Spina Bifida Outcome: A 25-Year Prospective

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Abstract
Background: Open spina bifida is the most complex congenital abnormality compatible with long-term survival. This report outlines the 20- to 25-year outcome for our original cohort of patients with a myelomeningocele treated in a nonselective, prospective manner. Methods: Of the initial 118 children, 71 patients were available for our most recent review. Nineteen patients have been lost to follow-up and 28 patients have died. Data were collected on: motor level, shunt status, education/employment, seizure history, mobility, bladder/bowel continence, tethered cord, scoliosis, latex allergy, posterior cervical decompression, tracheostomy and/or gastrostomy tube. Results: Mortality (24%) continues to climb into young adulthood. Eighty-six percent of the cohort have cerebrospinal fluid diversion, with 95% having undergone at least one shunt revision. Thirty-two percent have undergone a tethered cord release, with 97% having an improvement or stabilization in their preoperative symptoms. Forty-nine percent have scoliosis, with 43% eventually requiring a spinal fusion. Sixteen patients (23%) have had at least one seizure. Eighty-five percent are attending or have graduated from high school and/or college. More than 80% of young adults have social bladder continence. Approximately 1/3 of patients are allergic to latex, with 6 patients having experienced a life-threatening reaction. Conclusion: At least 75% of children born with a myelomeningocele can be expected to reach their early adult years. Late deterioration is common. One of the greatest challenges in medicine today is establishing a network of care for these adults with spina bifida.

Introduction
Open spina bifida is the most complex congenital abnormality compatible with long-term survival. The senior author (D.G.M.) has aggressively treated all children born with open spina bifida in a nonselective, prospective manner [1]. The medical goal always being to maintain stable neurologic functioning throughout the patient’s lifetime. Our original cohort, born between 1975 and 1979, has now reached early adulthood. This report outlines the 20- to 25-year outcome for this group of patients.
Methods

The original study group consisted of 118 children born between 1975 and 1979 with open spina bifida who underwent their myelomeningocele closure at Children’s Memorial Hospital, Chicago, Ill. The 5-, 10- and 15-year outcome for this group of children has previously been reported by the senior author (D.G.M.) [2–5]. This report focuses on the outcome 20–25 years after initial back closure. Currently, the average age of this cohort is 21.7 years with a range from 19.4 to 24.8 years.

Of the initial 118 children, 71 patients (38 females and 33 males) were available for our review. Nineteen patients have been lost to follow-up, 10 (53%) moved between the age of 6 and 19 years. Twenty-eight patients have died.

All patients included in this study completed a mailed questionnaire. Of the 71 patients, 59 are actively followed in our multidisciplinary spina bifida clinic. More detailed information was obtained from our clinic notes. For those patients who have moved, further information was obtained by a phone interview with the patient and/or parents. A chart review was conducted on the patients who have been lost to follow-up or died. Data was collected on: current motor level, shunt status, education and employment, seizure history, mobility, bladder and bowel continence, history of tethered cord and/or scoliosis, latex allergy and history of posterior cervical decompression, tracheostomy and/or gastrostomy tube.

Results

Motor Level

At our institution, a patient’s ‘myelomeningocele level’ is based upon his/her motor function. Each child undergoes a manual motor test immediately following birth, after back closure, following any spinal cord untethering and on a yearly basis. For this report, we have grouped together motor levels that function similarly. The levels are as follows: thoracic (all thoracic and L1), L2/L3, L4, L5, sacral and asymmetrical.

In the study group, 25/71 patients have a thoracic level, 5 are L2/L3, 14 are L4, 11 are L5, 15 are sacral and 1 has an asymmetrical motor exam. When comparing the motor level at birth to the current exam, 52 (73%) patients have remained stable, 8 (11%) have improved and 11 (16%) have worsened. Of the 19 patients who were lost to follow-up, 4 are thoracic level, 1 is L2/L3, 1 is L4, 4 are L5, 8 are sacral and 1 has an asymmetrical exam. In the cohort who died, 11/28 patients were a thoracic level, 4 were L2/L3, 3 were L4, 7 were L5 and 3 were sacral.

Shunt Status

In the study group, 86% (61/71) have cerebrospinal fluid shunts as compared to 47% (9/19) in those lost to follow-up and 86% (24/28) in those who have died. In the study cohort, a majority of patients have a ventriculoperitoneal shunt (97%); 1 has a ventriculopleural shunt, and another has a ventriculooatrial shunt. Fifty-three patients have 1 shunt and 8 have bilateral shunts. In the group that died, 2 children died within hours of their back closure; hence, the reported percentage shunted for this cohort may be falsely low.

