**Online Appendix**

**Table 1**. Population characteristics.

|  |  |  |  |
| --- | --- | --- | --- |
| **Patient characteristic** | Shunted (n=261) | Unshunted (n=32) | Overall (n=293) |
| Male gender (%) | 129 (49%) | 18 (56%) | 147 (50%) |
| Community ambulators (%) | 27 (10%) | 5 (16%) | 32 (11%) |
|  Unknown | 24 (9%) | 3 (9%) | 27 (9%) |
|  Community ambulators only among those who died after four years old (n=228) | 27 (12%) | 5 (56%) | 32 (14%) |
| Developmental delay (%) | 88 (34%) | 7 (22%) | 95 (32%) |
|  Unknown | 53 (20%) | 3 (9%) | 56 (19%) |
| History of epilepsy (%) | 88 (34%) | 4 (13%) | 92 (31%) |
|  Unknown | 49 (19%) | 1 (3%) | 50 (17%) |
|  History of epilepsy only among those who died after one year old (n=258) | 85 (35%) | 2 (13%) | 87 (34%) |
| Enrolled in National Spina Bifida Patient Registry (%) | 46 (18%) | 4 (13%) | 50(17%) |
| Median year of birth (IQR) | 1990 (1983-2000) | 2009 (1999-2015) | 1991 (1983-2004) |
|  |  |  |  |
| **Mortality data** |  |  |  |
| Median year of death (IQR) | 2012 (2007-2016) | 2014 (2010-2017) | 2012 (2007-2016) |
| Median age at death (years, IQR) | 20 (9-27) | <1 (<1-10) | 19 (5-27) |
| Infants (<1 year old) | 18 (7%) | 17 (53%) | 35 (12%) |
| Children (1-17.9 years old) | 94 (36%) | 8 (25%) | 102 (35%) |
| Adults (18 years old or older) | 149 (57%) | 7 (22%) | 156 (53%) |
| Location where death occurred (%) |  |  |  |
|  Hospital with multidisciplinary MMC care | 101 (39%) | 21 (66%) | 122 (42%) |
|  Home/group home | 39 (15%) | 7 (22%) | 46 (16%) |
|  Hospital without multidisciplinary MMC care | 34 (13%) | 1 (3%) | 35 (12%) |
|  Hospice | 11 (4%) | 0 (0%) | 11 (4%) |
|  Long-term care/nursing home | 2 (1%) | 0 (0%) | 2 (1%) |
|  Other (work, roadway) | 2 (1%) | 0 (0%) | 2 (1%) |
|  Unknown | 72 (28%) | 3 (9%) | 75 (26%) |
| Surgery within last 30 days of life (%) | 62 (24%) |  8(25%) | 70 (24%) |
|  Unknown | 87 (33%) | 4 (13 | 91 (31%) |
| Source of death data (%) |  |  |  |
|  Medical records | 238 (91%) | 31 (97%) | 269 (92%) |
|  Institutional databases | 29 (11%) | 2 (6%) | 31 (11%) |
|  Death certificate | 26 (10%) | 2 (6%) | 28 (10%) |
|  Parent or caregiver | 24 (9%) | 3 (9%) | 27 (9%) |
|  Primary care provider or other healthcare professional | 21 (8%) | 0 (0.0%) | 21 (7%) |
|  Obituary or other public announcement | 14 (5%) | 1 (3%) | 15 (5%) |
|  State or provincial database | 2 (1%) | 0 (0%) | 2 (1%) |
| Autopsy performed (%) | 23 (9%) | 7 (22%) | 30 (10%) |
|  Unknown | 124 (48%) | 9 (28%) | 133 (45%) |
|  |  |  |  |
| **Neurosurgical management** |  |  |  |
| Documented hydrocephalus (%) | 261 (100%) | 20 (63%) | 281 (96%) |
| Endoscopic third ventriculostomy (%) | 17 (7%) | 3 (15%) | 20 (8%) |
| Shunt revision (%) | 180 (71%) | n/a | 180 (61%) |
|  Unknown  | 27 (11%) | n/a | 27 (9%) |
|  Median shunt revisions (IQR) | 3 (1-5) | n/a | 3 (1-5) |
|  Median years from last revision to death (IQR) | 4 (1-12) | n/a | 4 (1-12) |
| History of spinal cord untethering (%) | 62 (25%) | 1 (3%) | 63 (22%) |
|  Unknown | 65 (25%) | 3 (9%) | 68 (23%) |
|  Median spinal cord untetherings (IQR) | 1 (1-2) | 2, 2 | 1 (1-2) |
|  |  |  |  |
| **Bladder and bowel management** |  |  |  |
| History of incontinent vesicostomy (%) | 46 (18%) | 1 (3%) | 47 (16%) |
