JNS PEDIATRICS

CLINICAL ARTICLE

J Neurosurg Pediatr 29:479–487, 2022

Number of surgeries performed during the lifetime of patients with myelomeningocele

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OBJECTIVE Patients with myelomeningocele often require multiple surgeries, but no study has clarified the kind of treatment given to these patients throughout their lives. The authors analyzed the type of surgery that was performed and at what age for Japanese patients with myelomeningoceles.

METHODS The Japanese health claims data of 556 patients with myelomeningocele for the period from January 2005 to March 2020 provided by the Japan Medical Data Center Co., Ltd., were examined to investigate the number of surgeries performed and the patient age at surgery for each specialty. The patients were divided into two groups (those ≤ 18 years old [group A] and those > 18 years old [group B]), and the way in which the types of surgery and the percentage of surgeries changed between these two groups was examined.

RESULTS The mean follow-up period was 4.4 years. The mean age at the end of the overall follow-up was 18.6 years (range 0–70.5 years), and 1033 surgeries were performed on 294 patients (0.42 surgeries performed per patient per year) during this period. The number of surgeries for patients in group A was 818 in 192 patients, with 0.62 surgeries per patient per year, and for patients in group B it was 215 in 102 patients, with 0.19 surgeries per patient per year. The number of surgeries and the mean age at the time of surgery were as follows: 313 neurosurgeries, 5.16 years; 280 orthopedic surgeries, 11.36 years; 70 urological surgeries, 14.57 years; and 202 dermatological/plastic surgeries, 16.19 years. In the surgeries related to myelomeningocele, the rates of CSF shunt placement, tethered cord release, muscle and tendon surgery, and other bone and joint surgery decreased significantly in group B, but they continued to undergo these surgeries. In group B, the rates of skin surgery, nephrostomy, ureterostomy, and cystostomy were significantly higher.

CONCLUSIONS A significant number of surgeries in multiple specialties related to myelomeningocele continue to be performed in adulthood, indicating that these patients require continuous care throughout their lives.

https://thejns.org/doi/abs/10.3171/2021.12.PEDS21535

KEYWORDS myelomeningocele; surgery; transition to adult; congenital

It is well known that patients with myelomeningocele undergo various surgeries throughout their lives[1,2]—not only neurosurgery,[3] but also orthopedic surgery,[4–6] urological surgery,[7] dermatological procedures, and others. However, how many surgeries are performed in patients with myelomeningocele over their lifetime is unclear. Surgeries often require hospitalization and often result in situations in which normal social activities are interrupted, including the need for rehabilitation for some time after surgery. We thought that by clarifying the types and the number of surgeries, it would be possible to get an idea of what kind of problems the patients face and what kind of medical care is needed for each age group.

Japan has a universal health insurance system, and claims for medical services such as surgery, medication, consultation, and medical treatment are billed to insurance associations from medical institutes. Whenever a medical procedure such as surgery is performed, the corresponding name of the disease is entered for the claim. By analyzing the Japan Medical Data Center (JMDC) claims database, which is an anonymized database of health insurance claims, it is possible to clarify what kind of treatment is being provided for myelomeningocele in real-world practice in Japan. In this study, the types of surgeries performed in patients with myelomeningocele in Japan were analyzed by patient age and surgical specialty.

ABBREVIATIONS ACE = antegrade continence enema; ETV = endoscopic third ventriculostomy; EVD = external ventricular drainage; JMDC = Japan Medical Data Center; NTD = neural tube defect; SB = spina bifida.

SUBMITTED November 15, 2021. **ACCEPTED** December 20, 2021.
INCLUDE WHEN CITING Published online February 18, 2022; DOI: 10.3171/2021.12.PEDS21535.

Methods

Database

This study design was a cohort study that used the claims database provided by the JMDC, one of the largest health insurance databases in Japan provided by the private sector. The JMDC claims database holds cumulative data on more than 10 million people (approximately 8.1% of Japan's population) from January 2005 through March 2020, including individuals who did not use any healthcare services. The JMDC claims database was acquired by the Research Committee of CNS Degenerative Diseases (Research on Policy Planning and Evaluation for Rare and Intractable Diseases; Health, Labor, and Welfare Sciences Research Grants; The Ministry of Health, Labor, and Welfare) for this study.

The database contains integrated medical and pharmacy claims data, including inpatient and outpatient data, from more than 90 health insurance associations covering company employees and their families.[8–10] The health insurance associations that contracted with JMDC do not include sole proprietors or welfare recipients; only those 75 years of age or younger are included in JMDC's database, because those older than 75 years of age are covered by the National Health Insurance system. Personal information is encrypted, and billing data are recorded by each individual's age under a unique encrypted identifier. The database records information on all visits, including diagnoses, tests, medication details, and surgeries from all medical institutions and healthcare providers. In addition, each receipt contains the date and time when each individual started visiting a medical institution during the period in question and the date of the last visit, which was used as the follow-up in this study. Therefore, the present study was not limited to patients born during this period.

Study Design

Diagnostic names for insurance claims are coded according to the ICD-10. In this study, the claims of patients whose disease was designated as myelomeningocele were extracted and analyzed. The following items were examined: age at the beginning of follow-up, age at the end of follow-up, age in March 2020, type of surgery undergone during the follow-up period, and age at the time of surgery. For surgery, myelomeningocele repair and untethering were included as neurosurgical procedures, and spinal deformity surgeries as orthopedic procedures. The distribution of age as of March 2020 and the distribution of age at the time each surgery was performed were examined. Each surgery was also classified into two groups by age (≤ 18 and > 18 years old: group A and group B, respectively), the percentage of each type of surgery in each group was calculated, and which items showed a percentage change was examined. By comparing the surgeries related to myelomeningocele in groups A and B, the issues that need to be taken into account in follow-up when the patients become adults were clarified.

