Treatment of myelomeningocele: a review of outcomes and continuing neurosurgical considerations among adults

A review

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Object. Myelomeningocele is the most severe congenital malformation of the CNS that is compatible with survival. From the time of the development of practical treatment for hydrocephalus in the late 1950s, affected individuals began to survive into adulthood in substantial numbers. Data on the neurological status of these individuals are sparse, as are descriptions of their continuing requirements for neurosurgical care.

Methods. A review of the literature was undertaken using the PubMed database maintained by the National Library of Medicine. Formal grading of the quality of evidence was not attempted, but methodological issues affecting validity or generalizability were noted.

Results. Observations from 2 major longitudinal studies of cohorts of patients treated without selection using contemporary neurosurgical techniques have been published at intervals beginning in the mid-1970s. Numerous cross-sectional, institutional reviews have focused on neurosurgical issues in adulthood: hydrocephalus, Chiari malformation Type II and syringomyelia, and secondary spinal cord tethering. The organization of medical services for adults with myelomeningocele has received limited study.

Conclusions. Surviving adults with myelomeningocele achieve a wide range of neurological and functional outcomes, the most critical and adverse determinant of which is symptomatic CSF shunt failure. From a neurosurgical standpoint, adults with myelomeningocele remain clinically active indefinitely, and they deserve periodic neurosurgical surveillance. *(DOI: 10.3171/2010.9.PEDS10266)*

KEY WORDS • **myelomeningocele** • **hydrocephalus** • **Chiari malformation Type II** • **spinal cord tethering** • **adult**

THE introduction of effective and practical CSF shunts in the late 1950s altered the natural history of myelomeningocele. Prior to that time, survival was the exception. Premature death has become the exception subsequently, but long-term survival for patients with myelomeningocele often comes at a high cost in terms of surgical procedures and their complications. Neurosurgical complications pose particular threats to functional outcomes as well as to life itself, so neurosurgical outcomes among patients with myelomeningocele reaching adulthood are of interest.

Once adulthood has been attained, what then? In North America and the United Kingdom, children with myelomeningocele usually find their way to specialized, multidisciplinary clinics staffed by pediatric neurosurgeons and other pediatric subspecialists. Thus, a small cadre of pediatric neurosurgeons has acquired a large experience

with the management of myelomeningocele in childhood, and indeed these experiences have been documented extensively in the literature. In early adulthood, however, patients with myelomeningocele scatter. Some receive continuing but disjointed care in the offices of various surgical specialists. A small fraction are enrolled in dedicated programs administered by physiatrists and staffed variably by therapists and social workers with surgical specialists available on an ad hoc consultative basis. Many receive no continuing care at all. The diffusion and disappearance of these patients once they graduate from children's medical centers has been an obstacle to the development of a body of knowledge concerning the ongoing neurosurgical needs of adults with myelomeningocele.

The literature review that follows attempts to address both of these issues: What has been the neurosurgical experience of patients with myelomeningocele who have reached adulthood? And what continuing neurosurgical issues do they face?

Abbreviation used in this paper: ICP = intracranial pressure.

Methods

The PubMed database maintained by the National Library of Medicine was searched on the terms "myelomeningocele" and "adult." Also searched were "Related Citations" designated by PubMed for relevant articles, bibliographies of lead or senior authors of relevant articles, and reference lists. Emphasis was placed on topics of practical neurosurgical concern. The substantial body of literature concerning the psychosocial status of adults with myelomeningocele and obstetrical issues among women with myelomeningocele was judged to lie outside the scope of this review and not considered in detail. Obstetrical management has been reviewed recently.[44] No attempt was made to assign formal grades to the evidence presented in the reviewed publications, but methodological issues affecting validity and generalizability were noted and are discussed.

Results

Survival

The probability that a newborn with myelomeningocele will survive to adulthood was greatly enhanced by the development of effective treatment for hydrocephalus in the late 1950s and, to a lesser extent, by introduction of clean intermittent catheterization and other measures for management of neurogenic bladder dysfunction in the 1970s. Survivors from this epoch are today's middle-aged adults with myelomeningocele. Their numbers are considerable, and concurrent with the swelling of their ranks, the incidence of new cases of myelomeningocele has plummeted by an order of magnitude to roughly 2:10,000 live births.[10] Adults with myelomeningocele now account for greater health care expenditures and more hospital admissions than children with this condition.[66]

Another factor impinging on the survival of newborns during this epoch was variation in criteria for treatment. A complete discussion of the historic controversy over selection of patients for active treatment is beyond the scope of this review, but understanding of contemporary survival statistics requires a minimum of context. The initial introduction of CSF shunts into neurosurgical practice allowed indiscriminate treatment of the hydrocephalus associated with myelomeningocele. The enthusiasm of neurosurgeons and other practitioners over this new capacity to treat a previously untreatable condition was dampened before long by discovery of the complications of shunt therapy, by new burdens of care for surviving disabled children assumed by families and by the health care system, and ostensibly by the suffering of the survivors themselves. This disillusionment was not limited to the United Kingdom, but it was articulated most systematically by John Lorber, a British pediatrician, who in the mid-1970s promoted criteria for selection for treatment and reported the outcomes of such selection. Lorber and Salfield[53] prescribed withholding active treatment from newborns with gross macrocephaly or myelomeningocele levels of L-3 or above to minimize the prevalence of severe disability among survivors. This policy was opposed vigorously by David McLone of Chicago, among others. McLone,[59,60] a leading figure in the development of the subspecialty of pediatric neurosurgery in North America, advocated aggressive, coordinated, multidisciplinary care of all newborns with myelomeningocele regardless of the severity of neurological involvement, and over time, he developed outcomes data that demonstrated much improved survival without any greater prevalence of severe disability. Contemporaneous cultural and political trends toward enhanced sensitivity to the rights of the disabled amplified the impact of McLone's work and established nonselective treatment of newborns with myelomeningocele as the standard of care in North America from the early 1980s onward. The first Chicago cohort of nonselectively and aggressively treated patients is approaching an average age of 35 years at the time of this writing.[7]

