FOCUSED REVIEW

Aging With a Disability

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This focused review highlights important issues in the care of persons who are aging with a disability. It is part of the study guide on geriatric rehabilitation in the Self-Directed Physiatric Education Program for practitioners and trainees in physical medicine and rehabilitation. This article specifically focuses on significant medical and rehabilitation issues pertinent to persons with cerebral palsy, spina bifida, postpoliomyelitis syndrome, and selected other neurologic and neuromuscular diseases. In addition to normal physiologic aging, people with these conditions often experience secondary complications and accelerated impairments because of aging itself. These complications are described, and monitoring strategies and treatment are recommended.

Overall Article Objective: To summarize issues in the care of persons aging with a disability.

Key Words: Aging; Cerebral palsy; Postpoliomyelitis syndrome; Multiple sclerosis; Rehabilitation; Spina bifida.

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PEOPLE WITH CONGENITAL OR early acquired disabilities are increasingly surviving into older ages. This fact requires a better understanding of the phenomenon of aging with a disability. Medical complications are related to the natural history of the original disability and often include nutritional problems, bowel and bladder dysfunction, respiratory complications, and progressive neurologic dysfunction. Muscle weakness and other biomechanic alterations that increase joint and muscle stress can result in decreased functioning and an acceleration of normal aging. Frequently, patients who are closely managed by their pediatric physicians are lost to regular follow-up when they become adults. This is unfortunate because regular physiatric follow-up may prevent or minimize the functional losses and progressive disability that is commonplace.

Depression and lower life satisfaction are more common in persons experiencing the later life effects of early life disabilities, but family functioning and attitudes toward disability are important mediating factors. Intellectual functioning influences vocational attainment more than physical disability, although some handicap in social and occupational integration is commonly reported. This review provides the practicing physiatrist with a basic knowledge of the issues of aging with the common childhood-onset disabilities.

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SPINA BIFIDA

Spina bifida, or myelodysplasia, is a group of developmental defects occurring during formation of the neural tube. This group of neural tube defects can range from anencephaly to spina bifida occulta. The terminology used is descriptive of the lesions. Spina bifida occulta refers to a posterior element defect of the spine with skin covering the lesion and no protrusion of neurologic elements. Generally, there are minimal neurologic deficits. Meningocele is a defect of the posterior spinal elements with a cystic, skin-covered lesion protruding from the back. Usually no neural elements are contained in the sac, and they have minimal neurologic defects. Myelomeningocele is a defect in the posterior spinal elements with neural structures found within the thecal sac leading to major neurologic deficits.

Adults with spina bifida must deal with the normal physiologic issues related to aging in addition to the associated secondary complications of their disability. This interrelationship has some similarities to that experienced by the population aging with spinal cord injury. It is now believed that these complications previously thought to be static in nature can have an increasing impact on functional ability with age. In treating the adult patient, one has to evaluate the usual age-related medical problems as well as the issues unique to this population.

Spina bifida is associated with abnormalities in the brain, including hydrocephalus and Arnold-Chiari type 2 malformations. Of the adults coming to a clinic, at least 90% will generally have ventricular-peritoneal shunts in place. Shunt malfunctions are not uncommon and present with chronic headaches, vomiting, personality changes, difficulty concentrating, and other neurologic symptoms. Shunt malfunction can lead to significant morbidity, mortality, and sudden death. Adults should be followed with routine neurologic examinations as well as periodic computed tomography scans. Current treatment includes the use of neuroendoscopic third ventriculostomy and shunt revisions. Adults can also develop signs and symptoms of tethered cord syndrome at any age. Any change in neurologic status, bowel or bladder changes, increasing orthopedic deformities, and gait variations warrants a thorough evaluation by the medical team. This assessment includes not only a complete motor and sensory test but also magnetic resonance imaging and urodynamics where indicated.

