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Quality of Life in Adolescents and Young Adults with and Without Spina Bifida: An Exploratory Analysis☆

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ABSTRACT

Purpose: The measurement of Quality of life (QOL) in adolescents and especially in adolescents with disabilities is limited, often by an assessment of function rather than perception. This analysis explores QOL in adolescents and young adults (AYA) with and without Spina Bifida (SB) from the perspective of AYA and their parents.
Design and methods: A descriptive study using content analysis was conducted as a component of a larger multi-site mixed-method study of secondary conditions and adaptation. Participants responded to a single open-ended question on the meaning of quality of life.
Results: Descriptive accounts from 209 families generated the following shared categories: an engaged family, a positive life, the goal of independence, being healthy, essential needs for living, having friends, relying on faith, and romantic relationships. A unique category emerged from parents, doing what AYA wants to do.
Conclusions: Family was the most frequently nominated component of QOL. The centrality of family in QOL is an important finding generally not assessed in measures of QOL or even less in health-related QOL instruments.
Practice implications: Findings illustrate the importance of evaluating overall QOL from the perspective of AYA and their parents.

Introduction

Quality of life (QOL) is an important component of living fully with a chronic health condition. Yet, there has been a lack of clarity in the literature about the concepts QOL and health-related QOL (HRQOL) and their meaning for individuals with disabilities and their families. QOL is defined by the World Health Organization as "an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations and concerns" (World Health Organization Quality of Life (WHOQOL) Group, 1998, p. 551). HRQOL is a sub-domain of QOL and captures a subjective perception of the impact of a health condition on the individual (Bakaniene, Prasauskiene, & Vaiciene-Magistris, 2016; Rofail, Maquire, Kissner, Colligs, & Abetz-Webb, 2014; Waters et al., 2009). Being clear about the concept being addressed will provide important information for individuals and their health care providers. The measurement of QOL and HRQOL in adolescents throughout the world and especially in adolescents with disabilities is particularly unclear, evaluated in a variety of ways and with different components or dimensions of QOL. To address this gap, we investigated the meaning of QOL in adolescents and young adults (AYA) living with the disability of Spina Bifida (SB) and their parents and a comparison sample of families with an AYA without SB.

Background

While there is an emerging literature on the concept of QOL in adolescents, it is complicated by measurement issues and types of measures. QOL is measured most often either with instruments that have multiple domains or a single item. Most studies have used instruments with multiple domains from measures based on typically developing youth or measures originally developed for adults with a disability with few capturing the meaning directly from the adolescent and their parents. Further, the previous measurement of QOL has often focused on the assessment of function rather than perception (Waters et al., 2009). In addition, it is not clear if there are different perceptions of adolescents who have disabilities and those without disabilities. When adolescents were interviewed directly to generate instruments,

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generally a small number participated, and there was evidence that domains of QOL or HRQOL varied across the age range (Szymanski et al., 2017). When QOL has been measured in adults with a disability, investigators have reported a "disability paradox" in individuals with disabilities often reporting relatively high QOL (Waters et al., 2009). This was originally seen as potential measurement error, as those without a condition could not imagine that individuals with a disability could perceive that their life was "good." Later investigators identified that individuals who acquired a disability may change the meaning of QOL or change the priority they give different domains of QOL. However, we do not know if these "response shifts" occur in those who grow up with a disability. Further, some investigators examining QOL in adults with disabilities have based their work on the assumption that QOL domains are the same for people with and without disabilities, although they acknowledge that adults living with lifelong physical disabilities may address those issues differently.

Internationally when QOL was investigated in AYA with disabilities, QOL was evaluated in a variety of ways and different components of QOL were identified. Development of autonomy and choice, purpose of life, positive relationships with others, and personal growth and self-acceptance were attributes of QOL identified in open-ended interviews of adolescents with physical disabilities living in inclusive communities of Zimbabwe (Mpofu, Sefotho, & Maree, 2017). Investigators using the World Health Organization Quality of Life-Shorter Version of the original instrument (WHOQOL-BREF) explored QOL in adolescents with disability in India (Abraham, 2013). They found that a majority had an average overall quality of life, but when examining perception of quality of life in specific domains, participants reported a moderate level of psychological factors (e.g., negative feelings) and were dissatisfied with the environment in which they are living (e.g., lack of transportation, money) (Abraham, 2013). AYA in Asia with physical disabilities were found to have different patterns of QOL scores when evaluating overall objective and subjective QOL when using the *Student Version of the Comprehensive Quality of Life Scale*; this is a global measure of QOL with the following domains: material well-being, health productivity, intimacy, safety, place in community, and emotional well-being (Lin et al., 2009). While no differences were identified between groups in objective QOL, subjective QOL was significantly higher (overall and domains) in adolescents with physical disabilities when compared to those without (Lin et al., 2009).

Measurement of overall QOL with a single item has been a component of several domain-specific QOL measures but has not been frequently reported. However, there has been preliminary work in adolescents with SB using a single item (How would you rate your QOL?) (Sawin, Brei, Buran, & Fastenau, 2002). The single item has the advantage of allowing the individual to include the domains important to them and then weigh the domains uniquely. However, the limitation is that health care providers working with an AYA do not know what is salient to each individual. This gap was addressed with the current study. Based on the literature and the authors' previous trajectory of work in adolescents living with SB, we conducted this study to explore the concept of QOL in AYA with and without SB from the perspective of both AYA and their parents.

Method

A descriptive study using content analysis was conducted as a component of a larger multi-site mixed-method study of secondary conditions and adaptation in families with AYA with and without SB. The larger study included in-depth 1–2 hour interviews of AYA with and without SB and their parent, neuropsychological testing, and teacher report of behavior in school.

