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Functional outcome in young adults with spina bifida and hydrocephalus

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Abstract *Introduction:* Quality of life was studied in 31 adult survivors of spina bifida, each with Verbal or Performance IQ score over 70.

Methods: Instruments measured physical and occupational function, cognitive/psychological function, somatic sensation, and social interaction in the context of the ability to live independently. *Results:* Domain group means except motor independence were in the average range (± 1 standard deviation). Variability within the group with respect to physical phenotype (high spinal lesions were associated with poorer motor independence) and medical

history [a greater number of shunt revisions (>4) were associated with poorer functional numeracy] was predictive of quality of life. Both spinal lesion level and number of shunt revisions influenced occupational status. *Discussion:* There was a relation between everyday mathematics, reading skills, and quality of life. The consequences of the physical, medical, and cognitive effects of spina bifida extend into young adulthood and have an impact on quality of life.

Keywords Spina bifida · Hydrocephalus · Quality of life

Introduction

Neurodevelopmental disorders, evident at birth and affecting physical, cognitive, and adaptive function throughout the childhood years, are also likely to influence quality of life outcome in adulthood. However, relatively, little is known about the adult outcomes of neurodevelopmental disorders, even those like spina bifida with hydrocephalus (SBH) that occur quite commonly.

Spina bifida with myelomeningocele is a neural tube defect and the most common disabling congenital condition affecting the central nervous system. Individuals with spina bifida have primary brain dysmorphology [15] as well as secondary brain effects from hydrocephalus, which develop in nearly all of these myelomeningocele cases. Treatment with a diversionary shunt restores the gross brain volume but does not repair the axonal and neuronal damage [7].

Although infants with spina bifida survive with early treatment [26], SBH continues to influence physical, cognitive, and adaptive function in infancy and childhood. Children with SBH have physical and motor deficits that can limit mobility and occupational function, a characteristic set of cognitive deficits [14] and difficulties in a range of other activities of social and everyday function. Children with hydrocephalus from mixed etiologies, including a large number with SBH, display a discrepancy between a Verbal IQ score, typically in the average range, and a reduced Performance IQ [7, 11]. These children have difficulty with higher order language, inferencing, and social discourse tasks [10]. They have neuropsychological deficits in visual perception [9] and specific academic difficulties in mathematics [2].

Little is known about adult function in individuals with SBH. Some of the consequences of SBH for cognitive function in young adults are now being identified, but

adaptive function and quality of life in young SBH adults are poorly understood. To the extent that physical and cognitive problems of SBH children persist into adulthood, the quality of life of adults with SBH would be compromised.

The World Health Organization defines health as the presence of physical, mental, and social well-being and not merely as the absence of disease. A universally accepted definition of quality of life may prove elusive [21]; however, there is general agreement that quality of life is multidimensional, that each dimension changes over time, and that it is a patient-perceived entity [30]. Much quality of life research has focused on four central domains: physical and occupational function, somatic sensation, cognitive/psychological function, and social interaction. Some information is available on the effects of SBH on quality of life in these domains in children and adults.

Physical and occupational Physical problems are common among children and adults with SBH not only from spinal lesions but also from brain dysmorphology. Problems related to spinal lesions include bladder and bowel problems, loss of skin sensation, spinal deformities, and muscle paralysis [22], which persist into adolescence [16] and impede emotional as well as physical maturity. In a large sample that included children with spina bifida, hydrocephalus, and SBH, all groups experienced reduced health-related quality of life relative to children with chronic physical disorders or psychiatric disorders; however, children with SBH fared more poorly than the other two groups with respect to mobility and continence [6]. Children with SBH have limited fine and gross motor skills, with many requiring mobility aids [17]. Lower limb deficits have been related to higher disability ratings and the physical aspect of quality of life [28] in a small group of adolescents with spina bifida. Adolescents with spina bifida also experience doubts about their health and physical condition, with over half of one group reporting physical problems [16] that are not related to objective markers of disease severity. Interestingly, parental hopefulness has been shown to contribute significantly to the variance in health-related quality of life for children and adolescents with spina bifida [23].

Employment is a significant concern for young adults with SBH [5]. Less than one third of adolescents and young adults with treated SBH are employable, while approximately three-quarters were judged employable with accommodations [19]. Vocational training and counselling are among the perceived needs of adults with spina bifida and their families [22, 33].

