

# Rare But Real: A Decade of Pediatric Bladder Tumors in a Single-Center Experience

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# Abstract

**Introduction:** Pediatric bladder tumors are rare and biologically distinct from their adult counterparts, exhibiting notable differences in histology, clinical behavior, and prognosis. Due to their low incidence and heterogeneity, standardized diagnostic and surveillance protocols for children are lacking. This study aims to present a decade-long, single-center experience, providing key insights into the diagnosis, treatment, and follow-up of pediatric bladder tumors, with the goal of contributing to the development of pediatric-specific management strategies.

**Observations:** Between 2013 and 2023, seven pediatric patients (ages 3 - 15 years) were diagnosed with bladder tumors at our institution. Histopathological diagnoses included papillary urothelial neoplasms of low malignant potential, low-grade urothelial carcinoma, papilloma, fibroepithelial polyp, inflammatory myofibroblastic tumor, hemangioma, and rhabdomyosarcoma. Clinical presentations ranged from incidental imaging findings to gross hematuria and urinary retention. Management strategies were individualized based on tumor type, location, and severity, including transurethral resection, laser ablation, partial cystectomy, or multimodal oncologic therapy. No recurrences were observed during a median follow-up of 3.5 years. Diagnostic imaging, cystoscopy, and histopathological evaluation were essential for accurate diagnosis, and treatment prioritized bladder preservation when feasible.

**Conclusion and Relevance:** Pediatric bladder tumors typically follow an indolent course and respond well to conservative surgical approaches, with excellent long-term outcomes. This experience underscores the need for pediatric-specific diagnostic pathways and follow-up protocols that minimize unnecessary invasiveness while ensuring oncological safety. The adoption of histology-adapted surveillance strategies could significantly enhance patient care by preventing overtreatment of low-risk lesions and ensuring vigilant monitoring of high-risk tumors.

*Keywords:* Pediatric Bladder Tumors; Papillary Urothelial Neoplasms of Low Malignant Potential (PUNLMP); Rhabdomyosarcomas (RMS)

# Introduction

Pediatric bladder tumors differ significantly from those of adults in terms of incidence, histology, behavior, and prognosis [1]. According to the SEER database, their incidence has increased over time, with 140 cases reported between 1973 and 2003 [2]. The most common subtypes include papillary urothelial neoplasms of low malignant potential (PUNLMP) (50.7%) and rhabdomyosarcomas (RMS) (36.4%) [2]. RMS, the most common mesenchymal tumor, mainly affects children under 10 years of age and accounts for 5% of all pediatric solid

malignancies [3]. Urothelial bladder cancer (UBC) is rare in children, occurring in 0.003% of those under 20 years of age, with a higher prevalence in adolescents over 13 years of age [4,5].

The diagnosis and management of these tumors vary. Cystoscopy remains the gold standard for detection, while transurethral resection (TURB) is preferred for urothelial neoplasms [6]. Treatment of bladder hemangiomas includes biopsy, fulguration, TURB, embolization, and laser ablation [7,8]. Inflammatory myofibroblastic tumors (IMTs) and RMS require biopsy for confirmation, and RMS is investigated using CT/MRI, chest CT and PET-CT [9]. Treatment follows risk stratification protocols (EpSSG, COG), with low-risk RMS treated by surgery and chemotherapy, while high-risk cases require multimodality therapy, including radiotherapy [1,10].

Despite these protocols, there remains a significant gap in empirical data on recurrence and relapse rates, particularly for urothelial and mesenchymal bladder tumors. Surveillance of RMS of the urogenital organs is well structured and includes regular clinical examinations, imaging and bone scans over five years (EpSSG RMS2005). In contrast, there are no standardized guidelines for urothelial bladder neoplasms (UNB), which are usually solitary, non-invasive lesions (pTa/pT1) with a five-year survival rate of 97.3% [11]. Follow-up strategies for other bladder tumors such as hemangiomas, IMTs, and fibroepithelial polyps remain undefined, leading to heterogeneous surveillance practices across centers.

This study presents a detailed analysis of pediatric bladder tumors treated in the Department of Pediatric Surgery of Santa Maria di Misericordia Hospital in Perugia (2013-2023). Seven patients (aged 0-18 years) were diagnosed with PUNLMP (2 cases), urothelial papillomas (1), fibroepithelial polyps (1), embryonal RMS (1), IMT (1) and hemangiomas (1). Given the rarity of these tumors and the lack of standardized follow-up protocols, this study aims to provide clinical insights into treatment approaches and long-term outcomes and contribute valuable data to an under-researched area of pediatric oncology.