Prior to the epoch of active treatment, long-term survival for newborns with myelomeningocele was uncommon indeed. Laurence[49] reported actuarial survival statistics for 426 newborns with myelomeningocele born in South Wales between 1956 and 1962. At that time and place, the incidence was very high by contemporary standards, roughly 4:1000 live births, and no patients received CSF shunts. Less than 20% of newborns survived past 24 months of age, but there was no subsequent mortality through 5 years of age. Davis et al.[13] presented actuarial statistics for 904 patients with myelomeningocele followed through the regional multidisciplinary birth defects program affiliated with the University of Washington in Seattle between 1957 and 2000. Dividing their cases into 2 groups based on dates of birth before or after the introduction of CT scanning in 1975, they documented survival to adulthood at rates of 54% and 85%, respectively, although continuing mortality was observed in early and mid adulthood. Hunt and Palmer[38] reported a median survival of 30 years in a remarkable cohort of 117 consecutive patients observed from birth into adulthood without any loss to follow-up. Past the age of 5 through the early adult years, there was a steady annual mortality rate of 3%. Oakeshott et al.[63] have now extended the observation of this cohort to 40 years. Their analysis shows a clear association of survival with spinal level. More than half of the deaths were unexpected and out of hospital; common causes were epilepsy, pulmonary embolism, CSF shunt failure, and urinary sepsis. Bowman et al.[7] have estimated 75% survival to adulthood for the first Chicago cohort with minimum follow-up of 20 years. They have observed continuing mortality in adulthood due to CSF shunt failure.

Prospective Studies

There have been 2 long-term, longitudinal cohort studies that have followed patients with myelomeningocele from birth into early and mid adulthood. They were initiated at Addenbrooke's Hospital, Cambridge, during the years 1963–1971 and at Children's Memorial Hospital, Chicago, during the years 1975–1979.[6,63] To survey the status of these 2 groups of patients offers the best perspective available on the contemporary neurosurgical condition of adults with myelomeningocele.

The Cambridge cohort was created by an agreement

among regional pediatricians to refer all newborns with open spine lesions to the neurosurgical service at Addenbrooke's Hospital, where 117 patients underwent closure of the spinal lesions within 48 hours of birth without any selection on the basis of severity of involvement. Ventriculoatrial shunts were inserted for active hydrocephalus manifested by macrocephaly or symptoms of intracranial hypertension. A conservative approach was taken to CSF shunt management. Revision was largely reserved for episodes of shunt failure with symptoms or signs of elevated ICP; there were no so-called "elective" revisions.[62] Surgery for hindbrain decompression or for tethered spinal cord is not mentioned in the publications of the experiences of the Cambridge cohort. Nor is there discussion of clean intermittent catheterization. Thus, despite initial nonselective treatment, there may have been differences between the management of this cohort and contemporaneous and current practices in North America. Follow-up studies were published at intervals by research general practitioners who were not involved directly in the care of the patients.[30–39,62–64] There was no neurosurgeon among the investigators. No patient was lost to follow-up. At the latest accounting, 60 patients had died. Causes of death as determined predominantly by death certificates were cardiopulmonary failure, renal failure and urinary sepsis, CSF shunt failure (10 patients), CNS infection (10 patients), epilepsy, and thrombotic thrombocytopenic purpura.[36]

Cognitive outcomes were powerfully related to individual histories of shunt treatment. Among 57 survivors assessed at an average age of 30 years, 9 patients had never had a shunt, 16 patients had a shunt that had never required revision, 10 patients had had revisions only before the age of 2 years, and 20 patients had undergone revisions at later times. Forty survivors (70%) had full-scale IQs greater than 80.[62] Hunt et al.[37] characterized the functional outcomes in this cohort on the basis of attainment of 1 or more of the following: independent, unsupervised community living; competitive employment; or regular driving of a motor vehicle. By these criteria, 27 of 57 survivors were "achievers." Eight of 9 patients who never had shunts were achievers, whereas only 20 of 48 patients with shunts attained this outcome. Among 16 patients with never-revised shunts, there were 11 achievers (69%); 5 (50%) of 10 patients who underwent shunt revision only in infancy were achievers; only 4 (18%) of 22 patients who required shunt revision after 2 years of age were achievers. The differences in rates of achievement among these groups of shunted patients were highly significant (p < 0.001). As there were no "elective" shunt revisions, categorization of patients by shunt history correlated highly with histories of clinical intracranial hypertension. Interestingly, normal intelligence as defined by full-scale IQ was unrelated to shunt history.[37] There were 2 cases of blindness and 4 cases of severe visual impairment attributed to episodes of CSF shunt failure with papilledema.[35]

Community ambulation, defined as the capacity to walk 50 m, was present in 32 patients (56%) at 9 years of age but in only 17 patients (30%) at 30 years of age. Only 2 of 17 patients with sensory levels at L-5 or below lost the capacity for community ambulation, but only 1 of 13 patients with myelomeningocele at L-4 or above who were community ambulators at age 9 years retained this capacity at late follow-up. No explanation for the observed decline in mobility was offered.

