Lower Urinary Tract Reconstruction in Childhood

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Neurogenic disorders, congenital anomalies, iatrogenic lesions and malignancies represent the main indications for reconstructive urogenital surgery in childhood. The goal of this surgery is to restore the individual to normal. The question arises which operative strategy is most suited to achieve this goal and at what point in time this intervention should take place.

Especially the adolescent patients and young adults have quite definite expectations pertaining to the outcome of this surgery. Voiding should be possible through the urethra under voluntary control, at convenient intervals and in the absence of urinary tract infections. Continence is of paramount importance as it affects every aspect of social and professional life. The impact and meaning of nighttime incontinence is often neglected. Many authors simply remark that this problem might easily be solved by the use of diapers at night, however, the consequences in puberty and later on in a partnership are detrimental.

The patient's wishes are, however, not always in line with the urologist's concern in planning reconstructive lower tract surgery. The procedure that is chosen must be adapted to the patients mental capacities. A continent internal reservoir necessitates CIC at regular intervals and in the first months after surgery regular irrigation for mucus evacuation has to be performed. This of course requires a certain level of intelligence and responsibility in order to assure compliance. Moreover, the patient's orthopedic status is a decisive factor. For wheelchair-bound patients, urethral CIC is nearly impossible or, to say the least, impractical. An almost invisible stoma located in the umbilical funnel is more advantageous.

Often renal function limits the amount of alternative procedures that are available. Continent diversion is preferable in many respects, nevertheless,

this type of diversion may not be benefical if renal function is severely compromised. Experience has shown that the borderline creatinine level is 1.5 mg/dl. If renal function is less, then continent urinary diversion is too risky.

Up to the point when the child enters school, urinary continence is a major issue as the home and parents create a protective and understanding environment. With increasing social contact in school or at a friend's house, this aspect becomes crucial. When puberty begins, body image is very important.

Intermittent catheterization, antibiotic prophylaxis and anticholinergic drugs have significantly lowered the rate of parenchymal infections during the first years of life and have thus substantially improved the changes for urinary tract reconstruction at a later time. When the adolescent or young adult leaves the protective and supportive environment of the family home compliance steadily decreases. These aspects must all be considered when planning reconstructive surgery in the child.

Therapeutic strategies

In order to stabilize renal function in an infection-free environment while permitting urinary continence, it is essential to correctly assess the surgery's limitations and risks. Despite all the progress in reconstructive surgery, we are not able to functionally reconstruct a sphincter. All attempts in this direction merely represent "balanced obstruction" and constitute a compromise with an unpredictable and all too often regrettably unfavorable outcome. The higher the degree of obstruction is, the more likely a final condition with CIC becomes, the more incomplete the obstruction is, the more unsatisfactory the result is because a life with diapers is the consequence.

Artificial sphincters are widely implemented to institute continence, but especially in children the

Table 1. Results of artificial sphincter implantation in children

| Author | No of pts | Complication rate | Continence rates | References |
|----------|-----------|-------------------|--------------------------------|------------|
| | | | | |
| Light | 132 | 24 % | 70 % continent | (28) |
| Gonzalez | 15 | 38 % | 67 % continent, 34 % explanted | (14) |
| Aprikian | 27 | 39 % | 88 % continent | (1) |
| Bosco | 36 | 61 % | 74 % continent, 25 % explanted | (4) |
| Grein | 39 | 56 % | 87 % continent | (15) |
| Barrett | 45 | 60 % | | (2) |

results of this surgery are quite depressing (Table 1).

Results are not much better in adults. In a series of 100 patients Nurse and Mundy report a 45 % complication rate ⁽³⁶⁾. Recently, a paper investigating children with the AMS 800 sphincter demonstrated that in 50 % of cases silicone shedding occurred ⁽³⁹⁾. The long-term effects are entirely unknown. Because of the high complication and moderate success rate, we feel that artificial sphincters should not be implanted before puberty.

