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Abstract

The World Health Organization definition of “health” has become all but a cliché. It is often used to highlight the need to address issues beyond the traditional medical model, often associated with deficits, disease, or disability. Most often the definition is all that is provided. The specifics of how a medical practitioner or a medical organization moves beyond medicine toward a focus on health are usually sketchy, at best. While health is certainly the outcome that medicine addresses, the emphasis is most often the reduction or elimination of deficits or deficiencies. This chapter will address the varied secondary conditions to which individuals with developmental disabilities are most vulnerable, and will identify the attitudes, approaches, and practices (the barriers) that undermine the health of individuals with developmental disabilities. We will balance this by identifying concepts, approaches, and practices that can encourage health. Health promotion is the process of enabling people to increase control over, and to improve, their health. This goal of helping individuals increase control over their health, with the requisite understanding and behaviors to improve their health is a critical one for individuals with developmental disabilities. Myers concluded that if individuals with developmental disabilities are to move toward health, there is a need for “greater professional humility to appreciate that people with both physical and intellectual impairments are able to experience and articulate their own satisfaction, pleasure, and joy”. Medicine is a crucial component of health, but health is more than medicine.

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Introduction

WHO definition of health: “Health is a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity” [1]. The World Health Organization definition of “health” has become all but a cliché. It is often used to highlight the need to address issues beyond the traditional medical model, often associated with deficits, disease, or disability. Most often the definition is all that is provided. The specifics of how a medical practitioner or a medical organization moves beyond medicine toward a co-emphasis on health are usually sketchy, at best. While health is certainly the outcome that medicine addresses, the emphasis is most often the reduction or elimination of deficits or deficiencies. “Health care” is the term used to connote what medical professionals do toward this end and covers diagnosis and intervention of medical conditions, with the best coverage including activities associated with “prevention” of medical conditions, but rarely the promotion of “physical, mental, and social well-being”; stated positively, health.

Medical practice has been improved specifically by including the prevention of secondary conditions associated with primary medical conditions as a component of practice. Even this addition, however, does not insure an emphasis on promoting health. If we are to address medicine, but allow it to be included in the broader notion of health, what concepts, approaches, and behaviors ought we to pursue? And can we assume that these components apply equally to individuals with developmental disabilities? For this chapter, we assume that (a) health is more important for this group than for the general population, and (b) the barriers to health are greater for this group.

We also assume that individuals with developmental disabilities will include individuals both with and without intellectual impairments. Boyle et al. [2] reported a developmental disabilities prevalence of 15 % among children aged 3–17 years, with autism, and attention deficit hyperac-

tivity disorder increasing, while hearing loss decreased over a 12 year period. The Administration on Intellectual and Developmental Disabilities (AIDD) estimated that between 7 and 8 million Americans across the life span (3 % of the population) experience an intellectual disability [3]. Given increased vulnerability of this population, health promotion and prevention of secondary conditions are particularly crucial for the individual, their family, and the health system. This chapter will address several pivotal issues:

- We will define and outline secondary conditions to which this population is most vulnerable.
- We will identify the attitudes, approaches, and practices (the barriers) by which “health professionals” unwittingly undermine health of individuals with developmental disabilities.
- We will identify which concepts, approaches, and practices can encourage health beyond, but including, medicine.
- We will identify environmental factors within and outside medical offices and facilities that can undermine or facilitate the health of this population.

The term “secondary conditions” has evolved since the early 1990s [4–6]. Basically, it can be defined as “any preventable condition to which a person or family is more susceptible by virtue of experiencing a primary diagnosis associated with disability” [4]. The term refers to outcomes for which a person is at greater risk due to the presence of a primary disabling condition. That is, the primary diagnosis is a risk factor for the appearance of a secondary condition. Secondary refers to timing—when the conditions occur—and does not mean that the conditions are less serious. The term “conditions” is used to suggest that the varied outcomes are not exclusively in the medical or physical domain of function but can also include emotional, social, and environmental dimensions.

Several examples of secondary conditions for individuals with developmental disabilities may clarify the concept. Urinary tract infections and

pressure sores are secondary conditions often seen in individuals with spina bifida or spinal cord injury. That is, they are conditions for which the person is at greater risk because of the primary disabling condition. Social isolation and depression are conditions experienced by individuals with various developmental disabilities. Without wishing to stretch the concept too far, one can entertain that reduced access to health care or injuries to family caregivers can also be considered secondary conditions for clinical purposes. In clinical practice, secondary conditions often can be more severe and disruptive than the stabilized primary diagnosis and have implicitly adverse consequences. Much, if not most, clinical time is spent addressing the secondary condition(s), beyond the primary medical condition creating body dysfunction. One of the clinician's tasks is to work with individuals with developmental disabilities and their families to prevent or reduce the impact of such secondary conditions on the person and family.