|  Unknown  | 29 (11%) | 0 (0%) | 29 (10%) |
| Bladder management at time of death (%) |  |  |  |
|  Incontinent vesicostomy  | 19 (7%) | 1 (3%) | 20 (7%) |
|  Incontinent diversion  | 18 (7%) | 0 (0%) | 18 (6%) |
|  Augmented bladder  | 71 (27%) | 6 (19%) | 77 (26%) |
|  Other | 137 (52%) | 25 (78%) | 162 (55%) |
|  Unknown  | 16 (6%) | 0 (0%) | 16 (5%) |
| Clean intermittent catheterizations  | 166 (64%) | 9 (28%) | 175 (60%) |
|  Per urethra | 100 (60%) | 6 (67%) | 106 (61%) |
|  Per channel  | 66 (40%) | 3 (33%) | 69 (39%) |
|  Still used at time of death  | 56 (85%) | 3 (100%) | 59 (86%) |
| Bowel care at time of death (%) |  |  |  |
|  Colostomy | 22 (8%) | 2 (6%) | 24 (8%) |
|  Malone antegrade continence enema | 53 (20%) | 5 (16%) | 58 (20%) |
|  Still used at time of death | 42 (79%) | 3 (60%) | 45 (78%) |
|  Neither  | 170 (65%) | 25 (78%) | 196 (67%) |
|  Unknown | 16 (6%) | 0 (0%) | 16 (5%) |
|  |  |  |  |
| **Skin and bone care** |  |  |  |
| Any previous surgery for skin ulcers or wound debridement (%) | 86 (33%) | 1 (3%) | 87 (30%) |
| Skin ulcers in the final 12 months of life (%) | 57 (23%) | 1 (3%) | 58 (20%) |
| History of scoliosis (%) | 153 (59%) | 5 (16%) | 158 (54%) |
| Bone fractures in the final 12 months of life (%) | 8 (2%) | 1 (3%) | 9 (3%) |
|  Unknown | 64 (25%) | 1 (3%) | 65 (22%) |
|  |  |  |  |
|  |  |  |  |
| **Baseline pulmonary function (among those who died after age one year old)** | n=243 | n=15 | n=258 |
| BiPap/CPap (%) | 33 (14%) | 4 (27%) | 37 (14%) |
| Supplemental oxygen (%) | 44 (18%) | 3 (20%) | 47 (18%) |
| Trachoestomy (%) | 29 (12%) | 2 (13%) | 31 (12%) |
| None (%) | 111 (46%) | 7 (47%) | 118 (46%) |
| Unknown (%) | 39 (16%) | 2 (13%) | 41 (16%) |
|  |  |  |  |
| **Sleep studies (among those who died after age four years old)** | n=227 | n=9 | n=228 |
| Any sleep study (%) | 53 (24%) | 1 (11%) | 54 (24%) |
| Sleep study in the last year of life (%) | 12 (5%) | 1 (11%) | 13 (6%) |
| Unknown | 61 (28%) | 2 (22%) | 63 (28%) |
|  |  |  |  |
| **Adult health and social characteristics** | n=149 | n=7 | n=156 |
| Obesity (%) | 43 (29%) | 2 (29%) | 45 (29%) |
| Hypertension (%) | 38 (26%) | 2 (29%) | 40 (26%) |
| Chronic kidney disease (%) | 32 (21%) | 2 (29%) | 34 (22%) |
| History of depression (%) | 31 (21%) | 2 (29%) | 33 (21%) |
| History of anxiety (%) | 15 (10%) | 1 (14%) | 16 (10%) |
| History of opioid addiction (%) | 5 (3%) | 1 (14%) | 6 (4%) |
| History of alcohol addiction (%) | 2 (1%) | 1 (14%) | 3 (2%) |
| History of suicidal ideation or attempts (%) | 4 (3%) | 1 (14%) | 5 (3%) |
| Unknown (%) | 25 (17%) | 0 (0%) | 25 (16%) |
| Living arrangement (%) |  |  |  |
|  Alone  | 4 (3%) | 0 (0%) | 4 (3%) |
|  With own family (i.e., with spouse/partner)  | 9 (6%) | 3 (43%) | 12 (8%) |
|  Parents/caregivers  | 65 (44%) | 1 (14%) | 66 (42%) |
|  Group home  | 11 (7%) | 2 (29%) | 13 (8%) |
|  Long-term care facility | 9 (6%) | 0 (0%) | 9 (6%) |
|  Unknown  | 51 (34%) | 1 (14%) | 52 (33%) |
| Had primary care provider (%) | 98 (66%) | 6 (86%) | 104 (67%) |
|  Did not | 3 (2%) | 0 (0%) | 3 (2%) |
|  Unknown | 48 (32%) | 1 (14%) | 49 (31%) |

Totals may not add up to 100% due to rounding.

IQR: interquartile range; MMC: myelomeningocele.