Bladder neck reconstructions (BNR) using the Young-Dees procedure or one of the many modifications have variable and often unsatisfactory results. Despite improvements, 30-50 % of patients remain incontinent and have many other voiding difficulties and complaints (23,26) (Table 2). Our own results were no better (Table 3).

A recent review from the Indiana University Hospitals demonstrated the detrimental damage this pro-

cedure can also cause to the upper urinary tract (Table 4) ⁽²³⁾. Only 57 % of patients had normal imaging studies of the upper urinary tract after BNR.

Philip Ransley has also had limited success in achieving continence using this technique (21), and has developed a low threshold to proceeding with bladder augmentation along with a suprapubic Mitrofanoff stoma for CIC (personal communication). In the face of long-term results, much of the enthusiasm associated with BNR surgery in early publications has given way to skeptical comments. Even though some centers still report that up to 80 % of their patients achieve "an excellent surgical result" (12,13), this is defined as one soiling/day and 3-h dry intervals.

In a large prospective urodynamic study comparing patients with closed exstrophied bladders before and after bladder neck reconstruction, Hollowell (18-20), as well as Kazachkov (24) have

Table 2. Results of bladder neck reconstruction (BNR) and bladder augmentation in children with bladder exstrophy (21)

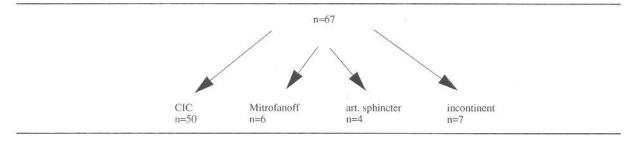


Table 3. Our results after Mainz Pouch augmentation and modified Young-Dees procedure in 8 children (age: 3 to 13 years)

| complete continence | 1 | |
|--|---|------------|
| daytime continence, partial nighttime continence | 2 | |
| daytime continence, nighttime incontinence | 2 | |
| complete incontinence | 2 | conversion |
| no spontaneous voiding | 1 | conversion |

Table 4. Results of the Young-Dees bladder neck reconstruction (23)

| 1978 - 1988 | (n=47) |
|---------------------------|---------|
| continence | 39-64 % |
| reoperation | 27-75 % |
| upper tract deterioration | 24 % |

shown that incontinence is mainly due to bladder instability and not solely a result of insufficient outlet resistance. The closed exstrophied bladder produces involuntary contractions that cause a substantial rise of intravesical pressure which easily overcomes the leakpoint pressure. Neuropharmacologic investigations we have performed at our institution indicate that alterations in the innervation and neurophysiology of these exstrophied bladders are responsible for these observations. The lack of response of these bladders to anticholinergic therapy seems to be attributable to functional abnormalities of the cholinergic receptor and the presence of non-adrenergic-non-cholinergic (NANC) innervation.

A substantial number of children have to perform CIC to evacuate their bladder or require CIC to be continent, at least to some degree. Data analysis has also shown that for some reason gender seems to play a role. While female patients with bladder exstrophy who are able to void without residual urine after bladder neck reconstruction demonstrated a low complication rate, male patients encountered significantly more problems.

The problem of producing continence is not specific to exstrophied bladders. Every form of incontinence is difficult to manage, especially in children. For practical purposes, a difference must be made between patients with a functioning sphincter (leak point pressure > 25 cm waterscale) and a small bladder capacity (< 100 ml) and patients with an incompetent sphincter (leakpoint pressure < 25 cm waterscale). The best results with bladder augmentation alone are achieved when iatrogenic bladder lesion or neurogenic bladder with an outlet resistance of more than 25 cm waterscale was the indication for intervention. These patients benefit from sole bladder augmentation, especially, if they are not wheelchairbound and even if spontaneous voiding is not possible and CIC is necessary. If the outlet resistance has to be increased, then sling-plastics (7,30,37) seem to produce the most satisfactory results and are most certainly to be preferred to artifical sphincters.

