

Adults with Childhood Onset Disabilities: A Focused Review of Three Conditions

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Abstract Adults with childhood onset conditions now have improved survival rates, and are experiencing health, medical, and performance issues related to their original conditions or treatments. The published science regarding the medical and performance issues of adults with cerebral palsy, spina bifida, and pediatric central nervous system cancer is increasing in quality, rigor, and clinical usefulness. This article is a systematized, focused review, with an appraisal of the literature, of these three conditions. Most of the published research is observational, from which recommendations can be made; however, there are few studies that demonstrate effective interventions. Although each of the three conditions has associated specific health and medical issues, they also have common problems and concerns related to health care, activity, performance, participation, and quality of life.

Keywords Cerebral palsy · Spina bifida · Pediatric brain neoplasm · Adults · Survivors · Health · Activity · Participation · Systematized review

Introduction

The number of published reports concerning aging and health conditions of adults with childhood onset disabilities

has increased since the early 1990s, but does not approach the number of published articles concerning the effects of disabling conditions during childhood. Because the USA has few national registries or databases that follow health and care needs of these individuals, long-term epidemiological studies of these conditions are difficult. Despite the increase in the number of publications about aging with childhood disability, there continues to be limited education about this topic for health professionals, even for those within rehabilitation professions. In addition, few clinicians receive knowledge updates or new information on this subject, and are unaware of the quality or rigor of scientific reports they may have read to support management choices. Not surprisingly, adults with disabilities, including childhood onset disabilities, report a lack of knowledge by health care providers related to their health concerns and issues [1].

This article will review the most recent information (2008–2012), with a focus on determination of design level and rigor, related to three conditions commonly encountered by physiatrists within adult and pediatric practices. Cerebral palsy and spina bifida compose the largest portion of diagnoses seen by pediatric physiatrists and those adult physiatrists willing to evaluate patients with childhood onset disabilities. Typically, practitioners who work with adults with disabilities have a limited awareness of the descriptive and observational reports regarding lifelong disability issues. Childhood cancer survivors, especially brain tumor survivors, are increasing in prevalence, and there are now more observational reports using national registries identifying longer-term issues these adults now face. Although physiatrists are not particularly engaged with this population at present, health and function-related conditions of these adults seem well suited to psychiatric specialty care.

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Methods

We have chosen to base our review of these topics on a rigorous formal quality appraisal review [2] of the literature from 2008 to 2012 to better help clinicians understand the significance of the science at present, which increasingly informs practice and evaluation of practitioners. In general, a comprehensive MEDLINE search was undertaken for each condition (cerebral palsy, spina bifida, and childhood or pediatric brain tumor) using that term then “adult” and/or “aging,” and then combining the results. Inclusion criteria were English language and publication year of 2008 forward. A gray literature search was also completed using Google and Google Scholar for each topic for relevant outcomes or observational research, again with limits of English language and publication year of 2008 forward.

Each article title and/or abstract was then initially reviewed to ensure the article represented observational, interventional, or review (with quality appraisal) research, and adults were represented in the article or adults were reported separately if comparisons were made with children or controls. Each article meeting that criterion was then reviewed fully, data were collected about the study design and outcomes, and the article was then rated according to “Reading the Medical Literature” (http://www.acog.org/Resources_And_Publications/Department_Publications/Reading_the_Medical_Literature) of the American Congress of Obstetricians and Gynecologists (with addition of reviews noting quality of evidence).¹ with an additional evaluation of rigor for the observational studies (since these were the most prevalent) using the STROBE statement on observational studies [3], identified by eight key items.² Qualitative research was assessed using a hierarchy of evidence approach [4].³ No commentaries, editorials, single case or case series reports, expert opinion, narrative reviews without quality assessment, or other articles of this type or of similar quality were included. There was crossover of the review process on all topics by the three authors to ensure

¹ Interventional studies: level I evidence randomized controlled trials; level II-1 evidence controlled trials. Observational studies: level II-2 evidence cohort and case-control studies; level II-3 evidence cross-sectional studies; level III evidence descriptive studies (case series, expert opinion). Other study designs: meta-analysis; decision analysis—modification to include systematic or other quality assessment reviews.

² STROBE items rated yes/no include Methods (setting, participants, variables, data sources, and statistical methods) and “Discussion” (key results, limitations, and interpretation).

³ Level I generalizable studies (methodologic sampling, analysis); level II conceptual studies (theoretical sampling, methodologic analysis); level III descriptive studies (practical sampling); level IV single case study (views of one subject).

consistency, with two authors involved for each topic; any discrepancies were decided by consensus. Descriptive tables were developed for articles fully reviewed and identified as level I or II; observational studies have the negative rigor ratings identified.

Results

In general, there were only a limited number of articles that were related to adults with these three conditions, and even fewer of methodological quality to be included in this report. Most of the articles concerned cerebral palsy. Most of the studies reported are observational, and most have a cross-sectional design, except those concerning pediatric cancers benefiting from registries; limitations in research were acknowledged, and most often related to small sample sizes, biases, differing levels of severity among subjects, and unvalidated measurement tools. Comparison of studies was limited since a variety of instruments were used. Case series describing outcomes from specific practices or regions and traditional literature reviews without literature appraisal, although serving a purpose for education and dissemination, are not included since their evidence level is rated low. We felt it was important to clearly identify the state of the science.

Cerebral Palsy

Cerebral palsy is a common diagnosis treated by physiatrists for rehabilitation or other health or functionally directed interventions to manage health and activity. Increasingly, it is recognized that function and health may change for adults with cerebral palsy over time and that there is a need for acknowledgement, recognition, and prevention (when possible) of issues related to aging [5].

The comprehensive MEDLINE search used the term “cerebral palsy” then “aging,” “adult,” and “pain” separately; each secondary term was separately combined with “cerebral palsy.” A total of 1,294 articles underwent title/abstract review, and 60 underwent more thorough review. Through a search of the gray literature using “cerebral palsy” and “adult,” an additional 19 articles were identified. A total of 28 articles were of sufficient level of evidence and rigor for presentation, and are noted in Table 1.

