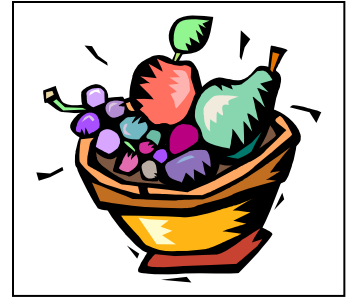


Laxative Diet



Foods that may produce a "laxative" effect:

- **Breast milk**
- **Fruit:** cherries, grapes, pineapple, strawberries, avocados, mango, papayas, plums, peaches, pears, raspberries, blueberries, oranges, etc.
- **Vegetables:** spinach, carrots, sauerkraut, cabbage, broccoli, peas, sweet potato, corn, green beans, cauliflower, etc.
- **Juices:** prune, apple, apricot, and cranberry
- **Dried fruits:** raisins, apricots, prunes, dates and figs
- **Spicy foods**
- **High fat dairy products:** known to produce laxative effect for some, however may cause constipation for others, trial these foods and if there is no benefit to bowel movements only provide enough dairy to meet calcium needs for age
- **Foods high in fat:** every persons digestive tract is different, for some fatty foods will provoke bowel movements and constipate others
- **Chocolate:** dark chocolate
- **Caffeine:** known to relax anal sphincter and produce a laxative effect for some patients

Functions of Fiber:

Soluble:

Prolong stomach emptying time so that sugar is released and absorbed more slowly, forms a gel when mixed with liquid which helps to soften stool

Insoluble:

Moves bulk through the intestines, promotes regular bowel movements, and helps prevent constipation

- If your child is taking laxatives soluble fiber will be help to bulk the stool
- If your child is not on laxatives and is trying to avoid constipation encourage sources of insoluble fiber
- Keep in mind most foods contain a combination of both soluble and insoluble fiber
- Examples of foods high in insoluble fiber: oranges, English muffin, whole-wheat bread, broccoli, corn, baked potato with the skin, and kidney beans

How much fiber is recommended for your child?

- Initial suggestion: age plus 10, example 4 years + 10 = 14grams/day
- Once child is consuming 1500 calories increase fiber consumption to 25-30grams/day



High Fiber Meal Ideas:

Breakfast:

- Whole wheat waffles w/fresh fruit (Brands to try: Kashi, or Eggo)
- Cereal choices: oatmeal, Frosted Mini Wheats, Kashi Mighty Bites, Raisin Bran, Wheat Chex, Cracklin Oat Bran, etc.
- Whole wheat bagel, or English muffin w/butter and jelly
- Bran muffins, add dried fruit to increase fiber content
- Add fresh fruit or juice to any breakfast

Lunch and Dinner:

- Vegetable soup w/whole wheat crackers
- Make a sandwich w/whole wheat bread or pita and add leafy green lettuce and tomato with meat of choice
- Use whole wheat macaroni to make macaroni and cheese and add peas
- Use whole wheat spaghetti; add sautéed zucchini and tomatoes to sauce, serve fresh fruit as a side or salad
- If it's pizza night, make your own, use store bought whole wheat crust, add sauce, small amount of cheese and load up with vegetables for toppings (green, red, yellow or orange peppers, mushrooms, tomatoes, olives, etc)
- Mexican Night; use whole wheat tortillas, add extra vegetables (tomatoes, lettuce, olives, etc), meat of choice, okay to add sour cream or try avocado's for a topping
- Prepare any meat, baked, broiled, grilled, etc; serve with whole wheat starch (brown rice, whole wheat pasta, legumes, beans, etc.) and add a vegetable as a side

Snack Ideas:

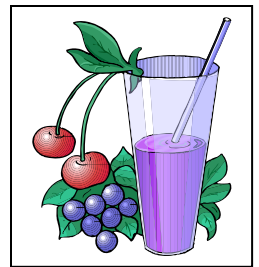
- Cut up green, red, yellow or orange peppers, cucumber slices, split peas, etc and serve with vegetable dip
- Sliced pears, peaches, strawberries, or cubes of cantaloupe with fruit dip
- 5" inch piece of celery, 2tbsp peanut butter, with small box of raisins
- Whole grain crackers, if high fat cheeses function as a laxative food add to snack

Why is Fluid Important?

- Important to drink adequate fluids while increasing fiber in the diet and once goal fiber level is achieved.
- Consuming more fluids can help avoid constipation
- The amount of fluid recommended for your child depends on child's weight, type of foods consumed and activity level

Fiber supplements:

- Benefiber
 - do not count Benefiber prescribed by MD for daily recommended amount
 - 1tbsp provides 3gm of soluble fiber
- Flax seeds
 - can be purchased as seeds or already ground; add to foods applicable
 - 1tbsp ground provides 1.9gm of fiber, 1tbsp whole provides 2.8gm of fiber
- Metamucil
 - 1tbsp provides 3gm of soluble fiber
- Do not recommend adding fiber supplements to beverages



Three Meals Rule: Ask your Dietitian if this rule applies to your child

- Every time food is eaten the body produces a gastric reflex, meaning the colon moves
- To help train the colon, offer three meals per day: same time, same quantity, similar foods, and no snacks in between
- Liquids are allowed in between meals only



Constipating Diet




Children with a tendency towards diarrhea need a diet and medications that will slow down the colon. Foods that further loosen the bowel movements are eliminated to help the colon move more slowly. The diet is very restrictive in the beginning, and as the diarrhea is controlled, more foods can be added to your child's diet. During this Phase I Diet you will need to give you child a multivitamin with mineral supplement. Anti-diarrheal medications are commonly used as well.

Your child can then choose one new food every 2-3 days and you will observe the effect on his/her colonic activity. If your child soils after eating a newly introduced food, eliminate that food from the diet. Continue to try other new foods, observing the effect on your child. The most liberal diet possible is the goal. If your child remains clean on the liberal diet, the dose of anti-diarrheal medication can gradually be reduced to the lowest dose effective to keep the child clean for 24 hours.

This type of diet is made up of foods to reduce the amount of stool in the lower bowel. Not everyone has the same reaction to the same foods, so learning to identify the foods that control your child's diarrhea best will be important. Using the Foods Recommended List should help you increase the variety of tolerated foods in your child's diet more quickly. Continue to give your child a multivitamin with mineral supplement each day.

Phase I Diet: Use to Constipate Your Child

Food Groups	Food Recommended	Food to Avoid
Milk and Milk Products	Rice Milk	All others
Vegetables	None	All
Fruits	Applesauce, Apples without skin, bananas	All others
Starches, Bread & Grain 	Bread, crackers and cereals made from refined flours, pasta and noodles made from white flours, white rice, pretzels, white potatoes without skin, dry cereals such as: Rice Krispies, Rice or Corn Chex, Corn Flakes, Kixx.	All others
Meat or Meat Substitutes	Baked, broiled, boiled or grilled meat, poultry or fish	All others
Fats and Oils	Limit amounts of butter, margarine and oils in food preparation during this phase, non-stick spray is allowed	All others
Sweets and Desserts	Made from allowed ingredients, plain cake, gelatin or popsicles, Rice Dream Frozen Dessert and limit amounts of concentrated sweets such as jelly and marshmallows	All others

Some additional tips when beginning to add foods into your child's diet

- Avoid extremely hot or cold foods. Foods served warm or at room temperature may be better tolerated.
- All vegetables should be well cooked
- Avoid raw fruits and vegetables
- Avoid foods and things that may produce gas or cause cramps, such as carbonated drinks, chewing gum, beans, cabbage, highly spiced foods, and swallowing air while talking and eating, or using a straw.

Phase II Diet: use to increase variety in diet

Food Group	Foods Recommended	Foods to Avoid
Milk and Milk Products	All milk products-limited to 2 - 8oz servings per day, Rice Milk, Soy Milk	Ice cream with nuts, cheese with nuts or seeds
Vegetables Cooked only 3 or more servings per day	Vegetable juice without pulp, soft cooked vegetables: yellow squash without seeds, green beans, wax beans, spinach, pumpkin, eggplant, potatoes without skin, asparagus, beets, carrots	Vegetable juices with pulp, raw vegetables, cooked vegetables not on the recommended list
Fruits 2 or more servings per day	Applesauce, apples without skin, bananas, fruit juice without pulp, melons, canned fruit except pineapple	Fruit juice with pulp, canned pineapple, fresh fruit other than those on the recommended list, prunes, prune juice, dried fruit, jam, marmalade
Starches, Bread & Grain 4 or more servings per day	Bread, crackers and cereals made from refined flours, pasta and noodles made from white flours, white rice, pretzels, white potatoes without skin, white rice, dry cereals such as: Rice Krispies, Rice or Corn Chex, Corn Flakes, Kixx.	Whole-grain breads, whole grain cereals, brown and wild rice, whole grain pasta, bran cereal, oatmeal
Meat or Meat Substitutes 5 to 6 oz per day	Meat, poultry, eggs, seafood. Baked, broiled, grilled or boiled are preferred methods of cooking	Chunky peanut butter, nuts, dried beans and peas, fried and greasy meats, salami and cold cuts, hot dogs, meat substitutes
Fats and Oils	All oils, margarine, butter, mayonnaise, salad dressings that do not contain foods from the Avoid List	Coconut, nuts, and seeds
Sweets and Desserts	Jelly, Rice Dream Frozen Dessert, sugar, marshmallows, angel food cake	Desserts containing nuts, coconut, whole grains or dried fruits, jams and preserves
Miscellaneous	Salt, sugar, ground or flaked herbs and spices, vinegar, ketchup, mustard, and soy sauce	Popcorn, pickles, horseradish, relish, jams and preserves

Phase I Sample Menus

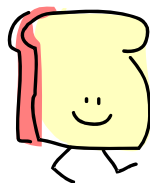
Meal	Day 1	Day 2
Breakfast	Rice Chex Cereal with Rice Milk Soft Boiled Egg White toast Jelly Tea	Grits- white Scrambled Eggs English Muffin Jelly Rice Milk
Lunch	Deli sliced turkey White Bread Applesauce Pretzels Rice Milk Sprite	Deli slice roast beef White Bread Applesauce Saltine crackers Rice Milk Sprite
Dinner	Baked chicken Ramen Noodles Banana Angle food cake Rice Milk Tea	Baked fish White Rice Peeled Apple Vanilla Wafers Rice Milk Sprite



Repeat Days 1 and 2 as needed to control diarrhea. Choose "single ingredient" food that your child wants most to increase variety in the diet. To increase calories try adding butter or margarine to bread, rice or potatoes as a new food. To improve nutritional content of the diet add new fruits and vegetables. If a food is not tolerated then do not try it again for a while. This does not mean it is permanently off the list; you may go back and try it again later if it is a highly desired food.

Adding to Phase II Diet: adding fats

Meal	Day 1	Day 2
Breakfast	Bagel with butter & jelly Scrambled Eggs Rice Milk Tea	Cream of Rice Cereal & butter Poached Egg White toast & butter & jelly Rice Milk Tea
Lunch	Deli sliced turkey White Bread Mayonnaise Banana Pretzels Rice Milk Sprite	Deli slice roast beef White Bread Mayonnaise Applesauce Saltine crackers Rice Milk Sprite
Dinner	Roast Beef Boiled Potatoes & butter Applesauce White Bread & butter Rice Milk Sprite	Baked Chicken Ramen Noodles Peeled apple Vanilla Wafers Rice Milk Sprite



Functions of Fiber:

Soluble: prolong stomach emptying time so that sugar is released and absorbed more slowly, forms a gel when mixed with liquid which helps to soften stool

Insoluble: moves bulk through the intestines, promotes regular bowel movements, and helps prevent constipation

- If your child is taking laxatives soluble fiber will be help to bulk the stool
- If your child is not on laxatives and is trying to avoid constipation encourage sources of insoluble fiber
- Keep in mind most foods contain a combination of both soluble and insoluble fiber

Foods High In Insoluble Fiber

Includes hemicellulose, cellulose and lignin.

- | | | |
|--------------------------|------------------------|----------------------|
| ▪ Cauliflower | ▪ Pinto beans | ▪ Bulgar |
| ▪ Spinach, raw | ▪ Lima beans | ▪ Popcorn |
| ▪ Cabbage, green | ▪ Kidney beans | ▪ Corn tortilla |
| ▪ Kernal Corn | ▪ Strawberries | ▪ Whole wheat breads |
| ▪ Broccoli, raw | ▪ Blueberries | ▪ Whole wheat bagel |
| ▪ Avocado | ▪ Cranberries | ▪ Wheat cereals: |
| ▪ Carrots | ▪ Cherries | Raisin Bran, |
| ▪ Cucumbers | ▪ Dates | Bran flakes |
| ▪ Tomatoes | ▪ Prunes | ▪ Rye |
| ▪ Green Peas | ▪ Prune juice | ▪ Barley |
| ▪ Baked potato with skin | ▪ Raisins | ▪ Millet |
| ▪ Green beans | ▪ Apricots | ▪ Almonds |
| ▪ Split peas | ▪ Pears | ▪ Sesame seeds |
| ▪ Chick peas | ▪ Guava | ▪ Brazil nuts |
| ▪ Lentils | ▪ Fresh pineapple | |
| ▪ Northern Beans | ▪ Chunky Peanut butter | |

Foods High in Soluble Fiber

Includes pectin, beta-glucans, fructans, guar and gums, psyllium

- Oatmeal
- Oat bran
- High fiber pastas
- Corn meal
- Yams
- Sweet potatoes
- Artichokes
- Rutabagas
- Parsnips
- Squash
- Mushrooms
- Chestnut
- Applesauce
- Blackberries
- Papayas

Supplements:

Name	Active Ingredient	Serving Size	Amount of Fiber
Metamucil Wafers	Psyllium 50% soluble	2 wafers	6 grams
Metamucil Powder	Psyllim 65% soluble	1 tbsp	3 grams
Ground Flax Seed	45% insoluble, 55% soluble	1 tbsp	3 grams
Benefiber	Wheat dextrin 100% soluble	2 tsp	3 grams
Citrucil	Methocellulose 100% soluble	1 scoop or 4 caplets	2 grams
Pectin	100% soluble	1.75oz package	4.3 grams

Sources:

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Nutrition Therapy for Patients with Anorectal Malformations

Allison Blicher RD, LD

Mary Pat Alfaro MS, RD, LD

Food and nutrition not only impact the growth of a child but also influence the developmental milestones a child achieves. Specific nutrition guidelines have been designed to improve the quality of life of children who are born with an ARM and can have a major impact. A dietitian's role with a patient who is born with an ARM depends on the patient's specific malformation and their overall prognosis for bowel control. Based on this information, nutrition intervention may be as simple as adding a few foods to the diet to avoid constipation or as dramatic as avoiding multiple foods to prevent diarrhea.

Patients are divided into two main categories, those with potential for bowel control and those with fecal incontinence. Nutrition recommendations are developed and advised based on each individual's case. Many patients who are born with an ARM do not have bowel control and suffer from fecal incontinence. These patients need Bowel Management, which is an artificial way to keep them clean. They usually benefit from some form of nutritional intervention.

CASE STUDY ONE:

5 year old female, born with an anorectal malformation, incontinent of stool, has no potential for bowel control, on bowel management receiving a daily enema to artificially remain clean.

Children who have no chance of bowel control with a tendency toward firm stool and receive a daily enema, do not require any specific nutrition modifications. If the child is successful at staying clean, (no accidents in-between enemas) the patient can eat whatever they desire and are counseled to eat an age appropriate healthy diet.

CASE STUDY TWO:

4 year old male, born with an anorectal malformation, incontinent of stool, no potential for bowel control with a tendency towards loose stools.

A small percentage of children who are born with an ARM have a tendency towards diarrhea and have a hyperactive colon. Nutrition modification for this group includes a strict diet along with medication to help slow down the intestines. The constipating diet is designed to decrease the transit time of food through the digestive tract. The goal is to slow the colon down so much that only the 24 hour enema empties the accumulated stool. The diet is very restrictive in the beginning but over time new foods can be carefully introduced. If there is a food the child is craving then parents are encouraged to introduce that food for three days and observe closely what changes occur with bowel movements. If the child soils after eating a newly introduced food, then that food should be eliminated from the diet. The process of liberalizing the diet is a continuous trial of adding new foods and observing stool consistency changes. Not everyone has the same reaction to the same foods. Learning to identify the foods that control an individual's bowels is helped by journaling to record the effect of a food on bowel control. Due to the limited food choices in Phase I of the Constipating Diet, a daily multivitamin with mineral supplement is recommended. [Table 1]

Table I: Phase I Constipating Diet

Food Groups	Food Recommended	Food to Avoid
Milk and Milk Products	Rice Milk	All others
Vegetables	None	All
Fruits	Applesauce, Apples without skin, bananas	All others
Starches, Bread & Grain	Bread, crackers and cereals made from refined flours, pasta and noodles made from white flours, white rice, pretzels, white potatoes without skin, dry cereals such as: Rice Krispies, Rice or Corn Chex, Corn Flakes, Kixx.	All others
Meat or Meat Substitutes	Baked, broiled, boiled or grilled meat, poultry or fish	All others
Fats and Oils	Limit amounts of butter, margarine and oils in food preparation during this phase, non-stick spray is allowed	All others
Sweets and Desserts	Made from allowed ingredients, plain cake, gelatin or popsicles, Rice Dream Frozen Dessert and limit amounts of concentrated sweets such as jelly and marshmallows	All others

The foods listed above are recommended in Phase I of the constipating diet. Once the child remains clean on this diet a new food can be added and tried for three days to observe the effect of that food.

CASE STUDY THREE:

4 year old male born with anorectal malformation with potential for bowel control, in need of laxatives to treat constipation.

Many patients present with fecal incontinence. Some of those patient once their constipation is adequately treated they become continent and can have voluntary bowel movements. To find these patients when we suspect this type of “pseudoincontinence,” we perform a laxative trial. Patients with a good prognosis original anorectal defect, with a good sacrum, and good muscles fall into this category. The patient is instructed to implement a high fiber/laxative diet in addition to the daily laxative medication. The high fiber portion of the diet uses a guideline for a daily total number gram of fiber equal to age plus 10. For example if the child is 5 years old, the grams of fiber recommended per day would be 15.

