There is no evidence that the usage of cognitive enhancers will increase life expectancy so therefore their utility in stabilizing and improving cognition in dementia should be justified for their continued usage on an individual basis.

**Non-Pharmacologic Management**

Persons with IDD may display increased neurobehavioral signs of dementia including an increase in impulsivity, reactivity to environment, and care handling techniques. Much of what we know about non-pharmacological approaches is drawn from work with typically aging individuals with dementia [25]. As is true of older persons in the general, adults with IDD may become more dependent upon others for care as they decline cognitively and functionally; they may also have less coping skills to deal with feelings of anger, fear and frustration that may accompany their experience of decline. Best practice in care management includes training family and paid caregivers on communication approaches that increase engagement with the person while lessening the demand for information processing and memory load. Communication approaches may include using multiple sources of cuing (visual and auditory aids), as well as simplifying instructions and task demands.

Family and staff caregivers who are knowledgeable about the person’s preferences and history of self-selected activities (when the person had been capable of independently making such choices) can utilize this information to keep the person engaged. Both social and cognitive stimulation in the form of therapeutic recreation activities is recommended to the point tolerated. Dementia has been characterized, phenomenologically, as an illness that deprives the individual of ‘personhood’. There is also an increasing trend towards emphasizing ‘personhood’ in the caregiver’s response to dementia. This movement is aligned with the person-centered philosophy prevalent among developmental disabilities service providers. Briefly, this can be characterized as basing the caregiving approach upon the values and preferences that guided the individual when he or she was able to make his or her own decisions independently. Critical to this approach is to communicate to caregivers that they need to be observant in recognizing if and in what ways an individual’s preferences regarding activities, interactions, foods, clothing, and other personal preferences change and then accommodate those preferences.

**Person-Centered Care**

Individuals may manifest psychiatric symptoms within the context of dementia, a phenomenon which has been termed “Behavioral and psychiatric symptoms of dementia” or BPSD. Individuals with Alzheimer’s disease, and Lewy body and frontal-temporal dementias may experience hallucinations and increased paranoia despite a history free of psychiatric symptoms prior to the onset of dementia. BPSD has been observed in adults with ID who present to emergency rooms for psychiatric inpatient admission. In general in addressing these symptoms, there is little support for the use of anti-psychotic medications [25] as the primary response. This applies to such psychiatric and behavioral symptoms both in the general aging population with dementia or those adults with IDD and dementia. There are instances when medication can help with severe agitation or aggression. However, therapeutic activities, staff handling techniques, and environmental management are usually among the more efficacious approaches. It is important to note that there are specific BPSDs for which antipsychotics should NOT be used, such as for memory loss, wandering, disorientation, apathy, and withdrawal.

In general, neurobehavioral problems are best managed with contingencies, environmental modifications, and caregiver training. For instance, wandering has been helped by offering a “wandering route” or “wandering garden” that allows the individual to safely walk within an enclosed or supervised setting. Individuals who awaken during the night and rummage through closets and drawers can be directed to a “rummaging space” where there are items for the indi-
viduals to investigate, towels to fold, or other simple repetitive activities to undertake apart from others so the person is not disturbing members of the household or other residents within a group home or long term care facility. Sensory rooms can be used either to help the adult decompress when agitated or to become stimulated when lethargic. One of the biggest challenges to caregivers is to keep the person safe, which includes anticipating potential environmental hazards because of the person’s perceptual problems and poor understanding of causal connections. Examples are when an adult might ingest something that is not edible or leave a cooking pot on the stove with the gas or electricity on without understanding the potential fire hazard involved.

It is possible to design behavioral shaping programs in early to middle stages of illness which include providing incentives for pro-social behavior, such as participating in recreation groups or staying in an area engaged in an activity. As a persons cognitive skills decline, so does his or her motivation to engage in and receptivity for programming; thus affecting the efficacy of behavioral programming. Clinicians have utilized applied behavior analysis on a limited basis in long-term care with individuals with dementia although there is limited information in the literature about extending this to persons with IDD. Studies are underway examining the efficacy of aromatherapy [26], music and movement therapy, exercise (such as walking, dancing) [27], gentle massage and the use of white noise [28] as non-pharmacological ways of addressing agitation among persons with dementia.

