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Healthy ageing in people with Down syndrome and dementia: the need to foster education and support programs for individuals, families and organizations

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Abstract

Most individuals with Down syndrome (DS) reach an advanced age which gives rise to specific senior-citizen needs. These must be considered and addressed, through anticipation and prevention. Difficulties are already emerging in services responsible for this population, as well as for the individuals concerned and their relatives. The inadequacy and unsuitability of policy planning and lack of adequate services are made apparent whenever a person with DS begins to develop cognitive deterioration or dementia.

The World Health Organization (WHO) has drawn up, in conjunction with the International Association for the Scientific Study of Intellectual Disabilities (IASSID) and Inclusion International, a report on the social and health care needs of aging persons with intellectual disabilities, including those with DS. This working document highlights as a priority the need for each country in the European Union to implement "Projects for Healthy Aging" that are naturally conducive to social inclusion while improving support and training for both formal and informal caregivers. The present article provides a rough outline for potential future programs targeting individuals with DS and cognitive deterioration or dementia.

Keywords: Care programs. Dementia. Down syndrome. Main caregiver overload.

Introduction

From time immemorial, aging has been a process that never fails to surprise and alarm. Aging and its consequences are an issue that affects us all. Moreover, it is a core issue in the sociodemographic structure of industrialized countries.

Despite this considerable interest, aging in people with intellectual disabilities is only a recent area of study. In the past, the living conditions endured by people with intellectual disabilities resulted in premature death. Planning for age- and health-related changes and enhancing or maintaining their adaptive competencies were never points for consideration.

Whereas adult life expectancy was once inexistent, this has changed dramatically in the last 50 years. An overall increase in care quality has boosted the life expectancy of this population, who can now look forward to reaching adulthood. In Catalonia, for instance, 10% of people with intellectual disabilities are over the age of 50.

With regard to life expectancy in people with Down syndrome (DS), around 80% now live beyond the age of 50, with some reaching 60 and even 70 (1). This increased life expectancy is fundamentally down to medical and social developments, the former consisting largely of such vital schemes as widespread use of antibiotics, immunization and the development of cardiac surgery for heart defects, and the latter – the social developments – consisting of schemes to phase out institutionalization and implement specialized community-based care and family support services.

Most people with DS now reach an age at which their needs as senior citizens must be considered and met, and this necessitates anticipatory and preventive measures.

This development is already putting a strain on the services in charge of caring for them as well as on their families and the individuals themselves.

When addressing aging and how it may affect people with intellectual disabilities (ID), it is important to bear in mind that this is a highly heterogeneous group. While all individuals with ID display signs of delayed or abnormal development with significant impairments, both intellectual and functional, the origin, development profile, nature and extent of these may differ enormously.

In fact, some such conditions may be related to the etiology of ID. Thus, for instance, people with DS are at greater risk of developing certain chronic illnesses than others whose etiology is different (2, 3). DS is well known to be caused by an overexpression of the genes in one segment of chromosome 21 – the Down syndrome critical region (DSCR), or Down locus. In addition to this cause of ID, there is Alzheimer-type age-dependent neurodegeneration. Those over the age of 40 are far more likely to experience onset of a cognitive deterioration process. In a study of people with DS (4) (x age: 46.5 years) with a total N of 285 participants, the prevalence of Alzheimer-type dementia (ATD) was estimated to be 13.3%. Although the primary cause of dementia in people with DS is ATD, typical pathological descriptions corresponding to Lewy bodies (LB) have been documented, and give rise to dementia with Lewy bodies (DLB).

Clearly, external manifestations will vary depending on whether the combination is DS-ATD or DS-DLB. Patients in the former group should present deterioration characterized by cortical cognitive impairment (memory defects, aphasic and apraxic changes, disexecutive syndrome – bearing in mind prior cognitive deficit) together with psychological and behavioral changes (irritability, increased anxiety, wandering) and functional changes (loss of previously learnt adaptive skills). Patients with DLB, on the other hand, should present a pattern of predominantly frontal neuropsychological involvement, more visual hallucinations and a marked presence of extrapyramidal symptoms, which could also explain the high prevalence of extrapyramidal impairment in DS (36%).

Primary caregiver overload

Caring for a patient with dementia in the home is a complex process in that it is a chronically stressful experience which affects both the mental and physical health of the primary caregiver. In both medical and psycho-social literature, the concept of "overload" is used to describe the emotional tiredness felt by the primary caregiver (PC).