Health and Health Care

A survival rate of 80 % for individuals with cerebral palsy to age 40 years and highest mortality under the age of 15 years is supported by a recent Australian registry study that confirms previous work. Predictors for mortality are

Table 1 Adults with cerebral palsy table of evidence

Authors, country	Description (design, <i>N</i> , measures)	Results	Level; rigor ^a
Health and health care			
Asdaghi Mamaghani et al. [7], Germany	Cross-sectional, comparison analysis <i>N</i> = 62, 18–78 years; young aged less than 36 years vs older aged more than 36 years, 31 in each group Clinical examination, dental impression measurement, demographic/sociologic information	Older adults had less orthodontia, less dental crowding; decayed/missing/filled teeth prevalence lower than published data from disability data in Germany; 2/3 have 3 dental appointments per year; increased dentition irregularities associated with more malocclusion	II-3; L = no
Balandin et al. [8], Australia	Mixed methods: qualitative—individual interviews; quantitative—cross-sectional <i>N</i> = 32, 30–69 years Clinical dysphagia evaluation, in-depth semistructured interview	Swallow/gastrointestinal changes as early as age 30 years; emotional response to changes; poor collaboration with providers regarding decisions/interventions	Qual II
Bloom et al. [66], USA	Systematic review <i>N</i> = 13 meeting criteria Defined criteria, tables identifying criteria	Inconclusive evidence about transition programs Only rated evidence: weak for meeting adult provider prior to actual care transition	Systematic review
Guettard et al. [9], France	Case-controlled cohort, 1997–2007 <i>N</i> = 54, 17 developed cervical myelopathy, 10 with CP (58 % of cohort); 13 controls with CP (35 % of controls) Dystonia and function assessment, movements video-recorded, clinical signs determined by research team, MRI results, age onset symptoms	Onset after 36 years of age; age and severity of neck dystonia main risk factors; gait changes/falls, hand muscle wasting, change in urinary function best clinical clues; authors recommend screening beginning in the 3rd decade	II-2; P = no, L = no
Nieuwenhuijsen et al. [10], Netherlands	Cross-sectional <i>N</i> = 29, 16–40 years Southampton Needs Assessment Questionnaire, Impact on Participation and Autonomy Questionnaire, nonstandard health care use questionnaire	Unmet needs reported mostly regarding long-term CP information, mobility problems, health care access; more motor impairment, more unmet needs and more visits to many different providers; increased GMFCS level related to higher unmet needs and more visits	II-3
Reid et al. [6], Australia	Prospective cohort, Victoria, Australia, registry <i>N</i> = 3,507 (birth 1970–2004) Mortality, GMFCS, additional health or associated conditions, cause of death	Similar to previous population cohorts; crude mortality 20 % at age 40 years, highest mortality under age 15 years; predictors related to severity; respiratory causes commonest; no change in survival since 1970	II-2
Watson et al. [67], UK	Scoping review, transition of care models <i>N</i> = 19 articles met criteria Normalization process theory by transition categories framework	14 models for diabetes mellitus, 4 for CP, 0 for autism spectrum disorder; descriptions or time-limited pilots; little/no evaluation, key elements to implementation	Scoping review
Pain			
Hirsh et al. [12], USA	Cross-sectional survey <i>N</i> = 83, mean (SD) age 40.3 (13.6) years Survey of pain-related issues	Many types of treatments, seen by many types of providers; rarely used treatments reported best effects	II-3; D = no, St = no
Lai et al. [14], Hong Kong	Uncontrolled intervention Intervention: carry 4-lb backpack at work for 60 min on weekdays for 4 weeks <i>N</i> = 9 with chronic low back pain; mean age 42 years (26–63 years) Pain level rated 0–10, surface EMG paraspinals	Significant improvement of pain immediate on doffing pack but not lasting, reduced EMG activity during carrying; overall pain ratings exhibited decreasing trend, but not significant	II-3; L = no, I = no

Table 1 continued

Authors, country	Description (design, <i>N</i> , measures)	Results	Level; rigor ^a
Malone and Vogtle [16], USA	Cohort comparison walk vs no walk, uncontrolled, repeated measures 3 times <i>N</i> = 26 (12 walk); mean age 42.3 years [23–63] Faces pain scale, Pediatric Quality of Life Inventory, Multidimensional Fatigue Scale, Pain Disability Index	Pain/fatigue in both groups, consistent and significant; affect ability to participate	II-3
Opheim et al. [13], Norway	Prospective cohort, 7-year follow-up (1999–2006) <i>N</i> = 149 (288, 1999–66 % response rate); mean age 40 years Pain sites/questionnaire, SF-36; nonresponder analysis	Comparisons with 1999: overall more pain sites, increased prevalence of neck pain and decreased prevalence of knee pain, poor correlation between pain sites and psychological health, but correlation with physical health	II-2
Riquelme et al. [11], Spain	Cross-sectional comparison <i>N</i> = 86 CP [35 (40 %), 18–30 years]; 115 no CP [46 (40 %), 18–30 years] CP assessment, pain questionnaire (interview), touch sensitivity (von Frey monofilaments)	Pain commoner in CP group than in no-CP group at any age, decreased touch sensitivity and QOL (unrelated to age) in comparison with the no-CP group (all related to age)	II-3
van der Slot et al. [15], Netherlands	Cross-sectional (part of larger study), some comparison with no-CP Dutch reference sample <i>N</i> = 56, spastic bilateral, mean age 36 years, 5 months (SD 5 years, 10 months) GMFCS, Fatigue Severity Scale, Multidimensional Fatigue Inventory, Center for Epidemiologic Studies Depression Scale, questionnaire interview	CP higher prevalence of chronic pain and fatigue, and depression to a lesser extent; pain/fatigue co-occurrence in 1/3, and all 3 in 1/6; chronic pain/fatigue not associated with daily function	II-3
Activity/performance			
Brown et al. [22], USA	Uncontrolled intervention: upper limb training, computer/Web-based, training 40 min/day, 5 days/week for 8 weeks <i>N</i> = 12 (21–57 years), GMFCS I–III Motor activity log, 9-hole peg test, grip strength, duration movement, sensorimotor tasks	Affected limb had decreased reach time for unilateral/bilateral tasks, sequential reaching; improved dexterity; safe, convenient, and effective	II-3
Hombergen et al. [20], Netherlands	Systematic review <i>N</i> = 21 articles met criteria Criteria: adults, comparative design, components of fitness	Health-related fitness showed 3 components of lower performance in CP: muscular strength, muscular endurance, cardiorespiratory endurance; best evidence for muscular strength with moderate level, remainder only limited	Systematic review
Nieuwenhuijsen et al. [18], Netherlands	Cross-sectional, part of larger study <i>N</i> = 42, mean (SD) age 36.4 (5.8) years, spastic bilateral Personal/CP characteristics, aerobic capacity, accelerometer, self-reported activity, Fatigue Severity Scale	Aerobic capacity, activity duration and intensity below population norms; low levels of fitness, less active, and 1/2 experienced fatigue; little evidence for relationships between fitness and activity or fatigue	II-3
Nieuwenhuijsen, et al. [17], Netherlands	Cross-sectional, part of larger study <i>N</i> = 56 spastic bilateral CP, mean (SD) age 36.4 (5.8) years Personal/CP characteristics, accelerometer	Duration/intensity lower compared with age-matched, no CP; gross motor function significantly associated with level of everyday physical activity	II-3
Opheim et al. [21], Norway	Case-control cohort, 7-year follow-up study <i>N</i> = 16 (149 in original study), mean age 38 years [31–40]; 8 with walk changes Activities-Specific Balance Confidence Scale, Falls Efficacy Scale—International, Balance Evaluation Systems Test	No differences between 2 groups; balance confidence correlated with postural responses, sensory orientation, gait stability, Balance Evaluation Systems Test total score	II-2

