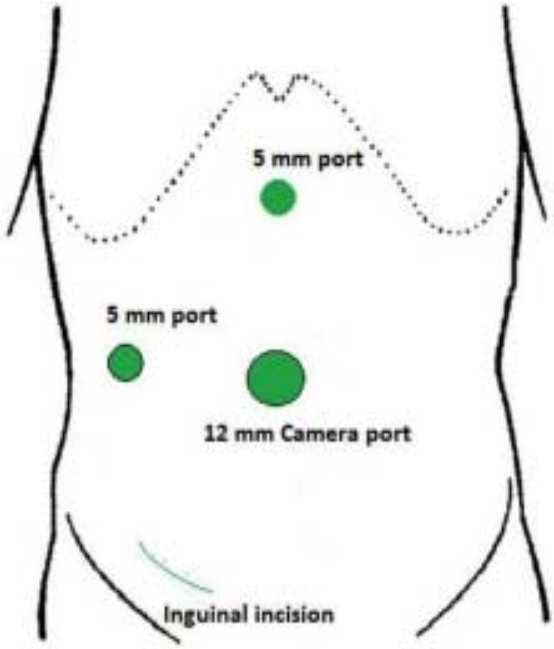

Fig. 2. Port site placement for three port laparoscopic nephroureterectomy for right sided Wilms' tumor

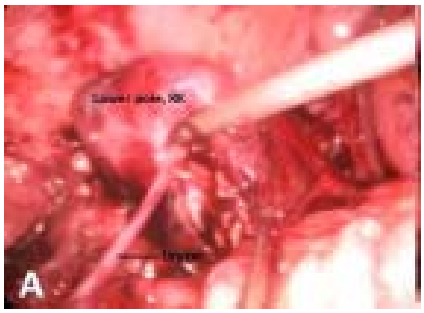

Fig. 3a

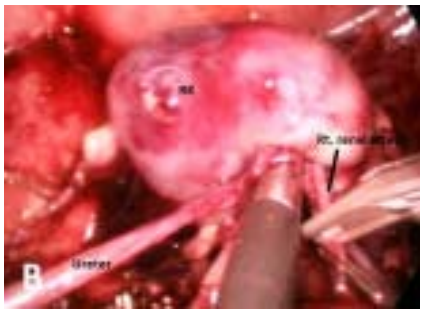

Fig. 3b

Fig. 3. Fig. 3a. Ureter identified, dissected and traced upwards to reach the renal hilum and the lower pole of kidney mobilized Fig. 3b. Right renal vessels clipped using ligaclips

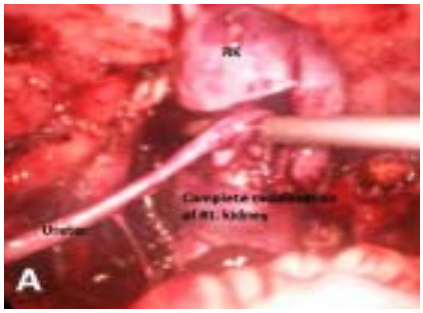

Fig. 4a

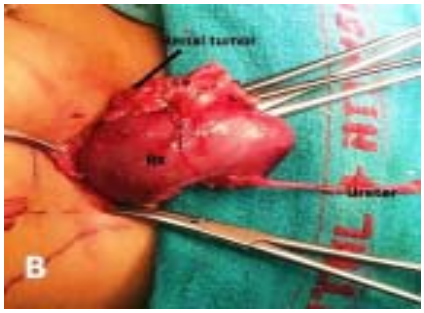

Fig. 4b

Fig. 4. Fig. 4a. Complete laparoscopic mobilization of the right kidney after complete mobilization of the upper pole Fig. 4b. Delivery of the resected specimen from right inguinal skin crease incision

the right kidney with tri-phasic histo-morphologic features, large areas of necrosis with fibrosis and approximately five percent of viable tumor. The renal sinus, resected end of ureter and the hilar vessels were free of tumor. The tumor extended beyond the renal capsule but the resected margins were free of tumor. There was no lymph node metastasis. The child is receiving adjuvant chemotherapy and is under regular follow up for the last three months.

Discussion

Open surgery has remained the standard surgical technique for Wilms' tumor with excellent survival for localized disease with favourable histology. Surgical procedure includes a liberal trans-peritoneal incision, complete inspection of the abdominal cavity, lymph node sampling, tumor resection and retrieval with no spill. Neoadjuvant chemotherapy can reduce the tumor size and can minimize the incidence of spill or rupture making resection easier and minimizing the recurrence rates. However, open conventional surgery carries the disadvantage of unsightly scars, greater postoperative pain and ileus, longer hospital stay and recovery period. Potential benefits of laparoscopic techniques include a more cosmetically acceptable scar, less postoperative discomfort and ileus, less pain, decreased bowel adhesions and shorter length of hospital stay.

