PERSPECTIVES

Primary Care for the Developmentally Disabled Adult

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The deinstitutionalization of developmentally disabled adults during the past 20 years has resulted in the majority of these persons being cared for by community physicians. Physician training in this area of practice, however, is limited. Primary care physicians often have only a few developmentally disabled adults in their practice, and it can be difficult to develop and maintain expertise while following only a small number of such patients.

Developmental disability is defined as a chronic mental or physical impairment that results in the delay or failure to achieve normal developmental milestones. Often such patients require a combination of special, interdisciplinary treatment, or provision of services for an extended period of time. Developmental disabilities include mental retardation, autism, cerebral palsy, spina bifida, and others. Most developmentally disabled adults are mentally retarded, and many have coexisting medical problems. With the deinstitutionalization of the developmentally disabled beginning in the 1970s, these persons began living in the community, where they are cared for by local, community physicians. Few primary care physicians have formal training in this area of adult medicine, and educational resources in this field for physicians are limited. Acquiring records of previous care and obtaining informed consent can be challenging. In addition, the regulation and delivery of medical care and reimbursement is changing, resulting in frustration for physicians, patients, and their caregivers. For all these reasons, physicians may shy away from assuming care of adults requiring special and sometimes more time-consuming office care.

This article reviews the medical problems frequently presented by developmentally disabled adults and offers an approach to managing them in the office setting. Topics addressed include the medical interview, preventive care, acute chronic illness, special situations, multidisciplinary coordination, and ethical issues. Although these topics are not inclusive, literature review and collective

clinical experience have shown them to be typical problems that face the physician provider.

THE MEDICAL INTERVIEW

The developmentally disabled patient may view the physician's office as a threatening environment (harboring needles and examinations of sensitive body areas) and react initially with agitation and restlessness. Conversely, the patient may withdraw and refuse to cooperate. The patient may become more cooperative if allowed to accompany a family member or group home member to one or more office visits in advance of his or her own initial visit. Should such behavioral interventions fail, premedication with a benzodiazepine such as lorazepam 0.5 mg orally 1 hour prior to an office visit is reasonable, especially if vaccinations, blood tests, or a gynecologic examination are anticipated.

The patient should be accompanied at each visit by a caregiver who knows the patient well and brings a list of current medications. At the initial visit, the caregiver should provide a copy of previous medical records or a written summary of the patient's past medical, surgical, and developmental history. Documentation of guardianship, if it exists, should be furnished to the health care provider for the medical record. Vaccination status, date of the last tuberculin testing, and the presence of the hepatitis B surface antigen are important, especially if the patient is living in a group home or working in close quarters with others (Table 1). Despite the advantages of a caregiver providing the majority of the medical history and taking instructions regarding medical care, the patient should participate in medical decisions when feasible. The patient also has a right to confidentiality and should be offered time alone with the physician to discuss any private matters pertaining to his or her physical or mental well-being.

It is helpful to establish the nature and adequacy of the patient's living situation at the initial visit. For example, caregivers at home may become emotionally and physically exhausted, necessitating intervention. Interventions in such circumstances might involve enlisting the aid of a social worker or visiting nurse, or referral to a day treatment program, which can give the caregiver a daily respite. In cases of abuse or neglect, county adult or child protective services should be notified. Disabled adults in all living situations—family, group home, or institution—are at risk of neglect and abuse. It is important

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for the practitioner and staff to have a low threshold for suspecting and reporting these problems.

PREVENTIVE CARE

Routine Vaccination and Testing

Guidelines for vaccination of developmentally disabled adults follow those of the general population,^{1,2} with attention to increased risk of exposure to contaminated body secretions and risk of living in a semiclosed environment (Tables 1 and 2). The majority of developmentally disabled adults in the United States have completed their childhood immunizations, but there is evidence that these patients have a lower immunization rate than the general population.³ Particular attention should be given to the immunization history of patients raised in underserved, rural communities or abroad. In addition to the usual adult vaccination schedule for tetanus, influenza, and pneumococcal pneumonia, a yearly influenza vaccine and possibly (one-time) meningococcal vaccine should be considered for all patients living in group homes. Group home patients and patients employed in workshops should have a yearly tuberculin test, usually a purified protein derivative. These group home patients benefit from serologic screening for hepatitis A and B, and vaccinations should be offered if the respective titers are negative. Varicella titers can be checked in a young adult without a history of chicken pox, and the vaccine should be administered if titers are low. Routine HIV testing is not considered necessary.

