Journal of Pediatric Surgery xxx (xxxx) xxx

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.sciencedirect.com/journal/journal-of-pediatric-surgery

Clinical Research Paper

Malignant transformation of sacrococcygeal teratoma versus presacral teratoma in Currarino syndrome: Results of 'The SCT-study'

Lieke J. van Heurn [a,*], Joep P.M. Derikx [a], Nigel J. Hall [b], Amr Abdelhamid AbouZeid [c], Simone de Campos Vieira Abib [d], Lohfa B. Chirdan [e], Shigehisa Fumino [f], Seong C. Kim [g], Oliver Muensterer [h], Maricarmen Olivos [i], Shawn D. StPeter [j], Jos Twisk [k], Nicolas Vinit [l], Tianyou Yang [m], Augusto Zani [n], L.W. Ernst van Heurn [a], The SCT-study consortium

[a] *Emma Children's Hospital, Amsterdam UMC, University of Amsterdam & Vrije Universiteit Amsterdam, Department of Pediatric Surgery, Amsterdam, the Netherlands*
[b] *University Surgery Unit, Faculty of Medicine, University of Southampton, Southampton, United Kingdom*
[c] *Faculty of Medicine, Department of Pediatric Surgery, Ain Shams University, Cairo, Egypt*
[d] *Pediatric Oncology Institute - GRAACC - Federal University of São Paulo, Brazil*
[e] *Pediatric Surgery Unit, Department of Surgery, Jos University Teaching Hospital, Jos, PMB 2076, Jos, Nigeria*
[f] *Department of Pediatric Surgery, Kyoto Prefectural University of Medicine, Kyoto, Japan*
[g] *Department of Pediatric Surgery, University of Ulsan College of Medicine and Asan Medical Center, Seoul, South Korea*
[h] *Department of Pediatric Surgery, Dr. von Hauner Children's Hospital, University Hospital, LMU Munich, Germany*
[i] *Pediatric Surgeon, Hospital de Niños Dr. Roberto del Rio-Universidad de Chile, Santiago, Chile*
[j] *Department of Surgery – Children's Mercy Kansas City, Kansas City, USA*
[k] *Department of Epidemiology and Data Science, Amsterdam Public Health Research Institute, Amsterdam UMC, Vrije Universiteit Amsterdam, De Boelelaan 1089a, 1081 HV, Amsterdam, the Netherlands*
[l] *Department of Pediatric Surgery and Urology, Necker-Enfants Malades Hospital, APHPand Université Paris Cité, Paris, France*
[m] *Department of Pediatric Surgical Oncology, Guangzhou Women and Children's Medical Center, Guangzhou Medical University, Guangdong, China*
[n] *Division of General and Thoracic Surgery, The Hospital for Sick Children, ON, Canada*

ARTICLE INFO

Article history:
Received 5 August 2025
Received in revised form
15 November 2025
Accepted 26 November 2025

Keywords:
Sacrococcygeal teratoma
Currarino syndrome
Recurrence
Malignant transformation

ABSTRACT

Background: The risk of malignant transformation and recurrence of Sacrococcygeal Teratoma (SCT) is relatively high, while it is possibly lower in cases associated with Currarino Syndrome (CS). However, the existing literature gives contradictory results. We aimed to examine the risk of malignant transformation in a large cohort of SCT and CS patients.

Methods: In a global retrospective cohort study, data of consecutive SCT patients and CS patients with presacral teratoma was obtained from 132 institutes in 62 countries. Malignant transformation, defined as malignancy at initial resection, malignant recurrence, or death due to malignancy was analysed for SCT and CS patients. The rate of malignant transformation was analysed with log-rank test and compared between groups.

Results: Of 3612 patients with presacral teratoma, 3388 entered analysis; 3183 SCT and 205 CS patients. The percentage of patients with malignant transformation at initial resection was higher in the SCT versus the CS group, 10·3 % and 31·9 % after one and two years, versus 4·2 % in CS patients after two years, respectively ($p < 0.001$). Histology in recurrent teratoma was malignant in 35·4 % ($n = 114$) of SCT patients and 5·9 % ($n = 1$) of CS patients ($p = 0.005$). Survival in both groups was equivalent at 94·9 % in SCT patients versus 96·9 % in CS patients ($p = 0·343$)

Conclusion: The SCT-study shows that malignancy is more often present in SCT than in CS patients with an increasing risk of malignant transformation with age compared to CS patients in whom malignancy is rare. Recurrence after resection was more often malignant in the SCT group.

