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Original Article

Long-Term Outcomes of Infantile Sacrococcygeal Teratoma: Results from a Multi-institutional Retrospective Observational Study in Japan

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ABSTRACT

Background: Tumor recurrence, anorectal and urinary dysfunction, and lower limb dysfunction after surgery are observed in infantile sacrococcygeal teratoma (SCT). In this paper, a multi-institutional retrospective observational study was conducted to clarify the long-term functional prognosis in Japan.
Methods: This study was conducted using a paper-based questionnaire distributed to 192 facilities accredited by the Japanese Society of Pediatric Surgeons, covering patients who underwent radical surgery at less than 1 year old and who survived for at least 180 days after birth from 2000 to 2019.
Results: A total of 355 patients were included in this analysis. Altman type was I-II in 248 and type III-IV in 107, and the median maximum tumor diameter was 6.1 (range: 0.6–36.0) cm. There were 269 mature teratomas, 69 immature teratomas, and 10 malignant tumors. Total resection was performed in 325, subtotal or partial resection in 27, and surgical complications were noted in 54. The median post-operative follow-up was 6.6 (0.5–21.7) years. Eighty-three patients (23.4 %) had functional sequelae, including 62 (17.5 %) with anorectal dysfunction, 56 (13.0 %) with urinary dysfunction, and 15 (4.2 %) with lower limb motor dysfunction. Recurrence occurred in 42 (11.8 %) at a median age of 16.8 (1.7–145.1) months old. Risk factors for dysfunction included preterm delivery, a large tumor diameter, Altman type III-IV, incomplete resection, and surgical complications. Risk factors for recurrence included immature teratoma or malignancy, incomplete resection, and surgical complications.
Conclusions: Postoperative dysfunction was not low at 23.4 %, and 11.8 % of the patients experienced recurrence occurring more than 10 years after surgery, suggesting the need for periodic imaging and tumor markers evaluations in patients with risk factors. It is necessary to establish treatment guidelines for best practice monitoring of the long-term quality of life.
Level of evidence: Level II Retrospective Study.

1. Introduction

Sacrococcygeal teratoma (SCT) is an uncommon extragonadal germ cell tumor in neonates, infants, and toddlers. This lesion consists of a solid and/or cystic component that develops from the tip of the sacrum, protruding outward from the buttocks, or within the pelvic cavity. The incidence of SCT is estimated to be 1 in every 40,000 births, and the male-to-female ratio is approximately 1:3, being more frequent in girls than in boys. Most cases are diagnosed at birth, and yolk sac tumors (YSTs) occur more frequently in children after infancy more than neonates [1].

Although most cases of infantile SCT are benign tumors in nature, including mature or immature teratomas with an excellent prognosis, some develop into extremely large lesions, leading to massive bleeding, high-output heart failure, and disseminated intravascular coagulopathy (DIC), and fatal outcomes during the neonatal period. In addition, even after successful tumor resection, some patients may present with tumor recurrence, malignant

Abbreviations: SCT, Sacrococcygeal Teratoma; QOL, Quality of Life; CIC, clean intermittent catheterization; DIC, disseminated intravascular coagulopathy; YST, yolk sac tumor; CNS, central nervous system; OR, odds ratio; CI, confidence interval; AFP, alpha-fetoprotein.

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transformation, anorectal and urinary dysfunctions, and lower limb palsy during long-term follow-up [2,3]. In 2017, the authors published clinical guidelines for SCT in Japan to provide physicians with information regarding the clinical practice of managing SCT [4]. However, no domestic multicentric cohort study involving the long-term follow-up of SCT has yet been performed, which prompted us to conduct the present study.

The incidence of and factors that contribute to these sequelae are relatively unknown, as is how patients experience these sequelae. In addition, the recurrence of SCT has become a major challenge that needs to be addressed. Identification of the risk factors for functional sequelae and recurrence may lead to better insight into the symptomatic and biological behavior of this tumor.

We, therefore, reviewed a multicentric cohort of SCT based on a questionnaire to clarify the Japanese trends in SCT sequelae, with the objective of improving the prognosis and quality of life (QOL) of this disease.

