のカテーテルを選択的に正中仙骨動脈に誘導し、手技を行い、手技終了後は左総頸動脈を用手的に圧迫止血した、との答えであった。

総頸動脈アプローチによるカテーテルインターベンションに関しては、小児循環器領域では、通常の方法で血管アクセスが困難な場合に選択される経路で54例中25例に総頸動脈が選択され、その約70%がより重症かつ緊急例で、合併症は全体で2例のみで、いずれも長期化するものではなかったと報告されている(4)。

Cowlesら(2)の症例では、合併症はなかったものの、シース挿入側の大腿動脈はシースより遠位が造影されていない。このことは、新生児期には大腿動脈が細径であることに加え、これより心臓側に位置する多血性の仙尾部奇形腫に多くの血流が奪われるため、大腿動脈がさらに細径になることから容易に理解できる。全国調査では、右大腿動脈アプローチの1例で後に下肢血流障害を生じている。

仙尾部奇形腫の栄養血管については、全国調査結果から、正中仙骨動脈のみならず内腸骨動脈も少なからず関与することが判明した。また、臍動脈アプローチならびに大腿動脈アプローチのいずれにおいても目的とする血管へのカテーテル誘導は容易でないことも判明した。正中仙骨動脈や内腸骨動脈へのカテーテル誘導は、臍動脈あるいは大腿動脈からのアプローチでは鋭角にカテーテルを進めなければならないが、左総頸動脈からのアプローチでは比較的直線状にカテーテルを下行させ、大動脈分岐部付近で左右あるいは前後にカテーテル先端を操作すれば、いずれの栄養血管にも比較的容易に誘導可能であると思われる。

術前動脈塞栓術は全ての症例に適応となるわけではなく、Cowlesら(2)が述べているように、腫瘍切除の際に腫瘍血管の先行遮断が行われる症例が潜在的な術前動脈塞栓術の適応といえる。全国調査結果からは手術例の少なくとも30%が潜在的な術前動脈塞栓術の適応であることが示唆された。しかし、実際の動脈塞栓術実施に関しては、それぞれの施設の状況が大きく影響するため、各施設において周産期診療にかかわるすべての医療者で適応に関して慎重に検討することが最も重要と思われる。

E. 結論

出生前に診断された仙尾部奇形腫に対するinterventional radiologyの位置づけについて検討した。全国調査結果から、手術例中、術前塞栓術が試みられたのは2.5%であった。腫瘍血管の先行遮断は手術例の約30%に行われ、これらが潜在的な術前塞栓術の適応症例と思われた。出生後行われた画像診断検査上、腫瘍への栄養血管は、正中仙骨動脈のみならず、内腸骨動脈も少なからず関与していることが判明した。腫瘍への栄養血管の種類ならびに解剖学的な分岐様

式からは、術前塞栓術を行う場合は、左総頸動脈アプローチが最も確実であることが示唆された。

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F. 健康危険情報

該当する健康危険情報はない

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表　出生後実施された画像診断検査による腫瘍への栄養血管の同定頻度と血管の種類

		腫瘍への栄養血管			
検査法	栄養血管	正中仙骨動脈	正中仙骨動脈+内腸骨動脈	内腸骨動脈	その他
US*	17/53(32%)	11	3	2	1
CT	17/46(37%)	8	6	2	1
MRI	4/29(14%)	3	0	1	0

*:US: 超音波検査

Ⅲ．研究成果の刊行に関する一覧表

研究成果の刊行に関する一覧表

雑誌

発表者氏名	論文タイトル名	発表誌名	巻号	ページ	出版年
Usui N, Kitano Y, Sago H, Kanamori Y, Yoneda A, Nakamura T, Nosaka S, Saito M, Taguchi T.	Outcomes of prenatally diagnosed sacrococcygeal teratomas: the results of a Japanese nationwide survey.	J Pediatr Surg	47(3)	In press DOI: 0.1016/j.jpedsurg.2011.08.020	2012
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Original articles

Outcomes of prenatally diagnosed sacrococcygeal teratomas: the results of a Japanese nationwide survey

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Abstract

Background/Purpose: Few large multicenter surveys have been performed on sacrococcygeal teratomas (SCTs) describing both the prenatal and postnatal courses. The aim of this study was to review and report on the prenatal surveillance and postnatal outcome of a large cohort of fetuses with SCTs in Japan.

Methods: A nationwide retrospective cohort study was conducted on 97 fetuses prenatally diagnosed with SCTs between 2000 and 2009. The prenatal course, perinatal data, and postnatal outcome were reviewed.

Results: Eleven pregnancies were terminated before 22 weeks of gestation. Of the 86 remaining fetuses, 3 died in utero, and 83 were delivered. Three infants died before surgery, and 8 infants died after excisional surgery. The overall mortality was 26%, with a mortality excluding terminations of 16%. The gestational age at delivery was younger than 28 weeks in 5, 28 to 31 weeks in 13, 32 to 36 weeks in 27, and 37 weeks or more in 37 cases, with mortality rates of 60%, 38%, 11%, and 0%, respectively. The tumor component was predominantly cystic in 54 and predominantly solid in 32 cases, with mortality rates of 2% and 33%, respectively.

Conclusions: The overall mortality of prenatally diagnosed SCTs excluding terminations was 16%. Early delivery and predominantly solid component tumors were associated with an increased risk of mortality.

