Quality of life in spina bifida: importance of parental hope

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Abstract

***Background and aims*—Prognosis in spina bifida (SB) is often based only on neurological deficits present at birth. We hypothesised that both parental hope and the neurophysical examination predict quality of life in children and adolescents with SB.**

***Methods*—A previously validated disease and age specific health related quality of life (HRQL) instrument was posted to families of children (aged 5–12 years) and adolescents (aged 13–20 years) with SB. We measured parental hope, determined the child's current physical function, and obtained retrospective data on the neonatal neurophysical examination (NPE). Regression analysis modelled HRQL firstly as a dependent variable on parental hope and NPE ("birth status"); and secondly on parental hope and current physical function ("current function").**

***Results*—Response rates were 71% (137 of 194) for families of children, and 54% (74 of 138) for families of adolescents. NPE data were available for 121 children and 60 adolescents. In children, the birth status model predicted 26% of the variability (R^2 hope 21%) compared with 23% of the variability (R^2 hope 23%)in the adolescents. The current function model explained 47% of the variability (R^2 hope 19%) in children compared with 31% of the variability (R^2 hope 24%) in the adolescents.**

***Conclusions*—In both age groups, parental hope was more strongly associated with the HRQL than neonatal or current physical deficits. A prospective study is required to determine whether a causal relation exists between parental hope and HRQL of children and adolescents with SB.**

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Keywords: spina bifida; health related quality of life; hope; newborn

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Spina bifida (SB) is a serious congenital anomaly with an incidence of 0.4–1.0 per 1000 births in the USA.[1] Universal folic acid supplementation will reduce, but not entirely eliminate neural tube defects.[2] The sequelae of SB include decreased mobility, bladder and bowel incontinence,[1] and neurosensory disabilities as well as social disadvantages such as decreased community acceptance and lack of a supportive peer network.[3–5] In addition, SB has major financial and employment implications for affected families.[4]

Improved operative interventions for SB (and particularly for hydrocephalus) during the neonatal period became available after 1960 and highlighted the need for the development of treatment guidelines. The therapeutic pendulum has swung considerably; from recommendations for active therapy regardless of the level of lesion[6–8]; to a selective treatment based on deficits assessed by a neurophysical examination (NPE)[7–11]; and back to aggressive therapy for virtually all patients.[1] This latter approach follows concerns about the predictive accuracy of the NPE, the ethics of selective practices,[12–15] and the survival of non-operated infants.[13–15]

Given the uncertainty about the best management for an individual infant with SB, it has been suggested that "parental directives for care" might resolve some of the dilemmas that arise when health caregivers make judgements about their patients' future quality of life.[16] This suggestion is in keeping with an increasingly heard request for more parental involvement in medical decisions for critically ill newborns.[17] Nonetheless, in order to make rational decisions, parents need accurate prognostic information; the NPE is used commonly to predict future physical and cognitive function.[18] Moreover, in clinical practice, limitations in mobility, bladder and bowel function, and cognitive development are often used to predict an individual's future health related quality of life (HRQL), despite the absence of empirical data on the predictors of HRQL in this population.

In our work with parents of children with spina bifida, it was suggested that factors other than NPE might predict children's HRQL. One hypothesised factor was level of parental hopefulness. Advisors from the Spina Bifida and Hydrocephalus Association of Ontario agreed with the importance of a construct such as hope in modifying outcomes. The construct of hope has been described in the nursing, psychological, and sociological literature.[19–31] It has been suggested that hope is essential for mental health,[19–21] for coping and adapting,[22] and for sustaining and energising a mother's maternal work in the care of her disabled child.[23] Hope is increasingly recognised as a disease modifier.[24–26] We therefore hypothesised that parental hope is an important determinant of HRQL in SB.

The objectives of our study were to evaluate the relative contributions of neonatal and current physical function, and of parental hope, to the HRQL of children and adolescents with SB.

