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Rectourethral and rectovesical fistula as serious and rare complications after Hirschsprung disease operation: Experience in seven patients

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ABSTRACT

Aim: To present the features and treatment of rectourethral fistula (RUF) and rectovesical fistula (RVF) after Hirschsprung disease (HD) operation.
Methods: A retrospective analysis was performed on patients with RUF and RVF after HD operation, who received repair surgery from a single surgeon between January 2005 and December 2019. Bowel function was assessed using the Rintala score.
Results: Seven patients were included. Six patients were referred to us after transanal endorectal pull-through (TEPT) in other centers; one RVF patient had fecal diversion at admission. Bladder-neck injury was detected during redo TEPT in our hospital in the remaining one patient and instant repair was given. 11 days later, RVF and sepsis were detected.
Fecal and urine diversion was performed immediately. The fistula openings were prostatic urethra (3), membranous urethra (2), bladder triangle (1), and bladder-neck (1). Anastomotic stricture (4), bladder stone (3), hydronephrosis and ureterovesical junction obstruction (2), pelvic infection (2), distal colonic dilatation (1) and other fistulas (1) were identified before repair surgery. Fecal and urine diversion was performed before repair surgery in one RUF and one RVF patient respectively, to treat pelvic infection owing to fecal or urine leaks and accompanying problems. Five RUFs were repaired by transperineal approach, and two RVFs were repaired using the transabdominal and transanal approach respectively. Four patients with anastomotic stricture underwent redo TEPT with simultaneous fistula repair. Aside from one RUF patient, the fistula in all patients successfully resolved. Median follow up time was 81 months (range, 5–116 months). No recurrence was observed. Median bowel function score was 17.5 (range, 17–18).
Conclusion: The location of fistula, presence of anastomotic stricture and the association of urinary complications have significant impacts on the treatment strategy in such rare complications after TEPT for HD.
The type of study: Treatment study.
Level of evidence: Level IV.

Rectourethral fistula (RUF) and rectovesical fistula (RVF) after Hirschsprung disease (HD) operation are less common complications. Related literature is limited, most of which are case reports [1–9]. Because of the relatively low incidence of RUF and RVF after HD operation, few pediatric surgeons have adequate understanding of this disease and lack the necessary experience to establish a standard approach for surgical repair. Moreover, surgical repair of postoperative RUF and RVF is a formidable challenge because of pelvic fibrosis and adhesions resulting from previous surgery and other accompanying complications [3].

By reviewing our cases, we aim to present the features and treatment of RUF and RVF after HD operation.

1. Materials and methods

RUF and RVF patients after HD operation who received repair surgery from a single surgeon between January 2005 and December 2019 were included in this study. Their clinical data, inclusive of clinical presentations, previous histories of pull-through (PT) operations, preoperative imaging and digital rectal examination (DRE) findings, presence of fecal or urine diversion, intraoperative findings, and operative procedures, were reviewed. Follow-up data were gathered via telephone interviews. Bowel function was evaluated according to the seven-item Rintala bowel function score (BFS). A BFS of ≥ 18 was considered to indicate normal bowel function [10].

Before repair surgery, RUF or RVF was diagnosed via voiding cystourethrography (VCU) and barium enema (BE). DRE, ultrasonography, X-ray and intravenous pyelography (IVP) were performed to detect the presence of accompanying complications.

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If anastomotic stricture was detected by DRE, redo transanal endorectal pull-through (TEPT) with laparotomy was performed with simultaneous fistula repair; otherwise, only fistula repair was performed. RUF was repaired using transperineal approach and RVF was repaired using transabdominal approach.

Before repair surgery, a urethral catheter was routinely inserted. A ureteral catheter was then inserted into the fistula through the urethral defect, which was pulled out through the anus for fistula localization (as shown in Fig. 1). If the fistula opening on the rectum could be exposed by anus, a ureteral catheter could also be inserted through it. If these failed, cystoscopy through the urethra was performed to locate and insert a ureteral catheter through the fistula.