In our patient material the patient's orthopedic status was of major significance. Non-wheelchair-bound patients demonstrate less upper urinary tract damage while wheelchair-bound patients show a tendency to gain weight as they become older and also experience increasing continence problems during physical activity which becomes evident during

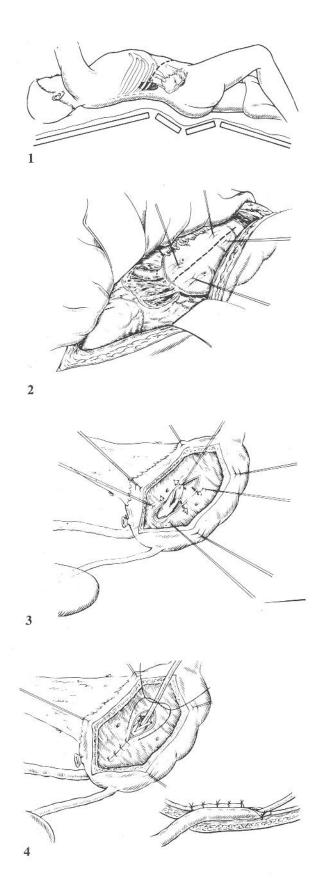
Table 5. Complications of the Mainz pouch I procedure at our institution: 1983-03/92: n=314

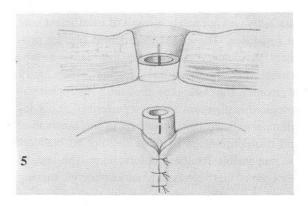
| Early complications | Treatment | n |
|--|------------------------|---------------|
| ileus | op. revision (n=3) | 4 |
| pouch tamponade | endoscopic evacuation | 2 |
| wound dehiscence | op, revision | 2 |
| intestinal fistula | conservative treatment | 2 |
| nipple necrosis | new ileum nipple | 1 |
| appendix necrosis spontaneous pouch | ileum nipple | 1 |
| rupture | op. revision | 1 |
| | | 13/314 (4.1%) |
| Late complications | Treatment | ņ |
| stones | endoscopic treatment | 27 |
| stomal stenosis | endoscopic treatment | |
| ureteral stenosis | neoimplantation | 18 |
| dividia dividia | nephrectomy | 5 |
| ileus | op. revision (n=3) | 4 |
| pouch perforation | op. revision | 4 3 3 |
| nipple gliding | op, revision | 3 |
| pouch tamponade (mucappendix stoma | | t 2 |
| dislocation | ileum nipple | 1 |
| | | |

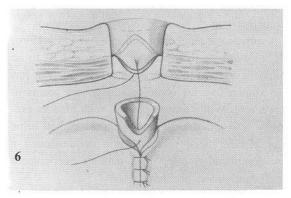
everyday activities such as getting in and out of a car. With increasing disproportion of the upper part of the body the umbilicus moves towards the symphysis. Especially female patients showing such changes have difficulty in performing urethral CIC. For all of these patients the Mainz pouch I with a nearly "invisible" umbilical stoma is the method of choice. This procedure yields beneficial results and produces a low complication rate (Table 5).

The last 10 years have taught us many lessons on how to avoid certain complications. Obstruction at the site of ureterointestinal anastomosis was primarily seen in patients with preoperatively dilated upper urinary tracts. An extraperitoneal approach to the pouch from a rightsided flank incision has proven very beneficial for repair of ureterointestinal strictures (Fig. 1). After the pouch is opened (Fig. 2), the ureter is reimplanted by means of a short submucosal tunnel (Fig. 3 and 4). When the pouch is of sufficient capacity, the risk of reflux is minute. Stoma stenosis, a frequent problem in our early series was effectively treated by V-plasty of the efferent segment (Fig. 5-7).