In daily appointments at clinics, little distinction might be made between primary symptoms and secondary conditions for the person with disabilities or his or her family. Diagnosis is diagnosis; treatment is treatment; intervention is intervention. The most relevant distinguishable clinical element of secondary conditions is that they are preventable. That is, although there is a greater risk for the occurrence of them, secondary conditions are not part of the primary manifestation(s) of a diagnosis. Depending on the clinician, evaluation and diagnosis of secondary conditions shows wide variations in the comprehensiveness. In addition, the less medical the secondary condition, the less attention may be given to assessment and interventions. This lack of comprehensive attention may be based on the assumption that the individual with developmental disabilities, with or without intellectual impairments, cannot adequately communicate concerns or problems—or that the information will not be accurate or relevant or helpful. Secondary conditions can take different forms. Crocker [7] developed a typology reflecting the range and complexity often found:

- **Complication:** An untoward occurrence, accidental but resulting from the primary condition (e.g., pressure sore in spina bifida)
- **Contingency:** An event involving another body system but ultimately deriving from the conditions of the primary condition (e.g., conductive hearing impairment associated with Down syndrome)
- **Unexpected progression:** A troubling extension of the potential continuing natural history of the primary condition (e.g., loss of ambulation in cerebral palsy)
- **Comorbidity:** Another parallel condition, deriving from the same background as that producing the first diagnosis (e.g., hydrocephalus and spina bifida).
- **Other health concerns:** Ill health from other origin but perhaps masked or confounded in some fashion by the primary condition (e.g., obesity in Down syndrome)
- **Effects of aging:** Liabilities or dysfunctions due to advancing years, often accelerated by a primary condition (e.g., overuse syndrome related to mobility problems)

To complement this typology, we could add social and emotional issues—those that emerge primarily because of the developmental disability experienced by the individual. As indicated earlier, secondary conditions should not be viewed only as medical conditions, but as any condition to which the individual is more vulnerable by virtue of their primary condition. This would include, for example, social isolation, identity issues, and emotional adjustment concerns.

In the field of developmental disabilities, the world of secondary conditions is a large and often complicated one. Varied therapeutic, educational, technological, environmental, and social interventions must be considered during clinical encounters with the individual and his or her family. As a matter of course, practitioners working with people with developmental disabilities usually are already aware of the breadth and complexity of secondary conditions and alternative interventions that are part and parcel of these interactions. Nonetheless, the skill and sensitivity of the

provider will be challenged beyond the medical issues to the health issues of the person and his or her family. Consider the following vignette:

Case 1: Elizabeth is an 18 year-old high school senior who is preparing to go off to college in the fall. She has cerebral palsy and a complex past medical history, including multiple surgeries, including soft tissue releases, two femoral osteotomies, and a spinal fusion. She has long-standing history of severe, debilitating migraines. Elizabeth is bright, at the top of her graduating class, and has already received a scholarship from a nearby liberal arts college. She has travelled internationally and is fluent in French. She is always well groomed and wears fashionable clothes. Elizabeth recognizes that many people see her in her wheelchair and draw immediate conclusions about her intellect; and that others encounter her intellect and cannot imagine the disabilities she has. Just as Elizabeth is preparing for the independence she anticipates in her college experience, her hip surgery begins to fail and she experiences unrelenting pain. On chronic pain medications, she struggles with sedation, gains weight, and develops severe bowel impactions. As Elizabeth makes the rounds of physicians in her senior year, her various specialists provide care and attention to her specific concerns, but do not address the secondary conditions now developing. None of her physicians ask about her weight gain or her bowel functions. None of her physicians encourage exercise or discuss nutrition. (Is depression identified? If so, is it just addressed with medication?)

Elizabeth's story may provide some insight into the care of young adults with disabilities. The great disappointment for Elizabeth and her family was the instability of her health at age 18 years. Most of what they had read suggested that cerebral palsy did not lead to additional health concerns. This very bright, engaging young woman's academic performance masked her problems so much that many teachers and peers could not imagine that someone so bright could

have so many disabilities. Moreover, the increasing health problems that demanded attention drew her physicians away from efforts to integrate health care, that is, to address depression, weight, conditioning, and bowel functioning, as well as to address her hip problems and her headaches.

Areas of Disparity

"Health disparity", usually suggests a negative difference in the health status of people with a certain characteristic when compared to the general population. For people with developmental disabilities, health disparities can create the conditions for developing secondary conditions. That is, if people with developmental disabilities are less able to access health care, their vulnerability to myriad secondary conditions is increased [8]. Haverkamp, Scandlin and Roth [9] reported results from the North Carolina Behavioral Risk Factor Surveillance System comparing adults with developmental disabilities with non-disabled adults. The data indicate numerous areas of disparity, beginning with the sedentary lifestyle of those with developmental disabilities; specifically, individuals in the developmental disability group were significantly more likely to have had no exercise in the previous month than those without disabilities. These individuals also were much more likely (seven times) to indicate poorer emotional support than adults without disabilities. In addition, this population had significantly poorer utilization of oral health care and the women with developmental disabilities reported having breast and cervical cancer screening much less often.

