Adults with myelomeningocele and other forms of spinal dysraphism: hospital care in the United States since the turn of the millennium

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OBJECTIVE The natural history and management of myelomeningocele (MM) in children is fairly well understood. There is a deficiency of knowledge regarding the care of adults, however, even though there are now more adults than children living with MM. The purpose of this study was to characterize the hospital care of adults with MM and hydrocephalus on a nationwide population base. Adults with other forms of spina bifida (SB) were studied for contrast.

METHODS The Nationwide Inpatient Sample for the years 2001, 2004, 2007, and 2010 was queried for admissions with diagnostic ICD-9-CM codes for MM with hydrocephalus and for other forms of SB.

RESULTS There were 4657 admissions of patients with MM and 12,369 admissions of patients with SB in the sample. Nationwide rates of admission increased steadily for both MM and SB patients throughout the study period. Hospital charges increased faster than the health care component of the Consumer Price Index. Patients with MM were younger than patients with SB, but annual admissions of MM patients older than 40 years increased significantly during the study period. With respect to hospital death and discharge home, outcomes of surgery for hydrocephalus were superior at high-volume hospitals. Patients with MM and SB were admitted to the hospital more frequently than the general population for surgery to treat degenerative spine disease.

CONCLUSIONS Patients with MM and SB continue to require neurosurgical attention in adulthood, and the demand for services for older patients with MM is increasing. Management of hydrocephalus at high-volume centers is advantageous for this population. Patients with MM or SB may experience high rates of degenerative spine disease.

http://thejns.org/doi/abs/10.3171/2015.9.SPINE15771

KEY WORDS adults; hydrocephalus; myelomeningocele; Nationwide Inpatient Sample; spina bifida; congenital
Methods

Data were extracted from the Nationwide Inpatient Sample (NIS) for the years 2001, 2004, 2007, and 2010. The NIS is an administrative database produced by the Healthcare Utilization Project (HCUP), a joint project of state agencies, industry, and the Agency for Healthcare Research and Quality (AHRQ). The NIS contains data from 20% of discharges from a scientific sample of non-governmental, acute care hospitals in the United States. The sampling is stratified, and the data are weighted to allow estimation of various parameters on a nationwide or regional basis or on the basis of hospital size, hospital location, or hospital teaching status. In this paper, “sample” refers to analysis of unweighted data, and “estimate” refers to analysis of weighted data.

In the 4 study years, the NIS contained up to 25 fields for ICD-9-CM diagnostic codes. Admissions of patients with MM with hydrocephalus were identified by diagnostic code 741.0x. In the following analysis “myelomeningocele” (MM) refers to this group of patients. Admissions of patients with other forms of SB were identified by diagnostic codes 741.9x, 742.51, and 742.59. The term “spina bifida” (SB) is used for these patients. Admissions coded both for MM with hydrocephalus and for another form of SB were analyzed with the MM group. Admissions of patients younger than 18 years were excluded. In addition to ICD-9 codes, the NIS includes Clinical Classifications Software (CCS) codes. CCS is a tool developed by AHRQ to condense the entire set of ICD-9-CM diagnostic and procedural codes into “a manageable number of clinically meaningful categories” (https://www.hcup-us.ahrq.gov/toolssoftware/ccs/CCSUersGuide.pdf). CCS codes were used to characterize reasons for admission and principal procedures and to identify complications of treatment. Hospital-acquired conditions (HACs) were identified with reference to the definitions of the Centers for Medicare & Medicaid Services (https://www.cms.gov/Medicare/Medicare-Fee-for-Service-HospitalAcqCond/index.html).