Table 1 continued

Authors, country	Description (design, <i>N</i> , measures)	Results	Level; rigor ^a
Participation			
Gaskin and Morris [19], Australia	Cross-sectional, some comparison with national norms <i>N</i> = 51, mean age 38.2 years (19–66 years) Demographics, GMFCS (self-assigned), Physical Activity and Disability Scale, SF-36, Profile of Mood States—Short Form, Physical Self-Efficacy Scale, Multidimensional Scale of Perceived Social Support	Low-level activity, below general recommendations; variation in reported activities; lower scores on physical functioning in general, but high social functioning and support, minimal role limitation, low levels of negative mood; reported activity not associated with health-related QOL	II-3; S = no, P = no
Magill-Evans et al. [23], Canada	Mixed methods: qualitative—semistructured interview; quantitative—cross-sectional <i>N</i> = 76, 54 CP, 22 SB; age 20–30 years Questionnaire related to employment; interview related to preemployment/employment situations	Employment less likely: for females, for lower IQ, with transportation dependence; 4 themes: transportation barriers, social reactions to disability, being “stuck,” employment programs	II-3
Nieuwenhuijsen et al. [24], Netherlands	Mixed methods: qualitative—semistructured interview using COPM quantitative—cross-sectional <i>N</i> = 87, mean (SD) age 19.9 (1.4) years, COPM, GMFCS, MACS	Most prevalent life problems: recreation/leisure, preparing meals, housework, dressing; most important: functional mobility, paid/unpaid work, socialization; higher GMFCS correlated with more mobility and higher MACS level correlated with more self care needs	Qual II
van der Slot et al. [26], Netherlands	Cross-sectional <i>N</i> = 56, mean (SD) age 36.4 (5.8) years, spastic bilateral Demographics through interview, physical examination, Assessment of Life Habits 3.0, SF-36, General Self-Efficacy Scale-12	Difficulties with social participation, low health-related QOL for physical functioning; better participation and higher physical/mental-health-related QOL related to higher general self-efficacy or greater willingness to persevere (self-efficacy measure)	II-3
Wiegerink et al. [25], Netherlands	Mixed methods: qualitative—semistructured interview; quantitative—cross-sectional <i>N</i> = 87, 18–22 years Demographics, GMFCS/MACS by PT, paper-version questionnaire of Dutch study on sexuality, interview regarding social activity, friendships	All had friends, less romantic experiences compared with Dutch population; peer group activities and dating associated with romance and sex, older age associated with sex, no associations GMFCS/MACS, education, gender	II-3
Basic science			
Maltais et al. [27], Canada	Theoretic, measure validation <i>N</i> = 15, GMFCS I–II, age 21–41 years General physiologic parameters, GMFM-D/-E, portable metabolic system measurement	Feasible to measure steady state and net VO_2 6MWT construct validity measuring walking ability; net 6MWT VO_2 strongly related to GMFM-D/-E scores with body mass/speed controlled; reliability not determined	NA
De Groot et al. [28], Netherlands	Theoretic, measure validation <i>N</i> = 20, 18–49 years, GMFCS I–15, II–5; unilateral/bilateral 10 each Isometric/isokinetic knee flexor/knee extensor strength, sprint power, aerobic capacity on 3 occasions	Good feasibility for strength, Wingate cycling, graded exercise bike test; all tests (except isokinetic) suitable for evaluating group changes, but not for individual studies for change—strength measurement can improve with a mean of 2 separate day tests	NA
Riad et al. [31], Sweden	Applied, muscle volume differences <i>N</i> = 46, mean age 17.6 years, 13–24 years; control group <i>n</i> = 14, sex/age-matched (achieved power) MRI hip, knee ankle muscle volume and concentric muscle work in sagittal plane with 3-D gait analysis; Winter’s scale to assess movement patterns and involvement	Hemiplegic side smaller all but gracilis, concentric muscles lower work plantar flexors, knee extensors, hip flexors and extensors compared with uninvolved side; hip extensor work higher on hemiplegic/uninvolved sides compared with controls	NA

Table 1 continued

Authors, country	Description (design, <i>N</i> , measures)	Results	Level; rigor ^a
Satonaka et al. [29], Japan	Theoretic, measure validation <i>N</i> = 16 completed both tests (9 did not achieve maximum effort), mean (SD) age 43.7 (14.5) years, “athetospastic” Maximal cycle ergometer test plus multistage submaximal test, VO_2 peak compared with predicted VO_2 max	No significant difference between VO_2 peak values and predicted VO_2 max values, and significant correlation power required 23, possible overestimate of VO_2 max	NA
Van Meeteren et al. [30], Netherlands	Theoretic, measure validation <i>N</i> = 83, mean age 19.9 years, no intellectual disability Interobserver reliability MACS—compared with GMFCS, limb distribution, educational level and Melbourne assessment, Abilhand questionnaire, Functional Independence Measure self-care domain, life habits questionnaire	Good inter-rater reliability, correlated moderately with outcome measures	NA

