HAYDARPAŞA NUMUNE MEDICAL JOURNAL

DOI: 10.14744/hnhj.2021.32659
Haydarpasa Numune Med J 2022;62(1):101–106

hnhtipdergisi.com

ORIGINAL ARTICLE

Evaluation of Urinary System Pathologies in Patients with Sacrococcygeal Teratomas

Hayriye Nihan Karaman Ayyıldız[1], Şafak Karacay[2], Ali Sayan[3], Ahmet Arıkan[4]

[1]Department of Pediatric Surgery, Umraniye Training and Research Hospital, Health Sciences University, Istanbul, Turkey
[2]Department of Pediatric Surgery, Faculty of Medicine, Yeditepe University, Istanbul, Turkey
[3]Department of Pediatric Surgery, Tepecik Training and Research Hospital, Health Sciences University, Izmir, Turkey
[4]Department of Pediatric Surgery, Tınaztepe Hospital, Izmir, Turkey

Abstract

Introduction: Sacrococcygeal teratomas (SCT) are the most common germ cell tumors. The incidence of voiding dysfunction is high in children with SCT and its origin is usually neurogenic. Pelvic innervation of the type and degree of teratoma is known that may cause undesirable effects on urinary continence.

Methods: In this study, a complete urological examination including urinary microscopy, abdominal ultrasound, voiding cystourethrography, and urodynamic study was performed in 11 patients who were operated for SCT in the neonatal and infancy period between 1996 and 2004.

Results: Three of our patients had Type I, six had Type II, and two had Type III tumors. Recurrence occurred in four patients during follow-up. Hydronephrosis was found as unilateral in two and bilateral in two patients. In urodynamic studies, we found low bladder compliance in four patients, detrusor sphincter dyssynergia in five patients, unstable detrusor contraction in five patients, and neurogenic bladder sphincter dysfunction in one patient.

Discussion and Conclusion: There was no significant relationship between the teratoma stage and urinary pathological findings and continence. The data about those who had problematic surgery, the type and number of the procedures, presence of invasion, the size of the tumor, recurrence, and undoubtedly the surgeon's technique have suggested that they are the effective factors in the occurrence of complications. The study suggests that the urodynamic studies should be performed alongside other studies in patients with SCT.

Keywords: Childhood; neurogenic bladder dysfunction; sacrococcygeal teratoma; urinary system findings; urodynamic studies.

Sacrococcygeal teratoma (SCT) is the most common solid neonatal tumor with an incidence of 1 in 30,000–40.000 live births, with a 3:1 female to male ratio[1]. They may cause post-operative neurological disorders due to the anatomical region they originate from. SCTs are classified according to a system developed by Altman et al.,[2] which divides the tumors into four anatomical types that differ in the degree of pelvic extension.

While the risk of urologic co-morbidity in SCT patients exists due to the pelvic involvement, the potential mass effect of the tumor and the need for surgical resection for negative margins, and it is also known that the type and the stage can have adverse effects on pelvic innervation, anal and urinary continence.[3,4]

Correspondence (İletişim): Hayriye Nihan Karaman Ayyıldız, M.D. Saglik Bilimleri Universitesi Umraniye Egitim ve Arastirma Hastanesi Cocuk Cerrahisi Klinigi, Istanbul, Turkey
Phone (Telefon): +90 505 293 50 58 **E-mail (E-posta):** nkaraman99@hotmail.com
Submitted Date (Başvuru Tarihi): 23.12.2020 **Accepted Date (Kabul Tarihi):** 13.07.2021

In the study conducted by the American Association of Pediatrics Section of Surgery, it was reported that the congenital anomalies related to the musculoskeletal system and kidney were found in 18% of the cases[2]. Urological complications have been observed in 22–28% of patients with SCT. In children with SCT, the incidence of voiding dysfunction is high, and its origin is usually neurogenic[5,6].

In this retrospectively study, we aimed to stress the early detection of bladder dysfunction and urinary system anomalies to start the treatment as soon as possible and to help determine the application standards prospectively.

Materials and Methods

We retrospectively examined the urodynamics studies of 11 patients who were operated at the SSK Tepecik Training Hospital Pediatric Surgery Clinic between 1996 and 2004 during the neonatal and in infancy period due to SCT. Patients were selected regardless of age, gender, SCT stage, clinical findings, and time elapsed from the last operation. We questioned the daily voiding frequency, painful voiding, incontinence, constipation and anal incontinence. The age of onset of the complaints, the type of surgical intervention, complications developing during and after the operation, radiological and serological examinations were also investigated.