There tends to be a greater deterioration of auditory and visual senses with age in developmentally disabled adults, compared with the general population. There is a need for careful vision and hearing testing in this population. Some states, for instance, require yearly vision and hearing screening for group home certification.

Routine dental care is difficult in many patients because of poor cooperation, especially for painful procedures. It may not be possible to get yearly dental examinations, but prophylaxis and restoration should be offered approximately every 2 years with conscious sedation or under general anesthesia. Patients with Down's syndrome or other chromosomal abnormalities associated with valvular

lar disease may require appropriate antimicrobial prophylaxis when receiving dental care.⁴ Unfortunately, lack of adequate insurance can be an obstacle to good dental care. Medicaid provides for routine dental procedures, but Medicare and most other insurance programs do not.

Exercise guidelines for the developmentally disabled are similar to those for the general population, with heightened awareness of safety. A large proportion of patients with chromosomal abnormalities have congenital abnormalities of the cervical spine, most importantly non-traumatic atlantoaxial subluxation.⁵ Permission to participate in sports that may result in stress to the spine, such as football, diving, and weightlifting, should be preceded by x-rays of the cervical spine.

Colon cancer screening is done the same way in the developmentally disabled patient as in the general population: fecal occult blood testing after the age of 50; and flexible sigmoidoscopy periodically after the age of 50, beginning earlier with a significant family history. The health care provider should decide on a regimen of screening that accommodates the patient's medical and family history, life expectancy, and ability to cooperate. If general anesthesia is necessary to perform a colon examination, the risks of frequent endoscopic screening studies may outweigh the benefits; however, this difficulty does not mean colon screening should never be performed.

Social History

Patients should be asked about alcohol, tobacco, and drug abuse. They should be counseled and offered smoking cessation programs or drug rehabilitation programs as indicated, although the data about effectiveness in this population are limited.

History of sexual activity should be ascertained by asking both the patient and the caregiver and using terms

Table 1. Initial Data Collection

Current medications
Past medical, surgical, and vaccination history
Active problem list
Documentation of guardianship
Home setting (family/group home/independent)
Date of most recent tetanus vaccination
Date of most recent PPD
Anti-hepatitis B surface antibody and antigen
Hepatitis A IgG
Varicella titers

Table 2. Health Maintenance

Basic health maintenance
Routine vaccination
Vision screening
Auditory screening
Dental prophylaxis
Colon cancer screening
Thyroid-stimulating hormone (in Down's syndrome)
Counseling: alcohol, tobacco, drug abuse, safe sex, diet, exercise
Female issues
Pap screening
Contraception counseling
Osteoporosis assessment
Postmenopausal hormone replacement
Breast exam
Mammography
Male issues
Testicular exam (under age 40)
Digital rectal exam for prostate cancer (over age 50)

the patient understands. Sexually active patients need reminders of safe sex and contraception at every routine visit, and should be questioned about symptoms of sexually transmitted diseases.

Male Issues

Developmentally disabled patients may be less likely to report testicular lumps and may not describe or localize inguinal pain well. A careful testicular examination should be done yearly on all male patients up to the age of 40 to screen for testicular cancer. Patients should be examined for inguinal and abdominal hernias, especially if there is a vague complaint of belly pain. Prostate cancer screening is controversial in the general population, but should be accomplished in developmentally disabled patients in a similar fashion. Guidelines should improve as more data become available on the efficacy of screening on morbidity and mortality.

Female Issues

In one survey, only 40% of women aged 18 years or older cared for by primary care physicians had a gynecologic examination within the preceding 3-year period.⁶ Consensus opinion is that gynecologic exams and Pap smears should be performed at least every 3 years in women over the age of 18.¹ Premedication with a benzodiazepine, conscious sedation, or even general anesthesia may be necessary. The current guidelines for breast examinations and mammography should be followed as well.