There has been a legitimate interest regarding the use of laparoscopic techniques for nephroureterectomy in Wilms' tumor. Increasing number of case reports and case series are available depicting the safety and feasibility of laparoscopy in Wilms' tumor. Initial reports described by Duarte *et al* in 2004 and 2006 have been encouraging.[1-2] They successfully treated two cases of unilateral non-metastatic Wilms' tumor in 2004 who received preoperative chemotherapy and then underwent laparoscopic nephrectomy. Later in 2006, they published a case series in which eight children with unilateral Wilms' tumor underwent laparoscopic nephrectomy after chemotherapy. They found it a safe procedure which provided a complete surgical approach required to treat this tumor. Similar results were published by Varlet F *et al* in 2009 who performed laparoscopic radical nephrectomy in five children with unilateral renal malignant tumor.[3] Four cases were of suspected Wilms' tumor who were treated with chemotherapy according to the International Society of Paediatric Oncology protocol and the fifth case was a juvenile renal cell carcinoma. Another case report by Patric J Javid in 2011 reconfirmed the feasibility of laparoscopic techniques for resection of Wilms' tumor provided oncological principles are carefully followed.[4] They performed laparoscopic resection of tumor with right radical nephroureterectomy, retroperitoneal lymph node dissection and resection of peritoneal metastases in a 2-year-old girl with a large right renal mass with peritoneal seeding and pulmonary metastasis.

In our case too, the oncologic principles followed were the same as that of open surgery. After neoadjuvant chemotherapy, the tumor shrunk in size and became amenable to resection. This case represents the subset of patients with Wilms' tumor who can be successfully treated with laparoscopy, especially when performed by beginners. We were able to reproduce all steps of open surgical approach including complete excision and lymph node dissection. The proper visualization and magnification provided by laparoscopy aided in tumor mobilization with minimal tissue trauma. Easy release of tumor adhesions and minimal bleeding was ensured during surgery with the use of new energy sources like the harmonic scalpel and ligasure. Finally the tumor was removed intact through an inguinal skin crease incision which was still smaller and less visible incision than the conventional laparotomy incision. We avoided morcellation and removal, as it would prevent proper histological staging and may have caused tumor spill. Post operatively, the child had decreased analgesic need, started early enteral feeding and was discharged on the third day. He received adjuvant chemotherapy as early as one week postoperatively.

While laparoscopy has been successful in the treatment of this case, the limitations pertaining to patient selection and

laparoscopic technique per se do prevail. Most cases selected were small, unilateral tumors, not crossing the midline, with absence of thrombus in the renal vein or inferior vena cava and had received neo-adjuvant chemotherapy, hence making the tumor less friable and decreasing the chances of tumor rupture. Patients with very large tumor with impending rupture or those with previous surgeries and extensive postoperative adhesions are not the ideal candidates for laparoscopy.[5] The absence of tactile feedback during surgery and the fear of tumor spillage during morcellation, inadequate retroperitoneal lymph node dissection, postoperative complications like atelectasis, port site recurrence, trocar site herniation and the effect of insufflation on biology of tumor cells have limited the use of laparoscopy till date.

Conclusion

Laparoscopic nephrectomy in Wilms' tumor is a feasible and a safe option in selected subset of patients with Wilms' tumor provided the principles of oncology are strictly followed. It reproduces all steps similar to an open surgical approach with the advantage of less postoperative pain, shorter hospital stay, a more acceptable cosmetic scar and early initiation of adjuvant chemotherapy. Proper patient selection, port placement and laparoscopic surgical experience contribute to the above. Careful controlled randomized studies are needed to help determine the benefits and drawbacks of this new evolving technique.

References

1. Duarte RJ, Dénes FT, Cristofani LM, Giron AM, Filho VO, Arap S. Laparoscopic nephrectomy for Wilms' tumor after chemotherapy: initial experience. J Urol 2004; 172:1438-40.
2. Duarte RJ, Dénes FT, Cristofani LM, Odone-Filho V, Srougi M. Further experience with laparoscopic nephrectomy for Wilms' tumour after chemotherapy. BJU Int 2006;98:155-9.
3. Varlet F, Stephan JL, Guye E, Allary R, Berger C, Lopez M. Laproscopic radical nephrectomy for unilateral renal cancer in children. Surg Laparosc Endosc Percutan Tech 2009;19:148-52.
4. Javid PJ, Lendvay TS, Acierno S, Gow KW. Laproscopic nephroureterectomy for Wilm's Tumor: Oncologic considerations. J Pediatr Surg 2011;46:978-82.
5. Bhatnagar S, Sarin YK. Scope and limitations of minimal invasive surgery in practice of paediatric surgical oncology; Indian J Med Pediatr Oncol 2010;31:137-42.

Case Report

Correction of Isolated Complete Penoscrotal Transposition: Point of Technique

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Abstract. Complete penoscrotal transposition also known as pre penile scrotum is a rare anomaly in which scrotum is transposed to a cranial position in relation to the penis. There are few cases reported in literature, and often it is associated with severe hypospadias. We report a novel operative technique which allows single stage correction of complete penoscrotal transposition.

Keywords: Complete penoscrotal transposition, Hypospadias, Penis, Pre-penile scrotum

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Introduction

Penoscrotal transposition (PST) is a rare anomaly characterized by malposition of the penis in relation to the scrotum. In complete PST, also referred to as pre penile scrotum, the scrotal folds are located cranially and fused in midline, while the penis emerges from perineum. Incomplete PST is a more common condition where the penis lies in the middle of the bifid scrotum. Both forms are most often associated with severe forms of hypospadias.[1] There are very few reports of complete PST with no chordee or hypospadias.

Several techniques have been described to correct PST of which Glen Anderson technique is popular.[1-5] We herewith report a rare combination of complete PST, glanular hypospadias and no chordee. We also describe a novel operative technique which allowed single stage correction with minimal lymphoedema and good cosmetic outcome.

