Research Paper

Outcome and life satisfaction of adults with myelomeningocele

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Abstract

\textbf{Background:} Myelomeningocele (MMC) commonly causes impairments in body structure and functions as well as cognitive disabilities that can have an adverse effect on adult life. Improved medical care has resulted in increased numbers of individuals with MMC surviving to adulthood, however little is known about the impact of MMC on the lives of adults age 25 years or older.

\textbf{Objective:} To gain a better understanding of outcomes in education, employment, relationships, reproduction and life satisfaction of adults with MMC.

\textbf{Methods:} A primarily quantitative multiple-choice questionnaire designed to capture outcomes in education, employment, relationships and reproduction, along with a previously validated life satisfaction checklist (LiSat-11), was completed by adults with MMC. Relationships between demographic variables, outcomes and life satisfaction were determined using cross tabulation analysis, logistic regression and linear regression.

\textbf{Results:} Ninety adults with MMC, age 25–85 years (median age 32), reported a diverse range of outcomes in education, employment, relationships and reproduction. The most consistent variable associated with difficulty attaining adult milestones was hydrocephalus, the presence of which reduced the likelihood of living independently ($p < 0.001$), having a partner ($p = 0.003$) and reproducing ($p < 0.001$), but did not contribute to reduced life satisfaction.

\textbf{Conclusions:} Adults with MMC, especially those without hydrocephalus, can obtain gainful employment, live independently, form partner relationships and have children, and these achievements contribute to life satisfaction. While MMC does not affect overall reported life satisfaction for adults, attention should be paid to specific domains with less reported satisfaction.

\textbf{Keywords:} Myelomeningocele; Spina bifida; Adults; Life satisfaction

Myelomeningocele (MMC), the most common type of spina bifida, is a neural tube defect (NTD) resulting from failure of the caudal end of the neural tube to close by roughly day 28 post ovulation.\textsuperscript{1,2} With the introduction of immediate spinal closure and shunting for hydrocephalus in newborns with MMC in the 1960s, the survival rate increased dramatically from approximately 10% to an estimated 46–75% of individuals with MMC surviving into adulthood today.\textsuperscript{3–7} Nearly 25,000 children and adolescents with spina bifida were estimated to be living in the United States in 2002, the majority of whom will reach adulthood in the coming years.\textsuperscript{8}

Despite improved medical treatment, individuals with MMC often have lasting cognitive disabilities, impairments in body structure such as scoliosis, and impairments in body functions including muscle weakness, paralysis and bowel and bladder incontinence, all of which can have an adverse effect on adult life.\textsuperscript{5,9} Predictors of long-term outcome, such as hydrocephalus, continence, lesion level, IQ, gender and mobility have been investigated in European and North American populations, though previous

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Abstracts of the data have been presented at the NSGC 30th Annual Education Conference and the 7th International Conference on Neural Tube Defects. A brief synopsis of results was posted on the Duke Center for Human Genetics website as part of a newsletter for study participants.

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research has been limited and results are conflicting. The presence of hydrocephalus, which occurs in approximately 80% of individuals with MMC, significantly increases the likelihood of cognitive impairment and is the most commonly reported variable associated with poor outcome.

The vast majority of MMC outcome research to date has been conducted either with populations of young adults, a distinct developmental group between the ages of 18 and 25 years, or in broad populations consisting of adolescents, young adults and adults. While these data are informative, these studies are not readily transferrable to adults with MMC. Adolescents and young adults may not have had adequate time to achieve life goals, have less medical complications and better preserved mobility than adults. Impairments in body structure and functions such as scoliosis, renal damage and pressure sores often worsen with age and the likelihood of independent mobility decreases, which may reduce the ability of adults to actively participate in society.

After extensive literature review, only three outcome studies were identified in which the study population consisted exclusively of adults with MMC, defined as individuals age 25 years or older. The purpose of this study is to gain a better understanding of outcomes in education, employment, relationships, reproduction and life satisfaction of adults with MMC.

As individuals with MMC reach adulthood, they face challenges in attaining milestones commonly used to indicate positive outcomes, such as obtaining postsecondary education, employment, living independently and forming partner relationships. Previous research indicates that a reduced number of young adults with MMC pursue postsecondary or graduate education. Individuals with MMC also have significantly higher unemployment rates than the general population and those who are employed tend to hold low-wage jobs. Individuals with MMC report a typical desire to live independently, have romantic relationships, get married and even have children. However, research has shown that the majority of individuals with MMC require some form of living assistance, have difficulty forming romantic relationships and have low rates of marriage. Though the majority of women with MMC do not have children, there is no evidence suggesting decreased fertility in this population. Women with MMC have normal menstruation, are able to become pregnant and have healthy babies, however, unique concerns for these women have been reported including recurrent urinary tract infections, worsening kidney function, premature labor and postoperative complications. In contrast to women, data show that men with MMC have reduced fertility but a proportion, typically those with lower level lesions, can successfully reproduce. Additionally, both men and women with MMC are at increased risk to have a baby with an NTD. Preconception folic acid supplementation may reduce this risk by up to 70%.