Mentally or physically disabled women who are unable to manage their menses in a reasonably hygienic, socially acceptable way can have a trial of medroxyprogesterone (Depo-provera) 150 mg administered intramuscularly every 3 months. In a woman over age 35 with a history of irregular menses, an endometrial biopsy or pelvic ultrasound is needed before initiation of medroxyprogesterone therapy to exclude endometrial carcinoma. On this regimen, 70% of women achieve amenorrhea; however, 30% of women develop irregular or heavy menses. Weight gain is another common side effect. When menses are suppressed, follicle-stimulating hormone levels should be checked yearly after the patient is 50 to ascertain the onset of menopause so that the hormonal injections can be stopped. Care should be taken in women at risk of osteoporosis, as long-term use of medroxyprogesterone may be associated with decreased bone mineral density.⁷

Contraception should be addressed in every sexually active woman. The same spectrum of birth control methods available to the general population of women can be offered. These include barrier methods, the intrauterine contraceptive device, oral contraceptives, and medroxyprogesterone. Issues of sterilization are discussed below.

Perimenopausal initiation of hormonal replacement therapy is indicated for women who do not have a strong

family or personal history of breast cancer. Continuous hormone replacement therapy for more than 5 to 10 years is currently controversial for the general population, and a decision about this therapy should be made jointly by the health care provider and the patient or the patient's guardian using available data.^{8,9} The decision should incorporate the patient's anticipated benefits (prevention of osteoporosis, vaginal atrophy, improved cardiovascular morbidity and mortality) and risks (breast cancer). Calcium and vitamin D supplements taken with food are usually indicated.

GENERAL MEDICAL ISSUES

Chronic Illness

As in the general population, middle-aged and elderly persons with developmental disabilities are more likely than younger persons to have chronic health problems such as arthritis, high blood pressure, and heart disease. In addition to these health care problems, many of these patients suffer more frequently from seizure disorders and possibly from depression and other mental disorders than does the general population.¹⁰ For instance, thyroid disease is a common problem in patients with Down's syndrome. One survey of Down's syndrome patients revealed that 20% had unrecognized hypothyroidism or borderline hypothyroidism while another 1.5% had unrecognized hyperthyroidism.³ A thyroid-stimulating hormone should probably be part of routine health evaluation in the Down's syndrome population (Table 2).

Care of the Elderly

Determining the onset of old age in the mentally disabled is difficult, and the literature varies. The Older Americans Act of 1965 defines elderly as age 60. Many persons with mental retardation have a decline in functional living skills beginning at age 50. There may be evidence of premature aging in individuals with Down's syndrome as early as 30 years of age.¹⁰ However, no studies currently provide an accurate estimate of the number of elderly persons with mental retardation in the United States.

In general, severely impaired, less mobile mentally retarded patients have a high mortality rate from respiratory infections. The majority of persons with mental retardation surviving to old age are higher functioning with milder deficits. As high-quality health care is provided, the percentage of older, developmentally disabled adults with more limiting disabilities in the general population will probably increase. Research also suggests that older adults with mental retardation living in the community have a high incidence of affective disorders, depression, and bipolar disorders, and may be twice as likely to show signs of mental illness or exhibit behavioral disorders as compared with the nonretarded population.¹⁰

Acute Illnesses

Acutely ill disabled patients can paradoxically be easier to deal with in the office because they are weaker and less resistive to examination. However, they require much more care at home. They may become dehydrated in the presence of fever, a painfully sore throat, or a gastrointestinal illness. Because symptoms are difficult to elicit, a thorough examination includes the skin in a nonmobile patient, looking for decubiti; the abdomen, to rule out peritonitis or diverticulitis; and the pelvis in the appropriate patient to check for a retained tampon or a sexually transmitted disease. Laboratory studies may include urinalysis, chest x-ray, and a limited computed tomography scan of the sinuses (or a Watter's view). Unfortunately, because the medical history of patients with developmental disabilities is often limited, they may receive tests more frequently than nondisabled patients whose history and physical examination is more straightforward.