Sacrococcygeal teratoma (SCT), which originates from the 3 germinal layers, is the most common congenital tumor, with a birth prevalence of 1 in 27,000 live births [1]. Sacrococcygeal teratomas diagnosed postnatally have been associated with an excellent prognosis after surgical excision [2-5]. In contrast, it has been reported that fetuses with a prenatally diagnosed SCTs still have a high risk of death even if the prenatal diagnosis may have made a contribution to improvement of the outcome [6]. The main reason for the

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poor prognosis in fetal SCTs is high-output cardiac failure caused by increased blood flow according to the amount of solid component present in the tumor [7,8] and rupture of the tumor during delivery with a massive hemorrhage [9,10]. However, the reported mortality rate excluding terminations in the fetuses with SCTs varied widely from 18% to 63% in different series [6-16]. Most of these studies were conducted in a single institution with a small number of patients, and there may have been selection bias [8] because some of the institutions were specialized centers for fetal treatment. The natural history of fetal SCTs has not been fully characterized because few large multicenter surveys have been performed describing both the prenatal and postnatal courses. The aim of this study was to review and report on the prenatal surveillance and postnatal outcome of a large cohort of fetuses with SCTs in Japan based on a nationwide survey.

1. Materials and methods

A nationwide retrospective cohort study was conducted on fetuses prenatally diagnosed with SCTs at major Japanese perinatal centers. We initially sent a preliminary questionnaire requesting the number of fetuses prenatally diagnosed with SCTs between January 2000 and December 2009 to 325 major perinatal centers in Japan and asked them to participate in our detailed survey. One-hundred ninety centers (58.5%) responded to the preliminary survey and reported that there were 138 cases with SCTs diagnosed prenatally during the past 10 years. We then sent a second form requesting further details about the fetuses from the centers that had corresponding cases and had accepted our offer to participate in a detailed survey. Forty-eight centers that had 101 cases of fetal SCTs consented to participate in our survey and returned the forms with further details. Four fetuses that had not been followed up until fetal demise or live birth owing to maternal transfer were excluded from the study, so 97 fetuses prenatally diagnosed with SCTs between January 2000 and December 2009 at 46 Japanese perinatal centers were included in the study and analyzed. The patient demographics, including the year of prenatal diagnosis, gestational age at diagnosis, occurrence of polyhydramnios, signs of hydrops fetalis, fetal interventions, prenatal outcome, mode of delivery, gestational age at delivery, sex of the fetus, and birth weight were reviewed. The type of the tumor component, tumor location, histology of the tumor, maximum diameter of the tumor, and postnatal outcome were also reviewed.

Polyhydramnios was regarded as positive if there was a finding of polyhydramnios either in the initial or final fetal ultrasonography. The presence of signs for hydrops fetalis was defined as positive if there was a finding of ascites, pleural effusion, or skin edema either in the initial or final fetal ultrasonography. The type of the tumor component was defined as 1 of 4 categories, such as cystic type (>90% of the tumor is cystic), predominantly cystic mixed type (50%-90% of the tumor is cystic), predominantly solid mixed type (50%-90% of the tumor is solid), and solid type (>90% of the tumor is solid), and the cases were classified according to the schema described in the questionnaire (Fig. 1). The type of the tumor component was determined by pathologic findings in surgical cases and by prenatal or postnatal diagnostic imaging in nonsurgical cases. The tumor location was categorized according to Altman's classification [2] determined by operative findings or diagnostic imaging. The maximum diameter of the tumor was defined as the maximum value of the maximum diameter of the resected tumor, the maximum diameter of the computed tomography performed after birth, and the maximum diameter of the magnetic resonance imaging performed after birth in the cases of live births and as the maximum value on fetal ultrasonography in cases of fetal demise.

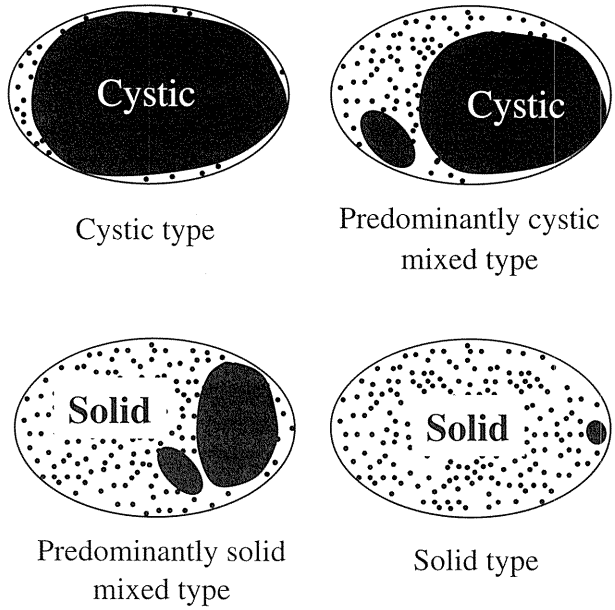

Fig. 1 A schematic diagram of the types of tumor components. Cystic type, more than 90% of the tumor is cystic; predominantly cystic mixed type, 50% to 90% of the tumor is cystic; predominantly solid mixed type, 50% to 90% of the tumor is solid; solid type, more than 90% of the tumor is solid.

The data were expressed as the medians (range). The frequencies and percentages were used to describe categorical data. The χ^2 test was used for the analysis of categorical data. $P < .05$ was considered to indicate statistical significance. Statistical analyses were performed with the JMP software program (version 8.02; SAS Institute, Inc, Cary, NC). This retrospective survey was approved by the institutional review boards of the 5 participating institutions (institutional review board approval no. 09392, National Center for Child Health and Development).