The comprehensive interview was used to administer instruments that addressed concepts delineated in the Ecological Model of Adaptation and Secondary Conditions in SB (Sawin et al., 2002; Sawin, Buran, Brei, & Fastenau, 2003). Three risk categories (SB Neurological Severity, Neuropsychological Functioning, Characteristics of the Sample), three protective factors (Adolescent Resilience, Family Resourcefulness, and Health Care Adequacy) and adaptation outcomes (QOL, Mental Health, and Self-Management) were assessed. This secondary analysis addressed a single open-ended question that followed the AYA and parent's survey assessment of the AYA QOL.

Ethics approval

IRB approval to conduct the larger study was obtained from the University of Wisconsin Milwaukee and four clinical settings. All participants provided written informed consent and/or assent before participation in any aspect of the larger study. If the adolescent declined assent, the family was not enrolled in the study.

Sample

Participants for this analysis represented 209 families (112 parents of AYA with SB, 97 parents of AYA without SB, 103 AYA with SB, and 94 AYA without SB) who reported on AYA's QOL (Ridosh, Sawin, Brei, & Schiffman, 2018). The convenience sample of families of AYA with SB was recruited for the larger multi-site study from four clinical programs in the Midwest and Eastern region of the United States (US) that had established SB programs. Inclusion criteria were: families with AYA 12 to 25 years of age, no intellectual disability documented by testing or parent report, English speaking, and no other major health conditions (except those associated with SB). Comparison families of an AYA without SB had similar inclusion criteria (AYA 12 to 25 years of age, no intellectual disability, English speaking, and no major health condition). They were recruited by referral from SB families participating in the study and from the four participating children's hospitals. Advertisements were displayed in each hospital and with primary care providers.

Data collection

The interviews, which lasted 1 to 2.0 h, were conducted via telephone by trained advanced practice pediatric nurses using computer-assisted direct entry relational databases to record interview data. Although studies of children with Spina Bifida have identified complex patterns of strengths and difficulties across domains, children and adolescents with SB generally have strong verbal skills and more visual perception problems (Dennis & Barnes, 2010; Dennis, Landry, Barnes, & Fletcher, 2006; Lollar, 1990; Wills, Holmbeck, Dillon, & McLone, 1990). Therefore, the use of interviews with adolescents to explore sensitive subjects (Boekeloo, Schamus, Simmens, & Cheng, 1998; Flanagan, Greenfield, Coad, & Neilson, 2015), and our history (Sawin et al., 2002) and those of others using interviews to study adolescents with chronic conditions (Austin, Huster, Dunn, & Risinger, 1996; Lyneham & Rapee, 2005), supported the use of telephone interviews over written surveys. Additionally, administration of multiple instruments in the larger study provided rationale for using interviews rather than having potential of survey burden.

The interviews included a Demographic and Clinical Information Form, reliable and valid instruments with Likert-like response patterns, and QOL items with response patterns of 0–100, and open-ended questions regarding QOL for AYA and parent. The measures of the larger study are fully described elsewhere (Ridosh, Sawin, Schiffman, & Klein-Tasman, 2016; Sawin, Buran, Brei, & Cashin, 2007). Each participant had the opportunity to have the interviews conducted in 2 sessions. Breaks were offered frequently. As a part of this comprehensive interview, participants were asked to rate a single question on AYA quality of life. Typical in QOL assessment, no definition of QOL was provided in order to let the participants use and weigh components of QOL important to them. After the rating was recorded, the interviewer asked the AYA a single open-ended question: "*What do you think QOL means for you*?" The follow-up probe, if the participant was unable to generate

a response, was "*What goes into making a good "quality of life*?"" No additional follow-up questions were asked once the AYA provided a response. Similarly, the question asked to the parents was, "*What do you think QOL means for your son/daughter*?" If the parent was not initially able to indicate a definition, the follow-up probe was "*What goes into making a good QOL for him or her*?" The AYA and parent's brief responses were recorded verbatim directly on the Microsoft Access® database by the data collector. Participants were not asked to expand on their description or provide a fuller explanation of the areas nominated.

Data analysis

Content analysis was used to capture and organize the responses (Ritchie, Lewis, Nicholls, & Ormston, 2013). The sorting of data by categories conducted by the authors was iterative. Two investigators collaboratively analyzed the AYA data first and created a coding sheet with the responses of each group. The responses were examined and assigned to clusters of common data. Preliminary categories were identified and refined. Agreement on categories was high and differences were resolved with discussion. The parent data were subsequently analyzed using the same process. Additional categories emerged from the parents of AYA without SB. The data from the parents of AYA with SB then were reviewed to determine if these additional categories were supported by the responses. Data consistent with several of the new categories were present in the parents of AYA with SB data. AYA data were rechecked for evidence of new categories and only limited evidence was identified. The third team member reviewed these data to assure confirmability. No new categories were identified. Once the final set of categories was determined, text for each category was organized by group of participants to identify the frequency of categories. Additionally, exemplar responses were selected to "tell the story" of the participants in each category. The first three investigators participated in the generation of the categories. The fourth investigator, who participated in the design of the larger study, reviewed the findings for accuracy and clinical relevance.

Results

Participants' characteristics

Of the 209 parents who reported on their AYA's QOL, 198 provided a response to the open-ended question (103 parents of AYA with SB, 95 parents of AYA without SB). Of the 197 AYA who reported on their QOL, 191 (97 AYA with SB and 94 AYA without SB) responded to the open-ended question.

The average age of AYA in both groups was approximately 15 years (range 12–25). The majority of AYA with SB (67%) had thoracic or high lumbar lesions and 74% had an Individual Educational Plan or 504 plan in school indicating they had a condition that impacted their learning. Only 3% of those without SB had such a plan. The only demographic result demonstrating a major difference between the families with and without SB was family income with 52% of families with SB having an income of $50,000 or more compared to 72% of the comparison families; income significantly different by group χ^2 (207) = 16.67, $p < .001$ (Ridosh et al., 2016; Ridosh et al., 2018). Additional details of the sample are described in Ridosh et al. (2016).

