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LIFE EXPECTANCY AND MORTALITY IN INTELLECTUAL DISABILITY

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ABSTRACT

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This thesis comprises the first population based study of life expectancy and cause specific mortality of persons with intellectual disability (ID). It is based on a 35-year (1962-1997) follow-up study of a nation representative cohort of 2369 persons with ID between ages 2 and 97 years.

Persons were classified by sex, quinquinneum of their birth and the level of ID in 1962 for the analyses. Age and sex matched general Finnish population was used as a reference population in all studies. Life expectancy was calculated. Observed and expected deaths and cause-specific mortality ratios were calculated giving standardised mortality ratios. Patient documents of suicide victims were examined. Standardised cancer incidence and prevalence between 1967 and 1997 were calculated.

The life expectancy had a positive correlation with intelligence quotient. Profound ID was connected with 30% lost of life span throughout the life, but persons with mild ID had a similar life expectancy with the general population. The mortality rate was 17.7 per 1,000 person years (CI 95% 8.4-27.0). Suicide rate for women was 12.4 suicides and for men 15.5 per 100 000 persons, while corresponding figures for the general population were 13.2 and 52.9. Three most common causes of death were cardiovascular diseases, respiratory diseases and cancer for both sexes. There were 173 patients diagnosed with cancer; the expected number was 188 (SIR 0.9, CI 95% 0.8-1.0).

The life span of persons with ID has lengthened generally, but it is depended on the level of ID. The cause specific mortality of people with ID differs from the general population. The improved life expectancy can be interpreted as a result of improved health care and social support and normalisation.

Key-words: intellectual disability, mental retardation, life expectanc, mortality, cancer

TIIVISTELMÄ

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Tämä tutkimustyö on ensimmäinen kehitysvammaisten henkilöiden elinajan ennustetta ja kuolleisuutta koskeva väestöpohjainen tutkimus. Se perustuu 35 vuoden (1962-1997) kohortin seurantatutkimukseen, jossa on mukana 2369 henkilöä (ikäjakauma 2-97 vuotta).

Tutkimuskohortti on luokiteltu ikäluokan, sukupuolen ja kehitysvamma-asteen mukaan. Vertailuväestönä on suomalainen väestö, joka on vakioitu iän ja sukupuolen mukaan. Odotettavissa oleva elinajan ennuste on laskettu. Kuolleisuuden vakioitu ilmaantuvuus kaikissa kuolinsyyluokissa on laskettu havaittujen ja odotettujen kuolemien määristä. Itsemurhan tehneiden henkilöiden potilaspaperit on tutkittu. Vakioitu syöpäilmaantuvuus ja vallitsevuus on laskettu vuosille 1967-1997.

Kehitysvamma-aste on merkittävin elinajan ennusteeseen vaikuttava tekijä. Syvästi kehitysvammaisten henkilöiden elinajan ennuste on 30% lyhyempi kuin kokoväestössä, mutta lievästi kehitysvammaisten elinajan ennuste on sama kuin keskimäärin väestössä. Kuolleisuus 1000 henkilövuotta kohden oli tutkimuksessa 17.7 (CI 95% 8.4-27.0). Kehitysvammaiset naiset tekivät itsemurhan 12.4 ja miehet vastaavasti 15.5 100 000 henkilöä kohti (väestössä vastaavat luvut 13.2 ja 52.9). Kolme yleisintä kuolinsyytä olivat sydän- ja verenkiertoelinsairaudet, hengityselinten sairaudet ja syöpä. Tutkimuksen aikana ilmaantui 173 syöpää, kun odotusarvo oli 188 (SIR 0.9 CI 95% 0.8-1.0).

Kehitysvammaisten elinaika on pidentynyt, mutta riippuu voimakkaasti kehitysvamma-asteesta. Kuolinsyyt poikkeavat kuitenkin väestöstä. Pidentynyt elinaika ja muutokset kuolleisuudessa heijastelevat tutkimusajanjaksona tapahtunutta terveydenhuollon ja kehitysvammapalvelujen kehitystä.

Kehitysvammaisuus, elinajan ennuste, kuolleisuus, kuolinsyyt, syöpä

Contents

1 List of Orginal Communications

This thesis is based on the following papers, which are referred to in the text by Roman numerals (I to IV).

I Kristiina Patja, Matti Iivanainen, Hanna Oksanen, Hannu Vesala, Isto Ruoppila.: *Life Expectancy Of Persons With Intellectual Disability: A Follow-Up Study From 1962 To 1997*. Journal of Intellectual Disability Research 45, pp. 591-599, 2000

II Kristiina Patja, Pekka Mölsä, Matti Iivanainen: *Cause Specific Mortality in Intellectual Disability: A Follow-Up Study 1962-1997*. Journal of Intellectual Disability Research (in press)

III Patja Kristiina, Eero Pukkala, Matti Iivanainen: *Cancer Incidence of Persons with Intellectual Disability*. Journal of Intellectual Disability Research (in press)

IV Kristiina Patja, Seija Raitasuo, Matti Iivanainen, Jouko Lönnqvist: *Suicides Of Persons with Intellectual Disability in Finland: a 35- Year Follow-Up Study*. Acta Psychiatrica Scandinavica (in press)

These results have been reprinted with kind permission of the copyright holders. The results have been presented in the 11th World Congress of International Association for Scientific Research of Intellectual Disability in August 2000. In addition previously unpublished results on Down syndrome and epilepsy are presented.

2 Abbreviations

CI 95%	95% confidence interval
CP	Cerebral palsy
DS	Down syndrome
E	Expected number of cases
GERD	Gastro-oesophageal reflux
ICD-8	International Classification of Diseases, eighth revision
ICD-9	International Classification of Diseases, ninth revision
ICD-10	International Statistical Classification of Diseases and Related Health Problems, tenth revision
ID	Intellectual disability
IQ	Intelligence quotient
O	Observed number of cases
OR	Odds ratio
RR	Risk ratio
SD	Standard deviation
SE	Standard error
SIR	Standardised incidence rate
SMR	Standardised mortality ratio

3 Introduction

Intellectual disability (ID), characterised by significantly subaverage intellectual functioning, is a common phenomenon in our society, affecting approximately 1% of the population, that is about 50 000 persons in Finland. However, it has an impact on much larger population through families, the educational system and various professionals. ID has multiple dimensions, from its emotional burden and ethical considerations to the economic hardships imposed, all which are in a continual state of flux, raising questions as to what influence healthcare has on the lives of people with ID.

Life expectancy and mortality provide a rough estimate of the quality and efficiency of the healthcare system. During the last four decades, several studies have reported an increased life expectancy in the ID population and in particular in young age groups, with a simultaneous decrease in mortality and indicated a change in cause-specific mortality compared to previous decades. It appears that the number of persons with ID is increasing due to ageing, but we neither know by how much nor over what time frame. Only scattered studies exist on selected cohorts from institutions or from registers of people with ID. Different kinds of data problems, the lack of common methods and definitions have therefore complicated international comparisons of life expectancy and mortality figures.

This study is based on a large, nationwide cross-sectional cohort of persons with ID in Finland. As far as the author could ascertain, it is the first published population based follow-up study on ID with a lifelong follow up period. We have examined the life expectancy and mortality with population-based methods, which offers several advantages. First, the general population has provided a reliable comparison population, allowing us to standardise the different qualities of the ID population. Secondly, the results describe the overall situation of the entire country, which is the main public health interest. Finally, many biases, such as selective inclusion, use of non-standardised methods and problems of comparability, are avoided. However, the material and methods used in this study are not unproblematic. Definition of ID is still under debate and taxonomy has changed during the follow-up period and together they influence the inclusion criteria. In

addition, the study is based on mortality statistics, which has its problems with coding of the causes of death.

This study combines life expectancy and cause specific mortality, since they are inter-dependent with a few special clinical features. Prolonging life expectancy is likely to change cause-specific mortality patterns, and correspondingly, the changes in the patterns will either improve or impair life expectancy. These results contain accurate information on the current number of adult persons with ID and estimates on the future number of ageing persons with, as well as changes in the mortality patterns. The information will benefit not only health professionals, but also service providers, in addition to persons with ID and their families.

4 Review of literature

This section provides an overview of the literature on life expectancy and mortality of persons with ID. It includes a short historical description on the prevalence of ID. The conditions and concepts covering ID, which have been defined differently across the decades, are reviewed. There are only few population-based studies of ID, and most studies are based on institutional patients carried out by non-comparable methods. This must be taken into account when comparing the results achieved in different studies. While drawing firm conclusions from the various findings of epidemiological research in the ID field is difficult, general information is needed in order to improve the quality and quantity of research in this field.

