# Successful Appendiceal Ureteral Substitution in a Rare Ureteric Involvement of Rhabdomyosarcoma in a Child - A Case Report

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***Abstract***

*Ureteral Rhabdomyosarcoma (RMS) in children is a rare malignancy that may involve the ureter, leading to obstruction and hydronephrosis. Management requires a combination of oncologic therapy and individualized surgical planning, particularly when tumor resection results in extensive ureteral defects. This case report describes an 8-year-old girl with pelvic embryonal RMS involving the right distal ureter, previously treated with pelvic radiotherapy and VAD (vincristine, actinomycin, and doxorubicin) chemotherapy. Imaging showed tumor compression of the distal ureter with grade III hydronephrosis. Surgical resection necessitated en bloc distal ureterectomy, creating a 10-cm ureteral defect. Due to prior pelvic radiation and concerns regarding bladder fibrosis and metabolic complications from bowel interposition, ureteral substitution using the appendix was performed. Postoperative recovery was uneventful, with stable renal function, normal electrolyte balance, and no urinary leakage. Follow-up imaging at 6 months demonstrated a well-functioning appendiceal conduit and no residual tumor. This case illustrates that appendiceal ureteral substitution can be a safe and effective reconstructive option for long distal ureteral defects in pediatric RMS, particularly when prior radiation limits alternative techniques. Careful surgical technique and close follow-up remain essential to ensure optimal outcomes*

***Keywords: Ureteral Substitution, Ureteral Reconstruction, Appendix, Embryonal Rhabdomyosarcoma***

## INTRODUCTION

Rhabdomyosarcoma (RMS) is a malignant tumor arising from primitive skeletal muscle cells and is the most prevalent soft tissue sarcoma in children (Chiloleti et al., 2024). Approximately 15%-20% of RMS cases originate in the genitourinary tract, most commonly in the vagina, urinary bladder, prostate, paratestis, and uterus (Wang et al., 2023; Chiloleti et al., 2024). The mainstay of treatment for RMS in children is a combination of radiation therapy and surgery, which can improve the 5-year survival rate from 55% to 71% (Chiloleti et al., 2024).

Surgical management typically begins with tumor resection after achieving tumor size reduction through radiation therapy (Xiong et al., 2020). However, treatment choice should be individualized according to the tumor characteristics and the patient's overall condition. Ureteral malignancy may cause ureteral obstruction either through direct invasion or external compression, leading to hydronephrosis and renal insufficiency (Artiles Medina et al., 2023).

However, urinary diversion in such cases is most commonly indicated in cases of acute kidney injury, and clinicians should carefully consider both the advantages and disadvantages of the available diversion methods (Artiles Medina et al., 2023).

Surgical resection of tumors affecting the ureter may result in ureteral defects, presenting significant reconstructive challenges for surgeons. Various approaches have been described for distal ureteral defects. Short defects can be managed with direct end-to-end anastomosis (less than 3 cm) or simple ureteroneocystotomy (3-4 cm) (Drake et al., 2025). Longer defects may require techniques such as vesicopsoas hitch (6-10 cm), Boari flap (10-15 cm), transureteroureterostomy, renal autotransplantation, or ureteral substitution (Drake et al., 2025).

However, each method carries substantial risks, such as metabolic disturbances and mucus production in ureteral substitution with ileum (Chen et al., 2023), as well as the high technical complexity that requires considerable surgical expertise in renal autotransplantation associated with renal autotransplantation (Chen et al., 2023; Drake et al., 2025). In our patient, the history of pelvic radiation further complicated decision-making and narrowed the available surgical options. One potential approach with relatively simpler technical demands and fewer complications compared with other reconstruction techniques for distal ureteral defects is ureteral substitution using the appendix, particularly in pediatric patients in whom the appendix is usually still present (Vageriya et al., 2020).

However, reports and data describing appendiceal ureteral substitution for long distal ureteral defects resulting from RMS resection are very limited. This case report aims to describe the clinical course, surgical management, and perioperative and 6-month outcomes of appendiceal ureteral substitution for a long distal ureteral defect. We present our surgical experience using the appendix to replace the right distal ureter following tumor resection for ureteral RMS in a child.