The parents interviewed were primarily mothers, about 94%. The average age of parents of AYA with SB was 43.08 years (range 27–63, SD 6.46) similar to the parents of AYA without SB who were on average 44.33 years old (range 32–67, SD 6.26). The majority of parents were married (70.3% of parents of AYA with SB vs 76.8% of parents of AYA without SB). Parents were mostly Caucasian (86%) and female (94%). Less than half of the parents had attended or completed college (47% parents of AYA with SB vs 43% parents of AYA without SB).

Content analysis

The content analysis process identified eight categories shared by participants across groups. However, one category, romantic relationships, was not endorsed by AYA with SB. Only parents nominated one unique category, doing what they want to do. The categories listed are presented in the order of frequency for the AYA with SB (see Table 1). An engaged family was the highest ranked category followed by a positive life, the goal to be independent, being healthy, essential needs for living, having friends, and relying on faith. Even though those with and without SB and their parents often used the same overall language to describe QOL (e.g., friends, health, independence) the meaning they detailed sometimes had different characteristics and patterns.

Twenty-two of the AYA with SB (23%) indicated, "*Do not know*" as a response to the open-ended question and follow-up prompt compared to only three (3%) of AYA without SB. Only three parents of AYA with SB and no parent of AYA without SB indicated, "*Do not know*." AYA with SB used less than half as many words to describe what QOL meant to them than AYA without SB. The order in the discussion of each category is based on the highest to lowest frequency endorsed by the four participant groups (AYA with SB, AYA without SB, parent of AYA with SB, or parent of AYA without SB).

Shared categories nominated across participant groups

The following eight categories/themes were most often shared by all four groups of participants: an engaged family, a positive life, the goal of independence, being healthy, essential needs for living, having friends, relying on faith, and romantic relationships. However, one theme category, romantic relationships, was not endorsed by AYA with SB.

An engaged family

In both groups of AYA, the positive contribution of the family was the most frequently nominated response for "What makes up QOL" (27% in SB and 35% without SB). Included in the descriptions of AYA with SB were characteristics such as family support, a good family, a healthy family, being able to talk to family, and function as a normal family. AYA without SB also indicated a family they can talk to and described a family that cares about you, gets along with each other, enjoying family, and unconditional love of family. Other descriptions of AYA without SB were having parents and family around, how the family lived together, having a loving family, the amount of fighting in the family, family time together, and quality of family relationships.

An engaged family was also a highly nominated response of the AYA's QOL from parents of AYA with SB (28%) and without SB (36%). Both parent groups described family love, family support, feeling loved for who AYA is, and families spending the time to be together. Additionally, parents described family being present, family relationships, and a family they can talk to as part of their AYA's QOL.

Positive life

A positive life was comprised of two descriptive accounts. First, the emotional aspect of happiness and, second, an overall appraisal of "*living a good life*." Both parent groups used the descriptor "*happy/happiness*." However, happy/happiness was most prominent in AYA without SB where it is the second most frequent category.

Having a good life was prominently nominated by AYA. It was nominated second in the group of AYA with SB and by slightly fewer AYA without SB. It was infrequently nominated by parents of AYA with SB and absent in those data from parents of AYA without SB. AYA in both groups described QOL as having a good life. Specifically, how well you are doing in life, what you do with your life, and how you live every day. Also, a good life meant without having to worry about too much, getting everything you need and should out of your life, and living life the way you are supposed to. A few AYA with SB defined a good life as follows: "*If you are alive that is all that is needed for QOL*" and "*To live a*

Table 1
Description of QOL: shared and unique categories.

Participants	Comments and/or exemplar statements
Shared categories	
An engaged family	
AYA with and without SB	Can talk to family
	Family that cares about you
Parents of AYA both with and without SB	Family, they can talk to
	Family spending time to be together
	Family support
A positive life	
Parents of AYA with and without SB	All groups used the descriptor "happy/happiness"
AYA with and without SB	
AYA with and without SB	Living a good life:
	How you are doing in life; how I live every day
	A good life without having to worry about too much
	Getting through issues in life OK
Parents of AYA with and without SB	Living a good life:
	Infrequent in parents of AYA with SB
	Absent in the parents of AYA without SB
The goal of independence	
Parents of AYA with and without SB	Independence was listed by both
	More prominent in parents of AYA with SB
Parents of AYA without SB	Identified building blocks: having autonomy making decisions, being organized and self-sufficient
Parents of AYA with SB	Focused on development needed toward future independence
	Hoping to move out
	Maintaining the ability to have normal responsibilities
AYA with SB	Learn to care for themselves
	Doing everything for yourself
AYA without SB	A general concept of how you believe in yourself
Being healthy	
Parents of AYA both with and without SB	Health; good health; being healthy
	Emotional health was a separate frequent component
Parents of AYA with SB	A substantial number of AYA specified health related to the condition such as better bladder care, mobility, or needing surgery
Parents of AYA without SB	Overall wellness such as nutrition, exercise, or being physically fit
AYA with SB	Healthy; good health; being healthy
AYA without SB	Same components of general good health and healthy but also endorsed emotional health
	Get through things with emotional ease
	Emotional feeling
Essential needs for living	
Parents of AYA with SB	A roof over their head; good home where AYA can get around; Having basic needs met
Parents with AYA without SB	Essential needs for living were more future-oriented;
	Cell phones; gas; transportation; clothes
AYA with SB	Generated less specification in their responses
AYA without SB	More articulate and generated more examples
AYA with and without SB	Good education; job; money/financial stability
	Good home
Having friends	
AYA and parents with and without SB	Identified by all groups as a component of QOL
AYA without SB and their parents	Having friends was more prominently identified
	Expanded on the characteristics of the friendship: acceptance of friends, time at friend's house, and the freedom to choose friends
Relying on faith	
Parents of AYA with and without SB	Faith; values; religion; spirituality; spiritual connections
AYA with and without SB	Mentioned only by a few
	Values; being religious; having a fulfilling purpose
Romantic relationships	
Parents of AYA with and without SB	Prospect of love in the future
	Full social life
	Being loved in relationships
	Comfort in surroundings
AYA without SB	Having someone you love mutually
	Not having to change who you are to be loved
AYA with SB	Not identified at all
Unique category nominated by parents	
Do what they want to do	
Parents of AYA with and without SB	Identified by both groups
	Belonging; doing what AYA wants to do
	Freedom to choose a lifestyle
	Being able to do things AYA wants to
	Being able to do desired activities
Parents of AYA without SB	Following her dreams; exploring options
	Having opportunities to pursue interest and activities
	AYA loves to do and doing them well
	Having time to relax and do what AYA wants to do
Parent of AYA with SB	Participating fully in society
	Being able to do the things AYA wants to do even with adaptations
	Being able to do what AYA wants to do with minimal adaptation
	Thankful AYA can do things that others in wheelchairs cannot do
	Being treated like a normal kid