Patients with RUF received repair surgery per the following. Patients were placed in the lithotomy position. An incision was made at the perineal body and anterior rectal wall, until exposure of the fistula opening was achieved (as shown in Fig. 2). An incision was made around the fistula opening on the rectum. The fistula tract was dissected until the urethral catheter was well exposed and then excised. Interrupted sutures with 5-0 absorbable stitches were used to close the defect (as shown in Fig. 3). The rectal wall was then sutured, or the colon would be pulled down to cover the repaired region. All incisions were sutured in situ, and the urethral catheter was left in place for 2 weeks.

This study was conducted in accordance with the declaration of Helsinki and approved by the Ethics Committee of our hospital (2020-Z-007).

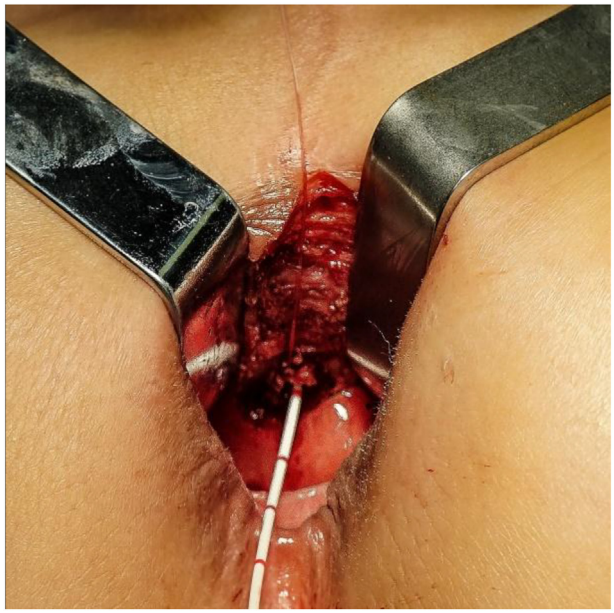

Fig. 2. Incision of the perineal body and anterior rectal wall until opening of the fistula (the place where a ureteral catheter passed through the rectum).

2. Results

Seven patients (RUF 5 and RVF 2) were included and were all boys. Their clinical details are summarized in Table 1. Six patients were referred to our hospital from other centers, and the incidence of RUF and RVF after HD operation could not be obtained. The remaining one patient developed RVF after redo TEPT carried out in our hospital, representing 0.13% of all HD operations.

Two patients (patient 1 and 2) had undergone 2 previous TEPT surgeries each, and the remaining five patients had all undergone 1 previous TEPT surgery. The median age at TEPT surgery was 1 year (range, 40 days to 8.5 years). The median time between TEPT and the onset of the first symptom of RUF or RVF was 3 months (range, 1 day to 5 years). The children presented with various characteristics and the details are shown in Table 1. One RUF patient (patient 4) had undergone prior unsuccessful fistula repair before referral.

Five patients developed accompanying complications before repair surgery, three of them required fecal and/or urine diversion. One patient (patient 1) received redo TEPT in our hospital, and bladder-neck injury was detected during redo TEPT. Instant repair was given via laparotomy without fecal diversion and suprapubic cystostomy. 11 days postsurgery, urine was discovered passing per the rectum and feces were discovered in the urethral catheter. Prior to that, the urethral catheter had been blocked by blood clots. An RVF in the bladder neck was detected, as well as sepsis caused by anastomotic leakage. An ileostomy and a suprapubic cystostomy were performed immediately to relieve sepsis. An anastomotic stricture subsequently developed in this RVF patient.