P participants, *D* data sources, *St* statistical methods, *L* limitations, *I* interpretation, *CP* cerebral palsy, *GMFCS* Gross Motor Functional Classification System, *EMG* electromyography, *QOL* quality of life, *COPM* Canadian Occupational Performance Measure, *MACS* Manual Ability Classification System, *GMFM* Gross Motor Function Measure (*D* and *E* refer to dimensions of the measure), *NA* not available, *SB* spina bifida, *SD* standard deviation, *6MWT* 6-min walk test, *PT* physical therapist

^a *Level* refers to level of evidence, following “Reading the Medical Literature” of the American Congress of Obstetricians and Gynecologists. Quantitative noted with numerics, qualitative noted by *Qual* and numeric. For rigor, only negative responses are documented with initial of review category and “no.”

strongly related to severity, especially “not walking” [6]. Additional health conditions are now better described:

1. Dental care has improved in recent years, and in Germany, where vigilant care is well provided, adults with cerebral palsy have similar recognition of dentition irregularities when compared with the German general public [7].
2. Swallowing changes are noted by adults with cerebral palsy with aging, in some as young as 30 years old. These changes with their subsequent dining modifications (with little involvement of the adults themselves in decision-making) engendered negative emotional responses [8].
3. Cervical myelopathy is associated with age (more than 36 years) and severity of neck dystonia, with the best clinical clues being gait changes/falls, hand muscle wasting, and changes in urinary function [9]; Guettard et al. [9] recommend screening beginning in the third decade.

There are now consistent reports of unmet health care needs for young adults with cerebral palsy. As has been reported in other studies of adults with disabilities, young adults with cerebral palsy have reported difficulties receiving information about lifelong health, mobility problems, and access to care, despite having seen providers (including rehabilitation physicians and physical therapists) routinely [10].

Pain

Pain continues to be commonly reported by adults (and children) with cerebral palsy [11]. Common areas of pain complaint are the back, legs, and neck. The number of pain sites increases with age. Many interventions are offered, although they are of modest to minimal effect, and no single provider type offers the best support [11–13]. A single uncontrolled interventional study suggested use of a backpack to improve back pain, but the effect proved to be only modest and over a short time [14]. Chronic pain and fatigue, and to a lesser extent depression, compose a complex set of interdependent factors that affect the performance of adults with cerebral palsy and may be unrelated to level of function [15, 16]. Activity and physical therapy modalities most consistently provide some level of improvement [12, 13]. However, chronic pain with or without fatigue does not affect daily function or psychological health, unlike in the general population, which reports decreased physical and mental health with pain [13, 15]. There is no new guidance on pain management other than a reiteration that pain symptoms may be expected, and should be identified, evaluated, and treated, even at younger ages. Adults with lifelong disabilities appear to have some accommodation of chronic conditions, however the pain–fatigue (and possibly depression) complex has yet to be unraveled.

Activity/Performance

Activity is generally decreased in adults with cerebral palsy, although fitness has no relationship to physical activity or fatigue [17, 18]. Activity is well below the general activity recommendations to promote health and wellness [19]. A higher Gross Motor Function Classification System level (severer motor impairment) is associated with lower activity and performance. There is moderate evidence that muscular strength is a component for lower fitness in cerebral palsy, with more limited evidence that muscular and cardiorespiratory endurance contribute to lower fitness; there is no good evidence related to flexibility and body composition in cerebral palsy [20]. Therefore, although activity levels are important, there are no clear directions to improve physical activity for adults with cerebral palsy.

A closer look at interventions and targets of interventions to improve performance reveals (1) balance measurement instruments do not seem to be correlated to deterioration in walking abilities [21], and not surprisingly, postural responses and adjustments seem to relate more to changes in walking, and (2) a novel upper limb program, using a computer/Web-based training and measurement system, can improve function, although long-term effectiveness has not been determined [22].

Participation

As expected, adults with cerebral palsy face difficulties engaging in social activities such as employment, recreation and leisure activities, and preparing meals, have limited access to transportation, and have limited dating and romantic experiences [23–26]. Higher severity may be related to lower employment: lower IQ and a need for transportation assistance (along with female gender) accounted for low employment in a mixed-methods study with adults with cerebral palsy and spina bifida [23]. However, there are opposing views regarding socialization; less socialization is reported with higher Gross Motor Function Classification System and Manual Ability Classification System category [24]; however, social functioning and support, friendships, and romantic experiences may have no associations with severity [19, 25]. The relationship of these social activities with health-related quality of life is also not clear [19, 26], and these conflicting reports may be related to differences in locales, available support, levels of function of the study participants, and the variety of measurements used. There is a further suggestion that self-efficacy, effort or perseverance, or adaptation may account for differing responses to barriers to socialization. This concept of resiliency continues to be elusive.

Basic Science

In the spirit of promoting interventional research, there is now standardization of routine measurements for adults with cerebral palsy. Many of these measures have been used in past research, and use of them (net VO_2 6-min walk test, strength measurement, predicted VO_2max , Manual Ability Classification System) was reported as a limitation of design and outcome interpretation [27–30]. More research is being directed at muscle parameters in cerebral palsy, possibly to better focus interventions. A single study measured muscle volume and concentric muscle work during walking to evaluate the value of a strengthening program; however, it was unable to identify exactly on which muscle groups a strengthening program should be focused to improve gait. However, the identified increased bilateral hip extensor work in hemiplegia demonstrated the importance of proximal strengthening [31].

Spina Bifida

There has been a significant increase in interest about the health and quality of life of adults with spina bifida, although the number of publications remains less than that for adults with cerebral palsy. Medical and surgical advances developed in the 1960s have now provided the first relatively large cohort of adults, who in some cases demonstrated unique health and aging issues.