There is a possibility that some of the cases designated as myelomeningocele may be closed dysraphism like lipomyelomeningocele, not myelomeningocele itself. If the patient had true myelomeningocele, the percentage of patients treated for hydrocephalus would be higher, and if more patients with closed dysraphism were included, the percentage would be lower. Therefore, we investigated the rates of hydrocephalus treatment performed in cases in which myelomeningocele repair was performed to estimate how many cases of closed dysraphism were mixed in with this group.

The protocol for this study was approved by the ethics committee of Kansai Medical University. Written patient consent was waived because data were de-identified. Raw data were generated by the JMDC. Derived data supporting the findings of this study are available from the corresponding author (M.N.) on request.

TABLE 1. Demographics in 556 patients with myelomeningocele

Characteristic	Value
No. of pts	10,284,139
No. of pts w/ myelomeningocele	556
≤18 yrs old (as of March 2020)	301
>18 yrs old (as of March 2020)	255
Mean age at start of observation, yrs (min–max)	14.2 (0–65.6)
Mean age at end of observation, yrs (min–max)	18.6 (0–70.5)
Mean observation period, yrs (min–max)	4.4 (0–15.2)
M/F ratio	270:286
No. of pts who underwent surgery	294
Pts ≤18 yrs old/>18 yrs old	192:102*
Total no. of surgeries	1033
Pts ≤18 yrs old/>18 yrs old	818:215
No. of deaths	4
Neurosurgical procedures	313
Orthopedic procedures	280
Dermatological/plastic surgical procedures	202
Urological procedures	70
Otorhinolaryngological procedures	47
Ophthalmological procedures	14
General surgery/other procedures	75
Obstetrical & gynecological procedures	32

Max = maximum; min = minimum; pts = patients.
* There was a significant decrease in the number of people 19 years and older who had surgery compared to those 18 years and younger ($p < 0.001$).

Statistical Analysis

For statistical analysis, demographic characteristics between groups were compared using Pearson's chi-square test for categorical variables and a 2-sample t-test for continuous variables. Statistical analyses were conducted using JMP 14.2.0, and p values < 0.05 were considered significant.

Results

An overview of the patients is shown in Table 1. The number of people with claims included in the analysis was 10,284,139. This represents approximately 8.2% of the total Japanese population of 125 million.

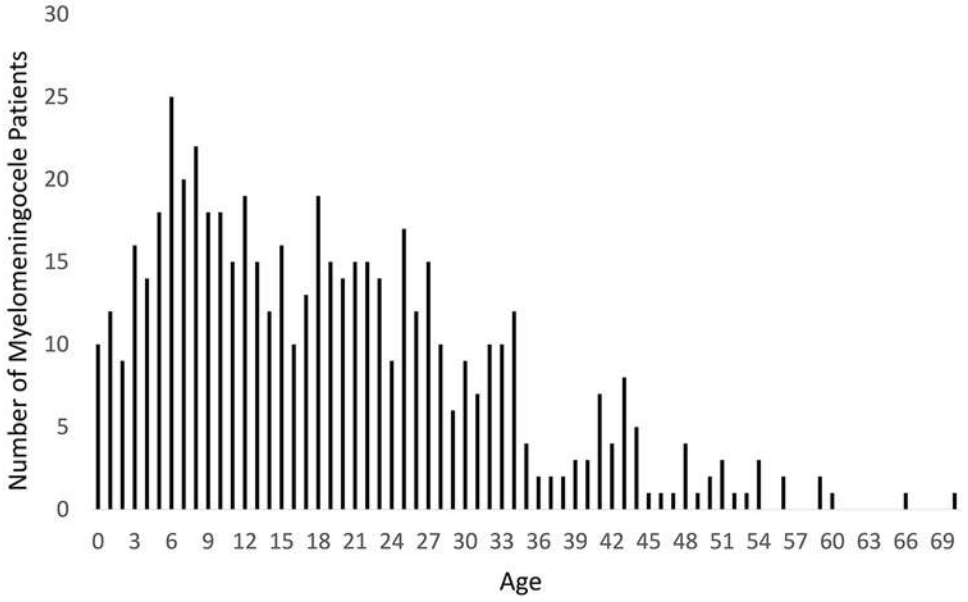

FIG. 1. Distribution of patients with myelomeningocele by age (as of March 2020). Horizontal axis: age (years); vertical axis: number of patients with myelomeningocele.

The number of patients with myelomeningocele was 556, which is 5.4 patients per 100,000 population in Japan. The distribution of patients by age as of March 2020 is shown in Fig. 1. The mean age of patients was 19.0 years, with a peak at 6 years. There were 301 patients in group A and 255 patients in group B. The mean follow-up period was 4.4 years, with 270 male and 286 female patients. The total number of patients who underwent surgery during this follow-up period was 294, with 192 patients in group A and 102 patients in group B. The number of patients who underwent surgery decreased significantly after age > 18 years ($p < 0.001$). The total number of surgeries was 1033, with 0.42 surgeries performed per patient per year. The number of surgeries for patients in group A was 818, with 0.62 surgeries per patient per year, and for patients in group B it was 215, with 0.19 surgeries per patient per year.