Despite physical burdens and social difficulties, research suggests that young adults with MMC are as satisfied with their lives as typically developing peers. However, certain aspects such as finances, self-care ability and partner relationships are often reported to be less satisfying. Unfortunately, life satisfaction research on adults with MMC is minimal, which leads to unclear expectations for the future.

Methods

Participants

Participants were recruited from the Hereditary Basis of Neural Tube Defects study conducted at the Center for Human Genetics at Duke University Medical Center. Ascertainment of participants occurred across the United States from 1994 until 2007 from a variety of sources including spina bifida clinics, support groups and self-referral in response to advertisements. Adults age 25 years or older with a reported diagnosis of MMC were contacted for participation in this study. From a pool of over 5000 participants, 325 were eligible to participate in this study. Consent was obtained from 124 individuals (38%) and 99 (30%) completed the questionnaire. Nine participants were excluded for reporting a diagnosis other than MMC, leaving 90 for analysis. There were no significant differences between responders and non-responders in regards to gender, race, age, lesion level or hydrocephalus status. IRB approval was obtained from Duke University Medical Center, as well as the University of North Carolina at Greensboro.

Measures

A primarily quantitative, 77-item multiple-choice questionnaire, which consisted of four outcome sections (education, employment, relationships and reproduction) was developed by the authors using published literature and previously developed questionnaires obtained from other research groups. Qualitative items were used to capture postsecondary degrees, job titles, partner disabilities and pregnancy complications. Additionally, participants were asked to describe why they were or were not concerned about having a child with an NTD. The questionnaire was pilot-tested on two adults with MMC and five health care professionals who either had experience caring for patients with MMC or in questionnaire development.

The LiSat-11, a self-administered life satisfaction checklist, was used to correlate outcomes in education, employment, relationships and reproduction to life satisfaction. Participants rated satisfaction with 10 life domains (life as a whole, employment, finances, leisure, contact with
friends and acquaintances, self-care, family life, partner relationships, physical health, and mental health) along a six-point scale from 1 (very dissatisfying) to 6 (very satisfying). The sex life domain was not included. The LiSat-11 was previously validated in a population of randomly selected Swedish adults, from which reference values exist.\textsuperscript{41}

**Procedure**

Participants completed the questionnaire either online or via mailed paper copy. Medical records were used to confirm the diagnosis of MMC, lesion level and presence or absence of hydrocephalus. If medical records and patient report contradicted, data from medical records were used. Medical records were not available on all participants (n = 24, 27\%) and, in those cases, patient report was not verified. Qualitative items were coded into categories by two independent researchers. Coding disagreements were resolved through discussion with a third researcher until a consensus agreement was reached. Job titles were coded based on U.S. Bureau of Labor Statistics categorizations.\textsuperscript{42}

**Statistical methods**

Data analysis was completed in SAS version 9.2 (SAS Systems, Cary, NC). Frequencies of each variable were calculated and summarized (PROC FREQ). Cross tabulation analysis was then used to determine the relationships between outcome and different variables (gender, lesion level, age (born before or after 1960), hydrocephalus, continence and mobility) in each section. Chi-square tests were used to determine statistical significance between these differences. Analysis of LiSat-11 was done using previously reported methods.\textsuperscript{41} Domain scores were summed and an average score was calculated for each. Analysis of Variance (PROC GLM) was used to determine significant differences in life satisfaction domains between demographic data and other variables.

To determine the effect, if any, of co-variance in the models, several variables were considered (age, gender, hydrocephalus, mobility, continence and lesion level). Bivariate regression was used to assess the association among these potential predictors and to limit collinearity. Consequently, age, mobility and lesion level were not included as they were all significantly associated with hydrocephalus. Multivariate analysis was subsequently performed with gender, hydrocephalus and continence included as co-variates. Logistic regression (PROC LOGISTIC) was employed for analysis of categorical outcomes while linear regression (PROC GLM) was used for continuous outcomes. A \( p \) value of <0.05 was used to determine statistical significance for all data analyzed. Unless stated, reported \( p \) values were derived from univariate analyses which remained significant after controlling for co-variates.

