

Ureterosigmoidostomy and obstructive uropathy

Ofer Yossepowitch* and Jack Baniel

SUMMARY

Background A 19-year-old mentally retarded man with failed exstrophy repair and ureterosigmoidostomy urinary diversion presented with high fever, vomiting and right-flank pain of 2 days' duration. Past medical history was notable for a left nephrectomy to treat an infected staghorn calculus in a poorly functioning kidney. Physical examination revealed pyrexia and right-flank tenderness.

Investigations Physical examination, renal function tests, electrolyte and metabolic assessment, urine and blood cultures, abdominal CT, ANTEGRADE PYELOURETEROGRAPHY, sigmoidoscopy and histopathology.

Diagnosis Ureterosigmoidostomy complicated by acute pyelonephritis, obstructive uropathy, recurrent urinary tract infections, renal impairment and the development of renal stones and metabolic acidosis.

Management Fluids, intravenous antibiotics, bicarbonate and potassium supplementation, and redirection of ureterosigmoidostomy to an ileal conduit.

KEYWORDS colon malignancy, metabolic disorders, postobstructive diuresis, ureterosigmoidostomy, urinary diversion

CME

This article offers the opportunity to earn one Category 1 credit towards the AMA Physician's Recognition Award.

THE CASE

A 19-year-old mentally retarded man presented with high fever, vomiting and right-flank pain of 2 days' duration. He had been born with classical bladder exstrophy and had previously undergone several early reconstructive trials at a different institute. At 5 years of age, a continent urinary diversion with bilateral URETEROSIGMOIDOSTOMY was created owing to severe anatomic and functional impairment of the lower urinary tract. The patient's mother reported that he had suffered frequent episodes of febrile urinary tract infections thereafter, requiring hospital admission and intravenous antibiotic treatment. According to his medical records, 4 years prior to his present referral the patient underwent left nephrectomy to treat an infected staghorn calculus in a poorly functioning kidney. Nevertheless, he continued to have recurrent urinary tract infections and was treated with antibiotics on a prophylactic basis. The current admission to the urology department was the result of a presumed recurring episode of pyelonephritis.

Physical examination revealed a high fever (39.5°C), right-flank tenderness, a lower-abdominal scar, and a distorted penis with no conspicuous urethral meatus. Laboratory results were remarkable for a raised white blood cell count of 22.5×10^9 cells/l, an elevated creatinine level of 2.4 mg/dl, and a slightly elevated anion-gap metabolic acidosis (pH 7.12, bicarbonate 10 mEq/l, base excess -16 mEq/l, chloride 121 mEq/l, sodium 139 mEq/l) associated with severe hypokalemia (2.9 mEq/l). Abdominal ultrasound followed by non-contrast-enhanced CT demonstrated significant right hydro-ureteronephrosis with a large amount of fluid filling a wide and tortuous sigmoid colon along with part of the descending colon (Figure 1A,B). A rectal tube was placed, which drained 800 ml of turbid urine, and a percutaneous nephrostomy

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GLOSSARY**ANTEGRADE****PYELOURETEROGRAPHY**

Radiologic study of the renal collecting system and ureter involving opacification by percutaneous antegrade injection of contrast media

URETEROSIGMOIDOSTOMY

A form of urinary diversion using the sigmoid colon as a reservoir and the anal sphincter as the continence mechanism

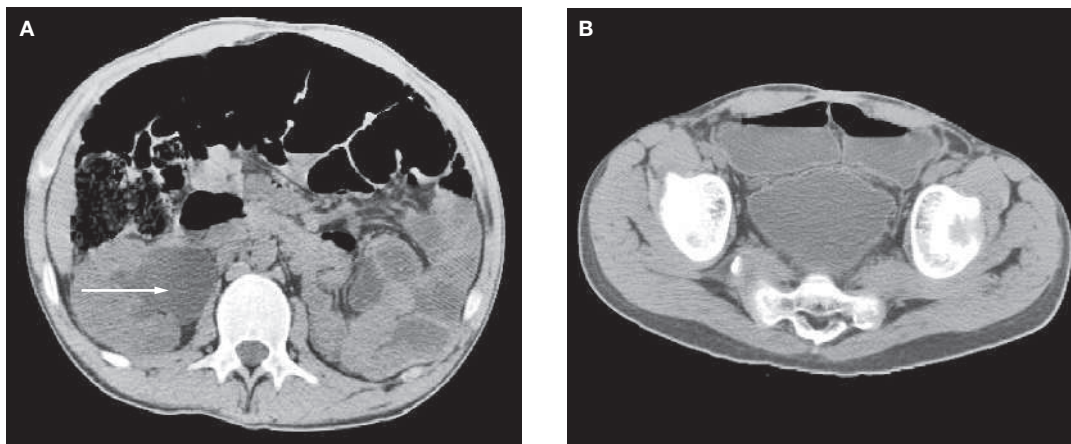


Figure 1 Abdominal CT scans of a patient with ureterosigmoidostomy and obstructive uropathy, showing (A) marked dilatation of the right renal pelvis (white arrow) and (B) a large amount of urine filling a wide and tortuous sigmoid colon along with part of the descending colon.