## METHODS

The study design was a retrospective case report involving a single subject (n = 1), an 8-year-old girl with an initial diagnosis of a right retroperitoneal mass involving the right distal ureter. The study was conducted at Dr. Sardjito Tertiary Hospital, Urology Division, Department of Surgery, from August 2023 to December 2023, in Yogyakarta, Indonesia. Clinical data and surgical details were collected from electronic medical records.

Data were curated from medical records and care monitoring sheets, including demographic characteristics, presenting symptoms, physical examination findings, laboratory

parameters (hematology, coagulation profile, liver and renal function, and electrolytes), and radiological assessments (abdominal MSCT and plain X-ray). The diagnosis of embryonal RMS was confirmed histopathologically following surgical resection of the mass.

The natural history of the disease and its management were presented chronologically, including patient presentation, surgical techniques, perioperative care, and postoperative outcomes. The primary outcomes assessed were kidney function and electrolyte balance over a 6-month follow-up period. This case report was prepared in accordance with the CARE Guidelines, with complete data anonymization and written informed consent for publication obtained from the patient's family. Ethical considerations were met according to the policies of the local ethics committee.

## RESULT

An 8-year-old girl was referred to the urology outpatient clinic for evaluation of an abdominal mass, with the chief complaint of intermittent abdominal pain. Three months earlier, she had undergone a laparotomy and biopsy of a retroperitoneal mass, which was diagnosed as pelvic embryonal rhabdomyosarcoma (RMS). Following the diagnosis, she received pelvic radiotherapy (10 cycles) and chemotherapy using the VAD regimen (vincristine, actinomycin, and doxorubicin) until now.

The patient had no significant past medical history, and her parents denied any congenital diseases. She was born at term with normal birth weight. There was no history of urinary tract surgery, trauma, or infection prior to the laparotomy biopsy. Her family history was unremarkable for renal, urological, hereditary diseases, malignancies, metabolic disorders, or congenital urinary tract abnormalities. However, the patient's father was a heavy active smoker since high school and routinely smoked inside the home.

Psychosocially, the patient was active and cheerful, and despite her symptoms, she received strong family support without evidence of psychological distress or socioeconomic difficulties. No relevant genetic predisposition to urological malignancies was suspected based on her clinical and family history.