The other RVF patient (patient 2) had undergone colostomy and suprapubic cystostomy to relieve sepsis caused by anastomotic leakage

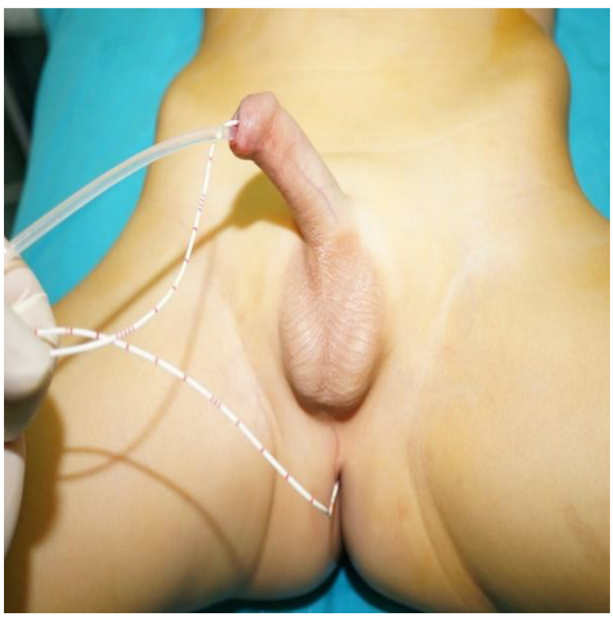

Fig. 1. A ureteral catheter placed to locate the fistula.

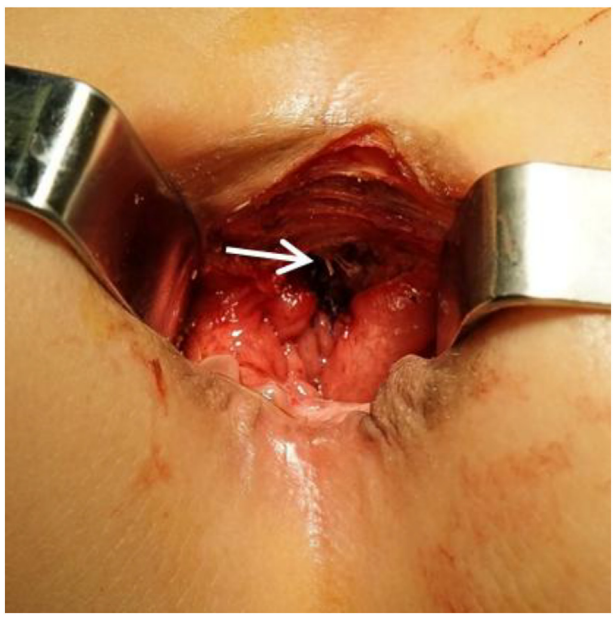

Fig. 3. Closing of the defect, the arrow showing the closed urethral defect.

Table 1
Patients' data.

Patient	Age at PT	Onset	Symptoms	Anastomotic stricture	Fistula locations	Stone	Hydronephrosis	Diversion	Age at repair	Repair surgery	Defect	Outcome	Age at follow-up	Rintala
1	3.5 yrs	11 ds	Sepsis, fecaluria and urinary passage [a]	Yes	Bladder-neck	No	No	Fecal, urine [b]	4.4 yrs	Transabdominal, TEPT	1–2 cm	Resolved	14.8 yrs	18
2	8.5 yrs	2 ds	Sepsis and urinary passage [c]	Yes	Bladder triangle [d]	Yes	Yes	Fecal, urine [e]	16.7 yrs	Transanal, TEPT	1–2 cm	Resolved	25.1 yrs	17
3	40 ds	1 yr	Pneumaturia	Yes	Membranous urethra	Yes	Yes	Fecal [f]	2 yrs	Transperineal, TEPT	≤1 cm	Resolved	9.1 yrs	17
4	1 yr	2 yrs	Fecaluria	No	Membranous urethra	No	No	No	13.5 yrs	Transperineal	≤1 cm	Resolved	20.8 yrs	18
5	11 mths	3 mths	Fecaluria	Yes	Prostatic urethra	No	No	Fecal [g]	8.3 yrs	Transperineal, TEPT	≤1 cm	Resolved	10.8 yrs	18
6	1.8 yrs	5 yrs	Urinary passage [a]	No	Prostatic urethra	Yes	No	No	9.8 yrs	Transperineal	1–2 cm	Failed to resolve	10.5 yrs	
7	9 mths	1 d	Fecaluria	No	Prostatic urethra	No	No	No	10.6 yrs	Transperineal	≤1 cm	Resolved	11.1 yrs	17