Stones were most commonly found attached to staples (Fig. 8,9). Using the appendix as a continence mechanism obviates this problem. After having created mesenteric windows and incising the taenia libera of the colon (Fig. 10), the appendix is embedded (Fig. 11 and 12). This method of creating







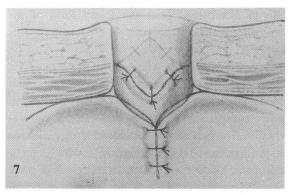
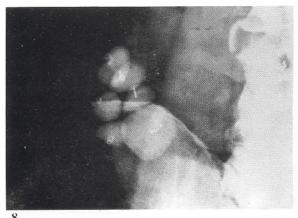


Figure 5-7. V-plasty of the umbilical stoma in order to prevent stomal stenosis. The suture line runs obliquely, thus avoiding concentric stricturing.

Figure 1. Optimal approach for pouch revision: right-sided flank incision. 2. Flank incision leads directly to the site of ureterointestinal implantation. The pouch is opened parallel to the ureteral anastomosis site (dotted line). 3. Cheation of a wide "button-hole" to comfortably accomodate the dilated ureter. After incision of the pouch mucosa, a wide, yet, short submucosal tunnel is prepared. 4. Two anchor sutures at 5 and 7 o'clock hold the ureter in position. The bowel mucosa is closed over the ureter with single stitch sutures (catgut 5/0).



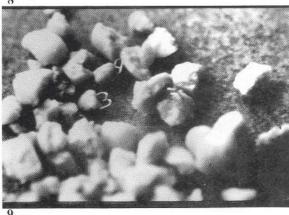


Figure 8-9. Plain x-ray film shows large stone development around staples in a Mainz pouch I. Stones attached to staples used to create an ileum nipple in a Mainz pouch I.

a continent outlet is preferred and used whenever possible. If the appendix is structurally altered and unusable, newer continence mechanisms such as a seromuscular tube ⁽²⁷⁾ (Fig. 13-15) or a tapered segment of ileum having been embedded in a taenia of the colon ⁽²⁹⁾ are used (Fig. 16-18).

To avoid diarrhea or frequent bowel movements in patients with myelomeningocele who undergo Mainz pouch I continent urinary diversion, the ileocecal valve is reconstructed (Fig. 19-24) (11).

For those patients with a competent anal sphincter and a functionally damaged or morphologically absent urinary sphincter, the Mainz pouch II (Sigma-rectum pouch) is a viable alternative. Preoperatively, anal sphincter competence is evaluated using a liquid enema. The patient has to be able to hold the saline enema for at least three hours during the day-time while walking about and for many hours at night. In addition a rectodynamic evaluation should render positive results (44). Previous or planned radiotherapy is a contraindication for the Mainz pouch

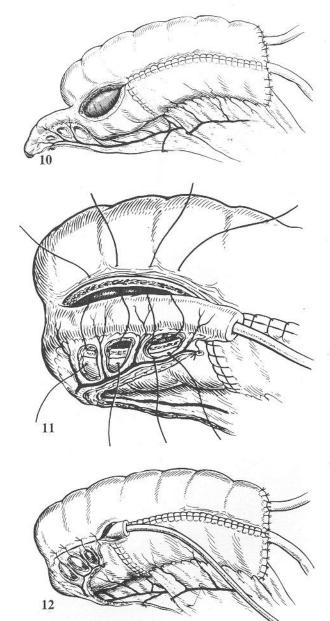


Figure 10. Excision of windows in the appendicular mesentery and incision of the taenia libera of the colon. 11. Placement of a 16 French silicone catheter through the appendix into the pouch. Seromuscular sutures (polygylcolic acid 4/0) are brought through the mesenteric windows in order to embed the appendix. 12. Closure of the seromuscular layer over the appendix with single stitch sutures (polyglycolic acid 4/0).

II procedure. The same holds true for pathologies of the sigmoid colon.