Methods

STUDY POPULATION

Following initial surgery, all children with SB in Ontario are referred to a regional, publicly funded specialist treatment centre for ongoing care. We obtained patient lists from two such regional centres. Chedoke-McMaster Hospitals

serves the Hamilton-Wentworth region of Ontario and follows approximately 90 SB patients; the Bloorview MacMillan Centre serves the South Central region of Ontario and follows approximately 650 SB patients. The study received ethical approval from these two institutions and from the Hospital for Sick Children in Toronto. A random sample was taken of all patients in the age groups 5–12 years and 13–20 years; their families received a postal enquiry as described below. This random selection ensured a representative sample of patients whose characteristics would cover the full range of disability and functional impairment.

PROCEDURE

Parents were asked for permission to retrieve their child's neurophysical examination findings from hospital records. In addition, parents were asked to complete a demographic data collection form, a form to describe current function, and the Miller Hope Scale. Parents of children aged 5–12 years were requested to complete the HRQL questionnaire on behalf of their child; adolescents aged 13–20 years completed this questionnaire themselves. Parental completion of the Hope Scale, and child completion of the HRQL occurred concurrently. To maximise response rates, we completed three mailings accompanied by two telephone calls which were spread over three months after the initial telephone call.

Demographic data and current functional status

Demographic data included family income, place of residence (urban or rural), and family composition. In addition, data were obtained on the current level of physical function, including mobility, and degree of bladder and bowel independence. Finally, the number of previous medical interventions was recorded, including all shunt revisions, and the number of surgical operations the child had received.

Parental hope: the Miller Hope Scale (MHS)

This 40 item scale was devised by Miller and Powers[20] and validated in 522 adults. This instrument has good measurement properties including test–retest reliability. In a pilot group of parents of both 5–12 year old children and 13–20 year olds with spina bifida, we found that the MHS was easily administered. In both age groups, the MHS was completed by the same parent who provided the data on current function and demographics.

Quality of Life Scales (HRQL)

This self administered reliable and validated age specific instrument was devised following the general methodology outlined by Kirshner and Guyatt,[32] and applied to HRQL.[33] We have previously reported the methodological development of this instrument.[34] The items for this instrument were generated through open ended interviews with families of children with SB. The items were then reduced, and validated by accepted techniques.[33] Two age specific questionnaires were developed: for children aged 5–12 years (44 items), and for adolescents aged 13–20 years (47 items). Items from several domains were included.

NEWBORN NEUROPHYSICAL EXAMINATION (NPE)

Data on the NPE were retrieved from the newborn hospital record where available. Details of the NPE included head circumference and percentile at birth, and presence or absence of other congenital anomalies (that is, chromosomal anomalies and congential anomalies not typically associated with spina bifida). Level of the spinal cord lesion was judged from the hospital record by references to sensory function, muscle innervation level, muscle power, and the presence or absence of an anal wink.[1] In a pilot study of 15 cases, there was 100% agreement of the information retrieved from the hospital record, as assessed independently by two individuals, a research assistant and a physician. For the entire study sample, two physicians independently rated the level of lesion for each study patient. (One physician is a neonatologist (HMK) and the other a tertiary care academic general paediatrician (PCP).) Inter-rater agreement was 85%. A third physician, a developmental paediatrician (PLR), arbitrated the 15% of cases where HMK and PCP did not agree.

STATISTICAL ANALYSIS

Statistical analysis was performed using SAS (SAS Institute Inc., Cary, North Carolina). We performed two separate forward selection regression models in each age group. A linear least squares regression model was used. Terms with a significance level less than 0.05 were deemed to be statistically significant, and were added to the model.

The first analysis in each age group was termed the "birth status" model. This examined the extent to which the current HRQL of the child could be predicted from two sets of variables: the neurological status at birth using the NPE, and parental hope measured by the MHS.

The second regression model was termed the "current function" model. This analysis asked how much of the variability in the HRQL could be explained by the current physical function of the child, and by parental hope.