There are two types of fiber; water-soluble and water insoluble. Water-soluble fiber prolongs stomach-emptying time. Sugars consumed are released and absorbed more slowly, forming a gel when mixed with liquids which helps to soften stool in the colon. Water insoluble fiber moves bulk through the intestines. This promotes regular bowel movements and helps to prevent constipation. If your child is taking laxatives, a combination of soluble and insoluble fiber will bulk and soften the stool. This is an ideal combination and allows the laxative to be more effective.

In addition to incorporating foods that are high in fiber we also recommend including foods that produce a “laxative effect.” [Table II] Keep in mind that each patient has their own unique digestive tract. Foods that may function as a laxative for some patients may cause constipation for others. Foods that may produce a laxative effect are high fat, highly spiced and high in caffeine. The colon is stimulated to move after eating, so it is recommended to eat only three meals a day and eliminate snacking. A diet with similar foods eaten at about the same time each day with approximately the same amount at each meal day after day will help promote regularity. Fluids are allowed in-between meals. To help toddlers follow this regimen, we suggest including a good source of protein and fat at each meal to prevent hunger pangs in-between meals.

If your child is a “picky eater,” there are alternatives to achieving fiber goals in the diet. Fiber supplements, flax seeds, and pectin can be added to foods that children like in order to increase the fiber content of the diet. Families already following a high fiber diet or who have tried this in the past meet with a dietitian to provide new ideas to achieve more variety at meal times and increase the fiber content of the meals. Using high fiber versions of “Kid Friendly” foods enhance acceptance of the diet long term. [Table III] As children get older, frequent education helps them understand how their food choices affect bowel function. Incorporating diet principals into lifestyles allows for freedom and flexibility and long term success.

Table II: Foods that may produce a “laxative” effect:

Age	Food
0-introduction of solids	Breastmilk
4-6 months	-Oatmeal or mixed grain cereal -Baby prunes
6-8 months	-Foods listed above -Fruit and vegetable baby foods -Prune juice -Apple juice
8-12 months	-Foods listed above -Finely chopped fresh or cooked vegetables: spinach, carrots, cabbage, broccoli, peas, sweet potato, corn, green beans, cauliflower, etc. -Finely chopped fresh fruits: cherries, grapes, pineapple, strawberries, avocado, mango, papayas, plums, apricots, peaches, pears, raspberries, blueberries, oranges, etc. -Finely chopped pieces of dried fruit: raisins, apricots, prunes, dates and figs -High fat dairy products: known to produce a laxative effect for some and constipation for others -High-fat foods: may function as a laxative food for some and a constipating food for others.
1 year and older	-Foods listed above -Dark chocolate, spicy foods and caffeine may be introduced if and when parents feel it is age appropriate.

Table III: High Fiber Kid Friendly Foods (note serving size may vary depending on age)

Type of Food	Serving Size	Grams of Fiber per serving
Multigrain cheerios	½ cup	1.5
Sliced fresh fruit with yogurt dip	1 cup	~3
Cut up fresh vegetables with veggie dip	1 cup	~3
Homemade trail mix:	¾ cup whole wheat chex small box raisins 1oz peanuts	9
Sun chips	11 chips (1oz)	2
Popcorn	2 cups	2.3
Whole grain wheat thins	17 crackers	2

Fiber Supplements:

Type of Supplement	Grams of Fiber	Serving Size
Benefiber	3	1 tbsp
Metamucil	3	1 tbsp
Ground flax seeds	1.9	1 tbsp
Whole flax seeds	2.8	1 tbsp

CASE STUDY FOUR:

Three month old female born with an anorectal malformation recently had her pull through and then her colostomy closure.

Infants that are born with anorectal malformations are encouraged to follow a “laxative diet” until they are ready to attempt potty training. Ideally, this means one to two well formed but soft bowel movements per day. If possible, mothers should breast feed their babies due to breast milk’s natural laxative effect. Once the infant is ready to start solids, between four and six months of age parents should introduce oatmeal cereal instead of rice cereal. Oatmeal has more fiber, and rice cereal may cause constipation. Parents can also add baby prunes to the oatmeal or offer mixed grain cereal mixed with fruit. Parents can give baby prune juice as well as apple and grape juice to help keep things moving. Between 6-8 months of age the parent can start fruits and vegetables. Making strained baby foods increases the quantity of fiber the infant will receive. Otherwise baby food fruits and vegetables with the highest quantity of fiber listed are best. Introduce one food per week to assess for food allergies. As the infant grows and starts trying solids, offering high fiber choices will help mold the child’s taste buds. Foods that should be avoided during early infancy and the toddler stage are any white starches, and bananas due to their reputation for causing constipation. A lot of parents ask the question, “Will my child become constipated once they transition to whole milk from formula?” The answer is: everyone is different. At this age toddlers are used to drinking the majority of their calories and are still in the process of trying new textures and tastes. Some children that drink excessive amounts of milk with minimal food intake may become constipated. To avoid this, provide enough milk in combination with other dairy products to meet the calcium needs for age [Table IV and V]. Keep in mind that large amounts of dairy products may constipate some children and have no effect on others.

Table IV: Calcium Recommendations

0-6 months	210 mg/day
7-12 months	270 mg /day
Ages 1-3	500 mg/day
Ages 4-8	800 mg/day
Boys and Girls Ages 9-18	1,300 mg/day

Table V: Which Foods are Good Sources of Calcium?

Food & Serving Size	Calcium (mg)	Food & Serving Size	Calcium (mg)
Yogurt, nonfat plain, 1 cup	400	Almonds, ¼ cup	94
Milk, skim, 1 cup	302	Bok Choy, ½ cup, cooked	79
Ricotta, part skim, ½ cup	337	Turnip greens, ½ cup, cooked	99
Swiss, 1 oz.	272	Orange, 1 medium	56
Mozzarella, part skim, 1 oz.	183	Collard greens, ½ cup, cooked	178
Macaroni and Cheese, ½ cup	180	Kale ½ cup cooked	90
Cheese Pizza, 1 slice	220	Broccoli, ½ cup, cooked	36
Canned sardines, w/bones, 3 oz.	330	Tofu, firm (calcium set), ½ cup	258
Canned salmon, w/bones, 3 oz.	181	Black-eyed peas, 1 cup	212
Calcium enriched orange juice, 1 cup	200	Navy beans, 1 cup	128
Calcium enriched soy milk, 1 cup	300		



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Urinary Reconstruction (Rather Than Diversion) for Continence in Difficult Pediatric Urologic Disorders

By Curtis A. Sheldon
Cincinnati, Ohio

Surgical reconstruction of the pediatric urinary tract for purposes of restoration of function is a complex and highly successful undertaking that holds the potential of dramatically enhancing a child's health and quality of life. Unfortunately, such reconstruction may carry with it adverse sequelae that may complicate subsequent therapy and may be difficult to reverse completely. Urinary tract function has several components, each of which are essential to ensure maximal renal preservation, freedom from clinically significant infection, and continence. Several surgical alternatives exist for each component. Representative alternatives and surgical judgment with respect to their application are reviewed.

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RECENT ADVANCES in surgical technique, the successful application of intermittent catheterization to the reconstructed urinary tract, and the lessons learned from the pioneering work on urinary undiversion¹ have allowed even the most anatomically devastated children to be reconstructed for continence as well as for preservation of renal function. Such reconstructive principles may now be applied to virtually all urinary tract anomalies with a good expectation for success. Reconstructive options are presently available even for those children with end-stage renal disease for whom renal transplantation will ultimately be required.

Urinary diversion, once popularized as the solution for many functional and structural urinary tract diseases, has now fallen into disfavor. In addition to an unacceptable effect of a urine collection bag on the body image of the child, an unanticipated high incidence of upper tract deterioration has been experienced. Loss of renal function from infection, obstruction, and urolithiasis has occurred at an alarming incidence. Furthermore, metabolic derangements such as acidosis and alterations in calcium homeostasis resulting in an adverse influence on bone growth and repair have been documented. Additionally, certain types of diversion appear particularly prone to malignant degeneration.

The goals of reconstructive surgery begin with the

attainment of a large capacity, low pressure reservoir. This not only is critical to the achievement of continence but also has, most importantly, been demonstrated to provide a protective influence on the upper urinary tract. Adequate bladder outlet resistance must be provided to prevent incontinence. If possible, the potential for spontaneous voiding is not sacrificed. Access for quick, easy, and painless catheterization is critical because failure of compliance with this modality when needed may undermine even the most successfully reconstructed urinary tract.

Those patients who are catheter-dependent should, if possible, have a "pop-off" mechanism whereby urine will ultimately leak out their urethra if they become unable or unwilling to catheterize. This is helpful in preventing the potentially devastating complication of bladder rupture that may otherwise occur. Finally, whenever possible, reconstruction should be performed with an effort directed at maintaining freedom from infection in both the upper tracts and the urinary bladder.

Those patients who have end-stage renal disease should, when possible, be reconstructed with techniques that are not at risk of precipitating acidosis, that minimize interference with effective dialysis and facilitate implantation of the allograft ureter on transplantation.² Experience with reconstruction of complex congenital anomalies such as urogenital sinus and cloacal anomalies clearly has demonstrated that the best outcome is associated with concomitant correction of urethral, vaginal, and anorectal anomalies when these anomalies coexist.^{3,4}

Before being considered for reconstruction, all children who are anatomically suitable undergo a vigorous trial of pharmacological therapy and intermittent catheterization. Only when failure of nonoperative modalities has been demonstrated is reconstruction entertained. Preoperative investigation includes measurement of 24-hour urinary output, intravenous urography (unless uremic or allergic), voiding cystourethrography including upright film of the bladder outlet, and detailed urodynamic investigation. Regardless of the reconstructive procedures anticipated, a full preoperative bowel preparation is performed because the surgeon must be prepared to respond to any of a number of unexpected anatomic findings that may be encountered.

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BLADDER OUTLET RECONSTRUCTION

Because a high pressure bladder requiring augmentation may alone be responsible for urinary incontinence, the need for bladder outlet reconstruction may be difficult to ascertain in this setting. Both urodynamic and radiographic findings such as leak point pressure, stress leak point pressure, urethral pressure profilometry, and upright cystography may indicate the need for bladder outlet obstruction.^{5,6,7}

Several excellent options for bladder outlet reconstruction exist. Several of the most commonly used are outlined in Table 1. The Young-Dees-Leadbetter bladder outlet procedure is demonstrated in Fig 1. This highly versatile procedure has been extensively applied to the reconstruction of the exstrophy patient but also is applicable to the iatrogenically deformed bladder outlet or the neuropathically incompetent bladder outlet associated with such entities as myelomeningocele or imperforate anus.⁸

The effectiveness of this procedure is enhanced by fashioning a long narrow mucosal tube. Generally, tubularization is performed over an 8F or 10F urethral catheter. Traditionally, the goals of this procedure are to attain continence, allow spontaneous voiding if the bladder is capable of contraction, and permit intermittent catheterization access if it is not. However, these goals are potentially divergent in that

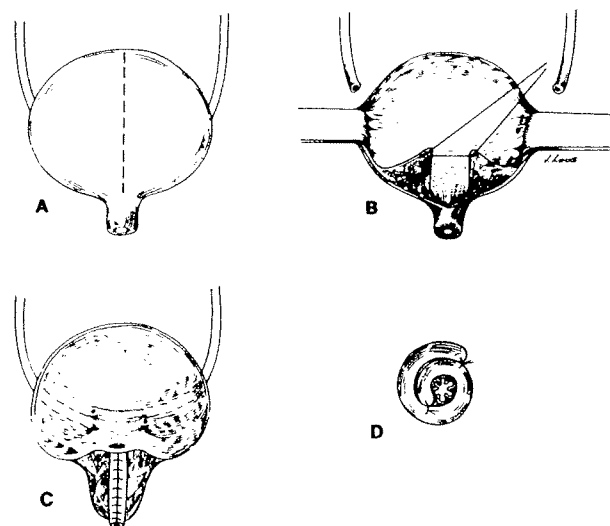


Fig 1. Young-Dees-Leadbetter bladder neck reconstruction. (A) The bladder is incised in the midline. Often a small "T" incision is created distally to enhance exposure of the proximal urethra. (B) Mucosal triangles are excised leaving a strip of bladder mucosa of approximately 10 to 12 mm in width and 3.5 to 4 cm in length. The ureters are detached from the bladder. (C) A series of incisions along the lateral aspect of the denuded detrusor relax and lengthen the neourethra. The edges of the mucosal strip are approximated over an 8F or 10F catheter. The ureters are reimplanted using a cross-trigonal technique. (D) The neourethra is reinforced by wrapping the mucosal tube with the denuded segment of detrusor. Not shown is the anterior suspension of the reconstructed urethra by either suture approximation to the rectus fascia or by placement of a fascial sling. From Sheldon CA, Bukowski T: Bladder function, in Rowe MI, O'Neill JA, Grosfeld JL, et al (eds): *Essentials of Pediatric Surgery*. St Louis, MO, Mosby Year Book, 1995, pp 731-747.

Table 1. Bladder Neck Reconstruction

Technique	Pros	Cons
Young-Dees-Leadbetter	Allows spontaneous voiding Uses native tissue Allows "pop-off"	Consumes bladder capacity May be difficult to catheterize Catheterization may injure
Kropp	Very reliable continence Uses native tissue	Difficulty with catheterization No "pop-off" Eliminates spontaneous voiding Consumes bladder capacity
Fascial sling	Technically fast and simple Easily reversible Allows spontaneous voiding Can be used as adjunct to Young-Dees-Leadbetter Allows "pop-off"	Excessive angulation may interfere with spontaneous voiding or catheterization
Artificial urinary sphincter	Allows spontaneous voiding	Erosion Infection Mechanical failures
Urethral closure	Definitive continence Technically simple	Increased risk of calculi Difficult to reverse

while a long narrow tube suspended anteriorly provides an excellent opportunity for continence and voiding, catheterization may be difficult and even injurious to the delicate continence mechanism. The provision of an alternate route for catheterization such as an adjuvant Mitrofanoff neurourethra (see below) prevents compromise and is easily reversible as an outpatient procedure if not needed.

The Kropp procedure (Fig 2) involves the creation of a tubular segment of detrusor attached to the urethra, which is then implanted submucosally into the bladder to create a one-way valve.⁹ The prospect for continence is excellent; however, catheterization may prove extremely difficult in some instances. The potential for spontaneous voiding is eliminated. This procedure has been applied most frequently to neurogenic incontinence.

Numerous variations have been reported on the fascial sling and have been recently reviewed in detail.¹⁰ Depicted in Fig 3 is one approach where the fascial sling is based on the pyramidalis and passed under the urethra. This has proved to be effective and is easily adapted to almost any major bladder reconstruction; it is an excellent adjunctive procedure to

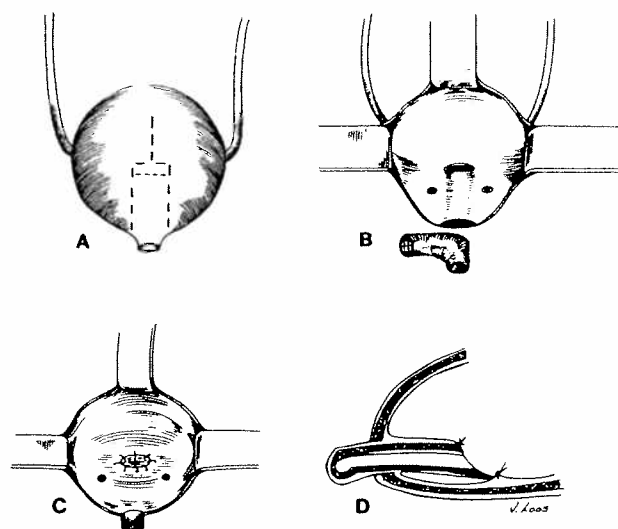


Fig 2. Kropp bladder neck reconstruction. (A) Bladder incision creates the urethral-based bladder flap. (B) The bladder flap is tubularized to create the neourethra; the submucosal tunnel is created in the bladder base. (C) Neourethral anastomosis is completed. (D) Functional result—increasing bladder pressure compresses and occludes the neourethra, producing continence. From Sheldon CA, Bukowski T: Bladder function, in Rowe MI, O'Neill JA, Grosfeld JL, et al (eds): *Essentials of Pediatric Surgery*. St Louis, MO, Mosby Year Book, 1995, pp 731-747.

the Young-Dees-Leadbetter procedure to secure anterior suspension. Care must be taken to avoid excessive angulation if either spontaneous voiding or intermittent urethral catheterization is anticipated.

The artificial urinary sphincter (Fig 4) is particularly well-suited to the patient with isolated bladder outlet incompetence whose bladder provides good capacitance and compliance. Although its use is

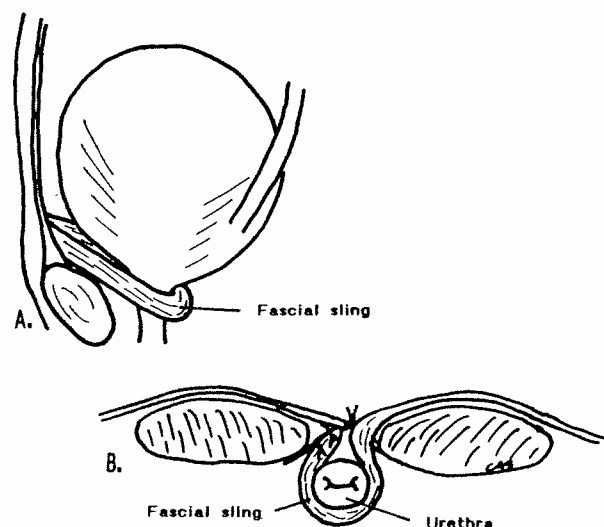


Fig 3. Fascial sling. (A) A 1-cm wide fascial strip is developed along the edge of the midline incision, based on the pyramidalis muscle. The sling is passed under the urethra, and the free edge is approximated to that contralateral edge of rectus fascia, creating anterior urethral suspension. (B) Appearance of sling in cross-section as it is passed posteriorly to the urethra.