Level of evidence: level III.

Abbreviations: AFP, Alpha-fetoprotein; CS, Currarino Syndrome; CT, Computed tomography; MRI, Magnetic resonance imaging; SCT, Sacrococcygeal teratoma; YST, yolk sac tumour.

* Corresponding author. Department of Paediatric Surgery, Amsterdam UMC, Meibergdreef 9, 1105 AZ Amsterdam, the Netherlands.
E-mail address: l.j.vanheurn@amsterdamumc.nl (L.J. van Heurn).

https://doi.org/10.1016/j.jpedsurg.2025.162848

Please cite this article as: van Heurn LJ et al., Malignant transformation of sacrococcygeal teratoma versus presacral teratoma in Currarino syndrome: Results of 'The SCT-study', Journal of Pediatric Surgery, https://doi.org/10.1016/j.jpedsurg.2025.162848

1. Introduction

Sacrococcygeal teratoma (SCT) is the most common germ cell tumour in neonates. Most tumours are diagnosed before or immediately after birth, but a small proportion of these tumours, particularly those with an internal position, is discovered at an older age. The risk of malignant transformation increases with age. In 11–35 % of patients, the tumour is malignant at the moment of initial resection [1]. Malignancy rates up to 70 % have been reported if SCT is diagnosed at the age of one year [2].

Currarino Syndrome (CS) is a hereditary disorder with a sacral bony defect, a presacral mass and an anorectal malformation or constipation [3]. Due to the phenotypic expression, the clinical presentation is variable, and a combination of all the defects is not necessary to make the diagnosis [4,5]. If a presacral tumour is present, 25–40 % are teratomas [6,7]. Presacral teratomas associated with CS are almost obligatory internal and are therefore rarely diagnosed prenatally or during early infancy [8]. As a result, CS associated presacral teratomas are often resected at an older age compared to SCTs [9]. Despite late diagnosis and late resection, malignancy in CS associated teratoma is rare and the risk of malignant transformation is low [9,10].

Recurrent SCT after resection occurs in 2–33 % of SCT patients and is often malignant with yolk sac tumour (YST) components in 22–56 % of recurrences [11–14]. Tumour recurrence decreases the overall 10-year survival to 60 % [15–17]. Recurrence after initial teratoma resection in CS patients has been rarely described [9,18].

Most studies of SCT and CS are relatively small due to the rarity of presacral teratomas and conclusions on malignant transformation remain difficult to draw. Therefore, we examined the risk of malignant transformation and tumour recurrence in SCT versus CS associated teratoma in a large, global cohort of patients.

2. Methods

2.1. Study design and participants

We performed a global retrospective cohort study of presacral teratoma patients: 'The SCT-study'. STROBE guidelines were followed [19].

We recruited as many patients from participating hospitals as possible regardless of geography. Paediatric surgeons and paediatric oncologists were invited to participate in the study through personal communication, the European Paediatric Surgeons' Association (EUPSA) Network Office, Pubmed publications, and a network of national and international study leads. Study information was provided in English, Spanish, French, and Russian. Participation was voluntary; no payment was made for data collection. Centres, which were anticipated to include ten or more patients were invited to participate. An exception was made for centres from low and lower-middle income countries [20]. For these centres, the minimum number of inclusions was set at five. The supplementary appendix provides an overview of the participating countries.

Due to the rarity of the condition, no sample size calculation was performed so all eligible patients were included. Exclusion criteria were (a) born before 1982 since Currarino Syndrome was first described in 1981, or (b) born after 2020 [3].

The Medical Ethical Board of Amsterdam University Medical Centre (Amsterdam UMC), determined that Medical Research Involving Human Subject Act (WMO) does not apply to the study and that official approval of the committee was not required (reference number W19_329 # 19.388). Participating centres obtained local approval to participate in the study following their own legal and ethical regulations. Data transfer agreements between the participating centres and Amsterdam UMC for data transfer approval were used to guarantee safe data use and storage.

Charts were reviewed by the local investigator. Data were validated with warning messages about possible errors when entered in Castor Electronic Data Capture (EDC). Furthermore, Castor files were structured so that out-of-range values could not be entered. Dependency fields were used for data and included initial resection, recurrence, death and follow-up. Furthermore, overall data distribution and frequencies in SPSS were checked to detect invalid entered data.

2.2. Procedures

Data was anonymized by the local investigator to transform personal data, which could be directly linked to an individual patient, into general information. Transformed patient data were uploaded in Castor EDC and encrypted [21]. Every participating centre had its own Castor link and had access only to its own transformed patient data.