2. Results

The annual number of the fetuses with SCTs was less than 7 before 2005 but increased thereafter and exceeded 14 cases

per year after 2007 (Fig. 2). The median maternal age was 30 years (range, 18-41 years), and median gestational age at diagnosis was 25 weeks of gestation (range, 15-36 weeks). Twenty-four cases (24.7%) were diagnosed before 22 weeks of gestation, the period in which a termination of pregnancy is legally permitted in Japan [17]. Eleven pregnancies were terminated before 22 weeks of gestation, and 86 cases intended to deliver (Fig. 3). Of the 24 cases diagnosed before 22 weeks of gestation, there were no significant differences in the size of the tumor, type of the tumor component, or incidence of the signs for hydrops fetalis between the cases that were terminated and the cases that were intended to be delivered (data are not shown).

The outcomes of the infants with prenatally diagnosed SCTs are shown in Fig. 4. Of the 86 nonterminated fetuses, 3 resulted in intrauterine fetal deaths, and 83 survived to be born. Of these survivors, 4 cases underwent fetal intervention, including radiofrequency ablation (n = 1), abdominal paracentesis (n = 1), and cyst aspiration (n = 2). All 4 of these cases underwent tumor resection, and 3 survived after the surgery. The patient who underwent paracentesis died of a massive hemorrhage during the surgery. Amnioreduction had been performed in 11 pregnancies to prevent preterm labor and maternal discomfort owing to polyhydramnios. After the live birth, 3 infants died before surgery on the day of birth, and 80 infants underwent excisional surgery at a median of 74 hours (range, 1-1581 hours). Twenty-five infants underwent surgery in the first 24 hours of life, and 6 of them (24%) died, whereas 55 cases underwent surgery after 24 hours, and only 2 (4%) of them died. Four infants died after surgery during the early neonatal period, 2 died during the later neonatal period, and 2 died later in infancy. Of the 9 neonatal deaths, 7 were related to massive hemorrhage from the tumors. Bleeding from the tumors was already recognized in 6 neonates at the time of cesarean section delivery, and 4 of them, including 2 neonates with a tumor that ruptured during the delivery, died on the day of birth. The median follow-up of survivors was 23 months (range, 0-113 months). The overall mortality was 26% (25/97), with a mortality excluding terminations of 16% (14/86).

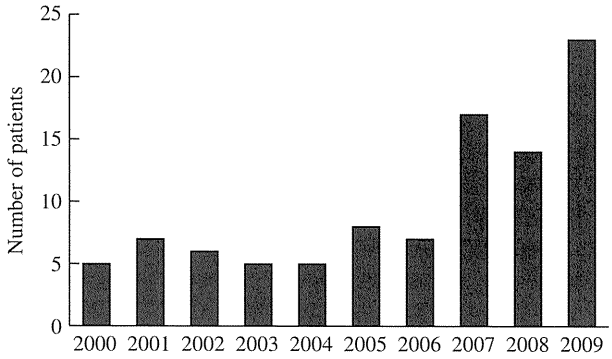

Fig. 2 The number of the fetuses with a prenatal diagnosis of SCT.

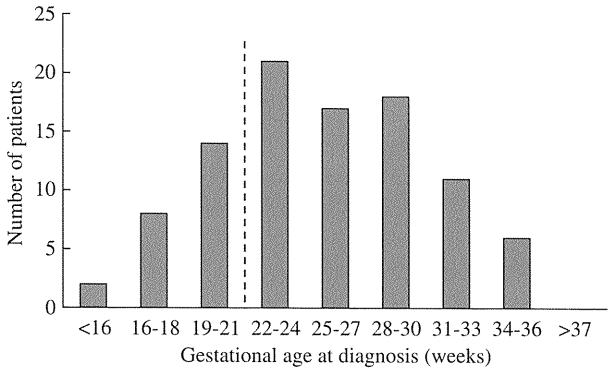

Fig. 3 The distribution of the gestational age at diagnosis. A termination of pregnancy is legally permitted in Japan before 22 weeks of gestation (broken line).

Table 1 reviews the demographics and mortality of the fetuses with SCTs that were intended to be delivered. The fetuses that had been diagnosed before 28 weeks of gestation had a significantly higher mortality rate compared with those diagnosed after 28 weeks of gestation. They were delivered at a median gestational age of 36.4 weeks (range, 26-41 weeks). Forty-five fetuses (55%) were born prematurely, and

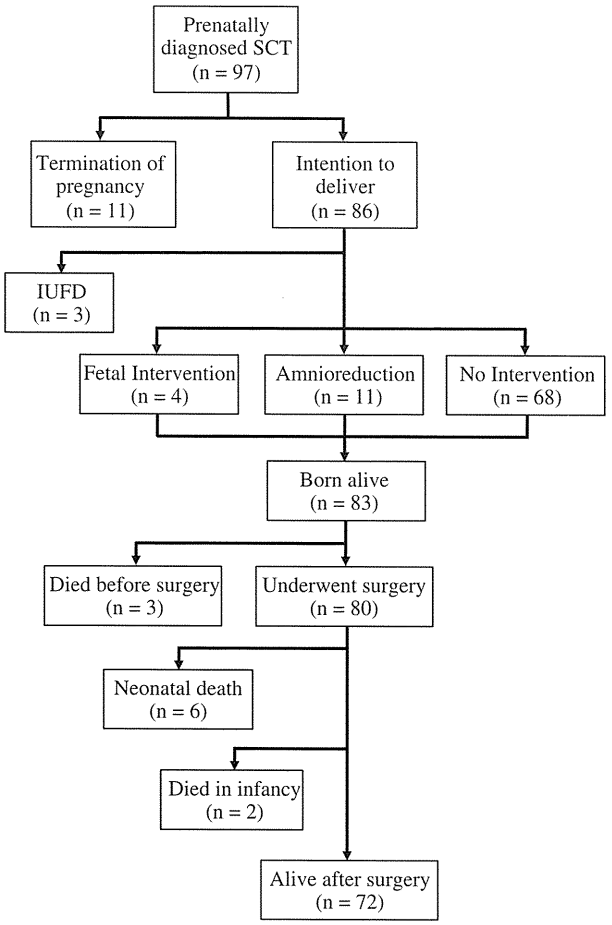

Fig. 4 The outcomes of prenatally diagnosed SCT (2000-2009). IUFD indicates intrauterine fetal death.