Somatic sensation Although many children and young adults with SBH have physical challenges, the natural course of the disease is not characterized by primary pain. Discomfort may arise as secondary to the use of mobility aids, pressure sores, or from procedures such as catheterization (e.g., [19]).

Cognitive/psychological function The cognitive profile evident in children with SBH appears to persist into adulthood. The profile of better Verbal than Performance IQ persists in young adults [18]. Hydrocephalus itself increases the chance of poor cognitive outcome in adults with spina bifida. Neuropsychological function of young adults with SBH was reduced relative to those with spina bifida only [31]. Childhood cognitive deficits that persist into adulthood have implications for educational achievement and likely for employment as well.

Behavioral and socioemotional problems are also of concern in the SBH population. Children with SBH are at risk for behavior problems and psychiatric disorders [13]. Children with spina bifida, with and without hydrocephalus, are also at risk for behavior problems [24]. Adolescents and young adults with spina bifida have poor self-esteem [16]. Spina-bifida-related stress was the most important predictor of quality of life in a sample of adolescents with spina bifida, the majority of whom had SBH [29].

Social interaction Spina bifida can result in social isolation for young adults and reductions in opportunities for relationships with members of the opposite sex [5]. Sociosexual adjustment is a concern. Few adolescents with SBH date regularly [16], and young adults with SBH express the need for sexual counselling [33]. Lack of opportunity to interact and compete with peers has been reported as a contributor to delayed maturation [16] and has been identified by adolescents and young adults with spina bifida and by their parents as a significant unmet need [22].

In brief, individuals with SBH reach adulthood with compromised physical function, impaired cognitive processes, reduced opportunity for full social integration and employment, and a variety of unmet needs. What contributes to quality of life in adults with SBH is unclear because of differences in the samples and designs of the published studies. Sources of variability include age, type of spina bifida, measures, and data presentation. Some studies have focused on adolescents only [16, 28], while others have included adolescents and young adults. However, the findings may be more representative of late adolescence (17–20 years of age) than adulthood, e.g., [5, 19]. Of the studies that have addressed outcome in adolescents and young adults with spina bifida or hydrocephalus, most have examined outcome in spina bifida with or without hydrocephalus [19, 28, 29] or hydrocephalus with spina bifida being one among several etiologies [18]. Level of cognitive impairment has been variable and sometimes has not been reported, blurring comparisons across studies. Employment, mobility, and independent living status are often reported as proportions of the sample studies rather than as continuous data based on more informative self-reports. Not all studies have considered outcome in relation to physical phenotype (e.g., level of spinal cord lesion) or to medical variables (e.g., number of shunt revisions). Most important, perhaps, is that no study has employed stand-

ardized self-report instruments to examine the subjective life experiences and quality of life of adults with SBH.

These issues were the focus of the present study, in which we explored the profile of functional outcomes in a group of adult survivors of SBH and examined relations between a physical phenotype (upper vs lower spinal lesions), a medical variable (number of shunt revisions), intelligence, functional literacy and numeracy, independent-living skills, quality of life, and range of experiences. The general hypothesis was that quality of life would be compromised because of restrictions related to physical phenotype and medical instability and also because of limitations of childhood skills required for adult function. Specific hypotheses were that:

- Physical phenotype (level of spinal cord lesion) would be related to motor independence and that degree of motor independence would influence quality of life through constraints on access to community, cultural, and employment resources, restricting the range of possible experiences.
- Medical instability (more lifetime shunt revisions) would produce a lower intellectual outcome, which in turn would be likely to influence the acquisition of functional literacy and numeracy skills in childhood and early adolescence. Acquisition of these functional skills, reflecting the ability to interact fluently within a cultural milieu, would be predictive of several aspects of quality of life, especially those involving social communication and community independence.

Methods

Participants

Data were gathered from 31 adults with spina bifida and shunted hydrocephalus who were taking part in a larger study examining neuropsychological and functional outcome. All participants gave informed consent for participation in the study. For inclusion in the study, all participants had a Verbal or Performance IQ score over 70.

The neural tube closes in successive stages at multiple sites, two of which may fail to close in spina bifida, an upper closure site, 1, and a lower closure site, 5 [34]. On the basis of birth records and medical chart reviews, two spinal lesion level groups were constituted, one ($n=8$) with upper lesions (L1 and above corresponding to van Allen's closure site 1) and the other ($n=23$) with lower lesions (L2 and below, corresponding to van Allen's closure site 5). The number of shunt revisions was recorded from the medical record and from interview data. Patients who had undergone four or more shunt revisions in their lifetime constituted the high revision group ($n=14$), and those with under four constituted the low revision group ($n=17$).