The NIS contains 15 fields for ICD-9-CM procedural codes. Five categories of neurosurgical admissions were defined as follows. Surgery for hydrocephalus included shunt insertion, shunt revision, shunt removal, ventricullocisternostomy, and external ventricular drainage (02.2, 02.31–02.35, 02.39, 02.41–02.43, and 54.95). Surgery for degenerative spine disease included cervical and lumbar disc surgery, cervical fusions excluding atlantoaxial fusion and lumbarosacral fusions (03.02, 03.09, 80.50–80.52, 80.59, 81.02, 81.03, 81.06–81.08, 81.32, 81.33, 81.36–81.38, 84.58, and 84.60–84.64). Surgery for spinal cord injuries included surgery for tethering and for syringomyelia (03.4, 03.51, 03.52, 03.59, 03.6, 03.7, 03.71, 03.72, 03.79, 03.97, and 03.98). Cross-tabulation demonstrated substantial overlap between degenerative spine surgery and spinal cord surgery, which proved to be due to supplemental coding for laminectomy for many cases of spinal cord surgery. Therefore, admissions coded for spinal cord surgery and for laminectomy were removed from the degenerative spine surgery category. Surgery for spinal deformity included thoracolumbar fusions (81.04, 81.05, 81.34, and 81.35). Surgery for pain or spasticity included implantation and revision of pumps and spinal cord stimulators (03.90, 03.93, 03.94, 86.06, and 86.94–86.98).

Hospital charges were analyzed in 2010 US dollars, adjusted for inflation using the health care component of the Consumer Price Index (CPI) (http://data.bls.gov/timeseries/CUUR000SAM?output_view=pct_12mths). Population estimates were taken from the website of the US Census (http://factfinder.census.gov/faces/tableservices/jsf/pages/productview.xhtml?pid=DEC_10_PL_P1&prodType=table).

Microsoft Excel 2007 and SPSS version 22 were used for data organization and for hypothesis testing. Variances for nationwide estimates were calculated using the code provided for SAS (version 9.4, SAS Institute) by the HCUP. Hypothesis testing of categorical data was performed using the Pearson chi-square test or Fisher’s exact test. Testing of quantitative sample data was performed using the Mann-Whitney U-test. A simple Z-test was used to compare estimated counts and means. Where p values have not been specified, denotation of significance implies α = 0.05.

This study was judged not to be human subjects research by the Nemours Institutional Review Board.

Results

There were 4657 admissions of patients with MM and 12,369 admissions of patients with SB in the sample. Table 1 presents the estimated nationwide counts of admissions and total charges for each year of the study in 2010 dollars. Each of these statistics demonstrated a significant increasing trend over the study period. Total charges and charges per admission increased more rapidly than the health care component of the CPI. Charges per admission tended to be greater for patients with MM than for patients with SB.

Based on data from the 2010 US Census, population-based regional admission rates were calculated (Table 2). Rates tended to be lower in the West and Northeast and higher in the Midwest and South. There was no apparent seasonal variation in admission rates for either MM or SB (data not shown).

The 10 most common reasons for sampled admission and the 10 most common principal procedures are listed in Table 3. This list reflects the burden of complications of hydrocephalus, paraplegia, and neurogenic bladder disturbance, and the measures needed to manage them, but degenerative spine disease is prominent as well. Degenerative spine disease was examined more closely for 2010. The CCS diagnostic category “spondylosis; intervertebral disc disorders; other back problems” accounted for 2.4% of principal diagnoses for the sampled admissions of MM and SB patients but only 2.0% of the entire 2010 sample (p = 0.0507). The CCS procedural category “laminectomy; excision of intervertebral disc” accounted for 2.1% of principal procedures for the sampled admissions of MM and SB patients but only 0.6% of the entire 2010 sample (p = 0.0005).

Comparison of MM and Other Forms of SB

Patients with MM were younger overall than patients with SB. The mean ages of patients with MM and SB in
the sample data were 38 and 43 years, respectively (p < 0.0005). As Fig. 1 indicates, there were relatively few admissions of patients with MM in the 5th and later decades of life. Patients older than 40 years accounted for 47% of sampled SB admissions but only 28% of MM admissions (p < 0.0005). Estimated nationwide counts of admissions of MM patients older than 40 years in each year of the current study are depicted in Fig. 2. A significant increasing trend was observed over the 10 years of this study.