4.1 Concept of intellectual disability

Part of the history of ID has been the search for a destigmatising terminology (Reid 1997). Historically, ID has carried multiple names, such as mental retardation, mental deficiency, mental handicap, mental subnormality, severe subnormality or oligophrenia, mental enfeeblement, insanity, and different levels of ID have been feeble-minded, imbecile, idiotia and moron (Gath 1992; King et al 1997; Williams 1996). In the literature, the following terms are used: mental retardation (emphasising the neurological dysfunction), learning disability (emphasising the difficulties in learning, widely used nowadays in British literature) and intellectual disability (emphasising the primary dysfunction, widely used in the United States). In this study, we prefer the term intellectual disability, which focuses on the aetiological factors leading to cognitive, adaptive and behavioural manifestations. The term was introduced in the Journal Mental Deficiency Research in 1992 (Frazer 1991) and after that used widely by associates of International Association of Scientific Study of Intellectual Disability (IASSID) in 1995 (Trevor Parmenter, the president of IASSID, personal communication).

Prior to the invention testing of IQ by Binet and Simon (Binet and Simon 1916), ID was defined with terms of the development of social and

vocational competence (Jacobson and Mulick 1996), but also with morphological qualification and quantification (Williams 1996). As the phenomenon became more measurable, the debate on the theory of the measures had begun (Detterman 1987). Attempts to achieve a comprehensive and consistent classification have been yet unsuccessful: taxonomy has changed continuously, classifications have remained unlinear and definitions have been understood with inconsistency (Detterman 1987; Williams 1996). The intellectual development of persons with ID has been reported to continue throughout the adult life (Eyman and Widaman 1987; Trower and Nicol 1996), but the IQ measures have been found rather stable with some increase for persons with mild and moderate ID (Fisher and Zeaman 1970), but rarely in severe and profound ID (Goodman 1976).

Mental retardation (MR), term used by the WHO definitions as synonym to ID, refers to substantial limitations in present functioning. It is characterised by significantly subaverage intellectual functioning, which exists concurrently with related limitations in two or more of the following applicable adaptive skill areas: communication, self-care, home living, social skills, community use, self-direction, health and safety, functional academics, leisure and work. By definition MR manifests before age 18. It is evaluated with psychological testing (IQ < 70) and adaptive behaviour (WHO 1995). Environmental aspects need to be considered as some individuals may behave normally in one environment and subnormal in another.

The WHO has divided ID into three basic areas: impairment, disability and handicap. Impairment is any loss or abnormality of psychological, physiological, or anatomical structure of

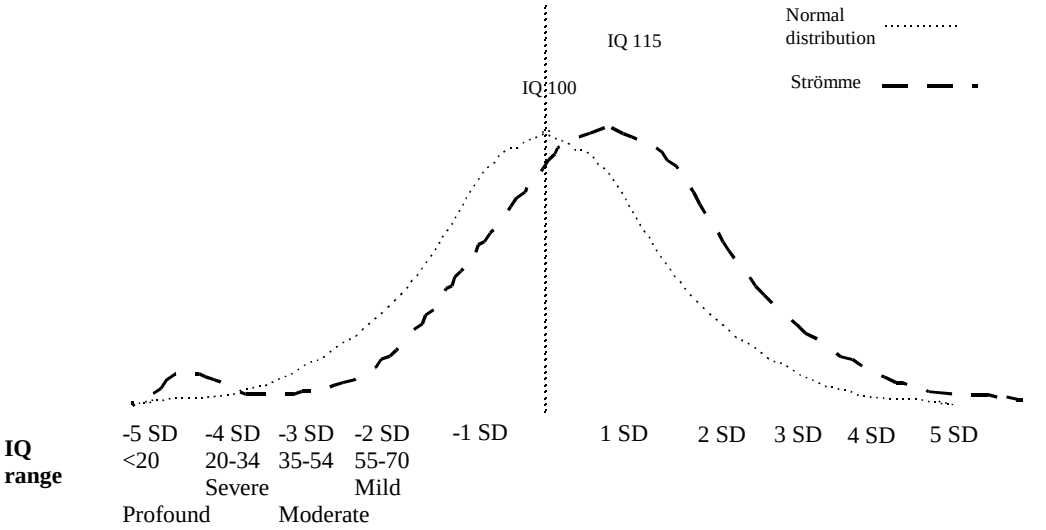

Figure 1. **Traditional normal distribution of intelligence suggested first by Binet 1916 and a new suggestion for distribution of intelligence by Strömme et al 1992.**

function. A disability is any restriction or lack (resulting from impairment) of ability to perform an activity in the manner or within the range considered normal for a human being. A handicap is a disadvantage for a given individual, resulting from an impairment or disability, that limits or prevents the fulfilment of a role that is normal (depending on an age, sex, social and cultural factors) for that individual (WHO 1980).

The degree of ID has been divided by IQ scores (Figure 1). The borderline intellectual functioning is defined with IQ 71-84 in the cutting point of -1 SD in the IQ deviation and normal intelligence as IQ over 84. There are though suggestions (Figure 1), that the cut point would be in 115, and an increase in profound ID due to the progressive neurological diseases (Strömme et al 1992).

4.1.1 Definitions of ID in the present study

In this study the WHO definition and classification of ID were used (WHO 1980; WHO 1995). The ID classes are six: normal intelligence, borderline intellectual functioning, mild ID, moderate ID, severe ID and profound ID (WHO 1980; APA 1994; WHO 1995). In medicine, in the clinical use the ICD-9, the term mental retardation (as synonym of ID) is used both in the heading and in the definitions of different levels (WHO 1975). ID is classified according to IQ as follows: 3170 (mild IQ 50-70), 3180 (moderate IQ 49-35), 3181 (severe IQ 34-20), 3190 (profound IQ under 20). The borderline intellectual function is excluded from this section, but it can be found in the section of V-diagnoses (APA 1994). In the latest ICD-10 (WHO 1995) mental retardation term is used with the definition of "a condition of arrested or incomplete development of the mind, which is especially characterised with impairment of skills manifested during the developmental period, which contribute to the overall level of intelligence i.e. cognitive, language, motor and social abilities" (WHO 1995). The level of ID is coded with similarly to ICD-9. In the present study, we are using the division of levels of ID of ICD-9 and define the ID with WHO 1980 and 1995 definitions and DSM-IV.

4.2. Care for persons with intellectual disability in Finland

Interest in ID in Finland began at the end of the 19th century, at the same time as a general interest in social well being was roused. The first committee on the welfare of persons with ID was set up in 1890 based upon the proposal of Bishop C.H. Alopaeus (Tarvainen 1966). The first school for children with hearing impairment or ID operated between 1877 and 1892 in Pietarsaari. The first institution for persons with ID was established in 1907 in Sortavala (nowadays in Russia). Legislation did not secure the rights of people with ID until 1927, when the first larger institutions were established. However the interest in ID was modest. More interest in ID began in the post-war period as the social policy quickly expanded to include people with ID, with institutionalisation following in the 1960s.

In the 1970s, the social and health care system started to change towards a more customer-based system, and the institutional population started to decrease. A goal for the service system was the realisation of the concept of normalisation (Wolfensberger 1972). Persons with ID were entitled to special services by law in 1977. Further, in 1995 the new constitution of Finland emphasised that discrimination on the basis of illness or disability on all levels of society was forbidden. This has diversified the participation of persons with ID in the communities, in spite of the simultaneous severe economic depression in Finland at the beginning of the 1990s. These changes have undoubtedly had an effect on life expectancy and mortality figures, which can be seen as outcome measures of the service system and society in general (Härö 1995).

4.3 Prevalence and incidence of intellectual disability

The prevalence of a phenomenon, as low intelligence quotient (IQ) in this study, has a great influence on all studies on it. If we assume that IQ is a normally distributed continuous variable, with ID defined as an IQ under 70, then 2.3% of population have ID. Its incidence has been estimated at 1% of new-borns and its cumulative incidence rates by 8 years of age at 9.1 and 8.3 per thousand for boys and girls, respectively (Katusic et al 1995). The prevalence of ID is even higher, in spite of the higher mortality rates for children in this population (Trower and Nicol 1996; Staruss et al 1998a). In developed countries,

Table 1. Studies of prevalence of ID per 1000 from Scandinavian countries 1936-2000 (Brask 1972; Åkesson 1967; Wallin 1975; Granat and Granat 1973; Bernsen 1976; Kääriäinen 1987; Rantakallio and von Wendt 1986; Hagberg et al.1987; Strömme 2000b).