Case Report and Technique

A term male neonate presented with complete PST and glanular hypospadias. There was no skin chordee. Both testes were normally descended and, the patient was able to pass urine well via the glanular meatus. There were no major anomalies involving other systems and karyotyping was normal 46XY. Hormonal evaluation did not reveal any abnormality. Since there was evidence of micropenis, monthly testosterone injections were given (three doses between 10-12 months).

At the age of 15 months there was good improvement in the shaft size and length (Fig. 1a). Since the hypospadias was glanular, parents elected to leave it uncorrected. Under general anaesthesia and caudal analgesia, surgical correction was performed. Initial catheterization with 6F feeding tube failed as the catheter could not be advanced beyond the posterior urethra. Cystoscopy revealed a prominent prostatic utricle and a supra pubic catheter was inserted under cystoscopic vision.

Fig. 1b shows the markings of the incision. Circumferential incision was made at the base of the penis. This was extended vertically at 12 O' clock in the midline between the fused scrotal folds. At a point where the penis has to be transposed the vertical incision was stopped and a horizontal incision was made (T shape) with either side of the horizontal limb going laterally to outline scrotal folds. Fig. 1c and Fig. 1d reveal deepening of 'T' incision and the circumferential incision around the base of penis, to release the abnormal soft tissue bands holding the scrotal folds in the cranial position. Once fully mobilsed, the scrotal folds are free to be moved caudally and penis free to move cranially to its natural position (Fig.1e). Soft tissue and skin approximation were obtained with 5-0 polyglactin sutures as shown in Fig. 1f. Soft compression dressing was kept for 8 days. The patient passed urine well on removal of dressing and there was no lymphoedema. Follow up at 3, 6 and 12 months revealed good cosmetic outcome. There were no late complications and parents elected to defer decision on surgery for glanular hypospadias till puberty.

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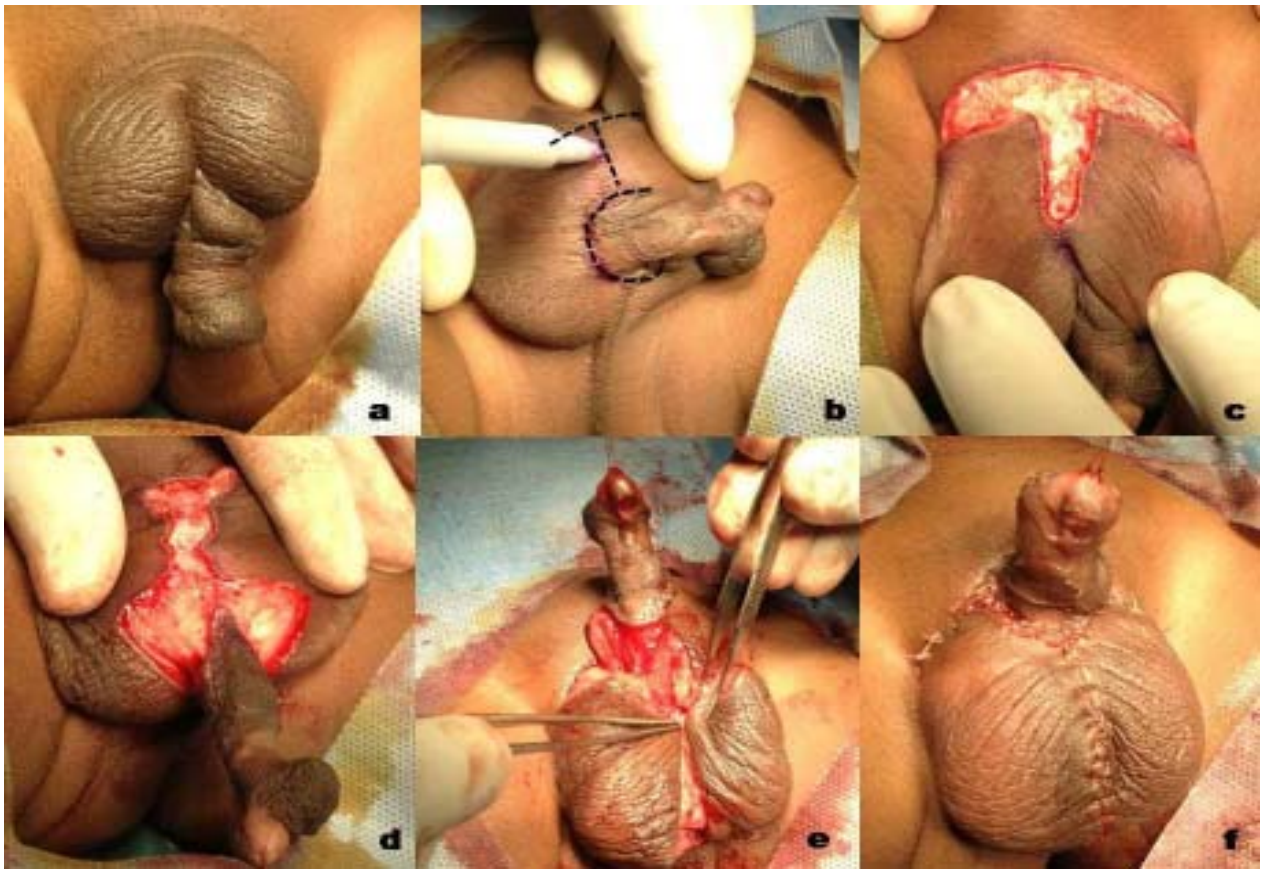