Overall female predominance was more pronounced among sampled patients with MM than sampled patients with SB (62% and 56%, respectively [p < 0.0005]). The difference in emergency admission rates for patients with MM and SB in the sample data attained statistical significance because of the large numbers of patients, but it was slight (51% compared with 49%). Among the 29 co-morbidities tabulated by the AHRQ, the following were significantly more common after Bonferroni correction in sampled admissions for SB patients: alcohol abuse; deficiency anemia; diabetes, uncomplicated; drug abuse; hypertension; hypothyroidism; liver disease; peripheral vascular disorders; chronic renal failure; and solid tumors. The predominance of many of these comorbidities is consistent with the older age of SB patients at admission. Rates of depression, obesity, and psychosis were identical among MM and SB patients. The mean number of comorbidities per sampled admission was greater among patients with SB than among those with MM, but the difference was small (1.6 compared with 1.4 [p < 0.0005]). Sampled admissions of patients with MM had more CCS codes for complications of treatment (which included CSF shunt failure and infection) than admissions of patients with

<table>
<thead>
<tr>
<th>Condition</th>
<th>Year</th>
<th>Admissions (SD)†</th>
<th>Total Charges (SD)†</th>
<th>Charges per Admission (SE)‡</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myelomeningocele</td>
<td>2001</td>
<td>4069 (355)</td>
<td>$152 M (16 M)</td>
<td>$37,560 (2178)</td>
</tr>
<tr>
<td></td>
<td>2004</td>
<td>5203 (393)</td>
<td>$192 M (23 M)</td>
<td>$37,404 (2931)</td>
</tr>
<tr>
<td></td>
<td>2007</td>
<td>6645 (397)</td>
<td>$287 M (28 M)</td>
<td>$45,082 (2753)</td>
</tr>
<tr>
<td></td>
<td>2010</td>
<td>7510 (517)</td>
<td>$350 M (34 M)</td>
<td>$47,641 (3015)</td>
</tr>
<tr>
<td>Other spina bifida</td>
<td>2001</td>
<td>11,435 (522)</td>
<td>$315 M (20 M)</td>
<td>$28,022 (1030)</td>
</tr>
<tr>
<td></td>
<td>2004</td>
<td>13,490 (556)</td>
<td>$434 M (25 M)</td>
<td>$32,617 (1149)</td>
</tr>
<tr>
<td></td>
<td>2007</td>
<td>17,987 (1722)</td>
<td>$720 M (116 M)</td>
<td>$40,458 (2921)</td>
</tr>
<tr>
<td></td>
<td>2010</td>
<td>18,777 (735)</td>
<td>$758 M (43 M)</td>
<td>$40,892 (1419)</td>
</tr>
</tbody>
</table>

M = million.
* Entries that do not share superscripts are statistically significantly different after column-wise Bonferroni correction. Increasing trends for admissions, total charges, and charges per admission were all highly significant.
† Mean (SD).
‡ Mean (SE).

<table>
<thead>
<tr>
<th>Region</th>
<th>Myelomeningocele</th>
<th>Other Spina Bifida</th>
</tr>
</thead>
<tbody>
<tr>
<td>Northeast</td>
<td>24.2 (4.8)a</td>
<td>61.8 (6.1)b</td>
</tr>
<tr>
<td>Midwest</td>
<td>27.0 (4.0)a</td>
<td>65.9 (6.4)b</td>
</tr>
<tr>
<td>South</td>
<td>25.4 (2.3)a</td>
<td>67.0 (3.8)a</td>
</tr>
<tr>
<td>West</td>
<td>21.7 (3.6)a</td>
<td>49.0 (4.2)a</td>
</tr>
</tbody>
</table>