**Results**

Most participants completed the questionnaire themselves (88\%) with the remainder completed by a parent (12\%). All questions were voluntary; therefore the sum does not equal 90 for all items.

**Demographics**

Demographic characteristics of the study group are summarized in Table 1. Participants were 25–85 years old with a median age of 32 years. Participants were primarily white and evenly split between male and female. The majority of participants had lumbo/sacral lesions and required a shunt for the treatment of hydrocephalus. Twelve participants (13\%) were born before 1960, none of whom had hydrocephalus. When controlling for co-variates such as hydrocephalus, the only difference between participants born before and after 1960, was a greater likelihood of the older group having children. Otherwise, there were no differences in outcomes based solely on age. Nearly half of all participants required a wheelchair for mobility. Most participants did not have complete bowel and bladder continence (defined as less than one bowel or bladder accident per month).

**Education**

Table 2 summarizes the highest level of education obtained for the study group. The vast majority of participants (94\%) completed high school or a high school equivalent (GED). The majority of participants (62\%) completed their high school education in a typical classroom setting, 13\% required an aide or extra one-on-one assistance, 10\%...
There was no association between gender and ability (35%). Most participants earned a salary of less than $25,000 per year, which is below the national average of $51–$75,000 per year (1%). Employment included health issues (42%) and job availability (35%). Most participants earned a salary of less than $25,000 per year, which is below the national average of $29,730. There was no association between gender and employment. Of those participants, only one was currently searching for employment. Of participants who never held a job, 82% attributed this to physical or medical limitations and 45% cited learning disabilities.

### Employment

Participants primarily received income from disability benefits (59%) and/or their own employment (48%). The majority of participants (85%) held a job at some point during their life with just under half (48%) currently employed. Participants reported a wide variety of job titles including front desk clerk, medical transcriptionist, cashier, social worker, certified public accountant (CPA) and elementary school teacher. Job categories were as follows: administrative support (28%), professional and technical (24%), executive, administrative, and managerial (11%), handlers, equipment cleaners, helpers and laborers (11%), service (10%), sales (10%), precision production, craft, and repair (4%), and transportation and material moving (1%).

Table 3 summarizes key characteristics of participants’ current or most recent jobs. Approximately half of working participants (51%) worked part-time (less than 40 h per week). The most common reported reasons for part-time employment included health issues (42%) and job availability (35%). Most participants earned a salary of less than $25,000 per year, which is below the national average of $29,730. There was no association between gender and employment. Of those participants, only one was currently searching for employment. Of participants who never held a job, 82% attributed this to physical or medical limitations and 45% cited learning disabilities.

### Relationships

Table 4 summarizes the living situation of the study group. Nearly half (43%) of participants still lived with their parents and 21% lived alone with or without assistance. Individuals who were independently mobile ($p = 0.003$) and those without hydrocephalus ($p = 0.0001$) were more likely to live independently (alone or with a partner, spouse or roommate). There was no association between participants’ living situations and continence, lesion level, age or gender.

Table 4 also summarizes participants’ romantic relationship histories. Over half (52%) of participants never had a serious partner or spouse, while 28% had been married. While the rate of marriage was low, for those who did marry, the age of marriage was consistent with the general population. Seven participants (17%) reported having a partner with a disability including NTDs, blindness and cerebral palsy. Individuals without hydrocephalus ($p = 0.003$) and with independent mobility ($p = 0.001$) were more likely to be in a relationship. There was no association between relationship status and age, gender, lesion level or continence.

Of participants who never had a partner, 20% reported a desire for a serious partner or spouse someday while 64% were unsure.

### Table 2

Highest completed education ($N = 90$)

<table>
<thead>
<tr>
<th>Education Level</th>
<th>n</th>
<th>%</th>
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<tr>
<td>No formal schooling</td>
<td>3</td>
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<tr>
<td>Some high school</td>
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<td>2</td>
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<tr>
<td>High school/GED</td>
<td>29</td>
<td>30</td>
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<tr>
<td>Some college</td>
<td>10</td>
<td>10</td>
</tr>
<tr>
<td>Technical or associates degree</td>
<td>18</td>
<td>19</td>
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<tr>
<td>College degree</td>
<td>33</td>
<td>35</td>
</tr>
<tr>
<td>Advanced/graduate degree</td>
<td>4</td>
<td>4</td>
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</table>

### Table 3

Job characteristics (current or most recent)