Individuals at the beginning stages of dementia may still have relatively intact communication skills. It is possible to offer individualized and in-group interventions to help individuals build upon their unique coping skills set and address their feelings and beliefs regarding their own experience of decline. Family and professional caregivers also benefit from the ability to process their thoughts and feelings with regard to care of individuals who are declining. There is increasing awareness of compassion fatigue and caregiver burden that affect both family and professional caregivers. Supports should be provided to help aid caregivers with relieving stress and fatigue and offering respite from continual care.

Organizing Management and Care

The care of someone with IDD and dementia often requires the careful coordination between family support and IDD agency or organizational support, as well the involvement and communication amongst specialists in medical, behavior health, and rehabilitative services. Among persons with IDD, the coordination of care is often a lifelong process that changes as individuals transitions through their various stages in life and development. Coordination of care needs to be person-centered, comprehensive, and proactive, and based on collaboration and continuous communication. The team members involved in this process may change and the addition of specialists may be required, including a nutritionist, occupational therapist, physical therapist, behaviorist, social worker, and palliative and hospice care provider. Additional resources may be the local Alzheimer’s organization, eldercare or agencies on aging, as well as university-based geriatric education centers. The goals and requirements of care will change as the individual progresses thru the various stages of dementia. Sharing relevant information, discussing realistic goals, reviewing and assessing the impact of medication management all are essential in ensuring that the individual’s safety and quality of life with dignity are maintained.

There are a number of factors complicating care decision-making, including but not limited to a much higher prevalence of neuropathology indicative of AD in most older adults with Down syndrome which has implications for course and duration. Studies show that given the average onset age of dementia symptoms is in the early 50s for adults with DS (compared to the late 60s or early 70s for adults with other etiologies of IDD), there is a need for greater surveillance for early symptoms of dementia in adults with DS. Such early-onset dementia also has a shorter course and leads to earlier mortality. Dementia
also presents with variable rates of decline (with duration ranging from 1 to 2 years for aggressive forms to 10–20 years depending on the type and other confounding factors). Given these variations, health care practitioners should note that some currently available residential settings may not offer a safe haven, while others may be well suited for stage-oriented care.

When affected adults live alone, safety and self-care may be compromised and may not be viable with progressive loss of function. When affected adults reside at the parental home, continued stay may not always be possible due to a number of factors (e.g., caregiver burden, physical care needs, home environment inadequacies, etc.). When affected adults reside in small groups (such as in a group home), some homes may not be prepared to provide long-term care, and the localities may not have viable in-community care support in all of these settings. Dementia elicits a change-of-care focus, with a shift from making gains to that of maintaining function and dealing with eventual loss and decline. Setting implications are that care staff trained in ‘developmental and growth’ techniques have to be reoriented to think in terms of maintaining capabilities, care situation planning has to consider progressive decline and greater inadequacies, and care environments have to offer structure, comfort, and familiarity. This is especially germane given the differential rate of decline evident in dementia – which makes attention to stage-related care a paramount consideration.

With this in mind, more local agencies are taking responsibility for the later-life care of aging adults with intellectual disabilities and are adapting and developing small dementia-care group homes [29]. Such group homes are designed to be ‘dementia-capable’ and provide for extended long-term care. Here, the health care practitioner can help by encouraging such forms of care and if tenable, provide on-site consultation to increase staff competencies and improve quality of care.

**Community Care Options**

As with the general population, adults with IDD with impairments due to dementia face limited options for long-term care. In most jurisdictions, dementia care options primarily include some variation of institutional care (such as nursing homes, old age homes, and dementia special care units), neighborhood group care (such as board and care homes, adult care homes, and old age home), and family care (e.g., living with family or other relatives, or informal caregivers). Among IDD care agencies, the latter two are also prevalent, but there is also an increasing use of specialized ‘dementia-capable’ group homes. Most IDD agencies or organizations try to avoid making referrals for admissions into generic nursing or other long-term facilities due to concerns with the quality of care given to persons with IDD and not wanting to expose some of their clientele to ‘re-institutionalization’. When continued care is needed most options include support for families that are committed to providing continued care at home, or admission to a general group home or specialized dementia-capable group home for adults with IDD [7, 30].