Since November 1990, 19 children have received a Sigma-rectum pouch at our institution. In these 19 children, 3 ureteroneoimplantation had to be performed in two children during two sessions. One child had an ileus 5 months after urinary diversion which was operatively revised. The obstruction at

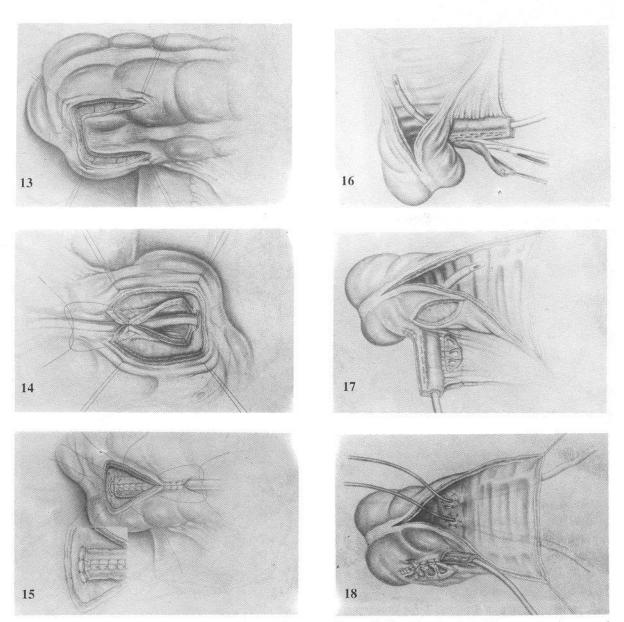


Figure 13. Seromuscular tube as continence mechanism. Incision of the seromuscular layer of the colon and careful separation of mucosa and seromuscularis laterally (1-2 mm). 14. After a 5 mm incision of the mucosa at the proximal end of the seromuscular strip, a tube is formed over a 18 French silicone catheter with single stitch sutures (polyglycolic acid 5/0). 15. Anastomosis of the newly created tube with the mucosa of the colon (5/0 single stitch sutures). Subsequently, the seromuscular layer is closed over the tube, thus submucosally embedding the continence mechanism.

the site of ureterointestinal anastomosis was caused by the use of slowly resorbable suture material. For this reason we now recommend the use of chromic catgut for ureteral implantation. The Mainz pouch II procedure is also indicated to correct failed ureterosigmoidostomy. For conversion surgery, a pre-

Figure 16. Tapered and embedded ileum as a continence mechanism. After insertion of a 14 French silicone catheter, the ileum is tapered and excess bowel discarded. 17. Oblique incision of the seromuscular layer between the taenia mesocolica and libera. Creation of mesenteric windows as described previously.

18. After placement of sutures through the mesenteric windows the seromuscular layer is closed using single stitch sutures, thus embedding the tapered ileal segment.

existing antirefluxive colonic conduit can be incorporated into the Sigma-rectum pouch, obviating the need for a new ureteral implantation. Fixation of the pouch to the promontory or the psoas muscle prevents excess mobility which might cause ureteral kinking and subsequent obstruction.

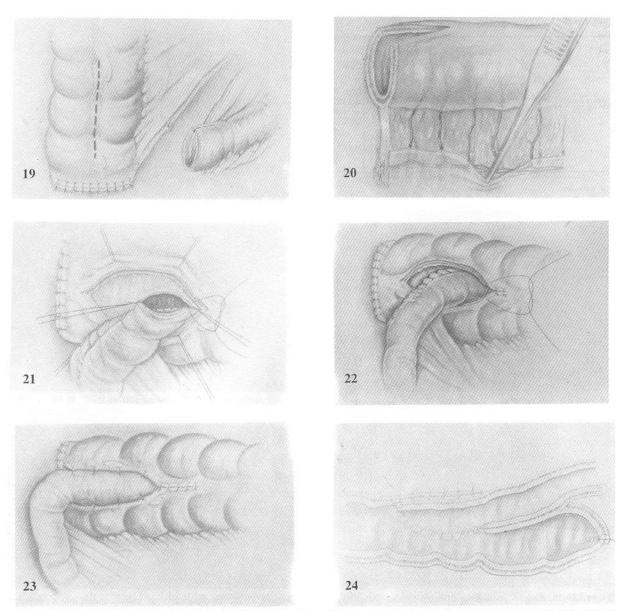


Figure 19. Reconstruction of the ileocecal valve. Terminal incision of the seromuscular layer of ascending colon over 6-7 cm and antimesenteric incison of terminal ileum. Incision of colonic mucosa over 2 cm (not shown). 20. Mesentery defatted and serosa removed.