2. Methods

2.1. Participants

A paper-based surgery questionnaire was administered to all 192 facilities accredited by the Japanese Society of Pediatric Surgeons and education-related facilities. The inclusion criteria for the survey were patients who had undergone radical surgery at less than 1 year old and who survived for at least 180 days after birth during the 20-year period from 2000 to 2019.

2.2. Survey items

Patient background characteristics, the presence of a prenatal diagnosis, Altman's classification, treatment, surgical complications (DIC, sepsis, intracranial hemorrhage, wound infection, organ injury, re-excision of residual tumor), tumor size, the pathological diagnosis, current functional disability, and recurrence were investigated. To evaluate the postoperative dysfunctions, symptoms were classified according to the definition of Masahata et al. [3]. The details of the bowel function were classified into five items (0: normal bowel movements, 1: fecal incontinence, 2: soiling, 3: constipation requiring laxatives or enema, and 4: severe constipation requiring bowel irrigation or stool extraction). Patients classified as 1–4 were considered to have anorectal dysfunction. The details of the urinary function were also classified into five items (0: normal urination, 1: urinary incontinence, 2: repeated urinary tract infection, 3: neurogenic bladder, and 4: clean intermittent catheterization [CIC]). Patients classified as 1–4 were considered to have urinary dysfunction. The details of the lower leg function were classified into three items (0: normal function, 1: limp, 2: walking difficulty). Postoperative scarring of the hip was evaluated to determine cosmetic acceptability. The sexual function, including the presence of erectile and ejaculatory dysfunctions, was also assessed, and the age at tumor recurrence and the pathology of recurrent tumors were investigated.

2.3. Statistical analyses

Univariate and multivariate relative risk analyses for the sequelae of functional impairment, including anorectal and urinary dysfunction, lower-limb motor dysfunction, and tumor recurrence, were performed using logistic regression. The investigated risk factors for the analysis of postoperative dysfunctions were as follows: premature birth (earlier than 37 gestational weeks), low birth weight (<2500 g), presence of a prenatal diagnosis (Yes/No), age at diagnosis (<8 days vs. ≥ 8 days old), maximum tumor diameter (<5.0 cm vs. 5.0–9.9 cm, 10.0–14.9 cm, or >15 cm), Altman classification (I-II vs. III-IV), pathology (mature vs. immature or malignancy), extent of resection (total vs. subtotal or partial), and presence of surgical complications. In addition, the risk factors investigated for the analysis of tumor recurrence were as follows: age at diagnosis (<8 days vs. ≥ 8 days old), maximum tumor diameter (<5.0 cm vs. 5.0–9.9 cm, 10.0–14.9 cm, or >15 cm), Altman classification (I-II vs. III-IV), pathology (mature vs. immature or malignancy), extent of resection (total vs. subtotal or partial), and presence of surgical complications.

Statistical analyses were performed using the JMP® software program, ver. 14.2.0 (SAS Institute Inc., Cary, NC, USA). Data are reported as the median and range or as frequencies and percentages. Proportions are presented with 95 % confidence intervals (CIs). Statistical significance was defined as $p < 0.05$.

3. Results

Of the 192 institutions asked for survey, 73 centers (38.0 %) responded and clinical data of 388 patients with a diagnosis of SCT in Japan during study period were collected. Ultimately, a total of 355 cases were included in the analysis after exclusion of duplicates and ineligible cases.

3.1. Background characteristics

Of the 355 cases with infantile SCT, there were 264 females and 91 males with a median gestational age of 38.6 (range: 26.0–42.3) weeks old and median birth weight of 3026 (range: 1020–5344) g. A total of 158 patients (44.5 %) were prenatally diagnosed, and the median age at diagnosis was 0 (range: 0–343) days. The Altman classification was type I in 175 (49.3 %), type II in 73 (20.6 %), type III in 38 (10.7 %), and type IV in 69 (19.4 %), and the median maximum tumor diameter was 6.1 (range: 0.6–36.0) cm. The pathology of the initial tumor was mature teratoma in 269 (75.8 %), immature teratoma in 69 (19.4 %), and malignancy in 10 (2.8 %). The median age at primary surgery for SCT was 15 (range: 0–364) days. Regarding the extent of tumor resection, total resection was performed in 325 (91.5 %), subtotal resection (≥90 %) in 22 (6.2 %), and partial resection (<90 %) in 5 (1.4 %). Perioperative complications were observed in 54 patients. The median follow-up period after birth was 6.6 (range: 0.5–21.7) years. The details of the patients' background characteristics are presented in Table 1.