SAMPLE SIZE

We calculated post hoc that the respective sample sizes of 60 and 121 had provided us with 80% power at the 5% level for detecting an association between the MHS measure of parental hope and an independent variable if the strength of the correlations were 0.33 and 0.23, respectively.[35]

Results

SUBJECT RESPONSES

The response rate was 71% (137 of 194 families) in the 5–12 year age group. The results of 121 matching NPEs (88%) could be obtained. The remainder of the children were either born out of province or had inadequate details recorded. Current physical function was determined in all 137 children and 131 responses were received for the parental MHS.

In the 13–20 year age group, the response rate was 54% (74 of 138 families). The NPE was available in 60 of these.

Table 1 Socioeconomic characteristics of the parents of children

	5–12 years (%) (n = 121)	*13–20 years (%) (n = 60)*
Income levels		
>$40 000/year	46	42
$20–40 000/year	36	43
$10–20 000/year	18	15
Size of community		
Large urban	50	38
Rural or small urban	50	62
Family structure		
Single parent families	17	18
Two parent families	83	83
Extended family support		
Yes	73	55
No	27	45

Table 2 Functional characteristics of children sampled

	5–12 years (%)	*13–20 years (%)*
Bladder		
Independent without catheter	10	8
Independent with catheter	44	66
Needs help	47	26
Bowel		
Independent (no enemas etc)	32	32
Independent, needing enemas	15	24
Needs help with bowels	53	43
Mobility		
Walks everywhere, no aids	18	12
Walks with aids	33	30
Household walker and wheelchair	12	16
Full wheelchair	37	42
Number of operations		
0	2	3
1–3	29	13
4–5	23	24
6–10	38	30
11–20	9	24
21–30	0	6
Maximum number of operations	20	30
Number of shunt revisions		
No shunt	50	33
1–4 revisions	40	51
>4 revisions	10	16

Table 3 Birth status model

Age group	*Steps*	*Variables*	*Coefficient*	*R^2*	*p value*
5–12 years	1	Parental MHS	0.4793	0.207	<0.0001
	2	Level of lesion	0.0227	0.051	0.0055
	3	Head circumference	−0.0230	0.005	0.3634
	4	Congenital anomalies	0.0080	0.001	0.7287
13–20 years	1	Parental MHS	0.5570	0.234	<0.0001
	2	Level of lesion	0.0170	0.017	0.2565
	3	Head circumference	−0.0091	0.029	0.8092
	4	Congenital anomalies	−0.0176	0.031	0.5990

Table 4 Current function model

Age group	*Steps*	*Variables*	*Coefficient*	*R^2*	*p value*
5–12 years	1	Parental MHS	0.4173	0.188	<0.0001
	2	Age	−0.0120	0.050	0.0007
	3	Number of operations	−0.0055	0.131	0.0014
	4	Bladder	−0.0688	0.102	0.0001
	5	Mobility	−0.0090	0.006	0.2522
	6	Bowel function	−0.0074	0.003	0.4316
	7	Number of shunt revisions	−0.0002	0.010	0.8267
13–20 years	1	Parental MHS	0.5171	0.238	0.0003
	2	Bowel function	0.0406	0.072	0.0171
	3	Age	−0.0052	0.004	0.5447
	4	Number of operations	−0.0003	0.008	0.8484
	5	Mobility	−0.0123	0.010	0.3269
	6	Bladder	−0.0569	0.041	0.0402
	7	Number of shunt revisions	0.0008	0.008	0.4049

CHILD AND FAMILY CHARACTERISTICS

Table 1 outlines the socioeconomic characteristics of the families in the two age groups. Table 2 presents details of the children's characteristics including the child's physical function as assessed by the families and by teenagers themselves, for bowel and bladder function, degree of mobility, and the number of operative procedures and shunt procedures.