Which data were collected was decided upon by a group of experienced paediatric surgeons with interest in SCT treatment and was based on main outcome variables used in previously published studies of malignant transformation of SCT and recurrent SCT.

Collected data included generic and condition-specific variables. Generic variables included: Castor EDC centre number, country, gender (male/female/unknown), age at diagnosis (days), pre-operative imaging modalities (none/ultrasound/computed tomography/magnetic resonance imaging/unknown), initial tumour resection at the participating centre (yes/no/unknown), age at initial resection (days), outcome (survival/deceased/unknown), age at follow-up (days), age at death (days), and cause of death. Condition-specific variables were Altman classification (I/II/III/IV/unknown) [22], CS (yes/no/unknown), initial SCT treatment (chemotherapy/surgery/no treatment/unknown), pathology (mature/immature/malignant/unknown), recurrence (yes/no/unknown), period between birth and recurrence (days), detection of recurrence (clinical examination/imaging/AFP/unknown), serum AFP-level at recurrence (μg/L), recurrent SCT pathology (mature/immature/malignant/unknown) and treatment of recurrent SCT (chemotherapy/surgery/no treatment/unknown).Cause of death was collected as a free-text category.

2.3. Statistical analysis and definitions

Data are presented as mean with standard deviation (SD) if normally distributed and median with interquartile range (IQRs) if skewed; count data are presented as numbers and percentages. Differences in patient demographics between SCT and CS patients were analysed with Fisher exact test for categorical variables, unpaired t-test for normally distributed continuous variables and Mann–Whitney U test for non-normal continuous variables. Kaplan–Meier curves and Cox proportional hazards regression analyses were used to evaluate risk of malignant transformation and recurrence. Hazard ratios (HR) with 95 % confidence intervals (CI) were reported, and a p-value of <0.05 was considered statistically significant.Statistical analyses were performed using SPSS for Windows version 25.0 software (SPSS) and GraphPad Prism 8 (GraphPad Software, Incl.).

2.4. Definitions

SCTs were classified according to the criteria proposed by the Surgical Section of the American Academy of Paediatrics [22]. In

Please cite this article as: van Heurn LJ et al., Malignant transformation of sacrococcygeal teratoma versus presacral teratoma in Currarino syndrome: Results of 'The SCT-study', Journal of Pediatric Surgery, https://doi.org/10.1016/j.jpedsurg.2025.162848

Currarino Syndrome, the tumour was associated with an anorectal malformation or anal stenosis and or/sacral bone defect [3]. Recurrence was defined as relapse of the teratoma at least three months after initial resection [10]. Tumour identification within three months after surgery is more likely due to an incomplete resection, therefore, these cases were excluded from data analysis. Malignancy free survival was defined as time from birth to malignancy or death due to malignancy and included initial malignancies, malignant recurrences and deaths due to malignant disease. Patients were censored at the number of days from birth to resection. In case of malignant recurrence or death due to malignancy, the number of days from birth to recurrence detection or death was used.

3. Results

3.1. Patient characteristics

In total, 145 centres from 62 countries participated and contributed data on 3612 patients treated for a presacral teratoma. In 224 patients, it was unknown whether the teratoma was associated with Currarino syndrome. These patients were excluded from analysis. The remaining 3388 patients (74.3 % female) were included; 3183 had a SCT and in 205 teratoma was associated with Currarino Syndrome. Patient characteristics are described in Table 1.

Patients with CS were more likely to be female and were diagnosed later compared with SCT (Table 1). Altman type IV SCT was more frequently found in the CS group than in the SCT group and patients with CS underwent initial resection at an older age compared with SCT.

SCT was treated with surgery in 2770 (87.0 %) children, with chemotherapy in addition to surgical resection in 374 (11.7 %) children. Fifteen (0.5 %) patients were treated with only chemotherapy. Eleven (0.3 %) SCT patients received no treatment and died immediately after birth due to bleeding or respiratory distress. In 13 (0.4 %) children, treatment was unknown. CS patients were also mostly treated with surgery (n = 186, 90·7 %). In eight (3·9 %) CS patients, chemotherapy was given in addition to surgical resection. Three (1·5 %) CS patients received no treatment and in 8 (3·9 %) treatment was unknown.