The mean age at surgery for each specialty and the significant differences between specialties are shown in Table 2. The mean age at the time of surgery was 5.16 years for neurosurgery, 9.02 years for otorhinolaryngology, 11.36 years for orthopedics, 16.19 years for dermatological and plastic surgery, 14.57 years for urology, 16.73 years for general and other surgery, 21.0 years for ophthalmology, and 31.09 years for obstetrics and gynecology. The mean age at the time of neurosurgery was significantly younger than that of other specialties. On the other hand, dermatological/plastic surgery occurred

TABLE 2. The mean age at surgery for each specialty and the significant differences between specialties

		p Value for Procedure (vertical vs horizontal column)							
Procedure	Mean Age in Yrs (SE)	Neurosurgery	Otorhinolaryngology	Orthopedics	Urology	Dermatology/ Plastic Surgery	General/ Other Surgery	Ophthalmology	Obstetrics & Gynecology
Neurosurgery	5.16 (0.68)	—	0.0423	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001
Otorhinolaryngology	9.02 (1.77)	0.0423	—	0.2228	0.0156	0.0003	0.0006	0.0011	<0.0001
Orthopedics	11.36 (0.72)	<0.0001	0.2228	—	0.0472	<0.0001	0.0006	0.0035	<0.0001
Urology	14.57 (1.45)	<0.0001	0.0156	0.0472	—	0.0472	0.2797	0.068	<0.0001
Dermatology/ plastic surgery	16.19 (0.84)	<0.0001	0.0003	<0.0001	0.0472	—	0.7355	0.1473	<0.0001
General/other surgery	16.73 (1.39)	<0.0001	0.0006	0.0006	0.2797	0.7355	—	0.223	<0.0001
Ophthalmology	21 (3.21)	<0.0001	0.0011	0.0035	0.068	0.1473	0.223	—	0.0089
Obstetrics & gynecology	31.09 (2.13)	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001	0.0089	—

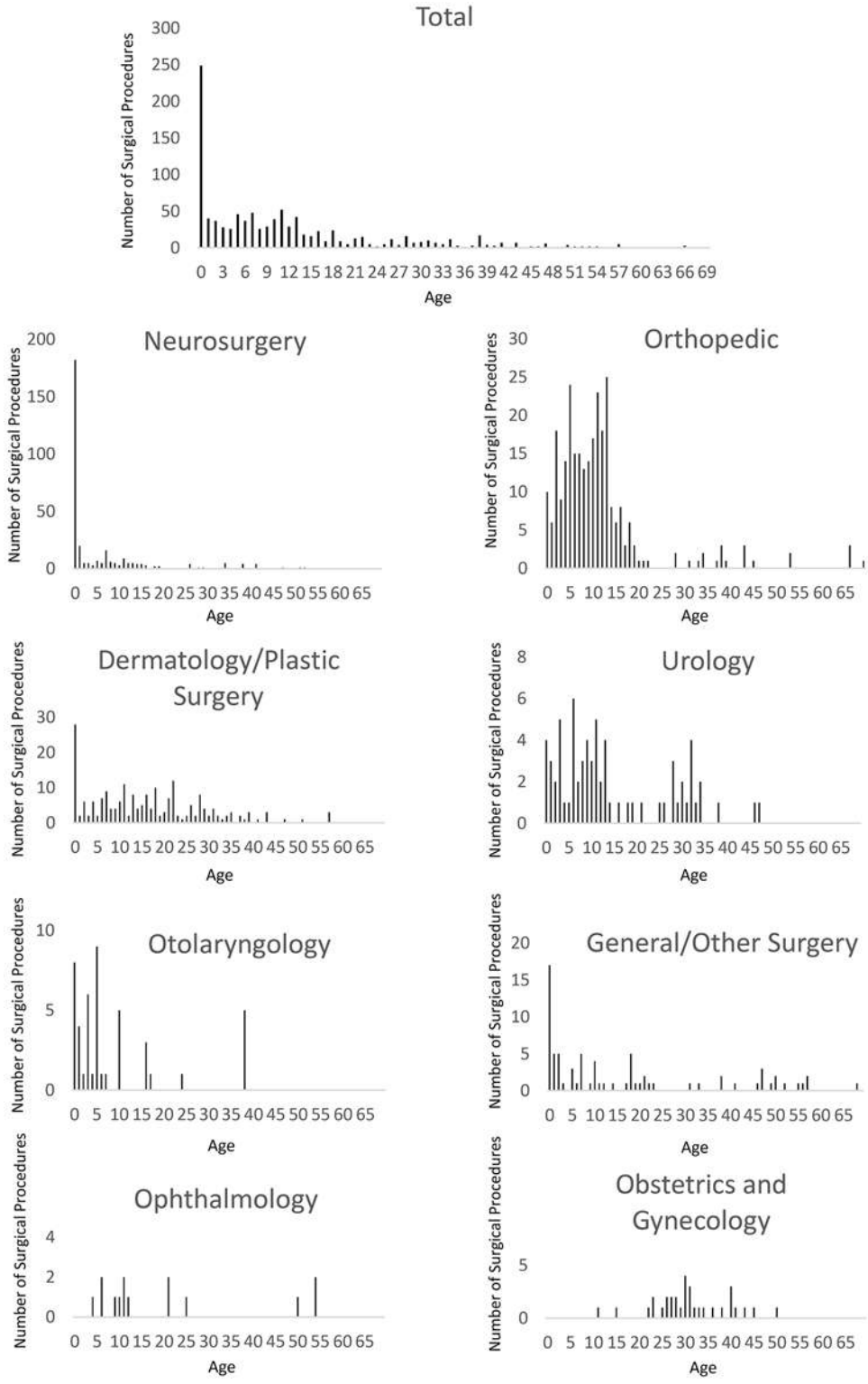

FIG. 2. Distribution of the number of operations for patients with myelomeningocele by age at the time of surgery. The overall number and the distribution for each specialty are shown. Horizontal axis: age (years); vertical axis: number of surgical procedures.