21. Ileocecal anastomosis (5/0 polyglycolic, single stitch sutures). 22. Closure of seromuscular layer creating a submucosal tunnel 4 cm long.

23. Lateral sutures fix ileum to ileocecal region in order to avoid kinking and obstruction. 24. Lateral view shows ileum smoothly diving into cecum.

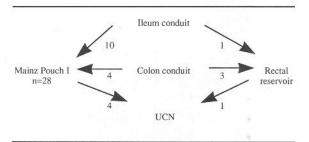
With increasing frequency we are confronted with aspects of converting incontinent form of urinary diversion into continent types. A total of 33 conversion procedures were performed in young adults at our institution (Table 6).

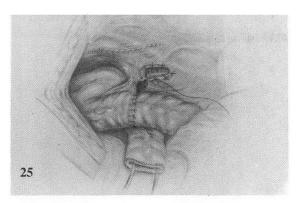
If, as mentioned above, a Sigma-rectum pouch is not indicated because of anal sphincter incompetence, an antirefluxive colonic conduit (Fig. 25-26) can also be incorporated into a Mainz pouch I continent reservoir. Figures 27-29 show examples of

how this can easily be achieved without the need for repeated ureteral implantation. Equally, the colonic conduit can be incorporated into a Mainz pouch II (Fig. 30-31).

The question of which intestinal segment to use for urinary diversion is fraught with difficulty. Virtually every segment of the intestinal tract has been used in diversion surgery and all have their specific advantages and disadvantages. Novel concepts such as incorporating an isolated gastric segment have

Table 6. Continent conversions performed at our institution (n:33) (43)





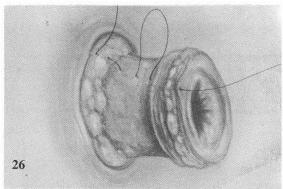
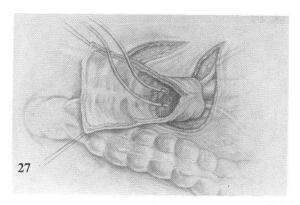
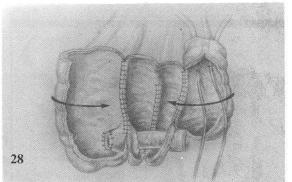


Figure 25-26. Correct positioning of colon conduit: After open, transcolic ureteral implantation the cranial portion of the conduit is retroperitonealized. Stoma creation: Suture grips the seromuscular layer and then the mucosa. Intracutaneous sutures avoid urine fistulas.

produced significant complications as increasing experience and follow-up is showing ⁽⁴²⁾. Hematuria, alkalosis, dysuria and peptic ulcers in a "dry" bladder have dampened most of the initial enthusiasm associated with using gastric segments ⁽³³⁾. Cases of perforation of the gastric segment of an augmented bladder secondary to peptic ulcer disease have even been reported ⁽³⁸⁾. The indications remaining are cloacal exstrophies and patients demonstrating compromised renal function ^(5,40).





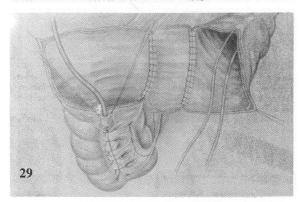
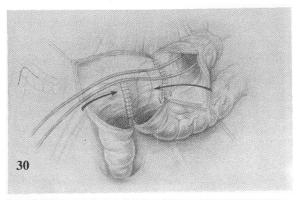


Figure 27-28. Incorporation of colon conduit into a continent reservoir (continent conversion). Antimesentic opening of colonic conduit. Ureters stented. Side-to-side anastomosis of split colonic conduit to ileocecal reservoir (Mainz pouch I). 29. As Fig. 28, only that appendix has been used as continence mechanism, thus saving one loop of ileum.