Overload refers to the psychological state brought on by the combination of physical work, emotional pressure, social restrictions and financial demands that arise when caring for the patient (5, 6). In recent years numerous studies have focused on attempting to identify the variables involved in the overload experienced by primary caregivers of people with dementia (7-10).

Some of the most common causes behind PC overload are personality changes in the patient, increasing difficulty exercising adaptive skills (11), lack of support (12, 13), not having the right strategies to manage care of the patient (14, 15), and behavioral disorders in the patient (16).

Measures devised to reduce caregiver overload abound for patients with ATD alone – that is, without prior ID (17). These range from psycho-educational measures (18) and emotional support (19) to telephone hotline support techniques, and even admission to specialist units known as 'respite' units (referring to the break for the family). An array of studies conclude that this latter resource is the most effective in cutting down on PC overload (20), although it is only immediately effective, as the improvement is short-lived (21).

In fact, medium-stay units specializing in dementia patients in crisis situations (intercurrent problems, behavioral disorders or family support issues) have been created in recent years and are on the up. Such units use qualified multidisciplinary staff and structural measures to enable safe care for patients with dementia.

But what about when the person is affected by dementia in a context of DS?

Policy planning in this case is clearly shown up as poor and inadequate, and the shortage of services to deal with people with DS and dementia becomes quite plain. It therefore falls to family members, particularly the mothers, to provide 'normal' care which can continue well into later life, forcing them to take responsibility for a family member with a disability at a time in their lives when the caregivers themselves need some kind of support.

Research has shown that most families see caring for people with DS – with no suspected dementia or added cognitive deterioration – as very rewarding. However, the experience becomes highly stressful, with high rates of emotional overload, when there is dementia in a moderate or severe phase, and this is compounded for very elderly caregivers, when the situation can become very difficult.

Furthermore, strain and stress in the PC have been proven to increase the care burden, which is greater when adequate support and information are lacking.

The World Health Organization (WHO) has drawn up, in conjunction with the International Association for the Scientific Study of Intellectual Disabilities (IASSID) and Inclusion International, a report on the social and health care needs of aging persons with ID, including those with DS (22). This working document highlights as a priority the need for each country in the European Union to implement "Projects for Healthy Aging" that are naturally conducive to social inclusion while improving support and training for both formal and informal caregivers.

Implementing schemes for people with DS and dementia: a possible reality in Catalonia?

Janicki et al (23) have put forward a four-phase procedure for planning initiatives in the field of aging in people with intellectual disabilities:

1. Identify the population.
2. Assess service users' needs and available services.
3. Define services, including problems and hindrances, particularly those that are needs-related.
4. Propose solutions to the problems

This outline has been used to put forward a model to implement schemes aimed at people with DS and cognitive deterioration or dementia added to ID:

Step one. Identify the population.

The target population must consist of people with DS and cognitive deterioration/dementia.

Step two. Assess service users' needs and available services.

The emotional disturbance and behavioral and psychiatric problems of service users with ID – and often masked by the disability itself – have created a new need, one already detected and addressed in other countries but only recently so in Catalonia: creation of a specialist care service network for service users with ID and mental health or behavioral disorders.

In Catalonia, in order to improve mental healthcare for people with ID, a new model has been set up involving the creation of a range of specialist services. Together with the networks run by the Health and Welfare & Family departments, these comprise a care circuit providing the most suitable services for each individual with ID at every stage in their development.

The work of these services is described in a document entitled "Mental Health Needs in People with Intellectual Disabilities" (24).

In fact, assessment of behavioral disorders is the chief ID-related demand experienced by mental health centers (MHC), and the primary cause of admission to psychiatric referral units.

Broadly speaking, needs are underdetected, so by the time a patient is admitted the situation has usually reached a breaking point, with increased behavioral disorder in the service user and overload beyond the caregiver's ability to cope. Some admissions are due to decompensation in the dementia process.

In the field of ID and dementia there is a need for customized tools and specific training, both for diagnostic guidance and intervention.

On the whole it is accepted that such a high level of specialization frequently cannot be offered by general services.

It is a well-known fact that service users with ID and mental or behavioral disorders – including those with DS and dementia – are frequently excluded from the generic mental health model for various reasons such as a lack of experience or the skills to channel and treat the needs of these people.