[a] Passage of urine per rectum.
[b] At 11 days after redo TEPT.
[c] Passage of urine per rectum and abdominal drainage tube.
[d] Located in the inner surface of the bladder base, between the ureteral orifice and the urethra.
[e] At 8 days after redo TEPT.
[f] Prior to repair surgery.
[g] At the time of repair surgery.

at 8 days after redo TEPT in other center. For reasons unknown to our hospital, the suprapubic catheter was removed six months later, leading to pelvic infection owing to urine leakage, as well as the development of fistulas from the bladder to the abdominal wall. Six years after removal of suprapubic catheter, the patient was referred to our hospital. An RVF in the bladder triangle was detected. Meanwhile two fistulas from the bladder to the abdominal wall (as shown in Fig. 4), bilateral hydronephrosis, ureterovesical junction obstruction, two big bladder stones (as shown in Fig. 5), and anastomotic stricture were detected. Redo suprapubic cystostomy was performed soon after the referral, about one year before repair surgery. The two big bladder stones were removed during urine diversion and the two fistulas from the bladder to the abdominal wall resolved spontaneously after urine diversion. Unfortunately, bilateral hydronephrosis and ureterovesical junction obstruction, possibly attributed to repeated inflammation affecting the ureteral orifice, did not resolve after urine diversion and necessitated ureteral reimplantation during subsequent RVF repair surgery.

Unilateral hydronephrosis and ureterovesical junction obstruction were also detected in one RUF patient (patient 3) before repair surgery. Pelvic infection, anastomotic stricture, and small bladder stone were also detected. The pelvic infection, owing to anastomotic leakage, compressed the ureterovesical junction, and led to ureterovesical junction obstruction and subsequent hydronephrosis (as shown in Fig. 6). An ileostomy was performed soon after the referral, about one year before repair surgery. Besides anastomotic stricture, the above symptoms resolved spontaneously after fecal diversion in this RUF patient. The small bladder stone did not receive intervention.

Although accompanying complications were detected in another two patients (patient 5 and 6), no surgical intervention was given before repair surgery. Anastomotic stricture and distal colonic dilatation (as shown in Fig. 7) were detected in one patient (patient 5). The distal colonic dilatation was removed during subsequent repair surgery. Small bladder stone, which was confirmed as fecal stone via subsequent cystoscopy, was detected in another one patient (patient 6) but did not receive intervention.

The median age at repair surgery was 9.8 years (range, 2–16.7 years). Five RUF patients received repair surgery using the transperineal approach described above. One RVF (patient 1) was repaired using the transabdominal approach. The other RVF patient (patient 2) underwent laparotomy but repair could not be made owing to

Fig. 4. VCU showing urine leakage from the abdominal wall (two fistulas, red arrow) and the rectum.