Coordinating Home Care

Developmentally disabled patients require more home health care than the general population. In one study, 57% of mentally retarded patients were reported to have a chronic condition requiring home care and treatment.⁶ The physician coordinates this care, which is provided in general by a variety of nursing agencies. The highest proportion (15%) is from nurses employed by their departmentally contracted residential or day program vendor. Only 3% receive services from Visiting Nurse Association or other generic home health care agencies. The nurse, in these situations, functions as a trainer, supervisor, and a monitor of lay health care givers, rather than as a direct care provider.

The number of studies done on the adequacy of home health care is limited. In the study referred to above, 30% of the subjects were judged to be receiving "adequate" care; 61% were receiving "inadequate" care; and 9% had care that was considered "mixed" in that some needs were met well, others less well. There may be a need for increased involvement of nursing care on a long-term basis to deal with diabetes control and other chronic medical problems. Other specialists, including social workers to provide individual or family counseling and management, occupational therapists, physical therapists, nutritionists, and psychologists, can make valuable contributions. The local county board of the Mentally Retarded and Developmentally Disabled can help the physician identify allied health care providers with training or experience with severe disabilities.

Inadequate insurance can be an obstacle to obtaining appropriate home care. The physician and service coordinator for the patient may need to work together to find reasonable solutions and alternatives.

SPECIAL SITUATIONS

In general, medical problems in the developmentally disabled are managed similarly to those in patients with-

out handicaps; however, some frequently recurring situations deserve special attention.

Obesity

Many adults with mental retardation who live in the community are overweight or obese, particularly women. In one study, 40% of mentally retarded women living in a group home setting were obese, compared with 18.9% in the Framingham (Mass.) population.¹¹ For women, predictors of obesity may be mild mental retardation concomitant with group home residence. For men, predictors may be the limitation of mobility concomitant with group home residence. Nutritional guidance is important; patients should be referred early for dietary counseling and receive frequent follow-up with reminders about food choice and exercise.

Weight Loss

Persistent, unexplained weight loss should prompt a careful physical examination plus the usual spectrum of laboratory tests when indicated, including blood counts, sedimentation rate, C-reactive protein, blood chemistry analysis, thyroid tests, urinalysis, chest x-ray, and fecal occult blood testing.¹² While the medical evaluation is proceeding, the patient should be offered nutritional supplements. In the event the workup is unrevealing, the patient should undergo a psychiatric evaluation for depression, anxiety, or psychosis.¹³ Effective appetite stimulants that can be applied safely in most cases regardless of the etiology of the weight loss are amitriptyline or doxepin in doses of 10–50 mg at bedtime.

Behavior

All persons deserve to live in the least restrictive environment possible and to be free of unnecessary physical and chemical restraints. The physician is often called on to treat or refer patients for behavior disorders, in particular impulsivity and aggressiveness. Neuroleptic and benzodiazepine drugs are commonly used but can cause sedation and cognitive deficits.¹⁴ Nonpharmacologic measures as suggested by a psychiatrist, psychologist, or behavior specialist should be considered and applied. Managing stress or providing rewards can be accomplished by the use of increased privileges, sugar-free candies, and music through headphones. Punishment or limit setting is best accomplished by the use of "time out" to one's room or adjustment of privileges. Research continues in finding pharmacologic agents to modify behavior while permitting the patient to function in a reasonably unrestricted environment.^{15,16} There needs to be caution, however, because most of the studies are either case series or uncontrolled, rather than random controlled trials. Also, some researchers combine studies in mentally retarded adults with studies in patients with organic brain syndrome or Alzheimer's

disease. The following list of medications offers alternatives to traditional neuroleptic medications:

- ◆ Selective serotonin reuptake inhibitors (SSRIs) may help reduce self-injurious and aggressive behaviors in autistic and mentally retarded individuals. Positive results have been observed with sertraline (Zoloft),¹⁷ with fluoxetine (Prozac),¹⁸ and by the author (DJR) using paroxetine (Paxil).
- ◆ Busipirone, an azapirone, at 15–60 mg/d, lacks serious side effects and treats mild chronic agitation, anxiety, and irritability. It should be started at low doses and titrated every 4 weeks to effect.
- ◆ Trazodone is a serotonergic drug that, in doses of 25–50 mg/d in divided doses, treats mild to moderate agitation (it should be used cautiously in men because of the risk of priapism).
- ◆ β -Blockers (propranolol at 80–240 mg/d) can help with aggression and tremulousness, with appropriate precautions regarding blood pressure, reactive airway disease, and heart failure.^{19,20}
- ◆ Anticonvulsants including valproic acid (Depakote) and carbamazepine (Tegretol) may control impulsivity and mood swings without sedation, although blood chemistry values (hepatic enzyme levels, blood counts, thyroid function tests, and electrolyte levels) need to be checked early after initiating treatment, then every 6 months.
- ◆ Intramuscular medroxyprogesterone (Depo-Provera) has been reported to eliminate previously intractable sexual outbursts in men.^{21,22}
- ◆ Risperdone (Risperdal) and olanzapine (Zyprexa) are now the antipsychotics of choice. The incidence of tardive dyskinesia with these drugs is significantly less than with conventional medications. Side effects often include weight gain, orthostatic hypotension, and edema.

In some cases these medications can play a dual role; valproic acid can be used to control both tonic-clonic seizures and behavior problems; β -blockers can help manage agitation, tremulousness, and hypertension; paroxetine can help with depression, anxiety, and obsessive compulsive symptoms, and reduce aggressive behavior.

Assessing mentally disabled patients for depression, bipolar disorder, or psychosis requires subjective information from the patient whenever possible and objective information from caregivers without exception. The more limited a patient's cognitive skills, the more reliance must be placed on the reports of the care provider. Self-report instruments like the Beck Depression Inventory, Zung Depression Scale, and the Hopkins Symptom Checklist are generally not helpful. Formal psychological testing depending on the psychologist can be extremely helpful. A patient or caregiver's reports of sleep, appetite, energy, behavioral disturbance, expressions of sadness, anhedonia, social withdrawal, or suicidal thoughts are reliable markers of depression. Mania and bipolar disorder are characterized by elevated energy above baseline; a decreased need for sleep; decreased appetite; agitation; rest-

lessness; frequent, loud, pressured vocalization; impulsivity; and intrusiveness. Manic periods lasting several days to weeks may alternate with depressive episodes also lasting days to weeks.

Psychotic symptoms are exhibited sometimes by evidence of extreme fearfulness as well as bizarre or odd behaviors like picking the air or speaking to people not actually present. Patients with the capacity to speak coherently can actually reliably report visual or auditory hallucinations or frank paranoia. Anxiety disorders such as obsessive compulsive disorder may be more common in the mentally disabled. Phobias, panic disorder, and generalized anxiety disorder, however, may be less common, though data are scanty and accurate assessment techniques are yet to be defined. However, separation anxiety from a primary caregiver can be extreme. Generally, these anxiety conditions respond best to typical doses of SSRIs, busipirone, or judiciously applied benzodiazepines.

Medication compliance by patients who live in group homes or who work in adult workshops can be challenging. Medications are best administered in the morning before the patient leaves for workshop, because patients are often more cooperative with the morning staff than with workshop or evening group home staff. These staff members are most familiar with patients' idiosyncrasies and are better able to convince them to take their medications. Psychotropic medications such as SSRIs, bupropion, monoamine oxidase inhibitors, and stimulants are usually best taken in the morning as they tend to be activating and may cause middle or terminal insomnia. Occasionally the reverse is true for SSRIs, in which case they may be given at night. For the most part, all other psychotropic medications may be given on a twice-daily schedule, working around daily workshop hours and giving the second dose on the day shift if possible. Nighttime dosing is especially recommended for sedating medications like neuroleptics, tricyclic antidepressants, and trazodone.

Cardiac Risk Factors

Cardiovascular disease ranks highest in mortality for the general population, in both men and women. Its role as a cause of morbidity and mortality in the developmentally disabled is less clear. Patients with severe impairments typically die from respiratory problems, complicated by aspiration and nutritional disorders. Persons with mild or moderate impairment living in the community may live longer and have significant risks of coronary artery disease.¹¹ As mentioned above, less restrictive environments seem to be associated with higher rates of obesity and thus increase the risk of diabetes and hypertension. At least 20% of men in group homes smoke. In the Framingham study, disabled men and women living in group homes were significantly more likely to have elevated cholesterol and low-density lipoprotein levels.