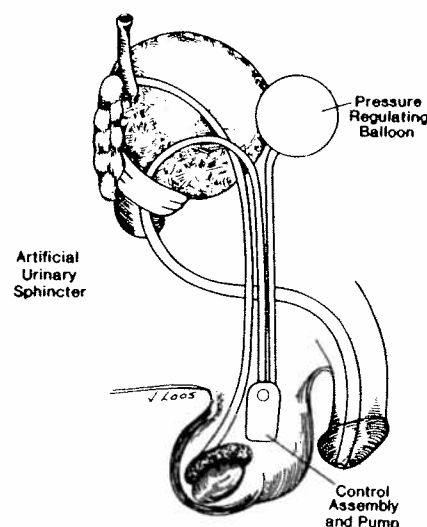


Fig 4. The artificial urinary sphincter. The cuff is positioned around the bladder neck, avoiding compression of the ureters. The cuff is connected to a control assembly, and the pump is positioned in the scrotum or labia majora that in turn is connected to a pressure-regulating balloon positioned so that it is exposed to intraperitoneal pressure. Fluid pressure from the pressure-regulating balloon applies sufficient pressure to occlude the bladder outlet without compromising blood flow. Increased intravesical pressure from movement, sneezing, or coughing is immediately met by the simultaneous application of increased cuff pressure caused by intraperitoneal pressure applied to the balloon. When the patient desires to void, the pump is activated, resulting in the transfer of fluid from the cuff, allowing the bladder outlet to open. The spontaneous redistribution of fluid back to the cuff via the control assembly results in resumption of bladder outlet competence. From Sheldon CA, Bukowski T: Bladder function, in Rowe MI, O'Neill JA, Grosfeld JL, et al (eds): *Essentials of Pediatric Surgery*. St Louis, MO, Mosby Year Book, 1995, pp 731-747.

hampered by its inherent risk of infection, erosion, and mechanical failure, excellent long-term results may be achieved, even in those patients requiring extensive additional reconstructive surgery, including those who have undergone previous bladder outlet procedures.^{11,12,13}

Closure of the bladder outlet is effective in assuring urethral dryness. However, it does preclude both urethral catheter access and spontaneous voiding. It not only potentiates the risk of bladder calculi but also makes their treatment difficult by severely compromising endoscopic bladder access. This procedure generally is reserved for those instances in which other modalities have proved unsuccessful or inapplicable.

BLADDER AUGMENTATION

A variety of donor tissue sources is available for increasing bladder capacity and enhancing compliance (Table 2). The most frequently used augmentation technique is that of ileocystoplasty (IC) in which ileum is used to augment bladder volume (Fig 5). Like the sigmoid colocolostomy (SCC) and the ileocecolocolostomy (ICC), the application of this

Table 2. Augmentation Modalities

Technique	Pros	Cons
Ileocystoplasty	Technically simple	Acidosis, infection, mucous Antirefluxing implantation unreliable
Sigmoid colocystoplasty	Technically simple Allows implantation of ureter or Mitrofanoff into tenia	Risks interruption of blood supply to rectum after reconstruction for imperforate anus Acidosis, infection, mucous
Ileocecostoplasty	Technically simple Allows antirefluxing implantation of dilated ureters with intussusception technique	Intussusception antireflux mechanism inherently unstable Risks devastating fecal incontinence in patients with marginal continence mechanism (eg, myelomeningocele or VATER)
Gastrocystoplasty	Reduced risk of infection and mucous production Allows easy implantation of ureter or Mitrofanoff	Hematuria-dysuria
Ureterocystoplasty	No risk of acidosis Metabolically inert No mucous May be performed extraperitoneally	Antirefluxing implantation not possible Dilated ureter only occasionally available Risk of failure to attain adequate compliance and capacity
Autoaugmentation	Technically simple Metabolically inert No mucous May be performed extraperitoneally	Antirefluxing implantation not possible Risk of failure to attain adequate compliance and capacity

modality is made effective by the concept of detubularization.¹⁴ This interrupts circular smooth musculature, dramatically decreasing its ability to apply pressure with peristaltic activity. Combined with reconfiguration, detubularizing enhances both augmented capacitance and compliance. The ICC has the unique feature of allowing easy antirefluxing reconstruction for massively dilated ureters without tapering, using an intussusception antireflux mechanism. These procedures have in common the absorption of urinary solute risking the development of systemic acidosis and its attendant effect on bone metabolism and development. This effect is particularly problematic in the patient with renal insufficiency and has been extensively reviewed.¹⁰

Gastrocystoplasty (GC) involves the augmentation of the bladder with a segment of gastric fundus,

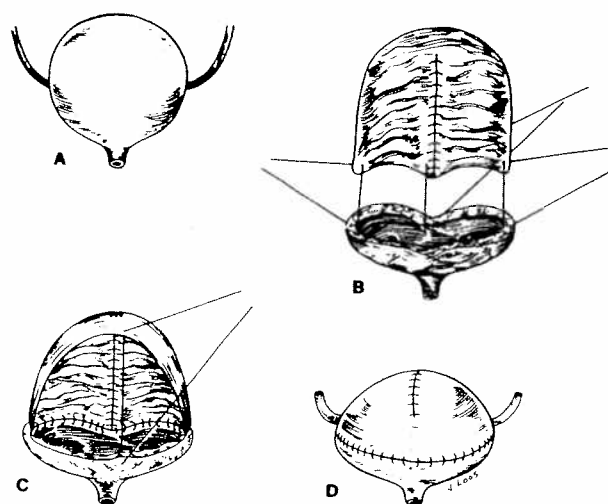


Fig 5. Bladder augmentation employing an intestinal segment. (A) The bladder is opened as a "clam shell." (B) The intestinal segment is detubularized by longitudinal incision along the antimesenteric border. A cup-patch is fashioned by suturing one edge of the resultant rectangle to itself. (C) The cup-patch is sutured to the remnant bladder plate. (D) Final appearance. From Sheldon CA, Bukowski T: Bladder function, in Rowe MI, O'Neill JA, Grosfeld JL, et al (eds): *Essentials of Pediatric Surgery*. St Louis, MO, Mosby Year Book, 1995, pp 731-747.

generally mobilized on a right gastroepiploic vascular pedicle (Fig 6).^{15,16} Like the ureterocystoplasty (UC), in which a dilated ureter is reconfigured to augment the bladder (Fig 7),¹⁷ and the autoaugmentation (AA), in which a large segment of detrusor is removed to enhance capacitance by creating a large area of unreinforced urothelium (Fig 8),¹⁸ the GC is

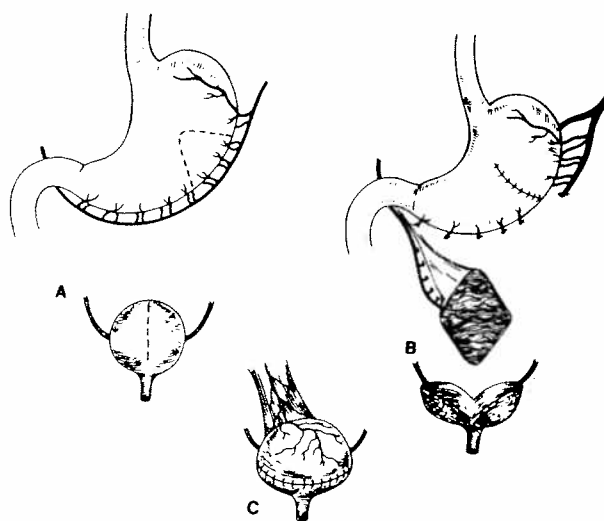


Fig 6. Bladder augmentation using a gastric segment: gastrocystoplasty. (A) A wedge of nonantral stomach is isolated, based on the right gastroepiploic vessels. (B) The bladder is prepared as a "clam shell." (C) The anastomosis is completed with a running double layer of absorbable suture. From Sheldon CA, Bukowski T: Bladder function, in Rowe MI, O'Neill JA, Grosfeld JL, et al (eds): *Essentials of Pediatric Surgery*. St Louis, MO, Mosby Year Book, 1995, pp 731-747.

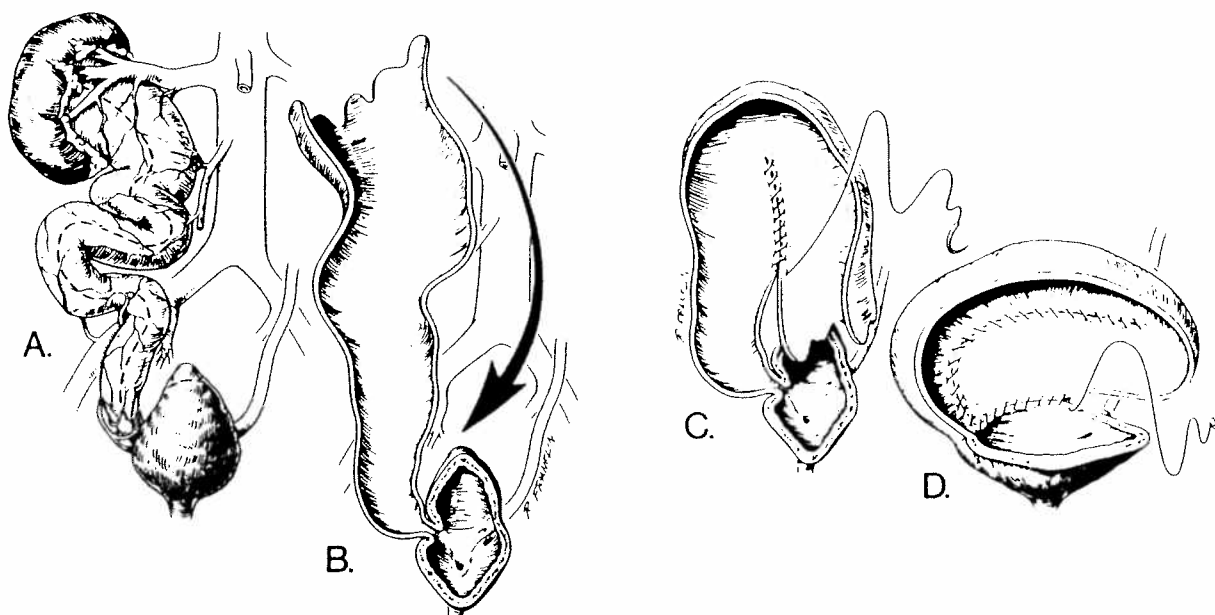


Fig 7. Ureteral bladder augmentation: ureterocystoplasty. (A) Normal blood supply to ureter. (B) Ureteral detubularization after mobilization. (C) Reconfiguration of ureter into "U-shaped" patch. (D) Ureteral patch is anastomosed to the bivalved native bladder. Reprinted with permission.¹⁷

well suited to patients with renal insufficiency. This is because urine solutes potentiating acidosis are not absorbed. Furthermore, there appears to be less risk of urolithiasis with GC, UC, and AA.

The UC and AA procedures are, in fact, totally metabolically inert because they use only urothelial

surfaces. However UC and AA are more likely to fail to attain adequate capacity and compliance because of an inherent restriction in surface area availability. In contrast, IC, SCC, ICC, and GC achieve inadequate reservoir parameters only if insufficient donor tissue is harvested. Of particular note is the fact that UC and AA can be performed retroperitoneally. This is of major benefit in the patient who has a ventricular-peritoneal shunt or is dependent on peritoneal dialysis.

Gastrocystoplasty carries a unique risk of peptic inflammation of the bladder outlet known as the hematuria-dysuria syndrome. This is generally controllable by hydration and occasionally by the use of histamine blockade or inhibition of gastric proton pump activity. The occurrence of this problem appears to be minimized by the avoidance of antral incorporation and ensuring a competent bladder neck because the urethra appears more vulnerable than does the native bladder remnant.

An additional consideration is the ability of the augment segment to allow implantation of a ureter (native or transplant) or a Mitrofanoff neourethra (see below). Such implantation is not possible in patients with UC and AA and is inherently difficult in patients with IC. SCC allows implantation along the course of the tenia. However, the resultant angulation of the ureter of Mitrofanoff may compromise outcome. In contrast, implantation into the GC segment is technically easy and versatile using tech-

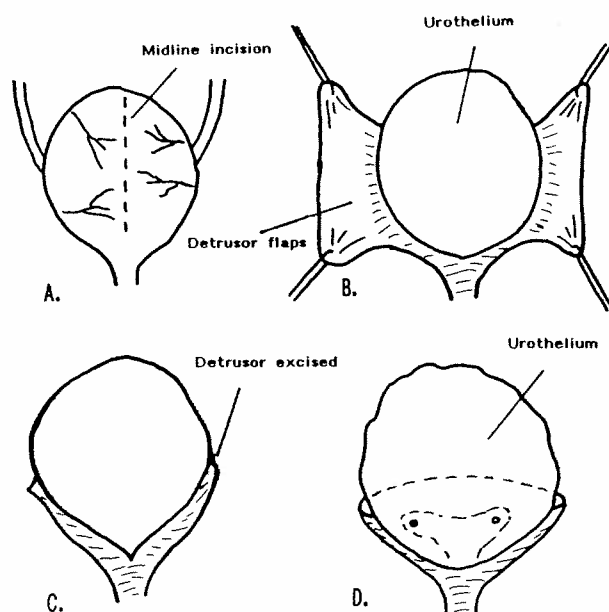


Fig 8. Autoaugmentation. (A) Midline incision in detrusor, leaving bladder mucosa intact. (B) Detrusor is dissected away from underlying urothelium. (C) Mobilized detrusor is excised. (D) Final appearance. Only a rim of detrusor surrounding the trigone remains. The remainder of the urothelium is devoid of its original muscular support.

niques that are directly analogous to those of implantation into native bladder itself.

It is imperative that the surgeon recognize the importance of the ileocecal valve in the attainment of adequate stool consistency to allow acceptable fecal continence in children with marginal continence mechanisms (eg, myelomeningocele or imperforate anus). The use of the ICC in such settings risks debilitating fecal incontinence and should be avoided whenever possible. Similarly, children with imperforate anus who have received anorectal reconstruction may have a rectal segment that is dependent on descending blood supply. Accordingly, the use of a SCC would appear unwarranted.

MITROFANOFF NEOURETHRA

The Mitrofanoff principle involves the creation of a continent catheterizable conduit that allows easy catheter access to the urinary bladder.¹⁹ As demonstrated in Fig 9, this involves the implantation of a tubularized visceral segment, (generally appendix, ureter, or tapered ileum) using a one-way flap valve mechanism into the urinary bladder with the exteriorization of the other end to the skin. Retrograde urine flow (incontinence) is prevented while antegrade passage of a catheter is readily accomplished. This technique is particularly well-suited to the patient who has difficulty with catheterization of the native urethra (urethra tortuous from reconstruction or congenital deformity, the patient who is restricted to a wheelchair, or the patient who has marginal control of upper extremities). Additionally, avoidance of the

pain associated with the catheterization of the difficult urethra may markedly enhance patient compliance.

Effective implantation has been demonstrated not only into native bladder but into colonic and gastric augment segments of the reconstructed bladder.²⁰ Further cutaneous exteriorization may be readily performed not to only the abdominal wall but also to the umbilicus and the perineum.²⁰ A recent review of 159 patients in six published series documented a 96% success rate with this powerful modality.¹⁰ The Mitrofanoff neourethra may be used either as a primary urethra, functioning as the only bladder outlet, or as an adjuvant neourethra facilitating safe bladder outlet reconstruction (see above). The relative merits of the most common Mitrofanoff segments used are outlined in Table 3.

Many conditions (eg, those associated with myelomeningocele and imperforate anus) have refractory fecal incontinence in addition to urinary incontinence. A patient dry of urine but in diapers because of fecal soilage is in no way a success. The incontinence of many such individuals is controllable by diet and by a variety of bowel regimens.¹⁰ However, many continue to soil because of a lax anus. Often, such individuals can be controlled by high-retention enemas administered by a balloon-sealed catheter. This often proves difficult because of an inability to achieve an effective seal or because of a latex allergy. When all other attempts prove fruitless, the antegrade continence enema administered via a catheter introduced into a continent cecostomy has proved to be

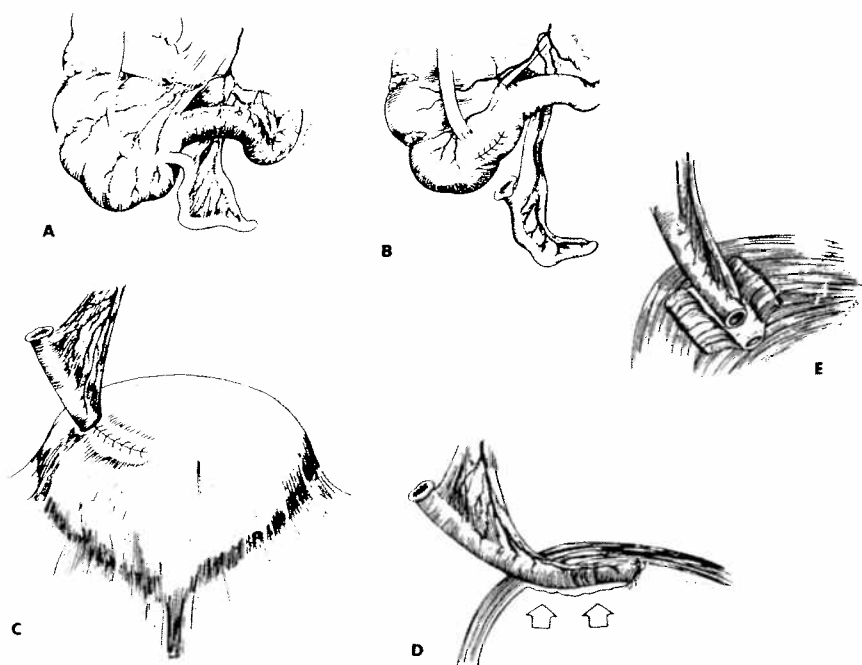


Fig 9. Appendiceal Mitrofanoff procedure. (A) Vascular anatomy of the appendix. (B) Appendix is divided from the cecum and mobilized on its vascular pedicle. (C,D) Appendix is implanted submucosally into the bladder. (E) Functional basis for Mitrofanoff continence. Reprinted with permission.²⁰

Table 3. Mitrofanoff Neourethra

Technique	Pros	Cons
Appendix	Technically simple	May be too short or absent
Tapered ileum	Excellent blood supply, length, and mobility	Mucous production Stone risk if stapled technique May be more difficult to catheterize
Ureter	May be performed retroperitoneally Technically simple if no reflux, and not massively dilated No mucous	Mucous production Increased risk of stomal stenosis Combination of need for reimplantation and tapering/tailoring may compromise blood supply

highly successful. A tubular visceral segment is implanted into the cecum in a fashion analogous to that of the Mitrofanoff procedure.²¹ Although most experience has involved the use of the appendix, I have found a short tapered ileal segment to suffice equally well (Fig 10).