A comprehensive MEDLINE search using the term “spina bifida” then “adult” and combining the results was completed. Forty-six articles were retrieved using this method; a gray literature search using the terms “spina bifida” and “adult” identified an additional 41 unique articles and proceedings. Fourteen articles met all the criteria and are included in Table 2.

Health and Health Care

A number of reviews or case series reports from clinics for adults with spina bifida have been published in the past 5 years [32–35], with neurosurgical, urological, musculoskeletal, and cardiopulmonary issues identified which demand specialty care beyond that of a typical primary care provider. A number of additional medical issues have emerged as well. Obesity appears to be a significant concern in adults, with rates in the 37 % range [36, 37]. However, sadly this is not greatly different from the 35 % for the general adult US population reported by the Centers for Disease Control and Prevention (<http://www.cdc.gov/obesity/data/facts.html>). Cardiovascular disease risk factors were reported in a Dutch sample, with 42 % of adolescents and young adults with spina bifida at increased risk, and of those, 61 % were nonambulatory [38]. Increased risk was

Table 2 Adults with spina bifida table of evidence

Authors, country	Description (design, <i>N</i> , measures)	Results	Level; rigor ^a
Bellin et al. [41], USA	Cross-sectional, data from year 1 longitudinal study <i>N</i> = 61, age 18–25 years Attitude toward illness, satisfaction with family functioning, patient assessment of chronic illness care, Hopkins Symptom Checklist	Pain, attitude, and family satisfaction correlated with depression. Only pain correlated with anxiety	II-3
Buffart et al. [38], Netherlands	Cross-sectional <i>N</i> = 31, age 16–30 years Biologic CVD risk factors, physical activity, aerobic fitness, body fat	Aerobic fitness associated with decreased CVD risk; 42 % of the total had increased CVD risk; 61 % of nonambulatory individuals had increased CVD risk	II-3
Chan et al. [48], USA	Cross-sectional <i>N</i> = 63, age 18–80 years World Health Organization Medical Outcomes Study 26-Item Short Form, CHART, questionnaire on friends and virtual socializing	Increased virtual socializing associated with self-reported increased number of friends; trend toward increased QOL with increased virtual socialization	II-3; V = no, D = no, L = no, I = no
Darrah et al. [49], Canada	Qualitative—open-ended questions/ interview <i>N</i> = 76, age 20–30 years; 22 SB, 54 CP Semistructured interviews with content analysis	Need for individualized service planning, transportation, and mainstream education	Qual II
Dennis et al. [46], Canada	Cross-sectional <i>N</i> = 26 SB with IQ > 70, <i>N</i> = 27 typical matched for age and gender, but not IQ 5 motor tests under 4 visual and cognitive challenge conditions; chart reviews	The SB group scored significantly lower on all tests, worse with distractions or dual tasks	II-3; P = no, D = no, St = no, L = no
Dennis et al. [45], Canada	Cross-sectional <i>N</i> = 32 SB, <i>N</i> = 17 typical matched for age, ethnicity, and SES but not IQ 8 tests or interview scales, some standardized	Prospective memory was poorer in the SB group and much worse in the older group	II-3; V = no, D = no, L = n, I = no
Dosa et al. [36], USA	Cohort, within clinical program—one-time measure <i>N</i> = 203, 3 age ranges: 94 aged more than 20 years BMI from records, self-report activity	Obesity rates were 18 % for children, 8 % for adolescents, and 37 % for adults; publicly insured individuals, females, and individuals with lower motor level were significantly more likely to be obese	II-3; D = no
Garcia et al. [37], USA	Cohort, within clinic program—one-time measure <i>N</i> = 232, mean (SD) age 35.1 (11.1) years Diagnosis of lymphedema in adults with SB; medical record review	9.2 % had lymphedema; significant associations were trauma, cellulitis, cancer, obesity, wounds, hypertension, higher lesion level, and mobility status; 38.3 % had hypertension and 37.5 % were obese	II-3
Mahmood et al. [39], USA	Cross-sectional, multisite <i>N</i> = 38, age 18–25 years; mean (SD) age 22.2 (2.0) years Chronic care model: PACIC; questionnaires: health care utilizations, preventable conditions; AMIS (structured interview); review of records	Increased urinary tract infection associations—no shunting, lower educational level, higher employment levels, lower AMIS scores; pressure sore associations—higher motor/education level; increased hospitalization associations—more wounds and lower AMIS scores	II-3

Table 2 continued

Authors, country	Description (design, <i>N</i> , measures)	Results	Level; rigor ^a
O'Mahar et al. [43], USA	Uncontrolled investigational Intervention: goal setting for independence; group sessions for cognitive strategies, catheterization, and hygiene; counselor monitoring goal attainment <i>N</i> = 74; adults 18–37 years, 27 of 33 participated GAS, SOSBMR, SBIS, KOSB, self-reports/parent reports	GAS highly significant and maintained after intervention, SBIS and KOSB were of moderate significance for adults, not younger age groups	II-3
Soe et al. [42] USA	Cross-sectional, compared with general population surveys/estimates <i>N</i> = 130, age range 12–31 years, 73 % aged 20–31 years Center for Epidemiologic Studies Depression Scale, health-related behavior survey; interview and self-completed questionnaires	Substance abuse highest for ages 25–31 years; depression 50 % across age groups; alcohol use correlation: depression, higher level of education; fair–poor health reports trend to increase with age; compared with age-matched general population health-risk behavior (exercise, diet, TV, and depression) higher and substance abuse lower at younger ages	II-3; D = no
Treble et al. [47], USA	Applied, clinical correlations, compared with typical <i>N</i> = 64, 27 (42 %) SB, remainder typically developing; age 8–28 years MRI software for cortical thickness and gyrification; IQ/fine motor scales	Increased or decreased thickness and gyrification was associated with lower IQ and poor fine motor outcome, suggesting functional maladaptation	
Young et al. [40], Canada	Cross-sectional <i>N</i> = 40 youths (mean 16 years), 13 adults (mean 26.6 years) Health Utilities Index, assessment of QOL, self-rated health	Self-rated health favorable, other measures lower and correlated with the level of lesion; adults rated health lower than youths	II-3; L = no
Zuckerman et al. [44], USA	Prospective cohort <i>N</i> = 68 each group initially, 52 SB and 60 controls at final observation Demographics, PPVT, Behavioral Rating Inventory of Executive Function, Cognitive Assessment System, Harter, parental intrusiveness, independence, academic achievement, employment status, romantic and peer relationships	Emerging adults with SB were less likely to leave home, attend college, maintain employment, have romantic relationships; had peer relationships; controlling for high school completion: only romantic relationships were less likely; executive functioning, intrinsic motivation and parental intrusiveness important predictors for achievement	II-2