**Family In-home Care**

Adults with ID affected by dementia who live with their families pose significant challenges for their aging parents and siblings as their function deteriorates [31]. In addition, as physical needs become more prominent, aging parents may be less capable of continuing to provide care at home (due to the burden of 24/7 care, their own aging, or the home no longer being functional), and the situation may lead to a crisis. Thus, families may need help in recognizing onset and understanding the progression of dementia in their son or daughter, their home may have to be physically adapted for dementia care, and they may need to be introduced to local support groups that share information about dementia and behavior management and supports. They should be advised that more significant problems may arise later as dementia progresses and care may change from ambulatory (i.e., the person walking about) to nursing focused care (i.e., the person needing bed care). Family caregivers should be helped with providing both physical and social care and given information about Alzheimer’s disease,
medical needs, behavior management, nutrition, and personal care, dementia-related behaviors, communication with people affected by dementia, and how to access outside help. The possibility that the aged caregiver may have concomitant cognitive and other physical challenges themselves must be appreciated as to how this may impact their ability to care for their son/daughter or other relative with dementia. Thus, consideration must be given to treating the whole family, i.e., the caregivers and the adult with dementia as a unit.

**Group Home Care**

Most community agencies use one of two community-living and support models for long-term dementia care [30]. One is “aging-in-place”, or supporting an individual for the course and duration of their infirmity. In this model the person affected progresses at home through the stages of the dementia and receive supports adapted at each stage. Agency resources are allocated as they would be to any chronic debilitating illness. The other is “in-place progression”, here a group of adults with dementia live in a specialty group home, with specially trained staff and a ‘dementia-capable’ environment. An agency may have several distinct homes for such care, and try to match the residents to the stage-determined level of care provided for in the homes. Such homes are generally set up to provide long-term care for adults in their mid- to later and advanced stage of Alzheimer’s disease (or other dementias). Generally staff with differential skills are assigned, depending on the mean stage presentation of the residents. For example, staff in homes specializing in late or advanced stage care tend to be more oriented to skilled nursing care, whereas in homes with ambulatory residents, staff are selected for their capability of engaging residual memories and capabilities, providing socialization, and engaging them in diversionary activities as well as providing personal care. Such homes usually have staff trained in knowing signs for changing capabilities and possess an elevated “index of suspicion”. The buildings are designed to accommodate the needs of residents with dementia (e.g., control for problems resulting from memory loss, disorientation, and wandering, while incorporating cues and familiarity). The staff-to-resident ratios generally are stage-dependent (e.g., more staff are assigned to homes with residents who are ambulatory and may be in the mid-to later stage — where personal care demands may be higher). Most local authorities are recognizing small group homes as practical solutions to providing quality neighborhood-based long term dementia care. When dealing with a patient living in one of these residences, liaising with the home’s manager and primary care staff is an important part of the intervention and treatment plan.

**Palliative and End of Life Care**

Participating in later life planning and in determining the aspects of advance directives and end-of-life planning is also crucial to respecting the dignity and rights of adults affected by dementia and should occur while the adult is cognizant, can benefit from counsel, and can still participate in life decisions. With respect to the latter, enabling decision-making is crucial to fulfilling the dignity and rights of adults affected by dementia. Choice and liability are best managed when providers enable the individual and family to be part of the decision-making process about dementia care and supports [7].

As physical and behavioral health begin to decline, especially during the latter mid-stage and progressing into late and advanced-stage dementia there will be a need to consider palliative and hospice care. Palliative care should be considered when the individual is showing signs of significant functional decline that may have associated with physical and emotional pain and discomfort. This type of care may reduce pain and suffering and allow the individual to have comfort as well as to those around him or her.