Table 1 Demographics and mortality of the fetuses that were intended to be delivered after a prenatally diagnosis of SCT

Characteristics	Distribution of patients (%)	Mortality (%) and *P* value
Gestational age at diagnosis (wk)	n = 86	*P* = .020
<22	13 (15.1)	3 (23.1)
22-27	38 (44.2)	10 (26.3)
≥28	35 (44.2)	1 (2.9)
Gestational age at delivery (wk)	n = 82	*P* < .001
<28	5 (6.1)	3 (60.0)
28-31	13 (15.9)	5 (38.5)
32-36	27 (32.9)	3 (11.1)
≥37	37 (45.1)	0 (0.0)
Sex	n = 85	*P* = .303
Male	23 (27.1)	2 (8.7)
Female	62 (72.9)	11 (17.7)
Birth weight (g)	n = 82	*P* = .404
<2000	7 (8.5)	1 (14.3)
2000-2999	42 (51.2)	8 (19.0)
3000-3999	27 (32.9)	2 (7.4)
≥4000	6 (7.3)	0 (0.0)
Mode of delivery	n = 82	*P* = .036
Vaginal delivery	12 (14.6)	1 (8.3)
Planned cesarean section delivery	39 (47.6)	2 (5.1)
Emergency cesarean section delivery	31 (37.8)	8 (25.8)

37 fetuses (45%) were delivered at full term. The mortality rates based on age at delivery were significantly different, and the younger infants demonstrated a higher mortality rate. The male-to-female ratio was 1:2.6. The median birth weight was 2893 g (range, 1020-5014 g). There was no significant difference in the mortality among the infants with different

Table 2 Characteristics of fetal ultrasonography and the tumor findings of the fetuses that were intended to be delivered along with their mortality

Characteristics	Distribution of patients (%)	Mortality (%) and *P* value
Polyhydramnios	n = 86	*P* = .078
Yes	26 (30.2)	7 (26.9)
No	60 (69.8)	7 (11.7)
Sign of hydrops fetalis	n = 86	*P* < .001
Yes	14 (16.3)	8 (57.1)
No	72 (83.7)	6 (8.3)
Altman's classification	n = 84	*P* = .734
I	48 (57.1)	10 (20.8)
II	26 (31.0)	3 (11.5)
III	5 (5.9)	0 (0.0)
IV	5 (5.9)	1 (20.0)
Type of tumor component	n = 86	*P* < .001
Cystic type	23 (26.7)	0 (0.0)
Predominantly cystic mixed type	31 (36.0)	2 (6.4)
Predominantly solid mixed type	25 (29.1)	8 (32.0)
Solid type	7 (8.1)	4 (57.1)
Maximum diameter of the tumor (cm)	n = 86	*P* = .213
<5.0	4 (4.7)	0 (0.0)
5.0-9.9	26 (30.2)	0 (0.0)
10.0-14.9	28 (32.6)	7 (25.0)
15.0-19.9	22 (25.6)	5 (22.7)
≥20.0	6 (7.0)	2 (33.0)

birth weights. Twelve fetuses (15%), including 2 cases who had undergone cyst aspiration for decompression before delivery, were born by vaginal delivery, whereas 70 fetuses (85%) were born via cesarean delivery: 31 because of the tumor size, 10 because of fetal cardiac failure, 9 because of the repetitive cesarean section, 5 because of the fetal distress, and 15 for other reasons. None of the fetuses delivered by cesarean section underwent ex utero intrapartum therapy (EXIT). The fetuses delivered by emergency cesarean section demonstrated a significantly higher mortality rate. Associated anomalies were detected in 10 (12%) of 83 infants and included congenital heart disease (n = 5), undescended testes (n = 2), an anorectal anomaly (n = 1), an ectopic ureter (n = 1), hydrocephalus (n = 1), and intestinal duplication (n = 1).

Table 2 reviews the characteristics of the fetal ultrasonography and the tumor findings in the fetuses that were intended to be delivered along with their mortality. Polyhydramnios was recognized in 30% of the patients, and some signs for hydrops fetalis were seen in 16% of the cases. The mortality of the fetuses with 1 sign of hydrops fetalis was higher than that of the fetuses without any sign of hydrops fetalis. Type I Altman's classification was the most common tumor location, and type II was the second most common. There were no statistically significant differences in the mortality rates among the infants with different Altman's classifications. The type of the tumor component was predominantly cystic (>50% cystic) in 54 cases (63%) and predominantly solid (>50% solid) in 32 cases (37%). There were significant differences in the mortality among the patients with different types of the tumor components, and the predominantly solid type was associated with higher mortality. Although there was no statistically significant difference in the mortality among the patients with different maximum diameters of the tumor, no fetuses that had a tumor with a maximum diameter less than 10 cm by any measurements died.