Fig. 1. **Fig. 1a. Preoperative appearance showing both scrotal folds fused in the midline, cranial to penis. Fig. 1b. Dotted lines show markings of the incision. Circumferential incision is made at the base of the penis. This is extended vertically at 12 O'clock in the midline between the fused scrotal folds. At a point where the penis has to be transposed the vertical incision was stopped and a horizontal incision was made (T shape) with either side of the horizontal limb going laterally to outline scrotal folds. Fig. 1c. After the incisions, deepening of 'T' incision and Fig. 1d. deepening of the circumferential incision around the base of penis, to release the abnormal soft tissue bands. Fig. 1e. Once fully mobilsed, the scrotal folds are free to be moved caudally and penis free to move cranially to its natural position. Fig. 1f. Final appearance at the end of the operation is seen.**

Discussion

The embryological sequence responsible for complete PST is unclear. During normal development in a male embryo, the labio-scrotal swellings, under the influence of di-hydro testosterone, migrate inferomedially, and fuse in the midline caudal to the penis, between 9th and 12th week of gestation. Abnormal positioning of the genital tubercule in relation to the scrotal swellings or incomplete/failed migration of labio-scrotal swellings have been suggested to cause complete PST.[1-3]

PST was first reported by Appleby in 1923. Patients with PST often have accompanying urological abnormalities such as chordee, hypospadias, vesicoureteric reflux, urethral atresia, and bifid scrotum. Mcllvoy and Harris first performed surgery to move the penis into a more cranial position through a subcutaneous tunnel beneath the prepenile scrotum.[2] Forshall and Rickham used a different technique in two patients in whom the cranially located scrotal flaps were elevated, rotated medially and caudally, and sutured beneath the penis.[3] Kolligian *et al* transferred the penis after straightening into a button hole designed in the skin of mons pubis. The authors feel this technique does not allow release of abnormal soft tissue bands well enough to achieve a good cosmetic result.[4]

Complications after surgery for PST include urethral and testicular injury, urinary fistula, flap necrosis, and penile edema. Circular incision at the root of the penis may partially compromise lymphatic drainage and cause lymphoedema. Observation of patients corrected by Glenn-Anderson technique showed gross edema that persists for long periods (6-9 months), and after resolution leaving the penile skin dusky

and darkly pigmented with appearance like the scrotal skin. Saleh felt that preservation of strip of skin at 12 O' clock position during correction of PST reduced postoperative edema and lowered complications,[5] however, we did not advocate this as we felt it results in excess skin causing dog ear. Despite this we did not encounter lymphoedema or late penile skin abnormalities reported earlier, as the penile blood supply and lymphatic drainage took care of the un-degloved skin attached over it.

The authors feel that whenever complete PST is associated with hypospadias, it should be staged to avoid complications associated with blood supply and lymphatic drainage. The technique we have described, is for isolated PST where there is no chordee requiring degloving. The described technique is an addition to existing techniques for correcting of complete PST.

References

1. Redman JF, Bissada NK. Complete Penoscrotal transposition. Urology 2007;69:181-2.
2. McIlvoy DB, Harris HS. Transposition of the penis and scrotum: Case report. J Urol 1955;73:540-3.
3. Forshall I, Rickham PP. Transposition of the penis and scrotum. Br J Urol 1956;38:250-2.
4. Kolligian ME, Franco I, Reda EF. Correction of Penoscrotal transposition: a novel approach. J Urol 2000;164:994-6.
5. Saleh A. Correction of incomplete Penoscrotal transposition by a modified Glenn-Anderson technique. Afr J Paediatr Surg 2010;7:181-4.

Case Report

Posterior Urethral Valves with Congenital Urethral Stricture

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Abstract. Management of stricture urethra remains a challenge to the treating surgeon irrespective of the etiology, age and site. Association of co-morbid conditions makes the task of surgeon even more difficult. We are sharing our experience in managing a case of posterior urethral valve with congenital stricture of posterior urethra.

Keywords: Posterior urethral valve, Preanal anterior coronal approach, Urethral stricture

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Introduction

Posterior Urethral Valve (PUV) is the commonest cause of lower urinary tract obstruction in male infants.[1-3] In the present era the most common mode of presentation is antenatal diagnosis. Postnatally, PUV can have a broad spectrum of presentations ranging from a life threatening pulmonary hypoplasia due to oligohydramnios, to mild obstruction or symptoms that may escape early detection and manifest only in later childhood, adolescence or even adulthood.[4-6] Universally accepted treatment protocol for managing PUV includes cystoscopic valve ablation using cold knife or electrocautery or LASER. PUV associated with congenital stricture urethra is very rare. We are sharing our experience in managing a case of posterior urethral valve with congenital stricture of posterior urethra.

