

Pediatric Urology: What's new?

Phichaya Sujjantararat, M.D.

Department of Surgery, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok 10700, Thailand.

Siriraj Med J 2007; 59: 259-263

E-journal: <http://www.sirirajmedj.com>

During the last two decades, there has been substantial advancement in the field of pediatric urology. Several studies have shed light on the anatomy and pathophysiology of various congenital malformations. This leads to new concepts, rational approach and improved surgical management of pediatric urologic anomalies. There has been a lot of change in surgical correction of congenital anomalies involving both urinary and genital systems. In the past, surgical outcomes were far from satisfactory. With new knowledge of the anatomy and pathophysiology, better surgical outcomes in terms of cosmesis and function can be obtained. The surgical repair of various genital disorders including hypospadias, exstrophy-epispadias and complex, congenital adrenal hyperplasia has evolved to the point that satisfies patients and their parents alike. Currently, interesting topics and what is new in pediatric urology can be summarized as the following.

1. Antenatal hydronephrosis
2. Hypospadias
 - 2.1 Urethral plate concept
 - 2.2 Tunica albuginea plication (TAP)
 - 2.3 One stage repair
3. Vesico-ureteral reflux (STING procedure)
4. Exstrophy epispadias complex (EEC)
 - 4.1 Single stage reconstruction
 - 4.2 Bladder neck reconstruction
 - 4.3 Complete penile disassembly
5. Continent urinary diversion
 - 5.1 Mitrofanoff principle
 - 5.2 Yang-Monti tube
 - 5.3 W-pouch with subserosal tunneling
6. Congenital adrenal hyperplasia
 - 6.1 Nerve sparing clitoral reduction
 - 6.2 Non-ablative clitoroplasty

1. Antenatal Hydronephrosis (ANH)

Prenatal ultrasound has been increasingly popular over the last few decades. The incidence of congenital abnormalities detected by prenatal ultrasound is 1%.¹ The rate of prenatal abnormalities detected by prenatal ultrasound varies with the gestational age. Detection rates increase when ultrasound is performed at midtrimester compared with earlier scanning.² Of all the congenital structural abnormalities detected by prenatal ultrasound, genitourinary tract ranks second only to central nervous system which account for about 20 and 50 % respectively.³ Hydronephrosis is the most common genitourinary tract abnormality

detected by prenatal ultrasound study.

However, not all cases of antenatally detected hydronephrosis are clinically significant. Persistent postnatal renal abnormalities are associated with a degree of dilatation of renal pelvis. Prenatal hydronephrosis is associated with various conditions with varying severity and prognosis, which range from transient physiologic dilatation of the collecting system that resolves spontaneously to life threatening urethral atresia. Mild dilatation, with anteroposterior diameter (APD) of less than 1.5 cm. tends to have spontaneous resolution postnatally. In addition to kidneys, the ureter, bladder wall thickness, amniotic fluid volume and fetal sex should be evaluated as well. Hydronephrosis without a demonstrable ureter suggests ureteropelvic junction obstruction (UPJO). Normally, the ureter is not visualized by ultrasound study. Dilated ureters are associated with a high grade vesicoureteral reflux (VUR), megaureter, ureterocele or ectopic ureter. Contralateral kidney should also be evaluated. If hydronephrosis involves only one kidney, the survival of the fetus is not compromised. On the contrary, bilateral hydronephrosis is more alarming, particularly in male infants with thickening bladder wall and oligohydramnios which indicates the posterior urethral valve. The major concern when facing a fetus with bilateral hydronephrosis is the presence of oligohydramnios which could adversely affect pulmonary development. Newborns with pulmonary hypoplasia rarely survive.

Amniotic fluid volume is also another important finding. It indicates fetal well being, particularly renal function and good respiratory system. Most of the amniotic fluid comes from urine which the fetus begins to form around the tenth week of gestation. Swallowing of the urine induces respiratory tract development. Therefore, oligohydramnios may be associated with pulmonary hypoplasia and affect fetal survival. In a fetus who has bilateral hydronephrosis, thickening bladder wall and oligohydramnios from the posterior urethral valve is a candidate for prenatal intervention. Before intervention for oligohydramnios, it is important to assess the likelihood of salvageable renal function and document a normal karyotype. The likelihood of salvageable renal function can be assessed by urine electrolytes. Urine electrolyte values associated with a good outcome include a urine sodium <100 mmol/L, a chloride <90 mmol/L, osm <210 mOsm/L and β 2 microglobulin <6 mg/L.⁴ The intrauterine intervention for a fetus with a posterior urethral valve and severe oligohydramnios is a vesicoamniotic shunt. (Fig 1) As this procedure