was inserted. The patient had lost 4.5 L of urine through the nephrostomy tube within four hours of drainage of his single kidney. Fluid maintenance with a solution of 5% dextrose in 0.45% saline was initiated, matching 50% of the patient's urinary output, with supplements of intravenous bicarbonate and potassium. The patient was followed up with repeated weighing at 12-hour intervals and serial measurements of venous blood gases, creatinine level and electrolyte level. Stability was achieved only 48 h later, after which time the patient could be maintained on oral intake. Urine and blood cultures grew *Escherichia coli*, and appropriate antibiotics were administered according to culture sensitivity. On the 4th day of hospitalization the fever subsided, creatinine level normalized, and adequate oral supplementation of sodium–potassium citrate was initiated.

To further investigate the patient's ureterosigmoidostomy, antegrade pyeloureterography was performed, disclosing a nondilated collecting system, a slightly tortuous ureter, and an intact ureterointestinal junction allowing rapid passage of the contrast medium into the bowel (Figure 2A,B). Short colonoscopy ruled out a mucosal abnormality at the anastomotic site or at other areas along the left colon. At this point, the patient and his family were advised that rediversion of the ureterosigmoidostomy to a noncontinent conduit was necessary because of a high likelihood of renal deterioration, recurrent urinary tract infections, metabolic abnormalities, and the potential risk of intestinal malignancy. Given the patient's intellectual

ability, a comprehensive urologic, psychiatric and social counseling session was held, and consent was provided by the patient and his parents. An ileal-conduit urinary diversion was created with complete excision of the previous ureterointestinal anastomoses along with a sigmoid cuff. Pathology was negative for dysplasia or malignancy and convalescence was uneventful.

DISCUSSION OF DIAGNOSIS AND TREATMENT

Discussion of postobstructive diuresis

The postobstructive diuresis (POD) phenomenon refers to the marked polyuria that can follow the relief of bilateral ureteral obstruction or the obstruction of a solitary kidney. Although there is no consensus regarding the degree of polyuria defining POD, it is generally agreed that patients with a urinary output of greater than 200 ml/h for two consecutive hours should be closely monitored for excessive fluid and electrolyte losses, which, if not properly replaced, can lead to severe dehydration and life-threatening electrolyte imbalance.

The mechanisms responsible for POD have been classified as physiologic and pathologic. Physiologic POD is attributed to the excretion of retained urea, sodium, and water, which mainly reflects an osmotic diuresis. The pathologic component is caused by the inability of the kidney to concentrate the urine, secondary to defective generation of a medullary solute gradient and to an impaired response of the distal convoluted tubules and the collecting ducts to endogenous antidiuretic hormone