* Entries that do not share superscripts are statistically significantly different after column-wise Bonferroni correction. Values are presented as the mean ± SD.
SB (27% compared with 16% [p < 0.0005]). Discharge to home was slightly more common among sampled patients with MM than those with SB (65% compared with 61% [p < 0.0005]), but there was no difference in hospital mortality rates (1.5%). Seventy-two admissions of patients with MM in the sample ended in death. Principal diagnoses for these admissions were infectious in 29% (predominantly septicemia but including urinary tract infection, osteomyelitis, and chronic skin ulcer), pulmonary in 25%, and central nervous system conditions in 24% (predominantly “complications of device; graft or implant”). There were 2 deaths attributed to complications of pregnancy.

Neurosurgical Admissions and Admissions for Other Indications

The estimated numbers of admissions for surgery for selected neurosurgical indications are presented in Table 4. As anticipated from the choice of diagnostic codes, there were many more admissions for surgery for hydrocephalus in the setting of MM than in SB. Figure 3 depicts the age distributions of patients with MM admitted for hydrocephalus and for other indications. The median age of sampled patients admitted for surgery for hydrocephalus was 25 years, much younger than the median age of the remaining patients in the MM cohort at 32 years (p < 0.0005). Admissions of MM patients older than 40 years for hydrocephalus were few in number, but they trended upward in the last 2 years of this study. Estimated nationwide counts are depicted in Fig. 4.

Admissions for surgery for degenerative spine disease were numerous in both groups. Sampled patients with MM admitted for surgery for degenerative spine disease were younger than patients with SB admitted for the same purpose, with median ages of 36 and 43 years, respectively (p < 0.0005). A comparison of Tables 1 and 4 indicates that between 3.2% and 6.1% of all admissions of MM and SB patients were coded for surgery for degenerative spine disease. The corresponding statistic for estimated nationwide admissions of patients 18 years or older in 2010 was 2.0% (p < 0.0005 for each year for both MM and SB). Surgery for degenerative spine disease accounts for a larger fraction of hospital admissions of patients with MM and SB than of the general population.

Annual estimated nationwide admission rates for neurosurgical indications seemed stable over the years of this

FIG. 1. Ages at admission of sampled patients with SB (left) and patients with MM (right). There were fewer admissions of patients older than 40 years in the MM group than in the SB group (p < 0.0005). Figure is available in color online only.

FIG. 2. Estimated counts of admissions of MM patients older than 40 years. Error bars represent standard deviations. Counts that do not share superscripts are significantly different after Bonferroni correction. Figure is available in color online only.
study with 2 exceptions: admissions among patients with SB for surgery for degenerative spine disease and spinal cord conditions trended upward over the study period. Close inspection of Tables 1 and 4 discloses that the variances for SB admissions in 2007 were much higher than in other years and much higher than the corresponding statistics for MM admissions. This aberration is attributable to reporting of very large numbers from a single outlying institution. The HCUP Data Use Agreement precludes further investigation. Admissions for surgery for hydrocephalus among patients with MM were flat in 2001 and 2004 but then trended upward in 2007 and 2010. Admissions for spinal deformity surgery and for surgery for pain or spasticity were few in number in both groups, and no observations regarding trends can be supported.

Estimated nationwide admissions for neurosurgical procedures and for other reasons are compared in Table 5 with respect to length of stay. Neurosurgical admissions for patients with MM were longer than nonneurosurgical admissions and longer than neurosurgical admissions for patients with SB.