Urological outcomes among the members of the Cambridge cohort have not been reported.

The Chicago cohort consists of 118 patients whose myelomeningocele was managed without selection by closure of the back lesion within the first few days of life, by insertion of CSF shunts for management of hydrocephalus, and by subsequent follow-up in a multidisciplinary clinic of the type that has subsequently become prevalent in North America. In 2001 Bowman et al.[7] described the status of this cohort at an average age of 22 years. At that time, 28 patients had died, and 19 had been lost to follow-up, leaving 71 patients available for assessment.

Among the surviving 71 patients, 61 (86%) had CSF shunts—precisely the same fraction as among the patients who had died. Only 9 (46%) of 19 patients lost to follow-up had had shunts, suggesting that the patients who had failed to return to clinic were as a group less involved. A total of 226 shunt revisions had been performed in the 71 patients available for review. Only 3 patients had not undergone a revision. The mode was 2 revisions; the median was 4. Three patients had had more than 12 revisions. Twenty-two patients had had shunt infections. Deaths due to CSF shunt failure were continuing to occur into young adulthood.[7]

Bowman et al. characterized mobility as the practice of ambulating "a majority of the time." By this criterion, 24 of 26 adults with myelomeningocele levels of L-5 or below were ambulatory. Despite careful follow-up and aggressive management of spinal cord tethering, 11 patients (16%) had declined by 1 or more spinal motor levels compared with early childhood.

Clean intermittent bladder catheterization was practiced by 60 patients (85%), and 54 of them (95%) performed this procedure without assistance. Among patients who were catheterized, 15% were totally dry, and 83% were dry "a majority of the time." Social bowel continence has been maintained in 53%.

Surgery for secondary spinal cord tethering had been required for 23 patients (32%) in the study group.[7] Two patients underwent 2 operations for tethered cord, and 1 patient had 3 operations. Only 3 operations for tethered cord have been performed in adulthood.[8] Four patients among the 71 continuing in follow-up had been treated by craniocervical decompression; 4 patients had had tracheostomies; and 6 patients had had gastrostomies. These procedures had been more prevalent among the patients who had died, but there had been no decompressions, tracheostomies, or gastrostomies among the patients lost to follow-up.[7]

Data on social competitiveness are not available to permit ready comparisons with the Cambridge cohort, but 2 patients were married and living with their spouses, and 11 lived independently. Fifty-five patients (77%) were living with a parent.[7] Data correlating competitiveness with CSF shunt history are likewise not available.

Hydrocephalus

The dominant neurosurgical issue in the management of children with myelomeningocele is maintenance of satisfactory CSF shunt function, and, regrettably, adults with myelomeningocele continue to experience symptomatic and even life-threatening episodes of shunt failure. The baleful effect on survival of continuing shunt dependence has been illustrated vividly by Davis et al., who analyzed a very large data set acquired prospectively beginning in 1957 through the regional multidisciplinary birth defects clinic at the Children's Hospital and Regional Medical Center in Seattle.[13] Among patients born before 1975, the rate of survival to 16 years of age was 54%. Among patients born later, actuarial survival to 16 years of age was 85%. The authors compared subsequent survival past the 16th birthday for 117 patients with CSF shunts and for 56 patients without shunts. Actuarial survival to age 34 years was 94% among patients without shunts but only 75% among shunt-treated patients (p ≤ 0.03). The cause of death was not determined, but the clear implication in this report was that the poorer survival among shunt-treated patients was causally related to shunt complications. Shunt dependence among patients with myelomeningocele is linked to spinal level, and Oakeshott et al.[63] have documented less frequent survival to adulthood among patients with higher spinal levels. In the data of Oakeshott and colleagues, however, this survival differential was established by 5 years of age, beyond which point survival plots for various spinal levels appear parallel. The excess mortality among adults with myelomeningocele and shunts observed by Davis et al.[13] is very likely attributable to complications of shunt treatment.

Other authors likewise have reported deaths and various catastrophic complications of shunt failure among adults with myelomeningocele. Tomlinson and Sugarman[80] tracked down 110 young adults (average age 21.5 years) with myelomeningocele and CSF shunts who had been students at a regional school for children with disabilities. There were 13 deaths in this study group: 3 patients had died of recognized shunt failure. In 2 instances, death had been preceded by chronic headaches for which medical attention had been sought. There were 4 sudden deaths in the community suspected to have been due to shunt failure. Sgouros et al.[75] reported the adulthood experiences of 70 patients followed from childhood for congenital hydrocephalus, including 23 with myelomeningocele. Thirty-three of 201 shunt revisions and 5 of 8 subtemporal decompressions were required after the age of 16 years. There were 2 deaths in adulthood from shunt failure, and 1 patient became blind. Acquisition of visual impairment and blindness in adulthood has been reported by other authors as well.[18]