Ileum has certainly enjoyed broad popularity for constructing continent reservoirs but here also long-term follow-up is demonstrating a significantly increased rate of complications at the site of ure-terointestinal anastomosis. Antirefluxive ureteral implantation into an isolated segment of large bowel which can also be used for augmentation is technically simpler and reliably prevents reflux.

When intermediary diversion or incontinent diversion has to be performed in order to stabilize or



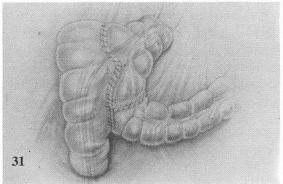


Figure 30-31. Side-to-side anastomosis of split colonic conduit to sigma-rectum pouch (Mainz pouch II),

improve renal function, the colonic conduit should be preferred. Our series demonstrated that the upper urinary tract condition at the time of incontinent diversion is of the utmost importance and is a significant factor in predicting the outcome. From 1967 to 1983 we constructed a sigma conduit in 94 children with a mean age of 7.2 years at the time of operation.

94 of 124 preoperatively dilated renal units showed a decrease in dilatation postoperatively, whereas in 10 units dilatation increased. 9 of 49 renal units which had pyelonephritic changes preoperatively showed further changes during follow-up. In an additional two renal units pyelonephritic changes were noted during the follow-up even though these changes were not present preoperatively. We believe that dilated ureters and ureters with a thick and hypertrophied walls are prone to stricturing at the implantation site. The risk for postoperative dilatation is severely increased. Therefore intermediate diversion should be performed at an early stage of disease and should not be considered as the ultimate solution.

Using the ileocecal segment for urinary diversion

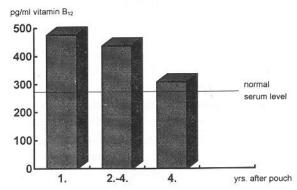


Figure 32. Vitamin B12 serum levels in 161 patients after Mainz pocuh I continent urinary diversion.

affords the opportunity to construct a large capacity, Iow pressure reservoir using very little bowel. Ureteral implantation is easily accomplished in an antirefluxive manner using a simple and reproducible technique. Moreover, a wide variety of continence mechanisms may be used. Any supposed disadvantage by resection of the ileocecal valve can be obviated by surgically reconstructing the valve (see above).

Looking at the metabolic consequences of urinary diversion, the question of vitamin deficiency development and metabolic acidosis are the most pertinent issues. We monitored the vitamin B12 levels in 161 patients having a continent Mainz pouch I reservoir over a period of 6 years. Results showed a value of vitamin B12 which was markedly reduced when compared to an age-matched control group. B12 levels were, however, still within normal values (Fig. 32).

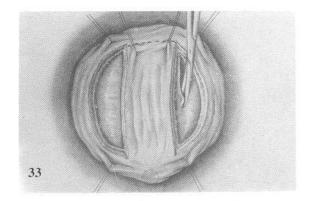
Similar results are reported by other authors ⁽⁴¹⁾. In a retrospective analysis of 130 patients after urinary diversion, Jahnson even found 11 patients with a vitamin B12 deficiency, however, all 11 patients had been irradiated preoperatively ⁽²²⁾. Routine evaluation of vitamin B12 serum levels should be carried out at follow-up. For such purposes B12 detection using the urinary methylmalonic acid (MMA) test seems to be superior to serum B12 radioassays ^(34,35).