In recent years, significant breakthroughs have been made in all matters relating to diagnostic evaluation, assessment and treatment of mental disorders (including dementia) and behavioral disorders in ID. New diagnostic tools have been developed and we have better knowledge of the manifestations of ID and psychiatric disease/behavioral disorders.

The new portfolio of services includes the SESM-DI (Specialist Mental Health / Behavioral Disorder and Intellectual Disability Service) which acts as a central backbone for all other resources, ensuring continuity of these processes, which tend to be drawn out (24) (Figure 1).

The SESM-DI offers clinical services (evaluation, diagnosis and intervention and follow-up plans – both outpatient and home care), advice and training. As a result of these new, more community-based and comprehensive care models, a need for new training schemes aimed at non-specialist professionals has arisen. In the UK, for instance, training has been a priority target for many years (and remains so), enabling psychiatrists, psychologists and nursing staff to receive lifelong and specialist training.

Step three. Define services, particularly those that are needs-related.

In the 18 months that the SESM-DI in the Martí i Julià Hospital (Gerona, Spain) has been running, more than 13% of cases have been people with DS who have been diagnosed with probable dementia following an intense assessment process. In most of these cases the deterioration had already reached the moderate phase. A further 9% of cases involved people with DS to be reassessed within 9 months for suspected cognitive deterioration.

The following needs have been noted in everyday clinical work:

– Identifying cognitive processes in people with DS over 35.

– Tools that enable staff to carry out strict assessment of needs and necessary support.

– Ensuring equal access to treatment (drugs, cognitive treatment and welfare services) as enjoyed by the rest of the population.

– Devising and supplying educational information that is intellectually accessible for older people with ID and their families.

Step four. Propose solutions to the problems.

Having reached this stage it is worth considering the need to set up a scheme that would enable us to:

– Improve information and develop schemes to screen for Alzheimer disease in people with ID and DS above 35 years.

– Take inventories with screening tools for primary care services (health and social welfare), and organizations that care for people with ID and their families.

– Identify and quantify the needs of people with DS and dementia.

– Identify and quantify the needs of caregivers (formal and informal) who live with or provide care for people with DS and dementia.

– Devise frailty indicators related to aging and dementia in people with DS and draw up relevant preventive health schemes.

– Improve family and caregiver support by means of a Lifelong Training and Information Plan in health education.

– Support and educate primary caregivers by fostering helpful attitudes/values through 'Caring for the Caregiver' programs.

– Enhance the primary caregiver's/family's ability to adapt to the deterioration process.

Running a scheme with these characteristics that addressed the wide range of functions related to DS and dementia would probably, among other things, enable us to:

– Identify dementia processes in people with DS, giving them equal access to drug treatment, rehabilitation and welfare services.

– Garner information on the needs of people with ID who are aging or have dementia which could be used to supply organizations and authorities with useful information for the planning and provision of services for users, families and centres.

– Increase social inclusion and encourage community living, reducing institutionalization.

– Reduce the burden on the main caregiver (family) in providing care for people with DS who are aging or have dementia.

– Improve training for the professionals who provide care for this kind of service user and their families.

In summary, this would enhance quality of life both for people with DS and cognitive deterioration/dementia and for their respective caregivers.

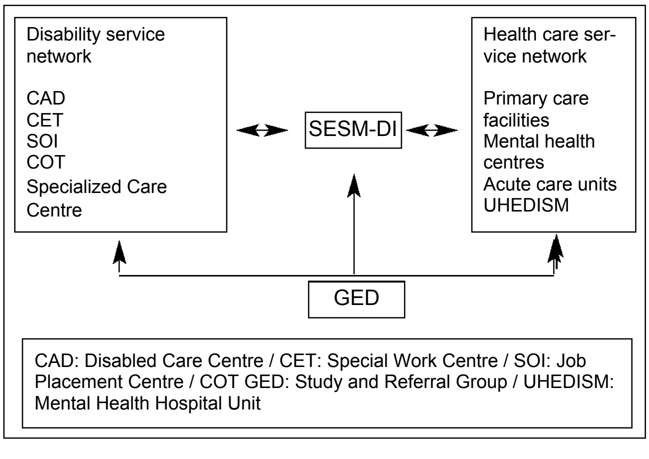

Figure 1. The SESM-DI acts as the central coordinator for all other services on both networks.

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