P participants, *V* variables, *D* data sources, *St* statistical methods, *L* limitations, *I* interpretation. *CVD* cardiovascular disease, *CHART* Craig Handicap Assessment and Reporting Technique Short Form, *QOL* quality of life, *SB* spina bifida, *SES* socioeconomic status, *BMI* body mass index, *PACIC* Patient Assessment of Chronic Illness Care, *AMIS* Adolescent Self-Management and Independence Scale, *SOSBMR* Sharing of Spina Bifida Management Responsibilities, *SBIS* Spina Bifida Independence Survey, *KOSB* Knowledge of Spina Bifida, *PPVT* Peabody Picture Vocabulary Test, *Harter* Harter Scale of Intrinsic Versus Extrinsic Orientation in the Classroom

^a *Level* refers to level of evidence, following “Reading the Medical Literature” of the American Congress of Obstetricians and Gynecologists. Quantitative noted with numerics, qualitative noted by *Qual* and numeric. For rigor, only negative responses are documented with initial of review category and “no.”

not associated with physical activity or body fat. Lymphedema frequency was reported by a single study as nearly 100 times the rate of that in the general population [37]. It was associated with trauma, cellulitis, cancer, obesity,

wounds, hypertension (38.3 %), level of lesion, and mobility status. Increased number of skin ulcers was associated with higher motor level and higher educational level in one study [39]. Higher number of hospitalizations

was associated with the number of wounds and lower self-management scores [39].

As in adults with cerebral palsy, pain was noted in a health-related quality of life survey more frequently in young adults than in youths [40]. High rates of depression, anxiety, and pain were found and were closely related to attitude and family functioning [41]. Similarly, Soe et al. [42] found that nearly half of the subjects studied reported depressive symptoms.

Researchers have addressed health promotion through descriptions of health risks. In general, physical activity is decreased in adults with spina bifida [38, 42], and walking was noted to decrease as adults aged [40]. Less healthy diets, less exercise, and more sedentary behaviors were reported in young adults with spina bifida, with a peak in substance abuse in their late 20s [42]. Self-management and early detection of wounds and urinary tract infections are suggested to decrease the incidence of complications [39], although no specific recommendations were offered. O'Mahar et al. [43] described a novel intervention targeting independence and self-management through a camp experience. Through goal identification and group session activities, campers and parents noted improvement in personal management and social goals, and to a lesser extent in taking responsibility and being independent with self-management tasks at the end of the period. Both studies acknowledged the need for approaches specifically focused on the cognitive needs of people with spina bifida.

Cognition

Cognitive changes are suspected earlier with aging and spina bifida. Many young adults with spina bifida demonstrate decreased executive function abilities [44]. In a comparison study of adults with and without spina bifida, memory problems were found at a younger age as well as less use of compensatory strategies, with no correlation of poorer memory with shunt history [45]. Upper extremity dexterity may be compromised in adults with spina bifida, particularly when distractions are present, which has implications for rehabilitation and eventual successful employment [46]. Treble et al. [47] looked at the effect of cortical thickness (measured by MRI) on IQ and fine motor dexterity in typically developing people and individuals with spina bifida. The more the thickness and level of gyrification deviated from typical (increased or decreased), the more impaired the subjects were on testing.

Participation

Most young adults with spina bifida do not live independently, have low rates of employment, and are less likely to have romantic relationships. A longitudinal study following

the transition from adolescence to adulthood for youths with and without spina bifida noted that youths with spina bifida have difficulty achieving adult milestones within the same time period as their peers. Typically developing subjects were more likely to leave home, attend college, become employed, and develop social relationships. When high school graduates were compared, there was no statistically significant difference except in romantic relationships, where development of romantic relationships for people with spina bifida was delayed. Youths with greater executive functioning are more likely to be successful [44]. Virtual socialization may contribute to the number of friends one identifies, but it does not affect quality of life [48].

As noted for cerebral palsy, 46 % of a sample of young adults with cerebral palsy and spina bifida were not employed, which was related to gender, cognitive scores, and transportation dependence. Qualitative themes in data analysis concerned transportation barriers, social reactions to disability, inability to advance within the job market, and employment programs that did not meet needs [23]. A further qualitative study was done with the same participants, and although transportation continued to be a roadblock, individualization and flexibility of services over time emerged as key requirements for effective support for people with disabilities [49].

Although more information describing the limitations is available, specific recommendations have been offered about interventions or support to improve levels of social participation.

Childhood Onset Brain Cancer Survivors

Advances in childhood cancer treatment have increased the number of survivors into adulthood, and with that, there has been increasing interest in the long-term health of survivors and late effects of their cancer treatments. Many countries, including the USA, have established registries to allow long-term follow-up. The Childhood Cancer Survivor Study (<http://www.cancer.gov/cancertopics/coping/ccss>), funded through the National Cancer Institute/National Institutes of Health, monitors survivors (and siblings) originally diagnosed more than 30 years ago by cohorts, and involves more than 20 US and Canadian centers.

As for the two previous conditions, a comprehensive MEDLINE search was completed from 2008 to 2012 using the term “childhood cancer” then “brain tumor” and then “adult” and “survivor” for a combined total of 86 articles. A gray literature search using “childhood CNS (central nervous system) cancer” and “survivor” found an additional 25 articles. Additional inclusion criteria were applied to focus on rehabilitation-related articles. Seventy articles were found; 15 met the detailed review criteria for reporting here, and are reported in Table 3.