Hospice care should be considered and called upon during the end stage of dementia. In the United States, according to the Medicare Benefit Policy Manual [33], criteria that may signal a
need for referral to hospice can include two or more episodes of pneumonia or other serious infections during the past 6 months; difficulty eating and swallowing, even with feeding help, that results in weight loss of 10 % or more over the preceding 6 months; and/or one or more skin pressure ulcers that are not healing. On an emotional level, the acceptance by families, support staff, and their agencies of the realization that the individual is within his or her last year of life is never easy to believe and accept. The coordination of care and having an open and honest discussion during the course of the disease can make this sad and trying time somewhat easier for all involved. Being sensitive to the feelings and concerns of involved caregivers and staff when someone is admitted to hospice is an important part of the consultation process when following a patient.

**Commentary**

The emergent acknowledgement of the increase rate of dementia among adults with IDD has led to the recognition of the import of a lifespan approach to securing and providing services and supports. Consideration of what is most needed in older age has taken on more prominence. Service organizations are responding to the emergence of age-associated neuropathologies as coincident to lifelong conditions and are attempting to adapt their services for continued community care until death. The historical focus on aging among adults with IDD [33] and the recent focus on dementia brought on by the National Plan to Address Alzheimer’s Disease [34] and its complement in the work of the National Task Group in Intellectual Disabilities and Dementia Practices [8] in the United States has heightened awareness of the latter age needs of adults with IDD – and in particular those at-risk of or already affected by dementia. The positive outcome of these public health initiatives is that more dementia-capable services are being developed, technologies are improving, and there is an increased interest in maintaining quality of life through to the end-of-life, irrespective of the nature and complexity of conditions prevalent in older age.

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**References**

Table 116.3 (continued)

<table>
<thead>
<tr>
<th>Name of measure</th>
<th>Instrument characteristic</th>
<th>Strengths/weaknesses</th>
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<tbody>
<tr>
<td>Test for Severe Impairment (Modified)</td>
<td>A 24 item cognitive test that takes 10 min to administer, it tests language, memory,</td>
<td><strong>Strengths:</strong> Most people with moderate and severe ID should be able to score on the</td>
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<tr>
<td></td>
<td>conceptual ability and spatial skills.</td>
<td>TSI unless they are at an advanced stage of dementia. Many report enjoying</td>
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<td>completing the scale.</td>
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<td><strong>Weaknesses:</strong> The small number of items</td>
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<td>within each subscale may not always be sufficient to detect subtle changes over</td>
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<td>time.</td>
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Source: NTG (2013) and adapted in part from British Psychological Society (2009)

Originally named the Dementia Questionnaire for Mentally Retarded Persons (DMR)

In discussion with the practitioner, the staff identified concerns regarding safety, well-being and quality of life. The team, which includes Jonathan, his family and professional caregivers, met and identified areas for increased support including developing a daily schedule with Jonathan, providing him with visual and auditory memory cues to assist his completion of daily routine. It was suggested that Jonathan visit an adult services center that offers social and recreational activities. The team members wanted to lessen task demands on Jonathan yet maintain opportunities for his mental and social stimulation and pursuit of preferred activity. Jonathan was scheduled for a direct psychological assessment which will include a baseline of his attention, memory, language and problems solving skills. He is scheduled for a follow-up with the neurologist in 6 months. Staff will be providing frequent, albeit brief, spot checks with Jonathan. The staff will be trained on communication skills and handling techniques; they will offer Jonathan skills retraining and more assistance during personal care. The agency is considering special program options for individuals whom they support who may need a modified day program consistent with their changing needs.