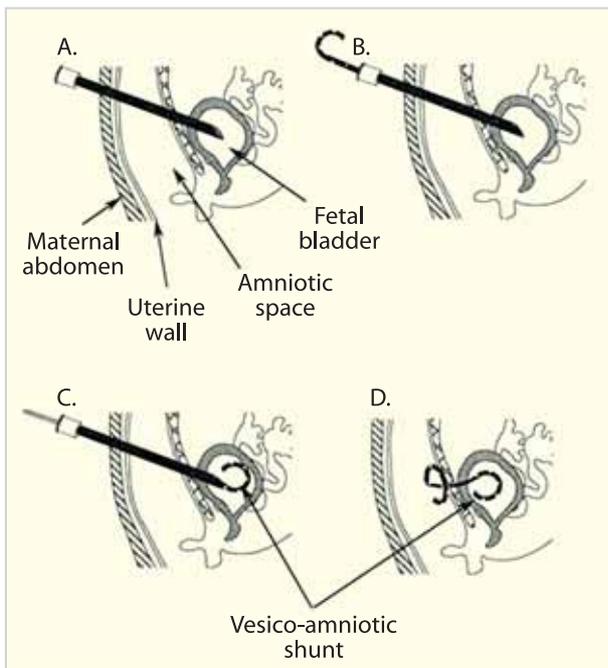


Fig 1. Vesico-amniotic shunt is indicated in a fetus that has severe oligohydramnios due to bladder outlet obstruction.

carries some morbidity and mortality rate, it is only recommended in a fetus that is between 28-32 weeks gestational age. For those who are more than 32 weeks, it is better to deliver the fetus in a center with high standard of neonatal care.

Postnatal evaluation starts immediately after birth. Complete physical examination is carried out. A palpable flank mass is usually associated with uretero-pelvic junction obstruction (UPJO) or a multicystic kidney. A palpable mass at the suprapubic area signifies a full bladder secondary to bladder outlet obstruction by posterior urethral valve (PUV) in boys and ectopic ureterocele in girls. Spontaneous voiding and urinary stream in the first 24 hours must be closely observed.

Neonates with antenatal diagnosis of hydronephrosis should be placed on prophylactic antibiotics immediately after delivery until follow up imaging studies and definite diagnoses are obtained. Amoxicillin (10-20 mg/kg) is the antibiotic of choice in newborns. After 3 months an infant's liver is mature enough, so trimethoprim-sulfamethoxazole or nitrofurantoin can be used instead of amoxicillin should continued prophylaxis be required.

Imaging study should be started with ultrasonography of the kidney and bladder. This should not be done within 72 hours after delivery as a false negative can occur due to initial low GFR and relative dehydration of the neonate

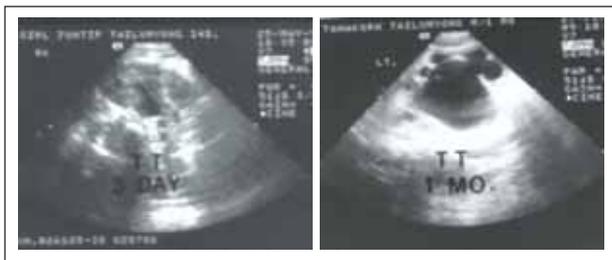


Fig 2. Ultrasonographic study of a neonate at 3 days (Left) and 1 month (Right) of age. Minimal hydronephrosis was seen at 3 days compared to severe hydronephrosis 1 month later.

(Fig 2). However in certain situations such as neonates with antenatally detected bilateral hydronephrosis, thick bladder wall and oligohydramnios which is usually associated with severe bladder outlet obstruction, prompt aggressive evaluation and management should be begun immediately.

