

Down Syndrome and Alzheimer Dementia —

Some common medical issues and implications for palliative and end-of-life care

The presence of symptoms of dementia in persons with Down Syndrome and with intellectual disabilities of other aetiologies poses challenges for physicians, nurses and other staff. The following observations reflect the growing literature on the health consequences of dementia, as well as the author's own clinical experiences in working with people with Down Syndrome and dementia and participating in multidisciplinary efforts to address the needs of these individuals. Challenges to be addressed include assessment and early diagnosis, medical surveillance, identifying and responding to common health problems, and care planning considerations in the later stages of dementia

Assessment and early diagnosis

The association of Alzheimer dementia with Down Syndrome is now well recognised among healthcare professionals, carers and families of people with Down Syndrome. Dementia screening is being increasingly used to diagnose Alzheimer dementia in people over 35 years old with Down Syndrome. Where screening is available, not only are more cases of Alzheimer dementia diagnosed, but the diagnosis is made earlier in the course of the disease.

Early diagnosis is important as it gives time to investigate various forms of intervention. Medications are now available which may slow the progression of the disease; a trial of these may be considered appropriate. In addition, carers can share and receive information regarding the course of the illness. It is important that this knowledge is availed of when deciding on the management of medical complications that arise during the progression of the disease.

Medical surveillance

The importance of medical surveillance among people with Down Syndrome cannot be over emphasised. Many medical disorders that occur in the general population also occur in people with Down Syndrome, but with greater frequency. Most of these are treatable disorders, but if left undiagnosed they could potentially pose a further handicap to this group of people. Medical monitoring of people with Down Syndrome should be life long. When a diagnosis of Alzheimer dementia is made the necessity of medical surveillance is all the more important to ensure that any treatable medical or mental health conditions are recognised and promptly managed.

Identifying and responding to common health problems

There are a number of medical problems that may be encountered in the person with Down Syndrome and Alzheimer dementia. While it is beyond the scope of this article to discuss all of these conditions it may be useful to highlight some of the more common problems.

Epilepsy:

While only 10% of people with Alzheimer dementia in the general population develop epilepsy, the figure for people with Down Syndrome is reported to be as high as 80%. In people with Down Syndrome the seizures tend to appear after the onset of other features of cognitive deterioration. Many different types of seizures may be seen, the most common being myoclonic seizures.

A myoclonic seizure is a brief contraction of a muscle, muscle group or several muscle groups. It can be single or

repetitive, varying in severity from tiny twitches to severe jerking. In people with Down Syndrome and Alzheimer dementia myoclonus can become quite troublesome, especially on awakening or as a startle response. As the dementia process progresses the myoclonic seizures may become quite resistant to treatment with medication.

When treating seizures in people with Down Syndrome and dementia an appropriate anti-epilepsy drug should be prescribed with the aim of optimum seizure control with the least side effects. A protocol for the use of emergency medication for prolonged seizures should be available to all.

Pressure ulcers:

Pressure ulcers or bedsores are not uncommon in people who are bed bound and particularly those with dementia. Pressure ulcers develop as a result of unrelieved pressure over certain parts of the body, usually bony prominences, such as the heels and the hip area. In people with Down Syndrome and Alzheimer dementia the combination of immobility, moist skin due to incontinence, possible nutritional deficits and the decreased perception of pain due to their cognitive deterioration can lead to the development of pressure sores.

Pressure ulcers can be difficult to prevent; however, if they occur there are a number of possible interventions that may help. The use of pressure relieving mattresses, specialist dressings, nutritional support and the institution of a programme for regular repositioning of the person may lessen the risk of pressure sores developing and also aid their treatment.

Infections:

Infections such as pneumonia, urinary tract infections and septicaemia are not uncommon in late stages of dementia and are the most frequent causes of death in the terminal stages of the disease.

Several factors contribute to the increased incidence of infections and their severity in people with advanced dementia. For example, the immune response may diminish with age and underlying medical conditions. Also, the presence of urinary or faecal incontinence may predispose to infection. Aspirating the contents of the stomach into the lungs, which can occur when the person is weak and immobile, may cause pneumonia. Bladder catheterisation, which may be unavoidable, can be another source of infection.

The person with Down Syndrome and Alzheimer's disease who has developed an infection may not present in the typical way. Even with pneumonia, the person may be unable to report their symptoms and may not present with a cough or temperature. The combination of all these factors emphasise the necessity for frequent medical and nursing assessment to detect signs and symptoms of an emerging infectious progress.