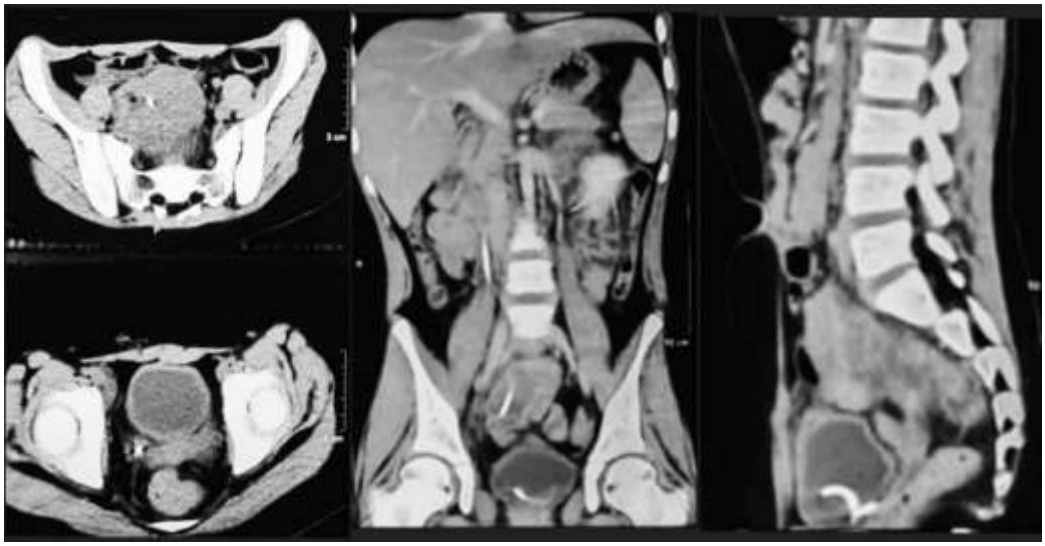

***Figure 1. Abdominal MSCT showing a retroperitoneal mass with involvement of the right presacral lymph node***

On physical examination, a suprapubic mass measuring approximately 8 × 8 cm was palpable, non-tender, and without signs of infection. Urologic ultrasound revealed grade III hydronephrosis of the right kidney, while the left kidney and bladder were normal, with a regular bladder wall. Abdominal MSCT demonstrated a reduction in tumor size to 7 x 6 cm, involvement of the right presacral lymph nodes, and compression/invasion of the right distal ureter resulting in grade III hydronephrosis (**Figure 1**). Laboratory findings were as follows: leukocytes 3.6 x $10^3$/µL, hemoglobin 10.5 g/dL, platelets 384 x $10^3$/µL, BUN 7.0 mg/dL, creatinine 0.56 mg/dL, sodium 136 mmol/L, potassium 4.0 mmol/L, chloride 105 mmol/L, PT 11.1/11.0 sec, aPTT 38.5/31.2 sec, INR 1.01, albumin 4.17 g/dL, random blood glucose 88 mg/dL, and non-reactive HBsAg. The patient was scheduled for surgical resection of the mass.

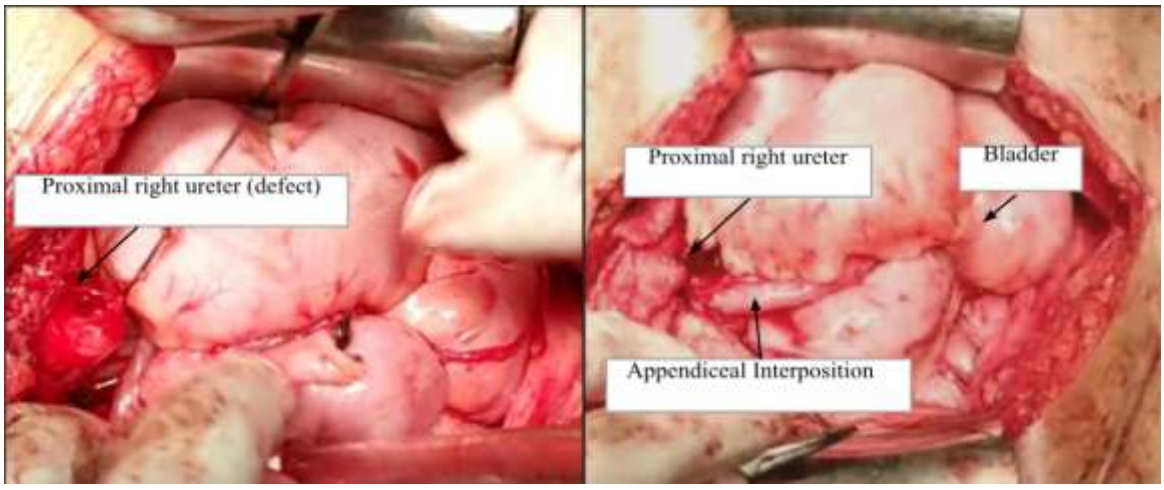


***Figure 2. Direct reimplantation of the ureter was not possible due to an extensive defect of the ureter after the distal uretectomy (left). An appendix was used as ureteral substitution and reimplanted into the bladder (right)***

Surgery began with a midline incision. Intraoperatively, the tumor was found to be enveloping and fused with the distal ureter, making separation impossible. Tumor excision was performed en bloc with a right distal ureterectomy, resulting in an approximately 10 cm ureteral defect (**Figure 2**). Considering this extensive defect, an appendiceal graft was selected for ureteral substitution based on the following factors: 1) History of pelvic radiation increasing the risk of radiation cystitis and bladder fibrosis, rendering a Boari flap technique unfeasible; 2) Ileal ureter substitution carries risks of metabolic acidosis and significant mucus production; 3) The surgical field on the right side facilitated easy visualization and mobilization of the appendix as a conduit. The appendix was harvested by carefully transecting it while preserving the mesoappendix. It was positioned in an isoperistaltic orientation and anastomosed to the proximal ureter using 4-0 braided synthetic absorbable sutures. A DJ stent was inserted antegrade, followed by neo-implantation of the distal appendix into the bladder. The resected mass measured approximately 10 x 7 cm. Histopathology examination confirmed embryonal rhabdomyosarcoma. The procedure lasted 180 minutes, with an estimated blood loss of 400 cc, and no intraoperative complications occurred.