#### **Case Presentation**

#### Case 1

A 12-year-old male patient was referred to our outpatient clinic after a bladder lesion was discovered incidentally during a routine ultrasound examination of the abdomen. The patient was asymptomatic and had no history of urinary tract infection, hematuria, dysuria, or urinary straining. He underwent annual abdominal and testicular ultrasounds as a follow-up for congenital conditions (left diaphragmatic hernia repair and orchidopexy). Ultrasonography revealed a 1 cm vascular lobular lesion with minimal internal flow on the right posterior bladder wall. Transurethral resection of the bladder tumor (TURBT) was performed 27 days later and confirmed a polypoid pedunculated lesion 1.5 cm in size. Histopathologic examination revealed a low-grade papillary urothelial neoplasm of low malignant potential (PUNLMP), which was graded as pTa, N0, M0. The patient had an uneventful postoperative course, and no adjuvant therapy was performed. Urine cytology, abdominal ultrasound and cystoscopy were performed as part of the follow-up and no recurrence was observed over a period of 5 years.

#### Case 2

A 15-year-old male with asymptomatic bladder diverticulosis diagnosed at 2 years of age presented with persistent microscopic hematuria discovered incidentally during routine urinalysis. He denied any associated symptoms such as flank pain, dysuria or fever and had no personal or family risk factors for urinary tract malignancy. Ultrasonography revealed small diverticula and a large left Hutch diverticulum with a hypoechoic, lobulated mass measuring 1.6 × 2.2 cm. Cystoscopy revealed a sessile lesion within the diverticulum, completely resected with TURB. Histopathologic analysis confirmed a low-grade papillary urothelial carcinoma (LG-Ta, N0, M0). Given the intradiverticular location and potential risk of recurrence, a strict follow-up protocol was implemented, including regular cystoscopy and urine cytology. No recurrence or disease progression was observed after 2 years.

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#### Case 3

A 13-year-old boy presented with intermittent, painless gross hematuria that had persisted for two weeks. He denied any history of trauma, infection, vigorous exercise, or bleeding disorders. Physical examination and laboratory tests were unremarkable, and urinalysis confirmed the presence of eumorphic red blood cells. Abdominal ultrasonography showed a 7 mm polypoid echogenic lesion on the postero-inferior bladder wall with minimal internal flow. Cystoscopy performed 25 days after the onset of symptoms showed a pedunculated vascular lesion at the bladder trigone, which was completely removed with TURB. Histopathology confirmed a benign urothelial papilloma without atypia or muscular involvement. The patient underwent a structured follow-up with cystoscopy and ultrasound. No recurrence or disease progression was observed during the 3-year follow-up period.

#### Case 4

A 7-year-old Albanian boy presented with a two-month history of intermittent gross hematuria, dysuria, and voiding strain. The symptoms were initially attributed to a post-operative complication following a circumcision performed the year before. There was no personal or family history of urogenital tumors or systemic disease. Ultrasound and CT scans showed a endoluminal bladder protrusion (7 x 7 mm), originating from the bladder neck and closely associated with the prostate. As a mesenchymal neoplasm (e.g. a rhabdomyosarcoma) was suspected, a cystoscopy was performed. A wide polypoid lesion was found and excised en bloc with a holmium laser. Histologic analysis revealed a benign fibroepithelial polyp without muscular invasion. Postoperative recovery was uneventful. During a 3-year follow-up, which included cystoscopy and imaging studies, neither recurrence nor urinary complications were observed.



Figure 1: Fibroepithelial polyp: US appearance of the lesion.

#### Case 5

A 4-year-old girl was admitted to the emergency department with acute hematuria and abdominal pain. The patient had no history of urinary tract infection, trauma, or congenital abnormalities of the urinary tract. Imaging revealed a pedunculated, exophytic mass (28 × 22 × 26 mm) on the anterior bladder wall and dome, which was suspected to be angiomatosis. Due to worsening bleeding and hemodynamic instability, an urgent open partial cystectomy was performed. Histopathologic analysis revealed an inflammatory myofibroblastic tumor (IMT) that was positive for ALK rearrangement and negative for other sarcomatous markers. The patient recovered well and had no postoperative complications. MRI, ultrasound, and cystoscopy were performed as part of the follow-up, all of which showed no recurrence or disease progression over a 4-year period.

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Figure 2: Inflammatory myofibroblastic tumor: US and TC appearance of the lesion. [A-B], intraoperative finding [C].

# Case 6

A 12-year-old boy was examined because of persistent macroscopic hematuria that had been occurring daily for a month, especially at the end of micturition. Ultrasonography was inconclusive, so cystoscopy was ordered, which revealed non-bleeding telangiectasias of the mucosa at the bladder neck, consistent with a suspected hemangioma. Endoscopic YAG laser ablation was performed until vascular sclerosis was achieved. The patient's medical history revealed a regressed infantile hemangioma on the buttocks. Genetic testing and Doppler studies ruled out syndromic or vascular malformations. Follow-up cystoscopy at 1 year revealed no recurrence. The patient continues to undergo routine pediatric urological monitoring.

