Dementia and intellectual disabilities

Why the concern about dementia among people with intellectual disabilities?

Everyone has to face the prospects of the increasing health risks that are associated with the expectation of living longer than ever before. Increased life expectancy is the result of discoveries that have led to major improvements in health over the past several decades.

However, with longer life the risk of dementia is expected to show a corresponding increase in the general population. Ageing individuals with intellectual disabilities (ID), a condition of lifelong limited intellectual, social and vocational capabilities, are also living longer and healthier lives because they share in the same benefits from medical discoveries. Consequently, they are also at increased risk of developing dementia.

How many people with intellectual disabilities are affected by dementia?

Population studies show that the prevalence of dementia among people with ID appears to be about the same as in the general population, which is about 5% of people aged 65 and older. However, the prevalence of Alzheimer’s disease, the most common cause of dementia, among adults with Down’s syndrome is about 25% for those who are 40 years of age and older and about 65% for those wo are 60 years of age and older. Thus, people with Down’s syndrome are especially vulnerable to developing dementia. This can be explained by the fact that people with Down’s syndrome have three copies of the portio of chromosome 21 which can contain a gene for amyloid precursor protein (APP). Over production of APP leads to its breakdown in such a way that amyloid and plaques are formed and eventually Alzheimer’s disease is manifested.

How are people with Down’s syndrome affected differently by Alzheimer’s disease?

Although only a fraction of people with Down’s syndrome show the symptoms of Alzheimer’s disease, upon autopsy nearly all older adults with Down’s syndrome show the brain lesions that are characteristic of Alzheimer’s disease. Men and women seem to be equally affected. The disease lasts on average for about eight years, with a range from one to twenty years. Average age of onset is about 50, followed by death at an average age of about 58 years. The natural history of Alzheimer’s disease in people with Down’s syndrome is similar to that of people from the general population with the notable exception that it starts about 20 years sooner in people with Down’s syndrome.

The symptoms of Alzheimer's disease show wide differences between people with Down's syndrome. In the early stage of the disease, memory loss is not always seen and not all the symptoms associated with Alzheimer's disease will be recognizable. Generally, changes in activities of daily living and work habits are noticed first. Epileptic seizures may occur early or late in the course of the disease. Cognitive changes are frequently present but they are difficult to evaluate because of limitations in the individual's language communication and related intellectual abilities.

How is dementia diagnosed in people with intellectual disabilities?

There is no test for dementia that can be used for ageing people with ID who are showing deterioration in function. They face the same situation as ageing people from the general population who are suspected of developing dementia. Diagnosis requires the exclusion of every other possibility. Many of the standard tests for this purpose are those used on the general population who are suspected of developing dementia. Diagnosis requires the exclusion of every other possibility but many of the standard tests used on the general population are inappropriate for people with ID, particularly for those with severe deficits in intellectual function.

It is recommended that comprehensive physical, medical, neurological, radiological and laboratory studies be combined with assessments of cognitive functions. Diagnosis of dementia should be reserved until after the evaluation of a similarly comprehensive follow-up examination six to twelve months later. Only the observation of significant changes during this six to twelve month period should be used for making a diagnosis of possible or probable dementia. Such evaluations are necessary to rule out conditions other than dementia and reversible causes of dementia.

Complete evaluations are the same as for other people suspected of having dementia. It is also important to discuss the individual's symptoms with members of his or her family or others who are in regular contact with them. Detailed guidelines are available from the American Association on Mental Retardation (AAMR) at www.aamr.org/ Reading_Room. These guidelines have been adopted by the AAMR and the International Association for the Scientific Study of Intellectual Disabilities (IASSID).

What services are needed by individuals with ID affected by dementia?

People with ID who are diagnosed with dementia can continue to live in the community if the right support and assistance are provided. A resource list is available from Dementia SA. Some service providers have effective specialist teams that set up individually tailored care services. They also provide essential home care. Since people with ID affected by dementia may not be able to continue to live on their own, so 'dementia capable' housing and support need to be provided. Use of daycare (or day activity services) is recommended to maintain their quality of life and adequate supervision. Home services can reduce the need for, and delay the time of, admission to expensive care institutions.

Principles that should guide the care decisions of people with ID and dementia.

The Edinburgh Principles were developed following a conference held in Edinburgh, Scotland in 2001. In summary, they are as follows:

The main principle guiding decision making should be that services should be aimed at meeting the individual needs of each person. This principle is summarized in the concept of 'person centered planning' which has been widely promoted as an effective framework for service delivery. The focus is on making decisions based on the individual strengths, capabilities, skills and wishes of each person. The individual's family, relatives and closest friends should be engaged with the person in all decisions with the aim of helping the person remain in his or her chosen home and community.