The National Core Indicators is a state-based survey specifically focusing on individuals receiving developmental disabilities services. The 2010–2011 NCI Annual Summary Report [10] indicated that while 75 % of adults in the survey had received a flu vaccination in the past year, only 39 % had ever had a pneumonia vaccine. Only 19 % of the respondents over 50 years had a colorectal screening in the past year. The report concluded that for almost all services, individuals with developmental disabilities living with par-

Table 2.1 Estimated prevalence of secondary conditions in adults with developmental disabilities

| Secondary condition/problem | Estimated prevalence/1000 |
|--|---------------------------|
| Physical fitness and conditioning | 590 |
| Communication | 573 |
| Mobility | 509 |
| Persistence or low frustration tolerance | 500 |
| Weight | 479 |
| Personal hygiene or appearance | 470 |
| Dental and oral hygiene | 451 |
| Fatigue | 422 |
| Depression | 369 |
| Sleep disturbance | 316 |
| Bowel dysfunction | 288 |
| Respiratory | 178 |
| Cardiovascular/circulatory | 156 |
| Osteoporosis | 112 |

From Traci et al. [12]; adapted by permission

Table 2.2 Adolescent spina bifida secondary conditions rank order

| | |
|--------------------------|--------------------------|
| 1. Bladder incontinence | 7. Recreational problems |
| 2. Learning/memory | 8. Self-esteem problems |
| 3. Physical fitness | 9. Fatigue |
| 4. Mobility problems | 10. Headaches |
| 5. Initiation/motivation | 11. Social isolation |
| 6. Bowel incontinence | |

Adapted from Lollar [13]

ents or relatives and those living independently were less likely to have received appropriate services than those individuals living in community-based housing and in institutional settings.

Emergency department (ED) use by working adults with disabilities provides a broader view of related to access and service needs. Rasch, Guley, and Chan [11] pooled the Medical Expenditure Panel Survey Data from 2006 to 2008 and found that while 17 % of the US working age adults report some type of limitation, this group accounts for 40 % of ED visits. They concluded that even given that people with disabilities may experience more health prob-

lems, ED use could be reduced with better access to regular medical care, and attention to the complexity of individual’s health profiles.

Traci, Seekins, Szalda-Petree, and Ravesloot [12] completed a study in Montana of the prevalence of secondary conditions among a group of 119 adults across a broad spectrum of living arrangements, from independent residences to residential care facilities. Communication difficulties were reported most frequently by the direct care providers who completed the survey, followed (in descending order of importance) by problems with physical fitness and conditioning, persistence or low frustration tolerance for task completion, weight, personal hygiene, dental and oral hygiene, fatigue, depression, mobility, and sleep disturbance. Traci et al. [12] concluded that these limitations all included significant behavioral or life style components; more medically oriented conditions, such as gastrointestinal dysfunction, bowel problems, or respiratory difficulties, were reported substantially less often. Looking closely at the list, one sees the correlation among several of the conditions—conditioning is related to weight, which is related to appearance, which is related to mobility, which is related to fatigue, which ultimately is related to sleep and depression. This relationship is, of course, just one of a number of cyclical groups that can be generated among the secondary conditions listed in Table 2.1. Although some of these conditions (e.g., depression, sleep disturbance) might have diagnostic codes, several do not (e.g., fitness, low frustration tolerance, personal hygiene). Thus, a clinician’s inquiring about some of these conditions would not necessarily be a part of routine patient-clinician encounters.

Lollar [13] amended the Secondary Conditions Surveillance Instrument [14] to identify secondary conditions among a sample of adolescents with spina bifida. Table 2.2 provides the rank order of the most problematic secondary conditions for this sample. Incontinence of bladder (ranked first) and bowel (sixth) were the two medically associated secondary conditions, although the definitions for these two conditions included in the instrument clearly related to the preventable negative social aspects of in conti-

nence rather than just the body dysfunction. In addition to limitations in learning (second) and mobility (fourth), secondary problems of everyday living were prominent. Physical fitness (third), motivation (fifth), self-esteem (eighth), and fatigue (ninth) overlapped with the secondary conditions of the adults in the Montana sample. This exercise showed, as might be expected, that young people with spina bifida contend with the social aspects of bowel and bladder incontinence as well as with the general problems of everyday life. Also, cross-cutting issues related to fitness, motivation, mental health, and fatigue emerge.

Havercamp [15] completed a population-based survey in North Carolina of health needs of adults with developmental disabilities. Information was collected from the adults and their case managers. Physical fitness and obesity were the major problems that emerged. A high rate of mental health problems was found, and more than half of those in the representative state sample were being prescribed medication for mental health problems. It is possible that this may represent over-reliance of medication, both for treatment of mental health problems, and for misattribution of difficulties to mental health problems. Finally, the survey indicated that access to health care services was often difficult, with particular problems in oral health services and in reproductive services for women. The study concluded that lack of physical activity was a risk factor for chronic conditions, such as cardiac disease, and that there was an alarmingly elevated rate of emotional problems associated with inadequate support and high stress. Moreover, adults with developmental disabilities have high rates of tobacco and alcohol use, thus creating additional risk factors for stroke, lung cancer, and respiratory disease.

Data clearly indicates that secondary conditions, including problems with access to care, are a part of the life experience of individuals with developmental disabilities. Together, the aforementioned studies indicate that physical fitness, obesity, and emotional issues are frequent secondary conditions among these groups. Associated with these conditions are motivation,

persistence or low frustration tolerance, poor communication, difficulty with personal hygiene, fatigue, mobility limitations, and sleep disturbance. Finally, obtaining oral health services and reproductive services is often difficult; however, services for emotional issues and associated medications seem to be rather well identified and readily available, contrary to anecdotal evidence.