The presence of a CSF shunt and complications of shunt treatment are associated with compromised cognitive function as well as poorer prospects for survival. Hetherington et al.[27] studied 31 adults with myelomeningocele and shunt-treated hydrocephalus who had verbal and performance IQs greater than 70. Patients were categorized according to myelomeningocele level: L-1 and above or L-2 and below. They were categorized according to shunt history as well: 4 and more shunt revisions or 3

and fewer shunt revisions. A battery of neuropsychological tests was administered together with a quality of life questionnaire. Shunt history but not spinal level was related to cognitive performance. Patients who had undergone 4 or more shunt revisions exhibited lower performance IQ and poorer functional math skills, which were in turn statistically associated with unemployment in this study group. Quality of life was modeled by logistic regression using all medical, demographic, and psychological variables. The model accounted for only 16% of the variance of the quality of life data, and shunt history failed to be retained as a significant predictor. In related work, Dennis et al.[16] studied upper-limb function in a similar group of adults with shunt-treated hydrocephalus and matched controls. The association of greater numbers of shunt revisions with lower performance IQ was confirmed, but there was no correlation with execution of upper limb motor tasks. Not surprisingly, higher spinal levels were associated with lower motor independence. Adults with spina bifida tended to do worse than controls on all tests of upper-limb function. Iddon et al.[42] studied neurocognitive function among adults with hydrocephalus with or without myelomeningocele, among adults with myelomeningocele but no hydrocephalus, and among matched healthy controls for both of these study groups. Verbal IQ was within the normal range for both patient groups. In the aggregate, adults with myelomeningocele but no hydrocephalus were no different from healthy controls, whereas the patients with hydrocephalus exhibited diffuse impairments of spatial recognition and working memory and verbal learning and delayed recall. A follow-up study by Iddon et al.[43] split out patients with hydrocephalus but no myelomeningocele as a third study group and confirmed that patients with hydrocephalus were diffusely impaired, especially with respect to executive functioning. Scores for patients with myelomeningocele without hydrocephalus generally fell within the normal range.

The question of so-called "arrested" hydrocephalus among older patients with myelomeningocele is vexed, at least in part because of inconsistencies in the usage of this term and related ones. Long periods of clinical quiescence are certainly no guarantee of shunt independence in the setting of myelomeningocele. In their study group of 117 young adults, Tomlinson and Sugarman[80] recorded 320 CSF shunt revisions. Twenty-four of these procedures were performed more than 10 years after the preceding episode of symptomatic shunt failure and 11 after more than 15 years. Edwards and Pople[18] examined decompensation among patients with arrested hydrocephalus, which they defined as asymptomatic ventricular enlargement compared with a previous postshunt baseline image. Among 173 patients with hydrocephalus of various etiologies, 17 (9.8%) met this definition of arrest. Arrest of hydrocephalus was a much more common phenomenon among patients with myelomeningocele than among patients with other diagnoses, occurring in 13 (26%) of 50 cases. Decompensation requiring additional surgical treatment occurred in 10 cases at an average age of 23.4 years and after an average interval of 16 years from the diagnosis of arrest. Eight of 10 patients exhibited new visual impairment from papilledema at the time of

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decompensation. There was residual optic atrophy in 4 patients, and 1 patient remained blind. Similar late decompensation of patients with myelomeningocele never previously treated for hydrocephalus has been reported as well.[81] Among the 9 adult patients with myelomeningocele presented by Oi et al.,[65] 3 exhibited "long-standing occult ventriculomegaly in adulthood" (LOVA). These 3 patients exhibited lifelong macrocephaly. Two of them came to attention because of severe headaches, which were replaced after shunt insertion by dizziness and postural headaches indicative of intracranial hypotension and requiring "delicate" adjustments of their programmable valves. Ventricular volume remained increased in both treated patients. One of the treated patients and the remaining untreated patient suffered severe depression as well. The challenge of chronic headaches has been taken up by Edwards et al.[19] Among their clinic population of adults with myelomeningocele and CSF shunts, they identified 42 patients who had undergone 51 evaluations for chronic headache. Twenty-two (43%) of 51 episodes were eventually attributed to some complication of hydrocephalus or its previous treatment. Early morning exacerbation, vomiting, and visual symptoms had fair specificity for intracranial hypertension. The authors stressed the utility of ICP monitoring. Mataró et al.[55] also endorsed ICP monitoring in the evaluation of adults with myelomeningocele and stable ventriculomegaly. They studied 23 patients with chronic, nonspecific symptoms such as sporadic headache and cognitive or behavioral complaints or with no symptoms at all. In 12 cases, a CSF shunt had been inserted earlier in childhood but was believed no longer to be functioning; the remaining 11 patients had never had any treatment for hydrocephalus. These authors categorized patients based on ICP criteria. Patients who exhibited mean ICP readings greater than 12 mm Hg were considered "active." Patients with mean ICP less than 12 mm Hg but with Lundberg A or B waves were considered "compensated," and all others were considered "arrested." All patients with active or compensated hydrocephalus underwent CSF shunt insertion after a battery of preoperative neuropsychological tests. Active patients performed worse on preoperative testing than compensated patients; symptomatic patients performed worse than asymptomatic patients. Postoperative testing at 6 months showed aggregate improvements on all neuropsychological tests. Active and compensated patients and symptomatic and asymptomatic patients responded equally well to intervention. A similar description of a smaller, exclusive group of never previously treated patients from the same institution has appeared as a separate report.[41]