Many of the risk factors for the development of infection in this group of people are an inevitable consequence of the progression of the dementia. The persistence of these risk factors explains why it is not unusual for the person to relapse after the infection has been treated. The issue therefore arises whether the treatment of infections in terminal stage dementia is indicated. Although there are no agreed standard guidelines at present, research suggests that aggressive

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treatment of infections in people with terminal stage dementia does not influence the progression of the dementia or the mortality rate. It has been shown that palliative management of infections in people with terminal stage Alzheimer dementia is associated with a lower discomfort rate than aggressive medical treatment.

Feeding difficulties:

People affected with Alzheimer dementia and Down Syndrome are prone to developing feeding problems. In the early stages memory impairment, poor judgment and difficulty feeding themselves may complicate meal times. In the mid stages, agitation and wandering may prevent the person from settling at a table to eat their meal. By the late stage, the progressive degeneration of the brain results in the person being unable to recognise the smell and taste of food. This problem is further complicated by the physical inability to chew and swallow effectively. Carers and families commonly report that this is the most traumatic issue in caring for people with dementia. Despite these difficulties many people, up to the terminal stage of the disease, do not develop any clinical or laboratory signs of malnutrition when specialist clinical input is available.

It is possible for a person with advanced dementia to be fed orally under the guidance of a speech and language therapist. The therapist will assess the person and then advise carers and families regarding the techniques of skilful feeding and food preparation. The importance of a medical assessment should not be forgotten to rule out any medical or dental condition that could be exacerbating the feeding difficulties. The input of a dietician will ensure that the person receives adequate nutritional input and advice on supplemental feeds if necessary.

The use of tube feeding (nasogastric or PEG/Gastrostomy tubes) had previously been thought to be the answer to feeding difficulties in people in the advanced stages of Alzheimer dementia. Tube feeding was thought to prevent aspiration pneumonia, pressure sores, dehydration and malnutrition and even prolong life expectancy. Research and published data now question whether tube feeding is appropriate. What has become apparent is that tube feeding in people with advanced dementia can adversely affect their quality of life. Restraints may be required to prevent the tube being pulled out, and infections may develop at the site of tube entry while the risk of aspiration still remains. In addition, account should be taken of the fact that the person is being deprived of the normal social interaction and pleasure that surrounds mealtime.

Care planning considerations in the later stages of dementia

Service providers for people with intellectual disability strive to achieve optimal care for people with Down Syndrome and Alzheimer dementia. Dementia care is clearly an area for services that will benefit from an active multidisciplinary and multifaceted approach. This is especially true when considerations for care for the end of life must be discussed, decided and implemented. That dementia is a progressive, degenerative disease requires that end-of-life considerations be discussed and preferably early in the disease process.

With a proactive dementia screening policy in the

Daughters of Charity Residential Services we aim for an early diagnosis of Alzheimer dementia in the Down Syndrome population. Education of families and carers commences from the time of diagnosis and continues throughout the course of the disease. As the person approaches the late stages of dementia, a formal meeting is organised with the family and those involved with providing care to the person, in order to discuss agreed care plans for the terminal stages of the disease. At this meeting a full and frank discussion with the family regarding the usual clinical course of Alzheimer dementia in Down Syndrome takes place. Therapeutic and management options of the different medical conditions that may arise are discussed. Families are fully informed regarding the options of a palliative care approach or an aggressive medical and therapeutic management approach to the disease. Following this meeting, the agreed care plan is signed by the clinical team and family members. The care plan is kept in the person's chart for the purpose of informing all staff involved. The families and carers may revisit and revise this plan at any time. Such individualised care plans ensure that clear goals of care are not only identified but also delivered. From our experience to date it appears that families who have been fully informed regarding the progression of this relentless disease usually opt for a hospice care approach.

Conclusion

The provision of care to a person with Down Syndrome and Alzheimer dementia involves families, carers and health professionals. The difficulties that present are emotive for all involved. The availability of a clinical and nursing team who are experienced in this area of medicine can ensure that patients with Down Syndrome and Alzheimer dementia are given appropriate care based on best clinical practice. Through sharing of information and support for decisions made, care can be optimised to ensure the best quality of life possible.

Jean Lane, M.B., FRCPI

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