REGRESSION ANALYSIS

The full details of the final regression models are presented by age group, in Appendix 1. Further detailed results are presented in the tables. Table 3 summarises the results for the birth status model for each age group. Table 4 summarises the results for the current function model.

Birth status model

This model explained 26% (total R^2) of the variance of HRQL in children aged 5–12 years and 23% in adolescents aged 13–20 years. In both age groups, the explanation provided by the parental MHS was far greater than that provided by any other variable.

Current function model

In the younger age group, this model explained 47% of the variance associated with HRQL. The variance was mainly explained by MHS, bladder function, and the number of surgical operations. In the older age group, this model explained 31% of the variance associated with HRQL, with MHS and bowel function contributing most importantly.

Discussion

In this study, we found a striking association between the child's HRQL and parental hope. In contrast, physical limitations which clinicians commonly use to predict HRQL appeared to be less important. Our data are consistent with the view that some of the ethical concerns surrounding the prediction of outcome for newborn infants with SB can only be solved by a better understanding of family desires, problems, and coping strategies.[16]

Previous work has shown that perceptions of HRQL in SB and other disabilities vary considerably between different observers. Cadman *et al* reported that parents of children with and without disabilities perceive physical and mental handicaps very differently from health care professionals.[36] Other investigators have shown that in contrast to physicians who care for children with SB, parents do not view those children as being sick.[5 37] In the present study, we have shown for the first time that differences in HRQL for children and adolescents can be attributed, at least in part, to differences in the degree of parental hope. Taken together, these findings suggest that health professionals should exercise extreme caution when attempting to predict an individual child's HRQL based on physical symptoms and signs. As patients and their families are the best judges of their own HRQL, we do not support the recent approach by Takayanagi and Suruga[38] to determine quality of life in neonatal surgical cases,

as it is based on the assessment of medical providers rather than of parents.

Hope is a construct that has been extensively explored in the nursing, psychological, and sociological literature, and reliable and valid scales for its measurement have been devised, including the one used in this study.[20] The Miller Hope Scale has been found to be correlated with religiosity, spiritual well being, hope, and other positive mood states[21]; the coping effectiveness[22]; social support and self esteem[27 29]; resolution of psychosocial issues and the perception that one is able to control one's life[28]; and psychological well being.[19 20] Larson[23] has found that hopefulness is critical for sustaining and energising a mother's maternal work in the care of her disabled child. Hope is increasingly recognised as a disease modifier,[24–26] although we are only beginning to understand how hope (and hopelessness) affect biological processes.

This study has limitations. We used proxy responses from parents to judge HRQL for children aged 5–12 years, while adolescents rated their own HRQL. Importantly, whether or not a parental proxy was used to determine HRQL, a strong association was found between parental hope and HRQL. Another limitation to the interpretation of the relation between parental hope and HRQL results from the cross sectional nature of the study. It is possible that parental hope is a reflection of the experiences of the family, or that parental hope directly influences the child's HRQL. The latter interpretation seems plausible, given that the construct of hope has been linked with positive psychological factors as noted above. Parents caring for a child with a physical disability may well empower their children with a similar sense of hopefulness, which may result in an improved perception of their HRQL.

It is not known whether hope is a static trait or a dynamic state. If hope is a trait, that is, a stable personality or temperamental characteristic, it follows that hope is likely to be a true determinant of HRQL. If hope is a dynamic state, it may potentially be responsive to targeted interventions. It has already been shown that a prospective nursing intervention reduced maladjustment in children with chronic disorders.[39] It is conceivable that nursing provision had in some way fostered hope. Wake and Miller have described several hope inspiring nursing strategies for treating hopelessness.[30 31] Furthermore, it is not known what the determinants of hope might be. It is possible that factors such as family income, family structure, or parents' level of education may in some way determine degree of parental hopefulness. Although several of these factors were measured in our study, we feel that determinants and dynamics of hope need new and prospective testing in future studies.