life and know that I can be normal." AYA without SB indicated the emotion of happiness more frequently whereas AYA with SB focused on the overall appraisal of having a good life.

The goal of independence

While independence appears in parent and AYA data it was a more prominent category in both the parents of AYA with SB and their AYA. Parents of AYA with SB were focused on development needed to acquire future independence, aspiring independence. These parents of AYA with SB indicated more intermediate responses such as being able to do things on AYA's own, maintaining the ability to have normal responsibilities, or hoping to move out. Their responses were hopeful but not fully confident about future independence. Unlike parents of AYA with SB, parents of AYA without SB seemed to assume children would be independent in college or living in the community. They focused on AYA continuing to function independently as they got older and identified building blocks such as having autonomy, making decisions, being organized, and becoming self-sufficient.

Independence-related concepts appear in both groups of AYA data. AYA with SB seem to be very aware of the need to learn to care for themselves as necessary for independence indicating: learning how to, being able to, or becoming capable of taking care of himself or herself. They spoke of doing everything for yourself and doing what you're supposed to do. In AYA without SB independence was present but not prominent and reflected a general concept of how you believe in yourself and treat or take care of yourself.

Being healthy

Being healthy, which included subcategories of general health and emotional health, was one of the top categories nominated by parents as a part of their AYA's QOL. However, health was more concerning for parents of AYA with SB (35% of parents of AYA with SB but only 22% of the parents without SB). In both parent samples, health was nominated with the general words "health," "good health," and "healthy" used most frequently. A few parents of AYA without SB generated additional comments (e.g., feeling well) but no specifics. A substantial number of parents of AYA with SB specified health related to the condition (e.g., physical, better bladder care, better outcomes than others with SB, mobility, or needing surgery) or to more general health (e.g., keep self-healthy, good health maintenance). Only parents of AYA without SB also endorsed concepts of overall wellness such as nutrition, exercise, or being physically fit in their definition of health.

Both parent groups frequently indicated that emotional health was a unique part of AYA QOL. Parents of AYA without SB nominated overall emotional health such as feeling good about themselves, having good self-esteem, being well-rounded, and being aware of strengths and weaknesses. The parents of AYA with SB, however, focused more on

strategies to achieve emotional health in the presence of a chronic condition such as having a sense of humor, not dwelling on problems, not getting depressed, sorting out the little stuff, developing positive attitudes and outlook on life. Also, included in this last category was having parents that facilitated positive coping. One parent indicated "*If we can see past this then he can too*." Taken together, general health and emotional health are one of the top nominated subcategories. However, health was clearly a concern for parents of AYA with SB.

AYA with and without SB did not mention health as frequently in their responses. When nominated, responses broadly referred to good health and being healthy. A difference noted in the AYA without SB is that responses mentioned emotional health such as "*get through things with emotional ease*" and "*emotional feeling*."

Essential needs for living

For the parents of AYA with SB, this category focused on a good home, a roof over their head, and a good home where the AYA can get around. Further descriptors were being able to get the things AYA needs as an adult to live, food, and having basic needs met. The QOL indicators for them were primarily present-oriented. They worried about basic things—"*How is my AYA going to have a roof over his head if AYA does not have a job*?" Parents of AYA without SB addressed some of the same elements but their perception of essential needs for living was more future-oriented. They also tended to define essential slightly differently and beyond the basics such as cell phones, gas, transportation, and clothes.

The category "essential needs for living" in the AYA data was defined or perceived somewhat differently by the two groups of AYA. AYA without SB were more articulate and generated more examples in their descriptive accounts. AYA with SB generated less specification in their responses. However, responses such as a good education, a job, money/financial stability, a good home, and a good environment were common.

Having friends

Having friends or friendships was a response generated in all groups, however, it was dramatically more prominent in AYA without SB (35%) and their parents (11%). AYA with SB (24%) identified friends as a part of QOL as opposed to 13% their parents. However, AYA without SB and their parents not only identified having friends but expanded on the characteristics of the friendship. These included the following: having friends who care for you, having good friends, having many friends, acceptance of friends, spending lots of time with friends, spending time at a friend's house, and having the freedom to choose friends.

Relying on faith

Both parent groups but only one AYA with SB and only a few without SB indicated relying on faith. This was made up of faith, values, religion, and spirituality as a part of AYA QOL. Parent responses included having faith, church beliefs, religion, spiritual connections, and spirituality. AYA were much more general addressing values, being religious, and having a fulfilling purpose.