About one half of ANH infants have a normal postnatal ultrasound or spontaneous resolve within 1 yr. Postnatal persistence of prenatally diagnosed hydronephrosis requires further evaluation. To predict the prognosis, the degree of hydronephrosis and parenchymal thickness should be classified according to the Society of Fetal Urology grading system.⁵ There is a strong correlation between the grade of hydronephrosis and the likelihood of surgical intervention being required. More than half of the children with APD of renal pelvis greater than 15 mm. require surgical intervention. Differential diagnosis of ANH includes multicystic and polycystic diseases of the kidney. The cause of persistent hydronephrosis can be determined by appropriate imaging studies such as diuretic renography, voiding cystourethrography (VCUG), IVP and magnetic resonance urography (MRU). Usually diuretic renography is obtained at 1 month of age when the kidneys of neonates are mature enough. The important information gathered from diuretic renography are split renal function (or differential renal function) and the half time ($t_{1/2}$) which is the time it takes for half of the radionuclides to leave the renal collecting system. Normal split renal function for right and left kidneys ranges from 40/60 to 60/40. Split renal function lesser than 40% indicates compromised renal function from real significant obstruction. Regarding the half time, obstruction is diagnosed when the figure is longer than 15 minutes.

After the obstruction is confirmed, careful review of postnatal ultrasound is helpful to delineate the location of obstruction. The absence of an ipsilateral dilated ureter confirms a UPJO, whereas the presence of ipsilateral ureteral dilatation suggests a mega-ureter, obstructive or nonobstructive. The exact diagnosis can be made by IVP and/or MRU. On the other hand, the presence of a dilated ureter and non-obstructive diuretic renography may be caused by VUR which is confirmed by VCUG.

In conclusion, persistence of ANH may be caused by the following diseases: (1) UPJ obstruction (50-60%), (2) VUR (35%), (3) Megaureter, (4) Posterior urethral valve and (5) Ectopic ureterocele. These diseases are treated accordingly by surgery.

2. Hypospadias

This is the most commonest malformation of male external genitalia. It is defined as an association of three malformations of the penis: (1) abnormal ventral opening of the urethral meatus that may be located anywhere from the glans penis to the perineum, (2) abnormal ventral curvature of the penis which is called chordee and (3) abnormal distribution of foreskin with a hood present dorsally and deficient foreskin ventrally. The aims of the surgery are: (1) correct ventral curvature or chordee so that the penis is straight upon erection (orthoplasty), and (2) reconstruct a new urethra so that the new external meatus is situated at the glans tip (urethroplasty, meatoplasty and glanuloplasty). In the old days, orthoplasty and urethroplasty were done separately with the latter being carried out 6-12 months after the first one. Nowadays, the surgical repair can be done in one stage, i.e. both orthoplasty and urethroplasty are carried out on the same occasion. Various changes in this field are summarized here.

2.1 Preservation of urethral plate

Formerly the urethral plate used to be removed as it was believed that this was dysplastic dartos fascia, Buck's fascia and corpus spongiosum. Several studies have shown that infant's urethral plate consists of abundant blood supply, nerve, smooth muscle and healthy connective tissue which can be used to reconstruct a new urethra⁶⁻⁸. From this knowledge, nowadays the urethral plate is preserved in all cases of hypospadias undergoing surgical repair, resulting in a lesser complication rate particularly fistula incidence.

2.2 Tunica albuginea plication (TAP)

After releasing skin and subcutaneous chordee, it is found, in some cases, that the penis still shows a certain degree of ventral curvature. This can be explained by disproportional length between dorsal and ventral sides of the corpora cavernosa. In the past, this remaining ventral curvature was left untouched or corrected by Nesbit procedure which might injure the dorsal neurovascular bundle and shorten the penis. Currently, this ventral curvature can be corrected by TAP.⁹

2.3 Onlay island flap

This one stage repair is very popular and widely done with satisfactory outcome in various reports. It has been done in Siriraj Hospital since October 1993. In this technique the urethral plate serves as the dorsal neo-urethral wall and the ventral neo-urethral wall is created by a vascular onlay flap from the inner prepuce. The technique is carried out by dissecting of an appropriate length of dorsal inner prepuce flap which is swung down and stitched to the urethral plate. Then the inner prepuce and the urethral plate are rolled to be a neourethra.

2.4 Tubularized incised plate urethroplasty (TIP)

In 1994, this technique was firstly described by Snodgrass.¹⁰ Many reports afterwards confirmed good outcomes of this technique. It is one of the single stage repairs which is widely accepted nowadays. For best result, the urethral plate has to be wide enough to form a neourethra after longitudinal incision along the plate.

All of these changing surgical repairs have been adopted and used in children with hypospadias who underwent surgical repair in Siriraj Hospital for more than one decade.