3. Discussion

We reviewed the prenatal course, perinatal data, and postnatal outcome in this Japanese nationwide retrospective cohort study conducted on 97 fetuses prenatally diagnosed with SCTs between 2000 and 2009. Of the 97 fetuses, 11 pregnancies were terminated, 3 died in utero, and 11 infants died after live births. The overall mortality including the termination of pregnancy was 26% (25/97), and the mortality excluding such terminations was 16% (14/86). The perinatal mortality and the neonatal mortality of this cohort were 12% (10/86) and 11% (9/83), respectively. Table 3 includes data from published series of fetal SCTs involving 10 or more cases, including our present results, which are the largest retrospective cohort study conducted for fetuses with a prenatal diagnosis of SCT [6-16]. Although the mortality among fetuses with SCT varied widely in different series, the outcome of the present study was better than that reported previously [6-11,14-16]. One of the reasons for this discrepancy is the advances that have been made in maternal or fetal management and perinatal care in recent years, as cases from more 10 years ago were included in some series. The other reason may be the effects of selection bias in some institutions, as some of them were highly specialized centers for fetal intervention and may be more likely to receive referrals of more serious cases associated with higher mortality.

Another possibility is that there was an increased population with a prenatal diagnosis of SCTs in our country likely owing to the advent of improved antenatal imaging techniques and screening. The Japanese Society of Pediatric Surgeons reported in their neonatal surgical survey performed every 5 years that the ratio of prenatal diagnosis in neonates with SCTs was 44% in 2003 [18] and 82% in 2008 [19]. The number of registered cases has increased rapidly in the past few years along with the increase in the ratio of prenatal diagnosis during the 5-year period from 2003 to 2008 (Fig. 2). Improvements in prenatal diagnosis may have

Table 3 The number of the patients and the outcomes reported in previous studies

Author	Study period	Study design	No. of cases	No. of TOP	IUFD	Postnatal death	Mortality excluding TOP (%)
Bond et al [7]	1990	M	45	11	11	6	17/34 (50%)
Sheth et al [11]	NA	M	15	2	5	2	7/13 (54%)
Holterman et al [10]	1980-1997	S	24	3	4	5	9/21 (43%)
Brace et al [9]	1992-1998	S	10	2	2	3	5/8 (63%)
Westerburg et al [8]	1986-1998	S	17	2	0	6	6/15 (40%)
Kamata et al [12]	1979-1999	S	14	0	1	2	3/14 (21%)
Hedrick et al [6]	1995-2002	S	30	4	5	7	12/26 (46%)
Benachi et al [13]	1983-2003	M	44	4	2	5	7/40 (18%)
Makin et al [14]	1993-2004	S	35	6	3	4	7/29 (24%)
Sy et al [15]	1991-2005	S	27	3	5	3	8/24 (33%)
Wilson et al [16]	2003-2006	S	23	4	3	4	7/19 (37%)
Present study	2000-2009	M	97	11	3	11	14/86 (16%)

NA indicates not available; S, single-institutional study; M, multicenter study; TOP, termination of pregnancy; IUFD, intrauterine fetal death.

contributed to the increased detection of milder cases that would not have been diagnosed previously. Although there were no significant differences in the prognostic factors between the 11 terminated cases and the 13 continued pregnancies for fetuses that were diagnosed before 22 weeks of gestation, termination of pregnancy may have contributed to the improvement of the mortality because the cases with earlier presentation demonstrated poorer prognosis, probably because of the larger tumor growth, compared with the cases with later presentation [8,16] (Table 1).

This is the first Japanese multicenter survey conducted for fetuses with a prenatal diagnosis of SCT. Our preliminary survey revealed that there were at least 138 fetuses with SCT during the past 10 years in Japan, and 70% of them were surveyed in detail. The population survey report of the Ministry of Health, Labour and Welfare of Japan reported that there were 11,155,608 live births and 329,757 stillbirths during the period between January 2000 and December 2009. Judging from the birth prevalence of SCT, the number of patients with SCT predicted for this period was estimated to be approximately 400 cases including postneonatally presented cases, and the patient number in our survey therefore corresponded to approximately one fourth of the estimated cases for that period. The Japanese Society of Pediatric Surgeons reported that 16 neonates with prenatally diagnosed SCTs were treated in 2003 [18], and 23 neonates with prenatally diagnosed SCTs were treated in 2008 [19]. Together with this demographic information, it is estimated that about half of the prenatally diagnosed cases of SCT in our country have been collected and surveyed from the 46 perinatal centers participated in the present study. In consequence, the results of this study accurately describe the current status of both the prenatal and postnatal courses of these infants and characterize the natural history of fetal SCTs in our country. This study will therefore provide useful information for prenatal counseling of parents.

The gestational age of the fetus at delivery had a major impact on the perinatal and postnatal mortality, as has been previously reported [6,9,10,16]. The mortality rates based on age at delivery were significantly different, and the younger infants demonstrated a higher mortality. This was presumably because of the synergistic effects of several factors, including the high mortality owing to prematurity, the high risk of preterm labor owing to the high-output cardiac failure and polyhydramnios, and the large tumor size that required early delivery. With regard to the mode of delivery, our data showed that emergency cesarean section was likely to be selected in the high-risk patients. The well-known Altman's classification into 4 types, depending on the relationship of the extrapelvic and intrapelvic parts, demonstrated no correlation with the outcome. In contrast, the type of the tumor component was well correlated with the outcome, which is consistent with the previous reports [6,8,10,13]. The solid component of the SCT is generally very vascular and has the potential for rapid growth, resulting in an increased risk of high-output cardiac failure and massive hemorrhage [6,13,15].

Although there were no statistically significant differences in the mortality rates among the cases with different maximum diameters of the tumors, no fetuses whose maximum diameter was less than 10 cm died. A recent study, which proposed a prognostic classification for the fetuses prenatally diagnosed to have SCT [13], defined 3 risk groups as follows: group A with tumor diameters less than 10 cm, absent or mild vascularity, and slow growth; group B with tumor diameter of 10 cm or greater, pronounced vascularity or high-output cardiac failure, and rapid growth; and group C with a tumor diameter of 10 cm or greater, predominantly cystic lesions with absent or mild vascularity, and slow growth. According to their prognostic classification, our cases were classified into 30 cases of group A, 28 cases of group B and 28 cases of group C, with mortality rates of 0%, 39%, and 11% respectively.