In total, 119 patients (3·5 %) died during follow-up. There was no difference in outcome between SCT and CS patients with 114 (3·6 %) and 5 (2·4 %) deceased patients in SCT and CS group, respectively. Median age at death was 114 days [2–1145] in SCT group and 210 days [15–506] in CS group. Overall survival after ten years was not different between SCT and CS patients with survival rates of 96.9 % and 94.9 % for CS and SCT patients, respectively (p = 0·343).

3.2. Malignant transformation

Histological diagnosis of recurrent masses after initial resection in SCT patients was mature teratoma in 2066 (64·9 %), immature teratoma in 591 (18·6 %) and malignant teratoma in 343 (10·8 %). In 183 (5·7 %) SCT patients histological diagnosis at initial

Table 1
Patient characteristics.

	Total (n = 3388)	Sacrococcygeal teratoma (n = 3183)	Currarino presacral teratoma (n = 205)	p-value
Sex				0·020
Male	857 (25·3 %)	791 (24·9 %)	66 (32·2 %)	
Female	2519 (74·3 %)	2381 (74·8 %)	138 (67·3 %)	
Missing	12 (0·4 %)	11 (0·3 %)	1 (0·5 %)	
Income country				0·023
LIC	58 (1·7 %)	57 (1·8 %)	1 (0·5 %)	
LMC	307 (9·1 %)	287 (9·0 %)	20 (9·8 %)	
UMC	684 (20·2 %)	657 (20·6 %)	27 (13·2 %)	
HIC	2339 (69·1 %)	2182 (68·6 %)	157 (76·6 %)	
Median age at diagnosis, days (IQR)	1 (0–96)	0 (0–60)	175 (2–611)	<0·001
Altman classification [22]				<0·001
Type I	1000 (29·5 %)	984 (30·9 %)	16 (7·8 %)	
Type II	1065 (31·4 %)	1035 (32·5 %)	30 (14·6 %)	
Type III	600 (17·7 %)	578 (18·2 %)	22 (10·7 %)	
Type IV	668 (19·7 %)	534 (16·8 %)	134 (65·4 %)	
Missing	55 (1·6 %)	52 (1·6 %)	3 (1·5 %)	
Median age at resection	14 (4–186)	13 (4–133)	273 (56–905)	<0·001
Pathology initial tumour				<0·001
Mature	2232 (65·9 %)	2066 (64·9 %)	166 (81·0 %)	
Immature	608 (17·9 %)	591 (18·6 %)	17 (8·3 %)	
Malignant	350 (10·3 %)	343 (10·8 %)	7 (3·4 %)	
Missing	198 (5·8 %)	183 (5·7 %)	15 (7·3 %)	
Recurrence				0·542
Yes	339 (10·0 %)	322 (10·1 %)	17 (8·3 %)	
No	2845 (83·9 %)	2673 (84·0 %)	170 (82·9 %)	
Missing	206 (6·1 %)	188 (5·9 %)	18 (8·8 %)	
Recurrence pathology				0·005
Mature	121 (35·7 %)	116 (36·0 %)	5 (29·4 %)	
Immature	38 (11·2 %)	34 (10·6 %)	4 (23·5 %)	
Malignant	115 (33·9 %)	114 (35·4 %)	1 (5·9 %)	
Missing	65 (19·2 %)	58 (18·0 %)	7 (41·1 %)	
Median time between initial resection and recurrence	347 (199–646)	347 (196–657)	349 (209–433)	0·949
Outcome				0·441
Survival	2921 (86·2 %)	2736 (85·9 %)	185 (90·2 %)	
Death	119 (3·5 %)	114 (3·6 %)	5 (2·4 %)	
Missing	348 (10·3 %)	333 (10·5 %)	15 (7·3 %)	

Data are n(%) or median (IQR).

Please cite this article as: van Heurn LJ et al., Malignant transformation of sacrococcygeal teratoma versus presacral teratoma in Currarino syndrome: Results of 'The SCT-study', Journal of Pediatric Surgery, https://doi.org/10.1016/j.jpedsurg.2025.162848

resection was unknown. In CS patients, the teratoma at initial resection was relatively more often mature compared to SCT patients (n = 166, 81·0 %). Immature (n = 17, 8·3 %) and malignant teratoma (n = 7, 3·4 %) were less frequent in CS patients compared to SCT patients (p < 0·001). In 15 (7·3 %) CS patients, the histological diagnosis after initial resection was unknown.