Demographic characteristics Sixteen males and fifteen females took part. Mean age of participants was 28.7, range 18.5 to 36.3. Six participants (four males and two females) were engaged, married or divorced and three had children. With respect to education, 11 participants had completed high school, 16 had completed all of a community college program or some of a community college program or undergraduate degree, 2 had completed undergraduate degrees, 1 had obtained a graduate degree, 1 had unavailable information. Participants were grouped on the basis of education into those who had some postsecondary education ($n=19$) and those who had completed secondary school only ($n=11$). At the time of the study, nine were unemployed, ten were students, working part-time or working as volunteers, nine were employed, and three had unavailable employment information. Three occupational groups were thus constituted.

Procedures

The following standardized instruments were administered over the course of a single testing session:

Wechsler Intelligence Scale for Adults–Revised [35]. This standard IQ test yields scores for Verbal IQ, Performance IQ, and Full-Scale IQ.

Kaufman Functional Academic Skills Test (K-FAST) [21]. The K-FAST tests functional academic skills in arithmetic and reading. The Arithmetic subtest includes 25 items that assess numerical reasoning, computation skills, and mathematical concepts. The topics covered in the items emphasize the everyday use of numbers for tasks such as baking, cooking, and reading graphical information. The Reading subtest consists of 29 items which require the examinee to read words, sentences, or paragraphs that are seen in everyday situations (e.g., Employees Only) or to interpret rebuses (e.g., the symbol for poison) or abbreviations (e.g., mo.). The Functional Academic Skills Composite score is a summation of the Arithmetic and Reading subtests. This score reflects the ability to use mathematics and reading to solve everyday problems and function effectively in their environment.

Scales of Independent Behavior–Revised (SIB-R) [4]. This standardized self-report assesses everyday living skills or adaptive behavior. Composite scores are derived for Motor, Social Language, Personal Living, and Communication, as well as an overall Broad Independence Index.

Quality of Life Questionnaire (QOL-Q) [21]. This self-report consists of 192 true/false items that yield composite scores for Material Well-Being, Physical Well-Being, Personal Growth, Marital Relations, Parent–Child Relations, Extended Family Relations, Extra-Marital Relations, Altruistic Behavior, Political

Behavior, Job Characteristics, Occupational Relations, Job Satisfiers, Creative Aesthetic Behavior, Sports Activity, Vacation Behavior, Social Desirability, and an overall Total Quality of Life score.

Life Experiences Checklist (LEC) [1]. This self-report measures the extent and range of life experiences. Composite scores are generated in Home, Leisure, Relationships, Freedom, and Opportunities domains as well as a Total score.

Data were analyzed using SPSS for Windows Release 11.01. Within each of the three relevant quality of life domains (physical function and occupation, cognitive/psychological function, and social function), relations among the appropriate indices from the standardized instruments (above) and medical and demographic variables were assessed using analysis of variance or nonparametric procedures. Effects of elements within each domain on overall quality of life were assessed by examining correlations. Finally, regression models were fitted to examine effects of multiple variables on overall life experiences and quality of life.

Results

Overview of results Tables 1, 2, 3, and 4 show major index and subscale scores for the cognitive and academic, independent living, quality of life, and range of life experiences instruments administered. On the majority of measures, the group scored within the average range (1 standard deviation of the mean). One notable exception was the Motor Independence Scale of the SIB-R, on which the group mean was very low.

Table 1 Cognitive ability, functional numeracy and literacy (Wechsler Intelligence Scale for Adults–Revised and Kaufman Functional Academic Skills Test)

| Variable | Total/♂/♀ | Lesion level | | Shunt revisions | |
|----------|--|------------------------------|-----------------------------|-----------------|-------|
| | | Lower/upper | <4/≥4 | <4/≥4 | <4/≥4 |
| <i>N</i> | 31/16/15 | 23/8 | 17/14 | | |
| VIQ | 95.4/95.1/95.7 ^a (9.1) (8.6) (9.9) | 94.3/98.9 (9.4)/(7.6) | 96.8/93.6 (8.8)/(9.5) | | |
| PIQ | 85.2/81.4/89.3* (10.2) (7.2) (11.6) | 86.6/81.4 (10.8)/(7.8) | 89.4/80.1** (10.5)/(7.4) | | |
| KF_A | 94.9/93.3/96.6 (12.6) (11.5) (13.9) | 92.9/100.5 (13.5)/(7.8) | 99.1/89.9* (11.2)/(12.8) | | |
| KF_R | 102.9/98.5/107.6* (12.8) (14.0) (9.9) | 100.4/110.3 (12.3)/(12.3) | 106.3/98.8 (10.0)/(15.0) | | |