Urine microscopy and urine cultures were obtained. The evaluation of the urinary system was made with lower abdominal ultrasonography (USG), voiding cystourethrography (VCUG) and urodynamics.

Urodynamic studies were carried out in the neuro-urology laboratory of our hospital's Urology Clinic, using the Laborie 600 brand urodynamic device.

Standardized uroflowmetry and cystometry with Electromyography (EMG) were performed to evaluate detrusor and sphincter functions during bladder filling and emptying. The parameters recorded were expected cystometric capacity, measured cystometric capacity, detrusor pressure, uninhibited contractions of the detrusor; residual volume, compliance on uroflowmetry[7].

Bladder capacity was calculated for age based on formula:

EBC=(age in years + 2)×30 ml, and (under the age of two) EBC=kg*7 ml[8].

International Childeren's Continence Society accepts compliance in children: If the expected functional capacity detrusor pressure is 10 cm H_2O or less, it is a normocompliance bladder, and if it is above it is a hypocompliance bladder[7].

In our study, uroflowmetry was performed only in compliant cases. In each case, urodynamic examination was performed by filling the bladder at least twice. The data were determined by calculating the average value of these two measurements. During this retrospective study, pre- and post-operative urological evaluations could be performed in only one patient with SCT who had just started treatment. Urodynamic examination was performed in other patients after SCT excision.

Findings

All patients who applied to our clinic were referred by obstetricians and pediatricians with the findings of swelling on the sacrum. Symptoms of urinary and rectal compression were not detected in the first admission of the patients. Three patients were diagnosed by antenatal USG. Eight (72.7%) of the patients were operated in the neonatal and 3 (27.3%) were operated in infancy period. Three (27.3%) of our cases were Type I, 6 (54.5%) were Type II, and 2 (18.2%) were Type III SCT.

The mean follow-up time was 39.5 months (3-84 months). During the follow-up, 4 patients (36.3%) had relapse. Two of these patients (50%) were Type II and 2 (50%) were diagnosed with Type III SCT. Two of these cases were operated once, 1 twice, and 1 four times in different centers. Abdomino-sacral excision was performed to the patient who was operated 4 times, and chemotherapy was given for teratocarcinoma.

Urodynamic examination was performed to the patients between 8 months and 9 years of age, with the mean of 56.3 months. The voiding complaints of 2 patients (18.2%) could not be evaluated because they were under 2-years-old and the parents did not report any complaints. There was no history of urinary infection in our patients younger than 2-years-old. While 8 of 9 patients (81.8%) older than 2 years had no complaints suggesting voiding dysfunction, only one patient had painful voiding sensation, frequent urinary tract infection and urinary incontinence. This particular patient who routinely performed clean intermittent catheterization due to detrusor sphincter dyssynergia (DSD), high residual urine and incontinence, was operated 4 times in various centers due to the development of malignancy and tumor recurrence. During the follow-up, urinary tract infection was not encountered in any of the patients except one.

USG revealed unilateral hydronephrosis in 2 (18.2%) and bilateral hydronephrosis in 2 patients (18.2%). In a case with unilateral Grade 1 hydronephrosis, no anatomical cause was found, and hydronephrosis regressed and kid-

ney size returned to normal in the six-month follow-up. In the other case with unilateral hydronephrosis, we detected Grade 3 vesicoureteral reflux (VUR) and horseshoe kidney. We found bilateral Grade 3 VUR in 1 case with bilateral hydronephrosis, and bilateral lower ureteric stenosis in the second case. Type I tumor was detected in the patient with unilateral hydronephrosis whom hydronephrosis regressed in the follow-up. Type II tumor was found in two cases: one with unilateral and the other one with bilateral Grade 3 reflux. We found Type III tumor in the patient with bilateral hydronephrosis.

VCUG revealed, two patients with VUR and one patient with irregular bladder contours and multiple diverticula. The degree of VUR was bilateral Grade 3 in one case and unilateral Grade 3 in the other.

The urodynamic examination results are summarized in Table 1. A patient was examined both in the pre- and at post-operative period (4–8 months of age). Both of these results are shown in Table 2.

In the examination performed while the bladder capacities of three patients (27.3%) (patients: 3, 8, 9) were found equal to the expected capacity, it was less than expected in six patients (54.5%) (patients: 1, 2, 6, 7, 10, 11) and more than the expected capacity in two patients (18.2%) (patients: 4, 5).While the measured bladder capacity of one of the three patients with Type I tumor was equal to the expected value (patient: 3), it was measured lower than the expected capacity in the other two patients (patients: 1, 2). The bladder capacity of two patients with Type II was equal to the expected capacity (patients: 8, 9), two were below the expected (patients: 6, 7), and the other two were measured above the expected capacity (patients: 4, 5). The bladder capacity of two patients (patients: 10, 11) with Type III tumors was measured below the expected value.