3. Vesicourethral reflux (VUR)

Surgery is the mainstay of severe grade VUR or



Fig 3.2 The same boy underwent single stage complete repair of bladder exstrophy. The bladder plate and urethra were closed as a continuous unit.

those who have multiple breaks through urinary tract infection. The results of open surgery for repair of primary VUR approach a 98% success rate. Otherwise medical treatment, i.e. antibiotic prophylaxis is indicated. Recently, an alternative treatment involving endoscopic subureteric injection of bulking agents is recommended in grade II, III, IV VUR. The bulking agent currently approved by the FDA of the USA is dextranomer/hyaluronic acid copolymer (Deflux[®]). The successful rate after a single injection varies between 50-70%. Regarding antibiotic prophylaxis, it is not further recommended in patients older than 5 yrs.

4. Exstrophy epispadias complex (EEC)

The most difficult surgical management of genitor-urinary system is EEC reconstruction. Never in the old days were satisfactory outcomes of functional bladder closure achieved. Frequently, bladder and abdominal wall closure was fraught with complications. Partial and complete dehiscence of the bladder and abdominal wall were not uncommon. To make children with EEC free from incontinence was nearly impossible. Fortunately, during the last few decades, better outcomes from functional bladder closure and reconstruction of bladder neck have been obtained in many pediatric urology centers including Siriraj Hospital. Functional bladder closure is successful thanks to iliac osteotomy and approximation of pubic symphysis in all children older than 72 hours. At the urology division, Siriraj Hospital, several cases of failed



Fig 3.1 Failed primary repair of bladder exstrophy as evidenced by the midline abdominal and bilateral inguinal incision. The bladder protruded from complete dehiscence of the wound.



Fig 3.3 External genitalia of the same boy 1 year postoperatively.



Fig 4.1 Appendix stoma of a girl with urogenital sinus who underwent appendico-vesicostomy.

functional bladder closure have been referred here.

4.1 Single stage reconstruction

Traditionally, EEC reconstruction is carried out in 2 stages. Firstly, bladder closure is done as soon as possible. At 3-5 years of age, epispadias reconstruction is carried out to complete the external genitalia repair. In 1999, single stage complete primary repair of exstrophy was reported by Grady and Mitchell in 18 patients with bladder exstrophy and 6 with cloacal exstrophy.¹¹ This technique is made possible by deeply incising the intrasymphyseal bands, complete penile disassembly and anterior approximation of the pelvic musculature over the bladder neck and urethra. This single stage technique was first done in Siriraj Hospital on 31 August 1999 with a satisfactory result. (Fig 3)

4.2 Bladder neck reconstruction (BNR)

BNR is performed to achieve continence in children with EEC. There are 3 surgical techniques commonly employed to make children with EEC void voluntarily and dry : (1) Young-Dees-Leadbetter (2), Kropp tunnel and (3) Pippi-Salle techniques. At the Urology Division, Siriraj Hospital, the first and third surgical techniques have been utilized in many patients with satisfactory results.

4.3 Complete penile disassembly

This surgical technique was done to reconstruct the epispadias for the first time in 1996.¹² In Thailand, the first boy with epispadias underwent complete penile disassembly repair at the Urology Division, Siriraj Hospital and was reported on 27 July 1999 in the Annual Scientific Meeting of the Royal College of Surgeons of Thailand.



Fig 4.2 The patient was catheterizing by herself through the Mitrofanoff stoma.

5. Continent urinary diversion

Occasionally children with urogenital sinus or EEC may not be dry even after functional bladder reconstruction and BNR. These children cannot hold urine and void at will.

Redoing BNR is less likely to achieve continence in children who fail from previous surgery. To cope with this problem, bladder neck closure and continent urinary diversion is currently the treatment of choice.

5.1 Mitrofanoff principle (Appendico-vesicostomy)

This principle involves bringing one end of a tubular structure to reimplant into the bladder while the other end is stitched to the abdominal wall creating a stoma for catheterization. A successful Mitrofanoff surgical treatment will make children dry. Bladder is emptied at intervals by clean intermittent catheterization through the stoma of the tubular structure. The first tubular structure used in the Mitrofanoff technique was the appendix. Later, uterine tube, ureter and tapered ileum were employed as conduits for continent urinary diversion. (Fig 4)

5.2 Yang-Monti

This is an alternative choice of conduit for continent urinary diversion. Frequently, the appendix in children with failed BNR is too short for the Mitrofanoff principle. This new technique makes continent urinary diversion possible in all children. A 2 cm. long ileum is opened along the antimesenteric border and rolled into a tubular structure 5-6 cm. long. The reconfigured ileum can be used in the same way as in the Mitrofanoff principle.

