

# 7 Individual ageing of people with an early onset of disability

This section focuses on individual ageing of people with a disability and reviews the literature on several important issues:

- How does the nature and timing of individual ageing differ between the population at large and people with an early onset disability?
- What are the differences in mortality and life expectancy between the population in general and people with an early onset disability?
- What, if any, is the main difference between the ageing population in general and ageing people with an early onset disability in terms of age-related health conditions, including major illnesses and diseases?
- What are the trends in prevalence of congenital malformations relating to early onset disability?

## 7.1 Early start of individual ageing

Theories of aspects of biological ageing may be divided into three broad categories: genetic, non-genetic and physiological (Section 1.2). Genetic theories are based on the belief that genetic factors are associated with particular congenital disorders that affect the rate of ageing in certain individuals. These genetically based congenital disorders include progeria such as Cockayne's syndrome (premature ageing in very young children), Werner's syndrome (which refers to the onset of ageing in late adolescent years) and the premature ageing of persons with Down syndrome (Aiken 1989; Bullock 1992; Cristofalo 1988 and Perlmutter & Hall 1985, cited in Suttie 1995:11).

Non-genetic theories generally state that ageing occurs as a result of changes to cells rather than as part of genetic development. A common conception is that body parts simply wear out over time.

Physiological theories of ageing mainly relate to illness and disease in particular body organs and systems.

While the three broad categories of theories view the ageing process from different perspectives, it is generally agreed that biological ageing is characterised by increased risk of death with age, an increase in incidence of disease and progressive deterioration of the body, and changes in the ability of the body to adapt to environmental variations (Suttie 1995: 12).

It has been suggested that to identify the onset of ageing, three factors relating to life change should be assessed in addition to chronological age (Janicki et al. 1985: 291). These factors are:

- Increasing physical frailty and decreasing physical reserves mainly attributable to chronological age rather than trauma or illness;

- Diminishing levels of functional skills, particularly in areas of self-care, personal hygiene and other basic activities of daily living attributable to chronological age rather than to trauma or illness; and
- For less mentally impaired individuals, the self-perception of ageing and desire to seek age-appropriate or normative roles and activities.

A number of US studies have suggested that the beginning of ageing for people with a developmental disability occurs during the individual's mid- to late 40s or early to mid-50s. The criteria used to define 'ageing' status in these studies often include chronological age, clinical observations of changing functional status and the individuals' own expectations of changes in normative aged-related activities (Dickerson et al. 1979; Segal 1977; Seltzer et al. 1982; Thomae & Fryers 1982, cited in Janicki et al. 1985).

There is empirical evidence indicating that people with intellectual disability resulting from certain conditions do age earlier. Signs of premature ageing have consistently been reported in people with Down syndrome and intellectual disability resulting from other chromosomal causes (e.g. Janicki et al. 1985; Suttie 1995; Williams & Chad 1998). For people with intellectual disability associated with certain chromosomal disorders or conditions such as Down syndrome, ageing may begin in their 30s, 40s or 50s (Bigby 1998; Janicki et al. 1985).

There is considerable documentation of earlier onset and higher incidence of Alzheimer's disease in people with Down syndrome (e.g. Bigby 1998; Gatter 1996). There are also suggestions that people with severe physical disabilities such as spinal cord injury and brain injury begin ageing earlier than the general population, and that a range of health conditions worsen with increased duration of disability (Fethney 1998; Gething & Fethney 1998; Menter et al. 1993 cited in Gething et al. 1999; Crewe 1990).

International studies have found that dementia occurs at much higher rates among adult and older people with intellectual disability (not related to Down syndrome) than among the general population. Some people with an intellectual disability may acquire dementia relatively early in life, at age around 50 (Cooper 1997).

A review of published research indicates that a very significant number of people with spinal cord injury do begin to experience various problems as they age. Fatigue is the single most common problem, followed by a number of other physical problems such as bones that break easily and skin that breaks down more readily than normal. These developments are considered to be more related to duration of disability than to chronological age. People who were injured in their teens often begin to experience problems in their 30s and 40s, much earlier than their peers without disabilities (Trieschmann 1987, cited in Crewe 1990).

Significant proportions of ageing people with polio have been found to experience a range of changes in functioning, such as unaccustomed fatigue, breathing problems, weakness in previously unaffected muscles and pain. These changes are called 'post-polio syndrome' and the most common explanation is that there has been premature ageing caused by over-work in the motor neurones that survived the polio virus (Trieschmann 1987, cited in Crewe 1990). The decrease in energy and strength that results from these changes requires these people to reprioritise and even drastically reduce their activities (Crewe 1990).