Apparently the first report in the medical literature describing the successful pregnancy of a woman with myelomeningocele was by Fujimoto et al.[22] in 1973. Women with myelomeningocele of this generation who survived to adulthood and achieved pregnancy generally did not have hydrocephalus, but for subsequent generations, the problem of the management of hydrocephalus during pregnancy has become salient. Reproductive issues among women with myelomeningocele have been subjected to formal literature review recently, but neurosurgical management was not considered.[44] In 2000, Ara-

ta et al.[3] reported the largest series of pregnancies among women with myelomeningocele to date. Seventeen women had experienced 29 pregnancies. Only 5 women had CSF shunts, and there were no episodes of shunt failure. There is a single case report of an episode of shunt failure during pregnancy in a woman with myelomeningocele,[28] but there is no other literature expressly devoted to neurosurgical matters during pregnancy in this setting. There are, however, several reviews and case series of pregnancies among women with hydrocephalus of unselected etiologies.[9,12,48,51,84] Cusimano et al.[12] described the case of a 21-year-old woman with hydrocephalus related to Dandy-Walker malformation who developed typical symptoms of shunt failure in the 30th week. Radiological evaluation was deferred, and her symptoms were managed by pumping of the valve and by aspiration of CSF from the shunt reservoir for 6 weeks until an elective cesarean section was performed. The authors attributed the patient's symptoms to functional obstruction of the shunt by high abdominal pressure. They reviewed the existing literature and found descriptions of only 17 pregnancies in 16 patients with shunts.

There were 5 other examples of functional shunt obstruction in the third trimester managed without surgery. Wisoff et al.[84] reviewed the literature and added 11 cases. Among 17 pregnancies in women with preexisting shunts, symptoms of shunt failure developed in 11. Diagnostic investigations indicated shunt patency in 7 patients who were treated variably by bed rest, fluid restriction, diuretics, steroids, shunt pumping, and CSF aspiration. In 4 of 17 pregnancies, CSF shunt revision was required during the pregnancy or within the following year. The authors recommended neurosurgical evaluations in advance of planned pregnancies to ensure optimal shunt function at the outset. Although there had been no reports of peripartum shunt infection, liberal administration of prophylactic antibiotics was recommended for both cesarean section and vaginal delivery.[84] Landwehr et al.[48] reported 25 pregnancies among 8 women with shunts—4 for hydrocephalus and 4 for pseudotumor. There were no complications related to the CSF shunts. No prophylactic antibiotics were given for the vaginal deliveries, and there were no shunt infections. Bradley et al.[9] surveyed 37 women with shunt-treated hydrocephalus, including 4 women with myelomeningocele, about their experiences during pregnancy. The patients were identified through the practices of the authors, through registries maintained by support groups, and through announcements in periodicals. In the course of 77 pregnancies, 10 CSF shunt revisions were required for 7 women during or within 6 months of the pregnancy, but there were no neurosurgical complications of delivery. There were no shunt infections. Forty-seven women, who accounted for 84% of the surveyed pregnancies, did not experience shunt-related complications. Five women gave histories of transient third trimester headaches. Interestingly, 6 of 19 women who breastfed their newborns reported headaches during nursing. This group subsequently extended their observations to 138 pregnancies among 70 women.[51] The authors drew attention to the months after delivery as a period of potential shunt instability.

Spinal Cord Tethering

A formal review of the literature concerning secondary spinal cord tethering among adults with myelomeningocele appeared recently.[25] George and Fagan[25] found only 7 case reports and case series reporting clinical experiences with this condition up through 2003.[21,52,65,67,74,79,82] The magisterial 1982 paper by Pang and Wilberger[67] on tethered spinal cord in adults includes a single patient with a sacral myelomeningocele who was investigated using metrizamide CT myelography. In addition to tethering at the site of the repair, this patient had a tight filum terminale. No other details were reported. Tamaki et al.[79] reported on 9 patients with neurological deterioration or progressive foot deformities after closure of myelomeningocele in infancy, but the 2 oldest patients were only 15 and 19 years of age. A case report by Logan et al.[52] described a 44-year-old sailor who had had an active and unrestricted career in the US Navy until retirement and who came to attention because of gait disturbance. He stated that he had undergone repair of a myelomeningocele in infancy. His neurological examination was notable for leg atrophy and long-standing ankle deformity. There was an old scar in the lumbosacral region. Imaging investigation showed spina bifida occulta at L-5 and a low conus. At surgery there was adherence of the conus to scar at the site of the previous operation, and there was a tight filum terminale. Ambulation improved. Filler et al.[21] reported on 2 young, independently ambulatory adults with precipitous but atraumatic onset of severe neurological impairment attributable to secondary tethering. Both patients made excellent functional recoveries after timely surgical treatment. The authors were struck by the fulminant preoperative courses in these cases, which they had not encountered in their experience with adult tethered cord syndrome from other causes. Oi et al.[65] reviewed an institutional series of adults with spinal dysraphism. The 18 cases were equally divided between myelomeningocele and other forms of spina bifida occulta. One patient who had undergone closure of myelomeningocele at birth required reoperation for secondary tethering at 17 years of age. Five of 9 patients with spina bifida occulta developed symptoms and signs of tethering in adulthood, 2 of them having undergone previous surgery for dysraphism early in childhood. The authors concluded that the development of active clinical tethering in adulthood is less common among patients with myelomeningocele than among patients with other forms of dysraphism. Selber and Dias[74] reviewed the status of 44 adults with sacral level myelomeningocele. All patients had been community ambulators in childhood, and at last follow-up at an average age of 23 years, only 3 had deteriorated to household ambulator or wheelchair status. Twelve patients had undergone tethered cord release, but the ages at which this intervention was performed were not stated. Van Leeuwen et al.[82] reported a large institutional series of adults operated for tethered cord syndrome that included 12 patients with myelomeningocele and secondary tethering. Three of 12 patients enjoyed postoperative improvements of 1 or 2 points on a modified Rankin Scale; no other data specific to myelomeningocele were reported. Thus, the review of George and Fagan[25] captured clinical data on precisely 18 patients older than 18 years of age with secondary tethering after repair of myelomeningocele in infancy. These data support very little generalization except to say that secondary tethering occurs with an incidence that cannot be estimated and that surgical intervention is sometimes beneficial.