Case Report

Thirteen months old boy, a case of antenatally diagnosed posterior urethral valve underwent suprapubic cystostomy (SPC) at day 2 of life in some other institute and came to our department at the age of 1 year. The child was investigated with micturating cystourethrogram (MCU) [Fig. 1] which showed dilated posterior urethra, irregular bladder, abrupt cut-off between posterior and anterior urethra and thin stream in anterior urethra. Urethrocystoscopy (7.5F) was done but scope was not negotiable 5cm beyond external urethral meatus due to tight stricture. The decision was taken to operate after obtaining consent for the same. We used the pre-anal anterior coronal approach described previously by us for managing posterior urethral strictures.[7] Lay open of the posterior urethral stricture (nearly 1.5cm) was done (Fig. 2) guided by urethral dilators both from external urethral meatus and SPC site. Posterior urethral valve was incised at 5, 7 and 12 O'clock positions through urethrostomy route. Stricture part was laid open and perineal urethrostomy was made. The postoperative period was uneventful and the child recovered well. Before discharge SPC was removed and strapped to close. During

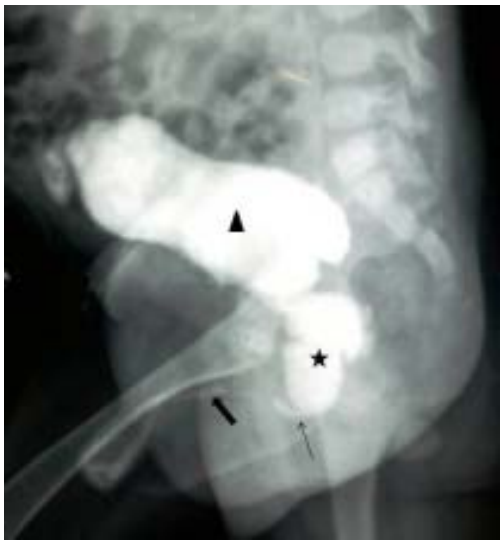

Fig. 1. Micturating cystourethrogram (MCU) showing dilated posterior urethra (asterix), irregular bladder (arrow head), abrupt cut-off between posterior and anterior urethra (thin arrow) and thin stream in anterior urethra (thick arrow)

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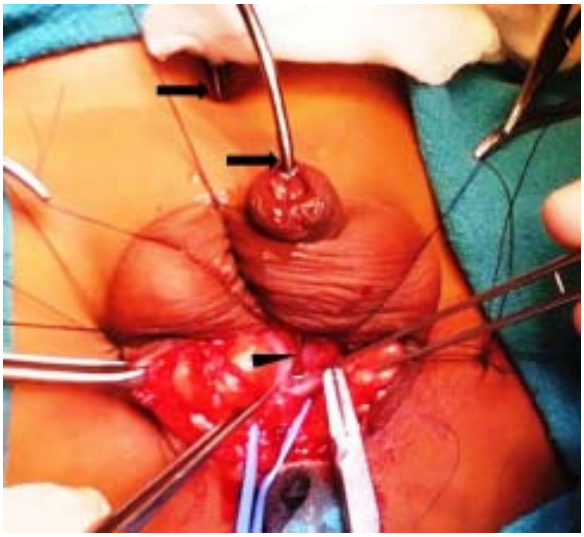

Fig. 2. Intraoperative photograph showing lay open of the posterior urethral stricture nearly 1.5 cm (arrow head) guided by urethral dilators (thick arrow) both from external urethral meatus and SPC site

follow up patient was advised to calibrate anterior urethra regularly. At last follow up, 6 months back, child was doing well without any urinary complaints and is waiting for closure of perineal urethrostomy.

Discussion

The most common cause of bladder outlet obstruction in boys is posterior urethral valve. PUV associated with congenital posterior urethral stricture is very rare. Various treatment options are available for managing cases with PUV, most widely accepted being the valve ablation.[8-10] Though posterior urethral stricture was described in PUV patients after valve ablation,[11-15] congenital posterior urethral stricture in PUV patients was not described in English literature to the best of our knowledge.

Posterior urethral stricture is considered the most debilitating because, if not managed properly, may lead to severe impairment of the quality of life, by affecting continence and potency. There are several treatment options for managing urethral strictures in children. In general they include, urethral dilatation, endoscopic visual internal urethrotomy, open urethral reconstruction (single stage or staged). We have used pre anal anterior coronal approach in our case to localize the stricture segment. Pre anal anterior coronal approach is an established terminology given by the senior author initially for managing cases of aphallia and then further extended to include posterior urethral strictures as well.[7,16] It is different from perineal approach in terms of incision, better exposure, minimal blood loss and ability to tackle and identify the stricture. In our case, as the stricture was congenital, long segment and associated with PUV, so authors decided to repair it in a staged manner by laying open it in first stage.

Conclusion

We would like to convey the message that the possibility of congenital posterior urethral stricture should be kept in mind while dealing with a case of PUV, where scope was not negotiable beyond anterior urethra into the dilated posterior urethra. Staged treatment is a possible and good option of repair of this type of stricture.