3.2. Postoperative functional sequelae

A total of 83 patients (23.4 %) had postoperative functional sequelae, including 62 patients (17.5 %) with anorectal dysfunction, 46 (13.0 %) with urinary dysfunction, and 15 (4.2 %) with lower limb motor dysfunction (there was some overlap) (Table 1). Fig. 1 shows a Venn diagram presenting the three overlapping dysfunctions. Among the 62 patients with anorectal dysfunction, 17 (27.4 %) suffered from soiling or fecal incontinence, 44 (71.0 %) required laxatives or enema, and 6 (9.7 %) had severe constipation requiring bowel irrigation or stool extraction. Of the 46 patients with urinary dysfunction, 8 (17.4 %) presented with urinary incontinence, and 36 (78.3 %) had severe urinary impairment, including repeated urinary tract infection, neurogenic bladder, and the need for CIC. Cosmetically unacceptable scars were noted in 55 of the 355 patients (15.5 %), 18 of whom underwent excision of the scar.

In the univariate analysis, premature birth before 37 weeks' gestation, a low birth weight <2500 g, the presence of a prenatal diagnosis, a maximum tumor diameter ≥10.0 cm, Altman classification type III and IV, incomplete tumor resection (subtotal or partial), and the presence of surgical complications, were

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Table 1
Demographics and clinical parameters of infantile sacrococcygeal teratomas.

Demographic data			Variable	
Patients' demographic				
Total number of patients		n (%)	355	(100.0)
Sex; female		n (%)	264	(74.4)
Gestational age at birth (wk)		median (range)	38.6	(26.0–42.3)
Birth weight (g)		median (range)	3026	(1020–5344)
Presence of prenatal diagnosis		n (%)	158	(44.5)
Age at diagnosis (day)		median (range)	0	(0–343)
Accompanied anomalies	urologic anomalies	n (%)	10	(2.8)
	spinal cord anomalies	n (%)	18	(5.1)
	cardiac anomalies	n (%)	12	(3.4)
	anorectal malformation	n (%)	17	(4.8)
	genetic disorders	n (%)	6	(1.7)
Status of tumor				
Maximum tumor diameter (cm)		median (range)	6.1	(0.6–36.0)
Altman' classification	I	n (%)	175	(49.3)
	II	n (%)	73	(20.6)
	III	n (%)	38	(10.7)
	IV	n (%)	69	(19.4)
Pathology	mature	n (%)	269	(75.8)
	immature	n (%)	69	(19.4)
	malignancy	n (%)	10	(2.8)
Details of primary surgery				
Age at surgery (day)		median (range)	15	(0–364)
Surgical approach	perineal	n (%)	268	(75.5)
	abdominal	n (%)	6	(1.7)
	abd + peri	n (%)	78	(22.0)
Extent of resection	total (100 %)	n (%)	325	(91.5)
	subtotal (≥90 %)	n (%)	22	(6.2)
	partial (<90 %)	n (%)	5	(1.4)
Surgical complications	DIC, sepsis	n (%)	4	(1.1)
	intracranial hemorrhage	n (%)	3	(0.8)
	wound infection, wound dehiscence	n (%)	18	(5.1)
	organ injury	n (%)	11	(3.1)
	re-excision of residual tumor	n (%)	18	(5.1)
Details of follow-up				
Follow-up period after birth (yr)		median (range)	6.6	(0.5–21.7)
Long-term complications	anorectal dysfunction	n (%)	62	(17.5)
	urinary dysfunction	n (%)	46	(13.0)
	lower limb motor dysfuction	n (%)	15	(4.2)
	CNS damage	n (%)	16	(4.5)
	cosmetically unacceptable scarring	n (%)	55	(15.5)
Reccurence of tumor		n (%)	42	(11.8)
	malignancy	n (%)	22	(6.2)
Age at reccurence (mo)		median (range)	16.8	(1.7–145.1)

DIC; disseminated intravascular coagulopathy CNS; central nervous system.

significant risk factors for postoperative dysfunctions. The presence of a prenatal diagnosis, large tumor diameter, Altman's classification, and surgical complications were also remained significant in the multivariate analysis (Table 2).