Compliance values were measured as normal in 7 patients (63.6%) (patients: 2, 3, 5, 7, 8, 9, 10) and as hypocompliant in 4 patients (36.4%) (patients: 1, 4, 6, 11). Among the 7 patients with normal compliance values, two had Type I, four had Type II, one had Type III tumor. Four hypocompliant patients had one Type I, two had Type II and one had Type III tumor.

The bladder capacity of the patient with Type II tumor who had undergone pre and post-operative evaluations was measured as 60 ml and 130 ml, and the compliance values were detected as 3.2 ml/cmH_2O and 2.5 ml/cmH_2O, respectively.

We observed unstable detrusor contraction (UDC) in 5 patients (45.5%). UDC was observed in 2 patients with Type I tumors, 2 patients with Type II tumors and one patient with Type III tumors.

The Pdet Qmax values obtained for all patients were be-

Table 1. Urodynamics findings

Case	Age (Month)	SCT Type	Expected Bladder Capacity (ml)	Calculated Bladder Capacity (ml)	Compliance	DI	LEAK	Pdetmax	DSD	EMG	Urine Residue (cc)
1	50	I	180	145	6.8	+	–	65	–	+	5
2	52	I	180	140	10.2	+	+	50	–	+	2
3	96	I	300	290	13	–	–	85	–	+	–
4	8	II	63	130	2.5	+	–	110	+	Dyssynergia	10
5	9	II	56	130	13.4	+	+	130	+	+/–	15
6	45	II	150	120	2.5	–	–	73	+	+/–	5
7	48	II	180	124	12	–	–	70	+	+/–	5
8	84	II	270	270	11.5	–	–	50	–	+	15
9	96	II	300	290	10.5	–	–	80	–	+	–
10	48	III	180	155	12	+	–	75	–	+/–	5
11	84	III	270	200	7.5	–	+	135	+	+/–	100

DI: Detrusor instability; DSD: Detrusor sphincter dyssynergia; EMG: Electromyography; SCT: Sacrococcygeal teratomas.

Table 2. Pre- and post-operative urodynamics findings

Case	Bladder capacity		Compliance		Residual urine		Sensitivity	
	Before	After	Before	After	Before	After	Before	After
1	60 cc	130 cc	3.2	2.5	10	10	Incompatible	Incompatible

tween 50 and 135 cmH_2O. We recorded 3 (27.2%) of the cases as pressures above 100 cmH_2O and evaluated them. These pressures were found to be high enough to affect the upper urinary system during both bladder filling and voiding. We observed VUR in two cases with Type II tumors, and DSD in one case with Type III tumor. Vesical pressure values of other 8 (72.8%) cases were measured within normal limits.

The relaxation of EMG activity with detrusor contraction occurred in 6 (54.5%) of 11 patients and these patients voided normally. In remaining 5 (45.5%) patients, partial sphincter relaxation was considered as the reason for the prolongation of urination time. DSD was observed in four patients with Type II tumors and in one patient with Type III tumors.

We detected residual urine which is more than 10% of the bladder capacity in only one case. This patient had neurogenic bladder sphincter dysfunction. The expected bladder capacity was 270 ml, and the maximum bladder capacity was measured as 200 ml. In this case, 100 ml of residual urine was detected after voiding.

In our study, the loss of motor function and defecation control of 2 patients (18.2%) could not be evaluated because they have uncompleted toilet training. Chronic constipation and weakness in lower extremity muscles were detected in a patient with Type III tumor. In one patient with Type II tumor, only chronic constipation was detected. We encountered no problems of walking or soiling in other patients. Anal incontinence was not detected in any patients. Statistical study could not be performed due to the small number of patient groups.

Discussion

SCT is one of the most common germ cell tumors in infants and children. Urological complications are observed in 22–28% of patients with this lesion[6]. Associated urological complications reported in children with SCT include VUR, ureteral and urethral obstruction, and neurogenic bladder[9].