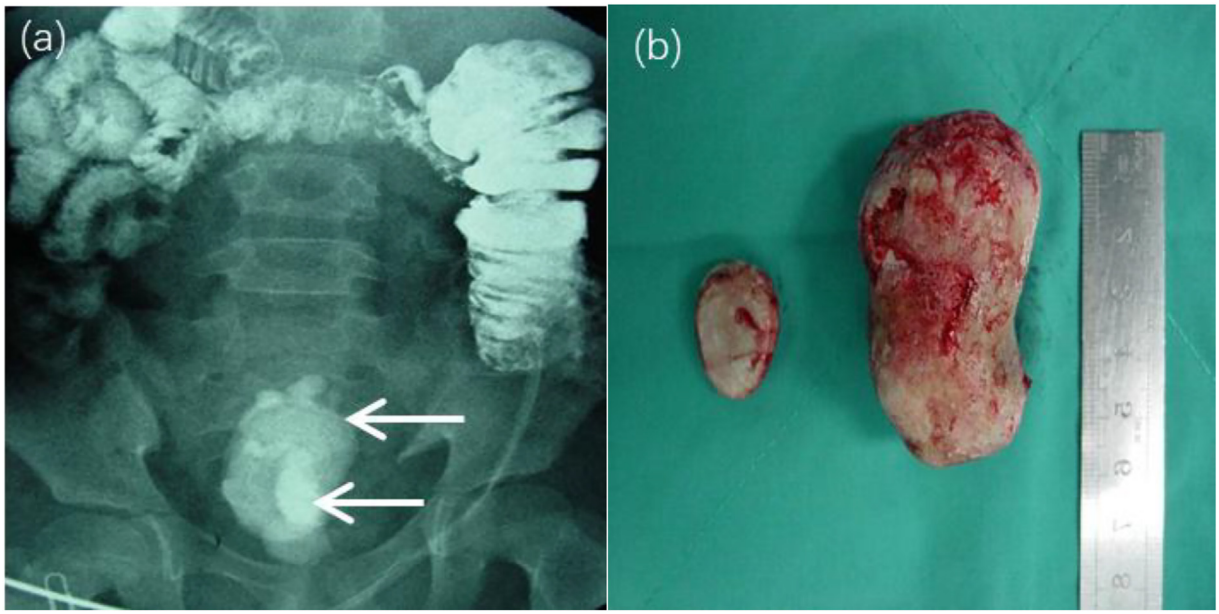

Fig. 5. X-ray demonstrating two bladder stones (white arrow) (a) and the two stones removed from bladder (b).

dense adhesions, which were aggravated by removal of suprapubic catheter before referral. After establishment of the pelvic tunnel, he received repair using the transanal approach. Defects of the urethra and the bladder identified during repair surgery are shown in Table 1.

Owing to the presence of anastomotic stricture, two RUF patients (patient 3 and 5) and two RVF patients (patient 1 and 2) underwent simultaneous redo TEPT with fistula repair. Ileostomy was performed in one of these patients (patient 5) during redo TEPT to act as a protective ostomy.

Six of the fistulas were successfully resolved after repair surgery in our hospital and one RUF repair surgery failed. Fecal diversion was closed in all, and a VCU or BE was obtained to confirm that no fistula was present before stoma closure. Median time to reversal of fecal diversion was 10.5 months (range, 7–14 months).

Except for one patient (patient 6) with a persistent fistula, the remaining six patients had a median follow up time of 81 months (range, 5–116 months). No recurrence was observed. Five patients had normal urination. One patient (patient 2) with rectobladder triangle fistula had occasional pain during urination. Seven years after stoma closure, he began to pass some urine from the abdominal wall again, after an unrelated wound infection. Median BFS was 17.5 (range, 17–18) in the six patients who were older than 9 years.

For patient 6, a redo repair surgery will be required via traditional transperineal approach. A midline perineal incision clearly exposing the urethra and fistula tract will be essential.

Fig. 6. IVP showing hydronephrosis, ureterovesical junction obstruction, and bladder displacement because of compression. The arrow shows the direction of bladder compression and displacement.