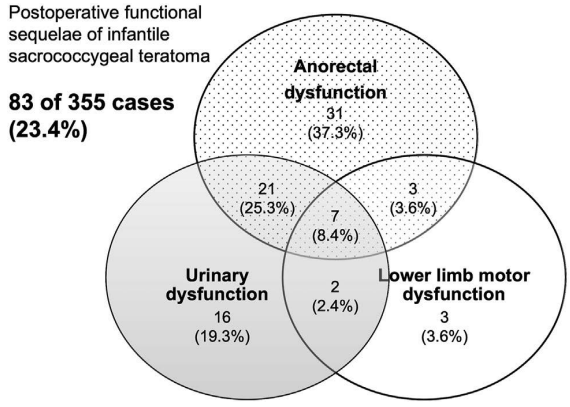

Fig. 1. A Venn diagram of postoperative functional sequelae of infantile sacrococcygeal teratoma in Japan.

3.3. Tumor recurrence

Tumor recurrence occurred in 42 patients (11.8 %) with a median age of 16.8 (1.7–145.1) months old. Nineteen patients were diagnosed through imaging studies, and 22 had elevated serum alpha-fetoprotein (AFP) levels at diagnosis of recurrence. Twenty-two of the 42 patients (52.4 %) were diagnosed with malignant germ cell tumors (immature teratoma, YST, or embryonal carcinoma), including 2 with a primary YST tumor at the initial diagnosis of SCT. The median age at malignant recurrence was 12.7 (1.7–36.9) months old. They underwent surgery with or without chemotherapy, and one patient eventually died of cancer. Regarding the initial surgery, 34 of the 325 cases undergone total resection had tumor recurrence (10.5 %), while among the 27 cases undergone incomplete resection (subtotal or partial), 9 underwent re-do surgery due to residual mass, and 8 cases had tumor recurrence including 3 malignancies eventually (29.6 %). Regarding the relationship between the initial and second pathology, patients with mature teratoma at recurrence (n = 16) also had mature teratoma (including unknown pathology) at the initial pathological diagnosis, while the patients with malignancy at recurrence (n = 22) had mature teratoma (n = 12), immature teratoma (n = 6), malignancy (n = 2), and unknown diagnosis (n = 2) at the initial pathological diagnosis.

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Table 2
Logistic regression analysis of postoperative dysfunctions.

Characteristics		Univariable			Multivariable		
		OR	(95 % CI)	*p*	OR	(95 % CI)	*p*
Premature birth	< 37 w	0.438	(0.240–0.797)	0.007*			
low Birth weight	<2500 g	2.036	(1.002–4.137)	0.049*			
Presence of prenatal diagnosis	Yes	1.791	(1.088–2.950)	0.022*	2.691	(1.076–6.728)	0.034*
Age at diagnosis	<8 d vs. ≥8 d	0.803	(0.459–1.404)	0.441			
Maximum tumor diameter	<5.0 cm	1.000			1.000		
	5.0–9.9 cm	0.647	(0.325–1.288)	0.215			
	10.0–14.9 cm	2.854	(1.397–5.834)	0.004*	2.972	(1.018–8.680)	0.046*
	≥15.0 cm	2.925	(1.293–6.618)	0.010*			
Altman' classification	III–IV vs. I–II	2.321	(1.392–3.869)	0.001*	4.494	(2.079–9.711)	<0.001*
Pathology	mature vs. immature or malignancy	0.581	(0.333–1.013)	0.056			
Extent of resection	subtotal or partial vs. total	2.504	(1.112–5.640)	0.027*			
Surgical complications	Yes	3.073	(1.629–5.798)	0.001*	3.506	(1.580–7.782)	0.002*

OR: odds ratio, CI: confidence interval.
p value was significant <0.05.

In the logistic regression analysis of tumor recurrence, pathology (immature teratoma or malignancy), incomplete tumor resection (subtotal or partial), and presence of surgical complications were significant risk factors in the univariate analysis, while incomplete tumor resection was the only significant risk factor in the multivariate analysis (Table 3).