**TABLE 4. Estimated nationwide annual admissions for various neurosurgical indications***

<table>
<thead>
<tr>
<th>Condition &amp; Surgical Indication</th>
<th>2001</th>
<th>2004</th>
<th>2007</th>
<th>2010</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myelomeningocele</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hydrocephalus</td>
<td>717 (93)</td>
<td>711 (101)</td>
<td>870 (121)</td>
<td>1084 (155)</td>
</tr>
<tr>
<td>Degenerative spine disease</td>
<td>252 (54)</td>
<td>234 (63)</td>
<td>298 (45)</td>
<td>263 (43)</td>
</tr>
<tr>
<td>Spinal cord conditions</td>
<td>105 (22)</td>
<td>147 (27)</td>
<td>148 (23)</td>
<td>145 (34)</td>
</tr>
<tr>
<td>Spinal deformity</td>
<td>—</td>
<td>9 (—)</td>
<td>5 (—)</td>
<td>35 (10)</td>
</tr>
<tr>
<td>Pain or spasticity</td>
<td>—</td>
<td>27 (10)</td>
<td>43 (8)</td>
<td>32 (10)</td>
</tr>
<tr>
<td>Other spina bifida</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hydrocephalus</td>
<td>98 (23)</td>
<td>120 (21)</td>
<td>170 (37)</td>
<td>136 (28)</td>
</tr>
<tr>
<td>Degenerative spine disease</td>
<td>455 (91)</td>
<td>437 (87)</td>
<td>886 (368)</td>
<td>691 (72)</td>
</tr>
<tr>
<td>Spinal cord conditions</td>
<td>481 (49)*</td>
<td>537 (65)*</td>
<td>1618 (1053)*</td>
<td>1046 (115)*</td>
</tr>
<tr>
<td>Spinal deformity</td>
<td>15 (9)</td>
<td>14 (—)</td>
<td>34 (—)</td>
<td>64 (5)</td>
</tr>
<tr>
<td>Pain or spasticity</td>
<td>41 (14)</td>
<td>57 (9)</td>
<td>44 (9)</td>
<td>78 (13)</td>
</tr>
</tbody>
</table>

* Entries that do not share superscripts are statistically significantly different after row-wise Bonferroni correction. Where there are no superscripts there are no significant row-wise differences. The only significant temporal trend was increasing admissions for surgery for spinal cord conditions among patients with SB in 2010 compared with previous study years. Values are presented as the mean (SD).

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**FIG. 3.** Ages of sampled patients with MM admitted for surgery for all other indications (left) and for hydrocephalus (right). There were fewer admissions of patients older than 40 years for hydrocephalus than for other indications (p < 0.0005). Figure is available in color online only.
for patients with SB, although after Bonferroni correction none of these differences reached significance. Protracted admissions for complications of CSF shunts may account for lengthier stays for neurosurgical MM patients. Sampled neurological admissions were much more common for patients with MM encountered more HACs than patients with SB (5.8% and 1.1%, respectively [p = 0.001]). On the other hand, sample neurological admissions were much more commonly coded for complications of treatment than nonneurological admissions (42% compared with 16% [p < 0.0005]). Discharge to home was significantly more common for sampled neurological admissions than for nonneurological admissions (77% compared with 59% [p < 0.0005]). Hospital mortality rates were also significantly lower for sampled neurological admissions than for nonneurological admissions (77% compared with 59% [p < 0.0005]). The mean numbers of comorbidities were lower for sampled neurological admissions than for nonneurological admissions (1.0 compared with 2.0 [p < 0.0005]). Among sampled neurological admissions, patients with MM encountered more HACs than patients with SB (5.8% and 1.1%, respectively [p = 0.001]). On the other hand, sample neurological admissions were much more commonly coded for complications of treatment than nonneurological admissions (42% compared with 16% [p < 0.0005]). Discharge to home was significantly more common for sampled neurological admissions than for nonneurological admissions (77% compared with 59% [p < 0.0005]). Hospital mortality rates were also significantly lower for sampled neurological admissions than for sampled nonneurological admissions (0.9% compared with 1.5% [p < 0.0005]).