Authoro	Year of the study	Country	Age	Profound ID (IQ <50)	Mild ID (50<IQ<70)
Brask	1963	Denmark	5-14	3.3	-
Åkesson	1964	Sweden	0-20	4.4 (IQ<52)	-
Wallin	1969	Sweden	5-19	4.2	-
Granat	1970	Sweden	20[1]	-	18.3
Bernsen	1976	Denmark	5-14	3.9	-
Kääriäinen	1978-81	Finland	7-9	6.3	7.5
Rantakallio	1980-81	Finland	14[2]	6.3	5.6
Hagberg	1984	Sweden	14-18[2]	3.3	3.9
Strömme	1992-97	Norway	12-15[2]	2.7	3.5

[1]Examination for military service
[2]Birth cohort study

prevalence has been estimated at 1% to 3% (Hodapp and Dykens 1996). In Scandinavia variation has been around 1% (Table 1). Finding all persons presents a challenge (Munro 1986). Since definitions and methods vary, some regional and temporal fluctuations exist in the number of persons reported to have ID (McDonald and MacKay 1996).

The most reliable figures are on the prevalence of persons with profound ID, which has been reported to vary from 3.4 to 6.3 per thousand (Kraus 1973; Abramowicz and Richardson 1975; Hagberg 1978; Gustavson et al 1977; McQueen et al 1987; Dupont 1989; Mallon et al 1991; McDonald and MacKay 1996; Rantakallio and von Wendt 1986; Roeleveld et al 1997; Fernell 1998). In contrast, for mild ID, the prevalence range is broader varying from 2 to 79.3 per thousand in developed countries (Hagberg et al 1981; Rantakallio and von Wendt 1986; Katusic et al 1995; McDonald and MacKay 1996; Roeleveld et al 1997). Roeleveld in 1997 encapsulates the problems of prevalence in his large review, when he states, that the calculation of a reliable average mild ID prevalence rate is virtually impossible. The identification of person with ID is not only based on psychological, physiological, social or economical factors, but a combination of these varying also by time and location. Therefore any criteria for ID is incomplete in some area and search for "true" prevalence or incidence is self-fulfilling process (Fryers 1993).

Over the first five decades of this century, ID was associated with extremely high mortality. For instance in two series of children with Down syndrome (DS) driven from service records in Great-Britain less than 40% of children survived beyond 5 years (Record and Smith 1955; Carter 1958). In developed countries, prevalence has risen this century due to the development of social support systems and medicine. The differences in prevalence rates probably reflect the improved standards of living and the improved health care especially in ante-, peri- and postnatal period (Diaz-Fernandez 1988; Louhiala 1995). We are also better equipped to deal with associated sequelae of ID (Crow and Tolmie 1998). For the same reasons, life expectancy is lengthening, and although less children are born with ID, prevalence of ID is increasing (Dupont et al 1987; Wolf and Wright 1987; Diaz-Fernandez 1988).

The first unpublished estimate of ID prevalence in Finland was made in 1906-07, and was reported to be 3.13 per thousand. A 1932 estimate yielded a prevalence of 4.43 per thousand and in 1939-40, the same figure was reported on the basis of the statistics from the newly established National Insurance Institution (Kaila 1942), but later in 1940s, the figure rose to 4.7 per thousand (Kaila 1943; Kaila 1944). In 1958, the prevalence of children under 16 was reported to be 3.03 per thousand (Yliruokanen 1959). In other countries, the prevalence had been found to be higher, which produced the new population-based sample in 1962 (Tarvainen 1966). From this sample, the prevalence of ID was calculated to be 6.62 per thousand (Amnell et al 1964). At present, the prevalence of ID in Finland is estimated at 1% (Sillanpää 1996).

4.4 Life expectancy

The length of a person's life is unpredictable, however, by studying populations it is possible to estimate life span or life expectancy for groups of people. Life expectancy is usually defined as the remaining lifetime in years for a person who has survived from the beginning of indicated time interval to the time of survey (Parkin and Hakulinen 1991). In survival analysis, the life-expectancy estimate is commonly set when 50% of a specified population has died. Life expectancy provides information on identification of risk factors for both scientific and practical reasons. In practice, service providers, families and professionals need estimates of life expectancy for different subgroups of persons with ID. There are only two study populations of ID (by Dupont in Denmark and from Eyman in California), which have been studied for life expectancy in its full definition.

Persons with ID are likely to have a shorter life expectancy than the general population. Historically, the survival of persons with ID has been of interest since only rarely did they reach adulthood (Richards and Sylvester 1969); in the 1960s, for instance, the life expectancy of a person with ID was 18.3 years (Collmann and Stoller 1963). Studies have repeatedly shown the inverse relation between the severity of ID and longevity, but they have used very heterogeneous populations and methods (Balakrishnan and Wolf 1976; Baird and Sadovnick 1987; Eyman et al 1990; Crichton et al 1995). In Denmark, Dupont found in her population-based study, that persons with mild ID have a life expectancy that is 10 years less than the overall Danish population (Dupont et al 1987). Nevertheless, life expectancy has been prolonged in all western societies, (Carter and Jancar 1983; Fryers 1986) and longevity has increased for all groups of persons with ID. This is particularly apparent in DS, where life expectancy has increased from 9 years in 1929 to 56 years in 1980s (Carter 1958; Collmann and Stoller 1963; Gallagher and Lowry 1975; Thase 1982; Fryers 1986; Baird and Sadovnick 1987; Devenny et al 1996; Steele and Stratford 1995).

Persons with more severe ID still have poor life expectancies, although low intelligence per se does not necessarily mean a shorter life. Underlying progressive disease, secondary handicaps and cco-morbidity present additional risk factors associated with non-mobility and lack of self-help skills (Kaveggia 1985; Eyman et al 1990; Eyman et al 1993a; Kastner et al 1994; Plioplys et al 1998). Epilepsy and cerebral palsy are the most common associated disorders reducing life expectancy (Coulter 1993; Crichton et al 1995; Jancar et al 1996). In the past only 35% of persons with profound ID survived into adulthood. However, today, two large studies based on service records in United States, have reported, that 70% persons with profound ID survive into adulthood (Eyman et al 1993b; Plioplys et al 1998).

4.5 Mortality

Mortality studies provide predictors for life expectancy. Mortality and survival are associated with a number of factors including aetiology of ID, age, level of retardation, associated disorders and self-help skills. Neurodegenerative and chromosomal diseases in aetiology of ID may be progressive, thus substantially

increasing mortality over time. Eyman et al have suggested the self-help skills are the most powerful predictors of mortality (Eyman et al 1990). Their study is based on the service registers and the majority of the population has profound or severe ID. Hence, the low self-help skills are more likely to be a consequence of aetiology of ID or associated disorders, rather than the level of ID or it being an independent risk factor, and are the outcome of neurological damage. Therefore self-help skills should not be used without information of aetiology of ID.

Studies of mortality of persons with ID are presented in table 2. It shows year of study, population and source of information, if these were given in the original article. Most studies report only overall percentages and only one study gives the risk ratios compared with the general population. Comparisons between studies are difficult, since they all use diverse methods and there is one population based study of children with ID (Similä et al 1986). Most studies are carried in institutions (eighth of 12 studies). Institutional population is more profoundly disabled and therefore respiratory mortality is most likely higher there than in cohorts with persons with mild ID living independently. Cardiac diseases show low prevalence for similar reasons. Non of the previous studies can be easily compared with the present study, because of different age range, different sampling and methodology.

The level and aetiology of ID are most frequently used mortality predictors (Balakrishnan and Wolf 1976; Forssman and Åkesson 1970; Carter and Jancar 1983; Eyman et al 1986; Dupont et al 1987; McGuigan et al 1995; Strauss et al 1998a; Conroy and Adler 1998). Because there are no studies presenting interactions between these mortality variables, they are not easily comparable. The relative impacts of development in medicine and society are also defined with difficulty. The multiple changes in the environments of the general population and this subpopulation increase the challenge in making comparisons. This is especially evident in DS, where the increase in life expectancy is from nine years to 56 years. This is likely the result of a combination of improved health care, social support and the heart surgery introduced in the 1970s to correct heart malformations, which has decreased mortality markedly (Thase 1982). Still the mortality is high DS beyond the age of 50. Virtually all adults with DS by the age of 35 to 40 years have neuropathologic hallmarks of Alzheimers´s disease. This is probably caused by an excessive production of [beta]-amyloid from a triplication of the [beta]-amyloid precursor gene located on chromosome 21 (Wisniewski et al 1985; Tanzi et al 1987; Lai 1999). This was only discovered, as there were aged persons with DS (Devenny et al 1996). The longer life span is expected to produce new morbidity and mortality predictors for this population.