Inherent in spina bifida are associated musculoskeletal and orthopedic complications. Spinal deformities, mainly kyphosis and scoliosis, may increase with aging. Chronic lack of sensation and muscle imbalances can lead to Charcot joints. Overuse syndromes are very common in people using wheelchairs and among community ambulators as well. Wheelchair users have a tendency to develop overuse symptoms in the shoulders, wrists, and hands, whereas community ambulators seem to develop knee and hip pain. Carpal tunnel and rotator cuff disease—well documented in the spinal cord literature—should have a similar incidence and etiology in this population. Knee pain originates from muscle imbalances in the lower extremities, along with knee flexion contractures. Weak hip abductors and extensors cause gait abnormalities, including genu valgum and a Trendelenburg pattern. Osteoporosis occurs in childhood and persists into adulthood. It is associated with...
an increased risk for fractures and is evaluated by using a dual-energy x-ray absorptiometer scan. Treatment, including calcium and vitamin D supplements and bisphosphonates, is available.6

Urologic issues are some of the most difficult for adults to manage. A very small percentage has normal function, whereas the majority requires some type of bladder management program. Despite these abnormalities, more than 80% of adults are able to develop social bladder continence.7 This is achieved by intermittent catheterization, use of indwelling catheters, urinary diversion surgeries, and artificial sphincters.8 Current data show that renal damage, leading to renal failure and its complications, remains a major contributor of morbidity and mortality in adults with spina bifida.9,10 An association also exists between the presence of a neurogenic bladder and the development of bladder cancer—an association that necessitates vigilant surveillance.11 The management of neurogenic bowel changes over time because gastric motility is altered with age. These modifications can be accomplished with medication changes, dietary modifications, and newer surgical interventions, such as cecostomy buttons and antegrade colonic enema procedures.12

The presence of insensitive skin and changes in fat and muscle distribution contribute to the high incidence of pressure ulcer formation. These ulcers are quite common in the lower extremities because bracing causes abnormal forces to be generated on bony prominences. The wheelchair-seated posture generates pressure in the ischial and sacral areas, placing them at high risk for skin breakdown. Proper cushion use and pressure mapping may alleviate some of the morbidity associated with pressure ulcers. The economic burden from this 1 entity alone is significant.13,14

Latex allergies are well documented in the literature and seem to have a multifactorial etiology. Adults may have a higher rate than children for reactions, including for anaphylaxis.15,16

Obesity has been documented in children with spina bifida and generally persists into adulthood. Nutritional studies indicate a decreased caloric expenditure when compared with caloric intake. Nutritional counseling should not only discuss the need for a balanced diet but also include information on fitness and exercise. There is a growing awareness that stress on young women to lose weight may cause a small cohort of adult women to develop eating disorders.14

Issues concerning sexuality are frequently overlooked in this population. It is a misapprehension to think that sexuality is not a critical part of a disabled person’s lifestyle. Data show that the majority of men and women with spina bifida have a desire for intimate relationships, including sexual contact.17 For men in this population, 72% report a capacity for achieving erections, with 67% experiencing ejaculation; however, satisfaction with penile rigidity is present in only about a third of the men who achieve erections.18,19 Studies with sildenafil (Viagra) show that it improves erectile function in 80% of men with spina bifida.20 Only 14% were able to father children; this success appears to be related to neurologic level of lesion.18

Women with spina bifida generally have normal menstruation, and during intercourse 88% have adequate vaginal secretions.19 The literature suggests that women with spina bifida are able to conceive and have children with a low complication rate.14

Women should have routine screening tests including Papnicolaou smears and mammograms; however, finding easily accessible clinics and equipment is often challenging. Pregnancy and risk for sexually transmitted diseases must be discussed openly.

CEREBRAL PALSY

Although cerebral palsy (CP) is described as a nonprogressive disorder of movement and posture that is attributable to a 1-time injury to the immature brain of a fetus or infant, cumulative evidence over the past 2 decades supports patients’ reports of functional declines associated with aging.21-24 The emergence of secondary conditions appears to be the consequence of life-long abnormal movements, altered postures, immobility, chronic medication consumption, and poor nutrition.21-24 Disorders associated with CP, such as seizures, mental retardation, visual and auditory impairments, learning disabilities, and communication disorders can also potentially contribute to the development of secondary conditions.25 These conditions may be unanticipated by the patient, family, or caregivers and may lead to diminished functional independence, decreased quality of life (QOL), changes in social and family roles, and altered vocational viability.21,22-24