Teaching hospitals were the venue for 82% of sampled neurological admissions but only 52% of nonneurological admissions (p < 0.0005). Sampled patients with MM underwent neurological procedures at teaching hospitals at slightly but statistically significantly higher rates than patients with SB (85% and 79%, respectively [p < 0.0005]). Mortality rates for sampled neurological admissions at teaching and nonteaching hospitals were nearly identical (0.87% and 0.97%, respectively [p = 0.8503]), as were rates of discharge home (78% and 77%, respectively [p = 0.8970]). Sampled neurological admissions at teaching hospitals were much more commonly coded for complications than admissions at nonteaching hospitals (45% and 28%, respectively [p < 0.0005]). Likewise, for the year 2010, 3.9% of neurological admissions to teaching hospitals were coded for an HAC, while the rate at nonteaching hospitals was only 0.7% (continuity adjusted p = 0.0993). Neurosurgical patients at teaching and nonteaching hospitals both had a median of 1 comorbid condition (p = 0.6270), and patients admitted to teaching hospitals were actually younger than patients admitted to nonteaching hospitals, with median ages of 34 and 40 years, respectively (p < 0.0005). At teaching hospitals the proportion of admissions for surgery for hydrocephalus was larger than for admissions for other neurological procedures (88% of cases compared with 78% [p < 0.0005]), and admissions for hydrocephalus accounted for 71% of all neurological admissions coded for any complication (p < 0.0005).

Of 773 sampled admissions for surgery for hydrocephalus, 675 (87%) carried a CCS code for a medical or surgical complication, as anticipated from the definition of surgical complication. The 13 busiest hospitals, which together accounted for 25% of sampled admissions for hydrocephalus, were designated “high volume” and the other hospitals were designated “low volume.” The median age of sampled patients admitted for surgery for hydrocephalus was 24 at high-volume hospitals and 28 at low-volume hospitals (p < 0.0005). Perhaps reflecting younger patient ages, the median number of comorbid conditions among sampled admissions at high-volume centers was 0, and the median at low-volume centers was 1 (p < 0.0005). The complication rate among admissions at sampled high-volume hospitals was 92.4% compared with 85.5% among admissions to low-volume hospitals (p = 0.0129). The reverse association held for HACs. In 2010 there were no HACs at sampled high-volume hospitals, while 9.2% of admissions at low-volume hospitals were coded for HACs (p = 0.0076). There were no deaths at high-volume hospitals, while 19 (3.3%) of 575 sampled admissions for surgery for

### Table 5. Mean lengths of stay (in days) for admissions coded for neurological procedures and for other admissions

<table>
<thead>
<tr>
<th>Condition &amp; Purpose of Admission</th>
<th>2001</th>
<th>2004</th>
<th>2007</th>
<th>2010</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myelomeningoele</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neurosurgical</td>
<td>8.62 (1.22)</td>
<td>9.88 (1.10)</td>
<td>8.65 (0.93)</td>
<td>7.49 (0.91)</td>
</tr>
<tr>
<td>Nonneurosurgical</td>
<td>7.88 (0.53)</td>
<td>6.34 (0.29)</td>
<td>6.50 (0.34)</td>
<td>6.77 (0.28)</td>
</tr>
<tr>
<td>Other spina bifida</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neurosurgical</td>
<td>6.63 (0.66)</td>
<td>6.59 (0.61)</td>
<td>6.30 (0.47)</td>
<td>5.13 (0.29)</td>
</tr>
<tr>
<td>Nonneurosurgical</td>
<td>6.91 (0.23)</td>
<td>6.81 (0.28)</td>
<td>6.70 (0.21)</td>
<td>6.55 (0.20)</td>
</tr>
</tbody>
</table>

* There were no significant temporal trends. Neurosurgical admissions for patients with MM were longer than those for patients with SB. Values are presented as the mean (SE).
hydrocephalus at low-volume hospitals ended in death (p = 0.0058). Sampled admissions to high-volume hospitals ended in discharge home in 86% of instances, while only 73% of admissions at low-volume hospitals ended in discharge home (p = 0.0004).