Table 3 Adult survivors of childhood central nervous system cancer

Authors, country	Description (research type, <i>N</i> , measures)	Results	Level; rigor ^a
Armstrong et al. [50], USA	Cohort, CCSS <i>N</i> = 2,821; age 20–24–years Medical records/mortality, chronic medical conditions, cognitive functioning	Survivors at greater risk of late mortality and developing subsequent neoplasms and chronic medical conditions (endocrine, neurological)	II-2
Armstrong et al. [53], USA	Cohort, 5 years or more of follow-up of CCSS registry <i>N</i> = 818 (523 with astrocytoma) Radiation region/dose (records); Neurocognitive Questionnaire; self-report cognition; SF-36	High radiation dose to temporal region—higher risk of memory impairment, social/general health problems; frontal region exposure—general health/social problems	II-2; St = no
Bowers et al. [57], USA	Cross-sectional clinical program survey Response rate 74 % Survey, not standard	Significant variations; neuro-oncology long-term follow-up programs used neuropsychologic testing after radiation therapy, longer imaging surveillance, growth hormone use; barriers: insurance, funding/dedicated time providers, survivors' preference not pediatric center, questioning need	II-3; S = no, V = no
Brinkman et al. [64], USA	Mixed methods: cohort survivors and population comparison, registry plus cross-sectional survey <i>N</i> = 78 (median 22 years), controls <i>N</i> = 78 (median 25 years) and matched Medical records, demographics/past history, 7 standardized tools, and nonstandard questionnaire	Survivors comparatively lower environment access/engagement, health-related QOL, social participation; limited environmental access associated with decreased health-related QOL	II-3; P = no
Ellenberg et al. [60], USA	Cross-sectional <i>N</i> = 802; minimal 16 years of survival Neurocognitive Questionnaire to assess task efficiency, emotional regulation, organizational skills and memory	Task efficiency, memory most impaired; increased risk with cranial radiation therapy, shunt placement, had stroke or persistent hearing/motor impairment or motor impairments; adverse affect on education, employment, income, and marital status	II-2
Ishida et al. [58], Japan	Cross-sectional, 3 comparison groups <i>N</i> = 185 survivors, 72 sibs, 1,000 population; mean age 23 years, mean 12 years since completion of treatment Nonstandard survey	Survivors maintained ties with original treating (likely pediatric) hospital; survivors with brain tumors/sarcomas, late effects maintained ties; married less likely to continue relationship	II-3
Kirchhoff et al. [62], USA	Mixed methods: cohort survivors and population comparison, registry plus cross-sectional survey <i>N</i> = 6,339 survivors, control <i>N</i> = 1967 sibs, age 25 years or older, stratified by ages; 13 % CNS Registry data; questionnaire nonstandardized	Childhood cancer survivors higher unemployment; those with CNS cancer most commonly unemployed owing to health; highest risk unemployment, but seeking work: high-dose CNS radiation therapy	II-3
Koch et al. [63], Denmark	Cohort, control; Danish registry (cancer and population) <i>N</i> = 1,877, 25 % CNS cancer; control <i>N</i> = 45,499, age/sex-matched Data analysis of registry items	Lower rate of marriage/cohabitation for those with childhood cancer, and more pronounced CNS tumors, especially males	II-2; P = no
Krull et al. [59], USA	Theoretic, measure validation <i>N</i> = 7,121 (6,739 survivors and 382 sibs) Questionnaire, comparison with cognitive/emotional measures on subset survivors 7 years prior	4 reliable measures: task efficiency, emotional regulation, organization, memory skills; demonstrated excellent reliability, and construct/discriminative validity	
Krull et al. [56], USA	Cohort, control; baseline and 7-year follow-up <i>N</i> = 1,652, 9 % CNS tumors; control <i>N</i> = 406 (sibs) Behavior Problem Index by parents, BMI, health behaviors survey, physical activity	In general: higher rates inattention, social withdrawal, emotional issues; stimulants/antidepressants use unrelated to behavior reports; for CNS tumor: higher risk inactivity	II-2

Table 3 continued

Authors, country	Description (research type, <i>N</i> , measures)	Results	Level; rigor ^a
Lancashire et al. [61], UK	Mixed methods: cohort survivors single survey, compared with national survey data <i>N</i> = 10,138; cohort—diagnosis 1940–1991 Questionnaire nonstandardized, compared with national survey comparable items	Poorest education attainment for those with leukemia/CNS cancer; high risk: brain radiation therapy, CNS tumor, older age completing survey, younger age at diagnosis, epilepsy, female gender	II-3; V = no
Ness et al. [54], USA	Mixed methods: cohort survivors and population comparison, registry plus cross-sectional survey <i>N</i> = 78 cohort: median age 22 years; median 14.7 years after diagnosis ; <i>N</i> = 78 control, median age 25 years Demographics, 6 standardized, not all validated for cancer survivors, open-ended questions	Survivors: weakness and poor exercise tolerance similar to performance at age more than 60 years; limited physical performance associated with poor outcomes at home/school	II-3
Ness et al. [55], USA	Cohort, control <i>N</i> = 9,301 survivors aged 18 years or older when 2003 questionnaire sent; control <i>N</i> = 2,886 sibs 2003 follow-up questionnaire with physical activity question plus routine standardized measures	Survivors less active and participated in fewer leisure activities than sibs and population comparison; medulloblastoma/osteosarcoma highest inactivity; associations: cranial radiation therapy, amputation, female gender, black race, older age, lower educational attainment, weight extremes, smoking, depression	II-3
Taylor et al. [52], UK	Cohort, UK childhood cancer registry <i>N</i> = 10,483 (79 % response), diagnosis 1940–1991; age 16 years or older Questionnaires regarding 2nd primary cancer diagnosis; cancer registry, link to national cancer/death registry	Commonest 1st neoplasm: CNS (47.4 %); 247 (95 %) CNS 2nd primary tumors—known consequence of brain radiation therapy; poor survival after 2nd primary glioma, but good after 2nd primary meningioma	II-2
Whelan et al. [51], USA	Mixed methods: cohort survivors and population comparison, registry plus cross-sectional survey <i>N</i> = 14,358 survivors; <i>N</i> = 4,023 controls; 72 % more than 18 years old Questionnaires regarding hearing problems/loss, tinnitus, deafness	13 % CNS tumors, 72 % cranial radiation therapy, 82 % no platinum chemotherapy; radiation therapy and use of platinum compounds significant increased risk of auditory complications	II-3

S setting, *P* participants, *V* variables, *St* statistical methods, *CCSS* Childhood Cancer Survivor Study, *QOL* health-related quality of life, *CNS* central nervous system, *BMI* body mass index

^a Level refers to level of evidence, following “Reading the Medical Literature” of the American Congress of Obstetricians and Gynecologists. Quantitative noted with numeric. For rigor, only negative responses are documented with initial of review category and “no.”