#### Case 7

A 3-year-old boy presented with acute urinary retention and a palpable pelvic mass discovered by his pediatrician. Imaging revealed a large (61 × 68 mm), vascularized mass infiltrating the bladder and displacing adjacent structures. An open biopsy by laparotomy

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Figure 3: Intraoperative view (cystoscopy) of hemangioma of bladder neck.

confirmed a fusiform high-risk rhabdomyosarcoma (RMS) of the bladder/prostate. The patient was treated according to the EpSSG 2005 protocol and received nine cycles of IVA chemotherapy followed by conservative, bladder-sparing surgery, and radiotherapy (50.4 Gy in 28 fractions). Post-treatment imaging showed no viable tumor, and the patient began maintenance chemotherapy. He achieved complete remission and has remained recurrence-free during the 5-year follow-up. Cystourethrogram showed no bladder dysfunction. Testicular monitoring showed testicular hypoplasia on the left and testicular retention on the right without further complications.



Figure 4: Fusiform cell rhabdomyosarcoma: US appearance of the lesion.

# **Results**

Seven pediatric patients (5 males and 2 females; age range 3-15 years) were diagnosed and treated for bladder tumors at our institution. Clinical presentation varied from incidental findings during routine imaging (cases 1 and 2) to symptomatic presentations such as macroscopic hematuria (cases 3, 5, and 6), acute urinary retention (case 7), and voiding symptoms including dysuria and straining

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(cases 4 and 5). Family history was negative for urogenital malignancies in all but one case (case 5, cousin with glioblastoma multiforme). Histologic diagnoses included low-grade papillary urothelial neoplasms of low malignant potential, low-grade urothelial carcinoma, urothelial papilloma, fibroepithelial polyp, inflammatory myofibroblastic tumor, bladder hemangioma and fusiform rhabdomyosarcoma. Tumor size ranged from 7 mm to 68 mm, and most lesions were in the posterior region or bladder neck. Treatment strategies included transurethral resection (TURB) for benign or low-grade tumors, laser resection, partial cystectomy, YAG laser ablation, and a multimodality approach with chemotherapy, surgery, and radiotherapy for RMS. No patient received adjuvant intravesical therapy. The follow-up period ranged from 14 months to 5 years. No recurrence was observed in any of the cases, and all patients remained in good health with no signs of disease progression. The full description can be found in table 1.

Case No.	Age (years)	Sex	Diagno- sis	Symp- toms	Family History	Histology	Size	Site	Treat- ment	Adju- vant Therapy	Follow- up Du- ration	Recur- rence
1	12	Male	PUNLMP	Asymp- tomatic	Negative	Papillary low-grade urothelial neoplasm	1.5 cm	Pos- terior bladder wall	TURB	No	5 years	No
2	15	Male	LG-Ta	Micro- scopic hematu- ria	Negative	Low- grade papillary urothelial carcino- ma	1.6 × 2.2 cm	Diver- ticulum bladder wall	TURB	No	2 years	No
3	13	Male	Papil- loma	Inter- mittent gross hematu- ria	Negative	Urothelial papilloma	7 mm	Postero- inferior bladder wall	TURB	No	3 years	No
4	7	Male	Fibro- epithelial polyp	Gross hema- turia, dysuria, voiding strain	Negative	Fibro- epithelial polyp	9 × 8 mm	Bladder neck	Laser resec- tion	No	3 years	No
5	4	Fe- male	IMT	Gross hematu- ria with pain	Cousin with glioblas- toma	Inflam- matory myofibro- blastic tumor	28 × 22 × 26 mm	Anterior bladder wall	Partial cystec- tomy	No	4 years	No
6	12	Male	Bladder heman- gioma	Per- sistent gross hematu- ria	Infantile heman- gioma (re- solved)	Presumed bladder heman- gioma	Not speci- fied	Bladder neck	YAG laser	No	14 months	No
7	3	Male	Rhabdo- myosar- coma	Pelvic mass, urinary reten- tion	Negative	Fusiform cell rhab- domyo- sarcoma	61 × 68 mm	Blad- der/ prostate	Chemo- therapy + con- serva- tive surgery	Radio- therapy + main- tenance	5 years	No

 Table 1: Legend: PUNLMP: Papillary Urothelial Neoplasm of Low Malignant Potential; LG-Ta: Non-Invasive Papillary Carcinoma Low

 Grade; IMT: Inflammatory Myofibroblastic Tumor.