In the study group, 95% have undergone at least 1 shunt revision (Fig. 1). Two revisions was the commonest occurrence (21%). Forty-one percent of the shunted study population have had 2–3 shunt revisions. Four patients have had more than 10 shunt revisions; 1 patient has had 31 revisions.

There were a total of 266 shunt revisions completed on this cohort of patients. Twenty-two patients have had at least 1 shunt infection throughout childhood, with 12 patients having had an infection with a gram-negative bacteria.

Education/Employment/Living Arrangements

Currently, 36% (26/71) of the study cohort are in high school or have recently graduated, and 49% (35/71) are attending college or have graduated. Two young adults are in a vocational-type setting and 8 patients are involved in workshop activities. Sixty-three percent (45/71) attend regular classes. Fourteen percent (10/71) require some additional help with their studies and 23% (16/71) are in

Fig. 1. The number of shunt revisions completed on the study cohort.
special education classes. Thirty-two of the 71 young adults are employed and 5 volunteer their services.

Two young adults are married and live with their spouses. Fifty-five patients in the study cohort live with their parents. Eleven live independently and 3 are in residential homes. There are no offspring from the study cohort.

Seizure History
Within the study group, 16 patients have had seizures. All have been on medication for a variable length of time. Six are currently off medication. All 16 patients have shunted hydrocephalus. Within this group, 7/16 patients have had a shunt infection, 5 with gram-negative bacteria. Seven patients have an IQ less than 70 and 5 have an IQ 70–80. Four have an IQ greater than 80 (normal intelligence).

Mobility
As previously demonstrated, mobility decreases from early childhood to the early teen years [4, 5]. Fortunately, the patients who remain mobile in their teens continue to ambulate a majority of the time (75–100%) in their young adult years (fig. 2). Forty-six percent of young adults (33/71) continue to ambulate. Nine patients (13%) ambulate 25–50% of the time. Twenty-nine young adults rely solely upon their wheelchairs for locomotion.

When grouped according to myelomeningocele level, those patients with the better lower extremity motor function have a higher percentage of walking 75–100% of the time (fig. 3). Fourteen of 15 patients with a sacral level (93%) ambulate 100% of the time. Only 1 of these patients uses a wheelchair for assistance around his college campus. Ten of the 11 young adults with an L5 motor level (91%) ambulate a majority of the time as compared to 57% of the patients with an L4 level. None of the patients with a thoracic/L1 or L2/3 level rely on ambulation for a majority of their locomotion, although a few ambulate on a minor basis. The young adult with the asymmetrical motor exam ambulates with braces 50% of the time.

Bladder/Bowel Continence
Sixty young adults (85%) are maintained on clean intermittent catheterization (CIC) of their bladder; 11 patients (15%) are not. Ninety percent (54/60) of the patients perform their own catheterization. For those patients on CIC, 83% are dry a majority of the time (75–100%). Fifteen percent always have urinary continence. Seven percent are dry 50% of the time and 10% are rarely, if ever, continent.

Continence, a majority of the time, decreases to 63% (7/11) for those not on CIC. Three patients not on CIC have no urinary continence; 2/3 have a vescostomy.

Only 38% of this cohort are actively involved in a bowel program. However, 52% of the patients report 100% social bowel continence. Eighty-nine percent of the young adults report bowel control a majority of the time (75–100%).

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Bowman/McLone/Grant/Tomita/Ito
**Tethered Cord/Scoliosis**

Within the study group, 23 patients have developed a symptomatic tethering of their spinal cords. Of those patients requiring a tethered cord release (TCR), 6/23 are thoracic level, 3 are L2/L3, 3 are L4, 5 are L5 and 6 are sacral. The average age at which these children developed a symptomatic tethered cord was 10.9 years. Three patients required multiple untetherings; 2 patients underwent 2 TCRs and 1 patient required 3 TCRs.

The commonest preoperative symptoms are worsening scoliosis (44%), gait changes (35%) and spasticity (26%) (table 1). Others developed back pain with or without radiation into their legs, decreasing muscle strength, lower extremity contractures or urinary bladder changes. Postoperatively, there is a 71% improvement in preoperative symptoms and 26% stabilization of symptoms. Two patients, or 3%, declined postoperatively. One patient has had a worsening of his gait following a tethered cord release at an outside institution. Another patient had an improvement in his preoperative scoliosis, but a decline in his motor level postoperatively (L3 to T).