Given the prevalence of cardiac risk factors, more cardiovascular disease will be detected in the aging population

of the developmentally disabled over time. The physician should intervene when indicated with dietary changes, cholesterol monitoring, smoking cessation in motivated patients, and appropriate cardiac evaluations to determine when medical versus surgical interventions are indicated.

GENERAL ANESTHESIA OR CONSCIOUS SEDATION

As alluded to above, a small but significant number of developmentally disabled patients will need routine preventive, diagnostic, or therapeutic procedures that cannot be accomplished in the office setting because of the patient's behavior. Commonly, these procedures include dental prophylaxis and restoration, pelvic examinations with Pap tests and endometrial biopsy, endoscopic procedures in the patient with dyspepsia, or cystoscopy in the patient with hematuria or recurrent urinary tract infections. Planning and coordinating the rotation of disciplines such as dentistry, gynecology, and others through the operating room is time-consuming and best accomplished via a "patient pathway" with as much of the procedure planned in advance as possible. Of all the extra tasks required of a primary physician and the office staff to care for these special-needs patients, this coordination is the most time-consuming.

ETHICAL ISSUES

Informed Consent

Confusion around guardianship and informed consent arises frequently during the care of the developmentally disabled person. It is surprising how many mentally retarded adults have no guardian if they have survived elderly parents and have no interested family members. Patients may have a trusteeship service to ensure their government benefits but no one to make medical decisions. Many mentally impaired adults can make reasonable decisions regarding their own routine health care without assistance. Others can make decisions with the assistance of their case manager, their family, or the nursing staff of their residential facility.

Major medical decisions (involving surgery, sterilization, cancer diagnosis, treatment, etc.) generally require a guardianship service of some kind. Guardianship services range from assisting with placement to guiding the patient in making major medical or surgical decisions. Alternatively, full guardianship can be provided to service the patient's benefits, pay bills, and make all medical and placement decisions. Appointed guardians are limited in their decision-making powers, however. They are expected to obtain consultation before making decisions about important issues such as major surgery. They cannot write a Living Will (Advance Directive) for a patient. Ethical decisions about sterilization, abortion, and Do Not Resuscitate orders may need to be reviewed by a probate court. The health care provider needs to know how to help a pa-

tient obtain appropriate protective services and what the limitations of a guardian are in making decisions.

Contraception and Sterilization of Young Women

The history of medical treatment of the mentally retarded in the United States parallels that of the development of civil rights for the mentally retarded. The major legislative act providing broad federal civil rights protection for the mentally retarded is the Americans with Disabilities Act of 1990. Protection of these rights took decades to develop. Early in the twentieth century, sterilization of mentally challenged persons was compulsory, based on "societal interests." Between 1940 and 1970 there was an increased recognition of the rights of the mentally challenged, and sterilization was prohibited. During the past two decades, social action groups determined that all persons have the right to live in the most integrated setting, prompting massive deinstitutionalization. The Rehabilitation Act of 1973 made it illegal for federal grant recipients to discriminate against persons with disabilities. In 1979, legislation was passed to deny federal funds for sterilization of mentally challenged patients. Currently, states differ in their legislation regarding sterilization. States may permit sterilization in some cases, prohibit it in others, or most commonly, offer no legal guidance.

In general, the form of contraception should be chosen after careful deliberation and with informed consent of the patient or guardian.²³ The least permanent and least intrusive methodology consistent with the lowest risk for the patient should be chosen. If sterilization is chosen, it must be clear that the risks of pregnancy and other means of contraception must outweigh the risk of sterilization. Care must be taken to ensure that the patient is not coerced. Typically, multiple interviews with the adult patient, over weeks to months, using the assistance of professionals such as special educators, psychologists, attorneys familiar with disability law, and the physician must be conducted. If the consultants agree that the individual is capable of consenting, court action is not usually required (this depends on the jurisdiction).

If the capacity to consent is uncertain, a court determination of mental competency to make this decision may be required. If an appointed guardian is asked to make a decision regarding sterilization of an adult patient, the probate court in that jurisdiction must approve the decision. Some states do not allow family members or parental guardians to authorize sterilization of an adult woman because there may be a conflict of interest. Because state laws vary, physicians need to know the laws in their own jurisdictions.