The general health of persons aging with CP is reported to be fairly good.25-27 However, an increased incidence of bowel and bladder dysfunction22,26 with urinary tract infections22 (UTIs), oral motor and dental disorders,22,27 fractures,22,23 fatigue,21 gatroesophageal reflux,27 problematic spas ticity,21 pain,21,23,26,28 progressive musculoskeletal deformity and dysfunction,21,22,26 deterioration of functional ambulation,22,24 and progressive cervical spine degeneration complicated by radiculopathy or myelopathy29 have been noted as people age.

Poor perineal hygiene, use of external catheters, and the presence of an indwelling Foley catheter correlate with an elevated risk of UTI in this population.22 Bladder incontinence can interfere with vocational pursuits.21 If related to bladder overflow or to environmental barriers, provision of a toileting schedule may successfully maintain continence.

Oral motor problems appear particularly common in persons with dyskinesias and/or spastic CP. Complaints include increased difficulty with chewing, eating, and swallowing, with resultant increased risk of aspiration, malnutrition, and dental decay.21,22,26-28

Osteoporosis may occur at an early age21 and has been attributed to decreased weight-bearing activity, diminished muscular forces on bone, poor nutrition, use of certain medications, and endocrine disorders.22,23,28 Significantly decreased bone mass in children and adults with severe spastic tetraplegia has been documented and is associated with an increased risk of nontraumatic fractures.30 An intervention trial with bisphosphonates has been advocated.90

Pain is commonly reported in adults with CP21-23,28 and is often related to underlying musculoskeletal dysfunction, degenerative arthritis, and overuse syndromes.22,23 Pain onset may be at a young age22,23,28; however, it is unclear whether pain is likely to increase with advancing age.22 Schwartz et al26 reported that 67% of 93 community-dwelling adults with CP complained of pain of more than 3 months in duration in 1 or more area. Of these patients, 58 had pain daily and 53% reported the pain intensity as moderate to severe. In a study26 of 63 community-dwelling women with CP, 84% reported pain. The most common sites of musculoskeletal pain include the hip, knee, ankle, lumbar, and cervical spine.22,23,28 In another sample of 76 adults with CP, Murphy et al22 reported the presence of cervical pain in 50% of the total group (75% of persons with dyskinesia), back pain in 43% of nonambulators, and pain in weight-bearing joints in 23% of the group (44% of nonambulators). Despite the apparent helpfulness of a variety of pain interventions, most people experiencing pain may not seek help from health care providers with their discomfort.31 Additionally, caregivers for persons with severe cognitive
and/or communication impairments may underappreciate or misinterpret nonverbal pain behavior.23 Particular vigilance for behavior changes is required.

Contractures,22 scoliosis,22,23 subluxation or dislocation of the hip,23 knee and foot deformities,29 and pelvic obliquity32 are all common in CP. These musculoskeletal deformities may be progressive, may be associated with pain, and may alter functional status.21–23 Lower-extremity contractures are particularly prevalent in nonambulators (up to 91%) and can be problematic for positioning, transfers, hygiene, and skin protection.22 When present, scoliosis may show significant progression over time and the progression rate appears to be related to the degree of curvature at the time of skeletal maturity.23,32 Curves greater than 50° at the time of skeletal maturity may show more rapid deterioration (up to 1.4°/y by 1 estimate).32,33 Progression of scoliosis can lead to difficulty with seating and positioning, with further effects on comfort, mobility, pelvic positioning, skin integrity, and independence.23,33 Neuromuscular scoliosis may be accompanied by significant cardiopulmonary compromise.34–36 Progressive spinal deformity can be associated with decreased vital capacity, diminished lung volumes, increased energy cost of breathing, hypoxemia, hypercapnia, pulmonary hypertension, and right-ventricular heart failure.35,37 Caretakers and clinicians should monitor for evidence of respiratory compromise. Nocturnal pulmonary dysfunction may manifest as behavioral signs of sleep disturbance, morning headache, and daytime somnolence.34 Serial measurements of vital capacity and respiratory muscle strength have been advocated.34,35 Treatment of scoliosis-related pulmonary disorders might include respiratory exercises, noninvasive methods of ventilatory assistance, or surgical correction of spinal deformity.35,38–42 Outcomes of surgical intervention are variable. Negative surgical outcome predictors appear to include preoperative hypertkyphosis, vital capacity less than 30% of predicted, history of recurrent pulmonary infections, and necessity of oxygen supplementation prior to surgery.35,43

