Long-Term Outcome of Sacrococcygeal Teratoma: A Controlled Cohort Study of Urinary Tract and Bowel Dysfunction and Predictors of Poor Outcome

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Objective To evaluate urinary tract and bowel function in children with sacrococcygeal teratoma, compare the findings with healthy children, and assess predictors of poor outcome.

Study design This was a controlled cohort study of all patients operated for sacrococcygeal teratoma at a tertiary pediatric surgery center, 2000-2013. Urinary and bowel function were compared with healthy control patients matched for age and sex. Perioperative and histopathologic risk factors were analyzed.

Results In total, 17 patients with sacrococcygeal teratoma and 85 healthy control patients were included in the study. Patients with sacrococcygeal teratoma more often were reported to have uncontrolled voiding (12% vs 0%, $P < .01$), difficulty in bladder emptying (24% vs 0%, $P < .001$), and pyelonephritis (18% vs 1%, $P < .05$). Constipation was more common in patients with sacrococcygeal teratoma (47 % vs 14%, $P < .05$), but the overall bowel function score was equal in the 2 groups. Children with large tumors and immature histology were more likely to have a dysfunctional outcome ($P < .05$).

Conclusions Uncontrolled voiding, difficulty in bladder emptying, pyelonephritis, and constipation were more common in patients with sacrococcygeal teratoma than in healthy children. Dysfunctional outcome was more prevalent in children with large and immature teratomas. *(J Pediatr 2018;■■:■■-■■).*

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Sacrococcygeal teratoma is the most common congenital neoplasm, with a reported prevalence of 1 in 10 000-40 000 live births.[1-5] The majority of the tumors are benign, and treatment is complete resection, including coccygectomy.[6] Most infants have a favorable neonatal prognosis, although fast-growing and highly vascularized tumors may compromise intrauterine growth and pose a challenge for obstetricians and pediatric surgeons.[5,7,8] Through multimodal treatment, survival has improved during the past decades, and focus among parents and caregivers is shifting toward increased awareness of long-term outcome.[6]

Follow-up studies of children with sacrococcygeal teratoma indicate that 7%-60% have some degree of long-term impairment of urinary tract and bowel function.[9-19] However, no previous study has compared such findings with a control group of healthy children matched by age and sex. Functional problems of the urinary tract and bowel are common in children[20,21] and correlate closely with age and sex.[20] Such symptoms may be due to tumor damage or surgical excision, but physiological immaturity of pelvic structures also may contribute. Increased knowledge of how tumor pathophysiology correlates with dysfunctional outcome would be valuable when risk-stratifying patients, counseling parents, and designing follow-up schemes.

The aim of this study was to assess long-term outcome in a consecutive cohort of children operated for sacrococcygeal teratoma and to compare these findings with healthy children matched according to age and sex. In addition, we aimed to evaluate perioperative predictors of urinary tract and bowel dysfunction.

Methods

We conducted a controlled cohort study of all survivors of sacrococcygeal teratoma in Southern Sweden between 2000 and 2013. With a catchment area of 1.8 million residents, the Children's Hospital in Lund, Sweden, is the single tertiary center for prenatal diagnosis, neonatal surgery, histopathologic classification, and

BFS	Bowel Function Score
UTI	Urinary tract infection

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follow-up. The prenatal and perioperative characteristics of the cohort have been summarized comprehensively.[5] In this study, each survivor of sacrococcygeal teratoma was matched with 5 children of the same age and sex. Controls were otherwise-healthy children, including siblings of patients or children of personnel and their acquaintances. Ineligible as controls were children with disorders or previous operations affecting urinary tract or bowel, as well as children with cognitive disabilities or neuromuscular diseases.

Data Collection, Questionnaire, and Structured Interview

Children and caregivers were invited to a follow-up visit regarding urinary tract and bowel function. Questionnaires were completed during a structured interview at the Children's Hospital and performed by a single resident in pediatric surgery not directly involved in the treatment of the patient. For children younger than 8 years of age, a parent proxy-report was used. Children aged 8 years or older answered the questions together with their parents.