VIQ significantly higher than PIQ

* $p < 0.05$; ** $p < 0.01$

^aStandard scores with a mean of 100 and a standard deviation of 15. Means and standard deviations

Table 2 Independent living skills (Scales of Independent Behavior–Revised)

| Variable | Total/♂/♀ | Lesion level | | Shunt revisions | |
|-----------------|---|-----------------------------|-------------------------------|-----------------|-------|
| | | Lower/upper | <4/≥4 | <4/≥4 | <4/≥4 |
| <i>N</i> | 29/14/15 | 22/7 | 16/13 | | |
| Broad Living | 86.3/85.2/87.1 ^a (26.9) (28.7) (26.4) | 92.1/65.7* 24.1/26.8 | 92.6/77.1 (25.7)/(27.1) | | |
| Motor | 54.7/55.6/54.0 (37.7) (41.8) (35.5) | 66.6/9.6** (32.0)/(13.2) | 61.6/44.7 (41.3)/(30.7) | | |
| Social | 101.3/99.9/102.7 (17.3) (17.7) (17.5) | 102.1/98.9 (18.3)/(14.6) | 104.1/98.0 (14.1)/(20.7) | | |
| Language | 97.8/94.4/101.1 (23.3) (20.7) (25.7) | 102.1/84.4 (21.2)/(25.9) | 107.9/85.5** (18.3)/(23.3) | | |
| Personal Living | 99.5/100.9/98.3 (24.3) (25.3) (24.2) | 100.5/94.3 (23.1)/(27.9) | 102.6/95.3 (20.2)/(29.3) | | |
| Communication | | | | | |

^aStandard scores with a mean of 100 and a standard deviation of 15. Means and standard deviations

* $p < 0.05$; ** $p < 0.01$

Physical function and occupation Physical function was compromised in the SBH group. Within the group, both physical phenotype and shunt revision history influenced physical function. Individuals with upper spinal lesions scored lower on the Motor Independence Index of the Scales of Independent Behavior [$F(1,27)=20.7, p < 0.0001$]. The Motor Independence Index [$F(2,23)=3.74, p = 0.04$] was related to employment status, and the lesion level was marginally related to employment status [$\chi^2(2)=4.98, p = 0.084$]. No individuals with high lesions were employed full-time. It is notable that while the mean score of the participants with lower lesions was still well below the average range of scores for those without handicapping conditions, they were nevertheless as a group able to work full-time. Occupational status was marginally associated with number of shunt revisions [$\chi^2(2)=0.50, p = 0.064$]. Only one individual with more than four shunt revisions was working full-time. Age differed across occupational status [$F(2,25)=8.50, p = 0.002$], with the mean age of the group employed part-time, volunteering or still in school (19), lower than that of those who were unemployed (25.1) or those who were employed full-time (22.6). Job Characteristics, Occupational Relations, and Job Satisfiers sub-indices from the QOL-Q were all rated in the normal range.

A greater number of shunt revisions were associated with a lower level of independence in Personal Living (eating, toileting, dressing, and self-care domestic skills) [$F(1,27)=8.43, p = 0.007$].

Physical function, as indexed by Motor Independence, level of lesion, or number of shunt revisions, was not related to the participants' overall experience of their quality of life or the range of their life experiences.

Table 3 Quality of life (Quality of Life Questionnaire)