A recent Australian survey of adults with cerebral palsy, known to agencies, reported that a majority of respondents considered that their physical condition was deteriorating, although cerebral palsy is not considered a progressive disorder. The survey results echoed findings of similar studies conducted in the United States and United Kingdom, that individuals with cerebral palsy experience negative changes in walking, digestion, bowel and bladder control, respiration, communication and swallowing. Depression, frustration, fatigue and anger were common among the people surveyed. In some cases, depression and anxiety

about ageing resulted in reduced social contact and affected independence and social interaction (Balandin & Morgan 1997).

Studies in the US and Canada showed that adults or ageing people with developmental or intellectual disability who had been living in long-term residential care had a lower level of functional ability and a higher rate of age-related decline than those living in the general community (Anderson 1989; Badry et al. 1989).

## **7.2 Decline in mortality and increase in life expectancy**

There is evidence to show that survival into older age is now a reality for some people with an intellectual disability, including some people with more severe disabilities (Eyman & Borthwick-Duffy 1994). However, international studies indicate that the mortality rate for people with an intellectual disability is greater than for the general population, especially for people living in institutions (Haveman et al. 1989; Haveman & Maaskant 1989). There are still large numbers of non-ambulatory individuals with severe or profound mental retardation whose life expectancies are very limited.

Until the 1950s, most studies showed that the majority of children with Down syndrome died before the age of 10 years, while in later studies at least half of them survived beyond the age of 30 years (Haveman & Maaskant 1989). A more recent birth cohort study of life expectancy for adults with Down syndrome was based on 1,610 affected individuals identified from over 1.5 million consecutive live births in British Columbia from 1908 to 1981. The study predicted that about 44.4% and 13.6% of live-born infants with Down syndrome would survive to age 60 and 68 years, respectively, as compared with 86.4% and 78.4% of the general population (Baird & Sadovnick 1988).

A 1990 United Kingdom study of trends in incidence and survival in Down syndrome also found that the number of young adults with Down syndrome was increasing, partly due to improvements in survival for those with congenital heart disease and decline in deaths from infection, and partly because of an increase of incidence in the 1960s (McGrother & Marshall 1990).

Studies conducted in the United States in the 1980s found that although the proportion of persons with mental retardation who live into old age was still lower than that for the population in general, the growth rate of this age group was higher than for other age groups with developmental disabilities (Anderson 1989:289).

With the exception of people with Down syndrome and certain other genetic conditions, and people with more severe disabilities, life expectancy and mortality rates for people with mental retardation in the United States are approaching those for the general population (Carter & Jancar 1983; Janicki 1986). A large proportion of adults with developmental disabilities now in their middle years are expected to survive into old age (Walz et al. 1986, cited in Anderson 1989: 290).

These changes are due to a number of factors, among which developments in medical technology, improvements in health care and social service programs and trends in community living are particularly important.

Over one-third of infants born with Down syndrome have congenital heart defects that are now repairable as a result of advances in medical technology. This may imply a potential further increase in the number of ageing clients with intellectual disability. However, some

people with Down syndrome who have benefited from early surgical treatment have not yet reached old age, so effects over time have not been fully documented (Suttie 1995).

### **7.3 Health conditions and major illnesses and diseases related to ageing: relationship to disability**

Among ageing people with an early onset disability, those with Down syndrome are more likely to have hearing and vision impairments, hypothyroidism, musculoskeletal problems and congenital heart disease. The prevalence of dementia of the Alzheimer type is particularly high in people with Down syndrome. It has been reported that neuropathological features of Alzheimer's disease are presented in all post-mortems of people with Down syndrome over 40 years of age, while clinical features may only be noted in a smaller percentage prior to death (Barcikowska et al. 1989 cited in Suttie 1995: 53).

It has been suggested that, excluding people with Down syndrome, people ageing with an intellectual disability do not differ significantly from the general ageing population in terms of the incidence of major illnesses and diseases attributable to biological ageing (e.g. heart disease, arthritis and higher blood pressure) (Suttie 1995).

However, a study of adults with intellectual disability aged 20–50 years in Sydney showed that these people have increased cardiovascular risk factors, more chronic diseases, and experience higher rates of morbidity and mortality compared with the general population. They also find it difficult to access health services because of low income and problems with communication and mobility (Beange et al. 1995).

Among the people reporting intellectual disability as their primary disabling condition in the 1993 ABS disability survey, 44% also reported associated physical impairments or disabilities and more than a quarter also reported speech problems. A high proportion (22%) of people reported associated psychiatric disabilities (Wen 1997).