at a significantly older age than neurosurgery, otorhinolaryngological, orthopedic, and urological surgery, and an older segment of patients with myelomeningocele underwent this surgery. Obstetrical and gynecological surgery was performed at the oldest age compared to the other surgeries.

Figure 2 shows the age at which surgery was performed in each specialty. In the case of neurosurgery, the peak age

was at 0 years, and although the number of surgeries decreased rapidly, surgeries were still performed after adulthood, indicating that the patients required lifelong treatment for the long-term cases. Orthopedic surgery peaked at 6 and 14 years of age, and although it declined thereafter, surgery was still performed after patients reached adulthood. Dermatological surgeries were performed mostly in the age group of 0 years, and thereafter the number of surgeries peaked gradually at approximately 20 years of age, and then decreased slowly until approximately 40 years of age. Urological surgery peaked at approximately 6 and 32 years of age. Otorhinolaryngological surgery peaked at approximately 5 years of age, and it was less frequently performed during the adolescent period. General surgery was most common at the age of 0 years and was performed until old age, although it decreased slightly. Ophthalmological surgery showed no consistent trend, and obstetrical and gynecological surgery began to increase at approximately the age of 20 years, reaching a peak at approximately the age of 30 years.

Table 3 shows the types of surgeries performed by specialty and the percentage of each type of surgery in group A and in group B. The surgeries considered to be related to myelomeningocele were the following: CSF shunt, endoscopic third ventriculostomy (ETV), myelomeningocele closure, untethering, external ventricular drainage (EVD), posterior fossa decompression, spinal deformity surgery, joint dislocation, bone fracture, muscle and tendon surgery, clubfoot surgery, bone and joint surgery, skin flap/transplantation, other skin surgery, bladder augmentation, nephrostomy/ureterostomy/cystostomy, vesicoureteral reflux surgery, dialysis access, cesarean section, tracheostomy/laryngotracheal separation, colostomy, and gastrostomy/enterostomy.

Among these surgeries, the proportion of patients in group B was significantly higher for skin flap/transplantation, other skin surgery, and nephrostomy/ureterostomy/cystostomy. CSF shunt, untethering, muscle and tendon surgery, and other bone and joint surgery were significantly more common in group A. The proportion of patients who underwent EVD, posterior fossa decompression, spinal deformity surgery, joint dislocation and bone fracture surgery, bladder augmentation, vesicoureteral reflux surgery, colostomy, gastrostomy/enterostomy, and tracheostomy/laryngotracheal separation was not significantly different between group A and group B.

Within the observation period of this study, 39 (68.4%) had been treated for hydrocephalus among 57 new patients who underwent myelomeningocele repair. CSF shunt placement was performed in 32 cases, EVD in 6 cases, and ETV in 1 case as the initial surgery for hydrocephalus.

During the follow-up period, at least 4 patients with myelomeningocele died. The ages of the patients were 0, 2, 22, and 50 years. The patients who were 2 and 50 years old had no record of medical treatment before death, suggesting that they died before obtaining medical attention. The 0-year-old (infant) patient had undergone cardiac surgery immediately before death, suggesting that the death was due to complications of cardiac disease. The 22-year-old patient had undergone multiple skin surgeries, including skin valvuloplasty, before death.

Discussion

In this study, the type and number of myelomeningocele surgeries and the age of the patients were clarified. The number of surgeries per person per year was 0.62 for group A but decreased to 0.19 for group B, although it was still high. Only Marreiros et al.[11] reported the number of surgeries (although only for patients younger than 21 years of age) within their institution. Based on their report, they sought to determine how many surgeries were performed per person per year. They considered 84 cases of myelomeningocele in patients younger than 21 years and found that 663 surgeries were performed, which gives an average of 7.9 surgeries per person and 0.56 surgeries per year, given that the average age was reported to be 14 years. In our study, patients ≤ 18 years of age had 0.62 surgeries per year, which is close to the value reported by Marreiros et al. On the other hand, there are no reports of the number of surgeries performed in adults, and our study is the first to report that information. Most of the surgeries performed in adult patients were related to myelomeningocele.

Studies have been conducted in the past to examine the situation of patients with myelomeningocele who have reached adulthood, such as the study of patients with myelomeningocele who transitioned to a medical home (i.e., a patient-centered care home),[12] the report examining the mortality rates of patients with myelomeningocele,[13,14] the study examining the changes in myelomeningocele across the US,[15] and assessment of health literacy and self-reported readiness for transition to adult care.[16] Myelomeningocele has also been suggested to benefit from continuous care throughout life.[17,18] Identifying the number of surgeries and the age at which the surgeries were performed will help us to examine these patients' medical needs for better transitional care.