| Variable | Total/♂/♀ | Lesion level | Shunt revisions |
|-------------------------------|---|------------------------------|------------------------------|
| | | Lower/upper | <4/≥4 |
| N | 31/16/15 | 23/8 | 17/14 |
| Total QOL | -0.64/-0.81/-0.47 ^a (1.2) (1.2) (1.2) | -0.51/-1.03 (1.3)/(0.90) | -0.39/-0.95 (1.2)/(1.1) |
| Material Well-Being | -0.95/-0.93/-0.98 (1.0) (1.0) (1.0) | -0.89/-1.13 (1.10)/(0.78) | -1.08/-0.80 (1.01)/(1.02) |
| Physical Well-Being | -0.26/-0.18/-0.34 (0.79) (0.75) (0.85) | -0.25/-0.27 (0.79)/(0.87) | -0.22/-0.29 (0.79)/(0.82) |
| Personal Growth | -0.42/-0.32/-0.53 (1.00) (0.98) (1.10) | -0.41/-0.45 (1.10)/(0.86) | -0.36/-0.48 (1.00)/(1.11) |
| Marital Relations (n=6) | -0.28/0.48/-0.66 (1.00) (0.50) (1.10) | -0.40/-0.05 (1.10)/(1.25) | -0.58/-0.01 (1.27)/(0.89) |
| Parent-Child Relations (n=3) | 0.41/0.02/0.61 (0.02) (0.61) (0.41) | 0.41/- (0.68) | 0.41/- (0.68) |
| Extended Family Relations | -0.19/-0.15/-0.25 (1.00) (0.88) (1.20) | -0.13/-0.38 (1.02)/(1.04) | -0.26/-0.12 (1.00)/(1.06) |
| Extra-Marital Relations | 0.22/-0.11/0.57 (1.10) (0.97) (1.10) | 0.26/0.12 (1.17)/(0.83) | 0.47/-0.08 (0.88)/(1.25) |
| Altruistic Behavior | -0.25/-0.42/-0.07 (0.99) (1.20) (0.74) | -0.28/-0.16 (1.03)/(0.95) | -0.29/-0.20 (0.99)/(1.03) |
| Political Behavior | -0.54/-0.37/-0.74 (1.10) (1.10) (1.01) | -0.34/-1.14 (1.10)/(0.91) | -0.24/-0.92 (1.10)/(1.01) |
| Job Characteristics (n=21) | 0.01/0.14/-0.09 (0.09) (0.82) (0.97) | 0.03/-0.07 (0.89)/(1.02) | 0.04/-0.03 (1.01)/(0.79) |
| Occupational Relations (n=19) | -0.12/0.19/-0.40 (0.96) (0.67) (1.13) | 0.008/-0.55 (0.96)/(0.97) | -0.13/-0.11 (1.14)/(0.78) |
| Job Satisfiers (n=20) | -0.79/-0.88/-0.73 (0.91) (1.05) (0.82) | -0.78/-0.88 (1.00)/(0.48) | -0.55/-1.11 (1.08)/(0.53) |
| Creative Aesthetic Behavior | -0.12/-0.53/0.35** (0.94) (0.90) (0.76) | -0.07/-0.19 (0.90)/(1.11) | 0.07/-0.32 (0.97)/(0.88) |
| Sports Activity | -0.49/-0.23/-0.77 (1.00) (1.10) (0.84) | -0.44/-0.63 (1.06)/(0.86) | -0.37/-0.63 (1.16)/(0.79) |
| Vacation Behavior (n=28) | -0.96/-1.27/-0.60 (1.12) (1.10) (1.08) | -1.05/-0.69 (1.20)/(0.91) | -0.96/-0.96 (1.14)/(1.15) |
| Social Desirability | -0.83/-1.06/-0.58 (1.04) (1.10) (0.95) | -0.84/-0.80 (1.06)/(1.03) | -0.89/-0.76 (1.20)/(0.83) |

^aZ-scores with a mean of 0 and a standard deviation of 1. Means and standard deviations

** $p < 0.01$

Cognitive/psychological function Cognitive abilities were below the theoretical population mean, with Verbal IQ in the average range and higher than Performance IQ which was in the low average range [$F(1,30)=29.11, p < 0.0001$]. Functional mathematics and reading skills were in the average range; however, reading skills were superior to functional math skills [$F(1,30)=17.03, p < 0.0001$]. Number of shunt revisions, but not level of spinal lesion, affected cognitive performance. A greater number of shunt revisions were associated with lower Performance IQ [$F(1,29)=7.75, p=0.009$] and functional math skills [$F(1,29)=4.58, p=0.04$]. Both Performance and Verbal IQ were correlated with functional mathematics skills [VIQ $r(29)=0.73, p < 0.0001$; PIQ $r(29)=0.58, p=0.001$].

Functional reading outcome was related to both VIQ and PIQ skills [VIQ $r(29)=0.64, p < 0.0001$; PIQ $r(29)=0.41, p=0.02$] and marginally to level of lesion [$F(1,29)=3.87, p=0.059$]. Interestingly, the group with upper lesions obtained a higher score than those with lower lesions. Women not only obtained better functional reading scores than men [$F(1,29)=4.32, p < 0.05$] but also obtained higher PIQ scores than men [$F(1,29)=5.18, p=0.03$].