SUMMARY

The current surgical armamentarium affords the surgeon an unprecedented opportunity to positively impact the life of a child who suffers from congenital or acquired, structural or functional, urinary pathology. Most require some combination of bladder outlet reconstruction, bladder augmentation, and creation of an alternate channel for catheterization.

However, such reconstruction comes with a price. Potential complications include urinary tract infection, urolithiasis, spontaneous bladder rupture, malignancy, loss of access for peritoneal dialysis, infection or dysfunction of ventricular-peritoneal (VP) shunts, and, rarely, disturbance of gastrointestinal function, metabolic derangements, and failure to achieve continence or upper tract stabilization.¹⁰

Of particular concern is the potential for subsequent surgery to cause injury to reconstructed organs or to their vascular pedicles. Most commonly, in my experience, such subsequent surgery takes the form of renal transplantation. Surgery for the acute abdomen (bladder perforation, small bowel obstruction,

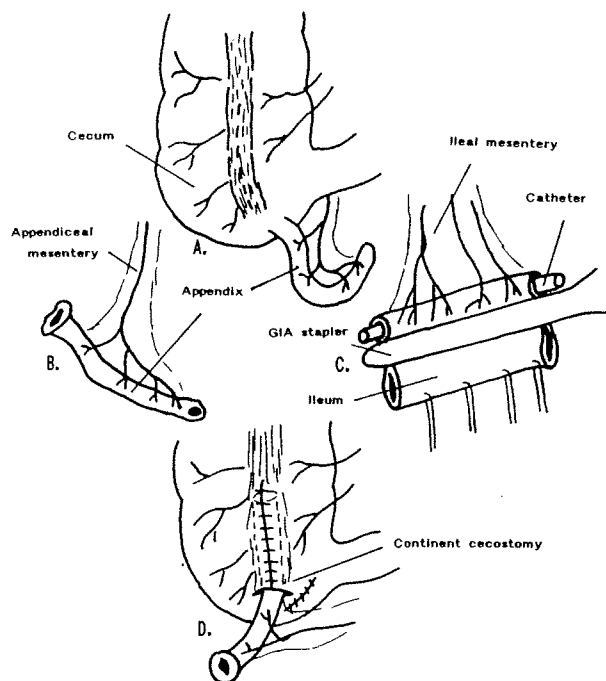


Fig 10. Continent cecostomy for performance of antegrade continence enema. (A) An incision is made along the anterior tinea of the cecum. (B) The appendix is divided from the cecum and mobilized on its blood supply. (C) Alternatively, a short segment of tapered ileum is prepared using a GIA stapler. Note that the creation of a tapered ileal segment for creation of a Mitrofanoff neourethra is performed with absorbable suture material to avoid the risk of urolithiasis. (D) The completed continent cecostomy.

or VP shunt complication) is also possible. Many reconstructed females will not be candidates for vaginal delivery and will require cesarean section.

Risk of a surgical disaster can be minimized or avoided by careful preparation for any procedure within or adjacent to the abdominal cavity. The surgeon should be familiar with the details of the preceding reconstruction, particularly the course of any vascular pedicles used. Whenever possible, a formal bowel preparation is performed. The patient is positioned, prepared, and draped so that the native urethra and the Mitrofanoff neourethra (if present) are accessible. Catheterization is performed on the field to allow both neourethral palpation and intermittent instillation of saline to facilitate identification of the boundaries of the reconstructed bladder.

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Occult Neurovesical Dysfunction in Children With Imperforate Anus and Its Variants

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● **Neurovesical dysfunction (NVD) is frequently encountered in children with imperforate anus and its variants. Such functional urologic problems are often compounded by associated anatomic urologic abnormalities that in combination may profoundly alter the course and prognosis of children with imperforate anus. Herein, we report 16 such cases. Management of NVD in children with imperforate anus offers several unique challenges that require important alterations in management. Specific recommendations are presented.**

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INDEX WORDS: Neurovesical dysfunction; anorectal anomalies; imperforate anus.

STRUCTURAL ANOMALIES of the urinary tract occur at high frequency in children with imperforate anus and account for significant morbidity.¹ However, functional abnormalities of the urinary tract (neurovesical dysfunction [NVD]) have remained poorly appreciated. In 1961 Duhamel² identified the concept of caudal regression. Variable degrees of anorectal, genitourinary, vertebral, and orthopedic anomalies are known to coexist in this population and may be associated with neurological deficit. More recently, occult spinal dysraphisms that include tethered cord, lipomas, neurenteric cysts, and diastematomyelia have been recognized as important associated anomalies in children with anorectal malformations.^{3,4} Although the natural history of such entities is incompletely known in children with imperforate anus, the tethered cord syndrome has otherwise been found to have significant urinary implications, to be potentially progressive, and may improve following surgical correction.⁵⁻¹⁰ Screening ultrasonography has proven useful in the diagnosis of occult spinal dysraphism in infants.¹¹⁻¹⁴

MATERIALS AND METHODS

A retrospective review of 90 patients with imperforate anus encountered between 1972 and 1987 was performed. Each case was evaluated for evidence of NVD. Patients were categorized into three groups according to the data available:

1. Clinically proven NVD. These patients either have their diagnosis proven by urodynamic investigation or have required intermittent catheterization (IC) for urinary retention in the absence of bladder outlet obstruction.
2. Presumed NVD on the basis of a documented tethered cord.
3. Presumed NVD on the basis of neonatal urinary retention in the absence of bladder outlet obstruction.

Patients were further classified according to the level of anorec-

tal deformity: (1) high imperforate anus; (2) low imperforate anus; (3) anal stenosis; (4) cloaca; or (5) cloacal exstrophy.

RESULTS

Table 1 outlines the characteristics of the patient population. Ages ranged from 1 to 11 years. The male-female ratio was equal. Patients presented with high imperforate anus (8), low imperforate anus (2), anal stenosis (1), cloacas (3), and patient cloacal exstrophy (1). Boys predominated in the high imperforate anus group; girls predominated in the remainder. Ten patients had clinically proven NVD either by urodynamic criteria or urinary retention requiring IC in the absence of bladder outlet obstruction. Two patients had NVD, presumed clinically on the basis of urinary retention compounded by urosepsis in the neonatal period. Both patients were managed with cutaneous vesicostomy and have thus not completed a urodynamic evaluation. Four patients were presumed to have NVD or to have been at risk for NVD on the basis of a documented tethered cord found as part of the screening protocol. Three of these infants are too young to allow accurate urodynamics and all have done well clinically. Within the entire group, only three patients had normal vertebral x-rays and in one patient, vertebral x-rays were not available for evaluation. Clearly, the majority of patients with NVD and imperforate anus will have abnormal vertebral radiographs; however, the presence of a normal radiograph does not exclude NVD. Six patients were documented to have tethered spinal cords and one patient was proven to have lumbar stenosis.

Seven of the 16 patients have required surgical intervention for their urinary tract and two have surgery scheduled. One patient has undergone augmentation, three patients had ureteral reimplantation, two patients had vesicostomy, one patient had cloacal exstrophy closure, and one of the patients who underwent reimplantation also received a Mitrofanoff neourethra. Two patients are scheduled for

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Table 1. Clinical Characteristics of 16 Patients With Imperforate Anus Variance With Proven or Strongly Suspected NVD

Case No.	Classification	Age (yr)/Sex	Group	Vertebral X-Ray Findings	Cord Abnormality	Urodynamics	Urologic Management
1	High	11/M	C	Narrow AP and transverse lumbar spinal canal, 6 lumbar vertebrae.	Lumbar stenosis	UDC, 75-mL cap, high UPP	Failed IC and drugs, awaits augmentation.
2	High	9/M	C	Normal	Tethered	—	IC
3	High	6/M	C	Sacral agenesis	—	—	Mitrof, IC, drugs
4	High	3/M	C	Absent S ₂ and S ₃ pedicles	Tethered	UDC, low compl	Awaits augmentation
5	High	4/M	C	Deformed S ₃₋₄ pedicle	—	UDC, 75-mL cap	IC, drugs
6	High	2/M	C	Abnormal S ₁	—	—	—
7	High	8/F	C	Normal	—	UDC, 50-mL cap	Augmentation, IC, drugs
8	High	2/M	PC	L ₄₋₅ abnormal	—	—	Vesicostomy
9	Low	1/F	Tethered	Normal	Tethered	—	—
10	Low	1/M	Tethered	—	Tethered	—	—
11	Low	5/F	C	S ₁ SBO	—	UDC, 75-mL cap, high UPP	Drugs
12	AS	5/F	Tethered	Partial sacral agenesis	Tethered	—	—
13	Cloaca	5/F	C	Partial sacral agenesis	—	—	Reimplantation, IC, drugs
14	Cloaca	7/F	C	Sacral dysgenesis	—	UDC	IC, drugs
15	Cloaca	2/F	PC	Hemivertebrae	—	—	Vesicostomy
16	CE	1/F	Tethered	Partial sacral agenesis	Tethered	—	Exstrophy closure

Abbreviations: AS, anal stenosis; CE, cloacal exstrophy; C, clinically proven; PC, clinically presumed; AP, anteroposterior; SBO, spina bifida occulta; UDC, uninhibited detrusor contractions; cap, capacity; UPP, urethral pressure profile; compl, bladder compliance; IC, intermittent catheterization.

bladder augmentation at the present time. Failure to recognize the significance of underlying NVD resulted in treatment failure of both patients who were managed by reimplantation. One patient was recovered with the institution of IC and anticholinergic drugs. The other patient required repeat ureteral reimplantation that was successful, with attention directed at appropriate management of the bladder. Two patients, now 8 and 11 years old, were never out of diapers and again had unrecognized NVD. One has been successfully managed with bladder augmentation and the second is scheduled for bladder augmentation. One patient in this series has end-stage renal disease that developed in part due to unawareness of his NVD associated with uninhibited detrusor contractions, low bladder compliance, and extremely high pressure storage. This child initially received vesicostomy with only a modest recovery of renal function and will require bladder augmentation and renal transplantation.

ILLUSTRATIVE CASES

Case 1

A 3-year-old boy born with hydronephrosis and imperforate anus had previously undergone bilateral ureteroneocystostomy. Intravenous urography (IVU) on presentation showed marked left hydronephrosis and cortical thinning. Renal scan showed delayed emptying on left. On voiding cystourethrography (VCUG) he was noted to have grade II vesicoureteral reflux (VUR) on the right and grade V VUR on the left. He had prominent uninhibited detrusor contractions (UDC) with intravesical pressures reaching 80 cm H₂O. His bladder capacity was 150 mL.

Treatment with anticholinergics and IC was instituted. Subsequent urodynamic studies showed no UDC and bladder capacity of 300 mL; an IVU showed marked improvement of left hydronephrosis and VCUG showed no reflux (Fig 1).

Comments. This case illustrates the important influence that the neurogenic bladder may have on management of VUR. It is important to realize that correct treatment of the neurogenic bladder, avoiding excessive intravesical pressures, will promote spontaneous resolution of reflux in such children. Equally important, failure to control excessive intravesical pressures in these patients is a common cause for failure of surgical correction of VUR. In this case, the bilateral ureteral neocystostomy was

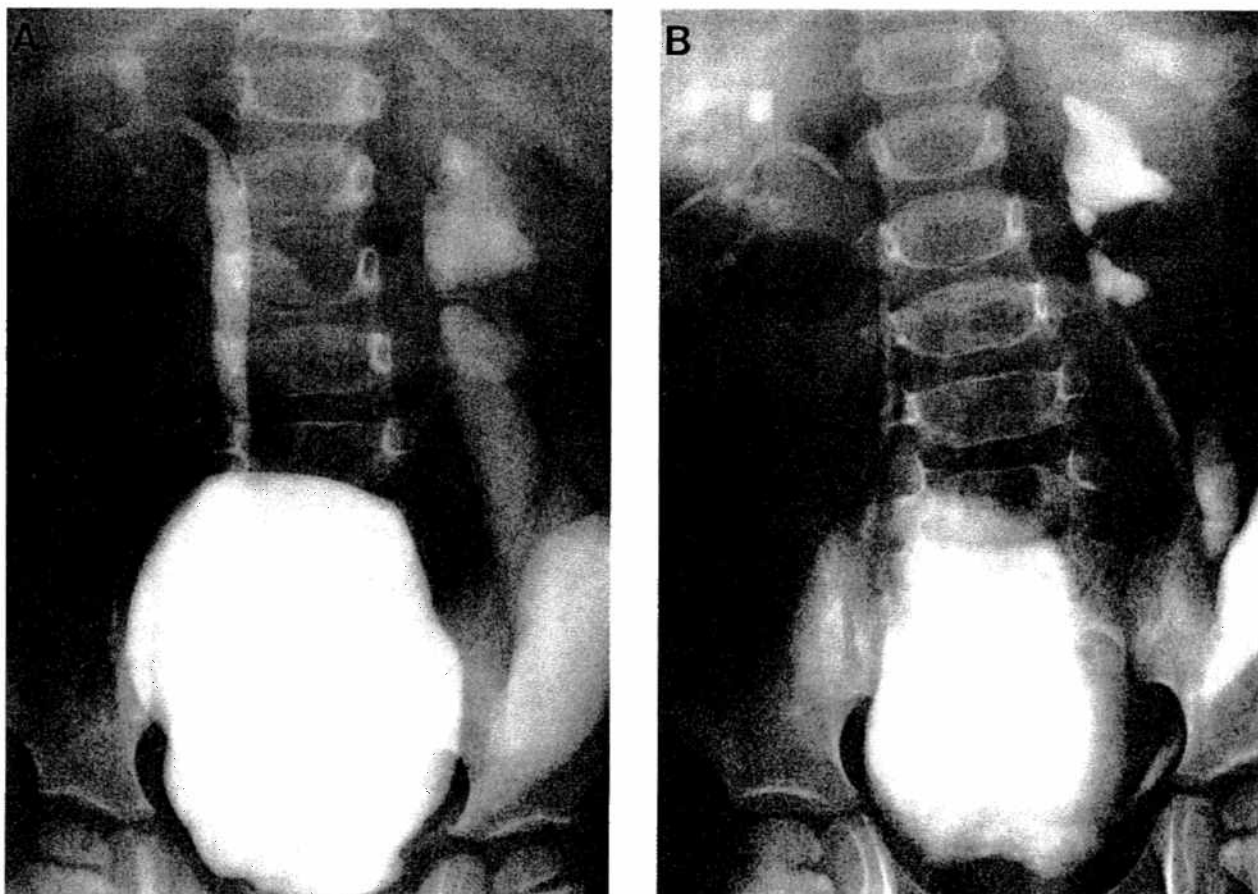


Fig 1. IVU of case 1. (A) The appearance of the upper tracts on presentation. At this point he had previously undergone bilateral ureteral neocystostomy but had failed due to lack of recognition of NVD. (B) The urograph taken after 4 months of anticholinergic medication and IC.

technically adequate but failed because of uninhibited detrusor activity. Once this was controlled, the reflux disappeared.

Case 2

A 5-year-old girl was born with a cloaca, ambiguous genitalia, and a duplicated left collecting system. She presented after having undergone bilateral ureteroneocystostomies with tapering of the left system, and two previous anorectal reconstructions.

Problems included continuous fecal incontinence, recurrent urinary tract infection (UTI), NVD necessitating IC, and bilateral hydronephrosis with a poorly functioning left upper pole. Examination showed no external vaginal orifice and marked clitoromegaly. Renal scan showed reduced function on the right and obstruction of both moieties on the left. Pelvic magnetic resonance imaging (MRI) showed lack of musculature around the rectum.

She then underwent repeat bilateral ureteral reimplantation (left common sheath). One year later her lasix renogram was normal with attention directed at controlling her NVD with anticholinergics and IC. Diverting colostomy was performed followed by posterior sagittal rectovaginal reconstruction with urethral lengthening. Currently she has a widely patent introitus.

Comments. This case again demonstrates the importance of controlling the neurogenic bladder when reconstructing the urinary tract. Of greater importance in this case was the previous surgeons failure to reconstruct the vagina and the anus concomitantly in the face of her cloacal anomaly. The performance of the pull-through for imperforate anus in this setting significantly compromises the prospect for future vaginal reconstruction.

Case 3

A 5-year-old boy was born with high imperforate anus, caudal agenesis, congenital urethral stricture, absent left kidney, and right megaureter (Fig 2). He had previously undergone urethroplasty, numerous dilatations, and right ureteral reimplantation.

Initial evaluation included cystourethroscopy showing a severely tortuous urethra that was extremely difficult to catheterize, along with a blind-ending ectopic left ureter draining into the proximal urethra.

He underwent posterior sagittal anoplasty, mobilization of the ectopic left ureter on its blood supply, and creation of a nonrefluxing Mitrofanoff conduit using this left ureter. Postoperatively he is totally continent to bladder volumes of 250 mL with an easily catheterizable conduit. Renal function is normal.

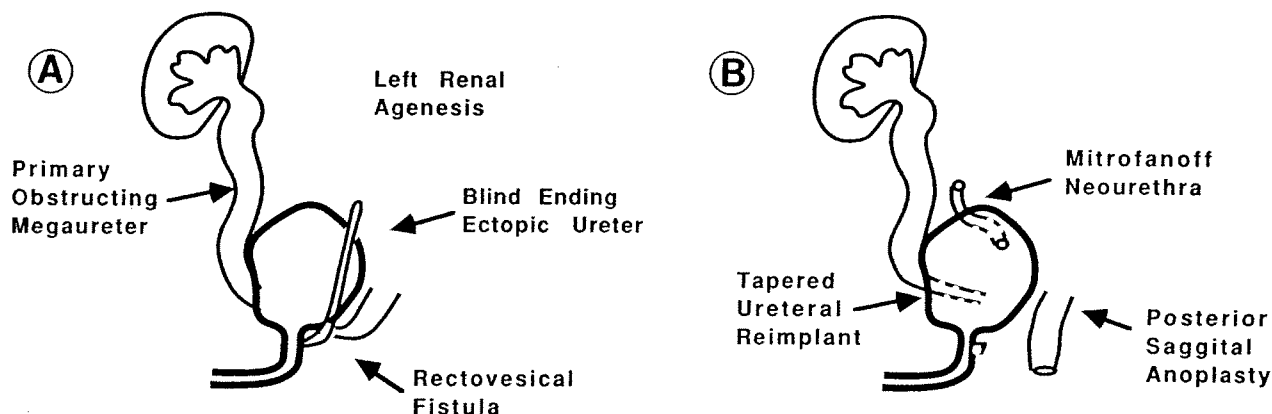


Fig 2. The (A) presenting and (B) reconstructed anatomy of case 3. A tapered ureteral reimplantation for primary obstructing ureter of a solitary right kidney was initially performed. A posterior sagittal anoplasty with division of rectovesical fistula was performed for high imperforate anus. A blind-ending ectopic ureter was mobilized on its blood supply and reimplanted into the bladder to create a continent catheterizable neourethra in this patient, who was very difficult to catheterize transurethrally.