In summary, the data is consistent with the Surgeon General's report [16]. The report included a list of under-recognized medical problems—including constipation and impaction, visual and auditory problems, recurrent ear infections, periodontal disease and infected teeth, osteoporosis, and neuropathies—to which individuals with intellectual disabilities are more vulnerable. This list, however, does not include the broader range of secondary conditions (emotional, familial, social, and environmental) encountered in clinical settings. The report, "Closing the gap: A national blueprint to improve the health of persons with mental retardation", concluded that individuals with cognitive impairments experience poorer health and have more problems "finding, getting to, and paying for appropriate health care."

Barriers to Diagnosis/Assessment/and Health Promotion

To highlight the potential secondary conditions or associated health difficulties, consider the following cases:

Case 2: Diego is a 45 year-old man with intellectual disability, who lives in a group home. He is mildly obese and mildly hypertensive but has no other known health problems. He presents to his primary care physician (PCP) with lower abdominal pain and mild fever of several days duration. His provider collects a urine sample. On dip urinalysis, a large number of white blood cells and red blood cells are noted. Diego is started on empiric antibiotics, pending results of urine culture. Urine culture grows several organisms, consistent with it

being an inadequate specimen (not a “clean catch”), and Diego’s care providers are instructed to have Diego complete the course of antibiotics. Diego’s abdominal pain does not resolve; in fact, it worsens, and his fever persists, over the next few days. Another urinalysis is obtained, again showing white blood cells, consistent with infection, and another course of antibiotics is prescribed. Two days later, Diego is found lying in bed, incoherent. His fever is 104°, and he is taken to the ER. His blood pressure, which usually runs high, is now running low, and continuing to decline, even on IV fluids. Blood counts show elevated white cell count, with relatively high neutrophils, consistent with sepsis. With blood cultures pending, Diego is diagnosed with septic shock, started on IV antibiotics. Diego is admitted to the intensive care unit, a central line placed, and pressors administered. With blood pressure continuing to drop, and history of abdominal pain with no positive urine cultures, Diego is taken to the OR for exploratory laparotomy. Diego’s abdomen drained large quantities of pus; he had ruptured diverticulitis, resulting in septic shock. After the operation, Diego spent several days in the ICU on pressors and IV antibiotics, then several weeks in the hospital. He was discharged home after several weeks, and continued to require in-home wound care for weeks after that.

In retrospect, it became clear that the white blood cells seen on urinalysis were not due to a urinary tract infection, but due to inflamed bowel resting on top of the bladder. Urinary tract infection, refractory to antibiotics in a 45 year-old man without significant past medical history, should have prompted curiosity and a more aggressive work-up at the time of first presentation to the PCP. Had Diego not had developmental disability, urinary tract infection in a healthy 45 year-old man surely would have prompted a more thorough work-up right from the start. But, because Diego has developmental disability, the apparent UTI did not stimulate the PCP’s curiosity enough to prompt more of a work-up, even when the urine culture and Diego’s response to a course of

antibiotics suggested that there was more going on here. The tragic thing, of course, is that a great deal of suffering and expense could have been prevented by more aggressive work-up earlier on.

Case 3: Temisha is a 4 year-old girl with autism spectrum disorder, presenting for comprehensive developmental assessment to an interdisciplinary child development referral center. She has long-standing history of chronic severe constipation, with associated stool withholding, and sometimes, some blood with passage of stool. She has episodic diarrhea, alternating with constipation. Temisha is severely irritable, and spends much of each day in fetal position. She participates little in daily activities at preschool or home. She misses a great deal of developmental preschool programming as a result of this chronic discomfort. Abdominal X-ray shows rectal stool impaction. Constipation is treated with dis-impaction, with large doses of stool softener, given orally, over 3 days. Soft stools are maintained with low-doses of stool softener daily for 1 year. Also, behavioral program of 5 min toilet sits, 4 times daily, is prescribed, to achieve regular pattern of soft, formed stools, 1–3 times per day. Over the next few months, behavior is greatly improved. Strong developmental progress is seen over the next few months, in association with improved bowel pattern. Family believes that Temisha’s very dramatic progress is due to the “cleansing of her body of toxins” by the stool softeners.

This case illustrates how a common secondary condition can greatly exacerbate functional limitations in a person with a developmental disability. It also illustrates the importance of thorough review of symptoms in children with severe functional limitations, and the dramatic improvements in function that may be seen when secondary conditions are addressed. At the time of Temisha’s presentation, she had already been suffering for years with chronic constipation, and had been presenting with classic physical and behavioral symptoms of this secondary condition. It is sobering to consider what the course of

Temisha's development, and of her life, would have been, had the severe chronic constipation continued to have been ignored.