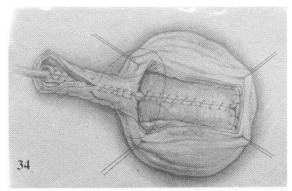
More than 197 reports of neoplastic change occuring after ureterosigmoidostomy have been published to date. However, secondary tumors are also found in other types of urinary diversions. 32 cases have been reported so far in ileum conduits, colonic conduits and ileo- and colocystoplasties. In our opinion, the hypothesis that the close contact of urethelial epithelium and bowel mucosa is the most likely pathomechanism involved in secondary malignancy development. Consequently, the same risk for secondary malignancy should exist in every form of urinary diversion and, equally, in patients having had bladder augmentation. Due to the stasis of urine in continent reservoirs and augmented bladders, the risk of malignant transformation might be higher in these cases. Of the 17 secondary malignancies that were found in bladder augmentations, 11 occured in ileocystoplasties and 6 in colocystoplasties indicating that no difference exist in this respect between ileum and colon. Recent research has suggested that metaplasia also occurs following gastrocystoplasty (25)

The latent period for detection in humans was on average 18 years ⁽¹⁰⁾. It is simply the fact that ure-terosigmoidostomy has been around for a lot longer than all other forms of diversion and, subsequently, the highest incidence of secondary malignancies has been reported in this type of urinary diversion. Whatever may hold true, regular examinations of the ureterointestinal anastomosis must be performed and, thus, those reservoirs are to be preferred that allow easy access for these investigations. Specially developed instruments facilitate this examination by variable degrees of magnification.

Should the suspicion of malignancy arise during optical evaluation of the ureteral implantation site, a biopsy can easily be performed. In case of grade 3 dysplasia, conversion to another form of continent urinary diversion is possible. Continent urinary diversion has proven to be advantageous in such cases as the patient is not confronted with an incontinent stoma causing additional dismay ⁽⁴⁶⁾.

Acidosis as a result of urinary diversion needs to be corrected if the Base Excess drops below - 2.5. This is prevention and not treatment. The situation is comparable to that of a diabetic patient. No one would wait until a diabetic coma develops before insulin is administered but looking at the follow-up programs of many institutions, this is exactly what is being done with respect to urinary diversion. Acidosis needs to be corrected early by administering alkalinizing drugs before dangerous levels are reached and most certainly before hyperchloremia develops.





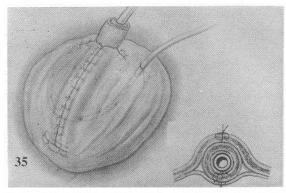


Figure 33. Technique of continent vesicostomy using bladder flap. Preparation of a 3x8 cm seromuscular "Boari"-flap and careful lateral mobilisation of mucosa and opening of the bladder lateral of the flap. 34. Closure of bladder mucosa using a running suture and adaptation of mucosa over a 18 French silicone catheter, and closure of seromuscular layer over tube. 35. Newly created tube subsequently laid back into submucosal bed, and the detrusor closed over the tube.

What will the future bring?

An increasing number of publications dealing with bladder augmentation using dilated ureteral tissue have been published in the recent past (3,6,16,45). Interestingly, none of these papers have cited the author who has first published ure-terocystoplasty. It was Herbert Eckstein who first

used a dilated ureter for augmentation and published this technique in 1973 ⁽⁹⁾.

The early results of ureterocystoplasty compare favorably with those of enterocystoplasty in selected cases, but further experience and follow-up is certainly required.

Similar advances have been made in constructing a continent vesicostomy. As an alternative to the Mitrofanoff principle ⁽³¹⁾ which is fraught with specific disadvantages ^(8,32,47), the use of a submucosally embedded bladder flap is a promising novel development (Fig. 33-35) ^(17,43).

In conclusion, we must realize that we are still far away from being able to construct a worthy alternative to the natural bladder. Nevertheless, substantial progress has been made. All reconstructive surgery of the lower urinary tract is by nature fraught with specific disadvantages. However, by carefully considering the advantages and disadvantages of each technique, a refined differential indication will permit us to identify the procedure that is most beneficial for the individual patient.

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