Among the cognitive and functional skills indices, only Verbal IQ was related to attainment of postsecondary education [$F(2,28)=4.35, p=0.046$]. Verbal IQ, Performance IQ, [VIQ and PIQ $F(2,25)=3.63, p=0.41$], and functional math skills [$F(2,25)=4.86, p=0.016$] were related to occupational status, with lower scores associated with unemployed status.

Table 4 Range of experience (Life Experiences Checklist)

| Variable | Total/ $\text{♂}/\text{♀}$ | Lesion level | Shunt revisions |
|------------------------|--|-----------------------------|----------------------------|
| | | Lower/upper | <4/ \geq 4 |
| <i>N</i> | 31/16/15 | 23/8 | 17/14 |
| Total Life Experiences | 0.59/0.20/1.00 ^a ** (0.81) (0.89) (0.44) | 0.62/0.52 (0.75)/(0.94) | 0.81/0.32 (0.63)/(0.94) |
| Home | 0.43/0.40/0.46 (0.60) (0.68) (0.52) | 0.40/0.48 (0.41)/(0.86) | 0.50/0.34 (0.47)/(0.73) |
| Leisure | 0.81/0.23/1.43** (1.23) (1.36) (0.68) | 0.83/0.79 (1.21)/(1.32) | 1.08/0.49 (1.15)/(1.28) |
| Relationships | 0.12/0.09/0.15 (0.62) (0.63) (0.63) | 0.18/0.02 (0.61)/(0.94) | 0.21/0.02 (0.69)/(0.52) |
| Freedom | 0.47/0.14/0.82** (0.73) (0.77) (0.51) | 0.39/0.61 (0.81)/(0.58) | 0.62/0.28 (0.55)/(0.89) |
| Opportunities | 0.23/-0.16/0.73** (0.88) (0.87) (0.65) | 0.44/-0.03 (0.83)/(0.94) | 0.46/0.05 (.81)/(0.94) |

^aZ-scores with a mean of 0 and a standard deviation of 1. Means and standard deviations

** $p < 0.01$

Cognitive skills were not related to overall Quality of Life (QOL-Q), but there was a relation between functional math skills and overall Quality of Life [$r(29)=0.36$, $p=0.047$]. Total range of experiences (LEC) was related to Performance IQ [$r(29)=0.45$, $p=0.012$], functional reading [$r(29)=0.56$, $p=0.001$], and functional mathematical skills [$r(29)=0.42$, $p=0.018$].

Somatic sensation Participants' sense of Physical Well-Being (QOL-Q) was well within the average range. Level of spinal lesion, number of shunt revisions, and score on the Motor Independence Index were not related to Physical Well-Being.

Social interaction On the QOL-Q, participants rated their Extended Family Relations and their Extra-Marital Relationships within the normal range. They also rated their Altruistic and Political Behaviors in the normal range. Their range of Relationship Experiences, as rated on the LEC, was average. Sex, level of lesion, and number of shunt revisions were not related to social interaction variables.

Regression models for Life Experiences and Quality of Life Females had a broader range of Life Experiences than did males [$F(1,29)=9.69$, $p=0.004$]; however, when a regression model was tested with the medical and demographic variables that may have been salient given univariate results, functional reading skill accounted for 32% of the variance (Adjusted R_2) in Total Experiences [$F(1, 26)=13.7$, $p=0.001$], and no other variable, including sex, would have contributed to the explanatory power of the model.

The Broad Independence Index of the SIB-R was marginally related to the Total Quality of Life Index [$r(27)=0.34$, $p=0.072$]. Total Quality of Life Index from the QOL-Q and the Total Experiences Index from the LEC were correlated [$r(29)=0.43$, $p=0.015$]. Under the assumption that overall independence and range of experiences contributes to quality of life, a regression model was tested including the Broad Independence Index and the Total Experiences Index with candidate medical and demographic variables. Only the Total Experiences Index had any explanatory power in the model, accounting for 16% of the variance (Adjusted R_2) in Total of Quality of Life [$F(1.26)=6.08$, $p=0.021$].