Neurosurgery

Patients with myelomeningocele undergo repair of the lesion immediately after birth, and many also undergo treatment for hydrocephalus in the neonatal period. All of these surgeries are more likely to be performed when the patient is younger than 1 year old, so the average age for neurosurgical procedures is lower than for procedures in other specialties.

In our study, 68.4% of patients who underwent myelomeningocele repair during the period received treatment for hydrocephalus. Adzick et al. reported that among patients who had postnatal myelomeningocele repair, 82% underwent surgery for hydrocephalus.[19] On the other hand, McCarthy et al. found that 77.5% of myelomeningocele cases had hydrocephalus, of which 24.6% were untreated.[20] Our result was consistent with previous reports. The slight differences between past reports and our data may be due to differences in the rate of hydrocephalus and the indications for shunting among centers, or it may be due to the inclusion of some cases of closed dysraphism in our study. However, even if closed dysraphism is included, the rate is considered to be low.

Although a total of 18 ETVs were performed, only 1 patient underwent ETV as initial treatment for hydrocephalus after myelomeningocele repair during the study

TABLE 3. The types of surgeries performed by specialty in patients ≤ 18 and > 18 years old

Procedure	Group A, No. of Pts ≤18 Yrs (%)	Group B, No. of Pts >18 Yrs (%)	% in A vs % in B	p Value
Neurosurgical interventions				
CSF shunt	93 (11.37)	7 (3.26)	A > B	0.0009*
EVD	42 (5.13)	7 (3.26)	A > B	0.2689
ETV	18 (2.2)	0 (0)	A > B	
Myelomeningocele closure	57 (6.97)	0 (0)	A > B	
Tethered cord release	53 (6.48)	3 (1.4)	A > B	0.0049*
Posterior fossa decompression	2 (0.24)	1 (0.47)	A < B	0.594
Other neurosurgical procedures	24 (2.93)	6 (2.79)	A > B	0.9138
Total	289 (35.33)	24 (11.16)		<0.0001*
Orthopedic interventions				
Spinal deformity surgery	24 (2.93)	5 (2.33)	A > B	0.6397
Muscles & tendons	64 (7.82)	3 (1.4)	A > B	0.0012*
Joint dislocation	9 (1.1)	6 (2.79)	A < B	0.0705
Bone fracture	21 (2.57)	6 (2.79)	A < B	0.8588
Clubfoot surgery	22 (2.69)	0 (0)	A > B	
Other bone & joint surgery	113 (13.81)	7 (3.26)	A > B	<0.0001*
Total	253 (30.93)	27 (12.56)		<0.0001*
Dermatological/plastic surgical interventions				
Skin flap/transplantation	14 (1.71)	12 (5.58)	A < B	0.0019*
Skin tumor resection	14 (1.71)	11 (5.12)	A < B	0.0052*
Nail surgery	5 (0.61)	4 (1.86)	A < B	0.0832
Other skin surgery	92 (11.25)	50 (23.26)	A < B	0.0001*
Total	125 (15.28)	77 (35.81)		<0.0001*
Urological interventions				
Bladder augmentation	5 (0.61)	3 (1.4)	A < B	0.2479
Vesicoureteral reflux surgery	9 (1.1)	0 (0)	A > B	
Nephrostomy/ureterostomy/cystostomy	6 (0.73)	5 (2.33)	A < B	0.0462*
Cryptorchidism surgery	9 (1.1)	0 (0)	A > B	
Urolithiasis surgery	7 (0.86)	9 (4.19)	A < B	0.0006*
Other urological procedure	13 (1.59)	4 (1.86)	A < B	0.7845
Total	49 (6)	21 (9.77)		0.0697
Otorhinolaryngological interventions				
Tracheostomy/laryngotracheal separation	10 (1.22)	0 (0)	A > B	
Ear surgery	21 (2.57)	0 (0)	A > B	
Nose surgery	5 (0.61)	5 (2.33)	A < B	0.0244*
Other otorhinolaryngological procedures	5 (0.61)	1 (0.47)	A < B	0.8029
Total	41 (5.01)	6 (2.79)		0.1811
Ophthalmological interventions				
Strabismus surgery	5 (0.61)	1 (0.47)	A > B	0.8029
Other ophthalmological surgery	3 (0.37)	5 (2.33)	A < B	0.0040*
Total	8 (0.98)	6 (2.79)		0.0477*
General surgery/other interventions				
Colostomy	11 (1.34)	1 (0.47)	A > B	0.2885
Gastrostomy/enterostomy	6 (0.73)	3 (1.4)	A < B	0.3579
Central venous catheter/access port placement	4 (0.49)	0 (0)	A > B	
Congenital malformation surgery (imperforate anus, cloacal exstrophy, etc.)	6 (0.73)	0 (0)	A > B	
Dialysis access	0 (0)	3 (1.4)	A < B	
Other gastrointestinal surgery	20 (2.4)	10 (4.65)	A < B	0.0979

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TABLE 3. The types of surgeries performed by specialty in patients ≤ 18 and > 18 years old

Procedure	Group A, No. of Pts ≤18 Yrs (%)	Group B, No. of Pts >18 Yrs (%)	% in A vs % in B	p Value
General surgery/other interventions (*continued*)				
Cardiac surgery	2 (0.24)	2 (0.93)	A < B	0.1521
Other surgeries	2 (0.24)	5 (2.33)	A < B	0.0011*
Total	51 (6.23)	24 (11.16)		0.0229
Obstetrics and gynecological interventions				
Cesarean section	0 (0)	20 (9.3)	A < B	
Other obstetrics surgery	0 (0)	6 (2.79)	A < B	
Gynecological surgery	2 (0.24)	4 (1.86)	A < B	0.006*
Total	2 (0.24)	30 (13.95)		<0.0001*
Overall total	818	215		

* Statistically significant at $p < 0.05$

period. The rest were performed after other hydrocephalus treatments such as CSF shunts. For hydrocephalus in myelomeningocele, ETV with or without choroid plexus cauterization as initial treatment,[21,22] and ETV in case of CSF shunt failure[23,24] have been reported. In Japan, ETV is rarely chosen as an initial treatment and tends to be performed after other hydrocephalus treatments.