For the patients who required multiple untetherings, 2 patients presented each time with the same symptoms that had improved after the previous untethering, and 1 patient had a new onset of symptoms with her second untethering.

There were 4 postoperative complications: wound dehiscence (3) and CSF leak (1). Two of the complications occurred in the patients requiring multiple releases.

Thirty-five young adults (49%) have scoliosis. Of the 35, 18 (51%) have a thoracic motor level, 2 are L2/L3, 5 are L4, 5 are L5, 4 are sacral and 1 has an asymmetrical motor exam. Within this group, 14 patients have undergone a TCR; 10 have had a TCR for a worsening of their scoliosis. Forty-three percent (15/35) of the patients with scoliosis eventually required a spinal fusion; 13/15 patients (86%) fused had a thoracic motor level.

**Latex Allergy**

In the study cohort, 23 patients (32%) have had an allergic reaction to latex. Forty-two patients are not allergic to latex and 6 patients have an unknown latex allergy status. Six of the 23 latex-allergic patients have had a severe, life-threatening anaphylactic reaction. The average age at which the patients became latex allergic was 12.5 years. When comparing the average number of shunt revisions, the patients who are latex-allergic have undergone 5.9 revisions, whereas those who are not latex-allergic have undergone 2.9 revisions.

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**Table 1. Preoperative tethered cord symptoms for the 23 patients who underwent a release and the postoperative results**

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<td>Scoliosis</td>
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<td>Decrease muscle strength</td>
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<td>Gait changes</td>
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<td>Spasticity</td>
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<td>Urinary changes</td>
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¹ Decline in motor function in 1 patient. LE = lower extremity.

**Posterior Cervical Decompression/Tracheostomy/Gastrostomy**

Within the study group, 4/71 (6%) patients have undergone a posterior cervical decompression. 4 (6%) have a tracheostomy and 3 (4%) have a gastrostomy tube. In the group of patients who died, 4/28 (14%) patients had a posterior cervical decompression, 8 (28%) had a tracheostomy and 7 (25%) had a gastrostomy tube. Of the group that has been lost to follow-up, there were no posterior cervical decompressions, tracheostomies or gastrostomies performed at the time of their last visit.

**Discussion**

Over the last 30 years, continual progress has been made in the outlook for patients with a myelomenigocele. Very little information is known about the long-term prognosis and late problems encountered by this group of patients. The purposes of this report is to review the outcome of young adults born with a myelomeningocele who were treated in a nonselective, prospective manner.

Twenty-four percent (28/118) of our original cohort have died. A majority of the deaths (18/28) occurred during infancy and the preschool years (fig. 4) [2, 4, 5]. Most of these children died secondary to hindbrain dysfunction (13/18). As expected, the cohort who died has a higher rate of posterior cervical decompression with tracheostomies and/or gastrostomy tubes. During school-age and early teen years, the death rate seemed to level off [4]. We felt the death rate had reached its peak. Unfortunately, this impression was wrong: the death rate has continued to climb into early adulthood with the commonest cause...
of death being an unrecognized shunt malfunction [McLone, unpubl. data].

In the series by Hunt et al. [6–8] of British patients with a myelomeningocele followed into early adulthood, 56% have survived into their 20th year. The commonest cause of death in this group of patients is renal failure. At a recent 30-year review of this cohort, Hunt [9] calculated a 1% risk of death per year from age 5 to 30 for all patients with open spina bifida.

We have previously assumed that the patients who were lost to follow-up tend to have fewer neurologic deficits, and, thus, less of a need for our multidisciplinary care. In the patients lost to follow-up, 67% have an L5 or sacral motor level as opposed to 1/3 of the children in the study group or those who have died. At the time of their last follow-up, only 47% had CSF diversion as compared to 86% in the study group and those who died. There were no reported posterior cervical decompressions, tracheotomies or gastrostomy tubes in the group lost to follow-up.

Within the study group, a majority of patients (73%) have maintained a stable motor exam into early adulthood. Eleven percent of patients have improved motor strength and 16% have declined by at least one motor level. As expected, those patients with the better motor strength (L5/sacral level) continue to ambulate into early adulthood. This finding is supported by Hunt and Poulton [6, 7], who found that all adult patients with an L5 or sacral level are ambulators.

As a group, 46% of young adults ambulate a majority of the time (75–100%). We have found that those patients who remain mobile in their teen years continue to ambulate in their young adult years. Although a number of investigators have also noted this relationship [10], McDonald et al. [11] found a correlation between decreased mobility and strong iliosposas/quadriceps with weak gluteus medius, with no relationship to age.