**Course of the Disease**

Alzheimer's disease is a progressive illness that will lead to loss of function, changes in behavior, growing needs for caregiver support, gross debilitation, and eventual death. It would be best to consider the type of care based on a stage-of-disease approach: early, mid-stage, late, and end-stage. The entire duration of the disease may be as long as 10 or more years, but there have been a number of cases in adults with Down syndrome who have had a rapid progression leading to death only within 1–2 years [21]. Anticipatory guidance is recommended to help prepare caregivers of what lies ahead so that expectations and preparations are made to help best deal with the increasing challenges and stressors of caring for the patient with dementia. During the early stage of disease it will be essential to provide counseling in regards to communication strategies; modification of expectations in the daily environments; safety; observation and assessment of behavioral changes; impact upon family, friends, and staff; and to determine how any applied therapies may be needed or altered to address the current health difficulties. During mid-stage dementia the individual may begin to change dramatically with progression of cognitive and behavioral difficulties. Dysphagia, seizures, gait instability, urinary incontinence may all begin to occur and progress requiring more caregiver support and supervision. Individuals who progress to late and end-stage dementia may need additional support that may not be available within their home or community living situation. Self-care, deterioration in hygiene, and greater dependence upon others greatly occurs during this time. Loss of control of bodily functioning, loss of mobility, and an increased risk of respiratory and urinary infections increases. Falls and risk for serious injuries can be problematic. Concerns by caregivers, when falls or other untoward behaviors occur, may lead to increased use of emergency room and other
urgent care. Agitation and aberrant behaviors can be exacerbated during this time. The healthcare provider will be asked to provide recommendations and guidance on how to best reduce and eliminate some of these challenges. As swallowing function deteriorates, recurrent aspiration pneumonia, malnutrition, and dehydration become predominant clinical concerns. A discussion on feeding tube placement, ventilator support, hospitalization all become prominent issues during the late and end-stage of the disease.

An open and honest discussion with the families and appropriate guardians needs to take place well in advance of these difficulties so that decisions regarding type and intensity of interventions are made in advance of the inevitable clinical crises. Caregiver burden greatly increases as the disease progresses and potential “burnout” can occur which can increase the chance for neglect and abuse. Looking out for these signs is essential as well as ensuring that the support team and family are provided the necessary recommendations and referrals for additional community-based services. Dementia and its related cognitive and challenging behaviors may become significantly exaggerated at any time during the illness if an environmental, social, or underlying health issue arises. If acute changes occur to the individual it is imperative that the underlying cause for these changes be determined and rectified if at all possible. Crisis intervention including emergency department and hospital admission may be thus avoided.

Pharmacologic Management

The mainstay of therapy is to provide cognitive enhancers which are meant to stabilize, improve, and help slow the progression of Alzheimer’s disease. Cholinesterase inhibitors; Donepezil (Aricept), Rivastigmine (Exelon), Galantamine (Razadyne) curb the breakdown of acetylcholine, a chemical in the brain important for memory and learning. These types of medications help increase the levels of acetylcholine in the brain. These drugs may slow the progression of symptoms for about half of people taking them but only for a limited time, on average 6–12 months. Common side effects are usually mild for these medications and may include diarrhea, vomiting, nausea, fatigue, insomnia, loss of appetite, and weight loss. Though generally mild, these side effects may be significant enough to prompt medication discontinuation in some individuals. The benefits of cholinesterase inhibitors in adults with DS have not proven to be clearly beneficial [22, 23].

Memantine (Namenda) is approved to treat moderate-to-severe Alzheimer’s disease. Memantine works by a different mechanism than other Alzheimer’s treatments; it is thought to play a protective role in the brain by regulating the activity of a different brain chemical called glutamate. Glutamate also plays a role in learning and memory. Brain cells in people with Alzheimer’s disease release too much glutamate. Memantine helps regulate glutamate activity. Memantine is the only drug for Alzheimer’s that works this way. It may improve mental function and performance of daily activities for some people. Memantine may have increased benefit when used with Donepezil, Rivastigmine, or Galantamine. Side effects of Memantine include tiredness, dizziness, confusion, constipation, and headache. The usefulness of Memantine with persons with IDD is still in question. When tested in a prospective randomized double-blind trial Memantine failed to show any significant benefit in cognition with adults with DS [24].

While these medications have been tested on older adults without IDD, there have been no large randomized controlled studies in adults with intellectual or developmental disabilities. There is very limited evidence from the current literature about the efficacy, safety, and tolerability of pharmacologic interventions for dementia in adults with IDD. If these medications are prescribed then there should be in place a serial assessment strategy to help determine whether these therapies are successful. Data and communication about cognitive function, levels of caregiver support, and severity of dementia related symptoms need to be reviewed at each office visit to help determine the course of the disease and the effects that therapies may have upon the decline of function.