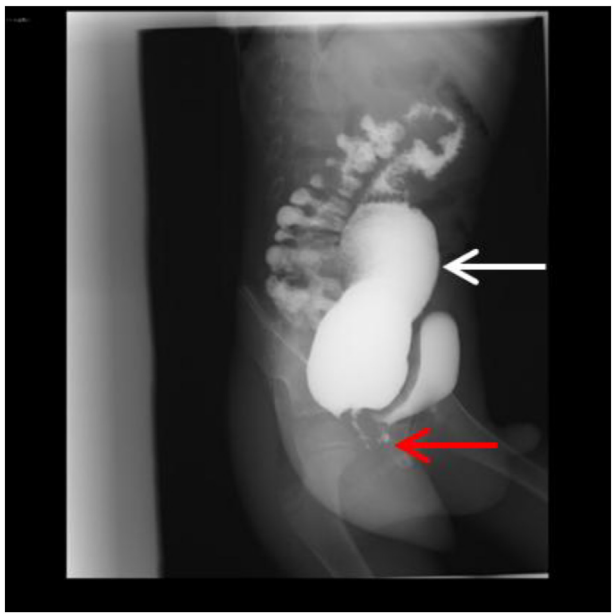

Fig. 7. Barium enema demonstrating distal colonic dilatation (white arrow) without aganglionic segment. The colon proximal to the dilated segment is of relatively normal caliber. Barium entered the urethra and bladder through the rectourethral prostatic fistula (red arrow).

Table 2
Published data.

Author (year) [ref]	No of cases	Location of fistula	Type of PT	Stricture	Diversion	No. of repairs	Successful surgery approach
Sarioğlu et al. (1998) [1]	1	Membranous urethra	Swenson	Yes	Fecal, urine	2	Redo Swenson
Langer et al. (1999) [2]	1	RVF	Soave	Yes	Fecal	1	Transabdominal and Duhamel
Kubota et al. (2003) [3]	1	RUF	Duhamel		Fecal	1	Posterior sagittal approach
Peña et al. (2007) [4]	2	RUF	Unknown		Unknown	1	Posterior sagittal approach
Sowande et al. (2008) [5]	1	RUF	Swenson	Yes	Fecal	2	Posterior sagittal approach
Liu et al. (2008) [6]	4	RUF	Unknown		No	1	York–Mason
Nguyen et al. (2009) [7]	1	RUF	TEPT		Fecal	1	Combined abdominoposterior sagittal approach
Vincent et al. (2013) [8]	1	RUF	Swenson		Fecal, urine	2	Combined abdominoposterior sagittal approach
Granéli et al. (2014) [9]	1	Seminal vesicle	TEPT		Fecal	2	Transverse perineal incision anterior to the anus

3. Discussion

RUF and RVF after HD operation are infrequent but very serious problems. Published literature is summarized in Table 2. Swenson is the most frequently reported HD operation leading to RUF or RVF [1,5,8] and TEPT is less frequently reported [7,9]. However, all 7 patients in our study had received a prior TEPT, which tends to be the surgery of choice for HD patients in China. RUF and RVF after HD operation are usually the result of surgical injury and can be avoided by adherence to basic surgical principles [4,9]. There is also another view suggesting that fistulas arise as a further complication of anastomotic leaks or disruption [8]. RUF could be because of a silent anastomotic abscess, which later breaks through the urethra [7]. Bladder-neck injury was detected during redo TEPT surgery in one patient, and we concluded that it was iatrogenic in nature. Despite treating four patients with anastomotic stricture, their clinical symptoms and medical history varied and we were unable to determine a causal relationship between anastomotic leakage and RUF (RVF).

Instant repair was performed for bladder-neck injury diagnosed during redo TEPT, but this ultimately failed and an RVF formed. Poor urine drainage owing to the absence of suprapubic catheter might be the reason for failure. However, we still maintain that intraoperative diagnosis and immediate repair are very important. Urologic injury that is diagnosed intraoperatively should be repaired immediately, leading to fewer postoperative complications [11].

Accompanying complications were detected in our patients before repair surgery. Most of these complications had a definite relationship with urethral or bladder injury (shown in Fig. 8). The other complications were related to improper treatment (shown in Fig. 9). These complications had significant impacts on the treatment strategy. As such, the combined application of various examinations including DRE, BE, VCU, ultrasonography, and IVP was necessary to provide the most accurate assessment before repair surgery.