For the 1st time, Kirk et al.,[10] examined 24 children with SCT in 1973 and reported 8 children with urological problems. 3 of these patients had hydronephrosis, 1 tumor-associated bladder outlet obstruction, and 4 patients had post-operative urinary retention due to per-operative nerve injury. It is stated that the full urological evaluation and follow-up is mandatory for all children with SCT.In his study in 1990, Malone et al.,[6] found fecal and urinary incontinence in 40% of the patients and weakness in the lower limb muscles in 2 patients. Both authors have been suggested that the grade of the tumor affects the frequency of urological sequelae[6,10]. In addition, Malone determined that in tumors with large presacral components, post-operative urinary and fecal incontinence increased up to 67% in the abdominosacral and up to 33% in the sacrococcygeal approach[6].

However, none of the reports mentioned above had detailed urodynamic studies. In a study by Boemers et al.,[5] on 11 patients, they have detected abnormal urodynamic findings in 9 patients. It has been stated that the main cause of neurogenic lower urinary tract dysfunction was due to traumatic injury to the pelvic plexus and sacral nerves during surgical dissection, and the primary cause was tense spinal cord anomaly in two patients.

Ozkan et al.,[4] in 2006 analyzed the UDS of 14 patients of operated SCT who presented with urinary infection or incomplete bladder emptying after SCT resection. Detrusor over-activity was seen in 8 patients, under-activity in 2 and normal activity in 4. Abnormal urethral sphincter EMG potentials were observed in 7 of 13 patients. They concluded that full evaluation, including UDS, is mandatory before and after surgery.

Berger et al.,[11] evaluated post-operative bladder and rectal function in 25 children with SCT and reported that 50% had bladder dysfunction. All four Altman types were represented, and bladder dysfunction was found in all tumor types. In additional Berger et al.,[11] reported that bladder dysfunction was common after SCT excision and the disorder was even higher in children who underwent multiple surgical interventions. Similarly, in their study on 27 patients, Güler et al.,[12] bladder dysfunction was determined in 87.5% of the patients undergoing two or more operations.

Khanna et al.,[13] evaluated a total of 57 patients clinic, radiologic and urodynamical. They detected urological complaints in 28 of 57 patients, and radiological abnormalities in 8 patients. Urodynamic study was done in 27 patients, 18 had abnormalities. Six patients without any clinical or ultrasonographic abnormalities had abnormal UDS. Three patients had overactive bladder, five dysfunctional voiding, one underactive bladder and one had giggle incontinence.

In our study, we detected UDC in 5 patients, DSD in 5 patients, neurogenic bladder sphincter dysfunction in 1 patient. While 2 of the patients with UDC had Type I, 2 had Type II and 1 had Type III tumors, 4 of the cases with DSD had Type II and 1 had Type III tumors. Although the number of intergroup patients was not sufficient to make statistical comparison, abnormal urodynamic findings were found

more frequently in all three types of patients with high tumor levels. These findings suggest that they are closely related to the nerve damage during the surgical intervention. Among the patients with UDC, the patient with Type III tumor underwent sacral and abdominosacral operations for 4 times, while one patient with Type II tumor had single operation and the other one had two operations.

In sacrococcygeal tumors, bladder and rectum dysfunctions are observed due to the location of the tumor and surgery[11]. In our study, unilateral hydronephrosis was detected in one patient in the pre-operative period. This complete regression of hydronephrosis in the post-operative period suggests that the hydronephrosis was due to the tumor compression. The VUR detected in the post-operative period in two patients may have developed due to surgical nerve damage. It is possible that a number of urinary system anomalies may have occurred due to the compression of tumors extending into the pelvis and the urinary system; however, since we could not perform sufficient pre-operative examinations in our patients, we did not find enough findings to support these claims.

In the literature, constipation has been reported in post-operative period[1,6,14,15]. In our study, chronic constipation was detected in two patients.

Surgical trauma alone cannot explain the high incidence of functional impairment in these children, and the root cause of bladder sphincter dysfunction is still unclear. Urodynamic evaluation of patients before and after surgery will help to examine this problem more precisely. However, since most of these children undergo tumor excision urgently after birth, urodynamics cannot be performed mostly in patients with SCT. In our series, a child had undergone pre- and post-operative urodynamic examination and tumor excision was performed with a sacral approach. Surgical treatment did not affect lower urinary tract function. Boemers reports that in two patients who underwent tumor resection by sacral way, there was no difference in pre- and post-operative urodynamics results and that the lower urinary system functions were not affected[5].

As a result, we found no significant relationship between the teratoma stage and urinary pathological findings and continence. Those who had problematic surgery, the type, number, tumor invasion and size and recurrence of the intervention, undoubtedly the technique and the patience of the surgeon seems to be the effective factors in the occurrence of complications.

Limitations of this study are the limited number of patients and therefore the inability to perform statistical analysis, the lack of detailed pre-operative urological examinations of the patients, the absence of patients with Type IV tumors, the short follow-up periods, and the retrospective evaluation.