Case 4: Oanh is a 27 year-old woman with intellectual disability, autism, and multiple congenital anomalies, associated with chromosomal deletion. She has long-standing history of self-injurious behavior. Specifically, she often moans and punches herself in the chest, frequently, all throughout the day. Various primary care providers throughout her adolescence and early adulthood continue to attribute this behavior to self-injurious behavior associated with autism. Various anti-psychotic medications and mood stabilizers had been tried, with various side effects, and without much reduction in the self-injurious behavior. At age 27 years, Oanh presented for the first time to an interdisciplinary center for a comprehensive assessment. The focality of the self-injurious behavior was suggestive of physical discomfort in the chest. Acid damage to the back molars and halitosis suggested possible gastro-esophageal reflux (GERD). A work-up was initiated. A Ph probe demonstrated severe gastro-esophageal reflux disease. Subsequent imaging showed a vascular anomaly restricting the duodenum, causing the GERD symptoms. Surgical intervention was successful in treating the GERD and produced resolution of the self-injurious behavior.

As this case, a 27 year-old woman finally being diagnosed with a severe, congenital vascular anomaly illustrates, shows, there are many barriers to the appropriate evaluation of and intervention for secondary conditions in individuals with developmental disabilities. Barriers to evaluation and intervention may be internal or external to the person. Barriers are often a result of the interaction of the person with his or her environment. Environmental barriers can include physical barriers, social, or attitudinal barriers, and policy or system barriers. Physical barriers are the most

visible ones, particularly if the individual has mobility limitations. The social and attitudinal barriers can be harder to recognize. For instance, a health care provider without adequate experience with people with disabilities might be impatient with someone with a disability for being slower at answering questions, completing basic reading tasks, or looking "different". Health care providers themselves can unwittingly be a barrier to the evaluation or and intervention for secondary conditions.

Attitude of Clinicians

The greatest of the barriers to evaluation and treatment of secondary and other conditions might well be the unexamined perceptions of providers. As is illustrated in the cases above, it is all too common for providers to attribute behaviors which might point to treatable secondary conditions to being mere "behavioral problems" or attributable to the primary condition, to be handled by mental health providers, or not requiring intervention at all. Unfortunately, when this happens, mental health providers all too often over-rely on the use of medications, which fail to address the underlying secondary condition, and may actually exacerbate the condition. In the case of Oanh, the use of anti-psychotic medications to treat her "self-injurious behavior" may actually aggravate the situation, by causing obesity, which increases GERD symptoms, or by causing sedation and/or prolactinemia, which worsens irritability. When individuals have developmental disabilities and unusual or problematic behaviors, it is all too common to attribute all behaviors to being "part of the disability", rather than being thorough and curious and taking the time to investigate secondary conditions that may be the cause of these behavioral symptoms [17, 18]. Particularly, as in the Oanh's case, it is tragic when other physical signs and symptoms, such as dental acid damage and halitosis, are ignored for decades, allowing tertiary conditions, such as Barrett's esophagus, to develop. As

discussed elsewhere, due to communication problems, which interfere with obtaining information by interview, and due to mobility problems, which interfere with the physical examination, patients with developmental disabilities need more, rather than less time, to allow for thorough investigation of secondary conditions. Unfortunately, due to unusual behaviors, which some health care providers may find off-putting, data gathering and examination may actually be more cursory for individuals with developmental disabilities than for patients without disabilities.

Health Insurance

Barriers to thorough exploration of potential secondary conditions may also be attributed to the policy of insurers. Although some time limitations for examinations and treatment, set by insurers and by demands on health care providers' time, are acknowledged to be necessary, the needs of the person with disabilities do not change because such limitations exist. In this context, it should be emphasized that small intervention strategies can prevent major secondary medical conditions from occurring and can improve function. For example, in Oanh's case, the tremendous suffering and expense of treating esophageal cancer can be prevented by early diagnosis and treatment of GERD. Comprehensive assessment is necessary for conditions to be identified and interventions to be implemented. In addition to comprehensive review of systems and examination, individuals with developmental disabilities must be evaluated and subsequently treated with an integrated approach, focusing both on the person and the context in which he or she lives and functions. This process requires attention to factors beyond the individual, including family or other significant individuals in their lives, neighborhood supports or lack thereof, and community resources. Medical conditions are often created and/or exacerbated by factors external to the person.

Professional Interactions to Overcome Barriers

Young people with disabilities may have less-developed social, communication, and/or decision-making skills, so the interpersonal skills of providers become more important, and more noted when they are absent. The most basic support for people with disabilities is the respect shown by a health provider. One of the most basic ways to establish respect is to address or question a person directly. Health care providers often find it easier to ask questions of a caregiver, family member, or person transporting a younger child or young adult than to ask that person directly. Young people, even children, can usually answer straightforward questions about their lives.

When compared with the amount of time routinely needed to see a person without disabilities, about twice as much time is necessary when seeing a person with disabilities or a member of that person's family [19]. Sometimes, including others for verification or elaboration is important, but the tendency all too often is to overlook the individual with disabilities in the name of efficiency and credibility. Unfortunately, there are significant problems with this all too common approach. For one, the opportunity for the health care provider to build a healing relationship with the young person with disabilities is lost. For another, the opportunity to help a young person with disabilities to develop competency in communication with their health care provider is lost. This, investment of extra time in making an effort to involve a young person with disabilities in the medical interview improves communication and care, and, in the long run, may actually save time and money, as illustrated in several of the cases described above.