Age is an important predictor of mortality. Children with ID have higher mortality rates than their age mates, (Similä et al 1986; Boyle et al 1994) with a marked decline in the adulthood population until the age of 50, when the mortality rate becomes comparable to the age group over 60 in the general population (Haveman and Maaskant 1989). It has been suggested that, age 50 might be a turning-point according age and mortality risk in the adult ID population.

Table 2. Studies of mortality of persons with intellectual disability in developed countries from 1930 to 1995.

Authors	Years Country *cause of death*	n of persons at risk	n of deaths *rate per 1000 person years*	Source of information	Method	Age	Level of ID	Causes of death (percentage of all deaths)
O'Brien and Zaharia 1998	1993-1995 U.S.	6,812	293 *16.57,*	Service Register	Cox, Poisson	0-99	All levels, by level also	Not mentioned
Strauss et al 1998b	1981-1995 U.S. *External causes*	733,705 person years	520 *7.08*	Service Register	SMR	15-59	From profound to moderate	Drowning (SMR 6.22), Fire (SMR 5.02), Falls (SMR 4.03), Suicide (0.31)
Strauss et al 1998a	1993-1995 U.S.	1,878	87	Institution and community	Descriptive	2-67	-	Respiratory diseases (36.7), Diseases of digestive system 13.8), Cardiovascular diseases (13.8), Epilepsy (12.6), Cancer (4.6)
Raitasuo et al 1997	1972-1993 Finland	not available	216	Institution	SMR	0-90	All levels, not reported by level	Diseases of nervous system (27), Disturbances of mental health (17), Vascular diseases (7), Cancer (6)
McGuigan et al 1995	1982-1990 Great Britain	1756	270	Service register	SMR, Kaplan-Meier survival curves	-	All levels, not reported by level	not mentioned
Cole et al 1994	1983-1991 Great Britain autopsies	88	49	Institution,	Descriptive	12-85	Not reported	Respiratory diseases (46.9), Cardiovascular diseases (28.6), Cancer (10.2), Diseases of digestive system (8.1)
McLoughlin 1988	1983-1987 Great Britain	not available	92 *23.1*	Institution	Age-specific death rates	-	All levels, not reported by level	Respiratory diseases (41.5), Cardiovascular diseases (24.2), Cancer (10.7), Diseases of digestive system (9.9), Neurological disorder (9.2)
Similä et al 1986	1966-1980 Finland	165	26 *157.6*	Birth cohort study	Death rates	0-17	All levels, not reported by level	Congenital abnormalities (53.8), Infectious diseases (26.9), Other causes (11.5), Injuries (7.7)
Jancar et al 1984	1966-1981 Great Britain *cancer*	not available	313 out of 496 deaths	Institution	Description	40-70	Not reported	Cancer (20.1), Myocardial infarct (8.9)
Carter and Jancar 1983	1930-1980 Great Britain		1,383		Annual death rates, average ages at death	-	All levels, not reported by level	Respiratory diseases (45.9), Tuberculosis (17.1). Sudden deaths (several ICD categories)
Richards and Siddiqui 1980	1968-1978 Great Britain	18,975	465	Institution	Annual death rates, average ages at death	5-89	All levels, not reported by level	Respiratory diseases
Chaney et al 1979	1944-1975 U.S. *Respiratory mortality*	not available	1,005	Institution	Cause-specific death ratio	0-30+	Age-sex and IQ matched groups	Respiratory diseases (profound ID 75 and other levels 38)
Polednak 1975	1958-1973 Canada	not available	707	Institution	Age-specific death rates	0-60+	Not reported	Respiratory diseases (47.7), Cardiovascular diseases (11.7), Diseases of nervous system (9.0), Accidents (8.1), Cancer (1.8)
Richards and Sylvester 1969	1929-1968 Great Britain	not available	1,196	Institution	Annual death rates, average ages at death	0-99	All levels, not reported by level	Respiratory diseases (25.8), Cardiovascular diseases, Neurological disorder

Another groups of mortality risk factors across the age range are associated disorders and chronic medical conditions. High mortality has been found with cerebral palsy (Hagberg et al 1989; Crichton et al 1995), epilepsy (Forsgren et al 1996) and congenital malformations (Chaney et al 1985; Frid et al 1999). People with ID have more chronic medical conditions and use more often regular medication, than the general population (Hand 1994; Beange et al 1995; van Schrojenstein Lantman-de Valk et al 1997). Chronic medical conditions, particularly if untreated, increase mortality, (Kapell et al 1998) as do some medications such as anti-epileptic and antipsychotic drugs, both of which have side-effects including drowsiness, constipation (Van Winckel et al 1999) and increased risk of infections (Coulter 1993; Forsgren et al 1996) and neurological side effects like tardive dyskinesia (Ko et al 1992). Nevertheless, untreated depression and undertreated epilepsy are common in this population (Göstason 1985; Raitasuo et al 1999a).

4.6 Cause specific mortality

Cause-specific mortality rates in people with ID have been reported in limited numbers and great variation in populations and methods and comparing them with the present study can be misleading (Table 2). It seems like in this study the prevalence of respiratory diseases is too low, but it is merely because we use population based sample with large age range with all levels of ID rather than having institutional population with profound ID or narrow age range. Obviously there is a high risk of respiratory disease in this population, which is associated with a low level of ID for all age groups (Chaney et al 1979). Respiratory function of persons with profound ID is more likely to be disturbed by gastro-oesophageal diseases (Kuruvilla and Trewby 1989), dysmorphias of the oral cavity (Gabre et al 1999), congenital heart diseases (Chaney et al 1985) and immunological deficits (Ugazio et al 1990) predisposing them to respiratory infections. An institutional setting may also increase the risk of infections (Schupf et al 1995).

In previous studies, prevalence rates of adult cardiac disease have been reported to be lower than in the general population (Carter and Jancar 1983; Chaney et al 1985; Eyman et al 1986; McLoughlin 1988; Raitasuo et al 1997). In the age group of 50 and older, only 14 to 26% is reported to suffer from cardiac disease (Janicki and Jacobson 1986; Hand 1994). It has been suggested that minimal use of alcohol (Clarke and Wilson 1999) and cigarettes (6.8-15.8%) (Minihan 1999) reduce cardiac mortality markedly (Beange et al 1995). However, children with ID do have more cardiac diseases because of more frequent cardiac malformations (Grech and Gatt 1999).

Diseases of the digestive system (Jancar and Speller 1994; van Schrojenstein Lantman- de Valk et al 1997) and neoplasms (Jancar 1990) have been reported with higher frequency in the institutional ID population than in the general population (Table 2). Persons with ID suffer more often from constipation (Van Winckel et al 1999) and chronic regurgitation (Rogers et al 1992), which increase the risks of

intestinal obstruction and peptic ulcer, respectively. High-risk groups for both diseases are severe ID, immobility, tube feeding and scoliosis (Jancar and Speller 1994). Of persons with severe ID living in institutions, 9.2% persistently eat non-nutritive substances (McAlpine and Singh 1986). Intestinal obstructions were found to be more common in this population, accounting for up to 2.4% of all causes in one study (Jancar and Speller 1994). These obstructions were associated with low intellectual capacity, cerebral palsy, epilepsy and psychiatric disorders, which may reflect the slow response of the autonomic nervous system and the side effects of some medications (Cole et al 1994).

There is only one report of external causes of death (violent causes, accidents, suicides etc.) by Strauss in 1998 (Table 2). They found lower external mortality of persons with ID than in the general population, although adult persons with ID did have an increased risk of falls, pedestrian accidents, drowning and fire accidents (Strauss et al 1998b). Male gender carried an increased risk for an external death (Strauss et al 1998b), but a lower risk for suicides, homicide or poisonings. Occupational accidents and traffic accidents are common in the general population, however as no studies exist for their frequency in the ID population, they are presumably rare.

4.7 Cancer

The incidence of cancer in people with ID is uncertain. There are many studies based on death rates reporting the prevalence of different types of cancer (Jancar and Jancar 1977; Achterberg et al 1978; Jancar et al 1984; Uno 1996) and case reports on various neoplasms (Jancar and Mlele 1985; Kamidono et al 1985; Miki et al 1999; Satge et al 1997). In addition, there are several reports on incidence of cancer for children with ID (Li et al 1984; Robinson et al 1984; Windham et al 1985; Mili et al 1993a; Mili et al 1993b; Ribeiro et al 1993; Mertens et al 1998). Many syndromes leading to ID have also been reported in conjunction with increased cancer incidence, e.g. (Stoller et al 1973; Scholl et al 1982; Robinson et al 1984; Braun et al 1985; Sasagawa et al 1986; Franceschi et al 1991; Korenberg et al 1992; Satge et al 1998), Cowden syndrome (Hanssen and Fryns 1995), Fragile X (Fulchignoni-Lataud et al 1997; Phelan et al 1988), Prader-Willi and Angelman syndromes (Nichollis et al 1998) and, most recently, tuberous sclerosis (Crino and Henske 1999).