Discussion

The range of challenges faced by individuals with SBH in the domains influencing quality of life is a significant public health concern. Establishing a network of care for adults with spina bifida represents an important challenge for health care systems [3]. Clinics dedicated to the care of adults with spina bifida and/or hydrocephalus have only been established recently, reflecting the survival of the first cohort who received intensive treatment of spina bifida and/or hydrocephalus during the 1960s. Surveys conducted at some of these clinics have demonstrated a range of unmet medical and psychosocial needs, including guidance and advice on work, education, social provision, and sexual counselling [33]. A survey of adolescents and young adults with spina bifida and their families using a standardized questionnaire supported the idea of unmet needs in these and other functional domains [22].

In the present study, cognitively capable young adults with SBH self-rated quality of life and range of life experiences within the average range in all major domains, despite the significant motor complications and milder cognitive complications associated with SBH. Objective markers of outcome were also encouraging for the majority in that all had completed secondary school, more than half the group had completed some postsecondary education, and the majority were employed or still in school.

Compromised motor function, typical of individuals with SBH, does not prevent development of a positive attitude towards sense of well-being. Despite a significant degree of compromise in Motor Independence for the majority of the group, they rated their Physical Well-Being in the average range. The tendency of some adolescents and young adults with significant physical challenges to self-report good levels of well-being or quality of life has been previously reported [16, 12].

Young adults with SBH rated the range and quality of their social interactions in the normal range. Only six in the group were engaged, married, or had been divorced at the time of the study; however, many members of the group were still in their early 20s and may not have had the

opportunity to become involved in serious relationships for reasons other than their medical status. The quality of social interactions for individuals with SBH into middle age and beyond remains to be studied.

Perhaps, the most important finding concerns the variability within the SBH cohort in outcome as a function of physical phenotype and medical history. In different ways, both spinal lesion level and number of shunt revisions affected adult outcome.

Upper spinal lesions produced poorer Motor Independence outcomes, in support of one of the specific hypotheses. Further, although self-reported quality of life or range of life experiences was not influenced by Motor Independence, employment status was related to lesion level and to Motor Independence. Approximately one third of the group was unemployed. No member of the high spinal lesion group had full-time employment.

Spinal lesion level is apparent at birth and is constant throughout life. Upper spinal lesions appear to affect employment through motor limitations, so presumably, the interventions required to ameliorate these effects could be instituted early in life. Future studies of outcome in SBH adults must take into account that the two spinal lesion level physical phenotypes might have different adult courses that could require different patterns of services to prepare for employment.

The effect of unstable medical control of hydrocephalus, as indicated by a greater number of shunt revisions, affected employment and personal living. As predicted, a larger number of shunt revisions were associated with reduced Performance IQ and difficulty in acquiring functional mathematical skills. Both variables were related to overall quality of life. Chronic medical instability, unlike spinal lesion level, cannot be identified until later in childhood or even in adult life. Other work has shown that shunt revisions after the age of 2 are associated with reduced achievement in independence and achievement in adulthood [19]. In terms of adult employment and quality of life, a medical priority is to minimize episodes of increased intracranial pressure by prompt shunt management.

The data also illustrate the importance of core literacy and numeracy skills, both of which were related to quality of life. Range of life experiences was associated with greater functional reading skills and range of life experiences predicted overall quality of life.

Numeracy was an important predictor of quality of life. While both reading and math are important for everyday function, it may be math that is the more relevant to personal independence. Unless one's job requires complex reading skills, a basic level of reading may be sufficient for everyday function. With poor numeracy skills, however, it is difficult to manage money, estimate amounts, or shop for oneself, which limits everyday independence. These data have implications for math instruction. The focus of academic math skills in the school-age years may not foster the degree of functional numeracy needed so that individuals with SBH may be independent in young adulthood. Whether interventions to promote acquisition of functional literacy and numeracy skills in childhood can enhance adult quality of life is an empirical question. Certainly, any such interventions should focus on mastery of basic math facts in the early grades and on math processes relevant to daily living skills, such as computation of sales tax and tips, estimation and comparisons, and deriving information from number graphics. Even with childhood interventions to ameliorate the effects of motor dysfunction or numeracy problems, it is possible that interventions for young SBH adults will continued to be required into adult life.

Finally, we have considered outcome in relation to several quality of life domains and have identified several relations among the variables analyzed. What remains to be studied is the causal relationships among these variables. As more individuals with SBH grow to adulthood, it will be possible to conduct studies with larger groups of participants, which will make it possible to assess the direction of causal flow among the variables relating to the quality of life in adults with SBH.

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