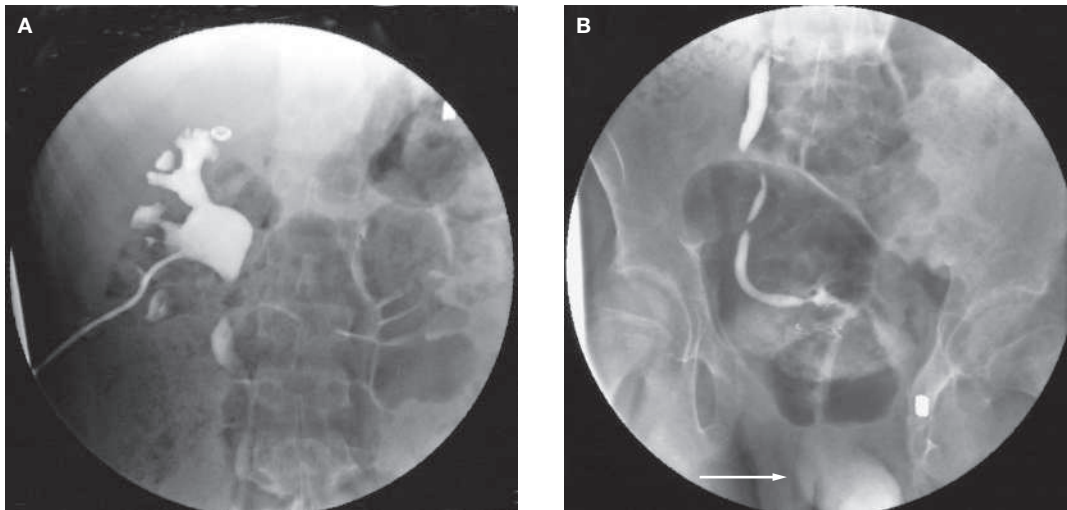


Figure 2 Antegrade pyeloureterography of a patient with ureterosigmoidostomy and obstructive uropathy, revealing (A) a nondilated collecting system and (B) an intact ureterosigmoid junction allowing rapid passage of the contrast medium into the bowel. Arrow indicates urine within the rectum.

(i.e. vasopressin-insensitive diabetes insipidus). The medullary osmotic gradient, which is essential for the regulation of water balance, enables the transport of water and electrolytes from a diluted filtrate within the ascending limb of the loop of Henle and the medullary collecting ducts into the hypertonic medullary interstitium. After relief of the obstruction, the increased medullary blood flow through the vasa recta 'washes-out' the medullary tonicity and impairs its ability to concentrate the urine. Other factors implicated in the pathogenesis of POD include a proximal tubular defect¹ and the augmented activity of atrial natriuretic peptide.²

POD requires careful assessment and management of fluid and electrolyte balance, including the correction of deficits of sodium chloride (with 0.45% NaCl), bicarbonate, potassium, and water. POD must be distinguished from the brisk physiologic diuresis of retained fluid and from iatrogenic diuresis resulting from excessive fluid administration. Therefore, when prolonged diuresis is observed, the rate of fluid replacement should be decreased, guided by clinical assessment of the intravascular volume and changes in urine output and body weight.

Our patient developed a marked diuresis that required prolonged intravenous fluid replacement following insertion of a nephrostomy tube with concomitant decompression of the lower intestinal tract. The obstructive uropathy in this case manifested as significant hydronephrosis

on presentation and protracted diuresis with normalization of serum creatinine subsequent to drainage of the kidney. Antegrade pyeloureterography failed to demonstrate a significant obstruction at the ureterointestinal anastomosis. Therefore, the obstructive uropathy might have been attributed, at least in part, to chronic fluid retention within the colon, resulting in compression of the ureteral outlet and exposure of the kidney to high filling pressures (similar to the obstructive uropathy associated with benign prostatic hyperplasia and chronic urinary retention). Tubular damage secondary to chronic pyelonephritic changes might also have contributed to the impaired ability of the single kidney to concentrate urine.

Discussion of ureterosigmoidostomy

Ureterosigmoidostomy enables the excretion of urine by means of rectal evacuation. Although this approach has now been abandoned by most urologists because of the complications associated with the mingling of the fecal and urinary streams, it is simpler to perform than other types of continent urinary reservoirs which require complex bowel refashioning. To ensure that only appropriate candidates are selected, several exclusion criteria for ureterosigmoidostomy have been advocated, including preoperative renal insufficiency, previous extensive pelvic irradiation, dilated ureters, an incompetent anal sphincter and colon disease.³⁻⁵ In addition,

Table 1 Ureterosigmoidostomy: long-term complications and management.

Complication	Management
Upper urinary tract deterioration	Assess for reflux nephropathy (DMSA scan for renal cortical scars) and anastomotic stricture (renal ultrasonography, IVP or antegrade pyeloureterography). Consider conversion to a noncontinent diversion.
Metabolic disorders	Routine assessment of serum electrolytes and venous blood gases. Administer alkalinizing agents and supplement potassium as required.
Colon carcinoma	Annual short colonoscopy beginning on the 10 th anniversary of the original operation.
Recurrent urinary tract infections	Treatment of asymptomatic bacteriuria is of debatable benefit. Assess for ureteral obstruction and stone disease and treat accordingly.
Nephrolithiasis	Complete stone eradication is mandatory.
Osteomalacia	Assess bone mineral density. Treat metabolic acidosis and administer dietary supplements of calcium and vitamin D.

DMSA, dimercaptosuccinic acid; IVP, intravenous pyelography.

owing to the long-term sequelae, most urologists reserve this solution for older patients. A recent modification of the classical technique, which utilizes a detubularized sigma (the Mainz-Pouch II procedure), has been reported to serve as an acceptable alternative to continent urinary diversion, providing satisfactory quality of life with reduced morbidity.⁶

Ureterosigmoidostomy patients require stringent follow-up to address the potential deleterious complications (see Table 1).