In most studies, over all cancer prevalence has been reported to be lower than in the general population varying, from 4.6% to 17.5% (Table 2) in different types of ID populations versus 20% in the general population (Carter and Jancar 1983; Jancar 1990; Cooke 1997). There are, however, a few types of cancer with higher incidence rates, such as oesophageal cancer in profound ID (Bohmer et al 1997). Deaths due to gastrointestinal neoplasms have been reported with a three-fold increase compared with the general population (Bohmer et al 1997; Cooke 1997). Some types of cancer have been reported with high prevalence, such as neoplasms of ventriculum, oesophagus, rectum and colon (Jancar 1990). Lung cancer shows a low prevalence (Jancar 1990). We know, for instance, that women with ID have many special features, which may influence cancer incidence, including

low sexual activity, (Calson and Wilson 1996) chemical castration, (Calson and Wilson 1996) and, short menstrual life, (Schupf et al 1997) and a low rate of sexually transmitted diseases and poor hygiene (Wingfield et al 1994).

There are groups of children and adolescents with ID with a high risk of developing neoplasms. Children with DS, congenital heart defect or digestive abnormality are found to be vulnerable to leukaemia in large population studies (Mili et al 1993a; Mili et al 1993b; Mertens et al 1998), but ID generally has not been reported as an independent risk of cancer. There are case reports with similar findings of the incidence of male reproductive cancers for person with ID, the incidence being highest in the teens (Dexeus et al 1988; Dieckmann et al 1997) and in syndromes with genetic background (Miki 1999; Cooper 1993; Al-Saleem et al 1998; Satge et al 1997; Kamidono 1985).

4.8 Mental disorders and suicides

The brain damage causing ID is often associated with reduced frustration tolerance and increased explosively, leading to behavioural problems and anxiety (Reid et al 1984). Conduct disorders and self-injuries (King 1993) are more common than in the general population (Göstason 1985; Lund 1985). It has been suggested that ID increases the risk of mental disorder, with the overall prevalence rates varying from 10% to 60% depending on the patient sample (King et al 1997). Affective disorders are less common than in the general population, varying from 1.7% to 8.9% (Lund 1985), as compared with 4.1% for major depressive episode, 1.7% for current dysthymia and 17% depressive mood in the general population (Isometsä et al 1997). However these disorders may simply be underdiagnosed among persons with ID. Symptoms of depression are often expressed in the form of somatic complaints, vegetative symptoms, or regression, making diagnosis of depression difficult (Raitasuo et al 1999b). Depression is a major risk factor for suicide in the general population.

In the general population, high suicide rates are associated with mental disorders, physical illness and social disintegration (Appleby et al 1999). Persons with ID form a special subpopulation with more physical illnesses, lower coping capacity and greater dependence on social support. The prevalence of mental disorders among the ID population is higher than in the general population (Lund 1985). Abuse increases the risk of suicide in the general population (Birhamer et al 1996). Persons with ID are particularly vulnerable to abuse, which can be encountered in public and private residences (Furey 1994). While the accumulative effect of these factors would lead to the assumption that people with ID have increased risk of suicide, no large-scale studies have reported on suicide rates of persons with ID (Harris and Barraclough 1998; Strauss et al 1998b). There are only a few reports on attempted and successful suicides, dating from 1899 (Carier 1899; Gammage 1902; Bellmann 1919/1920; Reynés 1920; Fribourg-Blanc and Scouras 1931; Menolascino et al 1989; Walters 1990; Hurley DesNoyers 1998). Suicides are regarded as a rarity among people with ID.

4.9 Health risks of persons with intellectual disability

The risk factor profile of ID population includes different groups of people with similar risks, but the additional risk of ID itself can be difficult to calculate over a time-period. Many syndromes carry an additional genetic risk of early death. DS is the largest group among the genetic syndromes and, as such, has the most information available on genetic risks. There are several abnormalities reported in cell differentiation in DS such as transient abnormal myelopoesis (Baschat et al 1998) and altered expression of genes (Chen and Antonarakis 1997). These are related to immunological deficiency leading to a vulnerability to infections (Thase 1982; Ugazio et al 1990) and to neoplasms of the immune system (Ford and Hanawalt 1997), especially to non-Hodgkin lymphomas, which are connected with acquired and congenital immune deficiencies as well as autoimmune disorders (Hardell et al 1998).

The population with ID differs from the general population by age- and sex-structure. The age structure of the former is weighted to persons less than 40 years of age, although life expectancy is steadily increasing (Patja et al 2000). There are more new born males than females with ID (1.3 versus 1.1 per thousand) (Crow and Tolmie 1998), but at lower levels of ID more females are born, especially with genetic disorders, where sex ratios are skewed at conception, or become so during embryonic development through differential intrauterine selection. For instance, the foetal male:female ratio estimate was 0.88 for trisomy 13, 0.90 for trisomy 18, and 1.16 for trisomy 21 (Huether et al 1996). This overall sex ratio difference predominates 25 years longer in the ID population than in the general population (Härö 1995; Patja et al 2000). The level of ID has a great effect on the age structure, and the health status of adults in general is better than in early years as the mortality mainly affects on children (Krauss and Seltzer 1986).

More than half of the persons with ID has an associated disorder such as epilepsy, CP or sensory impairment. Two thirds of adult persons with ID use regular medication (Hand 1994), of which antiepileptics and psychotropics are most common. For instance 25% of persons with ID have epilepsy in population based samples (Coulter 1993; Hand 1994; Forsgren et al 1996). Musculoskeletal diseases affect 13-54% of people with ID and every fourth person is seriously disabled (Sillanpää et al 1996), their condition complicated by cognitive problems and motor skill weakness (Dunne et al 1993). Low physical activity also leads to a high risk of osteoporosis, a risk further increased by antiepileptics (Center et al 1994) and overweight (Rimmer et al 1994).

Health promotion for ID populations is often inconsistent (Haefner and Elkins 1991; Beange et al 1995; Golden and Hatcher 1997). Although nutrition is depended upon environment, overweight is more common with persons with ID, the mildly disabled being more overweight than the profoundly disabled (Bell and Bhate 1992; Rimmer et al 1993). Diabetes is frequent, varying from 2.4% to 3.4%, and increasing to 8.9% in persons over 60 years of age (Hogg and Moss 1993; van Schrojenstein Lantman- de

Valk et al 1997). While smoking is less common in ID subpopulation overall (Hymowitz et al 1997; McDermott et al 1997), 37% of persons with mild ID living in the community have been identified as to smokers (Tracy and Hosken 1997). Screening of common cancers, such as breast cancer and uterine cancer, is not common (Cowie and Fletcher 1998).

4.10 Summary of the literature and justification of the present study

The biological features of persons with ID and environmental risks produce shorter life expectancy and high mortality for ID population in general, but what are the differences between the different subgroups with ID. The mortality and life expectancy studies in the past have been based on exclusive populations selected by their residence or their usage of special services (Balakrishnan and Wolf 1976; Eyman et al 1988; Strauss and Kastner 1996). There are no cause specific mortality figures for all levels of ID. Cancer and suicide rates in ID population are yet unknown. As the number of persons with ID living in the communities has increased, requests have been made for population-based samples (Strauss and Kastner 1996; Hayden 1998).

5 Aims of the study

The purpose of this study was to investigate the long-term survival of a nation-wide, population-based cohort of persons with ID in relation to sex, age, level of ID compared with sex and age-matched general population. The specific aims of this study are:

1. To analyse the life-expectancy and survival of persons with ID (Study I)
2. To investigate mortality and cause-specific mortality in this population (Study II)
3. To calculate the cancer incidence and prevalence in this population (Study II and III)
4. To describe and analyse the suicide mortality and suicide patterns and risk factors of suicide of persons with ID (Study IV)

6 Author´s own contribution in the study

The author has attended the study throughout recent data collection from 1997 onwards. Mainly the Finnish Association on Mental Retardation did the extensive data collection, but the author has participated in the identification and process data entries and in the converting the original data into computer. The author has been in contact with all registries needed in this study and she has linked the material used in this thesis. The original study material preserved in the National Archives of Finland has given to this thesis an insight into the lives of our study subjects during the beginning of the follow-up. There was a clinical sample collection in 1998. Although this material is not included, it has given valuable information of the present situation of the survived study population. The author has chosen the methodology of the study with the assistance of several competent specialists from different areas of epidemiological studies. The author has full contribution in all publications.