In some literature, fecal diversion was performed prior to repair surgery to offer a chance for spontaneous closure of the fistula but did not tend to be successful [3,5]. In our study, the purpose of fecal and/or urine diversion before repair surgery was different. Based on our cases, sepsis owing to anastomotic leakage, which only occurred in RVF patients, was an early postoperative complication that was most ideally dealt with by fecal and urine diversion. Diversion is of particular importance for

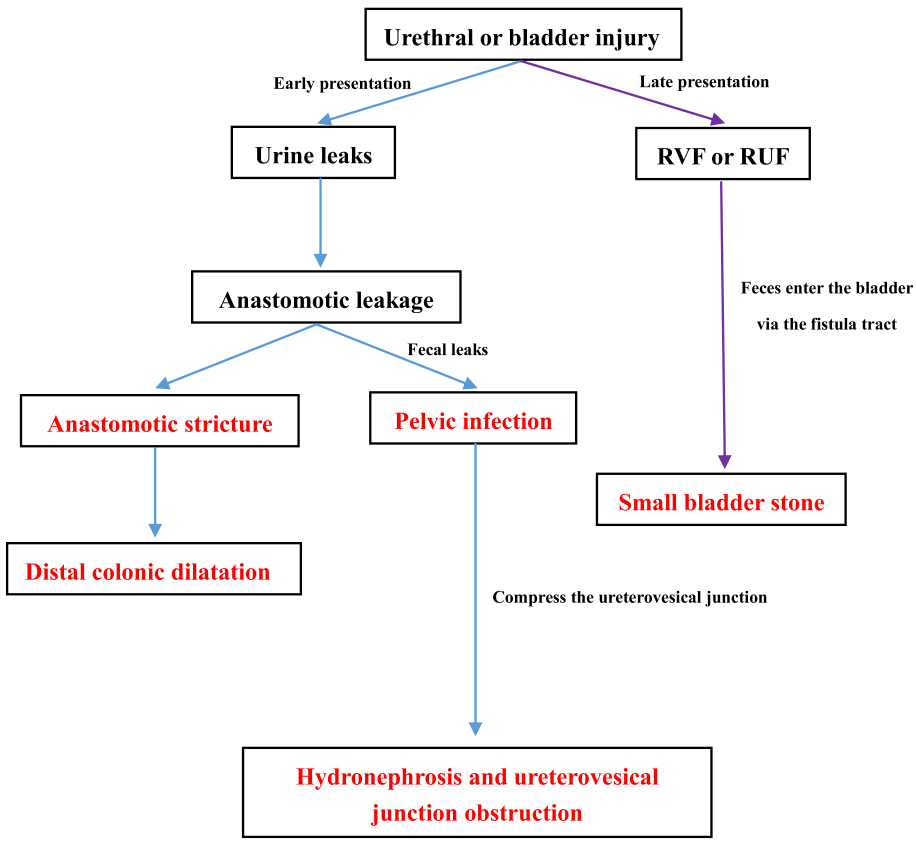

Fig. 8. The relationship between urethral or bladder injury and complications.

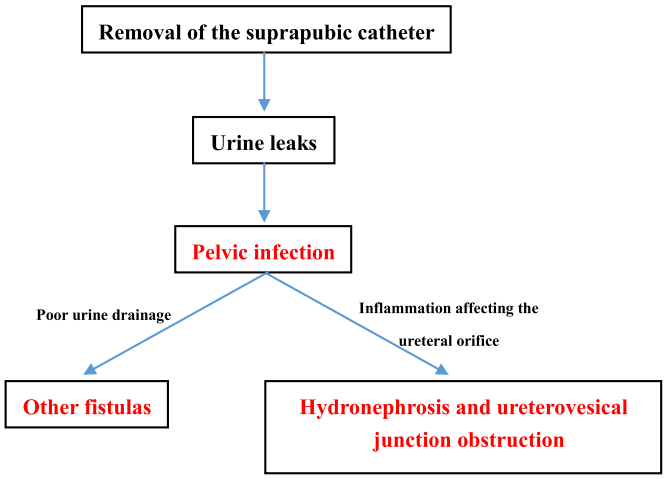

Fig. 9. Complications caused by improper treatment.

