Long-term Outcomes of Infantile Sacrococcygeal Teratoma: Results from Japan Nationwide Survey

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BACKGROUND & PURPOSE

- Sacrococcygeal teratoma (SCT)
  - during follow-up after tumor resection, some present sequela: **anorectal & urinary dysfunctions, lower limb palsy, tumor recurrence, malignant transformation**
- To clarify Japanese trends in SCT sequelae with aim of helping to improve prognosis & QOL

STUDY DESIGN

- Granted from the Ministry of Health, Labour and Welfare of Japan (20FC1042)
- Domestic multicentric cohort on a paper-based questionnaire survey for 192 facilities accredited by Japanese Society of Pediatric Surgeons (JSPS)
- Inclusion criteria**: 2000-2019, pts undergone radical surgery at < 1 yo and survived for at least 180 days after birth**
- Survey items
  - patient background, tumor size, Altman's classification (I-IV), pathology, primary surgery, follow-up

CONCLUSION

- **Postoperative dysfunction of SCT was not low at 23.4%**
- **11.8% had recurrence , some occurring > 10 yrs after surgery, need for periodic imaging and tumor markers**
- **It is necessary to establish treatment guidelines for best practice monitoring of the long-term QOL**

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, 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 dysfuction	**n (%)**	**15 (4.2)**
	CNS damage	**n (%)**	**16 (4.5)**
	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

Postoperative functional impairments with infantile sacrococcygeal teratoma

83/355 cases (23.4%)

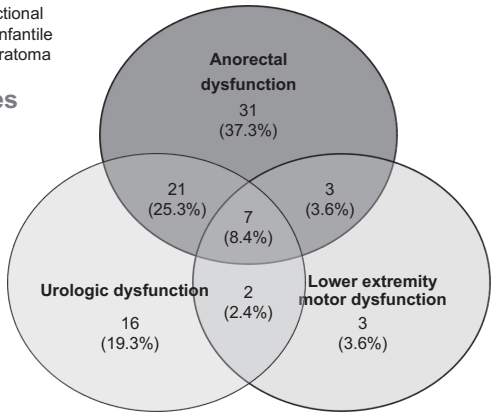

Table 2 Risk factors for functional sequelae

Characteristics		Univariable		Multivariable	
		OR (95% CI)	*p*	OR (95% CI)	*p*
Premature birth	<37w	0.438 (0.240–0.797)	0.007*		
low Birth weight	<2500g	2.036 (1.002–4.137)	0.049*		
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. ≥8d	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 excision	subtotal or partial vs. total	2.504 (1.112–5.640)	0.027*		
Surgical complication	Yes	3.073 (1.629–5.798)	0.001*	3.506 (1.580–7.782)	0.002*

Table 3 Risk factors for tumor recurrence

Characteristics		Univariable		Multivariable	
		OR (95% CI)	*p*	OR (95% CI)	*p*
Age at diagnosis	<8 d vs. ≥8d	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 excision	subtotal or partial vs. total	4.742 (1.964–11.451)	0.001*	4.979 (1.753–14.148)	0.003*
Surgical complication	Yes	3.078 (1.448–6.543)	0.004*		

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