Comments. The neurogenic bladder associated with imperforate anus may be especially difficult to manage secondary to the patient's inability to catheterize himself through the urethra due to angulation and tortuosity of this structure. Patients such as this who are dependent on IC can have an alternate abdominal neourethra created. A continent, catheterizable abdominal neourethra can be readily constructed from a ureteral segment, such as in this case, or the appendix. In either case an antirefluxing implantation of the conduit into the bladder is necessary.

Case 4

An 8-year-old girl was born with anorectal atresia and duplication of the vagina and uterus. She underwent fecal diversion and anoplasty at an early age. This patient presented with total urinary incontinence complicated by severe perineal candidal dermatitis.

Preoperative evaluation showed small bladder capacity (50 mL), severe UDC, and a normal urethral pressure profile (UPP). Her upper tracts were normal and grade I VUR was present on the right. Therapy with anticholinergics and IC was unsuccessful.

She underwent augmentation cystoplasty using jejunum (Fig 3). Postoperative functional bladder capacity is 120 mL. The patient is totally dry on a 2- to 3-hour voiding schedule along with anticholinergics and Lomotil and does not require catheterization.

Comments. This case illustrates the management of the child whose neurogenic bladder cannot be controlled by medication and IC and underscores the importance of urodynamic evaluation. Her urodynamic evaluation confirmed normal bladder outlet resistance. This predicted the success of bladder augmentation, which allows a marked increase in bladder capacity and a decrease in intravesical pressure. Note the fashion in which the augmentation was performed resulted in division of circular constricting muscular fibers throughout the entire length of the bowel segment. This not only geometrically promotes greater bladder capacity but significantly decreases any potential for the bowel segment to increase intravesical pressure due to its peristaltic activity.

Case 5

A 3-month-old boy had stigmata of VATER syndrome including imperforate anus, duodenal atresia, ventricular septal defect, and tricuspid atresia, as well as a multicystic kidney on the left. Surgical therapy had included a sigmoid loop colostomy. He had recurrent UTI in the nursery and at the time of initial evaluation had *Enterobacter* urosepsis and a VCUG showing poor bladder empty-

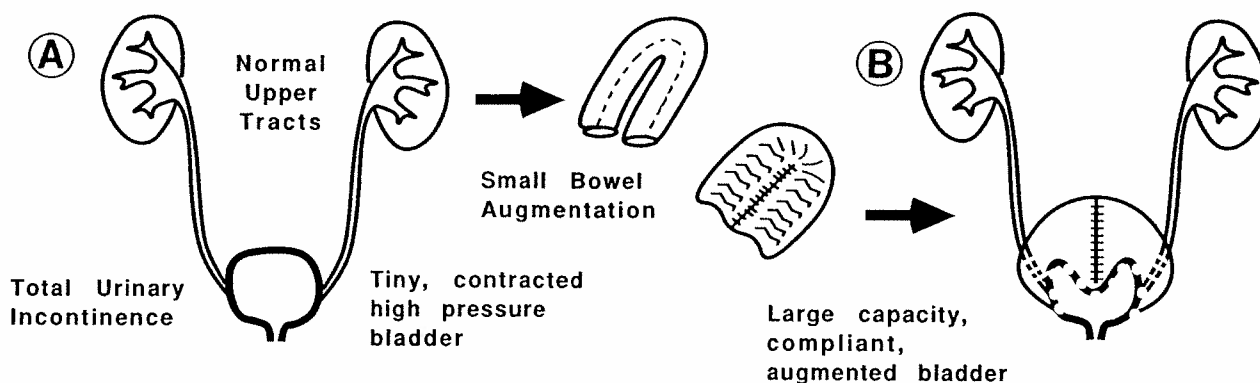


Fig 3. The (A) presenting and (B) reconstructed anatomy of case 4, who underwent small bowel augmentation for a small-volume, high-pressure bladder and achieved continence.

ing. He was treated with Foley drainage and antibiotics with improvement (Fig 4). When his cyanotic heart disease was stabilized he underwent vesicostomy and his colostomy was revised to a fully diverting configuration. His subsequent course has been stable.

Comments. This case illustrates the importance of modifying the management of imperforate anus in children with significant urinary disease. In this child who has a neurogenic bladder, poor bladder emptying, and a solitary kidney, failure to use a completely diverting colostomy allowed ongoing contamination of the urinary tract due to the presence of a rectourethral fistula. This child's life-threatening urosepsis was controlled by achieving complete bladder drainage and by converting the loop colostomy to a completely diverting colostomy.

DISCUSSION

This series underscores the importance of recognizing NVD in patients with imperforate anus and its variants. It also highlights several important and unique therapeutic considerations that alter the man-

agement of the imperforate anus and the management of NVD in these children. In our opinion, all patients with imperforate anus should be suspected as being at risk for NVD. Vertebral radiographs as well as screening ultrasonography of the spinal cord in infancy is mandatory. Children who present at an older age with evidence for a sacral mass, significant cutaneous dimpling, a sacral sinus, sacral pigmentation, or sacral hypertrichosis warrant careful neurological evaluation, which in our hands usually includes either MRI or computed tomographic myelography. Similarly, any patient with imperforate anus who presents with urinary retention, urinary incontinence, or deterioration of the upper tracts must be suspected as having NVD and evaluated accordingly.

It is our belief that all patients with imperforate anus should have a screening renal ultrasound and a radiographic VCUG. Bladder emptying must be documented and when a colostomy is performed for high imperforate anus a fully diverting colostomy should be performed to prevent bladder contamination through the rectourinary fistula. We recommend urinary suppression for all patients with rectourinary fistulas, and consider this mandatory for any patient found to have VUR or incomplete bladder emptying.

The management of NVD in patients with imperforate anus requires special considerations as well. In male infants with high imperforate anus lesions, IC may be quite difficult as it was in two patients in this series. The level of difficulty may preclude catheterization by the patient or parent. This is often related to a tortuous proximal urethra and the presence of small diverticuli in the proximal urethra at the point of division of the rectal fistula. This may be overcome with the use of a coude-tip catheter, but in one of the present patients the use of a Mitrofanoff continent catheterizable neourethra was essential. Additionally, bladder augmentation in patients with high

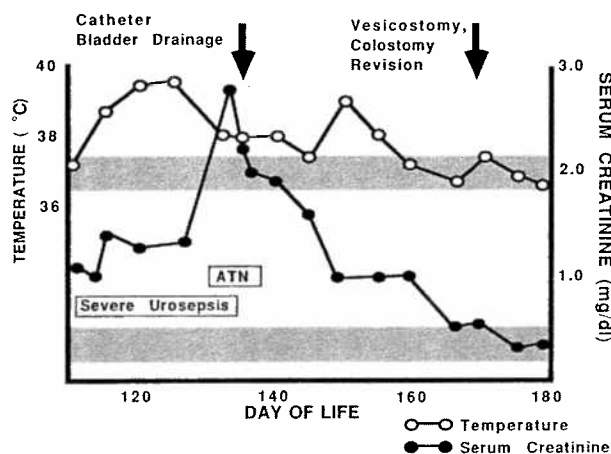


Fig 4. The hospital course with respect to serum creatinine and temperature of case 5. This demonstrates the sequelae of severe urosepsis in a child with imperforate anus in whom chronic urinary retention was unrecognized, and a partially diverting colostomy was performed for high imperforate anus. Urosepsis persisted despite antibiotics and resolved only after appropriate drainage had been instituted.

imperforate anus lesions requires specific consideration. Use of a sigmoid colon augmentation should be avoided due to the potential for devascularizing the distal bowel. In our opinion, the ileocecal valve should be maintained intact to avoid compromise of possibly marginal anal sphincter function. As a result, we recommend the use of small bowel or stomach for bladder augmentation in this instance.

In summary, it is well recognized that anatomic urinary pathology in children with imperforate anus may have a profound influence on the management and prognosis of these children. This study has illustrated the importance of NVD in patients with imperforate anus and its variants. All patients with imperforate anus must be considered at risk for NVD.

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SURGICAL IMPLICATIONS OF GENITOURINARY TRACT ANOMALIES IN PATIENTS WITH IMPERFORATE ANUS

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ABSTRACT

Genitourinary anomalies in patients with imperforate anus are a frequent source of significant morbidity. We report a retrospective study of 64 genitourinary procedures performed on 23 patients. Renal, collecting system, bladder and perineal anomalies were encountered in 65%, 83%, 87% and 65% of these complex cases, respectively. A total of 70% of the patients required intermittent catheterization due to bladder dysfunction. Surgical alternatives, complications of management and functional outcomes are reviewed in detail. Genitourinary reconstruction is best undertaken as an integral part of imperforate anus reconstruction. Failure to do so results in the loss of surgical alternatives, unnecessary reoperative procedures and compromised outcomes.

KEY WORDS: anus, imperforate; operative surgery; postoperative complications; urinary catheterization

Morbidity and mortality in cases of imperforate anus are largely attributed to associated structural and functional genitourinary anomalies.¹ Such morbidity often exceeds that of imperforate anus itself, and major genitourinary reconstruction is frequently required. We previously reported a retrospective review of 484 patients with imperforate anus anomalies.² Of these patients 6.4% with high imperforate anus anomalies and 1.1% with low anomalies died of renal failure. Nonfistulous genitourinary anomalies were encountered in 60% of high and in 20% of low imperforate anus cases. Significant upper tract anomalies were discovered in more than 33% of the cases and bilateral anomalies in 14% (7% symmetrical and 7% asymmetrical bilateral upper tract anomalies). We also reported a retrospective review of 90 consecutive cases of imperforate anus, of which 18% had neurovesical dysfunction.³ Again, the highest incidence of this pathological condition was encountered in patients with high imperforate anus anomalies.

Surgical management of such complex cases must be conceptualized and managed cohesively as a single complicated anomaly rather than as multiple independent anomalies. A fragmented approach to diagnosis, and medical and surgical management leads to unnecessary loss of therapeutic options and compromises outcome. Imperforate anus management may adversely affect subsequent genitourinary reconstruction and, conversely, genitourinary management may compromise the result of imperforate anus reconstruction. Surgery for imperforate anus has occasionally been implicated as the etiology of some associated anomalies, such as neurovesical dysfunction and urethral stricture or tortuosity.⁴⁻⁷ In this report we demonstrate the importance of neonatal screening and emphasize the essential role of an integrated surgical approach to operative planning.

MATERIALS AND METHODS

We retrospectively reviewed all cases of imperforate anus encountered from 1987 to 1992. Genitourinary tract anomalies with important surgical implications were present in 23 cases, which were classified as high or low imperforate anus anomalies depending on whether colostomy was performed as part of initial management. It was not possible to determine which cases would have been classified as intermediate lesions.

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RESULTS

A total of 64 genitourinary surgical procedures was performed on 12 male, 9 female and 2 genetically male patients 1 day to 23 years old. The 2 genetically male subjects had cloacal exstrophy and diminutive phallic structures, and underwent female reconstruction. Only 1 male patient with associated classical bladder exstrophy had low imperforate anus, while the remaining 22 had high imperforate anus anomalies, including cloaca in 6 and cloacal exstrophy in 3.

Figure 1 outlines the genitourinary diagnoses. Renal anomalies, most frequently dysplasia and agenesis, existed in 15 patients (65%). Collecting system anomalies were found in 19 patients, of whom 13 (57% of 23) had vesicoureteral reflux. Only 3 patients had unilateral reflux with a normal contralateral upper tract. Bilateral reflux was present in 5 patients and unilateral reflux with an abnormal contralateral collecting system in 5. Agenesis of the contralateral kidney was seen in 2 patients, obstructing megaureter in 1, a multicystic dysplastic kidney in 1 and a hydronephrotic ectopic ureteral insertion in 1.

A finding that further emphasizes the significance of vesicoureteral reflux is that only 2 patients with reflux had normal

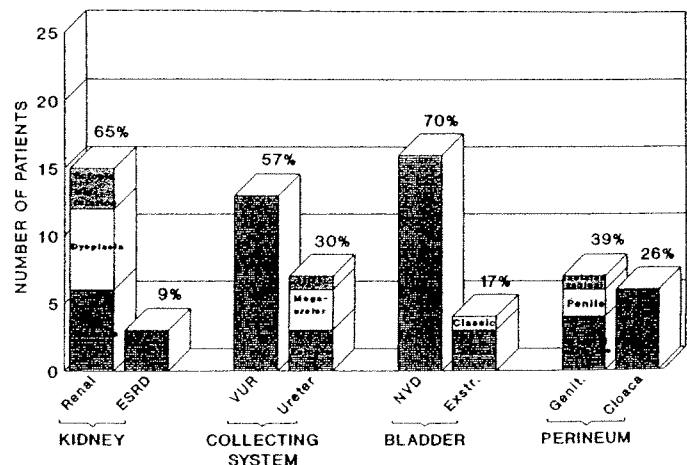


FIG. 1. Associated genitourinary anomalies. Renal, renal anomalies. ESRD, end stage renal disease. VUR, vesicoureteral reflux. Ureter, ureteral anomaly. NVD, neurovesical dysfunction. Exstr., exstrophy. Genit., genital anomaly. Cloaca, cloacal anomaly.

bladders. Neurovesical dysfunction or bladder outlet obstruction existed in 9 patients and cloacal or classical bladder exstrophy in 2. Only 1 patient had a protected upper tract system with a nonrefluxing normal contralateral kidney and a normal bladder. Functional or structural bladder abnormalities were frequent in this population. Evidence of neurovesical dysfunction was noted in 16 patients, many of whom were old enough to have this diagnosis proved urodynamically, while 7 had retention problems that were not urodynamically evaluable due to age or anatomy. Exstrophy anomalies were encountered in 4 patients (17%), of whom 3 had cloacal exstrophy and 1 classical bladder exstrophy.

Perineal anomalies were seen in 15 cases (65%) and were associated with cloaca in 6. Urethral strictures were noted in 4 cases, of which 3 proved to be congenital on radiography and/or endoscopy before surgical intervention. The remaining case was presumed to be congenital but not proved. Penile abnormalities existed in 2 patients (1 hypospadias and 1 epispadias) and vaginal anomalies were frequent. In addition to the vaginal anomalies in the 6 patients with cloaca, 1 with an otherwise uncomplicated high imperforate anus had isolated vaginal agenesis. In 1 genetically female patient cloacal exstrophy necessitated vaginal reconstruction for a duplication anomaly, and the 2 patients assigned a female gender will require future vaginal reconstruction.

Figure 2 outlines the genitourinary procedures performed. Of the 4 patients requiring nephrectomy 1 has undergone successful renal transplantation and maintains normal renal function 1.5 years later. Ureteral reimplantation was performed in 10 patients.

Cutaneous ureterostomy was done in 3 patients. Neonatal epididymitis secondary to a hydronephrotic ectopic ureter inserting into the seminal vesicle was encountered in 1 patient. Another patient with ectopic ureter associated with hydronephrosis and hydroureter with markedly diminished renal function and cloacal exstrophy required a temporary ureterostomy. The remaining patient, who had bilateral obstructing megaueters with neurovesical dysfunction, was about to undergo bladder augmentation and ureteral reimplantation when intraoperative ventilatory collapse necessitated termination of the procedure. Therefore, cutaneous ureterostomy was performed instead.

Surgical procedures pertinent to bladder abnormalities were particularly frequent. A total of 16 patients (70%) required

intermittent catheterization for retention dysfunction and 10 underwent bladder augmentation. Of the 10 patients gastrocystoplasty was done in 8, ileocystoplasty in 1 and sigmoid colonic augmentation in the patient with low imperforate anus. To date, 35% of the patients underwent a Mitrofanoff procedure, which involves creation of a continent, catheterizable neo-urethra that provides bladder access for intermittent catheterization when native urethral catheterization is impossible, excessively difficult or painful. The neo-urethra was constructed of ureteral segments in 4 cases, appendix in 2 and tapered ileal segments in 2.

Bladder neck reconstruction was required in 22% of the patients. Three patients with exstrophy anomalies underwent Young-Dees-Leadbetter reconstruction, 1 with neurovesical dysfunction underwent a Kropp procedure, and 1 with a complex cloacal anomaly and excessive perineal scar tissue due to previous surgery underwent closure of the bladder outlet. Cutaneous vesicostomy for temporary bladder decompression was done in 9 cases (39%), including 3 with neonatal urorepis and 4 with hydronephrosis due to urinary retention after intermittent catheterization failed. In 1 case of congenital urethral stricture and rectourethral fistula absorptive hyperchloremic metabolic acidosis developed due to retrograde passage of urine into the distal colon, which was immediately corrected following vesicostomy. In addition, a neonate with a cloacal anomaly who experienced massive, symptomatic urinary distention of the vagina could not be managed by intermittent vaginal catheterization at home because the parents were unable to negotiate the urogenital sinus consistently. Consequently, cutaneous vesicostomy was performed.

Posterior sagittal rectovaginal urethroplasty was done in 2 cases as a primary procedure and in 2 as a reoperation. Vaginal reconstruction by techniques other than posterior sagittal rectovaginal urethroplasty was performed in 4 cases, while hypospadias repair was done in 1, epispadias repair in 1 and urethroplasty in 1.

A total of 18 surgical complications was encountered in 14 patients in whom 44 genitourinary reconstructive procedures were performed. Nine major surgical complications due to surgery preceded referral and 9 complications (3 major) occurred under our care.