Subsequent work has brought the phenomenon of secondary tethering among adults into only slightly sharper focus. Begeer and Staal-Schreinemachers[5] mentioned 6 adult patients with myelomeningocele who required surgery for secondary tethering. Outcomes in this group were not presented separately from other patients with spina bifida occulta, but they stressed the continuing clinical activity of adults with myelomeningocele. Martinez-Lage et al.[54] reported the interesting case of a 20-year-old male who initially underwent closure in infancy but returned to attention with a claudication syndrome. Magnetic resonance imaging showed developmental lumbar stenosis and slight retrolisthesis of L-5 on S-1. He underwent L-5 laminectomy and release of tethering and subsequently returned to his functional baseline. This case illustrates the potential interaction of tethering with developmental and acquired, degenerative changes in spinal architecture. Quiñones-Hinojosa et al.[72] presented technical specifications for electrophysiological monitoring during surgery for adult tethered cord syndrome. These authors' clinical material included myelomeningocele with secondary tethering, but no specific details were provided. Lee et al.[50] reported long-term follow-up for 60 adults treated for tethered spinal cord, including 15 patients with myelomeningocele, 4 of whom had undergone surgery for secondary tethering previously in childhood. In general, the authors judged that untethering procedures for adults with myelomeningocele were more difficult than for other pathologies, and the outcomes were less satisfactory, although still worthwhile. Among the cases of myelomeningocele, patients presenting with back or leg pain improved in 67% of cases. Improvement was seen in 54% of cases presenting with new weakness and in 63% of cases presenting with deteriorating bladder function. Two patients had new postoperative weakness, and 1 had worse leg pain. In the review of surgery for secondary tethering presented by Al-Holou et al.,[2] there were 8 patients 20 years of age or older. Myelomeningocele and lipomyelomeningocele were represented among their patients, and adults were not discussed separately. The authors noted that neither original pathological diagnosis nor a history of previous surgery for secondary tethering affected the likelihood of benefit from intervention. Young age at first reoperation for secondary tethering affected long-term outcome adversely, however. Among the 29 consecutive adults who underwent surgery for tethering reported by Garcés-Ambrossi et al.,[23] there were 14 cases of myelomeningocele. Tethering pathology was not a significant predictor of response to treatment in this series, but the authors made the interesting observation that presentations featuring asymmetrical weakness or lower-limb hyperreflexia were associated with a higher rate of postoperative improvement in motor function. Postoperative recovery was largely complete within 6 months.

As adults are not just big children, there seem to be

distinctive features to tethered cord syndrome in older age groups. From the original work of Pang and Wilberger[67] onward, many authors have commented on the roles of trauma and vigorous activity in the precipitation of symptoms and signs;[1,20,26] however, this association seems to be less prominent among adults with myelomeningocele and symptomatic secondary tethering. Pregnancy and childbirth likewise are described as destabilizing events among women with cord tethering, although most of the examples in the literature have been women with various forms of spina bifida occulta.[1,15,20,40,45,71,76] Pang and Wilberger[67] proposed that assuming the lithotomy position for pushing and delivery places the tethered spinal cord under tension. Alternatively, the general ligamentous relaxation that occurs late in pregnancy may alter the mechanics of the spine, or changes in adiposity or in pelvic venous pressure may affect the marginally perfused tethered cord.[76] In a review written prior to the era of MR imaging, Shurtleff[76] opined that disc herniations were common among pregnant women with myelomeningocele. Accelerated intervertebral disc degeneration in the dysraphic spine has been attributed to abnormal mechanics of gait or to intrinsic disturbances of disc development,[14] and there has been speculation that spinal degenerative changes may destabilize the tight but previously asymptomatic tethered cord.[20,24,40,67,76,77] The case presented by Martinez-Lage et al.[52] may illustrate this phenomenon, but the paucity of descriptions of clear synergism between spondylosis and tethering raises the possibility that, as the spine shortens with aging, degenerative changes may actually have a protective effect. This concept has been realized surgically in the recent descriptions of spondylectomy for treatment of intractable tethering.[29,61]

In summary, there is only sparse information in the literature on secondary spinal cord tethering among adults with myelomeningocele. All of it is retrospective, descriptive, and uncontrolled. The magnitude of the problem is impossible to judge. The diagnosis of tethering is predicated on longitudinal assessments by a clinician familiar with the condition, but how many adults with myelomeningocele have access to such care is unknown. Furthermore, although practice variation in the diagnosis of tethering has never been measured, it is likely to be significant. All that can be said is that symptomatic secondary tethering occurs and that selected patients can benefit from surgical treatment.