<table>
<thead>
<tr>
<th>Number of years at job ($N = 74$)</th>
<th>n</th>
<th>%</th>
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<tr>
<td>&lt;1 year</td>
<td>19</td>
<td>26</td>
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<tr>
<td>1–5 years</td>
<td>27</td>
<td>36</td>
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<tr>
<td>6–10 years</td>
<td>15</td>
<td>20</td>
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<tr>
<td>&gt;10 years</td>
<td>13</td>
<td>18</td>
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<table>
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<tr>
<th>Hours worked per week ($N = 73$)</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
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<tr>
<td>1–9 h</td>
<td>14</td>
<td>19</td>
</tr>
<tr>
<td>10–19 h</td>
<td>10</td>
<td>14</td>
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<td>20–29 h</td>
<td>14</td>
<td>19</td>
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<td>30–39 h</td>
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<td>4</td>
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<tr>
<td>40 h</td>
<td>23</td>
<td>31</td>
</tr>
<tr>
<td>&gt;40 h</td>
<td>13</td>
<td>18</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Salary ($N = 71$)</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;$25,000 per year</td>
<td>51</td>
<td>72</td>
</tr>
<tr>
<td>$25,000–50,000 per year</td>
<td>12</td>
<td>17</td>
</tr>
<tr>
<td>$51,000–75,000 per year</td>
<td>4</td>
<td>6</td>
</tr>
<tr>
<td>$76,000–100,000 per year</td>
<td>4</td>
<td>6</td>
</tr>
<tr>
<td>&gt;$100,000 per year</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>
Reproduction

Table 5 summarizes participants’ reproductive histories. Sixteen participants (18%) had biologic children. All but one participant with biologic children, including all eight men, had lumbo/sacral lesions. Of the resulting 34 children, three (9%) had an NTD, born to two women and one man. This observed recurrence of NTDs in offspring is above the empiric recurrence risk due to recruiting participants from an NTD genetics study. The majority (80%) of participants (females or female partners of male participants) with biologic children were more likely to be concerned about the effects of pregnancy on their own health. Nearly half (43%) reported a desire to have a child due to physical or learning disabilities (15%) and infertility (7%). Nearly half (41%) reported a desire to have biologic children in the future.

Of participants with no biologic children, reported reasons for not having children were not having a partner/spouse (63%), choice (38%), not able to raise a child (14%), and infertility (7%). Nearly half (41%) reported a desire to have biologic children in the future.

Over half (57%) of participants were concerned about having a child with an NTD. Concerned participants fell into two categories; those that didn’t feel physically capable of caring for a child with a disability and those that didn’t want their child to suffer as they had suffered. Participants who were not concerned (43%) also fell into two categories. Unconcerned participants either felt that the risk of having a child with an NTD was low or that their own experiences living with MMC prepared them to parent a child with an NTD.

Life satisfaction

Table 6 summarizes LiSat-11 mean domain scores. Overall, satisfaction with self-care and family life were the highest, while satisfaction with partner relationships, finances and employment were the lowest. When compared to reference values from an adult population, participant satisfaction with life as a whole was equivalent. However, the domains of employment, contact with friends and acquaintances, self-care, partnership relationships, physical urinary tract and kidney infections and increased difficulty with mobility and continence. Of participants with biologic children, two men (25%) and two women (25%) reported difficulty conceiving. Two additional women (20%) and one man (11%) attempted to conceive without success. One participant adopted her spouse’s children from a previous relationship.

Of participants with no biologic children, reported reasons for not having children were not having a partner/spouse (63%), choice (38%), not able to raise a child due to physical or learning disabilities (15%) and infertility (7%). Nearly half (41%) reported a desire to have biologic children in the future.
health and mental health were all rated significantly lower than the reference group. Associations between participant characteristics and increased domain scores are reported in Table 6. No differences in life satisfaction were seen based on gender, lesion level or age.

**Discussion**

Participants were exclusively adults age 25 years or older with MMC with a median age of 32 years; which is a more homogeneous adult population than has often been previously described.\(^3\) The presence of hydrocephalus and lack of independent mobility contributed to difficulty attaining adult milestones while lesion level, gender and continence were not determining factors, which is consistent with some previous reports of young adults.\(^10\) Recent research demonstrated the effectiveness of in-utero surgical repair in reducing rates of hydrocephalus and improving motor outcomes.\(^4\) As these two variables seem to have the greatest impact on adult outcome, additional research into in-utero repair is warranted as the potential exists to significantly improve outcomes for future generations of adults with MMC.