Romantic relationships

Romantic relationships were nominated as a part of AYA QOL by both parent groups and by a similar percent of AYA without SB. However, it was not identified at all by AYA with SB. Parents of AYA without SB identified close relationships/companionship, the prospect of love in the future, having a full social life, and being loved in relationships. Parents of AYA with SB had similar responses including people who love you, feeling loved, enjoying people, and having a social life. They also indicated relationships hoped for but not yet achieved such as: having people to love AYA, wish for AYA to find the one good person that will love and respect AYA, and someone to love and care for AYA as AYA is. AYA without SB spoke generally about love in relationships to indicate having someone you love mutually and not having to change who you are to be loved.

Unique category nominated only by parents

Doing what they want to do

The predominant response in this category identified by both parent groups was "*Doing what they want to do*." Both groups of parents identified belonging, doing what AYA wants to do, freedom to choose a lifestyle, and doing all the things AYA would like. Further descriptions included doing things AYA enjoys, being able to do things AYA wants to, being able to do desired activities, doing everything AYA can, being involved, and going anywhere AYA would like. Parents of AYA without SB also addressed the larger "doing what AYA wanted to do" with following AYA's dreams, exploring options, having everything AYA needs for a bright future, having opportunities to pursue interest and activities AYA loves to do and doing them well, and having time to relax and do what AYA wants to do. A few parents of AYA with SB also specified the role of the condition indicating "*Being able to do the things AYA wants to do even with adaptations*," "*Being able to do what AYA wants to do with minimal adaptation*," or "*Thankful AYA can do things that others in wheelchairs cannot do*." One parent of an AYA with SB indicated a negative impact on QOL "*AYA cannot do things AYA wants to do*."

Some parents of AYA with SB but no others saw "participating fully in society" as an important component of "doing what you want to do." These parents indicated QOL meant participating in meaningful activities important to the AYA, being able to do adult things on their own, being treated like a normal kid, having equal opportunities as sibling without SB, keeping up with siblings, getting out of the house daily, or being able to do things family wants to do.

Discussion

This study elicited a beginning description of what makes up AYA QOL from their perspective and that of their parents. The salient categories identified by the AYA with and without SB and their parents are somewhat similar to those identified in the literature by adults and adolescents in the US and internationally. However, the priority and descriptors differed by participants with and without SB. For example, both AYA with and without SB nominated "family" most frequently while faith was rarely nominated, but when it was, it appeared to be of central value for the individual. All participants generated "friends" but the descriptors of having friends varied. Also, having a positive life, being happy or living a good life was frequently nominated with varying characteristics between groups.

The centrality of the "engaged family" in QOL for both AYA with and without SB was an important finding. The family seemed to be an anchor for AYA as they consider their QOL. Over time family functioning appears to be fairly stable during adolescence in families of AYA with SB and these families are not characterized with the increase in conflict which occurs in comparison families (Bakaniene et al., 2016; Jandasek, Holmbeck, DeLucia, Zebracki, & Friedman, 2009; Sawin & Bellin, 2010). It may be that when families are doing well this can be protective for the AYA. The lack of typical conflict may be due, in part, to the priority of SB condition-related shared responsibilities for care. However, even with evidence of the family's centrality in the life of the AYA, a family domain generally has not been included in many measures of QOL and even fewer HRQOL instruments (Bakaniene et al., 2016; Rofail et al., 2014; Sawin & Bellin, 2010).

While achieving independence is an expectation for all AYA, for many with chronic conditions achievement of independence skills is delayed (Davis, Shurtleff, Walker, Seidel, & Duguay, 2006). The meaning of independence as a category of QOL differed by the presence of SB in this study. Parents of AYA with SB focused on aspirations, not achievements, when reporting on what makes up QOL. The few responses from AYA that addressed independence were from those with SB who focused

on being capable of taking care of themselves. Adults with SB undergoing surgical reconstruction experienced more independence but did not have different perceptions of QOL when compared to those not having surgery. Understanding the role of independence in the meaning of QOL is important for future work (Coco et al., 2018). Perhaps aspiring independence is more prevalent in these parent data of youth with SB as they were acutely aware of the AYA's unique challenges to "catch up" on typical self-management or independence skills. When evaluating developmental milestones over time, AYA with SB were less likely at age 22–23 to achieve adult milestones such as independent living, attend college, or obtain full-time employment than their peers (Holbein, Zebracki, et al., 2017) . The latter study also determined parent expectations for independent living did not predict milestone achievements. Rather, parent expectations were related to the AYA's lesion level and cognitive abilities were associated with both independent living and attending college (Holbein, Zebracki, et al., 2017). Youth with SB have faced barriers finding employment that is accessible and matched to their abilities as well as securing accessible and reliable transportation to get to work (Lindsay, McPherson, & Maxwell, 2017). These are important factors to consider for achieving independence to enhance quality of life.

The difference in essentials for living, addressed by parents of AYA with and without SB, may be explained by the income difference between groups in this sample (Ridosh et al., 2016). Lower fiscal resources in families of AYA with SB may drive the present-oriented focus on basic needs for living. Opportunities for work were mentioned more frequently as a component of individual QOL by parents of AYA with SB and were also identified by AYA with SB (a job, money/financial stability). Therefore, targeted resources to build economic self-sufficiency are needed.

Income offers opportunities for social participation and may influence activities "beyond the essentials" mentioned by parents. Some of the activities (e.g., piano, dance) may be more easily supported by families who have more resources. Parents with AYA without SB, when describing "doing what they want to do" mentioned fun, ability to be active, and free time as making up QOL for the AYA. They also mentioned sports more specifically. For families of AYA with SB, programs that support adaptive sports participation are important to promote being active. Promotion of physical activity is supported by a recent study examining the relationship of lifestyle, participation in daily activities (e.g., personal care, housing, mobility, recreation), and HRQOL (measured by SF-36) in SB participants, when adjusting for sex and ambulatory status, those with higher levels of physical activity (min/day) had a higher physical HRQOL. No associations were found for mental HRQOL (Buffart, van den Berg-Emons, van Meeteren, Stam, & Roebroeck, 2009).