7 Ethical issues

The Ministry of Social Affairs and Health, and Ministry of Education approved the study. The ethics of this study has been considered in the permission by the ministries. The Data Protection Ombudsman approved of the data protection and linking the original material with national databases. The personal information of suicide cases is changed to encounter the protection of human rights of persons in this study.

8 Study design

This study was designed to be a prospective cohort study with a 35-year follow-up period. The study population included all intellectually disabled persons from the population study Finland in Miniature (in Finnish "Pienois-Suomi"), from 1962 forming a cohort of 2372 persons. In the study the persons were followed from 1st January 1962 to 31st December 1997 from national databases (Table 3). Since 1st January 1967 all residents of Finland have had a unique personal identification number, which enables the data linkage with the national databases. Information was collected of vital status, date of death, cause of death, cancer and suicides. Original data from 1962 was also utilised in analysis of life expectancy.

Table 3. National databases used in the present study.

1.	Original data of persons with intellectual disability: National Archives Service of Finland
2.	Death certificates: Statistics in Finland
3.	Pensions, medication and rehabilitation: Social Insurance Institution of Finland
4.	Social and health services: National Research and Development Centre for Welfare and Health

9 Subjects

9.1 Region of the study

In 1962 a large, nation-wide population-based study on ID was conducted. The cross-sectional, multidisciplinary study was undertaken to investigate the prevalence of ID (Table 4) and needs of services of people with ID. Within the country 57 municipalities were selected by socio-economical range to represent Finland. They were chosen in by their population, financial classification, social expenditure and occupational structure (Tarvainen 1966). There were 416,973 persons which was 9.4% of the total population (Amnell et al 1964). The regional distribution is shown in Table 4 (Tarvainen 1966).

Table 4. Prevalence (per 1000) of intellectual disability in the 1962 study[1] by level of ID in 1962 by Tarvainen[2].

Area	Severe ‰	Moderate ‰	Mild ‰	Total ‰
Total	**0.89**	**2.07**	**2.60**	**5.56**
Urban area	0.71	1.31	1.90	3.92
Rural area	0.98	2.47	2.96	6.41
Lounais-Suomi, South-West Finland	0.92	1.73	2.26	4.91
Eteläinen rannikkomaa, South Coast	0.71	2.25	2.36	5.32
Järvi- Suomi, Lake Region	1.12	3.22	3.57	7.91
Suomenselän suomaa, Nort East Finnish bogland	0.88	2.53	3.41	6.82
Pohjanmaan lakeus, Plain of Ostrobothnia	0.97	2.54	3.23	6.74
Vaara-Suomi, Eastern Finnish hill county	1.49	2.79	3.40	7.68
Lappi, Lapland	0.73	1.60	2.25	4.58

[1]Percentages calculated from population as registered in 1st January 1962. Those whose level of ID was unknown or had been diagnosed earlier were not included in the table. *Modified from Tarvainen 1966.*
[2]Prevalence of ID vary in three reports from the Finland in Miniature study. Psychological study is most reliable (6.62 per thousand). Medical part used persons examined by physician (prevalence 6.62) and social part the number of persons with social inquiry (prevalence 5.56).

The municipal officials (social workers, public health nurses, schools, child health centres, medical centres and other community officers) were asked to report all persons suspected or known to have an intellectual disability. There was an inquiry of persons a native of these municipalities in all Finnish institutions for intellectually disabled. They reported from 6.48 per thousand to 13.5 per thousand of population to the research group. The research team (for further information see Appendix I) examined reported persons.

9.2 Inclusion criteria of the subjects in 1962

Persons were included in the study if they met the criteria of ID (mental subnormality) by WHO in 1959. Psychological tests were used to determine whether persons met the criteria for ID (IQ < 70) and whether their learning capacities were eligible for schooling (Ruoppila 1966). In 1962 IQ was a sum estimate of a large set of tests (to increase the reliability) (i.e. Raven, Passive vocabulary test), which were standardised in Finland and were able to identify persons with ID (Ruoppila 1966). The tests included measures of verbal and non-verbal intelligence. Academic abilities, such as reading, writing and mathematics, were also measured. Clinical evaluation of level of the intelligence was done for all persons to increase the reliability and it also included evaluation of social skills, communicative skills and emotional status. The level of ID was clinically evaluated by psychologists and physicians for those unable to participate in the testing.

Physician performed clinical examination in standard form (see Appendix I). No laboratory or X-rays were done. The medical diagnostics have improved since year 1962, so diagnoses of aetiological factors and associated disorders are treated as background variables bearing in mind their limitations.

9.3 The study cohort in 1962

Municipalities reported a total of 4,013 persons between ages 2 and 64, of which 3,748 were examined and 84 were included on the basis of patient records. Physician studied 2,553 persons and patient records from institutions were obtained for 84 persons and 40 persons were included in only by the psychological examinations only. The prevalence of ID in the study was 6.62 per thousand and the distribution of levels of ID and age are presented in Tables 5 and 6 and in Figure 2.

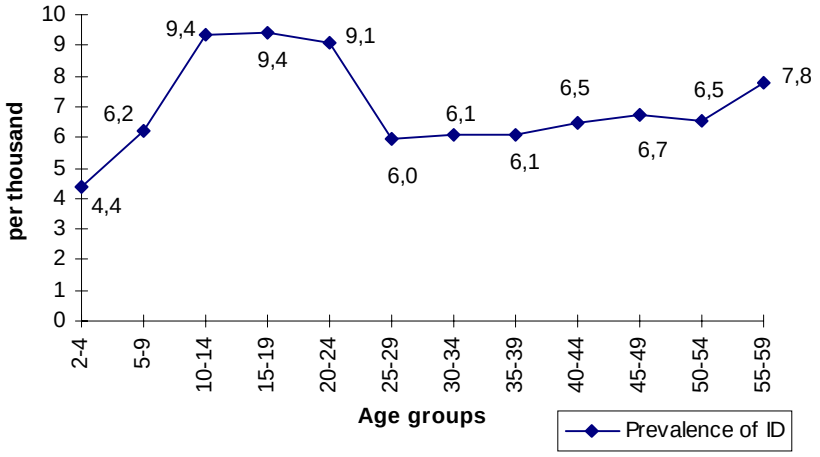

Figure 2. **Prevalence of intellectual disability in various age groups. Modified from Ruoppila 1966.**

Table 5 . **Persons with intellectual disability (ID) according to sex, age group and degree of ID in 1962. Down syndrome (DS), presented separately.**

Level of ID	Unknown level of ID	Profound n *(%)*	*%**	Severe n *(%)*	*%**	Moderate n *(%)*	*%**	Mild n *(%)*	*%**	Total n *(%)*	*%**
Total	**11** *(0.5%)*	**368** *(16%)*		**280** *(12%)*		**606** *(26%)*		**1,101** *(46%)*		**2,372**	
Men	4	213	*58*	143	*51*	267	*44*	556	*50*	1,187	*50*
Women	7	155	*42*	137	*49*	339	*56*	545	*50*	1,185	*50*
Age											
2-9	5	92	*25*	57	*20*	85	*14*	138	*13*	379	*16*
10-19	0	126	*34*	86	*31*	101	*17*	319	*29*	632	*27*
20-29	0	46	*13*	36	*13*	85	*14*	148	*13*	315	*13*
30-39	3	53	*14*	32	*11*	80	*13*	163	*15*	331	*14*
40-49	3	30	*8*	23	*8*	108	*18*	136	*12*	300	*13*
50-59	3	15	*4*	31	*11*	104	*17*	150	*14*	303	*13*

* Percentage within the group defined by the level of the ID.

Table 6. Prevalence of intellectual disability (ID) in Finland in Miniature- study in 1962 by the level of ID by Ruoppila[1].

Level of ID	IQ	Prevalence per 1000
Profound	< 20	1. 11
Severe	21-34	0.91
Moderate	35-49	1.63
Mild	50-69	2.97
Total		**6.62**

[1]Prevalence of ID varies in three reports from the Finland in Miniature study. Psychological study is most reliable (6.62 per thousand). Medical part used persons examined by physician (prevalence 6.62 per thousand) and social part the number of persons with social inquiry (prevalence 5.56 per thousand).