Discussion

Admissions analyzed for this study were identified on the basis of ICD-9-CM diagnostic codes. As is true of most administrative data sets, the accuracy of the coding is unknown. The intention was to identify admissions of adults with MM and hydrocephalus—the MM group (codes 741.0x)—and to compare them with admissions of adults with other forms of spinal dysraphism (the SB group). The MM group must be presumed to include both patients with continuing, active hydrocephalus who are at risk for symptomatic CSF shunt failure and patients with inactive or “compensated” hydrocephalus who never had shunts or whose shunts have failed without symptoms. Patients admitted for surgery for hydrocephalus during the study period must be among the former, but otherwise no distinction can be made between these possibilities in the NIS data. The SB group includes cases of lipomyelo-meningocele, filum lipoma and other spinal lipomas, and diastematomyelia in addition to patients with MM who never had hydrocephalus (codes 741.9x).

Numbers of admissions and charges per admission after adjustment for inflation increased steadily during the study period. The latter observation is not difficult to understand. The NIS captures hospital charges, but the health care component of the CPI is based on payments. Thus, what the current study shows is that hospital charges increased more rapidly than overall payments in the health care sector of the economy. An explanation that suggests itself is that hospitals have inflated charges to defend revenue streams against downward pressure on payments from insurers. This phenomenon is likely system wide and unrelated to the specific conditions examined in the current study.

The increase in numbers of admissions is more difficult to understand. Epidemiological considerations suggest that the total population of adults with MM is increasing, but the same rate of growth was seen for admissions of adults with SB. Both categories of admissions increased substantially faster than the overall population of the United States. As Tables 1 and 4 together indicate, only a small and relatively constant proportion of admissions was coded for neurosurgical procedures, so changes in neurological practices cannot be a major factor. Not all numbers increased; admissions for surgery for degenerative spine disease and for other spinal cord conditions were stable over the course of the study period for adults with MM. This observation weighs against the possibility of a systematic distortion arising from the sampling algorithm. The trends observed in this study probably reflect larger trends in the hospital care of adults with chronic conditions, explanations for which are beyond the scope of this report.

There are few actuarial survival data for adults with MM and hydrocephalus, but the evidence indicates that these patients die from various causes at higher rates than the general population in adult life as well as in childhood. Bowman et al. reported outcomes in early adulthood for the first cohort of patients with MM treated nonselectively and aggressively at Children’s Memorial Hospital in Chicago under the leadership of David McLone.3,18 Survival to adulthood was estimated at 75%. Among patients in follow-up, 85% had CSF shunts, and this cohort continued to be depleted in young adulthood by sudden death due to shunt failure. Davis et al. reported survival data from a very large clinic population at the University of Washington.7 Among patients born after 1975, the presence of a CSF shunt was not associated adversely with survival to age 16 years, but having reached a 16th birthday, patients with shunts were significantly less likely than patients without shunts to survive to 30 years of age. This discrepancy suggests a failure of transition to neurosurgical services in adult care settings. In 2010 Oakeshott et al. reported 40-year outcomes of a remarkable longitudinal study of nonselectively treated infants with MM initiated in the 1960s at Addenbrooke’s Hospital in Cambridge by Gillian Hunt.13,20 Survival to 5 years of age was 66%. Forty percent of patients who reached a 5th birthday were dead before 40 years of age. Half of these later deaths were unexpected and occurred out of hospital. Besides CSF shunt failure, late deaths were attributed to sepsis, epilepsy, and pulmonary embolism.