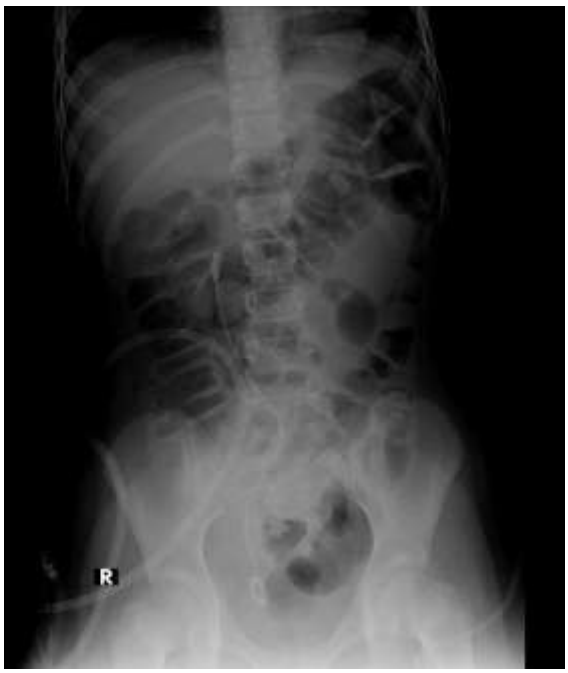


***Figure 3. Postoperative abdominal KUB confirming proper placement of the DJ stent in the right ureter***

Postoperative KUB confirmed proper placement of the DJ stent, with partial distal unraveling within the pelvic cavity (Figure 3). Within 24 hours, the drain yielded 200 cc of serohemorrhagic fluid, with no evidence of urinary leakage. During hospitalization, the patient experienced intermittent subfebrile fever for three days, which resolved spontaneously with antipyretics and antibiotics. The urinary catheter was removed on postoperative day 6, after

which she was able to void normally. No wound leakage was observed, and she reported decreasing postoperative pain. The patient was discharged on postoperative day 7.

**Table 1. Kidney function values during monthly follow-up**

| Postoperative timeline | BUN (mg/dL) | Cr (mg/dL) |
|---|---|---|
| 1st month | 7.2 | 0.61 |
| 2nd month | 6.4 | 0.62 |
| 3rd month | 7.3 | 0.43 |
| 4th month | 6.8 | 0.52 |
| 5th month | 9.0 | 0.64 |
| 6th month | 6.0 | 0.62 |

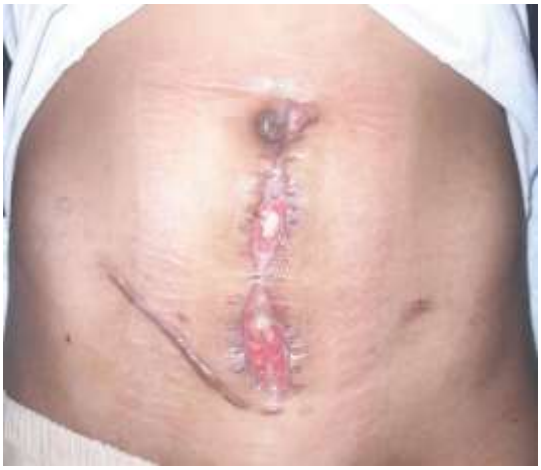

***Figure 4. Postoperative dry and clean surgical wound***

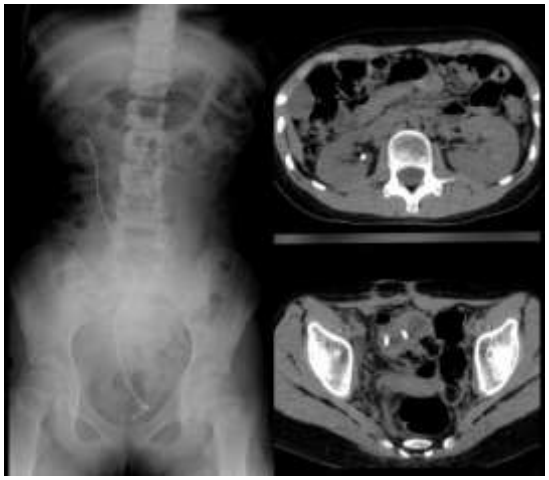

***Figure 5. Postoperative abdominal KUB (left) and abdominal MSCT at 6 months follow-up (right), showing no residual pelvic mass***

We monitored the patient's clinical status and kidney function monthly (**Table 1**). No significant clinical complaints were reported during follow-up. No fluid leakage or purulent discharge was observed at any time. Kidney function remained stable without signs of deterioration. The patient's parents reported complete resolution of abdominal pain. The

surgical wound showed a clean and dry wound after 6 months (**Figure 4**). Postoperative KUB and abdominal MSCT at 6 months demonstrated no residual mass in the pelvis (**Figure 5**).