With the strong belief that any neurologic, orthopedic or urologic decline is likely to be secondary to a shunt malfunction, our shunt revision rate is quite high [12]. Ninety-five percent of patients have undergone at least 1 shunt revision. Most patients (41%) have undergone 2–3 revisions. Twenty-two patients have experienced a shunt infection during childhood.

After ruling out a shunt malfunction, a tethered spinal cord is the second commonest reason for loss of function in a patient with a myelomeningocele [13–16]. Symptomatic retethering of the spinal cord at the original closure site occurred in 23/71 patients, or 32%. Three patients required multiple untetherings. There is no association between the myelomeningocele level and subsequent development of a tethered cord. The commonest presenting symptoms of a tethered spinal cord are scoliosis, gait changes and increasing spasticity. Following the TCR, 97% of patients were either improved or had a stabilization of their symptoms. Two patients or 3%, were worse after their TCR. One had a decline in his gait after a TCR at an outside institution, and another had an improvement in his scoliosis, but a decline in his motor level postoperatively.

Of the 71 patients, 35 young adults (49%) have some degree of scoliosis. Fifty-one percent of these patients have a thoracic motor level. Of the patients eventually requiring a spinal fusion (15/35), 86% (13/15) have a thoracic level. The overall percent of patients with scoliosis is lower than reported by Pigott [17] (82.5%); although, our results are in agreement that those with the worse neurologic condition tend to develop severer neuromuscular imbalances.

All 16 patients who have had seizures have shunted hydrocephalus. Although the numbers are small, there is a high shunt infection rate amongst this group of patients as compared to the shunted study group.

Eighty-five percent of young adults are attending or have graduated from high school and/or college. Sixty-three percent of patients attend regular classes; whereas, 37% require some or all special education classes. Thirty-two young adults are actively employed and 5 have volunteered their services. Most of the study cohort reside with their parents; however, 11 live independently and 2 are married.
In her 30-year follow-up report, Hunt [9] found a very interesting correlation between those young adults who were socially achieving (i.e. living independently, driving a car, employed or attending college) and had no prior history of raised intracranial pressure. She urges early shunt evaluation/revision to possibly prevent deterioration in the motivation and stamina necessary to overcome the disability hurdles these young adults face.

Achieving social bladder and bowel continence is a realistic goal. More than 80% of young adults are continent a majority of the time. CIC improves bladder continence. A majority of the patients on CIC perform their own catheterization (90%). We have had no deaths secondary to renal failure, and none of the patients have undergone a renal transplant.

Approximately 1/3 of the study cohort is allergic to latex. Six patients (8%) have had a severe life-threatening reaction to latex. As reported by others, the risk of becoming latex allergic increases as the number of surgical procedures increases [18–20]. Those study patients allergic to latex have had twice as many shunt revisions as those without a latex allergy. Currently, our policy is to treat all patients with myelodysplasia with latex precautions. For those patients who have had a life-threatening reaction, corticosteroids are administered preoperatively. We strongly encourage any patient who has had a severe latex reaction to carry an epinephrine auto-injection device at all times.

Conclusion

At birth, a child born with a myelomingingocele requires a life-long commitment by the patient, family and treating medical personnel. The medical goal is to always maintain stable neurologic functioning throughout the patient’s lifetime. At least 75% of children born with an open spina bifida can be expected to reach their early adult years.

Although it is not the natural history, late deterioration in patients with myelodysplasia is common. Once deterioration is noted, a rigorous search for the cause must be undertaken. For a majority of patients, the decline in neurologic function is secondary to a CSF shunt malfunction. However, this population of patients has a high incidence of multiple medical problems, including, but not exclusive to, the following: pressure sores, obesity, severe renal disease, hypertension, depression and visual impairment [9]. Physicians must be prepared to care for this unique group of young adults with chronic medical conditions.

One of the greatest challenges in medicine today is establishing a network of care for these adult patients with spina bifida. Currently, they are a population of patients without adequately coordinated medical care and poor medical insurance coverage. Kaufman et al. [21] have demonstrated that disbandment of multidisciplinary medical care for adults with spina bifida has lead to a much higher morbidity and rate of preventable operations, such as amputations and nephrectomies. Hopefully, the future generation of physicians and legislators will actively support and care for this growing number of young adults who have thus far survived this most complex congenital abnormality.

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References