4. Discussion

In 2009, the Japanese SCT Study Group was established with the support of a grant from The Ministry of Health, Labour and Welfare of Japan. As a precursor of this study, a nationwide retrospective cohort study was conducted on 97 fetuses prenatally diagnosed with SCT between 2000 and 2009. As a result, the overall mortality of prenatally diagnosed SCT excluding terminations was 16 %. Early delivery, predominantly solid component tumors, and histological immaturity were shown to be associated with an increased risk of mortality [5,6]. The next project of this group was the establishment of clinical guidelines for SCT, which will potentially contribute to the improvement of the prognosis and QOL of SCT patients [4]. However, these studies concentrated on the short-term outcomes of prenatally SCT, and there is a lack of information on long-term survivors in Japan, therefore, the present study was planned and conducted to determine the current status of patients with functional sequelae from a nationwide study, as well as the contributing factors associated with the development of functional sequelae. Judging from the birth prevalence of SCT, the number of patients with SCT predicted was estimated to be approximately 500 cases in Japan during 2000–2019, so the patient number in our study therefore corresponded to approximately 70 % of the estimated cases for that period.

In the present study, 83 patients (23.4 %) had postoperative functional sequelae, including 62 patients (17.5 %) with anorectal dysfunction, 46 patients (13.0 %) with urinary dysfunction, and 15 patients (4.2 %) with lower limb motor dysfunction, and these three major sequelae overlapped in 40.0 % of the cases. These findings indicate that these sequelae are complex complications with possible common causes. In this survey, the risk factors for these sequelae were premature birth (before 37 weeks' gestation), low birth weight (<2500 g), a prenatal diagnosis, maximum tumor diameter ≥10.0 cm, Altman classification type III and IV, incomplete tumor resection (subtotal or partial), and the presence of surgical complications. The origin of these dysfunctions is still debated, and the possible causes of them are myogenic and/or neurogenic impairment due to the compression effect of the large tumor and surgical damage to the pelvic organs and pelvic floor muscles. Therefore, premature infants and larger masses, especially those developing into the pelvic cavity (Altman III and IV), and the presence of surgical complications seem to be reasonable as the contributing factors to dysfunction.

The reported incidence of an impaired bowel function ranged from 19 % to 38 %, while that of urinary incontinence was approximately 50 % [7–12]. Other reported risk factors included obstruction of the urinary tract and intestinal tract in the prenatal imaging diagnosis, tumor recurrence, types other than Altman type I, and abdominosacroperineal resection for Altman type III and IV. Impaired movement of the lower leg has been reported in 5.0%–11.1 % of cases [4], but there has been little description of the extent of such impairment. These functional sequelae are relatively common in the patients with SCT, and it is recommended that these facts be mentioned to the patient's families before the course of treatment is decided. However, Cozzi et al. reported that there was

Table 3
Logistic regression analysis of tumor reccurence.

Characteristics		Univariable			Multivariable		
		OR	(95 % CI)	*p*	OR	(95 % CI)	*p*
Age at diagnosis	<8 d vs. ≥8 d	2.109	(0.855–5.201)	0.105			
Maximum tumor diameter	<5.0 cm	1.000					
	5.0–9.9 cm	0.970	(0.402–2.340)	0.946			
	10.0–14.9 cm	2.109	(0.820–5.421)	0.121			
	≥15.0 cm	1.565	(0.497–4.927)	0.444			
Altman' classification	III–IV vs. I–II	1.183	(0.596–2.348)	0.631			
Pathology	mature vs. immature or malignancy	0.477	(0.242–0.938)	0.032*			
Extent of resection	subtotal or partial vs. total	4.742	(1.964–11.451)	0.001*	4.979	(1.753–14.148)	0.003*
Surgical complications	Yes	3.078	(1.448–6.543)	0.004*			

OR: odds ratio, CI: confidence interval.
p value was significant <0.05.

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no significant difference in functional disorders between adult patients with SCT and the control group [13]. These facts imply that it is important to recognize these long-term complications and ensure careful and regular follow-up, as some symptoms can be ameliorated during follow-up.