In contrast to previous reports, when compared to the general population of adults 25 years of age and older in the United States, adults with MMC in the study group were more likely to complete high school, obtain technical or associates degrees and graduate college.\(^4\) This could partly be explained by the young age of individuals included in previous research, who may not have had adequate time to complete their education. Individuals who received special education in high school were less likely to pursue education beyond high school, indicating that this group may be in particular need of assistance with obtaining postsecondary education. Participants who pursued postsecondary education often obtained degrees in the social sciences or humanities. Learning disabilities, specifically in problem solving and mathematics, in individuals with MMC has been reported; which may account for the lower number of degrees in these areas.\(^4\) To optimize educational success, it may be advantageous for individuals with MMC to recognize common cognitive strengths and focus postsecondary education efforts in these areas.

Participants in the study group reported diverse employment experiences. Some participants held competitive, even executive, jobs and earned salaries well above the national average. However, the majority of adults with MMC in the study group reported an annual salary below the national average, which may be partly explained by the large proportion of participants in part-time employment. Participants who were employed reported significantly higher vocational and financial satisfaction. This highlights the importance of employment on adult life satisfaction and the need for additional resources to assist in finding employment which can accommodate individuals with cognitive disabilities and/or impairments in body structure and functions.

Participants with hydrocephalus and those lacking independent mobility experienced the greatest difficulty in living independently and forming romantic relationships. Several participants reported happy, healthy marriages and relationships, and this contributed significantly to life satisfaction. Overall, partner relationships were rated the least satisfying domain, indicating the need for additional attention to this aspect of adult life. Previous reports indicate that individuals with MMC have reduced social contact, which is clearly a barrier to developing romantic relationships.\(^2\) Adolescents and adults with MMC should be encouraged to participate in social activities both with non-disabled peers as well as other individuals with MMC or similar disabilities. Involvement in online groups, which were not available to adults with MMC in the past, is not limited by physical disabilities and may increase opportunities for social contact.

While men with MMC are reported to have reduced fertility, similar frequencies of successful pregnancies and infertility were reported by men and women, though the number of participants who attempted pregnancy was low.\(^3\) A reduced number of participants who conceived after the benefits of folic acid were known reported taking the therapeutic dose. Both women and men with MMC should be referred to a reproductive health specialist if they are considering pregnancy to discuss recurrence risks for NTDs, prenatal diagnostic options, and folic acid supplementation.

Women with MMC experienced increased frequencies of pregnancy complications, indicating a need for these
women to be monitored by high-risk obstetricians during pregnancy. Half of women in the study group delivered preterm, confirming previous reports of increased risk for preterm labor.\textsuperscript{34} There is mention in the literature that preterm, confirming previous reports of increased risk for pregnancy. Half of women in the study group delivered women to be monitored by high-risk obstetricians during

When compared to a reference group, adults with MMC in the study group were equally satisfied with life as whole, though several domains including partner relationships, finances and employment, were rated significantly lower. This suggests that while MMC does not affect overall life satisfaction for adults, actions are needed to increase satisfaction with specific aspects of adult life. Participants who lived with someone, including those residing in group homes, were significantly more satisfied with life as a whole than those participants living alone. While being self-sufficient is an important goal, living alone may contribute to decreased life satisfaction due to persistent social isolation. Isolated adults with MMC could consider seeking out roommates, perhaps by enlisting the help of local support group chapters.

There were several limitations in the present study. The twelve participants born before 1960 represent an especially fit group of individuals with MMC that may not translate to populations of adults born after 1960, when individuals with greater disabilities began to survive. However, multivariate analyses determined that the absence of hydrocephalus, rather than age alone, accounted for the generally positive outcome of these adults. Second, in some cases, a parent rather than the individual with MMC completed the questionnaire. Therefore, responses, especially in regards to life satisfaction, may not truly reflect those of the individual with MMC. However, a previous study examining quality of life in adolescents with MMC demonstrated that parent report closely mirrored self-report.\textsuperscript{38} Additionally, participants were recruited through an NTD genetics study and therefore often had an additional family member with an NTD. This study group may represent a population of individuals with more available resources or involvement in the community and may not represent all individuals with MMC. Lastly, the study had a relatively small sample size consisting of 90 adult participants with MMC.

This is one of the few outcome and life satisfaction studies on adults with MMC. Results of this study have practical implications for providing information about long-term outcomes for individuals with MMC. This information may be valuable for parents in making pregnancy decisions or to aid in developing realistic goals for adolescents with MMC in their transition to adulthood.\textsuperscript{25} This study also provides insight into areas of needed improvement in the care of adults with MMC. In particular, there is a clear need for information regarding reproductive issues for both men and women with MMC. Future research should focus on areas of reported difficulty and those domains with reduced life satisfaction to better understand the needs of adults with MMC.

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References