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#### Discussion

This case series provides a critical and structured overview of pediatric bladder tumors and illustrates the clinical and histopathological heterogeneity of seven cases treated over a ten-year period. The spectrum of diagnoses ranges from urothelial lesions (PUNLMP, low-grade papillary carcinoma, papilloma) to benign mesenchymal tumors (fibroepithelial polyp, inflammatory myofibroblastic tumor), vascular malformations (hemangioma) and malignant soft tissue sarcomas (rhabdomyosarcoma). Our aims were: (1) to highlight diagnostic complexity, (2) to evaluate treatment strategies and outcomes, and (3) to assess the applicability of adult diagnostic and surveillance models in pediatric patients.

A consistent theme across all cases is diagnostic delay, often due to non-specific symptoms. Gross hematuria, typically terminal, intermittent, or mild, was the most common symptom but was often incorrectly associated with benign causes. Our findings reflect the literature, with Paner., *et al.* [12] reporting delays of more than one year in up to 25% of pediatric patients. Despite evidence from Greenfield., *et al.* (2000) and Castagnetti., *et al.* [13] urging timely investigation of persistent hematuria, clinical inertia remains prevalent. We advocate a change in clinical perspective: persistent hematuria, regardless of its severity, should prompt a comprehensive evaluation including imaging and, if indicated, cystoscopy.

Pediatric bladder tumors are biologically and clinically distinct from their adult counterparts. All urothelial lesions in our series were low-grade and non-invasive, without common molecular mutations such as TP53 and FGFR3 [1,12]. This molecular profile favors conservative treatment strategies and less intensive surveillance in children. Case 2, a low-grade carcinoma in a bladder diverticulum, illustrates the limitations of adult-derived prognostic models. While diverticula in adults are associated with a higher risk of recurrence, our patient remained recurrence-free, highlighting the need for pediatric-specific risk stratification. The accurate diagnosis of non-urothelial tumors, such as IMT, fibroepithelial polyps, and hemangiomas, depended heavily on histopathologic and molecular profiling. ALK rearrangement was crucial for the diagnosis of IMT and emphasizes the importance of advanced diagnostics even when imaging appears benign [14]. Ultrasonography was the first choice in all cases and offered high sensitivity and specificity [15,16], although it failed to detect flat telangiectasias in one patient. MRI has been essential for surgical planning and staging in complex cases and remains the modality of choice for pediatric pelvic tumors due to its superior soft tissue contrast and lack of ionizing radiation [14]. Cystoscopy remains the gold standard for definitive diagnosis and treatment. It allows visualization of the lesion, biopsy and resection. Laser-assisted techniques (holmium and YAG) were advantageous in anatomically sensitive areas and offered precision with minimal thermal injury [13,17].

Treatment was individualized according to histology, morphology, size, and location, reflecting the diversity of pediatric bladder tumors and the lack of standardized pediatric guidelines. TURB alone was effective for non-invasive urothelial lesions, as reported by Caione., *et al.* [18]. No patient received intravesical chemotherapy, consistent with pediatric concerns regarding toxicity and unclear benefit [15]. IMT required partial cystectomy due to the hemorrhagic presentation and size of the lesion. The RMS case followed multimodality pediatric oncology protocols [10].

Tumor behavior was consistent with previous reports: No recurrences of urothelial tumors occurred, confirming recurrence rates below 10% in pediatric cohorts [18,19]. IMTs and hemangiomas have also been successfully treated with complete surgical excision [8,14]. RMS required bladder-sparing therapy with chemotherapy, surgery and radiotherapy, with good long-term disease control. Follow-up protocols in our series varied widely, reflecting both the heterogeneity of the tumor and the lack of pediatric consensus. Most institutions adapt adult protocols, which often include invasive procedures such as frequent cystoscopies that may be excessive in low-risk pediatric cases [13]. Surveillance strategies in our cohort had a balance between safety and invasiveness, with ultrasound, urinalysis, cystoscopy, and MRI used when warranted. RMS remains the only tumor type for which there are specific pediatric surveillance guidelines, but even these may need to be updated. For rare lesions such as IMT or hemangiomas, longitudinal data is lacking, justifying more cautious follow-up.

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We strongly support the development of histology-specific, risk-adapted surveillance protocols. Patients with solitary, low-grade, non-invasive tumors can be safely monitored with regular ultrasonography and urinalysis, whereas patients with high-risk histology or incomplete resection require closer surveillance, possibly including cystoscopy and MRI. The lack of recurrence after five years in low-grade cases, as reported by Castagnetti., *et al.* and Paner., *et al.* further calls into question the need for long-term invasive surveillance in all patients.

# Conclusion

Although pediatric bladder tumors are rare and often indolent, optimal management remains hampered by the lack of dedicated guidelines. Our series, together with current evidence, supports a shift from adult-modeled strategies toward individualized pediatric care. Central to this evolution is the recognition of hematuria as a potentially significant finding in children. The development of international, minimally invasive, and histology-specific surveillance protocols must become a shared priority to ensure long-term, evidence-based care for this unique patient population.

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