References

1. Nasir AA, Ameh EA, Abdur-Rahman LO, Adeniran JO, Abraham MK. Posterior urethral valve. World J Paediatr 2011;7:206-16.
2. Imaji R, Dewan PA. The clinical and radiological findings in boys with endoscopically severe congenital posterior urethral obstruction. BJU Int 2001;88:263-7.
3. Atwell JD. Posterior urethral valves in the British Isles: a multicenter BAPS review. J Paediatr Surg 1983;18:70-4.
4. Bomalaski MD, Anema JG, Coplen DE, Koo HP, Rozanski T, Bloom DA. Delayed presentation of posterior urethral valves: a not so benign condition. J Urol 1999;162:2130-2.
5. Hendren WH. Posterior urethral valves in boys. A broad clinical spectrum. J Urol 1971;106:298-07.
6. Schober JM, Dulabon LM, Woodhouse CR. Outcome of valve ablation in late-presenting posterior urethral valves. BJU Int 2004;94:616-9.
7. Singh A, Panda SS, Bajpai M, Jana M, Baidya DK. Our experience, technique and long-term outcomes in the management of posterior urethral strictures. J Pediatr Urol. 2013 Jun 19. pii: S1477-5131(13)00151-4. doi: 10.1016/j.jpurol.2013.05.018.
8. Farhat W, McLorie G, Capolicchio G, Khoury A, Bägli D, Merguerian PA. Outcome of primary valve ablation versus urinary tract diversion in patients with posteriorurethral valves. Urology 2000;56:653-7.
9. Walker RD, Pardon M. The management of posterior urethral valves by initial vesicostomy and delayed valve ablation. J Urol 1990;144:1212-4.
10. Yohannes P, Hanna M. Current trends in the management of posterior urethral valves in the paediatric population. Urology 2002;60:947-53.
11. Myers DA, Walker RD. Prevention of urethral strictures in the management of posterior urethral valves. J Urol 1981;126:655-7.

12. Lal R, Bhatnagar V, Mitra DK. Urethral strictures after fulguration of posterior urethral valves. J Pediatr Surg 1998;33:518-9.
13. Nijman RJM, Scholtmeijer RJ. Complications of transurethral electroincision of posterior urethral valves. Br J Urol 1991;67:324-6.
14. Crooks KK. Urethral strictures following transurethral resection of posterior urethral valves. J Urol 1982;127:1153-4.
15. Bruce J, Stannard V, Small PG, Mayell MJ, Kapila L. The operative management of posterior urethral valves. J Pediatr Surg 1987;22:1081-6.
16. Bajpai M. Scrotal phalloplasty: A novel surgical technique for aphallia during infancy and childhood by pre-anal anteriorcoronal approach. J Indian Assoc Pediatr Surg 2012;17:162-4.

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Introduction to Volume 1

The past few years have seen rapid changes in the understanding of congenital malformations and acquired diseases. Paediatric Urology has not remained untouched by these developments. Presently, there are numerous journals which address these problems with authors presenting their points of view. A personal bias in the results of the study, albeit inadvertently, becomes unavoidable in such publications. Also, at one point of time several articles exist on the same subject, with as many points of view.

On the other hand, some textbooks attempt to review the subject. But, unfortunately, sometimes several years may pass before a new edition of the textbook is printed. Thus, the reader is left with exhaustive reviews of the journals needed to keep pace with the developments. The shortcomings of such alternatives cannot be overemphasized.

The present attempt is to provide to the reader with a balanced and comprehensive review of the problem, with clear instructions and key information, on an annual basis. The chapters will be followed by additional commentaries where necessary.

New Delhi, India
1 February 1998

EDITORS

Editorial

Disorders of Sex Development: The Quintessence of Perennial Controversies-II (Age, Timing & Type of Surgery)

The question-**Who is the right person to take decision regarding choice & time of surgery- Parents or Patients?-** continues to stare,[1] the birth prevalence of genital anomalies remains as high as one in 300 births[2] However, the extent of genital ambiguity may depend on the expertise of the observer, and prior to presentation to a clinical expert, the label of ambiguous genitalia is often assigned to newborns where the most appropriate sex of rearing is not immediately clear to those present at the child's birth.

What defines DSD and how it is managed and treated is a political and controversial area with individuals, families, clinical practitioners, mental health professionals, ethicists, lawyers, advocates and activists often having differing views.

The sex of a newborn baby is usually the first question asked by & from the parents. There is enormous pressure for people to conform to cultural and social expectations.

In the past, most medical experts believed the birth of an infant born with an DSD condition is a medical and social emergency usually requiring surgical intervention and that living with ambiguous genitalia is psychologically traumatic for the child. Early normalization was, therefore, sought so that children could be given a successful gender identity and bonding with parents would become easy.

More recently, the medical sense of urgency to correct the situation has decreased. Babies born with DSD conditions are regarded as having a significant medical condition, but more emphasis is placed on getting an accurate diagnosis of the cause of the ambiguous genitalia and making a careful decision about assigning gender before making any decisions about surgery.

Age & Timing for Surgery

Three age periods can be distinguished in the problem of sex assignment:

1. The newborn period, extended to early infancy, when parents decide
2. From 2 to 10 years of age, when sex re-assignment is generally not recommended
3. From 11 years of age up to late adolescence or young adulthood, when patients decide. When sex assignment becomes necessary, particularly in newborns, the aim is to choose that sex that will allow the best future functional adaptation that better goes along with the biological sex. Decisions might be difficult and controversial because in some cases there is no "good" solution.

In general recommendations in infants are based on:

1. Etiologic diagnosis (if available, molecular diagnosis), diagnosis of type of DSD is useful because disorder evolution partially depends on etiology, known from both personal experiences and scientific publications
2. Development of external genitalia and potential future sex function
3. Possibility of surgical correction

4. Development of internal genitalia, and fertility potential
5. Parental acceptance
6. Psychological evaluation of parents and family, including social environment

Understanding the Social and Cultural Background of the Family is Crucial for A Fruitful Relationship Between Parents and Doctors

However, it is sometimes difficult to synchronize delivery of information to parents by different members of the team. Special efforts should be made by the informing professional to ensure appropriate understanding by the family of the situation, including chromosomal constitution, gene function, heredity, gonadal development, as well as external and internal genitalia differentiation. During follow up patients will go through all stages of growth and development and information has to be delivered according to patient mental developing and understanding. The medical team should constantly update itself with new developments that might arise along many years, and to give answers to questions, which are frequently, influenced by a high emotional impact.