Of the complications that occurred secondary to surgery performed before referral 4 resulted from performing urethral and vaginal reconstruction as separate procedures subsequent to imperforate anus repair. All 4 of these patients were referred with cloaca, and had previously undergone anorectal pull-through operations. The complications may have been avoided if a comprehensive surgical approach had been followed. In 2 of these cases magnetic resonance imaging (MRI) suggested that fecal continence could be improved by repeat posterior sagittal rectovaginal urethroplasty due to an ectopic course of the rectum through the striated sphincteric muscular complex. Both patients underwent this procedure and recovered well without complication. In the 2 remaining patients the MRI scan suggested that fecal continence was unlikely to be improved by repeat anorectal surgery. Both patients underwent urethrovaginal reconstruction using perineal inlay flap techniques. However, urethrovaginal fistulas developed in both cases, which accounted for 2 more complications. Three patients had undergone ureteral reimplantation for the correction of reflux before referral, which failed because associated high intravesical pressure was not recognized. Repeat reimplantation was successfully performed after normalization of bladder function. In 2 cases bladder augmentation was done, and 1 case was managed by anticholinergics and intermittent catheterization.

The complications that occurred secondary to surgery performed after referral include 2 bowel obstructions (1 adhesions and 1 hindgut stomal stenosis) that were successfully corrected,

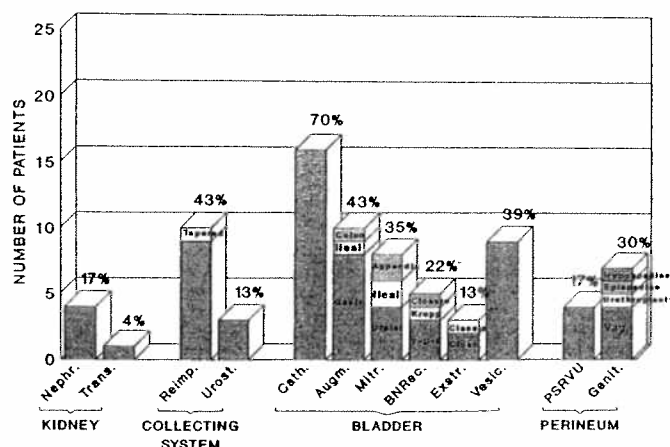


FIG. 2. Procedures performed. *Nephrect.*, nephrectomy. *Trans.*, transplantation. *Reimp.*, ureteral reimplantation. *Urost.*, cutaneous ureterostomy. *Cath.*, intermittent catheterization. *Augm.*, bladder augmentation. *Mitr.*, Mitrofanoff neo-urethra. *BNRec.*, bladder neck reconstruction. *Exstr.*, exstrophy anomalies. *Vesic.*, cutaneous vesicostomy. *PSRVU*, posterior sagittal rectovaginal urethroplasty. *Genit.*, genital reconstruction.

and 1 ventilatory collapse during bladder reconstruction that necessitated abrupt termination of the procedure. The etiology of this complication was subsequently demonstrated to be latex allergy. A lymphocele that developed after creation of a gastric neo-bladder was successfully treated by percutaneous aspiration. A urethral stricture that recurred after urethroplasty for a severe congenital urethral stricture resolved with urethral dilation. A urethral stone that formed on a nidus of nonabsorbable suture used to approximate the pubis following closure of cloacal exstrophy was removed endoscopically. Surgical revision for Mitrofanoff stomal stenosis restored normal function in 1 case in which the Mitrofanoff neo-urethra was constructed of ureter and in 1 in which it was made of tapered ileum. In the remaining case late vaginal stenosis after primary posterior sagittal rectovaginal urethroplasty for cloaca was caused by injury from vaginal dilation.

The degree of success from the perspective of renal function and urinary continence is of greatest importance. End stage renal disease exists in 3 of our patients and diminished renal function in 5. Urinary continence was evaluable in 16 patients, of whom 13 are totally continent, 2 are partially continent with 2 to 3-hour dry intervals after voiding or catheterization and 1 is totally incontinent. Of the remaining 7 patients 2 are too young for evaluation of urinary continence and 5 continue with diversion.

DISCUSSION

Our series highlights 3 important characteristics regarding patients with imperforate anus: 1) there are a large number of associated genitourinary anomalies;^{2,3,8,9} 2) the risk of death and major morbidity from these anomalies exceeds that of the imperforate anus,¹ and 3) many of the abnormalities previously attributed to surgical intervention for imperforate anus are congenital. The latter fact is particularly true of patients with neurovesical dysfunction and urethral stricture.

Several important reconstructive principles may be identified. Neonatal screening for genitourinary anomalies is essential for all patients with imperforate anus. Renal ultrasound and radiographic voiding cystourethrography should be obtained in all instances, preferably before colostomy. In addition, vertebral radiographs and ultrasound of the spine should be obtained for all neonatal patients. Patients who are old enough to have undergone calcification of the vertebral spine will require MRI to identify associated spinal cord anomalies. The assessment of bladder emptying is also critical, and is generally demonstrable on voiding cystourethrography, post-voiding ultrasound or post-voiding catheterization before the patient is discharged home.

The combination of recto-urinary fistulas and neurovesical dysfunction or bladder outlet obstruction predisposes to critical neonatal illness.⁶ Neonatal urosepsis, absorptive hyperchloremic metabolic acidosis and progressive pyelonephric renal injury exacerbated by the presence of reflux are of critical importance. Early, maximal decompression of obstructive hydronephrosis is essential to minimize the risk of chronic renal insufficiency.

Reflux in cases of imperforate anus is a common and serious finding.^{2,10} Recto-urinary fistula exacerbates the risk of pyelonephritis, as does the high incidence of associated neurovesical dysfunction or bladder outlet obstruction. Correction of vesicoureteral reflux by ureteral reimplantation is prone to failure if the surgeon does not detect and correct an associated high pressure dysfunctional bladder. Urodynamic investigation is indicated in all such cases and aggressive medical or surgical management is recommended when bladder dysfunction is detected.³ One should consider performing a fully diverting, relatively low colostomy with complete evacuation of the distal limb. Prophylactic antibiotics are necessary, as is the need to ensure effective bladder emptying by spontaneous voiding, in-

termittent catheterization or occasionally performing a cutaneous vesicostomy.

If bladder outlet obstruction or neurovesical dysfunction is detected or suspected, incidental appendectomy is contraindicated.¹¹ An appendiceal Mitrofanoff neo-urethra provides excellent bladder access for decompression and continence in even the most devastating cases. While the appendix may be congenitally absent in cases of cloacal exstrophy, 3 patients in our series were unable to receive an appendiceal Mitrofanoff neo-urethra due to previous appendectomy.

If bladder augmentation is necessary, several important points should be considered. The use of colon for bladder augmentation is generally inappropriate for high imperforate anus pathology. Marginal fecal continence is often present in such cases and fecal incontinence may become incapacitating if colonic physiology is disturbed. The use of an ileocecal segment may diminish the consistency of the stool, thereby exacerbating a tendency to incontinence. In addition, it is our impression that the use of a section of sigmoid colon may result in the loss of accommodation and worsen preexisting compromised fecal continence. Patients who have undergone anorectal mobilization could experience devascularization of the anorectum if a distal colonic segment were used because of the descending nature of its blood supply. If ureteral reimplantation or a Mitrofanoff neo-urethra is necessary and cannot be placed in the native bladder segment, small bowel augmentation presents a poor alternative due to an inability to achieve a consistent antirefluxing mechanism.

Cases of cloaca should never be managed by initial anorectal reconstruction alone,^{6,10} which necessitates extensive and complicated subsequent surgery. In our experience posterior sagittal rectovaginal urethroplasty appears to be the most appropriate surgical procedure in such cases. The complication rate associated with posterior sagittal rectovaginal urethroplasty was extremely low in primary and reoperative cases. In contrast, all patients in whom we attempted urethrovaginal reconstruction without disturbing the previously reconstructed anus experienced complications in the form of urethrovaginal fistulas, which appeared to be caused by perineal and perirectal scar contracture, and the resulting impairment of healing.

Patients who will experience chronic latex exposure must be tested preoperatively for latex allergy and appropriate precautions should be taken during subsequent reconstructive surgery.¹² Long-term exposure includes intermittent catheterization, a suprapubic tube and a gastrostomy tube. In our series 1 patient experienced intraoperative ventilatory collapse that necessitated termination of surgery.

The midline intestinal segment of the cloacal exstrophy plate need not be used to achieve bladder physiology compatible with renal preservation and urinary continence. This segment should be incorporated into the fecal stream by tubularization since this procedure in conjunction with preservation of the entire length of hindgut helps promote nutrient absorption and growth.

In conclusion, the presence of genitourinary anomalies in patients with imperforate anus markedly influences management. However, a comprehensive, integrated approach to the reconstruction of these anomalies yields excellent results.

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INSTRUCTIONS FOR INITIATING GENTAMICIN BLADDER IRRIGATIONS

1. Obtain order from physician – use attached order protocol
2. In consultation with parents, choose a home care, not outpatient, pharmacy and send orders. They will typically precert the insurance, prepare the drug in unit dose syringes, provide catheter tip syringes for drug administration, and deliver the drug to the patient's home.

If the patient lives out of your area, you may need to contact their local pediatrician, explain what our urologist would like to order, and enlist the local doctor's assistance in arranging the gentamicin locally through a local home care pharmacy (usually one which handles IV drugs) or a local hospital pharmacy.

Occasionally, a patient will have a relationship with a local, independent pharmacy that is willing to mix and provide the drug. Usually they will take a verbal order in the same way that you would call in a script to a pharmacy.

3. Arrange lab monitoring. Ask the parents where they would like to have labs drawn and send lab orders per protocol. Remind the parents to call your office to check the results.
4. Teach procedure per protocol. With many parents you can do this verbally. You can also demonstrate with a catheter, water filled syringe, and a cup rather than actually catheterizing the patient. If the family desires or you think it is advisable, you can order a home care nurse visit to do a one-time teaching visit. Choose a skilled nursing agency and process order per their protocol

All orders must be written in the metric system and include date, time, physician's signature and pager/phone number. Use ball point pen.

Date _____ Time _____ Weight _____ KG Height _____ CM _____ M²

Allergies: ☐ No Drug/Contrast Allergy ☐ No Food Allergy ☐ No Product/Latex Allergy ☐ Unable to Obtain Allergy Information
Specifics: _____

Pediatric Urology Gentamicin Bladder Irrigation Protocol Orders

Solution: 120 mg gentamicin in 250 mL normal saline

Stability: 4 days refrigerated, 1 month frozen

(1) **Dose:** (Choose amount, & treatment or maintenance)

Amount: ☐ 30 mL OR ☐ _____ mL

☐ **Treatment** – ordered amount of gentamicin solution BID for _____ days

☐ **Maintenance** – ordered amount of gentamicin solution at bedtime

(2) **Frequency:** (Choose frequency of maintenance dose):

☐ Daily

☐ Every other day

☐ _____ Days per week

(3) **Duration:** (Choose length of maintenance therapy)

☐ 3 months

☐ 6 months

☐ Other _____

(4) **Monitoring:** ☐ Baseline BUN, creatinine _____

☐ **Treatment therapy:** Creatinine, random gentamicin level after 3rd dose
AND/OR

☐ **Maintenance therapy:** (Choose risk level)

☐ **High Risk** (Patients with renal insufficiency, renal scarring) –

Obtain: BUN, creatinine, random gentamicin level twice/week for 2 weeks,
then once/week for 2 week, then every 2 weeks for duration of therapy

☐ **Moderate Risk** (Patients with bladder augmentations)

Obtain: BUN, creatinine, random gentamicin level once/week for 2 weeks,
then every 2 weeks for 1 month, then once/month for duration of therapy

☐ **Low Risk** (All other patients) – Obtain random gentamicin level after one week. If no
absorption of gentamicin documented, no further monitoring

Procedure: Transfer gentamicin solution from leur tip syringe to catheter tip syringe. Catheterize the bladder per patient's usual procedure, drain urine from bladder and leave catheter in bladder when drained. Then attach gentamicin solution-filled syringe to catheter and gently push medicine through catheter into bladder. Remove syringe and hold catheter end up to allow drug to drain by gravity into bladder. Then pinch catheter and withdraw from bladder, allowing gentamicin to remain indwelling in bladder until next scheduled catheterization.

Physician Signature/Credentials

Pager #

HOME CARE COMPANY
GENTAMICIN BLADDER IRRIGATION PREPARATION INSTRUCTIONS

Solution – 120 mg gentamicin in 250 ml normal saline

Stability – 4 days refrigerated
One month frozen

Delivery – Please prepare and dispense the drug in unit dose syringes which can then be frozen safely for one month. The parent then daily moves the next day's dose from the freezer to the refrigerator. If you dispense the drug in leur-tip syringes, an equal number of catheter-tip or funnel-end syringes should be provided. The parent can slip the tip of the leur-tip syringe into the tip of the funnel-end syringe to transfer the drug prior to administration. Dispensing the drug in bags from which the parent withdraws a daily dose should be avoided as a new bag of solution will be required every 4 days since the solution is only stable for 4 days thawed.

Procedure – catheterize the bladder per patient's usual procedure, drain urine from bladder and leave catheter in bladder when drained. Then attach gentamicin solution-filled syringe to catheter and gently push medicine through catheter into bladder. Remove syringe and hold catheter end up to allow drug to drain by gravity into bladder. Then pinch catheter and withdraw from bladder, allowing gentamicin to remain indwelling in bladder until next scheduled catheterization.

Bladder Irrigation Instructions

1. Irrigate Mitrofanoff bladder catheter every morning and every bedtime with ordered amount of normal saline for irrigation. The amount should be no more than 1/2 the patient's bladder capacity but should be a large enough amount to dilute the mucous.

- allow the irrigation solution to drain from catheter
- then attach catheter-tip syringe to catheter and gently pull back on the syringe to aspirate or pull back any remaining fluid or mucous. Continue reattaching syringe and aspirating back until you meet resistance. Do not force – once you meet resistance, stop aspirating.
- Once all fluid and mucous have been removed from the bladder, attach the catheter to the drainage bag, if the catheter is indwelling, or remove the catheter if child is catheterized intermittently.

2. If necessary, the bladder may also be irrigated at lunch and dinner times with 60 ml of normal saline for irrigation. Then aspirate out fluid and mucous as described above.

The catheter may be irrigated any additional times when urine is not flowing easily. Call the Urology office if you have difficulty irrigating the bladder or if urine does not drain into the bag even after irrigation.

CATHETERIZATION .TIPS

ENGAGE CHILD AND FAMILY

- Relaxation techniques
- Explanation
- Allow parent or child to cath if able to

CHOOSE CATHETER FOR CHILD AND SITUATION

- Size, type
- History difficult cath
- Anatomy — vesicostomy, Mitrofanoff

MALE TIPS

- Intra urethral lidocaine
- Tight sphincter
- Technique

FEMALE TIPS

- Lidocaine, emla
- Locating urethral meatus
- Anatomical differences

FACILITATING SELF ESTEEM

- Reinforce feelings of competence and learned coping skills
- Help child to normalize experience

GENTAMICIN BLADDER IRRIGATION THROUGH INDWELLING CATHETER

Procedure: Provide drug in unit dose syringes. If gentamicin solution is prepared in leur tip syringes, transfer gentamicin solution from leur tip syringe to catheter tip syringe. Disconnect drainage bag from catheter and cover bag tubing with a cap or sterile gauze. Attach gentamicin solution-filled syringe to catheter and gently push medicine through catheter into bladder. Plug catheter and leave medicine to dwell in bladder for one hour. If there is more than one catheter in the bladder, all catheters must be clamped for the one hour dwell period. After one hour, reconnect drainage bag to catheter and allow gentamicin to drain. Unclamp any other catheters also.

MITROFANOFF CATHETERIZATION OF THE BLADDER WITH HYDROPHILIC CATHETER

Definitions

MITROFANOFF - A continent, catheterizable tube, leading from the bladder to the outside abdominal wall. A Mitrofanoff is often created with the appendix, but may also be constructed with a portion of small bowel or with a ureter

CATHETERIZATION - Catheterization is removing urine from the bladder by placing a tube, called a catheter, into the bladder. This is done when children are unable to empty their bladders on their own, when a child's bladder leaks urine, or when high pressures have developed in the bladder.

Supplies Needed:

1. Catheter – size _____; coude _____ or straight _____
2. Lubricant - water soluble
3. Betadine swabs
4. Urine collection container - an emesis basin or cup
5. Gloves, if ordered

Procedure:

1. Wash hands well with soap and water.
2. Prepare supplies. Open Betadine swabs and place on a clean paper towel. Peel open hydrophilic catheter pack about 3 inches. Pour sterile water into catheter pack so that the water rises about 3 inches from the bottom of the catheter. If water is included in catheter pack, follow instructions to dispense water. Let catheter with water sit while preparing child for catheterization.
3. Cleanse Mitrofanoff opening. Starting in the center of the Mitrofanoff opening, wipe the Betadine swab in a circular outward direction around the opening. Repeat this procedure a total of three times
4. If directed to wear gloves, put the gloves on now. Pull the catheter out of the pack, squeezing the catheter to firmly grasp the slippery surface. Ask your child to take deep breaths or blow out so that the muscles will be relaxed when you put the catheter in.
5. Insert the catheter straight into the Mitrofanoff opening. If using a curve-tipped catheter, point the curve tip toward the middle of the body and diagonally downward as you insert the catheter. Continue to push the catheter in a downward direction toward the middle of the body. You will be pushing the catheter in a diagonal direction. You may meet a feeling of resistance as you push the catheter in - continue gently, but firmly pushing the catheter until it passes the point of resistance and urine starts to flow. If you are unable to push the catheter in, do not force it.
6. If the bladder is overfull, the Mitrofanoff may become very tight and be difficult to catheterize. If this seems to be happening, ask your child to urinate if able, or catheterize the child through the urethra if you have been taught to do this. Emptying some urine from the bladder by urinating or catheterizing from below will relieve some pressure from the Mitrofanoff and make it easier to catheterize. If you are still unable to catheterize, stop and notify the doctor.
7. When urine stops flowing, slowly remove the catheter. Clean the Betadine off the skin with a wet washcloth. Note the amount and color of the urine you have collected.
8. Call the urology office or the urology doctor on call if you cannot put in the catheter, if there is blood on the catheter or draining from the Mitrofanoff opening once you remove the catheter, if your child has fever, pain over the lower abdomen, pain with catheterization or urinating, or new or increased urine leakage.