Renal failure

Renal deterioration has been reported to afflict as many as 70% of patients with bladder exstrophy treated with ureterosigmoidostomy, with 10% of the patients dying of renal failure and 23% losing a kidney.⁷ Other studies have found a lower incidence,^{4,5,8} which may reflect a variance in preoperative renal function among the different series. The mechanisms leading to upper-urinary-tract compromise have not been completely elucidated, and include pyelonephritic changes, the formation of renal stones, and partial obstruction due to ureterointestinal anastomotic stricture. It is imperative that these factors be assessed and corrected when progressive renal failure is encountered.

Metabolic disorders

The hallmark metabolic abnormality in patients with ureterosigmoidostomy is hyperchloremic, hypokalemic metabolic acidosis. The acidosis is caused by the excessive reabsorption of ammonium chloride through the intestinal mucosa into the blood, whereas the hypokalemia

and the depletion of total body potassium are believed to result from renal potassium wasting as a consequence of renal damage and osmotic diuresis.⁹ Unlike the small-bowel mucosa, the colonic mucosa lacks the ability to blunt the potassium loss. This explains the high incidence of hypokalemia in ureterosigmoidostomy patients compared with patients with urinary diversions using ileal segments. Treatment includes alkalinizing agents, chloride-transport blockers, chlorpromazine, and potassium supplementation. Metabolic acidosis should be balanced in these patients cautiously (particularly if acquired over a long period of time) to avoid an overly rapid correction, which can result in life-threatening alkalosis. It should also be emphasized that the acidosis might mask a true depletion of total body potassium due to potassium shift from within the intracellular compartment. Therefore, if the acidosis is corrected without attention to potassium replacement, serum levels of potassium may rapidly decrease, leading to significant morbidity.

Colon cancer

Neoplasia at or adjacent to the anastomosis of the ureters and colon may develop in as many as 40% of patients on long-term followup.^{10–14} Approximately half of the tumors are adenocarcinomas of the colon and the rest are benign polyps.¹¹ The latency period between ureterosigmoidostomy formation and tumor diagnosis ranges between 3 years and 53 years (mean 20 years).^{11,15} The pathways of carcinogenesis are not completely understood, but changes in mucin production, an increase in ornithine decarboxylase

activity, chronic infection with formation of nitrosamines, and chronic inflammation are all considered to be contributing factors.¹⁰

A recent consensus report on colorectal cancer screening in patients with uretero-sigmoidostomy (or the more recent variants) recommended that all patients undergo flexible sigmoidoscopy to visualize the colon up to and just beyond the higher ureteric orifice.¹³ Screening should begin 10 years after the original operation and be repeated annually thereafter. Patients in whom the uretero-sigmoidostomy was subsequently converted to an alternative diversion should still be followed up by sigmoidoscopy unless the ureteric anastomoses were completely excised.¹³

Urinary tract infections and nephrolithiasis

Interposition of the bowel into the urinary tract is associated with an increased risk of bacteriuria, bacteremia, and septic episodes. These are particularly prominent when the fecal and urinary streams are combined.³ Deterioration of the upper urinary tract is more likely to occur when urine cultures are dominant for *Proteus* or *Pseudomonas spp.* Therefore, patients with pure cultures of these organisms should be treated aggressively, whereas those with mixed cultures may generally be observed.³

Renal stones are commonly composed of struvite and form consequent to recurrent infections with *Proteus mirabilis* or other urea-splitting bacteria.¹⁶ Additional mitigating factors for the formation of calculi after ureterosigmoidostomy include metabolic acidosis with resultant hypercalciuria and hypocitraturia.¹⁶ Renal stones in these patients often serve as a nidus for recurrent infections, and so every effort to ensure their complete eradication must be made.

The patient's present hospital admission, combined with his past medical history (remarkable for the entire spectrum of adverse effects of ureterosigmoidostomy), indicated that rediversion was necessary for his long-term well being. Given the patient's failed exstrophy repair, previous nephrectomy, recurrent infections and metabolic imbalance, we considered an orthotopic or continent cutaneous diversion to be inappropriate. We also felt that his impaired intellectual status would make a change in voiding habits impractical. From

a technical point of view, we could have made use of the long and redundant sigmoid colon as a conduit in order to separate the fecal and urinary streams; however, to diminish the risk of future colonic malignancy, we proceeded with the creation of an ileal conduit urinary diversion with complete excision of the ureterosigmoid anastomoses.

CONCLUSION

The introduction of contemporary forms of urinary diversion has led to the general disuse of classic ureterosigmoidostomy. This case illustrates the potential long-term problems encountered in these patients, and emphasizes the importance of close medical surveillance.

Competing interests

The author declared he has no competing interests.

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