The probability of malignant transformation at initial resection started to increase directly after birth in SCT patients and increased further with age. This risk was 3·3 %, 5·1 %, 10·3 % and 31·9 % at three months, six months, one year and two years, respectively (Fig. 1) [23]. The risk of malignant transformation at initial resection was lower in CS patients: 1·4 %, 3·0 %, 4·2 % and 4·2 % at three months, six months, one year and two years (p < 0.001), respectively.

In univariable Cox regression, SCT had a significantly higher risk of malignancy at initial resection compared to those with CS (HR 10.59, 95 % CI 4.38–25.63; $p < 0.001$).

This association remained significant after adjusting for age at diagnosis, gender, and Altman classification in a multivariable model (HR 10.25, 95 % CI 4.18–25.12; $p < 0.001$), indicating that isolated SCT is an independent risk factor for malignancy at initial resection.

The malignancy-free survival was higher in CS patients than in SCT patients with 96.9 % and 96.2 % at age one year and two years versus 94.7 % and 88.6 % in SCT patients (HR 5·74, 95 % CI 2·38–13·87; $p < 0·001$) (Fig. 2). In CS patients, malignancy free survival stabilized around the age of one year and around the age of four years in SCT patients.

The malignancy free survival remained higher in the CS group when compared with only internal SCTs (HR 19·69, 95 % CI 4·87–79·62; $p < 0·001$) (Fig. 3).

3.3. Recurrence

A total of 339 (10·0 %) patients developed a recurrence at a median age of 11·4 months (IQR 6·5 months - 1·8 years) after initial resection. The percentages of recurrence were equivalent in both groups (SCT 322 patients (10·1 %), CS patients 17 (8·3 %), [p = 0·542]).

Also, recurrence-free survival was equivalent in both groups (HR 0·91, 95 % CI 0·55–1·50; $p = 0·700$) and stabilized in both groups four years after initial resection with recurrence-free survival of 87·7 % in CS patients and 86·7 % in SCT patients (Fig. 4).

In the SCT group, the treatment of the recurrence was resection in 130, a combination of surgery with chemotherapy in 145, and in 29 patients with chemotherapy only. Treatment was unknown in 14 patients and 4 patients received no treatment due to an inoperable recurrent tumour with metastases. In CS group, 12 patients were treated with surgical resection with 6 receiving additional chemotherapy. One patient was treated with chemotherapy only. One patient received no treatment with an unknown reason. In 3 patients, treatment of the recurrence was unknown.

The histology of the recurrent tumour in SCT patients was mature teratoma in 116, immature teratoma in 34, malignant teratoma in 114 and was unknown in 58 patients. In CS patients, the histology of the recurrent tumour was mature teratoma in 5, immature teratoma in 4 and malignant in one patient, respectively. In 7 patients, the histology of the recurrent tumour was unknown.

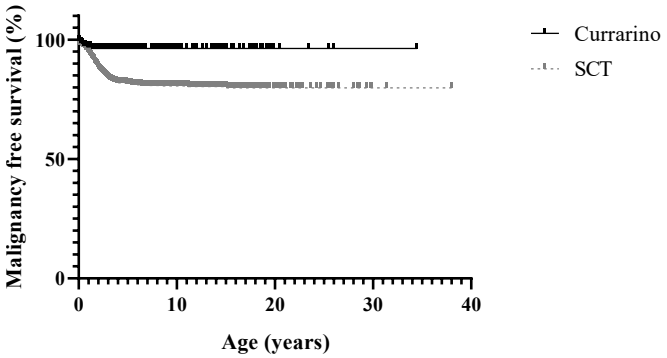

Fig. 2. Malignancy free survival of patients with sacrococcygeal teratoma (SCT) and patients with a presacral teratoma associated with Currarino Syndrome (CS).

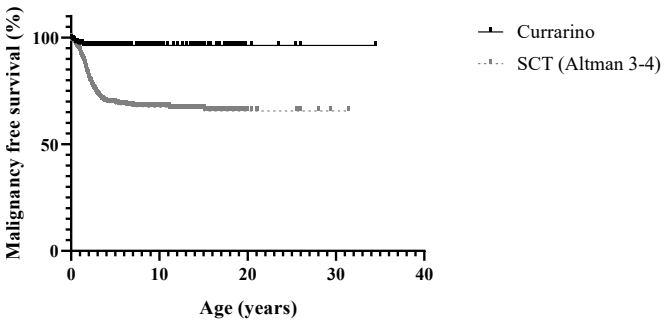

Fig. 3. Malignancy free survival of patients with a complete internal (Altman stage III and IV) sacrococcygeal teratoma (SCT) and patients with a presacral teratoma associated with Currarino Syndrome (CS).