MITROFANOFF CATHETERIZATION OF THE BLADDER

Definitions

MITROFANOFF - A continent, catheterizable tube, leading from the bladder to the outside abdominal wall. A Mitrofanoff is often created with the appendix, but may also be constructed with a portion of small bowel or with a ureter

CATHETERIZATION - Catheterization is removing urine from the bladder by placing a tube, called a catheter, into the bladder. This is done when children are unable to empty their bladders on their own, when a child's bladder leaks urine, or when high pressures have developed in the bladder.

Supplies Needed:

1. Catheter – size _____; coude _____ or straight _____
2. Lubricant - water soluble
3. Betadine swabs
4. Urine collection container - an emesis basin or cup
5. Gloves, if ordered

Procedure:

1. Wash hands well with soap and water.
2. Prepare supplies. Open Betadine swabs and place on a clean paper towel. If catheter is disposable, peel open pack and leave catheter in pack. If catheter is self sterilized, place catheter on a clean paper towel. Squeeze lubricant onto open catheter pack or paper towel.
3. Cleanse Mitrofanoff opening. Starting in the center of the Mitrofanoff opening, wipe the Betadine swab in a circular outward direction around the opening. Repeat this procedure a total of three times
4. If directed to wear gloves, put the gloves on now. Pick up the catheter and roll the tip of the catheter and the first 2-3 inches lubricant. Ask your child to take deep breaths or blow out so that the muscles will be relaxed when you put the catheter in. It is also helpful to offer your child distractions, such as a toy or video.
5. Insert the catheter straight into the Mitrofanoff opening. If using a curve-tipped catheter, point the curve tip toward the middle of the body and diagonally downward as you insert the catheter. Continue to push the catheter in a downward direction toward the middle of the body. You will be pushing the catheter in a diagonal direction. You may meet a feeling of resistance as you push the catheter in - continue gently, but firmly pushing the catheter until it passes the point of resistance and urine starts to flow. If you are unable to push the catheter in, do not force it.
6. If the bladder is overfull, the Mitrofanoff may become very tight and be difficult to catheterize. If this seems to be happening, ask your child to urinate if able, or catheterize the child through the urethra if you have been taught to do this. Emptying some urine from the bladder by urinating or catheterizing from below will relieve some pressure from the Mitrofanoff and make it easier to catheterize. If you are still unable to catheterize, stop and notify the doctor.
7. When urine stops flowing, slowly remove the catheter. Clean the Betadine off the skin with a wet washcloth. Note the amount and color of the urine you have collected.
8. Call the urology office or the urology doctor on call if you cannot put in the catheter, if there is blood on the catheter or draining from the Mitrofanoff opening once you remove the catheter, if your child has fever, pain over the lower abdomen, pain with catheterization or urinating, or new or increased urine leakage.

BLADDER AUGMENTATION

UZAY YASAR
CURTIS A. SHELDON

Bladder augmentation is a complex yet highly successful option in the field of surgical reconstruction of the pediatric bladder. Refinements of surgical technique, the application of clean intermittent catheterization (CIC), advances in urodynamics, and review of our experience in urinary diversion and undiversion have led to major advances in this dynamic field. Despite these successes, there remain adverse sequelae and long-term complications that must be considered before undertaking augmentation cystoplasty.

There are several goals in the surgical restoration of bladder function. First and foremost is the protection of the upper urinary tracts with the creation of a large-capacity, low-pressure reservoir for urine storage. Patients should be able to go 4 hours between catheterizations while maintaining a low intravesical storage pressure. When bladder pressure is greater than 40 cm H₂O, patients are at high risk for upper tract deterioration. Urinary continence is another important goal of reconstruction. This is often a difficult problem to diagnose and treat because of its multiple causes, ranging from bladder neck incompetence to uninhibited detrusor contractions to an areflexic detrusor muscle, resulting in overflow incontinence. If possible, the preservation of spontaneous voiding should be attempted. When spontaneous voiding is not feasible, access for easy catheterization becomes a necessity, whether by the native urethra or by the creation of a continent, catheterizable conduit. Keeping these fundamentals of bladder reconstruction in mind, augmentation may be applied for the correction of many urinary tract anomalies with good expectation for success.

WORKUP

Before surgical reconstruction on the bladder is pursued, more conservative measures must be attempted. Initial treatment for any patient with a dysfunctional bladder being considered for augmentation should first consist of pharmacologic therapy, often in combination with CIC. Anticholinergics can be used to increase the capacity and compliance of the bladder to an acceptable degree. Behavioral modifications also may play a part in conservative therapy. If these measures fail, surgical therapy might be warranted.

Meticulous preoperative evaluation of the patient is critical to determine if the patient is a candidate for bladder augmentation. A careful history and physical examination are performed. Preoperative evaluation of the patient should assess renal function, a 24-hour urine collection for volume measurement, and glomerular filtration rate if renal function is in question. Determining urine output is essential for creating an augmented bladder with a large enough capacity to store at least 4 hours of urine output. This is especially true for children in high-output renal failure who may produce up to 4 L of urine in 1 day. *Nuclear renography* is indicated if elements of obstruction or renal scarring are present. *Voiding cystourethrography* is an important study to determine the presence of vesicoureteral reflux, whereas formal *urodynamics* provide crucial information on leak point pressure, bladder capacity, bladder compliance, and the presence of uninhibited detrusor contractions. Lastly, *upright cystography* can be performed to evaluate the competency of the bladder neck.

PREOPERATIVE PREPARATION

Once the decision for augmentation is made, the patient must be properly prepared. The full nature of the operation and its inherent risks and potential complications must be discussed. The patient must understand that he

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or she may lose the ability for spontaneous voiding and that intermittent catheterization may become a permanent part of life postoperatively. Because postoperative compliance with management is critical to the success of any bladder reconstruction, a careful assessment of the child's willingness and ability to comply is important. A full bowel preparation must be performed preoperatively. This must be done regardless of the reconstructive procedure planned, for the surgeon must have the ability to tailor the case intraoperatively for any eventuality. The preparation can be performed in an outpatient setting with less complicated patients, but often requires a full 2 days of in-hospital mechanical preparation for those children with a neuropathic bladder and bowel. Performing an adequate bowel preparation in this patient population can be a challenge.

Cystoscopy should be performed immediately before the augmentation to assess native anatomy such as bladder size and morphology, location of ureteral orifices, and competency of the bladder neck. This can provide important anatomic insight and aid the surgeon in determining which reconstruction to perform. Additionally, a urine culture should be sent a few days before the operation and a positive culture should be treated preoperatively to prevent seeding of the peritoneal cavity with infected urine.

TECHNICAL CONSIDERATIONS

Several important concepts are pertinent to bladder reconstruction. Detubularization of an intestinal segment consists of opening an isolated bowel segment along its antimesenteric border. Detubularization of a bowel segment results in an interruption of its circular muscle fibers, causing an interruption of its basic electrical rhythm. This interruption results in blunting of bowel contractions and an increased compliance of the reconfigured bowel segment. This isolated segment is then reconfigured into a more spherical shape to fit the bladder. This is done by folding the intestinal segment upon itself in the shape of a U, S, or W and anastomosing the bowel wall together. In addition, reconfiguring the segment into a spherical shape maximizes the volume achievable for any given length of bowel segment. This results in a dramatic increase in effective bladder capacity when used to augment the bladder.

Another concept important in bladder reconstruction is management of the native bladder. If the detrusor muscle of the native bladder is left intact, it generates high pressures that can cause the augmented bowel segment to act as a diverticulum. There are two ways to avoid this problem. The first involves excision of the supertrigonal detrusor originally described by Gil-Vernet, leaving just a cuff of bladder to be anastomosed to the reconfigured intestinal segment. The second involves preserving the

native bladder and opening it with a wide sagittal incision starting just cephalad to the bladder neck anteriorly. This incision is carried posteriorly to the trigone of the bladder. The augmenting segment is then anastomosed to this "clam-shell" bladder. This seems to be a more advantageous technique for management of the native bladder. It is technically easier to anastomose the reconfigured bowel segment to the remaining bladder, and the supertrigonal bladder adds to the overall capacity of the augmentation. Additionally, both appendicovesicostomy (Mitrofanoff principle) and ureteral reimplantation are more easily performed into the detrusor muscle than into a bowel segment. This becomes especially important if vesicoureteral reflux exists preoperatively, necessitating ureteral reimplantation.

The Mitrofanoff principle was introduced in 1980 and is currently an important option for the surgeon performing augmentation cystoplasty in the pediatric population. It involves the creation of a continent, catheterizable conduit that allows easy catheter access to the bladder. This involves the implantation of a tubularized visceral segment using a one-way flap valve mechanism into the urinary bladder with the exteriorization of the other end to the skin. This exteriorization is usually to the abdominal wall, but also may be to the umbilicus or the perineum. The conduit is commonly fashioned from appendix but also can be created using a segment of ureter or tapered ileum. It is the one-way flap valve mechanism that provides continence, and effective implantation of the valve has been demonstrated not only into native bladder but also into colonic and gastric augmented segments of the reconstructed bladder. Antegrade passage of a catheter is easily accomplished, making this technique particularly well suited to the patient who has difficulty catheterizing the native urethra. This is a frequent finding in this population and includes children bound to a wheelchair, those who have a tortuous urethra from congenital deformity or reconstruction, and children who have only marginal control of their upper extremities. In addition, catheterizing the conduit is less uncomfortable than catheterizing the native urethra and may enhance patient compliance.

Various donor tissue can be used to augment the bladder, including ileum, an ileocecal segment, stomach, sigmoid colon, and ureter (Table 129.1). Recently, autoaugmentation, either alone or with the use of an intestinal seromuscular patch overlying denuded bladder epithelium, has been used. However these newer techniques have not replaced the gold standard of augmenting bladder with a segment of the intestinal tract.

The most common segment of bowel to be used in bladder augmentation is ileum. In the ileocystoplasty, a segment of ileum approximately 20 to 40 cm long is isolated and detubularized (Fig. 129.1). It is important that the mesentery of the isolated segment is long enough to reach the bladder. An ileoileostomy is performed and the

TABLE 129.1. SIGNIFICANT CHARACTERISTICS OF AUGMENTATION MODALITIES

Technique	Pros	Cons
Ileocystoplasty	Technically simple	Acidosis, infection, mucus Antirefluxing implantation unreliable
Sigmoid colocolocystoplasty	Technically simple Allows implantation of ureter or Mitrofanoff into tenia	Risks interruption of blood supply to rectum after reconstruction for imperforate anus Acidosis, infection, mucus
Ileocecolocystoplasty	Technically simple Allows antirefluxing implantation of dilated ureters with intussusception technique	Intussusception antireflux mechanism inherently unstable Risks devastating fecal incontinence in patients with marginal continence mechanism (e.g., myelomeningocele or VATER)
Gastrocystoplasty	Reduced risk of infection and mucus production Allows easy implantation of ureter or Mitrofanoff No risk of acidosis	Hematuria-dysuria
Ureterocystoplasty	Metabolically inert No mucus May be performed extraperitoneally	Antirefluxing implantation not possible Dilated ureter only occasionally available Risk of failure to attain adequate compliance and capacity
Autoaugmentation	Technically simple Metabolically inert No mucus May be performed extraperitoneally	Antirefluxing implantation not possible Risk of failure to attain adequate compliance and capacity

From Sheldon CA. Urinary reconstruction (rather than diversion) for continence in difficult pediatric urologic disorders. *Semin Pediatr Surg* 1996;5:11.

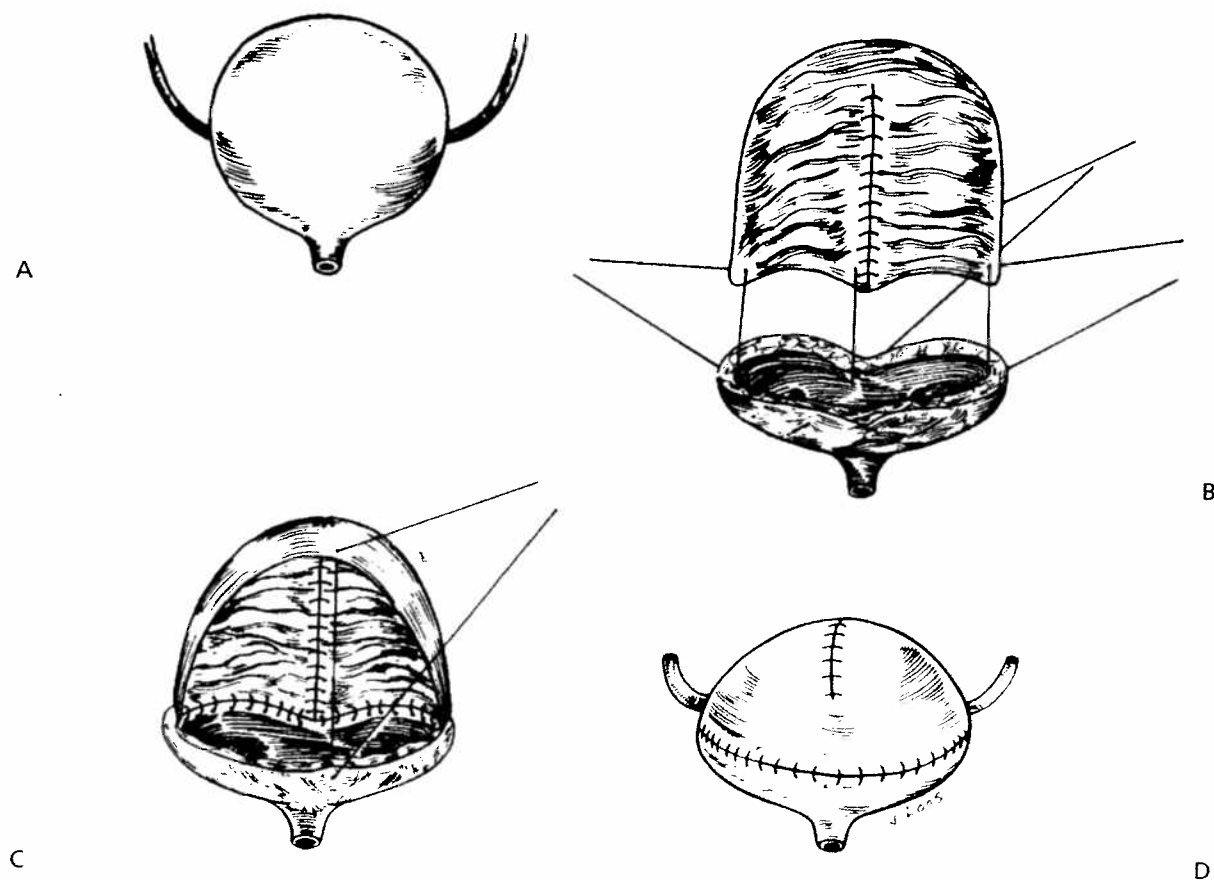


FIGURE 129.1. Bladder augmentation using an intestinal segment. **A:** The bladder is opened like a clam shell. **B:** The intestinal segment is detubularized by longitudinal incision along the antimesenteric border. A cup patch is fashioned by suturing one edge of the resultant rectangle to itself. **C:** The cup patch is sutured to the remnant bladder plate. **D:** Final appearance. (Reprinted from Sheldon CA, Bukowski T. Bladder function. In: Rowe MI, O'Neill JA, Grosfeld JL, et al., eds. *Essentials of pediatric surgery*. St. Louis: Mosby Year Book, 1995:731-747; with permission.)

mesenteric window closed to prevent internal herniation. Once the segment of ileum is reconfigured for maximum capacity, it is anastomosed to the clam-shelled bladder. The closure is started at the posterior aspect of the native bladder, and I prefer to close with two separate layers of running absorbable suture, the first layer interlocking. Before the closure is complete, a Malecot suprapubic tube (SPT) is brought out through a separate incision in the native bladder. Drains are left in place, and the urethral catheter placed preoperatively is left as a secondary means for post-operative bladder drainage.

The ileocececystoplasty is performed in much the same manner as the ileocystoplasty. The right colon is mobilized, and an ileocecal segment is isolated. Approximately 20 to 30 cm of terminal ileum is isolated. The segment is opened in its entirety along the antimesenteric border and reconfigured for anastomosis to the bladder. The advantage of the ileocececystoplasty is that it allows construction of an ileal antirefluxing nipple to which dilated

ureters can be attached, thus preventing the transmission of pressure and refluxing urine to the upper tracts. If necessary, the ureters also can be tunneled through the cecal segment, creating an antirefluxing mechanism. When the ureters are too short for reanastomosis to the bladder, a limb of ileum can be left intact to reach the shortened ureters. The ileocececystoplasty is rarely performed in the pediatric population, however, because removal of the ileocecal valve in children can result in intractable diarrhea. This is especially true in children with marginal continence mechanisms (myelomeningocele and imperforate anus patients), the very population most likely to need the bladder augmentation procedure.