Chiari Malformation and Syringomyelia

There is substantial literature describing the presentations of Chiari malformation Type II and associated syringomyelia in childhood,[68,70,83] although contemporary practice has trended away from earlier emphasis on craniocervical decompression toward confirmation of satisfactory CSF shunt function. Reports of experiences among adults are far fewer. Disturbed hindbrain physiology certainly persists into adulthood. For example, Swaminathan et al.[78] studied pulmonary function in 14 young adults with myelomeningocele and 14 healthy controls. Although the study patients were not actively symptomatic, hypercapnic ventilatory responses were significantly depressed. McDonnell et al.[58] attempted to assess the

prevalence and the clinical significance of the Chiari II malformation by canvassing patients attending a program for adults with myelomeningocele in Belfast, Northern Ireland. Of 76 patients evaluated, 63% had symptoms or signs consistent with the Chiari II malformation or syringomyelia, but only 27% reported developing new relevant symptoms in the preceding 12 months. Among 25 patients who had undergone MR imaging in the course of this investigation, 76% had Chiari malformations, and 48% had syringomyelia. Clinical assessment exhibited a specificity of 100%, but many patients with MR findings were asymptomatic. Craig et al.[11] reported 5 patients from among 220 adult patients followed through this same program in Belfast who required surgical treatment for the "Chiari/hydrosyringomyelia complex." Upper-limb weakness and wasting was the presentation in 4 patients; only 1 patient had a bulbar syndrome. Treatment was craniocervical decompression in 2 patients, CSF shunt revision in 2 patients, and both interventions in the remaining patient. Only 1 patient enjoyed significant improvement; 2 stabilized, 1 suffered further deterioration, and 1 died of pneumonia about 10 weeks after surgery. These discouraging results may be related to the protracted histories that averaged 3 years before treatment. Piatt and D'Agostino[69] mentioned 2 adult patients with myelomeningocele who underwent craniocervical decompression with exploration of the fourth ventricle. Headaches, gagging, and vomiting were the symptoms of 1 patient whose MR imaging study showed a 2-cm cyst of the uvula of the vermis. Symptoms resolved with decompression and excision of the cyst, which appeared to be gliotic cerebellar tissue on microscopic examination. The second patient, who had associated syringomyelia, came to attention with upper-limb weakness with intrinsic muscle atrophy. At surgery for decompression, multiple small nodular lesions were found among the fronds of the choroid plexus in the roof of the fourth ventricle. Histological examination was consistent with subependymoma. His clinical condition did not improve. (A second example of subependymoma was reported in a child in this series of 7 patients.) Rahman et al.[73] described 4 adult patients with recurrent hindbrain symptoms after craniocervical decompression in childhood. Presentations included headache, neck pain, diplopia, dysphagia, dysphonia, upper-limb paresis and hypertonia, and gait instability. Retroflexion of the dens seemed to contribute a degree of ventral cervicomedullary compression in 3 of 4 cases. After careful evaluation of CSF shunt function including surgical exploration in several cases, these patients were treated by re-decompression and posterior occipitocervical fusion with some benefit in each instance. The authors posited a role for chronic, progressive craniocervical deformity in late development of symptoms among patients who have undergone decompression in childhood, although none of their patients exhibited instability on dynamic radiographs. This review thus has identified published descriptions of the treatment of only 11 adults with myelomeningocele symptomatic from the Chiari II malformation or associated syringomyelia. As is true for spinal cord tethering, the representativeness of these observations is compromised by inconsistent access for adults with myelomeningocele

to services of sufficient clinical sophistication to recognize and to treat this condition.

Systems of Coordinated Medical Services

Limitation of access to longitudinal, multidisciplinary care for adults with myelomeningocele probably leads to adverse health outcomes, although the data are few. Kaufman et al.[46] described the health consequences of disbandment of a coordinated, multidisciplinary spina bifida clinic in a case-control study. Patients were interviewed 3 years after the closing of their program in St. Louis, and their experiences were compared with a control group matched for diagnosis, age, and distance from home to hospital who had continued care in a Chicago program. Roughly twice the fraction of study patients had had no follow-up specialty care as in the control group. Older patients in the study and the control groups were more likely to disappear from follow-up than younger patients. The authors focused on surgical procedures, which they categorized as operations for shunt complications, operations for "preventable" conditions, and "proactive" operations. The 2 populations had roughly the same rate of shunt surgery. Twenty-seven percent of patients from the disbanded program had undergone operations for preventable complications—predominantly surgery for cutaneous ulcers. The rate from the patients continuing in care was only 1%. The patients who continued in care had a much higher rate of "proactive" operations—predominantly tendon releases, clubfoot repairs, and release of spinal cord tethering. This study was limited to children and adolescents. It was limited as well by its use of surgical procedures as a surrogate measure for health, but it demonstrates clearly the critical role of care coordination.

Kinsman and Doehring[47] addressed the question of preventable morbidity among adults with myelomeningocele as part of an institutional audit of resource utilization by this population. Over an 11-year period, there were 353 admissions of 98 adults with myelomeningocele to the Johns Hopkins Medical Institutions. Neurological conditions, predominantly CSF shunt complications, accounted for 56 admissions. The authors judged that 166 admissions (47%) were for potentially preventable conditions, such as urinary tract sepsis, renal calculi, cutaneous ulcers, and osteomyelitis. The average length of stay for management of these preventable conditions was 12.6 days.