Transition from Pediatric to Adult Care

A related issue is the transition from pediatric to adult care. Pediatricians, both primary care and sub-specialists, often are torn between continu-

ing care for young people as they mature to adulthood and acknowledging the limits of their own training, experience, and professional comfort. Crucial in the transition from child to adult health care providers is the reconstruction of resources by the young person and his or her family. This process often requires looking for different medical and health providers, emotional supports, and hospitals because many pediatric medical centers will not admit anyone older than 21 years of age. This change can be particularly difficult for a young adult with disabilities whose pediatric team has not made transition plans and is not on the staff of an adult facility [20]. Again, a case study may help to elucidate this issue.

Case 5: Keith is a 21 year-old man with history of mild intellectual disability and chronic lung disease. His mother is alarmed to find that Keith is becoming progressively more and more fatigued. His academic functioning, in his last year of his transition program, and his hygiene, are beginning to suffer, as his energy flags. Suspecting that these symptoms may reflect an exacerbation in his lung disease, Keith's mother calls the Pulmonary clinic where Keith has been a patient for two decades, to make an appointment. To her great surprise and shock, she is told that Keith will need to transfer care to an adult Pulmonologist, now that he is an adult. Pediatric hospitals often do not provide appropriate preparation for transition to adult care.

In the meantime, while waiting for that appointment, Keith followed up with his developmental-behavioral pediatrician, whose clinic did not have such strict age limits. Keith's mother, accustomed to being the spokesperson for her quiet son, launches into a detailed history of Keith's pulmonary symptoms. The developmental-behavioral pediatrician gently interrupts Keith's mother and asked Keith how he has been doing lately. With some prompts and a lot of patience, the pediatrician eventually elicits from Keith a 2 year history of obsessive compulsive symptoms. These symptoms have been gradually escalating in severity, up until the present. Keith explains that he is up

every night for hours, contorting his body and freezing his body into bizarre positions at the side of his bed, unable to sleep. Even without pulmonary function testing, the cause of Keith's escalating fatigue quickly becomes clear.

Medical staff and family, along with the young person, must establish mutual respect, communication, and a relationship that will allow straightforward analyses of issues related to independence during adolescent development. These professional behaviors allow for thorough and complete assessment and evaluation related to secondary conditions of individuals with developmental disabilities. The foundation for promoting health is now established.

Principles of Health Promotion

The WHO defines "health promotion" as "the process of enabling people to increase control over, and to improve, their health. It moves beyond a focus on individual behavior towards a wide range of social and environmental interventions" [21]. The Ottawa Charter for Health Promotion [21] declared that the ability to reach a state of physical, mental, and social well-being, an individual or group "must be able to identify and to realize aspirations, to satisfy needs, and to change or cope with the environment". This goal of helping individuals increase control over their health, with the requisite understanding and actions to improve their health is a critical one for individuals with developmental disabilities. While individuals with developmental disabilities may have impairments in learning, cognition, emotion, or behavior that make these goals challenging, health/medical professionals have the opportunity to promote the health and well-being of these individuals. The final goal of all health care, medical care, and health promotion is that the individual be able to participate in society.

Health promotion services really begin with helping a person with developmental disabilities to become increasingly responsible for his or her own health, consistent with their capacity. Two terms are relevant in this regard. Selfdetermination is usually a long-term goal that may not be breached,

whereby the individual with developmental disabilities is able to make most choices for themselves, whether health, education, community-living, or as simple as choosing clothes to wear or food to eat. Self-advocacy, however, is a process by which individuals with developmental disabilities are encouraged to make their wishes, desires, and preferences to be through empowerment. Health care professionals may not keep this goal of self-determination utmost in their clinical thinking. Health promotion, when included, is viewed as providing health-related information—from the professional to the individual. Sound bites of accumulated wisdom are presumed to provide the knowledge and incentive for healthy behaviors to begin and continue. A clinician can encourage self-advocacy in any appointment or interaction by willingness to engage the individual on their level, even if it takes more time.

Although people with developmental disabilities are usually quite willing to listen to authority figures such as medical professionals, as are most of the general population, they may have difficulty comprehending the message. If comprehension is not a problem for the individual, the person still may experience memory problems or difficulty keeping a sequence of instructions in the order necessary for them to be of help. Video or written guidance may be used as an adjunct to any verbal interaction or direction. Frustration tolerance can be sufficiently low that, if barriers arise, the whole message can be lost. Yet, guidance toward healthy behaviors provides the greatest opportunity for health care providers to be oriented beyond just medicine and toward health.

Counseling is, by definition, two-way in nature—that is, an exchange between or among individuals. Counseling connotes mutuality, rather than more directed terms such as guidance or teaching. In view of the secondary conditions discussed earlier—poor conditioning, obesity, oral health problems, and so forth—counseling to promote health behaviors and prevent secondary conditions is extremely important, particularly in the areas of physical activity, healthy diet, and tobacco use. Unfortunately, the time required for appropriate counseling interaction on health

behaviors is extremely limited during routine clinical visits. It is all but prohibitive if an individual requires more attention, as is often the case with people with developmental disabilities. In addition, such health promotion services are not always covered by insurers, despite the fact that many insurers use prevention as a major thrust of marketing for their plans.