Early onset of degenerative arthritis, particularly in weight-bearing joints, may be associated with “…excessive physical stress, strain, biomechanical abnormalities, and compensatory functional overuse.”23 Murphy et al.22 reported an association between the presence of pain in weight-bearing joints and a cessation of ambulation around age 45. However, 75% of the group who had stopped walking had done so by age 25 because of fatigue, inefficiency of gait, or improved access to functional activities via wheelchair mobility.22 Other investigators27,44 have reported loss or deterioration of functional ambulation in adults with CP, although it less clear how pain and/or musculoskeletal decompensation contributed. Intervention studies are needed to help identify strategies for management of painful and functionally limiting degenerative orthopedic changes in ambulatory adults with CP.

Persons with atethoid CP are at an increased risk for developing cervical spondylotic myelopathy at a relatively young age (as early as the fourth decade of life), which is thought to result from excessive abnormal involuntary head movement.29 Immediate and short-term outcomes of surgical decompression and anterior or posterior fusion are favorable.29,45,46 Recent reviews of longer-term (>5y) surgical outcomes, however, suggest a relatively high risk for later (>5y postsurgery) development of cervical kyphosis, instability at adjacent segments, or neurologic deterioration, necessitating vigilant follow-up.29,47,48

Decreased use of preventative care has been reported in adults with CP.21,22 In a study of 101 adults with CP, Murphy22 reported that over 90% of the study group lacked preventative general health evaluations, including surveillance for cardiovascular risk factors. In this same study, 90% of women did not receive regular gynecologic screening examinations and fewer than 10% of men received regular prostatic evaluations. Possible explanation for the relative paucity of preventive care include limited education of patients and caregivers regarding the importance of preventative measures, restricted availability of informed and responsive health care providers, problems with access to health providers, funding constraints, perceived or real environmental barriers to adequate examination, and physical barriers (including spasticity and contractures) to proper positioning during routine physical examinations.21,22

**POSTPOLIOMYELITIS SYNDROME**

Postpoliomyelitis syndrome (PPS) is a constellation of signs and symptoms that appear in people 30 to 40 years after the original onset of their poliomyelitis. The syndrome includes weakness, fatigue, joint pain, and aching or cramping muscle pain, and, frequently, a related functional decline. People with PPS may experience a wide range of difficulties.

Presence and severity of symptoms appear to correlate with the severity of the original poliomyelitis. Although the etiology of PPS is not known, the following theories have been postulated for it: (1) weakened muscles: controlled by 10% to 20% of the original number of motor units (because of initial poliomyelitis) affected muscles have a further strength decline with aging; (2) viral load: there exists a continued viral presence and ongoing effects, as evidenced by identified poliovirus genome sequences and inflammatory cytokine production in the cerebrospinal fluid of only those polio survivors with PPS; and (3) concomitant conditions: normal aging processes, including arthritis, spinal stenosis, disuse deconditioning, obesity, and accrete to produce the postpolio constellation.

Rehabilitation management follows the general principles of maximizing functional abilities through energy conservation. Interventions include standard ones such as walkers, crutches, and wheelchairs and an increasingly technologically oriented arsenal of adaptive equipment. Reduction of muscle overuse through the application of selective light-weight splinting, body-weight reduction, and counseling also may be pursued. Several studies51 now suggest that moderate conditioning can result in significant strengthening in an older age group with PPS, without adverse consequences.