McDonnell et al.[56] described the organization of a "Clinic for Adults with Spina Bifida and Hydrocephalus" (CASBAH) in Belfast, North Ireland, and they surveyed the health status of its registrants. Among 211 surviving adults with myelomeningocele, only about 40% had CSF shunts in place. As an indication of the value of the CASBAH program to the patients who had been referred to it, the authors noted that 179 (73%) continued in regular follow-up with no or infrequent missed appointments. There had been 18 deaths since the inception of the clinic. Renal failure accounted for 4 deaths; 1 patient had died of CSF shunt failure; and 2 patients had died of complications of the Chiari malformation.[57] As other writers have stressed subsequently, cancer of the urinary tract was lethal in 2 instances.[4] Sepsis, cardiac failure and myocardial infarc-

tion, basilar artery aneurysm rupture, postpartum pulmonary embolism, peptic ulcer disease, and status epilepticus accounted for the remaining deaths. The authors emphasize the continuing medical activity of this population and the potential for preventative interventions.

The potential value of prevention was noted as well by Begeer and Staal-Schreinemachers[5] in their report of the experience of a spina bifida program with intake of new adult patients who, apparently, had not had continuing specialized care previously.[5] Among 22 adults with myelomeningocele, 9 were seeking a general evaluation, 8 had experienced recent gait deterioration, and 3 needed attention for cutaneous ulcers; there was 1 case of meningitis and 1 case of shunt failure. The initial evaluations led to 9 neurosurgical procedures: 6 operations for spinal cord tethering and 1 each for cervical stenosis, shunt revision, and shunt insertion. The authors were impressed by the unnecessary and, in many instances, irreversible health damage experienced by these unattended patients, and they remarked on the lack of awareness—among patients and in the medical community—of the progressive nature of myelomeningocele in adulthood.

Most recently, Dicianno and Wilson[17] queried the National Inpatient Sample (NIS) from the Healthcare Cost and Utilization Project of the Agency for Healthcare Research and Quality to assess reasons for hospitalization and causes of in-hospital death among adults with myelomeningocele. The NIS is a data set representing discharge data from roughly 20% of nonfederal, acute-care hospitals in the US stratified to permit extrapolations of estimated national statistics. The most common primary diagnoses were urinary tract infection, complication of device, implant, or graft, chronic cutaneous ulcer, cutaneous or subcutaneous infection, and sepsis—all potentially preventable conditions. Preventable conditions were listed for 33.8% of admissions and an estimated $364 million in hospital charges in the 2004–2005 study period. Remarkably, 35.7% of hospitalizations associated with primary diagnoses of preventable conditions ended in death. Sepsis, pneumonia, and respiratory failure were the leading causes of death. Hospitalizations for preventable conditions were more common at rural hospitals and at urban nonteaching hospitals. The authors underlined the potential health benefits and cost savings of preventative measures and the importance of coordination of care.

This review has not discovered any population-based data regarding access to multidisciplinary, longitudinal care for adults with myelomeningocele. Dicianno and Wilson[17] knew of only 4 programs devoted to adult patients in the US. A search of the web for this writing identified only 9 institutions in North America with spina bifida programs that accommodate adults (Appendix). Funding for such programs is notoriously precarious. The kernel of the model spina bifida program is care coordination, but there is no insurance payment mechanism for care coordination outside the context of the office visit. Even within that context, actual payments are grossly insufficient for the expenditure of time that is required typically. Spina bifida programs therefore must have alternative sources of support such as institutional cost-shifting or programmatic grants, both of which tend to be unstable

over time. For children, the historical source of grant support has been federally funded, state-administered Social Security Act Title V Maternal and Child Health Services Block Grants, but no such entitlements extend to adults. Nor are the chronic care requirements of patients with myelomeningocele accommodated readily by most general neurosurgical practices, which are small businesses devoted to acute, monophasic illnesses. In theory, the cost of care coordination might be recouped by savings from avoidance of preventable complications. As health care is currently organized in the US, such costs and savings are not often realized by the same entity, but the decision by Kaiser Permanente of Northern California, a closed-panel health maintenance organization, to organize an exemplary spina bifida program for adults suggests that the economics can be favorable (http://mydoctor.kaiser permanente.org/ncal/specialty/genetics/specialty_clinics/ SBC/index.jsp).

Conclusions

To call for more research is customary at the conclusion of a literature review, but more research cannot be the first priority. The more urgent necessity for adults with myelomeningocele is access to longitudinal, multi-specialty clinical care like what most of them experienced as children. This review has demonstrated a continuing requirement for neurosurgical follow-up, and comparable data are readily available for urology and orthopedics. If programs of this kind existed in greater numbers, clinical research would follow without any exhortation, as it has historically for children with myelomeningocele. As befits the specialty most instrumental in the survival of these patients to adulthood, neurosurgeons must support efforts to organize longitudinal, multispecialty care for this population.

Appendix

Institutions with programs that offer longitudinal care for adults with myelomeningocele.

Gillette Lifetime Specialty Healthcare, St. Paul, Minnesota
Glenrose Rehabilitation Hospital, Edmonton, Alberta, Canada
Kennedy Krieger Institute, Baltimore, Maryland
Loma Linda University, Loma Linda, California
Moss Rehabilitation Center for Adults, Elkins Park, Pennsylvania
Kaiser Permanente Northern California, Oakland, California
Rehabilitation Institute of Chicago, Chicago, Illinois
University of North Carolina School of Medicine, Chapel Hill, North Carolina
University of Pittsburgh Medical Center, Pittsburgh, Pennsylvania

Disclosure

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