9.4 The identification process and follow up from 1995 to 1998

The follow-up study was initiated in 1995. The original 1962 study forms of medical examinations and psychological tests were re-reviewed. The names and dates of birth of subjects and their parents were collected for identification, since the personal identification number was not introduced until 1966 in Finland. There were the original research documents, the special blank-forms, from the psychological and medical examinations available for this study in the National Achieves of Finland and the punch cards for 2,372 persons with ID. Medical data and psychological estimates were entered to punch cards (n=2372) and were the most reliable source of information. Author went over the medical forms and psychologist the psychological forms, respectively. The original forms of persons with DS were missing, but the data was in the punch cards. Scanning the cards individually transformed the punch cards into digital coding. In three original reports, the numbers of persons with ID vary, but we have chosen to include those persons, who were in psychological testing found to have ID and have an original form or in case of DS a punch card. The different populations of the study in 1962 and the present study are shown in Figure 3 and Table 7.

The retention rate was therefore 97.4%. All persons from the 1962 cohort were not identified, but it was possible to analyse the dropouts by the original material. The dropouts did not differ significantly from the study population by sex or level of ID, but were generally born in the first two decades of the century.

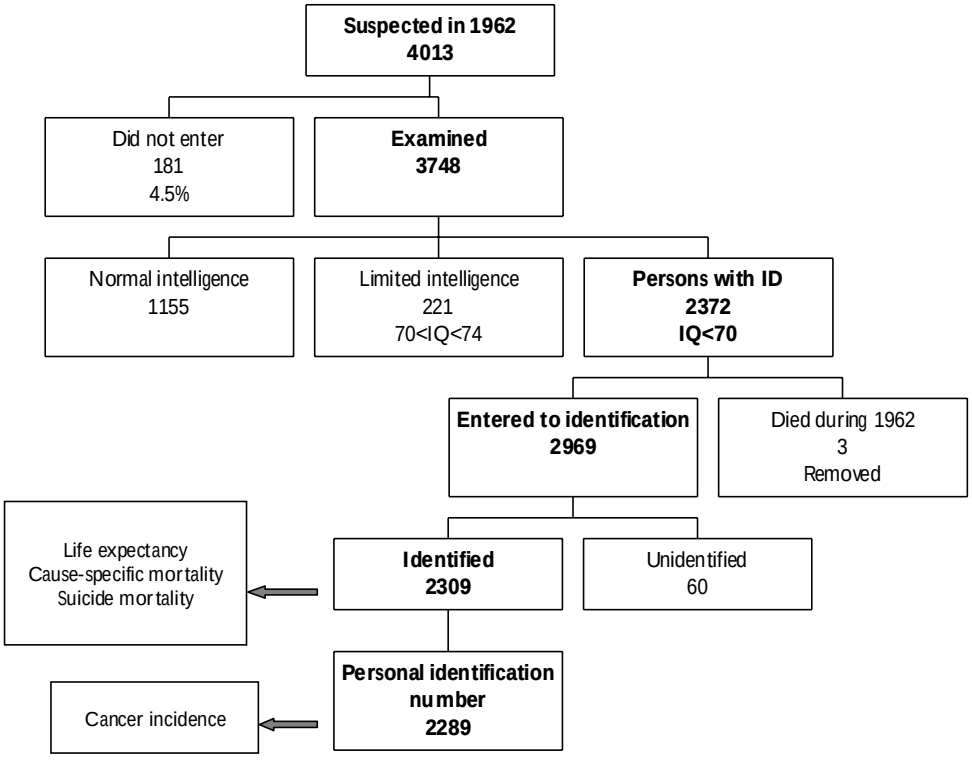

Figure 3. **Flowchart of the population in the present study from 1962 to 1997 from numbers of suspected persons with intellectual disability to study populations and the present studies conducted on these populations. Identification was done in Population Register Center (86.5%), The Parishes of Finnish Lutheran Church (4.2%) and Administrative Council of Helsinki (7.8%)**

Table 7. Persons with intellectual disability in 1997: survived, deceased and unidentified groups by sex, age, age at death and level of intellectual disability.

	Unknown	Profound	Severe	Moderate	Mild	Total
Alive						
Mean age	*56.2*	51.5	51.9	56.8	55.4	54.7
(SD)	(22.6)	(9.9)	(9.9)	(13.5)	(11.8)	(11.9)
Men	*1*	*92*	*83*	*130*	*312*	*630*
Women	*2*	*64*	*63*	*134*	*317*	*579*
Total	**3**	**156**	**146**	**403**	**618**	**1,198**
Age group						
37-39	0	11	0	18	27	56
40-49	2	72	84	73	242	473
50-59	0	47	39	80	174	340
60-69	0	16	12	44	91	163
70-79	0	8	8	28	71	115
80+	1	2	3	21	24	51
Deceased						
Mean age	*65.2*	42.1	56.4	61.7	61.9	57.7
(SD)	(11.8)	(19.2)	(18.3)	(17.0)	(16.2)	(18.7)
Men	*2*	*117*	*63*	*132*	*232*	*546*
Women	*6*	*83*	*65*	*195*	*216*	*565*
Total	**8**	**200**	**128**	**327**	**448**	**1,111**
Age group						
2-9	0	7	1	1	1	10
10-19	0	23	4	6	7	40
20-29	0	29	9	17	18	73
30-39	0	36	11	16	21	84
40-49	1	32	14	26	45	118
50-59		26	32	58	75	191
60-69	3	33	23	91	124	275
70-79	3	9	25	77	114	228
80+		5	9	35	43	92
Identified						**2,309**
Unidentified		13	6	15	26	60
Total	**11**	**368**	**280**	**606**	**1,101**	**2,369**

The follow up included a new sample to evaluate the predictability and reliability of the 1962 study and stability of IQ during lifespan (Ruoppila and Numminen 2000).

10 Methods for present study

The data collection was carried out in years 1995-1997. The Population Register Centre of Finland, the Parishes of Finland and the Administrative Council of Helsinki provided information of vital status (Study I) and the date of death (Study II). Statistics Finland provided the death certificates, which were possible to receive also for the persons, who deceased before 1967, since the death registry in Statistics Finland is organised by the year of death and names before 1967. Death certificates were collected and reviewed by authors. In the Finnish Cancer Registry the personal identification numbers were used as a key to their cancer statistics (Study III). From the death certificates the persons who committed a suicide or whose death was classified as undetermined were collected and their case records were reviewed, except for two persons, who did not have any case report, but the caregiver of these persons were interviewed (Study III). The data classification is described later.

10.1 Medical disorders

The original study included information on aetiological and associated disorders (see Appendix I). The diagnoses based mainly on the information in the clinical examination and were accompanied with patient records desultorily. Therefore most of clinical information of the 1962 study was rejected. Variables consisting important clinical value were included in the analysis of the background variables: DS, epilepsy, CP, hearing and visual impairment (the classification and prevalence of associated disorders presented in Appendix I).

10.2 Classification and diagnoses of deaths

Cause of death depends on manner of inquest of death and medical practice of era. Form of certificate and diagnostic criteria have changed over this period, but classification of death and practice of inquest has remained same through this period. Classes of death were

classified into six categories according to the Finnish practice of inquest: disease, accident, suicide, homicide, war and undetermined causes. The practice of inquest and a residency of a person were coded.

Causes of death were divided into primary, immediate and contributing causes. The primary cause of death, is a disorder, disability or cause initiating the set of diseases leading to the immediate cause of death. The immediate cause of death is a disease, defect or disability, according to signs of which patient had died. Contributing cause of death is an additional disease, defect disability or cause contributing to the death. All these causes of death were classified into 18 categories according to ICD-9 (WHO 1975). Causes of death were coded from ICD-10 and ICD-8 according to ICD-9 codes, since largest population had died 1987-1995 and changes in coding from version ICD-8 to ICD-9 were modest (WHO 1965).

10.3 Suicide predictors

All cases of death classified as suicide or undetermined deaths were included in the suicide study. Author reviewed patient documents of these persons and in two cases the caregivers were interviewed due to lack of patient documents. The diagnoses, medication, associated disorders, signs of depression, suicide attempts, and emotional losses and social network were recorded. Case reports were formed without any information on identity. The undetermined cases were evaluated for their suitability for suicide. The suicide methods were classified by international classification of suicide methods (Öhberg et al 1995). The method of suicide was defined as active, if means of suicide needed any acquisition; and passive if no requirements of skills were needed to acquire.

10.4 Classification of neoplasms

Finnish Cancer Registry is a population-based registry covering the whole Finland. It receives information from all levels of healthcare. Various surveys have indicated that the coverage is almost complete (Teppo et al 1994). The coding is always supervised and checked by a physician.