Regarding cosmetically unacceptable scarring, 15.5 % of patients with SCT complained of unacceptable surgical wounds in this study. Derikx et al. found that 40.3 % of SCT patients complained surgical wounds problems, and the diagnosis before the age of 8 days old and large tumors were significant risk factors [8]. Although cosmetic wounds do not prioritize the prompt treatment for the life-threatening condition of SCT, physicians should listen to the patients' complaints in the outpatient clinic during follow-up and consider plastic surgery for disfigurement, if indicated.

The recurrence rate of SCTs is reported to be 8.3 %–13.0 % within 5–6 years after surgery [6,14,15]. A meta-analysis study of 1516 patients with SCT showed that the pooled proportions of mature and malignant recurrences were 3 % and 5 %, respectively, and many instances of recurrences occurred within 1–6 years, with some cases occurring as long as 20 years after the initial diagnosis [16]. In this study, the recurrence rate was 11.8 %, and the median age at occurrence was 1 (0–12) years old. The recurrence rate was 6.2 % for malignant cases, with a median age at occurrence of 1 (0–3) years old. The risk factors associated with SCT recurrence included immature of malignant pathology at the initial surgery, incomplete tumor resection, and the presence of surgical complications. Other reported risk factors were unresected coccyx bone, tumor spillage, and failure to detect malignant components within the tumor, which were not elucidated in this study because of the nature of the questionnaire study. To eliminate the chances of tumor recurrence and improve the survival, physicians should endeavor to perform complete surgical resection without surgical complications, understand the importance of regular and careful follow-up, and be mindful that regular measurement of serum AFP is recommended for the early detection of malignant recurrence for at least three years after the treatment, as AFP is highly sensitive for malignancy [4].

Several limitations associated with the present study warrant mention. This study was conducted retrospectively using a paper-based questionnaire administered to physicians. The individual patient records were not centrally reviewed, so the quality of data was not guaranteed. Many of the institutions had a small number of cases, and the surgical procedure and management were determined according to the clinical decisions of each institution. Regarding the definition of dysfunction, the missing comments were considered to indicate no dysfunction, so we may have underestimated the frequency of dysfunctions. Anorectal dysfunction was defined as constipation requiring laxatives or an enema, which may have been caused by factors other than surgery for SCT, and possibly overestimated. This study included a questionnaire on the sexual function; however, we did not receive any responses concerning this subject. One possible reason was that the target age group was relatively young, even up to 20 years after surgery. Another reason is that questions related to sexuality tend to be particularly sensitive to answer for Japanese people. Therefore, a long-term prospective study including sexual problems in SCT patients is needed to establish a comprehensive treatment strategy.

In conclusion, this study was a multi-institutional retrospective observational study on the long-term outcomes of infantile SCT in Japan. The rate of postoperative dysfunction was not low at 23.4 %, and 11.8 % of the patients experienced recurrence occurring more than 10 years after surgery, suggesting the need for periodic imaging and tumor marker evaluations in patients with risk factors. Unacceptable cosmetic scarring was a common problem. Given these findings, it is necessary to establish treatment guidelines for best practice monitoring of the long-term QOL.

Ethical statement

This national survey was approved by The Japanese Society of Pediatric Surgeons, and the institutional board review approval (ERB-C-1953). Informed consent was obtained in the form of opt-out on the website. This study was performed in accordance with the Ethical Guidelines for Clinical Research published by the Ministry of Health, Labour and Welfare of Japan on July 30, 2003 (revised 2008) and complied with the Helsinki Declaration of 1964 (revised 2013).

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Conflicts of interest

The authors declare there are no conflicts of interest associated with this study.

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References

[1] Rescorla FJ. Teratomas and other germ cell tumors. In: Coran AG, Adzick NS, Krummel TM, Laberge JM, Shamberger RC, Caldamone AA, editors. Pediatric surgery. 7th ed. Philadelphia: Elsevier Saunders; 2012. p. 947–57.

[2] Braungart S, James EC, Powis M, Gabra H, , CCLG Surgeons Collaborators, Losty PD. Sacrococcygeal teratoma: long-term outcomes. A UK CCLG Surgeons group nationwide study. Pediatr Blood Cancer 2023;70. https://doi.org/10.1002/pbc.29994.

[3] Masahata K, Ichikawa C, Makino K, Abe T, Kim K, Yamamichi T, et al. Long-term functional outcome of sacrococcygeal teratoma after resection in neonates and infants: a single-center experience. Pediatr Surg Int 2020;36:1327–32. https://doi.org/10.1007/s00383-020-04752-7.