5.3 Bladder augmentation

Children with EEC and small bladder plate are not good candidates for functional bladder closure. The small bowel is used for bladder augmentation, resulting in a pouch. The continent pouch is made possible by either the Mitrofanoff or Yang-Monti methods.

6. Congenital adrenal hyperplasia (CAH)

This disease is caused by deficiency of adrenal 21- or 11- hydroxylase for more than 95% of the cases. As these enzymes are necessary in producing adrenal corticosteroid and mineralocorticoid, low levels of these two hormones are anticipated, resulting in no negative feedback at the pituitary gland. Consequently, the pituitary gland secretes a high level of ACTH which constantly stimulates the hyperplastic adrenal gland to produce a high level of androgen. The newborn who has these enzyme deficiencies will have clitoromegaly, no labia minora, rugae of labia majora and a variable degree of labioscrotal fusion. Frequently, the ambiguous genitalia confuses the doctors regarding gender assignment at delivery.

Historically, these poor girls underwent either clitorotomy or clitoral relocation. The clitoris should not be resected anymore as it is a sensitive organ and necessary for reaching orgasm. For clitoral relocation, part of the clitoris was put under the pubic symphysis resulting in a unsatisfactory cosmetic result. Even worse, when these girls grew older, they would experience pain after the clitorides were stimulated during sexual activity.

At the Urology Division, Siriraj Hospital, nerve-sparing clitoral reduction has been employed for almost 15 years with satisfactory results. On 9 February, 1993 the first girl with CAH underwent nerve sparing clitoral reduction. Last year a new surgical technique called non-ablative clitoroplasty was mentioned in the annual scientific meeting

of the European Society of Pediatric Urology, held in Athens, Greece. The concept behind this new technique is that no or minimal tissue of the patient is removed.

In the future, the management trend will constantly change. New surgical techniques are introduced for better outcomes. Currently, the hottest issue is robotic-assisted surgery such as pyeloplasty and ureteral reimplantation. Series of children undergoing robotic assisted pyeloplasty have been reported in many journals and scientific meetings. As Siriraj Hospital has possessed the latest Robot da Vinci[®] S since a month ago, we hope to use this machine in pyeloplasty in the near future.

REFERENCES

1. Grisoni ER, Gauderer MWL, Wolfson RN, Izant RJ Jr. Antenatal ultrasonography: the experience in a high risk perinatal center. *J Pediatr Surg* 1986; 21: 358-61.
2. Taipale P, Ammala M, Salonen R, Hiilesmaa V. Two-stage ultrasonography in screening for fetal anomalies at 13-14 and 18-22 weeks of gestation. *Acta Obstet Gynecol Scand* 2004; 83: 1141-6.
3. Elder JS. Antenatal hydronephrosis: fetal and neonatal management. *Pediatr Clin North Am* 1997; 44: 1299-321.
4. Glick PL, Harrison MR, Golbus MS, Adzick NS, Filly RA, Callen PW, et al. Management of the fetus with congenital hydronephrosis. II. Prognostic criteria and selection for treatment. *J Pediatr Surg* 1985; 20: 376-87.
5. Fernbach SK, Maizels M, Conway JJ. Ultrasound grading of hydronephrosis: introduction to the system used by the Society for Fetal Urology. *Pediatr Radiol* 1993; 23: 478-80.
6. Baskin LS, Erol A, Li YW, Cunha GR. Anatomical studies of hypospadias. *J Urol* 1998; 160: 1108-15.
7. Erol A, Baskin LS, Li YW, Liu WH. Anatomical studies of the urethral plate: why preservation of the urethral plate is important in hypospadias repair. *BJU Int* 2000; 85: 728-34.
8. Baskin LS, Erol A, Li YW, Liu WH. Anatomy of the neurovascular bundle: is safe mobilization possible? *J Urol* 2000; 164: 977-80.
9. Baskin LS, Duckett JW. Dorsal tunica albuginea plication for hypospadias curvature. *J Urol* 1994; 151: 1668-71.
10. Snodgrass W. Tubularized, incised plate urethroplasty for distal hypospadias. *J Urol* 1994; 151: 464-5.
11. Grady RW, Mitchell ME. Complete primary repair of exstrophy. *J Urol* 1999; 162: 1415-20.
12. Mitchell ME, Bagli DJ. Complete penile disassembly for epispadias repair: The Mitchell technique. *J Urol* 1996; 155: 300-4.