One of the striking differences in the descriptions of QOL between groups was in the "having friends" category. Having friends was endorsed by all groups as a part of QOL, but not as often by AYA with SB and their parents. When AYA with SB identified having friends they did not include any characteristics of friendship quality, while others did. In a longitudinal study of AYA with SB and peers, in the context of "best friends", there was no difference in self-reported friendship quality (e.g., companionship, security, and closeness) between groups over time, except for increased help from friends (Stiles-Shields, Driscoll, Rausch, & Holmbeck, 2019). When evaluating predictors of social skills over time, Holbein, Peugh, and Holmbeck (2017) confirmed that neuropsychological skills (attention and executive function), not family or health, was most important for social skill development. Opportunities for social skill development will be dependent on targeting specific neuropsychological deficits. Promoting prosocial behaviors in youth with SB may be useful to enhance the AYA's ability to establish friendships, enhance friendship quality, and contribute to QOL.

The number of "do not know" responses might also affect the priority of categories. For example, friends were nominated more in the AYA without SB, but that difference might disappear if the participants replying "do not know" had nominated friends. While the salient categories were identified from a rigorous process of content analysis, comparison of categories should be interpreted with caution based on this study alone.

Limitations

Although providing important information regarding the meaning of QOL for AYA, there are limitations in this analysis that need to be considered. First, the ethnic and geographic diversity of the sample are limited. The meaning might differ in a more diverse sample. Second, the sample represented a large age range. In this sample where the average age was 15, there was not sufficient data to segregate by developmental stage (early, middle, late adolescents and emerging adults). Using a larger sample or targeting specific age range, such as emerging adults, would allow us to better determine if there were differences by developmental stage. In addition, the first two authors coded the information together. It is possible that separate analysis might have identified different codes. However, the third team member review decreased this likelihood.

A major limitation of this study was the use of a single open-ended question embedded in a broader interview. This format, without the possibility of in-depth, follow-up questions, limits the quality of the data collected. Although the most salient categories were probably identified, there was no opportunity to expand on the initial brief response. In addition, the format used while appearing to work well for AYA without SB might have created challenges to the participants with SB who are known to have cognitive learning challenges in working memory, expressive language, and executive functioning. All participants were easily able to respond to structured items on instruments with defined response patterns. Generally, the participants were able to spontaneously articulate what QOL meant to them. However, we do not know if the AYA with SB that responded "do not know" did not know what the concept "quality of life" was or could not articulate what made up quality of life for themselves. The open-ended question may have been too unstructured or ambiguous for some AYA with SB. In a HRQOL instrument-development study using a more structured qualitative interview appeared to enhance the generation of important concepts (Szymanski et al., 2016). However, the identification of the salient categories generated from this project forms an important base for future studies of QOL in AYA.

Implications for research

AYA with SB, as research participants, may need accommodations to more fully articulate what QOL means to them such as a more focused in-depth interview, orientation to the definition of QOL, and the use of more concrete questions. In-depth qualitative interviews with sensitive probes are indicated to better describe the AYA perception of QOL and provide a rich description of the meaning and context of QOL for those with and without SB. Current measures do not use inductive methods to understand the meaning of QOL and none of the measures allow the participants to "weigh" or prioritize domains important to them. Expansion of existing comprehensive measures to include a ranking of importance may be helpful.

Further research can help us to understand the nature of the family, and whether it continues or diminishes as the AYA ages. Preliminary evidence, using the WHOQOL-BREF measure in a longitudinal analysis of young adults with SB, indicated that individual differences in family satisfaction predicted QOL (physical health, psychological, and environment domains) (Bellin et al., 2013).

Implications for practice

Assessment of QOL in clinical practice environments can be an important aspect of care delivery that is overlooked in many current

practices today. QOL was the most important outcome for the health care transition process as identified by international researchers and multi-disciplinary clinician members of the Health Care Transition Research Consortium for youth with special health care needs (Fair et al., 2016). Measuring QOL specifically remains important since HRQOL accounts for only a portion of QOL (69–75% of the variance in overall QOL) (Palimaru & Hays, 2017). Evaluating HRQOL alone does not allow the provider to get the whole picture. Also, evaluating QOL from multiple reporters (parent and child) remains important given the differences in meaning.

Providers can ask a single question to understand the meaning of QOL and a follow-up question to focus on areas of concern. A single question such as "*What is the meaning of quality of life for you*?" or "*What makes up QOL for you?*" can provide valuable information. Similar categories may be identified across reporters and the meaning and priority may be different or change for individuals. Thus when assessing QOL, asking a follow-up question "*What would make your QOL better*?" could be useful to understand "what is missing" and identify unmet needs. Parent information can be helpful for clinicians to provide anticipatory guidance in areas of discrepancy or concern. Both primary and secondary care providers can integrate QOL assessment in all adolescents' contacts.

An assessment made up of these data would facilitate providers to assess QOL based on what is most important to the individual and the family. Since an engaged family was most frequently reported as a component of the meaning of QOL in adolescents, it is important to assess family relationships in all families with AYA. Providers can inform parents of these findings so they can respond to the needs of their AYA such as "*Feeling loved and cared for*," "*Being able to talk to family*," and "*Feeling supported by family*." The PROMIS® *Family Relationships* instrument (Bevans et al., 2017) may be useful to assess baseline and changes in the quality of family relationship experiences from the perspective of children and their parents.