In this report, EVD was performed 49 times, which is a relatively large number. Eight of 57 patients (14%) who had myelomeningocele repair underwent EVD as the initial treatment for hydrocephalus. It has been reported that simultaneous myelomeningocele repair and EVD reduces wound complications in patients with myelomeningocele,[25] and there may be a certain number of Japanese neurosurgeons who accept this. Moreover, some Japanese neurosurgeons may still be cautious about performing a CSF shunt at the same time, although some reports, such as that of Radmanesh et al., have shown no difference in complication rates between myelomeningocele repair alone and CSF shunt placement performed simultaneously.[26] Finnegan et al. reported that 13% of patients with myelomeningocele in Ireland were treated simultaneously with myelomeningocele repair and EVD,[27] and this percentage was similar to that in our report. In other EVD cases, shunt infections may have occurred in relatively large numbers, and many patients may have undergone EVD until the infection subsided.

Hydrocephalus associated with myelomeningocele is a relatively common cause of infection and shunt malfunction.[3,28] Some cases of hydrocephalus including myelomeningoceles that were shunted in childhood require shunt revision in adulthood and lifelong follow-up.[29,30] In the present data, as in previous reports, hydrocephalus-related surgeries were performed 14 times in group B, suggesting the need for follow-up in adulthood. There is a report that tethered cord release was performed in 11% and 21.5% of myelomeningocele cases.[3,31] In the present report, 56 of 556 patients (10.0%) underwent tethered cord release during the follow-up period. Although most of the patients who underwent tethered cord release were in group A (53 patients), there were 3 cases of tethered cord syndrome in group B. Tethered cord syndrome is still present in adults, although few cases require surgery, and follow-up is important.

Orthopedics

A total of 280 orthopedic surgeries were performed, of which 253 were performed for patients in group A and 27 for patients in group B. The rates of spinal deformity, bone fracture, and joint dislocation surgery did not decrease in group B, indicating that these conditions continue to be a problem even after 18 years of age.

Children with spina bifida (SB) develop a variety of congenital and acquired orthopedic deformities. Some of these include deformities of the hip joint such as contractures, subluxations, and dislocations. There are also problems with the knee joint, including knee flexion/extension contractures, inward deformity of the knee, and late-onset knee instability and pain.[4,6] In addition, rotational deformities (inward or outward) of the lower extremities are common.[4,6] Regarding scoliosis, the results of the literature review indicated that the prevalence of scoliosis in patients with myelomeningocele was 53%.[32] However, there were no detailed data on the age at which these orthopedic surgeries were performed.

Urology

As for urological surgeries, cryptorchidism surgeries were lower in patients in group B, but the rates of bladder augmentation and vesicoureteral reflux surgery did not decrease in patients in group B. The number of urological surgeries also showed a bimodal pattern, with the first peak in childhood and the second peak in adulthood. Therefore, urinary tract management, especially in adulthood, was also shown to be important. A review of the literature on the urological management of SB from prenatal diagnosis to adulthood stated that surgical intervention is indicated when there is a risk of decreased renal function, and is also considered in children who do not have satisfactory voiding with medical management.[7] The rate of a successful transition from adolescent SB treatment to

adult SB treatment is also an issue,[33] so there is a need to ensure that there is an adequate system in place to see a specialist urologist in adulthood.

Dermatological and Plastic Surgery

There were 202 surgeries in dermatology and plastic surgery. This number was the second highest after neurosurgery and orthopedics. Most of the surgeries were for skin wounds, and it is assumed that most of them were for the treatment of skin ulcers (bedsores). The average age of patients at surgery in this area was higher than for other surgeries performed in patients with myelomeningocele, indicating that a significant proportion of adult patients are undergoing these surgeries. Myelomeningocele is a risk factor for the development of skin ulcers in adult and pediatric patients. However, there is a lack of information on the development of skin ulcers in patients with myelomeningocele. A review of the literature on pediatric neural tube defects (NTDs) found that only 11 papers published between 1975 and 2014 addressed skin ulcers in patients with NTDs, and more research is needed to guide ulcer prevention and treatment strategies in pediatric patients with NTDs.[34]

General Surgery and Other Surgery

In the area of gastrointestinal surgery, a total of 12 colostomies and 9 gastrostomies/enterostomies, probably for defecation management, were performed. The proportion of these surgeries did not change in group B, indicating that the problem of defecation continues into adulthood.