Pulmonary impairment is also quite common in PPS. It occurs primarily because of respiratory muscle weakness but also is attributable to the high incidence of scoliosis, obesity, sleep-disordered breathing, and bulbar muscle dysfunction that postpolio patients experience. Because pulmonary impairment may lead to acute respiratory failure, with consequent hospitalization, tracheal intubation, and bronchoscopies, it is important that clinicians monitor the respiratory status of patients with PPS. A significant decline in ventilatory function may warrant pulmonary therapy or assistive ventilatory devices. Timely introduction of mouthpiece or nasal intermittent positive pressure ventilation, manually or mechanically assisted coughing, and home blood gas monitoring decreases the need for subsequent hospitalizations for acute respiratory failure and tracheal intubation.52

**MULTIPLE SCLEROSIS**

Multiple sclerosis (MS) is a chronic inflammatory, demyelinating disease of the central nervous system, characterized by multiple lesions in the white matter of the brain and spinal cord. Although MS primarily manifests in young adults between the ages of 20 and 40 years, it may occur in children and elderly persons as well.53 Resulting functional loss includes weakness, fatigue, spasticity, and impairments of speech, swal-
lowing, vision, cerebellar functions, bowel and bladder function, and cognition.\textsuperscript{54}

Both inpatient and outpatient rehabilitation programs are of value in both progressive MS and in the more common relapsing, remitting forms.\textsuperscript{54-57} Research\textsuperscript{58} has shown the effectiveness of rehabilitation in reducing disability and improving QOL in patients with MS. Other studies\textsuperscript{57} have shown that physical fitness of MS patients can be improved by exercise and can also help them achieve a lower level of perceived fatigability.

Assessment of the patient with MS should include evaluation of his/her physical, psychologic, vocational, and social functioning. A wide variety of physical measures, including stretching of tight and spastic muscles, resistive exercises to increase strength, and aerobic conditioning to increase cardiovascular fitness, are effective in MS patients.\textsuperscript{53,58} Pharmacologic interventions for spasticity, including oral medications, intramuscular botulinum toxin, and intrathecal baclofen, may be helpful.\textsuperscript{59} If significant impairment is present, orthotic assistance, such as a plastic ankle-foot orthosis to substitute for weak ankle dorsiflexion, or gait aids, such as a cane or walker to assist in stability and function, can be used. A myriad of assistive devices and technology may be applied over time, as a changing spectrum of impairments occurs. Devices as simple as weighted utensils to assist persons with upper-body mobility problems and personal digital assistants for those with cognitive impairments limiting memory may be useful.\textsuperscript{60} Assistive technology is a field still in its infancy, and in no disease is it put to more of a test than in MS, in which impairments fluctuate and disabilities loom as ever-changing challenges.

A multidisciplinary approach to the care of the MS patient, with a team that includes patient, physiatrist, neurologist, physiotherapist, occupational therapist, speech therapist, nurse, psychologist, urologist, and family members, has been shown to be most effective in approaching the disability of MS patients, although not changing their underlying impairments.\textsuperscript{58} This effectiveness is present whether used alone or in conjunction with intravenous steroid treatment.\textsuperscript{61} Indeed, as the disease progresses, which is, unfortunately, frequently the case, a multidisciplinary approach aimed at the impairment, functional activities, and the patient’s environment becomes more urgently needed.\textsuperscript{59} Individual modifications, including home setup, ramps, lifts, level access showers, bath aids, and widened doorways, can help improve mobility and maintain functional independence. Similarly, societal changes, such as accessible buildings and transportation, help facilitate this as well. Unfortunately, many people with moderate to severe disability receive very little in the way of community services, and these services are often felt to be inadequate and poorly coordinated.\textsuperscript{62}

\textbf{AMYOTROPHIC LATERAL SCLEROSIS}

Amyotrophic lateral sclerosis (ALS) is the most common degenerative disorder of the motoneuron system. Most cases have onset after the age of 40 years, and the mean age of onset is 58 years, with a predominance of men afflicted.\textsuperscript{63} ALS is characterized clinically by muscle atrophy, weakness, fasciculations and cramps, hyperactive reflexes, and increased muscle tone. Bulbar onset, with dysphagia or dysphonia, occurs in 20\% to 30\% of cases. The disease is generally not accompanied by dementia, pain, decreased sensation, or incontinence. Unlike MS, typically there are no relapses or remissions, and the disease is slowly progressive.\textsuperscript{64} Average disease duration is 3 to 4 years, but 10\% of patients survive more than 10 years. Because there is no cure for ALS at this time, symptomatic care, again using the multidisciplinary team, is the mainstay of treatment.