Primary Outcomes

The questions regarding urinary tract function were based on a 10-item questionnaire published by Stenström et al[22] and the International Children's Continence Society's definitions of incontinence, emptying habits, and urinary tract infections (UTIs).[23] Urinary tract dysfunction was defined as having at least 1 of the following symptoms: uncontrolled voiding, difficulty in bladder emptying, or incontinence. Neurogenic bladder was defined as incapacity to store urine or empty the bladder during a urodynamic study. The questions concerning bowel function were based on the validated Bowel Function Score (BFS) developed by Rintala and Lindahl.[24] The questions evaluated bowel frequency, emptying habits, soiling, fecal accidents, and social problems. Soiling was defined as small amounts of fecal staining in the underwear, whereas fecal accidents were defined as the involuntary loss of feces.[25] Bowel dysfunction was defined as 1 or more of the following symptoms: inability to hold back defecation, soiling, fecal accidents, or constipation requiring medical treatment.

Predictors

Medical records of patients with sacrococcygeal teratoma were reviewed for data regarding gestational age at birth, birth weight, age at surgery, tumor size, Altman type,[1] and histopathologic classification. Tumor size was recorded as the largest diameter and volume calculated through preoperative imaging.[26] Extension of the tumor was recorded according to the Altman type, ranging from mainly external (type I) to entirely internal localization (type IV).[1] The tumors were classified as mature, immature, or malignant histologically.

Statistical Analyses

Comparison of functional outcome in patients and control patients was performed by logistic regression for dichotomous outcome variables and linear regression for continuous outcome variables. Assessments were made with adjustment for both age and sex, and ORs and β-coefficients were calculated with 95% CIs. For outcomes with zero cases in the reference group, exact logistic regression was used. Predictors of outcome were analyzed with the Mann–Whitney *U* test for continuous variables and reported as median and [range]. Categorical variables were analyzed with the Fisher exact test. *P* values <.05 were considered significant. Regression models were performed with STATA, version 12.0 (StataCorp LP, College Station, Texas). Remaining statistical analysis was performed with SPSS Statistics, version 22.0 (IBM Corp, Armonk, NY). Statistical analyses were approved by a medical statistician.

Ethical Approval

The study was approved by the regional research ethics committee (registration number 2016/289), and written parental consent was obtained for all patients.

Results

Patient demographics are presented in **Table I**. All 17 patients eligible for inclusion agreed to participate. The median age at the time of study was 7.3 years, and the girl:boy ratio was 3.25:1. Prenatal diagnosis was made in 12 cases (71%), and diagnosis was obvious in another 2 patients at birth. These children were operated at a median age of 3 days (range 0-15 days), whereas the remaining 3 patients were diagnosed later and operated at 2 months, 1.9 years, and 4 years, respectively. One child was diagnosed late due to a misdiagnosis at the local hospital, and 1 child was diagnosed after prolonged constipation. The median size of the teratomas was 9.0 cm, and median tumor volume was 147 cm^3. All children were operated with tumor resection and coccygectomy, and no children were treated with adjuvant therapy. In 1 patient, the teratoma ventrally displaced the bladder and compressed the urethra with a resulting

Table I. Demographics and tumor characteristics of patients born with sacrococcygeal teratomas

Demographics/characteristics	
Age, y*	7.3 [3.5-16.0]
Sex, female/male	76%/24%
Birth weight, g*	3790 [2280-4320]
Gestational age, wk*	38.7 [28.4-40.9]
Tumor diameter, cm*	9.0 [3.5-20]
Tumor, volume, cm^3*	147 [11-2072]
Altman classification	
Type I	41%
Type II	18%
Type III	29%
Type IV	12%
Immature histology	18%
Radicality	
Complete	24%
Uncertain	29%
Nonradical	47%
Reoperation†	29%

*Median [range].
†Reoperation due to recurrence or residual tumor.

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