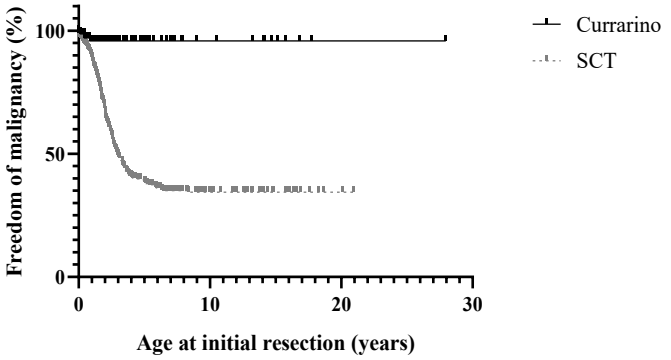

Fig. 1. Malignancy at initial resection of patients with sacrococcygeal teratoma (SCT) and patients with a presacral teratoma associated with Currarino Syndrome (CS).

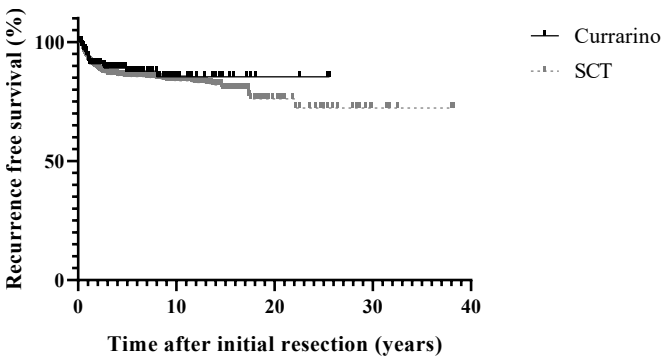

Fig. 4. Recurrence free survival of patients with sacrococcygeal teratoma (SCT) and patients with a presacral teratoma associated with Currarino Syndrome (CS).

Please cite this article as: van Heurn LJ et al., Malignant transformation of sacrococcygeal teratoma versus presacral teratoma in Currarino syndrome: Results of 'The SCT-study', Journal of Pediatric Surgery, https://doi.org/10.1016/j.jpedsurg.2025.162848

4. Discussion

This large global study in 62 countries worldwide shows a difference in tumour behaviour between patients with SCT and patients with presacral teratoma associated with CS. CS is a rare disorder that is associated with a teratoma in 25–40 % of the patients [6,7]. The diagnosis was only recognized after description in 1981 [3]. Therefore, patients with a presacral teratoma before 1982 were excluded from this study.

In this study, age at diagnosis and initial resection was older in CS patients than in SCT patients. This was also found in another comparative study of SCT and CS patients with a median age at initial resection of six months in CS patients compared to 8 days in SCT patients [10]. Presacral teratomas associated with CS are internal and can therefore be difficult to diagnose [24–26]. Furthermore, an underlying genetic component can be found in CS patients both in familiar but also in sporadic cases [27]. Due to differences in genetic penetrance, there is a wide variety in symptoms, even in familial cases, with a relatively high proportion of clinically asymptomatic patients [28].

Malignancy at initial resection in CS is rare; in a previous study of 16 CS patients, there was no malignant transformation in CS patients aged between 6 days and 60 years. The risk of malignant transformation had been estimated at 1 % [10]. This was lower than the 3·4 % found in the current study but still much lower than the risk of malignancy at initial resection in the SCT group of 10·8 % despite the older age at initial resection in the CS group. If the risk of malignant transformation at resection is compared in similar age groups, the difference is even more evident with malignancy rate of 4·2 % after one and two years of age in the CS group versus malignancy rates of 10·5 % and 39·9 % at age one and two years. Moreover, malignancy free survival was higher in CS patients with malignancy free survival rate of 96.2 % after two years of age and 88.6 % in SCT patients. Malignancy free survival stabilized earlier in CS patients at one year compared to four years in the SCT group.

Recurrence after initial resection was equivalent in CS and SCT patients. However, recurrences were more often malignant in SCT patients and malignant recurrences were rarely found in CS patients. A literature review and case report described three malignant recurrences in CS patients [9,18].

4.1. Limitations

Limitations are primarily attributed to its retrospective study design and long inclusion period. However, the low incidence would make a prospective study prohibitively long. Second, the number of collected variables was limited. Third, the use of anonymized data made data validation not possible. Finally, there is bias because an unknown proportion of patients has not been included in the study. Due to poor CS recognition in LICs and LMICs because of lack of imaging modalities, CS patients could be incorrectly classified as SCT in this study.