Nulsen and Spitz reported the first modern CSF shunt for hydrocephalus in 1951,19 but some years passed before improvements in valve technology and dissemination of surgical practices made CSF shunts widely available and created the first cohort of surviving infants with MM. The Addenbrooke’s Hospital study, for instance, was begun in the 1960s. Infants born with MM in the early 1960s entered their 5th decades during the years of the current study. If this cohort is surviving in substantial numbers down to the present, its leading edge ought to have been manifest as an upward trend in admissions of patients older than 40 years during the years of this study. This trend was indeed evident—highly significant for overall admissions (Fig. 2) and credible, at least, for admissions for hydrocephalus (Fig. 4). Numbers of admissions were much smaller for patients older than 50 years, and no leading edge was detected for this cohort (data not shown). These observations are consistent with the concept of a wavelet of aging survivors at an early stage in its passage through the health care system. The neurosurgical needs of this cohort can be anticipated to increase. For the purposes of this study, the definition of a neurosurgical admission was restricted to neurosurgical procedures that were indicated by the study conditions, MM and SB, and their complications. Craniootomy for brain tumor, for example, was not considered. With respect to hospital death and discharge home, neurological patients fared no better at teaching hospitals than at nonteaching hospitals, and with respect to complications they fared considerably worse, even though patients at teaching hospitals were younger. This paradox is explained by the concentration of surgery for hydrocephalus at teaching hospitals.

The association of higher clinical volume with better outcomes has been established for a number of com-

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mon neurosurgical conditions, including hydrocephalus. The association seems to hold for surgery for hydrocephalus among adults with MM and SB in particular. HACs and deaths were recorded exclusively at low-volume hospitals, and discharge to home was not as frequent as from high-volume institutions. Death and discharge to home are coarse but important measures of quality in the treatment of hydrocephalus. Clinicians who supervise the transition of patients with MM and hydrocephalus to adult care settings must consider this association between volume and quality.

Surgery for degenerative spine disease seems to account for a larger proportion of hospital admissions of patients with MM and SB than for the general population. Abnormal mechanics of posture and gait may accelerate degenerative changes in this population. Alternatively, patients with MM and SB are under continuing neurosurgical observation for their congenital conditions and may thereby have better access to neurological services than the general population. The younger distribution of ages of MM patients admitted for surgery for degenerative spine conditions may relate to a higher prevalence of paraplegia and scoliosis in this population than among patients with SB, or it may reflect limited survival into later adult life. The literature concerning degenerative spine disease in the MM and SB population is extremely limited. To what degree coexisting hindbrain herniation, syringomyelia, or spinal cord tethering have implications for the surgical management of degenerative disease is unknown, but anecdotal evidence encourages further investigation of this question.

The problematic reliability of clinical data extracted from administrative data sets is widely recognized. The current study provided an unexpected illustration: Diagnoses corresponding to the definitions of HACs are essentially absent from the sampled discharge data from 2001, 2004, and 2007. They appear in substantial numbers only in the data from 2010. The Centers for Medicare & Medicaid Services began restricting payments for treatment of HACs in 2008. Payment considerations clearly sensitized hospital discharge coders to HAC-related diagnoses that they had neglected in earlier years.

The current study provides support for regionalization of surgery for hydrocephalus at high-volume centers among patients with MM and SB, and the other neurosurgical needs of this population are likely best served at busy centers as well. The multidisciplinary clinic is the obvious mechanism for concentrating patients at sites with high levels of expertise. Historically, SB clinics have been the major source of knowledge about the care of children with MM, and the multidisciplinary clinic is the logical venue for quality improvement in the care of adults as well. Neurosurgeons have an obligation to promote multidisciplinary clinics dedicated to the care of adult patients with MM.

Conclusions

Patients with MM and SB continue to require neurosurgical attention in adulthood, and the demand for services for older patients with MM is increasing. Management of hydrocephalus at high-volume centers is advantageous for this population. Adult patients with MM and SB may experience high rates of degenerative spine disease and may benefit from increased availability of multidisciplinary clinics such as have been widely available for the care of children.

Acknowledgments

I thank J. Sandford Schwartz, MD, for helpful comments.

References


Disclosures
The author reports no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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