Providers can share with families of AYA with conditions such as SB, the trajectory of developmental task achievement. These families may need to build in additional time and provide support to address the development of independence and friendships. Findings from the International and Interdisciplinary Health Care Transition Research Consortium also indicated having a social network of friends was a highly rated transition outcome (Fair et al., 2016). It is important to understand these non-health or condition-related aspects of youth's lives. Particularly important is the development of social skills. Given the importance of neuropsychological skills (attention and executive function) for the development of social skills (Holbein, Peugh, et al., 2017), providers need to refer children for neuropsychological testing and programs targeted at promoting social skills training. Prosocial skills can be promoted to facilitate youth establishing friendships. In addition, language enrichment experiences, early in childhood, may be especially useful with the cognitive profile of AYA with SB. Addressing these areas have the potential to address AYA QOL by focusing on the building blocks of social skills early in life.

Finally, identifying ways to enhance social relationships such as friendship formation and peer relationships are essential to achieving developmental milestones in both AYA with or without chronic health conditions. Perhaps social media use may be leveraged for socialization, connectedness, and development of peer groups with similar interests. Since evidence does support the use of technology to improve self-management of chronic conditions (Parmanto et al., 2013), perhaps a pragmatic approach is to deliver targeted interventions for AYA which integrate the use of social media to facilitate social relationships while enhancing self-management.

Conclusions

In conclusion, the large number of participants in this study provided a context of meaningful findings from comparison groups of parents and AYA with and without SB. Similar categories were identified but often the descriptions of the categories varied. Engaged family as a central component of QOL for all was prominent. Clinicians and researchers need to better understand the definition of QOL for AYA, and how that may be different for youth with chronic health conditions as they transition to adulthood. The results can provide a basis for further qualitative studies to more fully explore the meaning of QOL, especially for AYA. Integrating what the parents and AYA themselves identify as components of their QOL provides needed direction for research and practice.

CRediT authorship contribution statement

Monique Ridosh: Conceptualization, Methodology, Data curation, Formal analysis, Writing - review & editing.**Kathleen Sawin:**Conceptualization, Methodology, Data curation, Formal analysis, Investigation, Writing - review & editing, Funding acquisition.**Gayle Roux:**Methodology, Formal analysis, Validation, Visualization, Writing - review & editing.**Timothy Brei:**Validation, Writing - review & editing, Funding acquisition.

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References

Abraham, S. (2013). Quality of life among adolescents with disability undergoing integrated education. *Research Journal of Recent Sciences*, *2*, 1–5.

Austin, J. K., Huster, G. A., Dunn, D. W., & Risinger, M. W. (1996). Adolescents with active or inactive epilepsy or asthma: A comparison of quality of life. *Epilepsia*, *37*, 1228–1238.

Bakaniene, I., Prasauskiene, A., & Vaiciene-Magistris, N. (2016). Health-related quality of life in children with myelomeningocele: A systematic review of the literature. *Child: Care, Health and Development*, *42*, 625–643.

Bellin, M. H., Dicianno, B. E., Osteen, P., Dosa, N., Aparicio, E., Braun, P., & Zabel, T. A. (2013). Family satisfaction, pain, and quality-of-life in emerging adults with spina bifida: A longitudinal analysis. *American Journal of Physical Medicine & Rehabilitation*, *92*, 641–655.

Bevans, K. B., Riley, A. W., Landgraf, J. M., Carle, A. C., Teneralli, R. E., Fiese, B. H., ... Forrest, C. B. (2017). Children's family experiences: Development of the PROMIS® pediatric family relationships measures. *Quality Life Res*, *26*, 3011–3023.

Boekeloo, B. O., Schamus, L. A., Simmens, S. J., & Cheng, T. L. (1998). Ability to measure sensitive adolescent behaviors via telephone. *American Journal of Preventive Medicine*, *14*, 209–216.

Buffart, L. M., van den Berg-Emons, R. J., van Meeteren, J., Stam, H. J., & Roebroeck, M. E. (2009). Lifestyle, participation, and health-related quality of life in adolescents and young adults with myelomeningocele. *Developmental Medicine and Child Neurology*, *51*, 886–894.

Coco, C. T., Meenakshi-Sundaram, B., Eldefrawy, A., Henry, M. L., 3rd, Watts, T., Aston, C. E., ... Slobodov, G. (2018). A cross sectional single institution study of quality of life in adult patients with spina bifida. *Neurourology and Urodynamics*, *37*, 1757–1763.

Davis, B. E., Shurtleff, D. B., Walker, W. O., Seidel, K. D., & Duguay, S. (2006). Acquisition of autonomy skills in adolescents with myelomeningocele. *Developmental Medicine and Child Neurology*, *48*, 253–258.

Dennis, M., & Barnes, M. A. (2010). The cognitive phenotype of spina bifida meningomyelocele. *Dev Disabil Res Rev*, *16*, 31–39.

Dennis, M., Landry, S. H., Barnes, M., & Fletcher, J. M. (2006). A model of neurocognitive function in spina bifida over the life span. *Journal of the International Neuropsychological Society*, *12*, 285–296.

Fair, C., Cuttance, J., Sharma, N., Maslow, G., Wiener, L., Betz, C., ... Ferris, M. (2016). International and interdisciplinary identification of health care transition outcomes. *JAMA Pediatrics*, *170*, 205–211.

Flanagan, S. M., Greenfield, S., Coad, J., & Neilson, S. (2015). An exploration of the data collection methods utilised with children, teenagers and young people (CTYPs). *BMC Research Notes*, *8*, 61.

Holbein, C. E., Peugh, J. L., & Holmbeck, G. N. (2017). Social skills in youth with spina bifida: A longitudinal multimethod investigation comparing biopsychosocial predictors. *Journal of Pediatric Psychology*, *42*, 1133–1143.