4.2. Recommendations

We recommend an individualized approach in the treatment of SCT and CS patients. In SCT patients, early and complete resection is recommended because of the higher risk of malignant transformation. On the other hand, the risk of malignant transformation in CS patients is very low compared to SCT patients so that time can be taken to evaluate the other associated anomalies.

Furthermore, an additional anterior meningocele is often found in Currarino patients leading to possible neurological damage with resection. Because of the low risk of malignant transformation in CS patients, it may be justified to accept the risk of residual tumour or recurrence as complete resection may be associated with severe neurological damage such as faecal incontinence, voiding problems and sexual dysfunction [29,30]. Therefore, delayed or conservative treatment could be considered in this group, especially in asymptomatic patients, taking the small risk of malignant transformation later in life into account. In these patients, yearly follow-up with serum alpha fetoprotein concentration, ultrasound, and MRI is recommended to monitor possible tumour growth and malignant transformation. For both patient groups, oncological follow-up for at least four years after initial resection is recommended to detect possible recurrent disease.

5. Conclusion

Despite similar clinical appearance, CS and SCT show different risks of malignant transformation and tumour recurrence. Therefore, treatment in CS patients can be more conservative compared to SCT patients in which resection early in life and close follow-up is essential.

Data sharing

Following publication of the study results, the full, anonymous de-identified patient dataset will be made available. Proposals should be directed to sct-study@amsterdamumc.nl to gain access. Data requestors will need to sign a data access agreement.

Funding source

This study received funding from Kika Children Cancer-free Foundation. Kika Children Cancer-free Foundation had no involvement in collection, analysis and interpretation of data or in writing and decision to submit the manuscript for publication.

Conflicts of interest

All authors declare no conflicts of interest.

Acknowledgements

We thank the EUPSA Network Office for their help in raising awareness for the study and contacting centres for possible participation. We would like to thank Mikhail Povarov for helping with the Russian translation of the study documentation.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jpedsurg.2025.162848.

References

[1] Rescorla FJ, Sawin RS, Coran AG, Dillon PW, Azizkhan RG. Long-term outcome for infants and children with sacrococcygeal teratoma: a report from the childrens cancer group. J Pediatr Surg 1998;33(2):171–6.
[2] De BA, Madern GC, Hakvoort-Cammel FG, Haentjens P, Oosterhuis JW, Hazebroek FW. Study of the factors associated with recurrence in children with sacrococcygeal teratoma. J Pediatr Surg 2006;41(1):173–81.
[3] Currarino G, Coln D, Votteler T. Triad of anorectal, sacral, and presacral anomalies. AJR Am J Roentgenol 1981;137(2):395–8.
[4] Emans PJ, Kootstra G, Marcelis CL, Beuls EA, van Heurn LW. The currarino triad: the variable expression. J Pediatr Surg 2005;40(8):1238–42.
[5] Martucciello G, Torre M, Belloni E, Lerone M, Pini Prato A, Cama A, et al. Currarino syndrome: proposal of a diagnostic and therapeutic protocol. J Pediatr Surg 2004;39(9):1305–11.
[6] Samuel M, Hosie G, Holmes K. Currarino triad–diagnostic dilemma and a combined surgical approach. J Pediatr Surg 2000;35(12):1790–4.
[7] Köchling J, Pistor G, Märzhäuser Brands S, Nasir R, Lanksch WR. The currarino syndrome–hereditary transmitted syndrome of anorectal, sacral and