Several pioneering research teams have studied the wide variety of sexual development disorders with a view to changing the way they are treated.

One example is Congenital Adrenal Hyperplasia (CAH). In this condition, due to enzymatic deficiency, there is a surfeit of the precursors of testosterone. As a result there are distinct changes in the female genitalia. Most notable is the enlargement of clitoris which resembles the penis. The most frequently done surgery is to reduce the size of the clitoris. However, some researchers found, that, these patients may have reduced sensuality during sex.[3,4,5] It led to a growing trend towards a more conservative approach with increasing emphasis being placed on the preservation of sexual function later in life. 'Endocrine Society' guidelines of 2010 state "Surgical guidelines emphasize early single-stage genital repair for severely virilized girls, performed by experienced surgeons.[6]

The international Consensus statement on management of DSD cites gender dissatisfaction occurring more frequently with individuals with an DSD condition. However Crawford *et al*'s long term outcome study, found that early intervention yielded acceptable cosmetic results while producing minimal impairment in quality of life.[7]

Type of Surgery

Among the most contentious issue in regard to those with CAH involves XX females. Females with CAH are generally born with an enlarged clitoris and often a vagina that is atypically formed. Questions arise as to whether they should have genital surgery or not-it is often recommended. Sometimes, dependent upon the degree of masculinization of the genitals, there is question of whether they should be reared as males or females.

Not infrequently, for various reasons such as the disease's late onset, some will be reared as males and some will request such assignment. Recommendations as to the management of severely masculinized infants with CAH are controversial. When it was first suggested that severely masculinized females be raised as males the idea was generally rejected. Lately, however, the idea is being revived.[8,9]

Regarding the management of infants with DSD conditions and ambiguous genitalia in a survey[10,11] the Paediatric Urologists overwhelmingly favored female gender assignment for females with CAH even if they were extensively masculinized. They recommended feminizing surgery-reducing the size of an enlarged clitoris-and considered that preservation of female fertility was of foremost importance and the masculinization of behaviors or inclinations, was of lesser importance. There was a great difference of opinion as to the age it would be best to do the surgery.

(***To be continued in part III)***

New Delhi
May 1, 2013

Prof. M. Bajpai
Editor

References

1. Bajpai M. Editorial: Disorders of Sex Development: The quintessence of perennial controversies-I (Who is the right person to take decision regarding choice & time of surgery- Parents or Patients?). JIAPS Jan-Mar 2014;Vol 19 (1):3-4.
2. Ahmed SF, Dobbie R, Finlayson AR, *et al*. Regional & temporal variation in the occurrence of genital anomalies amongst singleton births, 1988-1997, Scotland. Archives of Disease in Childhood. 2004;89:F149-F151.
3. Creighton SM, Farhat WA. Early versus late intervention of congenital adrenal hyperplasia. J Pediatr Adolesc Gynecol. Feb 2005;18(1):63-9.
4. Creighton S. Early versus late intervention of congenital adrenal hyperplasia. J Pediatr Adolesc Gynecol. 2004 Dec;17(6):411. Corrected and republished in: J Pediatr Adolesc Gynecol. Feb 2005;18(1):63-9.
5. Crouch NS, Minto CL, Laio LM, Woodhouse CR, Creighton SM. Genital sensation after feminizing genitoplasty for congenital adrenal hyperplasia: a pilot study. BJU Int. Jan 2004;93(1):135-8.
6. Speiser PW, R. Azziz, L. Baskin, L. Ghizzoni, Hensle TW, D. Merke, H. Meyer-Bahlburg, W. Miller, V. Montori, S. Oberfield, M. Ritzen, and W.P. C., Congenital adrenal hyperplasia due to steroid 21-hydroxylase deficiency: an Endocrine Society clinical practice guideline. Journal of Clinical Endocrinology & Metabolism, 95 (2010)4133-4160.
7. Crawford, Warne, Grover, Southwell, Hutson; Results from a paediatric surgical centre justify early intervention in disorders of sex development, Journal of paediatric surgery, 2009;44:413-16.
8. C.P. Houk, and P.A. Lee, Approach to assigning gender in 46, XX congenital adrenal hyperplasia with male external genitalia: replacing dogmatism with pragmatism. Journal of Clinical Endocrinology & Metabolism 95 (2010);66[66]: 4501-8.
9. J.C. Jorge, C. Echeverri, Y. Medina, and P. Acevedo, Male Gender Identity in an XX Individual with Congenital Adrenal Hyperplasia. Journal of Sexual Medicine 5 (2008); 122-131.
10. M. Diamond, Pediatric management of ambiguous and traumatized genitalia. The Journal of Urology 162(1999); 1021-1028.
11. Diamond DA, Burns JP, Mitchell C, Lamb K, Kartashov AI, Retik AB. Sex assignment for newborns with ambiguous genitalia and exposure to fetal testosterone: attitudes and practices of paediatric urologists. J Pediatr. Apr 2006;148(4):445-9.