There have been some report of fetal interventions, such as maternal-fetal surgery to resect the tumors [6,14], radiofrequency ablation [20], major vessel laser ablation [14], and vessel alcohol sclerosis [14], to prevent the high-output cardiac failure. In some cases, there were indications for prenatal intervention, such as amniodrainage, to prevent preterm labor [6,14,16] and cyst decompression before delivery to prevent tumor rupture [6,9,14,16]. There were 4 cases of fetal intervention and 11 cases of amniodrainage in the present study, and all of the cases survived to be born, and 3 cases of fetal intervention and 7 cases of amniodrainage survived to discharge. However, detailed validation may be necessary to evaluate and definitively conclude the efficacy of these prenatal interventions. Of the 9 neonatal deaths, 6 infants had already developed hemorrhage from the tumor at the time of cesarean section, including 2 cases of tumor rupture during the cesarean delivery. Most of them were premature infants who had huge and predominantly solid tumors. An early delivery with an EXIT-to-resection strategy [21] or emergency preoperative tumor embolization [22] may have some benefits for such critical cases.

A major limitation of this study is that this survey was conducted in a retrospective manner by using a questionnaire requesting details about the patients. Many of the centers had a small number of cases, and the maternal and fetal management, including the criteria for fetal intervention, were determined according to the clinical decisions of each institution. Moreover, prognostic factors were analyzed only by a descriptive study. A more detailed analysis of the data and a prospective study will therefore be needed to establish a comprehensive treatment strategy, including preoperative tumor embolization, EXIT procedures, and fetal interventions.

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Usefulness of exchanging a tunneled central venous catheter using a subcutaneous fibrous sheath

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ABSTRACT

Objectives: The reserve of the venous route to the central veins is important for long-term parenteral nutrition (PN). Frequent catheter-related bloodstream infection (CRBSI) induces occlusion of the venous routes. Therefore, a modified exchange procedure using a tunneled central venous catheter (CVC) with a fibrous sheath was developed to preserve the route to the central veins.

Methods: Seven patients who required long-term PN received the modified exchange procedure and the outcome of exchanged CVC was retrospectively reviewed.

Results: The procedure was performed 10 times in seven patients. The venous routes were either the subclavicular or the internal jugular vein in all patients. The exchange of the catheter was due to CRBSI or occlusion in almost all patients. The mean duration of new catheter use was 296.2 days following the exchange. Four catheters continued to be used, and the remaining ones were removed. The reasons for removal were severe CRBSI and occlusion, each of which occurred in two catheterized patients, while the reason for removing the remaining catheters was because the patients no longer needed the catheters.

Conclusion: The modified catheter exchange using fibrous sheath, even in patients with CRBSI, appears to be an effective procedure for reserving the venous route to the central veins in patients who require either long-term PN or other treatments.

Introduction

The placement of a tunneled central venous catheter (CVC) or implantable infusion port in the central veins is an effective method for providing venous access in various clinical situations [1–3]. In fact, these devices can be effectively used for long-term parenteral nutrition (PN), aggressive chemotherapy, and stem cell or bone marrow transplantation, particularly in pediatric patients [1,2]. It is occasionally necessary to reinsert or exchange the catheter or port during the long-term use of CVC for several reasons, which include catheter-related bloodstream infection (CRBSI), occlusion, or catheter damage [3–6]. Usually the occluded or damaged CVC associated with CRBSI is removed and the new CVC is inserted from a different cutaneous site [3,4]. However, when the long-term use of venous access for long-term PN is required, the number of available venous routes to the central veins is often limited due to infection, inflammation, or thrombosis caused by the frequent uses of these routes [4,7,8]. Therefore, clinicians must maintain the venous routes to the central vein in patients who need the long-term use of venous routes.

Brevetti et al. first showed that the use of an existing subcutaneous route around the existing catheter is useful to maintain venous access routes when the catheter is damaged or inadvertently displaced during the long-term use [8]. Some reports also recommend a similar technique for reinsertion of a new central venous catheter following accidental catheter removal [9–14]. In almost all reports, the use of the insertion technique is not recommended for patients with CRBSI [8–13].

Based on the previously reported exchange or reinsertion procedure of a CVC [7–13], a modified exchange procedure of CVC for long-term use of the venous route was developed using the subcutaneous route composed of a fibrous sheath (FS) that was used in patients with CRBSI. This procedure helped to maintain the venous route to the central vein, even in patients with CRBSI.

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Patients and methods

Patients

Seven patients, including five pediatric patients, who were treated between April 2007 and March 2009 and required long-term CVC use, received the exchange of a Broviac (Bard, Salt Lake City, Utah, USA), a Hickman catheter (Bard), or a Port-type catheter (5 or 6 F, P-U Celsite Port; Toray Medical Co. Ltd., Tokyo, Japan) using subcutaneous FS. In the patients, one patient underwent this procedure three times and one patient underwent catheter exchange twice. Therefore, this procedure was performed 10 times in seven patients. Table 1 shows the basic characteristics of each patient. In seven patients, five were infants and two were adults. The mean age at the time of exchange of CVC was 20.6 ± 13.3 mo in the five pediatric patients. The minimum age was 9 mo at the time of CVC exchange using this procedure. In adult patients, the age at the time of exchange of CVC was 26 and 29 y. The mean body weight was 6.5 ± 4.0 kg in pediatric patients. The minimum body weight was 4.1 kg at the time of the exchange of the CVC. In adult patients, the body weights were 32 and 43 kg. The primary underlying diseases included extremely short bowel syndrome, intestinal hypoganglionosis with or without Hirschsprung's disease, chronic idiopathic intestinal pseudo-obstruction syndrome, and chronic granulomatous disease. The main reason for the long-term use of the CVC in all patients was to receive long-term PN.