In our study, the lack of striking results in terms of complications can be attributed to the small number of patients as well as the concentration of patients at lower stages and the application of meticulous neonatal intervention principles. On the other hand, we will have the chance to discuss the findings more comfortably at the end of satisfactory urodynamic studies in both pre- and post-operative periods in such patients. Clinic, radiologic, and urodynamic studies may be useful in elucidating the causes of urinary system pathologies that occur in the post-operative period, as well as it will be useful in early prevention and treatment. We recommend early urological evaluation for all patients with SCT to protect their kidney function and increasing the quality of life.

Ethics Committee Approval: Since there was no Ethics Committee at the time of this study, it was done with the approval of Health Sciences University Tepecik Training and Research Hospital Chief Physician.

Peer-review: Externally peer-reviewed.

Authorship Contributions: Concept: H.N.K.A., A.A.; Design: H.N.K.A., A.S.; Data Collection or Processing: H.N.K.A., Ş.K.; Analysis or Interpretation: H.N.K.A., A.A.; Literature Search: H.N.K.A.; Writing: H.N.K.A.

Conflict of Interest: None declared.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Shalaby MS, O'Toole S, Driver C, Bradnock T, Lam J, Carachi R. Urogenital anomalies in girls with sacrococcygeal teratoma: A commonly missed association. J Pediatr Surg 2012;47:371–4.
2. Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: American academy of pediatrics surgical section survey-1973. J Pediatr Surg 1974;9:389–98. [CrossRef]
3. Lahdenne P, Wikström S, Heikinheimo M, Marttinen E, Siimes M. Late urologic sequelae after surgery for congenital sacrococcygeal teratoma. Pediatr Surg Int 1992;7: 195–8. [CrossRef]
4. Ozkan KU, Bauer SB, Khoshbin S, Borer JG. Neurogenic bladder dysfunction after sacrococcygeal teratoma resection. J Urol 2006;175:292–6; discussion 296. [CrossRef]
5. Boemers TM, van Gool JD, de Jong TP, Bax KM. Lower urinary tract dysfunction in children with benign sacrococcygeal teratoma. J Urol 1994;151:174–6. [CrossRef]
6. Malone PS, Spitz L, Kiely EM, Brereton RJ, Duffy PG, Ransley PG. The functional sequelae of sacrococcygeal teratoma. J Pediatr Surg 1990;25:679–80. [CrossRef]

7. Nevéus T, von Gontard A, Hoebeke P, Hjälmås K, Bauer S, Bower W, et al. The standardization of terminology of lower urinary tract function in children and adolescents: Report from the standardisation committee of the international children's continence society. J Urol 2006;176:314–24. [CrossRef]
8. Koff SA. Estimating bladder capacity in children. Urology 1983;21:248. [CrossRef]
9. Reinberg Y, Long R, Manivel JC, Resnick J, Simonton S, Gonzalez R. Urological aspects of sacrococcygeal teratoma in children. J Urol 1993;150:948–9. [CrossRef]
10. Kirk D. Urinary complications of sacrococcygeal teratoma. Z Kinderchir 1976;18:294–304.
11. Berger M, Heinrich M, Lacher M, Hubertus J, Stehr M, von Schweinitz D. Postoperative bladder and rectal function in children with sacrococcygeal teratoma. Pediatr Blood Cancer 2011;56:397–402. [CrossRef]
12. Güler S, Demirkaya M, Balkan E, Kırıştıoğlu İ, Kılıç N, Sevinir B. Late effects in patients with sacrococcygeal teratoma: A single center series. Pediatr Hematol Oncol 2018;35:208–17. [CrossRef]
13. Khanna K, Agarwala S, Bakhshi S, Srinivas M, Jana M, Devasenathipathy K, et al. Need for urodynamic evaluation as a regular follow-up tool in assessment of long-term urological outcomes in patients with sacrococcygeal teratoma. J Pediatr Surg 2019;54:2107–11. [CrossRef]
14. Kremer ME, Derikx JP, van Baren R, Heij HA, Wijnen MH, Wijnen RM, et al. Patient-reported defecation and micturition problems among adults treated for sacrococcygeal teratoma during childhood--the need for new surveillance strategies. Pediatr Blood Cancer 2016;63:690–4. [CrossRef]
15. Rintala R, Lahdenne P, Lindahl H, Siimes M, Heikinheimo M. Anorectal function in adults operated for a benign sacrococcygeal teratoma. J Pediatr Surg 1993;28:1165–7. [CrossRef]