## DISCUSSION

Rhabdomyosarcoma (RMS) is the predominant connective tissue sarcoma in paediatric populations, constituting approximately 50% of all pediatric soft-tissue sarcoma (Martin-Giacalone et al., 2021). RMS therapy necessitates a complete removal of a local primary tumour, typically accomplished through a multimodal therapy combining surgery, radiation and chemotherapy (Evans et al., 2024). In RMS tumors affecting the ureter, the approach to managing the ureteral defect after excision depends on the length of the removed ureter. At the moment, a wide variety of methods have been documented for treating ureteral defects, such as intestinal onlay/interposition, psoas hitch, Boari-flap, trans-ureteroureterostomy, renal auto-transplantation, and, as a last resort, renal autotransplantation (Chen et al., 2023; Drake et al., 2025).

A suitable technique for repair will depend on the specifics of the injury. The main principles of repair include watertight spatulated anastomosis, low tension on anastomotic sites, blood supply preservation, appropriate debridement, and enough drainage (Ade-Ojo & Tijani, 2021). For long ureteral defects, surgical options include transureteroureterostomy, renal autotransplantation, or ureteral substitution (Xiong et al., 2020). In our case, direct reattachment was not possible due to the length of the excised ureter. Upon evaluating the dimensions and quality of the appendix, we opted to utilize it as a substitute for the absent segment of the distal right ureter. The reason for using appendiceal interposition was that the appendix's anatomical position allowed for its mobility, placement, and anastomosis in a tension-free manner without compromising its vascularization (Bilotta et al., 2021). Since the ureter and appendix have comparable luminal diameters, a technically feasible anastomosis is possible. Additionally, the appendix absorbs only a small amount of urine, reducing the effect of electrolyte imbalance caused by excess urine absorption (Wang et al., 2022).

The appendix is considered an advantageous option due to the minimal morbidity associated with its incorporation and its high success rate in comparison to alternative methods (O'Rourke et al., 2022). We found that numerous studies have demonstrated the effectiveness of using the appendix for ureteral reconstruction in both adult and pediatric populations. A study presented by Dal Moro et al., (2020) highlighted cases involving adult patients with colorectal cancer where the substitution led to favorable outcomes without significant complications. Similarly, pediatric cases, such as those reported by Vageriya et al., (2020),

showcased successful recoveries with minimal complications, reinforcing the technique's applicability across different age groups.

A Boari flap demands sufficient bladder capacity and compliance to harvest the long, wide-based flap necessary for bridging defects. Our patient has a history of pelvic radiation and is at risk of suffering from reduced bladder capacity and compliance due to fibrosis or radiation cystitis, making the creation of a viable Boari flap technically challenging (Srikanth et al., 2022).

Ileal ureter substitution is associated with risks such as electrolyte imbalance, urolithiasis, mucus production causing obstruction, increased infection risk, and longer recovery (Drain et al., 2021). Furthermore, renal auto-transplantation necessitates performance in specialized centers and carries the risk of complications that may arise from vascular and urinary tract anastomosis (Lin et al., 2025). The appendix has inherent limitations that need to be considered. Its length may be insufficient for large ureteral defects, limiting its use in complex reconstructions. Along with this, stenosis, fistulas, and complete dehiscence were listed as possible inherent issues with surgical anastomosis (Bilotta et al., 2021).

From an oncologic perspective, the reconstruction did not compromise surgical principles. An en bloc resection was achieved, and the histopathology confirmed negative surgical margins, which is critical for RMS surgical therapy. The patient was managed in a multidisciplinary setting to address adjuvant therapy for his nodal disease.

## CONCLUSION

This case demonstrates that appendiceal ureteral substitution is a feasible and effective option for reconstructing long distal ureteral defects following pelvic rhabdomyosarcoma resection in children. Despite prior pelvic radiotherapy, the appendix provided a safe conduit with minimal metabolic risk and good functional outcomes. Postoperative follow-up showed stable renal function, preserved electrolyte balance, and no residual tumor. This case underscores the importance of individualized surgical planning, careful intraoperative technique, and close monitoring to achieve optimal results while minimizing complications.

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