One facet of counseling that is often overlooked involves the discussion of exploitation. From childhood onward, people are vulnerable to being manipulated by others who are more cognitively adept and more emotionally hardened. Whether the outcome of the exploitation involves money, time, work, or even sex, individuals with developmental disabilities are of a more trusting nature, without guile, and therefore more vulnerable to manipulation. Health care providers can offer both the perceived authority and the sense of personal safety to allow someone with a developmental disability to disclose information about being taken unfair advantage of by others.

Another aspect of health promotion emphasizes the need for clinical preventive services. Clinical preventive services traditionally include screening tests, immunization, and counseling [22]. Because counseling can, and often does, overlap into health promotion services, it is discussed in that context in this section. A study by Jones and Kerr [23] indicated, however, that individuals with cognitive impairments did not receive annual health screenings. The Guide to Clinical Preventive Services [19] from the US Preventive Services Task Force should be applied to people with developmental disabilities. The recommendations cover 80 primary conditions for which all individuals are at risk. It concludes that, if a segment of the population does not receive the services detailed therein, they are at greater risk for the conditions identified—a straightforward conclusion to draw, but particularly poignant for the population of individuals with developmental disabilities. Of those preventive services provided by the Task Force, most relevant for individuals with developmental disabilities would include coronary disease, cancer, metabolic and nutritional disorders, vision and

hearing disorders, emotional problems, and substance abuse.

Screening for emotional problems in both children and adults with developmental disabilities is critical given the magnitude of this problem. The Healthy People 2020 chapter called “Disability and Secondary Conditions” provided two objectives aimed at reducing depression among children and adults with disabilities [24]. Data for that report from the National Health Interview Survey indicated that 17 % of children without disabilities are reported to be sad, unhappy, or depressed, whereas 31 % of children with disabilities report these emotional problems. Likewise, 28 % of adults with disabilities report that depression prevents them from being active, whereas only 7 % of adults without disabilities report the same.

Providers may mistakenly conclude from a routine visit that nothing untoward is occurring emotionally for an individual because the problem is not evident. Observational skills notwithstanding, providers need to ask about emotional issues. Although screenings for emotional problems might be routine in most practices for most children or adults, people with developmental disabilities often seem to slip through the clinical cracks. One problem is that commonly utilized standardized screens for emotional conditions are not standardized for individuals with disabilities. These screens often depend on literacy, insight about feelings, and/or ability to describe these feelings, abilities which may be impacted in some disabilities.

Case 6: Luisa is a 20 year-old woman with Down syndrome, who was quite high functioning, conversant, frequently meeting with friends, enjoying a number of hobbies, until she started a slow decline over the last couple of years. Parents, alarmed by this decline, brought her to a Genetics specialist for a consultation, 2 years previously. The consultant concluded, without the benefit of any tests, that this decline in function represented early onset Alzheimer’s disease. The loss of function has persisted and parents pursue a second opinion through an interdisciplinary Down Syndrome Center. The specialists at the Down Syndrome Center

note that Alzheimer’s disease is more common in Down syndrome than in the rest of the population, but that this is due to the fact that Alzheimer’s disease presents about 20 years earlier in Down syndrome than in the rest of the population. It is noted that Alzheimer’s is virtually unheard of in 20 year-olds with Down syndrome, prompting a more aggressive work-up. Screening of emotional state yields symptoms consistent with depression and obsessive compulsive disorder. Sleep study finds quite severe obstructive sleep apnea. Treatment of these conditions produces significant improvement in functioning over the next year.

Enhancing Function

A relatively new classification addressing function which complements the diagnostic classification, ICD, has been approved by the World Health Organization. The International Classification of Functioning, Disability, and Health (ICF) [25] outlines three dimensions of function and analogous levels of difficulty—body functions/impairments, activities/activity limitation, and social participation/participation restrictions. The ICF provides the conceptual and coding to operationalize these concepts. All of these, however, are influenced by the novel dimension added to the ICF by WHO—environmental factors.

Environments Can Be Either Barriers or Facilitators

Environmental facilitation to reduce or prevent secondary conditions can be seen as the outer boundary of clinical practice. Personal assistive technology, however, is routinely provided for vision and hearing impairments in the general population. These devices are so ubiquitous that they are not commonly viewed as disability related, but simply a part of functioning. For example, assessing what assistance should be

provided to keep impairments (body function problems) such as refractive vision impairment from becoming personal activity limitation (seeing clearly enough to read) or participation restriction (seeing well enough to drive in the community) is considered standard medical care.

In contrast, other assistive technologies, which have the capacity to ameliorate impairments of people with developmental disabilities, and thus to reduce limitation and improve participation of these individuals in the community, are not as universally accepted as part of health care. Newer technologies, such as smart-phones, with various applications, are used by a growing segment of the population to organize their lives. Use of these devices and applications by individuals with developmental disabilities, including cognitive impairments, has the potential to increase community participation substantially. Features such as basic social cues, simple directions (perhaps based on global positioning features), mathematical computation abilities, or emergency procedures can be including, using symbols, signs, oral instructions, or written material, according to level of functioning of the individual. These devices have applications for what is generally considered to be “health-care”, such as reminders to take medications, or reminders of doctors’ appointments. In a broader conceptualization of “health care”, though, these devices and applications can be seen to foster health in people with developmental disabilities by increasing community participation, which, in turn, reduces secondary conditions, such as depression, agoraphobia, and physical deconditioning, which impact physical health as well.