Holbein, C. E., Zebracki, K., Bechtel, C. F., Lennon Papadakis, J., Franks Bruno, E., & Holmbeck, G. N. (2017). Milestone achievement in emerging adulthood in spina

bifida: A longitudinal investigation of parental expectations. *Developmental Medicine and Child Neurology*, *59*, 311–316.
Jandasek, B., Holmbeck, G. N., DeLucia, C., Zebracki, K., & Friedman, D. (2009). Trajectories of family processes across the adolescent transition in youth with spina bifida. *Journal of Family Psychology*, *23*, 726–738.
Lin, J. H., Ju, Y. H., Lee, S. J., Lee, C. H., Wang, H. Y., Teng, Y. L., & Lo, S. K. (2009). Do physical disabilities affect self-perceived quality of life in adolescents? *Disability and Rehabilitation*, *31*, 181–188.
Lindsay, S., McPherson, A. C., & Maxwell, J. (2017). Perspectives of school-work transitions among youth with spina bifida, their parents and health care providers. *Disability and Rehabilitation*, *39*, 641–652.
Lollar, D. J. (1990). Learning patterns among spina bifida children. *Zeitschrift für Kinderchirurgie*, *45*(Suppl. 1), 39.
Lyneham, H. J., & Rapee, R. M. (2005). Agreement between telephone and in-person delivery of a structured interview for anxiety disorders in children. *Journal of the American Academy of Child and Adolescent Psychiatry*, *44*, 274–282.
Mpofu, J., Sefotho, M. M., & Maree, J. G. (2017). Psychological well-being of adolescents with physical disabilities in Zimbabwean inclusive community settings: An exploratory study. *Afr J Disabil*, *6*, 325.
Palimaru, A., & Hays, R. D. (2017). Associations of health-related quality of life with overall quality of life in the patient reported outcomes measurement information system (PROMIS®) project. *Applied Research in Quality of Life*, *12*, 241–250.
Parmanto, B., Pramana, G., Yu, D. X., Fairman, A. D., Dicianno, B. E., & McCue, M. P. (2013). iMHere: A novel mhealth system for supporting self-care in management of complex and chronic conditions. *JMIR mHealth and uHealth*, *1*, e10.
Ridosh, M. M., Sawin, K. J., Schiffman, R. F., & Klein-Tasman, B. P (2016). Factors associated with parent depressive symptoms and family quality of life in parents of adolescents and young adults with and without Spina Bifida. *Journal of Pediatric Rehabilitation Medicine*, *9*, 287–302.
Ridosh, M. M., Sawin, K. J., Brei, T. J., & Schiffman, R. F. (2018). A Global Family Quality of Life Scale: Preliminary psychometric evidence. *Journal of Pediatric Rehabilitation Medicine*, *11*, 103–114.
Ritchie, J., Lewis, J., Nicholls, C. M., & Ormston, R. (2013). *Qualitative research practice: A guide for social science students and researchers*. Thousand Oaks, CA: Sage Publications Inc.
Rofail, D., Maquire, L., Kissner, M., Colligs, A., & Abetz-Webb, L. (2014). Health-related quality of life is compromised in individuals with spina bifida: Results from qualitative and quantitative studies. *European Journal of Obstetrics, Gynecology, and Reproductive Biology*, *181*, 214–222.
Sawin, K. J., Brei, T. J., Buran, C. F., & Fastenau, P. S. (2002). Factors associated with quality of life in adolescents with spina bifida. *Journal of Holistic Nursing*, *20*, 279–304.
Sawin, K. J., Buran, C. F., Brei, T. J., & Fastenau, P. S. (2003). Correlates of functional status, self-management, and developmental competence outcomes in adolescents with spina bifida. *SCI Nursing*, *20*, 72–85.
Sawin, K. J., Buran, C. F., Brei, T. J., & Cashin, S. E. (2007). Individual and family factors associated with health-related quality of life in adolescents and young adults with spina bifida. *SCI Nursing*, *23*, 1–13.
Sawin, K. J., & Bellin, M. H. (2010). Quality of life in individuals with spina bifida: a research update. *Developmental Disabilities Research Reviews*, *16*(1), 47–59.
Stiles-Shields, C., Driscoll, C. F. B., Rausch, J. R., & Holmbeck, G. N. (2019). Friendship quality over time in youth with spina bifida compared to peers. *Journal of Pediatric Psychology*, *44*(5), 601–610.
Szymanski, K. M., Misseri, R., Whittam, B., Casey, J. T., Yang, D. Y., Raposo, S. M., ... Cain, M. P. (2017). Validation of QUALAS-T, a health-related quality of life instrument for teenagers with spina bifida. *Cent European J Urol*, *70*, 306–313.
Szymanski, K. M., Misseri, R., Whittam, B., Yang, D. Y., Raposo, S. M., King, S. J., ... Cain, M. P. (2016). Quality of Life Assessment in Spina Bifida for Children (QUALAS-C): Development and validation of a novel health-related quality of life instrument. *Urology*, *87*, 178–184.
Waters, E., Davis, E., Ronen, G. M., Rosenbaum, P., Livingston, M., & Saigal, S. (2009). Quality of life instruments for children and adolescents with neurodisabilities: How to choose the appropriate instrument. *Developmental Medicine and Child Neurology*, *51*, 660–669.
Wills, K. E., Holmbeck, G. N., Dillon, K., & McLone, D. G. (1990). Intelligence and achievement in children with myelomeningocele. *Journal of Pediatric Psychology*, *15*, 161–176.
World Health Organization Quality of Life (WHOQOL) Group (1998). Development of the World Health Organization WHOQOL-BREF Quality of Life Assessment. *Psychological Medicine*, *28*, 551–558.