presacral anomalies. Case report and review of the literature. Eur J Pediatr Surg 1996;6(2):114–9.
[8] Bartels SA, van Koperen PJ, van der Steeg AF, Deurloo EE, Bemelman WA, Heij HA. Presacral masses in children: presentation, aetiology and risk of malignancy. Colorectal Dis 2011;13(8):930–4.
[9] Yoshida A, Maoate K, Blakelock R, Robertson S, Beasley S. Long-term functional outcomes in children with currarino syndrome. Pediatr Surg Int 2010;26(7):677–81.
[10] Dirix M, van Becelaere T, Berkenbosch L, van Baren R, Wijnen RM, Wijnen MH, et al. Malignant transformation in sacrococcygeal teratoma and in presacral teratoma associated with currarino syndrome: a comparative study. J Pediatr Surg 2015;50(3):462–4.
[11] 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(11):1189–94.
[12] Wang Y, Wu Y, Wang L, Yuan X, Jiang M, Li Y. Analysis of recurrent sacrococcygeal teratoma in children: clinical features, relapse risks, and anorectal functional sequelae. Med Sci Monit 2017;23:17–23.
[13] Pauniaho SL, Tatti O, Lahdenne P, Lindahl H, Pakarinen M, Rintala R, et al. Tumor markers AFP, CA 125, and CA 19-9 in the long-term follow-up of sacrococcygeal teratomas in infancy and childhood. Tumour Biol: J Int Soc Oncodevelopmental Biol and Med 2010;31(4):261–5.
[14] Yao W, Li K, Zheng S, Dong K, Xiao X. Analysis of recurrence risks for sacrococcygeal teratoma in children. J Pediatr Surg 2014;49(12):1839–42.
[15] De Backer A, Madern GC, Hakvoort-Cammel FG, Haentjens P, Oosterhuis JW, Hazebroek FW. Study of the factors associated with recurrence in children with sacrococcygeal teratoma. J Pediatr Surg 2006;41(1):173–81. discussion -81.
[16] Schmidt B, Haberlik A, Uray E, Ratschek M, Lackner H, Hollwarth ME. Sacrococcygeal teratoma: clinical course and prognosis with a special view to long-term functional results. Pediatr Surg Int 1999;15(8):573–6.
[17] De CF, Sarnacki S, Patte C, Mosseri V, Baranzelli MC, Martelli H, et al. Prognosis of malignant sacrococcygeal germ cell tumours according to their natural history and surgical management. Surg Oncol 2012;21(2):e31–7.
[18] Hage P, Kseib C, Adem C, Chouairy CJ, Matta R. Atypical presentation of currarino syndrome: a case report. Int J Surg Case Rep 2019;57:102–5.
[19] von Elm E, Altman DG, Egger M, Pocock SJ, Gøtzsche PC, Vandenbroucke JP. The strengthening the reporting of observational studies in epidemiology (STROBE) statement: guidelines for reporting observational studies. J Clin Epidemiol 2008;61(4):344–9.
[20] Hamadeh Nrv C, Metreau E. New world bank country classifications by income level: 2022-2023 2022 [Available from: https://blogs.worldbank.org/opendata/new-world-bank-country-classifications-income-level-2022-2023.
[21] Castor EDC. Castor electronic data capture 2019 [27 Aug. 2019]. Available from: https://castoredc.com.
[22] Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: american academy of pediatrics surgical section Survey-1973. J Pediatr Surg 1974;9(3):389–98.
[23] van Heurn LJ, Derikx JPM, Hall N, Aldrink JH, Bailez MM, Chirdan LB, et al. Malignant transformation and tumour recurrence in sacrococcygeal teratoma: a global, retrospective cohort study. Int J Surg 2024;110(11):7177–86.
[24] Chirdan LB, Uba AF, Pam SD, Edino ST, Mandong BM, Chirdan OO. Sacrococcygeal teratoma: clinical characteristics and long-term outcome in Nigerian children. Ann Afr Med 2009;8(2):105–9.
[25] Sitkin NA, Ozgediz D, Donkor P, Farmer DL. Congenital anomalies in low- and middle-income countries: the unborn child of global surgery. World J Surg 2015;39(1):36–40.
[26] Chirdan LB, Ameh EA, Abantanga FA, Sidler D, Elhalaby EA. Challenges of training and delivery of pediatric surgical services in Africa. J Pediatr Surg 2010;45(3):610–8.
[27] Dworschak GC, Reutter HM, Ludwig M. Currarino syndrome: a comprehensive genetic review of a rare congenital disorder. Orphanet J Rare Dis 2021;16(1):167.
[28] Lynch SA, Wang Y, Strachan T, Burn J, Lindsay S. Autosomal dominant sacral agenesis: currarino syndrome. J Med Genet 2000;37(8):561–6.
[29] 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(4):690–4.
[30] Kremer ME, Derikx JP, Peeters A, Ter Kuile MM, van Baren R, Heij HA, et al. Sexual function after treatment for sacrococcygeal teratoma during childhood. J Pediatr Surg 2016;51(4):534–40.

Please cite this article as: van Heurn LJ et al., Malignant transformation of sacrococcygeal teratoma versus presacral teratoma in Currarino syndrome: Results of 'The SCT-study', Journal of Pediatric Surgery, https://doi.org/10.1016/j.jpedsurg.2025.162848