In Japan, the actual number of antegrade continence enema (ACE) procedures is unknown because surgical procedures for ACE are described as colostomy and enterostomy. Regarding the optimal management of neurogenic bowel, a report from North America stated that 17.3% were ACE/cecostomy tubes and 2.8% were ileostomy/colostomy.[35] Although ACE surgery does not appear to be as prevalent in Japan as it is in North America, factors such as race and treatment facility have also been associated with the likelihood of neurogenic bowel intervention in previous reports.[35]

Obstetrical and Gynecological Surgery

In the current study, 20 cesarean sections were performed for patients with SB, which suggests that some female patients with myelomeningocele deliver by cesarean section in Japan. According to previous reports, women with SB had a significantly higher rate of cesarean delivery than women without SB,[36,37] which suggests that young women may also need obstetric support.

Limitations of the Study

This study was based on a claims database, which is anonymized and unable to identify an individual, so that detailed information of individual cases, such as lesion level, was not available. Thus, no details regarding the patients or surgical trends over time are available. The data may be biased by patients transitioning into or out of this one health insurance database over time. In Japan, myelomeningocele is a designated intractable disease, and without this disease name on the claim, the patient cannot receive support through the recognition of intractable disease. Therefore, it is likely that there are only a few cases of myelomeningocele that do not have that name on the claim, but it is possible that some do exist. In addition, patients who did not visit a medical institution within the period covered were not included. There is a possibility that some of the cases designated as myelomeningocele may be closed dysraphism such as lipomyelomeningocele, not myelomeningocele itself.

Conclusions

The mean age of patients who underwent surgery by specialty was different, with the youngest receiving neurosurgery. The number of surgeries per person per year was 0.62 for those ≤ 18 years old, whereas the number of surgeries for those > 18 years old decreased to 0.19, but it was still found to be high. In the group of patients > 18 years old, the rate of skin flap/transplantation, other skin surgery, nephrostomy/ureterostomy/cystostomy, and dialysis access increased significantly. The results showed that conditions related to myelomeningocele continue to be problems in adulthood. Patients with myelomeningocele may require follow-up by a multidisciplinary team in adulthood, especially including skin wound care specialists.

Acknowledgments

This work was supported by grants-in-aid from the Research Committee of CNS Degenerative Diseases (Research on Policy Planning and Evaluation for Rare and Intractable Diseases; Health, Labor, and Welfare Sciences Research Grants; The Ministry of Health, Labor, and Welfare, Japan [20FC1049]).

References

1. Copp AJ, Adzick NS, Chitty LS, Fletcher JM, Holmbeck GN, Shaw GM. Spina bifida. *Nat Rev Dis Primers*. 2015;1:15007.
2. Patel SK, Staarmann B, Heilman A, Mains A, Woodward J, Bierbrauer KS. Growing up with spina bifida: bridging the gaps in the transition of care from childhood to adulthood. *Neurosurg Focus*. 2019;47(4):E16.
3. Talamonti G, D'Aliberti G, Collice M. Myelomeningocele: long-term neurosurgical treatment and follow-up in 202 patients. *J Neurosurg*. 2007;107(5)(suppl):368-386.
4. Swaroop VT, Dias L. Orthopedic management of spina bifida. Part I: hip, knee, and rotational deformities. *J Child Orthop*. 2009;3(6):441-449.
5. Thomson JD, Segal LS. Orthopedic management of spina bifida. *Dev Disabil Res Rev*. 2010;16(1):96-103.
6. Swaroop VT, Dias L. Orthopaedic management of spina bifida-part II: foot and ankle deformities. *J Child Orthop*. 2011;5(6):403-414.
7. Snow-Lisy DC, Yerkes EB, Cheng EY. Update on urological management of spina bifida from prenatal diagnosis to adulthood. *J Urol*. 2015;194(2):288-296.
8. Kimura S, Sato T, Ikeda S, Noda M, Nakayama T. Development of a database of health insurance claims: standardization of disease classifications and anonymous record linkage. *J Epidemiol*. 2010;20(5):413-419.
9. Uno S, Asakura T, Morimoto K, et al. Comorbidities associated with nontuberculous mycobacterial disease in Japanese adults: a claims-data analysis. *BMC Pulm Med*. 2020;20(1):262.
10. Fujimoto S, Tsuruoka N, Esaki M, et al. Decline incidence