Professor Stuart B. Bauer

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Born and raised in Brooklyn, NY, Dr. Bauer earned a bachelor's degree (Magna Cum Laude) in Chemistry at Brooklyn College in 1964 and a medical degree from the University of Rochester School of Medicine & Dentistry in 1968. After interning at King County Hospital in Seattle (1969) and serving 2 years as a captain in the US Army during the Vietnam War, he completed his postgraduate training in Surgery and Urology at Tufts-New England Medical Center in Boston in 1975. During his Urology residency he had an elective at the Middlesex Hospital in London with Mr. Richard Turner Warwick (1974). After completing his Urology residency he became a member of the Pediatric Urology service at Tufts-NEMCH for 2 years and then joined the Division of Urology at Children's Hospital in 1977. He advanced up the academic ladder being promoted to Professor of Surgery (Urology) at Harvard Medical School in 2000.

Within 6 months of his appointment at Children's Hospital Boston, the first urodynamics laboratory in the world entirely dedicated to the diagnosis and treatment of neuro-urological disorders in children was organized under his direction and guidance. This facility is unique in its scope for it includes the expertise of a neurophysiologist, a behavioral medicine therapist and a gastroenterologist as part of the evaluation team. Our urodynamics laboratory has attracted patients from all 50 states and at least 15 foreign countries. Hundreds of physicians and associated health care personnel from the United States and world-wide have been observers in order to learn how to properly perform and interpret urodynamic studies in children. Twenty foreign and American urologists and neurologists have spent 6 months or more of fellowship in our Urodynamics laboratory. This has provided them with basic knowledge for recommending and evaluating treatments such as behavioral therapy and biofeedback training, proper drug selection and dosing, alternative therapies, and surgical options for improving (1) upper urinary tract drainage and function, and (2) lower urinary tract continence and drainage in children with a variety of urological conditions.

The major emphasis of Dr. Bauer's clinical research has involved the study of bladder function in children with neurologic, anatomic and functional disorders. The major breakthroughs are listed as follows:

1. Characterizing the bladder in newborns with myelodysplasia and its effect on kidney function and ultimate continence as the child grows has been his primary focus. As a result of this work there has been a change in the way newborns with myelodysplasia are assessed and managed. No longer is it acceptable to provide only expectant treatment for these infants. Urodynamic studies are now an integral part of the initial newborn evaluation. If testing of the bladder shows that the child is at risk for upper urinary tract deterioration, he or she is begun on prophylactic therapy (intermittent catheterization and anticholinergic agents) to lessen the chance of damage and reduce the need for augmentation cystoplasty later in life. An added benefit of this proactive approach has been that children accept catheterization easily as they grow and a greater number become continent at a younger age without the need for medicines or surgery.

2. Children with occult spinal dysraphism and a tethered cord syndrome (*i.e.*, lipoma, lipomeningocele, diastematomyelia, etc.) have less morbidity if they undergo corrective surgery early in life rather than waiting until the disease manifests itself with changes in lower extremity or lower urinary tract function, as it invariably will do, later in life.

3. Boys with posterior urethral valves develop various patterns of bladder dysfunction in the face of urethral obstruction on urodynamic studies. We have characterized these alterations in function to better manage the children, improving both upper urinary tract drainage and urinary continence in very efficient ways.

4. Neurologically normal children who have recurrent urinary infection or intractable urinary incontinence have been studied urodynamically in order to identify specific patterns of behavior. As a result we have proposed effective treatment protocols based on their type of dysfunction.

Currently, Dr. Bauer is working with the Division of Gastroenterology to understand the neurophysiologic relationship between the lower gastrointestinal and lower urinary tracts. The aim is to determine how dysfunction in one organ system influences the behavior in the other.

His entire career has been devoted to advancing the understanding of lower urinary tract function in children. As such, he has authored 155 original scientific articles, written chapters in 60 books, coedited a book on Pediatric Urology for Primary Care physicians, been a visiting professor at 20 academic institutions (8 in foreign countries), a lecturer or invited speaker at 68 meetings and courses (23 in foreign countries) and have delivered 120 papers at national and international meetings on a wide range of subjects relating to his research efforts.

Involvement on a national and international level:

1. Section on Urology of the AAP Executive Committee member, Secretary, then Chairman and subsequently, Executive Council member
2. President, currently (since 2008), of the International Children's Continence Society, after having first served 4 years on its Executive Board where he has worked tirelessly to advance the objectives of this global society
3. Program chair of 2 postgraduate courses
4. Reviewer on 10 peer-reviewed journals
5. Assistant Editor of the Pediatric Section of the Journal of Urology for two consecutive 5 year terms (1996 – 2006)
6. Assistant Guest Editor for the Pediatric Supplement of the Journal of Urology for the combined AAP Section on Urology and ICCS meeting in Boston in 2008

Awards Received

1. The Jonas E Salk Scholarship from the City of New York Board of Higher Education (1964)
2. Recognition as an outstanding teacher from the Urology Program at Harvard Medical School (1991)
3. First prizes for clinical and basic science research from the AAP Section on Urology (1998 & 1999)
4. Recipient of the Paediatric Urology Progress Medal from the World Federation of Societies for Paediatric Urology (2010)
5. Pediatric Urology Medal from the Section on Urology of the American Academy of Pediatrics for his outstanding contributions to his field of interest during his career (2011)
6. The Lifetime Achievement Award from the Spina Bifida Association (of America) for all the contributions that have been made to improve the health and welfare of these children (2012)

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