[4] Fumino S, Tajiri T, Usui N, Tamura M, Sago H, Ono S, et al. Japanese clinical practice guidelines for sacrococcygeal teratoma, 2017. Pediatr Int 2019;61:672–8. https://doi.org/10.1111/ped.13844.

[5] Usui N, Kitano Y, Sago H, Kanamori Y, Yoneda A, Nakamura T, et al. Outcomes of prenatally diagnosed sacrococcygeal teratomas: the results of a Japanese nationwide survey. J Pediatr Surg 2012;47:441–7. https://doi.org/10.1016/j.jpedsurg.2011.08.020.

[6] Yoneda A, Usui N, Taguchi T, Kitano Y, Sago H, Kanamori Y, et al. Impact of the histological type on the prognosis of patients with prenatally diagnosed sacrococcygeal teratomas: the results of a nationwide Japanese survey. Pediatr Surg Int 2013;29:1119–25. https://doi.org/10.1007/s00383-013-3384-7.

[7] Shalaby MS, Walker G, O'Toole S, Hammond P, Carachi R. The long-term outcome of patients diagnosed with sacrococcygeal teratoma in childhood. A study of a national cohort. Arch Dis Child 2014;99:1009–13. https://doi.org/10.1136/archdischild-2014-306414.

[8] Derikx JPM, De Backer A, van de Schoot L, Aronson DC, de Langen ZJ, van den Hoonaard TL, et al. Long-term functional sequelae of sacrococcygeal teratoma: a national study in The Netherlands. J Pediatr Surg 2007;42:1122–6. https://doi.org/10.1016/j.jpedsurg.2007.01.050.

[9] Partridge EA, Canning D, Long C, Peranteau WH, Hedrick HL, Adzick NS, et al. Urologic and anorectal complications of sacrococcygeal teratomas: prenatal and postnatal predictors. J Pediatr Surg 2014;49:139–43. https://doi.org/10.1016/j.jpedsurg.2013.09.042.

[10] Braungart S, James EC, Powis M, Gabra H, , CCLG Surgeons Collaborators, Losty PD. Sacrococcygeal teratoma: long-term outcomes. A UK CCLG Surgeons group nationwide study. Pediatr Blood Cancer 2023;70. https://doi.org/10.1002/pbc.29994.

[11] Salim A, Raitio A, Losty PD. Long-term functional outcomes of sacrococcygeal teratoma – a systematic review of published studies exploring 'real world' outcomes. Eur J Surg Oncol 2022. https://doi.org/10.1016/j.ejso.2022.09.002. S0748798322006539.

[12] Hambraeus M, Al-Mashhadi A, Wester T, Svensson P-J, Stenström P, Lilja HE. Functional outcome and health-related quality of life in patients with sacrococcygeal teratoma – a Swedish multicenter study. J Pediatr Surg 2019;54:1638–43. https://doi.org/10.1016/j.jpedsurg.2018.10.044.

[13] Cozzi F, Schiavetti A, Zani A, Spagnol L, Totonelli G, Cozzi DA. The functional sequelae of sacrococcygeal teratoma: a longitudinal and cross-sectional follow-up study. J Pediatr Surg 2008;43:658–61. https://doi.org/10.1016/j.jpedsurg.2007.10.066.

[14] Derikx JP, De Backer A, van de Schoot L, et al. Factors associated with recurrence and metastasis in sacrococcygeal teratoma. Br J Surg 2006;93:1543–8.

[15] Padilla BE, Vu L, Lee H, MacKenzie T, Bratton B, O'Day M, et al. Sacrococcygeal teratoma: late recurrence warrants long-term surveillance. Pediatr Surg Int 2017;33:1189–94. https://doi.org/10.1007/s00383-017-4132-1.

[16] Kops AL, Hulsker CC, Fiocco M, Zsiros J, Mavinkurve-Groothuis AMC, Looijenga LH, et al. Malignant recurrence after mature Sacrococcygeal teratoma: a meta-analysis and review of the literature. Crit Rev Oncol Hematol 2020;156:103140. https://doi.org/10.1016/j.critrevonc.2020.103140.

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