Organ Transplantation

Organ transplant decisions do not arise routinely in the care of patients in general, or developmentally disabled patients specifically. Nevertheless, they represent an issue of allocating scarce resources and deserve men-

tion. In general, selection of transplant recipients is based on medical urgency and waiting time. However, low intelligence may decrease the potential recipient's ranking. There was a good deal of publicity in 1995 highlighting Stanford University Medical Center's rejection of Sara Jensen, a woman with Down's syndrome, for a heart transplant. Ms. Jensen, a high school graduate and an advocate of rights for the disabled, began a campaign to heighten awareness of this bias. She successfully underwent a heart-lung transplant at Stanford in January 1996 (N. Peters, *The Times of London*, Oct. 15, 1995; R. Cole, *The Associated Press*, Jan. 23, 1996; C. Fremon, *Los Angeles Times Magazine*, April 14, 1996).

The U.S. Task Force on Organ Transplantation has a patient-centered approach to ranking potential transplant beneficiaries,²⁴ which could be utilized in all decision making involving expensive or scarce resources. A successful outcome is a function of the additional length and quality of life that someone will have as a result of the transplant, compared with what he or she would have had without it. Benefit to society is not an issue; the issue is the relative benefit to the patient. This approach should prompt the primary care clinician not to hesitate to refer high-functioning mentally retarded patients for organ transplant or other aggressive procedures if they have a high likelihood of benefit.

Limiting Care Without Abandoning Care

Physicians and caregivers may set limits, explicitly or implicitly, on the care of a developmentally disabled adult. Decisions to treat pneumonia with intravenous antibiotics, or to operate emergently on an acute appendix may be straightforward. Major interventions such as coronary bypass or long-term dialysis may appear less in the patient's best interest. Many difficult decisions fall between these extremes. Physicians are often pressed to try to clarify their own conceptual guidelines to help the patient and family deal with these issues.

One approach to dealing with these difficult decisions is to view treatment of medical and surgical problems as sustaining life, relieving suffering, or both. Treatment itself may impose suffering, which then must be weighed against the benefit of treatment. The discomfort of draining an abscess almost invariably is outweighed by the benefit; the prolonged burden of chemotherapy may not be. A highly functioning patient may accept a burdensome treatment because he or she understands the long-term goals. A person who cannot comprehend the long-term goal cannot understand the reason for the burdens, such as frequent blood draws, intravenous medications, or disfiguring side effects. To these discomforts are added panic and fear. In fact, with prolonged, painful interventions, patients who are severely mentally impaired see a previously nurturing environment in the doctor's office as a hostile environment, and their remaining days may be clouded by distrust.

The health care provider should try to explain, as explicitly as possible, treatment goals for each patient and use those goals to guide the patient and family through their treatment options for medical and surgical problems. It is important to elicit the patient's wishes. If the patient's will is unknown and basically unknowable, the family, the guardian, and members of the health care team must deliberate over the medical problem and the psychosocial implications of both the problem and the treatment, and attempt to identify the patient's values. One hopes the best answer will emerge. When serious disagreement persists, an ethics consultant or an ethics committee can help guide deliberation and aid in negotiation among family members. Ethicists and ethics committees never assume ultimate responsibility for decisions; that rests with the physician.

CONCLUSIONS

It is important to appreciate that as the U.S. population matures, the number of middle-aged and elderly developmentally disabled adults living in the community will increase. This population may constitute a larger proportion of the primary care physician's practice. Although this population carries a heavier burden of chronic illness than their nondisabled counterparts, studies indicate that the vast majority of their problems can be handled well by primary care physicians. We hope more information will become available to clinicians over the next few years as more controlled studies are performed to define the physical and mental health needs of the developmentally disabled. Providing primary care for the developmentally disabled is challenging but rewarding. Physicians need to accept the challenges of this aspect of medicine and contribute their efforts and expertise to this field.

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REFLECTIONS

Lightness

The rooms in heaven
all have flowers in them,
and Ming vases, bluebells, a ship rose, sweet
william. Blooms in eternity
beginning to bloom.
Water
and bees can get there, and the pale
breeze that unfurls
the scent of the lilac. Mirrors
fix you at any age. I dream my father is always
almost 30. What in heaven
do they pray for.

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