Gastrocystoplasty involves the augmentation of the bladder using a segment of stomach, usually from the body. The segment is isolated by taking a wedge of stomach along the greater curvature, being careful to avoid the lesser curvature and thus injury to the vagus nerve (Fig. 129.2). This segment is brought down on a right gas-

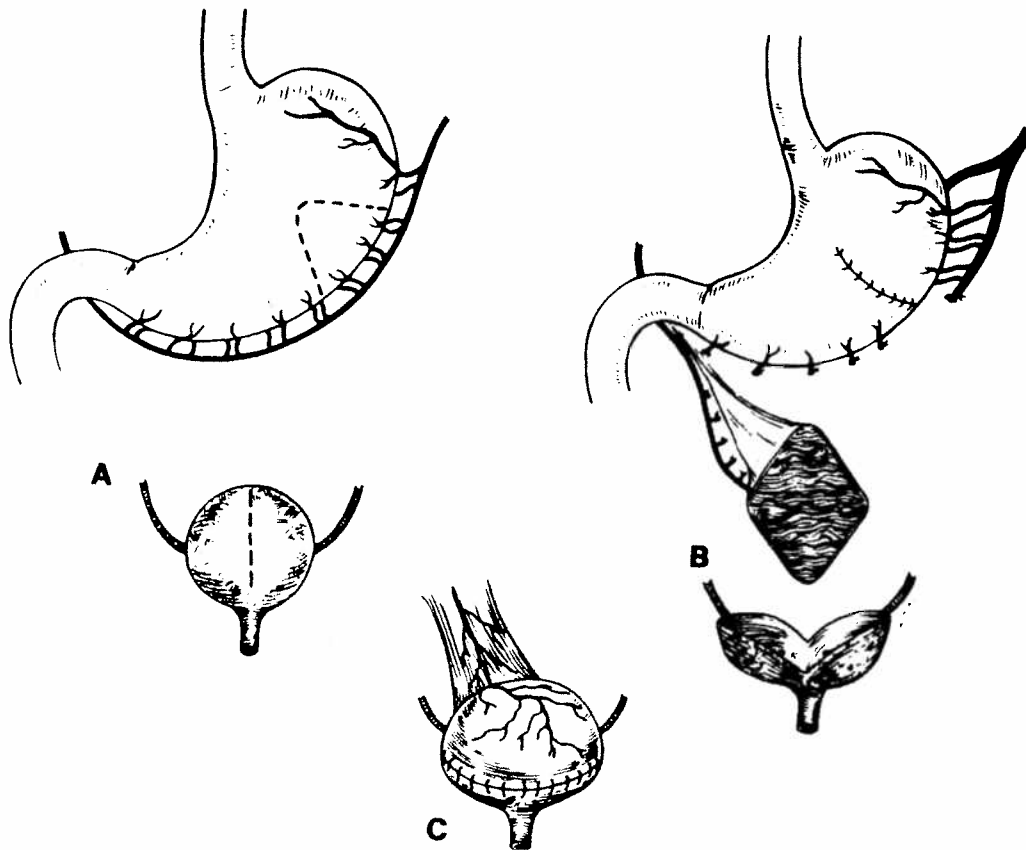


FIGURE 129.2. Bladder augmentation using a gastric segment: gastrocystoplasty. **A:** A wedge of nonantral stomach is isolated, based on the right gastroepiploic vessels. **B:** The bladder is prepared to open like a clam shell. **C:** The anastomosis is completed with a running double layer of absorbable suture. (Reprinted from Sheldon CA, Bukowski T. Bladder function. In: Rowe MI, O'Neill JA, Grosfeld JL, et al., eds. *Essentials of pediatric surgery*. St. Louis: Mosby Year Book, 1995: 731-747; with permission.)

troepiploic vascular pedicle, although infrequently the left gastroepiploic artery is used. This segment is then anastomosed to the opened bladder with a running double layer of absorbable suture. Using a segment of stomach has some distinct advantages over using small or large bowel. Compared with intestinal segments, it is less permeable to urinary solutes, specifically chloride, and presents no risk for causing acidosis, which is better suited for patients with renal insufficiency. Because the stomach segment secretes acid, children with renal insufficiency may see a reversal of their baseline acidotic state. In addition, gastrocystoplasties are less prone to mucus production, have a reduced risk for infection, and allow easy implantation of ureters or Mitrofanoff neourethras.

Colocystoplasty using sigmoid colon is a common option for pediatric bladder augmentation. A 20-cm segment of sigmoid colon is selected and isolated. A colocolostomy is performed and the mesenteric window closed. In order to disrupt the strong contractions that the sigmoid colon tends to have, the segment of sigmoid is detubularized and reconfigured to provide maximal compliance and capacity. Anastomosis to the bivalved bladder is performed in a similar fashion as other intestinal segments, and a suprapubic tube and drains are left in place before closing. The advantage of the colocystoplasty lies in the fact that ureters can easily be reimplanted by an antirefluxing manner by tunneling the ureters through the teniae.

Ureterocystoplasty and autoaugmentation are alternatives to using a gastrointestinal segment for bladder augmentation. These two options are well suited for children with renal insufficiency. In both cases the tissue used to augment the bladder is urothelium, a metabolically inert tissue that consists of transitional epithelium. This epithelium is relatively impermeable and does not lead to electrolyte changes or acidosis secondary to urinary solute absorption, and it does not produce mucus. Additionally, both procedures can be performed retroperitoneally. This avoids the complications associated with entering the peritoneal cavity and is advantageous for the patient who has a ventriculoperitoneal shunt or is dependent on peritoneal dialysis.

In the ureterocystoplasty, dilated ureter is used to augment the bladder. Massively dilated ureters are often seen in the pediatric population. Not uncommonly, such ureters may be associated with kidneys that have minimal or no function. When this is the case, the dilated ureter is mobilized and detubularized. Often the entire renal pelvis of the nonfunctioning kidney can be carefully mobilized along with the ureter. Because the blood supply to the ureter approaches medially above the true pelvis and laterally in the true pelvis, care must be taken not to devascularize it during its mobilization. After it is detubularized, the ureter is reconfigured into a "U-

shaped" patch. This patch is then anastomosed to the bivalved bladder with a double-layer of running absorbable suture, the first layer interlocking. The bladder is opened in a similar manner as the ileocystoplasty. The difference is that the sagittal incision is extended posteriorly to the ureteral orifice of the dilated ureter. In this manner, the ureter is never detached from the bladder, helping to preserve its blood supply. When the kidney of the dilated ureter still functions, the distal segment of the ureter can be used to augment the bladder while the proximal ureter is either reimplanted into the bladder or anastomosed to the contralateral ureter. A major disadvantage to this technique is the small number of children requiring a bladder augmentation that fit the criteria for this procedure.

Autoaugmentation involves the excision of the detrusor muscle overlying the bladder dome, leaving the underlying bladder mucosa intact (Fig. 129.3). With time, this bladder mucosa bulges like a wide-mouthed bladder diverticulum, increasing bladder capacity and compliance. A thick, fibrous layer replaces the excised detrusor. There are two variations in the surgical technique, both with equally good results. The first involves simple excision of the detrusor muscle to create the diverticulum. With the second technique, the detrusor muscle is incised to allow the mucosa to bulge. In both cases, the detrusor edges are secured to the psoas muscle bilaterally to prevent reapproximation of the muscle. The advantages of the autoaugmentation are those of maintaining a native urothelial surface. Additionally, it is an extraperitoneal procedure with complete maintenance of bowel integrity. Complications from this operation are few, and reoperative bladder reconstruction does not seem to be hindered. However, there are concerns over the limited improvement in bladder capacity and compliance, and it is generally not recommended for children with low-capacity, high-pressure bladders.

Because of concerns over minimal improvements in bladder volume and compliance secondary to fibrosis after the autoaugmentation procedure, some surgeons have been covering the exposed bladder mucosal surface with a demucosalized enteric segment in an attempt to decrease the amount of scarring. Different enteric segments have been tried with varying success. Demucosalized stomach and sigmoid colon placed over the bladder mucosa after autoaugmentation seem to offer some promise, but further investigation is needed.

POSTOPERATIVE CARE

As with any patient that undergoes major abdominal surgery, postoperative care of the bladder-augmented patient can be complex. Immediately after surgery, elec-

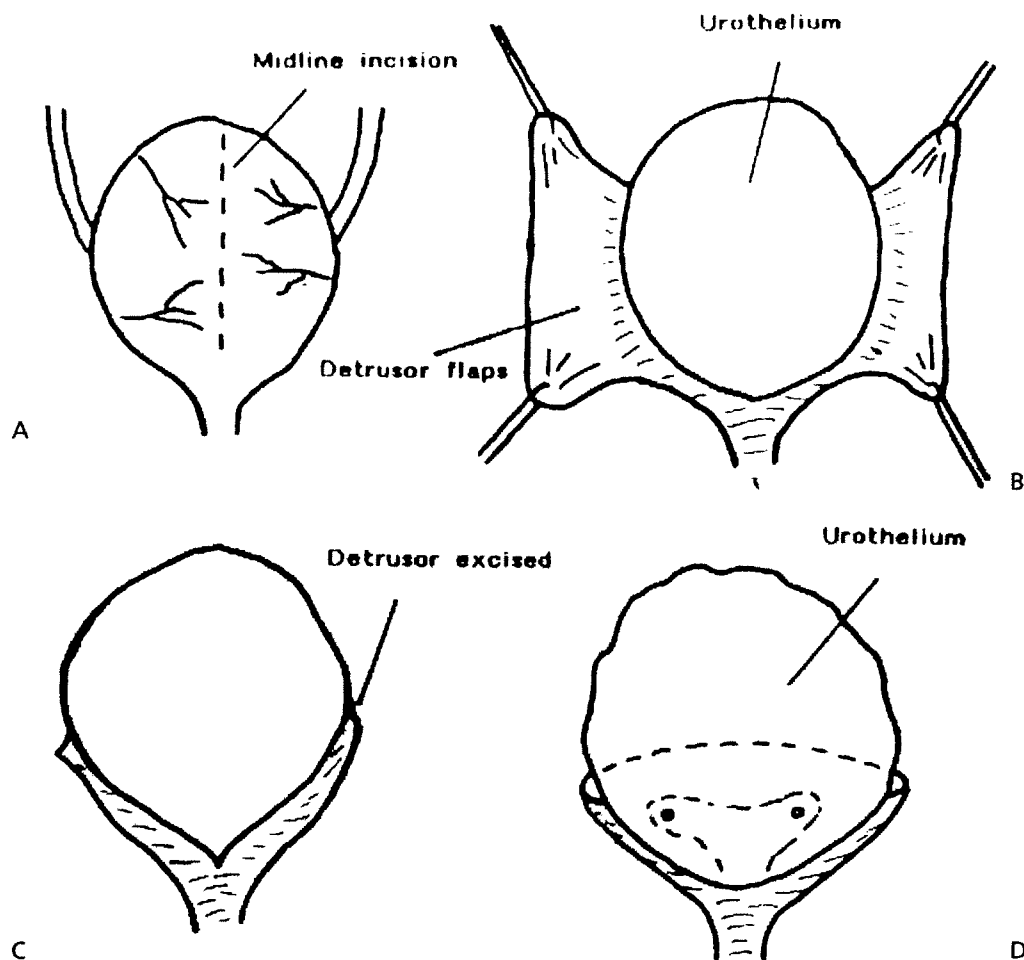


FIGURE 129.3. Autoaugmentation. **A:** Midline incision in the detrusor muscle, leaving bladder mucosa intact. **B:** The detrusor muscle is dissected away from underlying urothelium. **C:** The mobilized detrusor muscle is excised. **D:** Final appearance. Only a rim of detrusor muscle surrounding the trigone remains. The remainder of the urothelium is devoid of its original muscular support. (Reprinted from Sheldon CA, Bukowski T. Bladder function. In: Rowe MI, O'Neill JA, Grosfeld JL, et al., eds. *Essentials of pediatric surgery*. St. Louis: Mosby Year Book, 1995:731-747; with permission.)

trolyte and blood counts must be followed carefully, especially in the face of bleeding and third-space fluid shifts. Intravenous fluids should be sufficient to compensate for intravascular deficits. The patient should be maintained on bowel rest, with nasogastric tube (NGT) decompression and broad-spectrum antibiotic coverage. Urine output from the urethral catheter, SPT, and Mitrofanoff catheter (if present), as well as drain and NGT output, should be monitored carefully. Parenteral nutrition should be considered, because some of these children are often without oral nutrition for 5 days or more. Depending on which augmentation technique is used, it may be necessary to gently irrigate the bladder free of mucus several times each day. The NGT is removed after the resolution of ileus, and the diet is slowly advanced. The patient can be discharged from the hospital only after being without fever, tolerating a regular diet, and receiving the supplies and teaching needed for

proper tube care. They go home on oral antibiotics, with their SPT, urethral catheter, and Mitrofanoff catheter to drainage. Typically, our patients are followed carefully postoperatively by telephone, and ultrasonography and a panel of renal tests are performed at the 2-week follow-up visit. Approximately 4 weeks postoperatively, indwelling catheters are removed and intermittent catheterization is resumed. The time between catheterizations is gradually increased from every 3 hours to four times a day.

COMPLICATIONS

Augmentation cystoplasty can be a highly rewarding undertaking for both patient and surgeon. However, it can be challenging and is fraught with many potential complications. These include metabolic derangements, alterations in

gastrointestinal function, urinary tract infection (UTI), urolithiasis, malignancy, bone demineralization, and spontaneous bladder rupture. These complications are specific to bladder augmentation, and are in addition to those normally encountered with major abdominal surgery. The risk of a complication varies according to the type of augmentation performed and is outlined in Table 129.1.

Metabolic derangements are commonly encountered after bladder augmentation with a gastrointestinal segment. The determinants of the character and severity of these metabolic derangements are a consequence of the nature of the segment used, the absorptive surface area, the dwell time, and the metabolic reserve of the patient. Intestinal segments tend to absorb many urinary solutes that can lead to alterations in acid-base status, electrolyte abnormalities, hyperammonemia, and eventually bone demineralization. The major solutes absorbed by ileum and colon are ammonium and chloride. Patients with normal renal function compensate by excreting this additional acid load. However, in patients with compromised renal function, this absorption can quickly lead to an acidotic state, marked by hyponatremia and hyperchloremia. Buffering of even a slightly acidotic state is provided by bone, which can lead to demineralization and secondary hyperparathyroidism. Jejunum is rarely used for augmentation cystoplasty. This is because it often leads to the unique metabolic state of a hyponatremic, hypochloremic, hyperkalemic, and metabolic acidosis. Patients with such metabolic derangements present with fatigue, weakness, anorexia, and polydipsia. However, perhaps the most worrisome effect seen in children is the retardation of growth and development, especially in those children with compromised renal function.

Gastrocystoplasty leads to a distinct metabolic state. The stomach segment secretes chloride ions and acid into the augmented bladder, occasionally leading to a hypochloremic metabolic alkalosis. This often has the beneficial effect of improving the acidotic state seen in children with renal insufficiency. However, there are consequences to using stomach. These children seem to be vulnerable to dehydration during episodes of viral gastroenteritis. This is especially true in children with renal insufficiency. Additionally, gastrocystoplasty can result in the hematuria-dysuria syndrome, a syndrome marked by pain and urinary bleeding due to erosion of urothelium by acidic urine. It is more commonly seen in children who are incontinent and have exposure of the urethra to acidic urine. It is also seen in children with renal insufficiency who do not produce enough urine to dilute the acid load secreted by the stomach segment. This syndrome is usually mild and is treatable with histamine blockers or proton pump blockade. Infrequently, the symptoms can be intractable, and may require consideration of another augmentation modality.

Many of the complications brought on by augmentation cystoplasty are secondary to resection of the bowel. Bowel obstruction is uncommon after augmentation cystoplasty,

occurring in approximately 3% of patients. The incidence of obstruction is the same regardless of which bowel segment is used. In addition, vitamin B₁₂ deficiency can occur with bladder augmentation in two separate ways. Intrinsic factor (IF) is responsible for binding vitamin B₁₂ in the gut and its eventual absorption in the terminal ileum. Because IF is produced by the parietal cells in the fundus of the stomach, gastrocystoplasty can in theory cause a shortage of IF production and thus vitamin B₁₂ deficiency. Because distal ileum is the sole site of vitamin B₁₂ absorption, its resection during an ileocystoplasty also can result in a malabsorption of vitamin B₁₂. A deficiency of vitamin B₁₂ can lead to megaloblastic anemia, although this may take years to occur.

Diarrhea can become a significant problem in children after bladder augmentation using a segment of intestine. Resection of the ileocecal valve is most often implicated. Resection of the valve can do this in two ways. First, it causes a decreased transit time from the ileum to the cecum. Second, it allows backflow of bacteria from cecum to ileum, causing bacterial overgrowth and malabsorption of fat and vitamin B₁₂. This is especially true in children with neurogenic dysfunction of bladder and bowel who typically rely on chronic constipation for fecal continence. It is estimated that up to 10% of children undergoing resection of the valve will have problems with chronic diarrhea. Resection of large segments of ileum also can result in diarrhea as a consequence of decreased bile acid reabsorption. This overflow of bile acid into the colon results in both a secretory diarrhea as well as steatorrhea. It would seem prudent not to resect large segments of ileum or the ileocecal valve for augmentation cystoplasty in children, especially those with preexisting risk factors.

Another risk of augmentation cystoplasty is that of tumor formation. Most of our experience comes from studying the ureterosigmoidostomy model, where the risk of malignancy near the site of ureterocolonic anastomosis is increased several hundred times above normal. These tumors are usually adenocarcinomas, although benign tumors, and rarely a transitional cell carcinoma, have been documented. This is believed to be a result of urinary nitrosamines, known to have a carcinogenic influence on the colon, which originate from the mixing of feces and urine. Tumor development has been seen in the augmented bladder as well. The exact incidence is unknown at this point, but it is clearly much greater than in the general population, and may not be influenced by which gastrointestinal segment is used to augment the bladder. Because of the long latency period of these tumors (>20 years), periodic endoscopy is warranted for the lifetime of the patient.

Patients have a higher incidence of both upper tract and reservoir bladder calculi after augmentation. Although varied in composition, struvite stones are most commonly seen. There are several reasons for this increased incidence. Stasis of urine in a dependent reser-

voir is a contributing factor, and the presence of mucus in the reservoir can further act toward stone propagation. Mucus also can serve as a nidus for infection, often by urea-splitting bacteria such as *Proteus* or *Klebsiella*. Metabolic changes in these patients and periods of dehydration also contribute to stone formation. Another causative agent is a foreign body, such as nonabsorbable sutures or staples used during the reconstruction. Care should be taken to avoid as many of these factors as possible, and the patient should be instructed to completely empty their bladder with CIC. UTIs caused by stone-forming organisms should be treated aggressively with antibiotics. Metabolic abnormalities and bouts of dehydration should be addressed and treated appropriately.

One of the most treacherous complications of bladder reconstruction is perforation of the augmented bladder. It is one of the most common causes of an acute abdomen in these patients, and suspicion should be high when a child with an augmented bladder presents with abdominal pain, distension, fever, decreased urine output, and sepsis. The etiology of perforation often goes undiscovered, and no single bowel segment seems to be at greater risk than another. Cystography can be diagnostic, demonstrating extravasation of contrast from the augmented bladder. However, there is a significant incidence of false-negative results with cystography, and if suspicion remains high, computed tomography with bladder contrast can be more definitive. Surgical explo-

ration and repair are the standard course for bladder rupture management after augmentation cystoplasty.

SUGGESTED READINGS

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