All parents of the patients were informed of this procedure before the exchange and provided informed consent for their children to undergo the exchange.

Modified procedure for exchange of CVC

The modified procedure was based on Brevetti et al.'s [8] and Inoue et al.'s [14] procedures. The procedure herein was conducted for CVC exchange via the subclavicular or internal jugular vein. Patients with CRBSI were given a broad spectrum antibiotic over the 1–2 d prior to the exchange procedure. Thereafter, the procedure was performed if the CRBSI-associated fever improved. In addition, the CRBSI diagnoses were made based on using bacterial culturing findings of the arterial blood, the aspirated blood through the CVC, and the discharge at the CVC exit site. We finally decided that the CRBSI was positive when the bacterial findings in the aspirated blood culturing were matched with that in the arterial blood, giving no evidence of exit site or tunnel infection. The patients with fungal infection were excluded as candidates for the exchange procedure.

The patient was placed in a supine position under general anesthesia and the skin was scrubbed and sterilized using povidone-iodine under the maximum barrier precautions in the operation room. After localizing the subcutaneous Dacron cuff of the existing catheter, a skin incision was made 1 cm proximal to the palpable cuff (Fig. 1A). The subcutaneous tissue was gently opened until the FS around the existing CVC was found. The FS was lifted by forceps or vinyl tape (Fig. 1B). Two or three supporting sutures were placed on the both sides of anterior wall of the FS and then the anterior wall of the FS was opened. At that time, the posterior wall of the FS was not intentionally cut to prevent the retraction of remaining FS to the venous site. The existing tunneled CVC was cut at the opening site of the FS and the proximal catheter was removed (Fig. 1C). A length of new sterile tunneled CVC was trimmed to the length from the new cutaneous exit site to the superior vena cava or right atrium using x-ray imaging. The new tunneled CVC was inserted from the new exit to the wound at the FS lifting site using a tunneler attached to the new catheter (Fig. 1C). Thereafter, the new CVC was smoothly placed through the FS on the superior vena cava or right atrium without a guidewire using x-ray imaging. The posterior wall of the FS was cut and removed with the remaining CVC. All wounds were closed and the new inserted CVC was fixed on the skin at the new exit site (Fig. 1D).

Results

The details of the exchange of the tunneled CVC are shown in Table 1. All of the existing tunneled CVCs were 4.2-F Broviac catheters, other than three port type catheters. The insertion was on the right side in all pediatric patients and on the left side in adult patients. The venous routes were either the subclavicular or the internal jugular vein in all patients. The subclavicular vein was used six times in five patients, while the internal jugular vein was used four times in three patients. The new tunneled CVCs were 4.2-F Broviac catheters in all pediatric patients but one patient, who received a 7-F Hickman catheter. In adult patients, the new catheters were 6.0-F Port type catheters. In eight of the catheters in six patients, the new CVC was similar in

Table 1
Patients exchanged tunneled CVC using fibrous sheath

Patient	Age	Sex	BW (kg)	Primary underlying diseases	Existed CVC	VAR used for exchange	Inserted new CVC	Reason for exchange	Duration of new CVC use	Reason of new CVC removal	Remaining no. of VAR
1	12 mo	G	4.1	SBS (10 cm)*	4.2-F Broviac	Internal jugular	4.2-F Broviac	CRS	86 d	Died (HF)	5
2	11 mo	B	4.2	Hirschsprung's disease + hypoganglionosis	4.2-F Broviac	Subclavicular	4.2-F Broviac	CRS	120 d	CRS	5
	20 mo	B	5.7	Same as above	5-F Port	Internal jugular	4.2-F Broviac	CRS	168 d	CRS	4
	25 mo	B	7	Same as above	4.2-F Broviac	Internal jugular	4.2-F Broviac	CRS	425 d†	—	4
3	12 mo	B	4.3	SBS (5 cm)*	4.2-F Broviac	Subclavicular	4.2-F Broviac	CRS	100 d	Occlusion	5
	27 mo	B	5.2	Same as above	4.2-F Broviac	Subclavicular	4.2-F Broviac	CRS	301 d	Occlusion	5
4	9 mo	B	5.3	SBS (4 cm)*	4.2-F Broviac	Subclavicular	4.2-F Broviac	CRS	82 d	Discontinuation of PN	5
5	49 mo	B	16	CGD	4.2-F Broviac	Subclavicular	7-F Hickman	Treatment	428 d†	—	6
6	26 y	M	32	CIIPS	6-F Port	Subclavicular	6-F Port	Local infection	833 d†	—	4
7	29 y	M	43	Hypoganglionosis	6-F Port	Internal jugular	6-F Port	Occlusion	419 d†	—	1

B, boy; BW, body weight; CGD, chronic granulomatous disease; CIIPS, chronic idiopathic intestinal pseudo-obstruction syndrome; CRS, catheter-related sepsis; CVC, central venous catheter; G, girl; HF, hepatic failure; M, male; mo, months old; no., number; PN, parenteral nutrition; SBS, short bowel syndrome; VAR, venous access route; y, years old

* The parentheses mean the length of remaining small bowel.

† The CVC is still being used.