Health and Health Care

Survival of childhood central nervous system (CNS) cancers has high risk of late mortality beyond 5 years, and survivors are noted to have late effects or secondary conditions related to neurologic, sensory (especially auditory), endocrine, and musculoskeletal systems [50, 51]. Additionally there is an increased risk, compared with a general population of siblings, of developing other neoplasms, especially second primary brain tumors [50, 52]. The incidence of neurocognitive impairments is high, and they relate to radiation doses and sites of radiation therapy [50, 53]. Physical performance is limited for adult survivors of childhood brain tumors as noted in a cross-sectional study [54]; in particular, survivors of medulloblastoma and osteosarcoma had the highest rates of inactivity, and

cranial radiation therapy, amputation, female gender, black race, older age, lower educational attainment, extremes of weight, smoking, and depression had high associations with inactivity [55]. Muscle strength and fitness values for the survivors were similar to standard and comparative values for individuals aged 60 years or older, with limited physical performance and poorer management of home and school activities. Survivors of CNS tumors are at risk of obesity, inactivity, and an increasing number of chronic conditions [56]. Natural target areas for rehabilitation activities would be within the realms of activity level, weight management, and performance. Adult survivors often maintain their follow-up services within pediatric center delivery networks [57, 58]; although this was a preference for some survivors, a number of barriers were noted in maintaining that activity.

Cognitive and Psychological Issues

Cognitive impairments are common, and pediatric oncology centers often include neuropsychologic testing in their routine follow-up. The tool used within the US registry studies, the Neurocognitive Questionnaire, has been standardized for this population [59]. Through the Childhood Cancer Survivor Study network, associations have been found among radiation dosing, site of radiation therapy, neurocognitive issues, and social participation; exposure of temporal brain regions to radiation is related to increased risk of memory and social functioning impairments and general health problems, whereas frontal brain exposure is associated with physical performance and general health problems [53]. Additional factors (e.g., shunt placement, gender, and diagnosis before 2 years) are implicated in affecting cognitive function [60]. Survivors of pediatric leukemia and CNS tumors are at higher risk of depression and anxiety, attention deficits, oppositional behavior, and social withdrawal than individuals with other pediatric cancer diagnoses [56].

Participation

As cancer survivors age, there is a need for monitoring of long-term social consequences of tumors and treatment. A registry survey study from the UK has documented that survivors of childhood cancers are at a higher risk of lower educational attainment with cranial radiation therapy, CNS tumor diagnosis, older age at completion of the questionnaire, younger age at diagnosis, having epilepsy, and being female [61]. Again, cranial radiation therapy has been found to be associated with higher unemployment rates [62]. Pronounced reductions in marriage/cohabitation are seen in survivors of CNS tumors [63]. Adult survivors of pediatric brain tumors tended to limit their participation and interaction with the environment, which is associated with reduced health-related quality of life [64]. In general, low physical and cognitive performance, and less social participation, is common for adult survivors of pediatric onset CNS cancer. Many recommendations to facilitate social and performance success are offered within these publications that involve rehabilitation and psychiatric strategies, although there have been no controlled interventional studies to show effectiveness.

Models of Transitional Care

Health care transition is a topic of interest for individuals with complex care needs and their care providers, and deserves a separate section. Although there is much interest in this topic in the literature, there is very little evidence to define the key components, evaluation structure, or

outcomes of such programs. There are publications discussing this theme for all three childhood onset conditions. A qualitative study with a small group of adults with a variety of childhood onset disabilities identified challenges to transition of care—lack of access to health care, lack of professionals' knowledge, lack of information regarding the transition process—and offered the solutions of providing early detailed information about and extensive support for the process [65]. Two review articles about health care transitions for adolescents with complex medical needs note there is inconclusive evidence about key components for transition programs and what constitutes effective transitional care [66, 67]. A survey of long-term follow-up programs in the USA for survivors of childhood brain tumors noted considerable variations in services and organization across the country. Additionally, the survey offered barriers to establishment of programs: lack of insurance, lack of funding or dedicated time for professionals, patients' uncertainties about the need for follow-up, and patients' desire not to be followed in a pediatric program [57]. Transition programs may theoretically make sense and consumers may report high satisfaction conceptually, but there are no clear guidelines or outcome studies to promote their implementation.

Conclusion

Survival of people with these three childhood onset conditions into adulthood has enabled cohort studies of associated health and performance issues. Although these reports have increased in quality and rigor, especially when national registries or networks are available, most are observational, and there is little interventional research. Some of the reported studies have validated established measures for these special populations. The availability of these tools should facilitate future interventional studies.

Health providers should anticipate secondary conditions/late effects, and health risks for each condition. Adults with childhood onset disability require increased complexity of services and specialized understanding by providers. Current published information shows that there are many commonalities among these three conditions. These consumers are requesting more information from their health care providers and many are seeking participation in care decisions. Low levels of activity, performance, and participation are prevalent, and they may relate to the severity of the impairment. Observational studies often recommend improved accessible exercise opportunities, and more self-management, cognitive rehabilitation, pain management, and health promotion programs. These resources are, of course, key components of comprehensive rehabilitation, and consumers, advocacy groups, and practitioners now

recognize the usefulness of psychiatric and rehabilitation strategies. These suggested interventions await further rigorous research to demonstrate effectiveness.

Several key concepts remain elusive, despite the many observational studies. What are the relationships among chronic pain, fatigue, and mood? What role does resiliency and self-efficacy play in long-term outcome, and can resiliency be taught or modeled? What are the relationships among cognitive impairments, “motivation,” and self-efficacy? How can exercise influence fitness, fatigue, pain, and performance over a lifetime with a disability? What strategies are useful to modify present activity and social outcomes? Although we have ever more information, much research remains to be done.

Disclosure

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