curtailing or controlling sepsis [12]. In addition, fecal and urine diversion was performed respectively before repair surgery in two patients to treat pelvic infection caused by fecal or urine leaks, and its accompanying problems; and to allow the tissue to become more pliable by relieving inflammation owing to irritation from fecal or urine leaks. In our opinion, the most important purpose of diversions before repair surgery was to control infection. Prior to any attempt at fistula repair, the surgeon must ensure that infection and local inflammation have resolved [13].

There are four common categories of repair: transanal, transabdominal, transsphincteric, and transperineal (mainly used in adult) [14]. Transabdominal approach is our preferred approach for RVF repair because it provides optimal exposure. It has been reported for repairing RVF after HD operation [2], and was successfully applied in one of our RVF patients. However, the other RVF patient had to receive repair surgery using the transanal approach because of severe adhesions. Although transanal approach has been reported for repairing intraoperative bladder injury, it is rarely used now because it is often impossible to expose tissue properly to excise and repair the fistula [11,14]. In our study, two RVF patients had anastomotic stricture, which made it harder to expose the fistula opening on the rectum by transanal approach. However, after establishment of the pelvic tunnel, the bladder defect was relatively easily exposed. When the transabdominal approach is restricted owing to severe adhesions, the transanal approach may be the next best choice.

Most RUFs after HD operation were repaired by transsphincteric approach, such as posterior sagittal approach and York–Mason [3–8]. In our study, the transperineal approach was performed in all RUF patients. It is a relatively minimally invasive approach compared to the transsphincteric approach and differs from the traditional transperineal approach. The fistula tract can be dissected from the rectum to urethra without injury to the urethra. The urethra was seldom freed from its surrounding tissues. Only the defected part of the urethra, whereby interaction with the fistula tract occurred, was disturbed. There was no dysuria or urine incontinence at follow up time. Moreover, the incision was sutured in situ with little damage to the bowel function. Median BFS was 17.5 (range, 17–18), and half of the patients indicated normal bowel function. All but one RUF with a defect larger than 1 cm healed in our study. This approach proved effective for RUF, but might only be more suitable for RUF with small defect.

Stricture formation secondary to anastomotic leakage of the initial pull-through was the most common cause for performing a redo PT [15]. For RUF or RVF after HD operation, urine leaks caused by bladder or urethral injury can lead to anastomotic leakage, and anastomotic leakage can give rise to anastomotic stricture. The presence of anastomotic stricture in RUF or RVF patients made redo PT necessary, and this view has also been supported in other literature [1,2]. Tissue interposition flaps, although previously used in the repair of RUF after HD operation and commonly used in adults, were not used in our repair [8,9,14]. Instead, the healthy colon was pulled down during redo TEPT just behind the region of the repaired fistula, which is equivalent to tissue interposition. Simultaneously repairing the fistula during redo TEPT also aided in the resolving of the fistula. Sarioğlu A et al. [16] thought that it was difficult to cope with RUF with repair alone, and raised the importance of a second PT. Based on our experiences, 66.7% of patients without anastomotic stricture healed completely after fistula repair alone. If patients did not have anastomotic stricture, there was no indication for redo PT.

After single repair surgery in our hospital, only one RUF patient failed to resolve. This result was superior to other reports, in which success was generally only obtained after two repairs [1,5,8,9]. In the long-term follow up, there was no recurrence of RUF and RVF. However, there was an unforeseen accident in one RVF patient. After an unrelated wound infection seven years later, he developed another bladder fistula that was connected to the abdominal wall. The cause was not defined.

In conclusion, the location of fistula, presence of anastomotic stricture and the association of urinary complications have significant impacts on the treatment strategy in such rare complications after TEPT for HD.

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