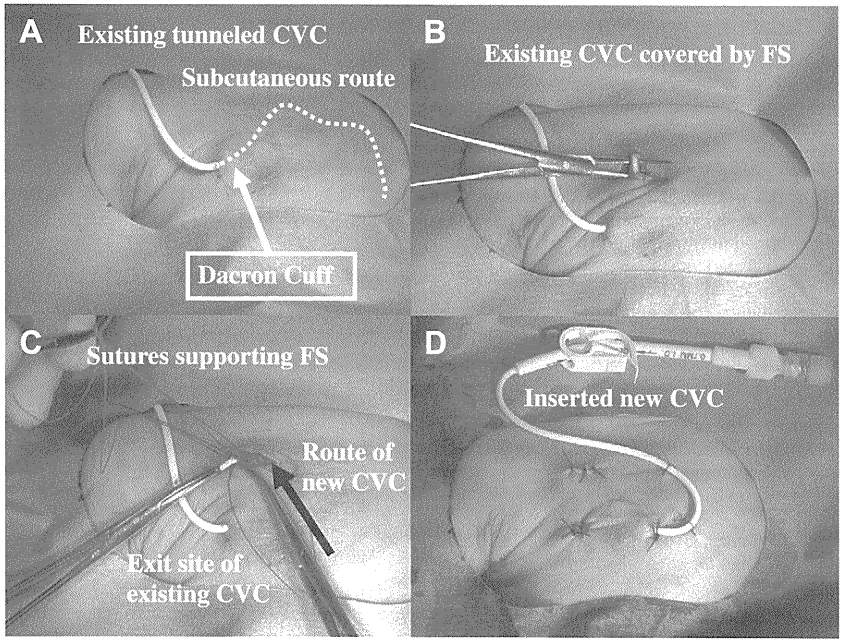

Fig. 1. Exchange procedure for a tunneled central venous catheter (CVC). (A) The subcutaneous route of the existing tunneled CVC. The dotted white line shows the subcutaneous route. The skin incision was made 1 cm proximal to the palpable Dacron cuff. (B) The fibrous sheath (FS) around the existing tunneled CVC. (C) The stump of the existing tunneled CVC and the route of insertion for the new tunneled CVC. The anterior wall of the FS is supported by sutures. (D) The newly inserted tunneled CVC using the FS. The CVC is inserted through the existing FS route from a new exit site.

size to the existing CVC. In one patient (patient 5), the new CVC catheter size was larger than the existing CVC.

The reason for exchanging the CVC was CRBSI in four pediatric patients. The remaining pediatric patient underwent the exchange to receive further treatment with long-term PN for chronic granulomatous disease. In adult patients, the exchange was due to local infection of the insertion site or to catheter occlusion. The new tunneled CVCs were used for a mean duration of 296.2 d following the CVC exchange. The four new CVCs are still in use, while 6 of the 10 exchanged CVCs were removed. The reason for removal was severe CRBSI, which antibiotics did not resolve, in two pediatric patients with two new catheters. In three patients with new catheters, the reason for removal was due to the occlusion of the catheter. The remaining two CVCs were removed because the pediatric patients no longer required the CVC, due to either death or PN discontinuation. After the exchange procedure, the number of venous routes to the central vein was more than four in all pediatric patients. In the patients with CRBSI prior to the exchange, the CRBSI improved immediately after CVC exchange. In addition, no complications were associated with the catheter in any patients during the 3-mo period following the exchange procedure.

Discussion

In general, a tunneled CVC should be removed if it becomes occluded or infected, and a new CVC is then reinserted [5]. However, in adult and pediatric patients who require long-term CVC use for the treatment of diseases including intestinal failure, the frequent removal of tunneled CVCs due to occlusion or infection often causes a decrease of the venous routes to the central vein [3–8]. This may cause limitations for the administration of conservative or surgical treatments for the disease leading to the death of the patients. Therefore, clinicians who treat patients requiring long-term treatment, including PN, usually try to preserve the number of venous routes to the central vein during the long-term treatment period [8,9]. Numerous clinical reports have addressed CVC exchange or reinsertion procedures that preserve the number of venous routes to the central vein [8–14]. CVC exchange or reinsertion using the FS formed around the existing tunneled CVC is effective and safe [8–14]. However, this procedure is not yet universally accepted because it is limited only to cases where the CVC is occluded or accidentally removed [8–13]. Therefore, the tunneled CVC exchange procedure was modified to confirm if it could be used in patients with CRBSI.

The important conclusion herein was that we were able to perform CVC exchange in patients with CRBSI. Since the introduction of the procedure by Brevetti et al. [7], similar tunneled CVC exchange procedures using the FS have been proposed, but the procedures were contraindicated by CRBSI. CRBSI is assumed to originate from a bacterial bank, such as an infected biofilm or fibrous materials, which are attached to lumen of the CVC [7,15,16]. To confirm this origin of CRBSI, a wide broad spectrum antibiotic was administrated via the existing tunneled CVC 1–2 d prior to performing the exchange procedure in this report. If the symptoms were improved after the antibiotic treatment, the origin was thought to be associated with the intraluminal biofilm or fibrous materials associated with the existing CVC. Therefore, the exchange of an intraluminally infected CVC with a new sterile one would be advantageous for patients with CRBSI. Furthermore, in the current procedure, the distal FS was completely removed to prevent the bacterial invasion from the existing exit. There were no infectious complications immediately after the modified exchange CVC procedure, although the procedure was performed seven times in four CRBSI patients whose symptoms gradually improved after 1–2 d of antibiotic treatment. In addition, Inoue et al. reported that a similar exchange procedure was effective in two adult patients with CRBSI [14]. Therefore, this modified exchange procedure might be useful for preserving the venous route to the central vein, even in patients with CRBSI. However, to elucidate the