We would argue that more attention to the understanding and nurturing of hope is warranted in the long term care of children with SB and probably also in other childhood disorders. The importance of hope has been recognised by nurses and psychologists. It should be equally recognised by physicians caring for children.

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Appendix 1

The final models are as follows.

BIRTH STATUS

5–12 year age group: HRQL = 0.32 + 0.48 (parental MHS) + 0.02 (level of lesion)

13–20 year age group: HRQL = 0.41 + 0.56 (parental MHS)

CURRENT FUNCTION

5–12 year age group: HRQL = 0.74 + 0.42 (parental MHS) − 0.01 (number of operations) − 0.07 (bladder function) − 0.01 (age)

13–20 year age group: HRQL = 0.53 + 0.52 (parental MHS) − 0.04 (bowel function)

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Why us? Why me?

Guiding parents through the diagnosis and management of rare and devastating conditions is difficult and draining. Perhaps the most tricky question to tackle is "why us?". Most people are unable to assess probability and, even if they can understand how often in the population an event might occur, this rarely translates into an appreciation of the chances of such an event happening to them.[1] The rarity of such an event makes it difficult to believe and make sense of why it has happened to any one individual.

Having been a consultant paediatrician for ten months, I have found counselling parents facing these diagnoses one of the hardest aspects of my job. I am, however, getting a surprising amount of practice. Recent discussions in the medical and lay press, combined with a somewhat warped sense of fascination with probability and increasing disbelief relating to my experiences this year, led me to wonder if I am getting a predictable amount of practice, or if is this a highly unlikely sequence of events.

I am part time, so carry about a tenth of the workload. Thus, I can calculate the probability that I might be the consultant responsible for any patient newly diagnosed with each condition in any one year in my district (table 1). If I am right, and each event is independent of the other, multiplying these probabilities together should give the probability of children with all of these conditions diagnosed in one year in Portsmouth being admitted under my care.

This makes the probability of a paediatrician with a tenth of the workload in Portsmouth seeing all these conditions in one year, 6.1×10^{-16} or 1 in 1633 billion.

We like to rely so much on measurement to help us make sense of what is happening in medicine. In this case, as the numbers get smaller (or bigger), they seem increasingly ridiculous. The reality of my caseload reminds me that this is really happening but the figures suggest it almost certainly isn't. I haven't spent too long wondering "why me?". I have learned such a lot in this crash induction into consultant practice, perhaps most of all, the value of supportive colleagues. Of course for me the question "why?" doesn't intrude on my life and belief structure at all, but for parents it often remains the most difficult part to come to terms with. Knowing the chances of a rare event happening are no comfort once it has done so and the question being asked is "why?".

The author would like to thank those who checked her maths.

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Editor's note

As a new consultant nearly 25 years ago, I came across a similar improbability list for GPs, written by the late Professor R S Illingworth. I used it in teaching for a decade or so, then lost the reference. Does anyone have it?

Table 1 Incidence and probability of disease occuring in Portsmouth

Condition	*Incidence/ birth prevalence*	*In Portsmouth (6000 deliveries/year)*	*Probability of seeing in any one year on 1 in 10 rota*
Ondine's/Hirschsprung's /Neuroblastoma[2]	1in 1.4 million	1 in 233 years	0.00043
Apert's (normal twin)[3]	1 in 80 000	1 in 13 years	0.00769
MCAD[4]*	1 in 22 000	1 in 3.6 years	0.02732
Galactosaemia[5]	1 in 450 000	1 in 7.5 years	0.01333
Prader-Willi[6]	1 in 16 000	1 in 2.6 years	0.03846
Pelizaeus-Merzbacher's (conatal form)[7]	1 in 770 000	1 in 128 years	0.00078
Munchausen syndrome by proxy (<1 year old)[8]	1 in 35 700	1 in 5.9 years	0.01695

MCAD, medium chain acyl-CoA dehydrogenase deficiency.