in upper gastrointestinal bleeding in several recent years: data of the Japan claims database of 13 million accumulated patients. *J Clin Biochem Nutr*. 2021;68(1):95-100.
11. Marreiros H, Loff C, Calado E. Who needs surgery for pediatric myelomeningocele? A retrospective study and literature review. *J Spinal Cord Med*. 2015;38(5):626-640.
12. Aguilera AM, Wood DL, Keeley C, James HE, Aldana PR. Young adults with spina bifida transitioned to a medical home: a survey of medical care in Jacksonville, Florida. *J Neurosurg Pediatr*. 2016;17(2):203-207.
13. Borgstedt-Bakke JH, Fenger-Grøn M, Rasmussen MM. Correlation of mortality with lesion level in patients with myelomeningocele: a population-based study. *J Neurosurg Pediatr*. 2017;19(2):227-231.
14. Dicianno BE, Sherman A, Roehmer C, Zigler CK. Co-morbidities associated with early mortality in adults with spina bifida. *Am J Phys Med Rehabil*. 2018;97(12):861-865.
15. Kelly MS, Thibadeau J, Struwe S, Ramen L, Ouyang L, Routh J. Evaluation of spina bifida transitional care practices in the United States. *J Pediatr Rehabil Med*. 2017;10(3-4): 275-281.
16. Rague JT, Kim S, Hirsch JA, et al. Assessment of health literacy and self-reported readiness for transition to adult care among adolescents and young adults with spina bifida. *JAMA Netw Open*. 2021;4(9):e2127034.
17. Hopson B, Rocque BG, Joseph DB, et al. The development of a lifetime care model in comprehensive spina bifida care. *J Pediatr Rehabil Med*. 2018;11(4):323-334.
18. Alford EN, Hopson BD, Safyanov F, et al. Care management and contemporary challenges in spina bifida: a practice preference survey of the American Society of Pediatric Neurosurgeons. *J Neurosurg Pediatr*. 2019;24(5):539-548.
19. Adzick NS, Thom EA, Spong CY, et al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. *N Engl J Med*. 2011;364(11):993-1004.
20. McCarthy DJ, Sheinberg DL, Luther E, McCrea HJ. Myelomeningocele-associated hydrocephalus: nationwide analysis and systematic review. *Neurosurg Focus*. 2019;47(4):E5.
21. Warf BC, Campbell JW. Combined endoscopic third ventriculostomy and choroid plexus cauterization as primary treatment of hydrocephalus for infants with myelomeningocele: long-term results of a prospective intent-to-treat study in 115 East African infants. *J Neurosurg Pediatr*. 2008;2(5): 310-316.
22. Adebayo BO, Kanu OO, Bankole OB, Ojo OA, Adetunmbi B, Morgan E. Early outcome of endoscopic third ventriculostomy with choroid plexus cauterization versus ventriculoperitoneal shunt as primary treatment of hydrocephalus in children with myelomeningocele: a prospective cohort study. *Oper Neurosurg (Hagerstown)*. 2021;21(6):461-466.
23. Talamonti G, Nichelatti M, Picano M, Marcati E, D'Aliberti G, Cenzato M. Endoscopic third ventriculostomy in cases of ventriculoperitoneal shunt malfunction: does shunt duration play a role? *World Neurosurg*. 2019;127:e799-e808.
24. Furtado LMF, da Costa Val Filho JA, Holliday JB, et al. Endoscopic third ventriculostomy in patients with myelomeningocele after shunt failure. *Childs Nerv Syst*. 2020;36(12): 3047-3052.
25. McDowell MM, Lee PS, Foster KA, Greene S. The use of external ventricular drainage to reduce the frequency of wound complications in myelomeningocele closure. *Pediatr Neurosurg*. 2018;53(2):100-107.
26. Radmanesh F, Nejat F, El Khashab M, Ghodsi SM, Ardebili HE. Shunt complications in children with myelomeningocele: effect of timing of shunt placement. Clinical article. *J Neurosurg Pediatr*. 2009;3(6):516-520.
27. Finnegan R, Kehoe J, McMahon O, et al. Primary external ventricular drains in the management of open myelomeningocele repairs in the neonatal setting in Ireland. *Ir Med J*. 2019;112(5):930.
28. White MD, McDowell MM, Agarwal N, Greene S. Shunt infection and malfunction in patients with myelomeningocele. *J Neurosurg Pediatr*. 2021;27(5):518-524.
29. Paulsen AH, Lundar T, Lindegaard KF. Pediatric hydrocephalus: 40-year outcomes in 128 hydrocephalic patients treated with shunts during childhood. Assessment of surgical outcome, work participation, and health-related quality of life. *J Neurosurg Pediatr*. 2015;16(6):633-641.
30. Dupepe EB, Hopson B, Johnston JM, et al. Rate of shunt revision as a function of age in patients with shunted hydrocephalus due to myelomeningocele. *Neurosurg Focus*. 2016; 41(5):E6.
31. Dias MS, Wang M, Rizk EB, et al. Tethered spinal cord among individuals with myelomeningocele: an analysis of the National Spina Bifida Patient Registry. *J Neurosurg Pediatr*. 2021;28(1):21-21.
32. Mummareddy N, Dewan MC, Mercier MR, Naftel RP, Wellons JC III, Bonfield CM. Scoliosis in myelomeningocele: epidemiology, management, and functional outcome. *J Neurosurg Pediatr*. 2017;20(1):99-108.
33. Szymanski KM, Cain MP, Hardacker TJ, Misseri R. How successful is the transition to adult urology care in spina bifida? A single center 7-year experience. *J Pediatr Urol*. 2017; 13(1):40.e1-40.e6.
34. Pandey A, Gupta V, Singh SP, Kumar V, Verma R. Neuropathic ulcers among children with neural tube defects: a review of literature. *Ostomy Wound Manage*. 2015;61(12): 32-38.
35. Routh JC, Joseph DB, Liu T, et al. Variation in surgical management of neurogenic bowel among centers participating in National Spina Bifida Patient Registry. *J Pediatr Rehabil Med*. 2017;10(3-4):303-312.
36. Shepard CL, Yan PL, Hollingsworth JM, Kraft KH. Pregnancy among mothers with spina bifida. *J Pediatr Urol*. 2018; 14(1):11.e1-11.e6.
37. Shepard CL, Yan PL, Kielb SJ, et al. Complications of delivery among mothers with spina bifida. *Urology*. 2019;123: 280-286.

Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: M Nonaka, Yabe, Zaitsu. Acquisition of data: M Nonaka, Yabe, Nakashima. Analysis and interpretation of data: M Nonaka, Isozaki, Komori. Drafting the article: M Nonaka. Critically revising the article: M Nonaka, Asai. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: M Nonaka. Statistical analysis: M Nonaka, Zaitsu. Administrative/technical/material support: all authors. Study supervision: M Nonaka, Nakashima, Asai.

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