A number of factors need to be considered when interpreting study findings (Suttie 1995). Firstly, there is a possibility of under-reporting of diseases and illness among people with intellectual disability because of their poor communication skills or possible insensitivity to pain and illnesses (Anderson 1993).

Secondly, for many types of illness and disease, comparisons between ageing people with intellectual disability and the general ageing population are not available.

Finally, studies have found considerable variations in factors affecting health status. For instance, people with intellectual disability may be exposed to additional risk factors such as non-mobility. Long-term institutional placement may affect health status through poor self-care. Additional complicating medical conditions could also affect health conditions (Eyman & Borthwick-Duffy 1994; Suttie 1995). Hence, further study is needed to properly understand differences in health status between people ageing with intellectual disability and the general ageing population.

It has also been pointed out that the variations in the health and functional status of the older population cannot be explained simply using the dichotomy of acute or lethal and chronic degenerative diseases. Nor can these variations necessarily be explained using the simple distinction between 'age-dependent' diseases (i.e. those diseases viewed as arising as a result of ageing processes) and 'age-related' diseases (diseases related to particular ages). This distinction may reflect more about our current level of knowledge of disease mechanisms than about disease processes. It is particularly difficult to apply this distinction to chronic and degenerative diseases. For instance, Alzheimer's disease, a disease process

usually characterised as 'age dependent', appears to have some genetic determinants and may be treatable and preventable (Manton 1990).

## 7.4 Trends in incidence and prevalence of congenital malformations

A review of epidemiological studies found that Down syndrome is the most common genetic cause of severe intellectual disability (IQ <50). Chromosomal etiologies (the majority of which are Down syndrome) were estimated to account for 20% to 40% of all cases of severe intellectual disability (Alberman 1978; McGrother & Marshall 1990; McLaren & Bryson 1987).<sup>6</sup> Therefore, changes in incidence and the prevalence of these conditions have direct implications for services.<sup>7</sup>

A study that looks at present estimates and future projections of the UK population with Down syndrome estimated an overall prevalence rate of 6.7 per 10,000, or 30,000 affected individuals. Results, based on a study population of over 7 million, gave no indication of a sizable reduction in the future Down syndrome population. Recent reductions in prevalence among the youngest age groups are likely to be explained by changes in the maternal age distribution for general population births together with a reduction in numbers of all births (Steele & Stratford 1995).

Prenatal diagnosis is another important factor affecting the incidence and prevalence of Down syndrome and other congenital malformations that may have resulted in early onset of disability. In 1992, Australian rates of several important congenital malformations, including Down syndrome and spina bifida, ranked in the top half of rates for developed countries (de Looper & Bhatia 1998: 35).

In Australia, incidence rates for Down syndrome in births remained relatively stable at around 12.8 per 10,000 births during the period 1987 to 1996, ranging between a high of 14.0 per 10,000 births in 1993 and a low of 11.9 per 10,000 births in 1987. The number of babies with Down syndrome surviving beyond the neonatal period (within 28 days of birth) increased from 249 in 1987 to 304 in 1990 and dropped to 246 in 1996. The reported number of induced abortions performed after prenatal diagnosis of trisomy 21 increased substantially during this period, reaching a peak of 130 in 1994. In the years 1987–1996, induced abortions accounted for 21.1% of all recorded notifications of Down syndrome, increasing from under 15% in 1987 to over 20% in the 1990s (Hurst et al. 1999: 82).

Incidence of spina bifida in Australia declined gradually from 7.1 per 10,000 births in 1987 to 3.0 per 10,000 births in 1994, increased to 3.5 in 1995, and then dropped again to 3.0 in 1996. Among 1,279 infants with spina bifida and for whom short-term outcome was known, 22.0% were stillborn and 19.8% of those born alive died during the neonatal period (Hurst et al. 1999: 38).

The 1998 ABS disability survey, unlike previous surveys, enables information on disabling condition to be related to responses to the survey screening questions (a series of questions

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<sup>6</sup> The higher estimates tend to come from studies that included non-survivors from the relevant populations (McLaren & Bryson 1987).

<sup>7</sup> An AHIW report critically reviewed the definitions and estimates of prevalence of intellectual disability with special reference to Australia and provided refined estimates of prevalence from national population surveys. The report also presented a preliminary analysis of patterns of intellectual disability in Australia (Wen 1997).

about specific impairments, activity limitations or restrictions). This additional information may allow analysis of the associations between a disability and a particular impairment or disease. However, the difficulties of attributing disability to particular impairments and diseases using cross-sectional survey data must be appreciated when interpreting the data. Also, the time between onset of illness and development of disability may vary depending on the nature of the disease and other factors (Campbell et al. 1994).