\textbf{Symptoms}

Weakness is the major symptom of ALS. Many of the principles outlined previously, for the MS patient, are used to treat the ALS patient. A regular, moderate physical exercise program, at least in the early stages of the disease, helps reduce disability in the ALS patient.\textsuperscript{65} Dysarthria is a common consequence of ALS. Speech therapy specialists may be helpful initially and subsequently may help in furnishing communication devices.

Dysphagia, caused by decreased motility of the tongue, pharynx, and esophagus, is also very common and can lead to choking and aspiration. Initially, dietary alterations emphasizing easy to chew, calorie rich foods, as well as instruction in swallowing techniques, may be helpful. If, however, the patient continues to lose weight and eating becomes intolerable, placement of a percutaneous endoscopic gastrostomy tube is advised before the patient reaches a state of respiratory distress and while vital capacity remains over 50\% of predicted value.\textsuperscript{66}

Dyspnea, or respiratory insufficiency, occurring as the degenerative process and involving the inspiratory, expiratory, and bulbar muscles is the leading cause of death in ALS.\textsuperscript{67} Nocturnal hypoventilation results in symptoms that include fatigue, sleep disturbances, headaches, decreased concentration, and resulting mood disturbances. Nocturnal noninvasive positive-pressure ventilation (NPPV) relieves the symptoms of hypoventilation and improves QOL for ALS patients.\textsuperscript{68} Unfortunately, NPPV is not universally offered to patients, for various reasons, including concerns about prolongation of life at the expense of QOL and reimbursement.\textsuperscript{68,69}

\textbf{MUSCULAR DYSTROPHIES}

The muscular dystrophies are a group of progressive disorders of variable severity. The most well known is Duchenne’s muscular dystrophy (DMD), which results from a complete deficiency of dystrophin. Characterized by progressive wasting and weakness of proximal limb-girdle muscles, DMD is usually evident by the age of 5 years, is followed by inability to walk by the age of 8 to 12 years, and results in death by the early twenties from respiratory failure.\textsuperscript{70} In Becker’s muscular dystrophy (BMD), some dystrophin is present; therefore, the age of onset and the rate of progression are more variable than in DMD. Although most cases still have onset in childhood, occasionally the disease will not manifest until early adulthood and typically patients do not require full-time use of a wheelchair until their twenties. Other forms of muscular dystrophy include the dominant and recessive limb-girdle muscular dystrophies and restricted myopathies, including the fascioscapulohumeral types. All are incurable and generally progressive, and rehabilitation goals are to limit the disability and to maintain function for as long as possible.

BMD is characterized by a lordotic posture and waddling gait, much like DMD. Muscle contractions and spinal deformities, however, are not common, as they are in DMD. Muscle training exercise programs, especially if initiated early in the disease, may be beneficial for patients with BMD.\textsuperscript{71} Devices to allow continued ambulation, and then wheelchair mobility, as well as continued habilitation are extremely important to the patient who is aging with muscular dystrophy.\textsuperscript{72} Indeed, persons with less severe forms of muscular dystrophy seek ways that they can continue to function efficiently and not just simply function, as they remain in the work force, and face the time constraints of modern existence.
CONCLUSIONS

People with spina bifida, CP, and neuromuscular diseases are commonly treated by pediatric physiatrists, but with improvements in medical care, this population is aging and its needs are increasingly being met by practitioners of adult rehabilitation medicine. Although poliomyelitis is now rarely acquired in the United States or other industrial countries, its delayed effects, as seen with PPS, are increasingly managed by physiatrists. An awareness of the common medical, physical, psychologic, and functional needs associated with normal aging of people with early life disability is important if rehabilitation professionals are to lead their patients to the best possible outcomes.

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