Case 7: Billy is a 4 year-old boy with quadriplegic cerebral palsy. He arrives to developmental pediatrics clinic with his mother pushing him in an umbrella stroller. During the interview, he is constantly sliding out of his seat, and his mother must keep adjusting his position to keep him from falling out of the stroller. During assessment, he is placed in chair with appropriate trunk supports. His mother is amazed to see how well he is able to use his arms when his trunk is thus stabilized.

Mother explains that prescription for wheelchair has been denied by insurance yet again. When mother spoke with person in authorization office, she was advised to “just go get a used wheelchair from a nursing home”.

Sadly, this case, like all the others in this chapter, is not fictionalized, except for the name of the child. For children with cerebral palsy, and other children who lack postural control, wheelchairs function as more than just a set of wheels for carting around the child. The postural support offered by a quality, customized wheelchair, by stabilizing the child, allows the child to develop upper body coordination, eventually toward a goal, perhaps of controlling their own power chair and increasing independent mobility and inclusion. The postural support also helps to prevent aspiration, which may occur with less support, and, of course, may prevent injury that may result from falling out of an inappropriate chair or stroller. Ironically, a chair may be paid for many times over for the same cost as hospitalization for aspiration pneumonia, let alone head injury.

Germane to this discussion is the issue of funding for technology to assist people with developmental disabilities. Medical necessity is the term used by health care professionals to describe the need for various kinds of assistance for restoring function, reducing disease, or restoring physical equilibrium, such as a wheelchair for a person with a mobility limitation or a voice synthesizer for somewhat with limitations in vocal communication. Determinations are based on the notion that such assistance is needed for improved function—at the level of body functions and structures, using ICF descriptions. The definitions used by third-party payers, however, often differ from those used by health professionals in practice [26]. As commonly used, the use of medical necessity as a criterion denies needed services to individuals with disabilities. For example, though an individual with limitations in vocal communication may need an augmentative communication device to be able to communicate with their health care provider, as well as for broader communication which sustains overall

mental and physical health, requests for such technology are often not authorized by payers. Similarly, people who rely on wheelchairs for long-distance mobility are often refused authorization of payment for wheelchairs if they have limited ability to ambulate, such as within their home. However, failure to authorize such equipment impacts community participation and reduces opportunity for social interactions which are, ultimately, essential to health and well-being.

Many people with cognitive impairments do not need the kinds of assistance routinely covered by the medical necessity criterion. They might, however, need accommodations at home to increase safety or independence or health maintenance programs. These and other needed services are important for might be called health necessity. Health necessity describes the services for maintaining function, preventing secondary conditions, increasing functional independence, and equalizing opportunity for participation. Rehabilitation, even, is at times not deemed a medical necessity except as it serves to restore function. In cases of developmental disability, where rehabilitative services, including therapy and equipment, may serve to maintain function at current level, but cannot be reasonably expected to restore or improve function, payers often refuse to cover such services and equipment. Tragically, this may lead to loss of function that otherwise would have been preventable. Clinical practice, therefore, should be vigilant that all possible health necessities are explored for people with developmental disabilities.

Maintaining function and increasing independence to decrease secondary conditions also should be part of the prescription for well-being for these individuals. Care should be taken, however, because the network that maintains assistive technology often is fragmented, the result being technology that is present but nonfunctioning. Under these circumstances, the environmental facilitator becomes a barrier, capable of creating secondary conditions for the individual or family. Fundamental to the success of any environmental facilitator is access, whether to a medical service

coordinator, a technological device, or a personal assistance device. Creativity is a needed characteristic for health providers, finding ways to connect the individual with the needed supports. Over time, the financial benefits as well as the individual functioning are validated.

Interdisciplinary Approach

Finally, the concept of teamwork has focused on one of two notions—the “team” is all of the varied medical specialists involved in a an individual’s care or the “team” is a group of individuals from varied disciplines (including possibly nursing, social work, speech language pathology, occupational therapy, physical therapy, psychology, recreation therapy, and nutrition) who contribute to the individual’s well-being and the well-being of the individual’s family. In the current environment, both approaches to teamwork have suffered. For coordinated interdisciplinary teams to function, time must be allocated for the team members to actually coordinate care. Billing for medical care is usually authorized only for face-to-face patient care, so that funding is not provided for team coordination. In the current climate, with stretched budgets, coordination of care is often cut. As is illustrated in the case of Luisa, these teams are well worth funding, as such teams can be essential to the discovery of treatable secondary conditions, promote health, save money, and prevent suffering.

Conclusions

Allan Myers [27] reported studies showing a direct relationship between the number of secondary conditions reported and happiness, thus highlighting the relationship between health and happiness. He concluded that if individuals with developmental disabilities are to move toward health, there is a need for “greater professional humility to appreciate that people with both physical and intellectual impairments are able to experience and articulate their own satisfaction,

pleasure, and joy". Medicine is a crucial component of health, but health is more than medicine.

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