Classification of neoplasms is based on topographic classification arranged according to the anatomical site of tumour, except for few histological types such as melanomas, lymphomas and leukemias (Muir and Percy 1991). The neoplasms in this study are classified as they appear in the ICD-9, which comprises the categories running from 140 to 239 and they are divided into six groups according to the behaviour of the neoplams: malignant neoplasms, malignant neoplasms of lymphatic and haematopoetic tissue, benign neoplasms, carcinoma in situ, neoplasms of uncertain behaviour and neoplasms of unspecified nature (WHO 1975). The benign neoplasms are excluded from this study.

11 Data analyses

11.1 Background variables

Background variables are presented in table 8. There were also information of birth, family and self-regulating skills and evaluation of need of care. Statis-tical packages SPLUS 5.0 (MathSoft Inc), SPSS 8.0 (SPSS Inc.) and Glim (Hakulinen et al. 1988) were used.

Table 8. Background variables used in the present study to describe risk factors for life expectancy and cause specific mortality modified from the original data (further information in Appendix I).

Variable	Classification	Study I	Study II	Study III	Study IV
Age	In years	+	+	+	+
Sex	male, female		+	+	+
+					
Age groups	In 5-year age groups	+	+	+	+
The level					
in 1962	Dichotomy	+	+	+	+
Down					
syndrome	Dichotomy	+	+		+
Epilepsy	Dichotomy	+			
Cerebral palsy	Dichotomy	+			
Visual					
impairment	Dichotomy	+			
Hearing					
impairment	Dichotomy	+			
Need of care	institutional care and				
in 1962	semi-independent living				
	or independent living	+			
Mobility and	Three-step scale:				
self-helping	highly dependent,				
skills	moderately dependent,				
	independent	+			

11.2 Life expectancy (I)

The proportion of expected life lost and life expectancies were calculated with the survival analysis package developed by the Finnish Cancer Registry (Hakulinen et al 1988). This package produced extensive life tables for all groups, including standard life-table information and also the baseline probability of surviving over the period calculated from mortality figures of the general population. An age, sex, and quinquinneum-specific general population survival probability was obtained from the published statistics for every person entering the interval period together with the number of those who died during the interval. The data were analysed as binomial variables with effective number as a total number. The main purpose in using this model instead of Kaplan Meier (Kaplan and Meier 1958) or Cox´s model (Cox 1972) or other regression models, was to prevent the bias with comparison population.

In order to compare the total mortality of different groups, Kaplan Meier curves were calculated (Kaplan and Meier 1958). Cox's proportional hazard models were used to quantify the differences between groups (Cox 1972). Variables in the variables were age, square of age, sex, and level of ID, epilepsy, visual and hearing impairment. Interaction terms sex/age and sex/level of ID were left out from the final analysis as insignificant.

11.3 Cause-specific mortality (II, IV)

Death certificates (n=1095) were received from Statistics Finland and reviewed by the author. An indicator of relative mortality differences was obtained by calculating the age-adjusted relative mortality rates of persons with ID compared with the general population. The absolute differences between the age-standardised death rates were analysed. The age-standardised cause-specific death rates were needed to calculate the absolute mortality differences; and the share of each cause of death in the total excess mortality of persons with ID, was derived by indirect age standardisation, using 5-year age bonds. In order to compare the mortality of different diseases Cox's proportional hazard models were used to quantify the differences between groups (Cox 1972). Variables in the equation were: age, square of age, sex, level of ID, epilepsy, visual and hearing impairment, visual impairment. Interaction terms sex/age and sex/level of ID were left out from the final analysis as insignificant.

11.4 Cancer (III)

The expected numbers of cases for total cancer and for specific cancer types were calculated from age and sex matched general population for sexes, all levels of ID and DS. The SIR´s were calculated by dividing the observed number of cases by expected number of cases. The exact 95% confidence intervals were given on the presumption that the number of observed cases followed the Poisson distribution. Though there is some regional variation of cancer within Finland, it has a only slight effect on incidence rates, so countrywide rates have been used in this study (Pukkala 1992).

12 Results

12.1 Population characteristics

The cohort for the present study included 2,309 persons, 1,165 (50.4%) men and 1,144 (49.6%) women producing a total of 60,969 person years during the 35-year follow-up period (Table 7). The mean follow-up time was 26.9 years (SD 10.9) and 59% of the study population was followed over 30 years. At the end the follow-up period 50.6% of the original study population was alive, 13% with profound, 12% with severe, 22% with moderate and 53% with mild ID. The mean age of men alive was 53.2 years (SD 10.8), and respectively for women 56.4 years (SD 12.7). At the end of the follow-up period oldest man was 99 years old with mild ID and woman 97 years old with moderate ID. The mean age of women death was 59.3 years (SD 17.8) and respectively for men 55.5 years (SD 19.5). At death the oldest man was 93 years old and woman 97 years old.

12.2 Life-expectancy

Life expectancy had positive correlation with the level of ID. For persons with mild ID, the proportion of expected life lost did not differ from that of the age-sex stratified general population during the first three decades of life (Table 9). In the moderate ID group, the proportion of life lost increased slightly and in the severe and profound ID group, it increased significantly to 20% for all age groups. The survival of persons with ID had improved as the number of persons aged from 40 to 59 years had increased over 30% from 1962 to 1997 (Figure 4).

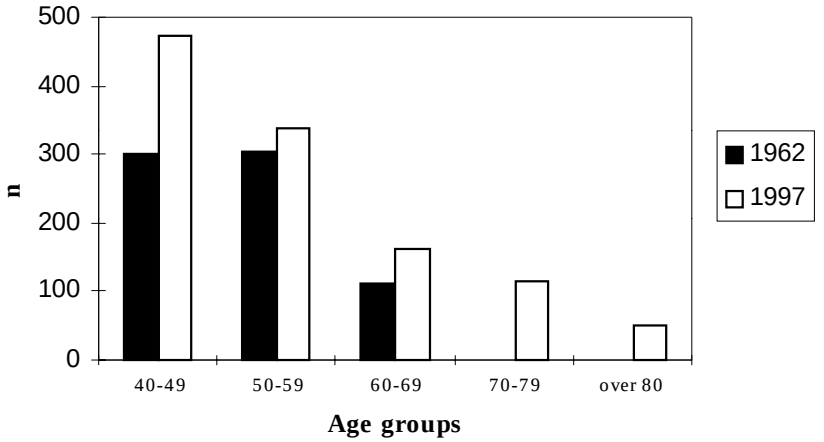

Figure 4. **Number of persons over 40 years with intellectual disability in 1962 and 1997 in the study cohort. (Note that in 1962 persons older than 64 were excluded from the study).**

The expected life span in 1963 calculated for ID population, showed a difference between sexes compared to the general population, albeit of smaller magnitude, except for profound ID than in other levels of ID, where no definite difference was shown (Table 9). Women with profound ID had poorer life expectancy than men with profound ID after age 20, but this was reverse in the age group under 20 years (Table 9).

Table 9. **Proportion of expected life lost in 35-year follow-up study by age group in 1963, sex and intelligence quotient calculated with the survival analysis package developed by the Finnish Cancer Registry. When expected life lost is negative, the group has x% better survival than the general population.**

Age band in 1963		*Men*	*Women*
0-10	Mild (50-69)	-4.8	-3.3
	Moderate (35-49)	0.6	13.6
	Severe (20-34)	-7.7	4.0
	Profound (0-19)	35.3	24.8
11-20	Mild (50-69)	-4.1	-5.7
	Moderate (35-49)	1.6	0.9
	Severe (20-34)	2.2	11.3
	Profound (0-19)	26.0	18.5
21-30	Mild (50-69)	-0.04	4.7
	Moderate (35-49)	16.0	13.8
	Severe (20-34)	4.9	37.6
	Profound (0-19)	19.0	33.3
31-64	Mild (50-69)	5.5	14.0
	Moderate (35-49)	-1.7	20.9
	Severe (20-34)	15.1	22.0
	Profound (0-19)	23.7	42.9

Table 10 presents significant explanatory variables for high mortality risk (variables in table 8). Each risk factor contributes to the logistic regression equation by percentage given in the table. Most risk factors are disorders associated with ID, but in older groups, age itself was a significant predictor of mortality (Table 10).

Table 10. Significant factors (p< 0.05) contributing to low survival using Cox´s regression by age groups.

Age group in 1963	Risk factor	coefficient of determination[1]
2-9	Profound ID	10.6
	Epilepsy	2.2
	Hearing impairment	4.2
	Young age within the group	26.9
10-19	Profound ID	6.9
	Epilepsy	7.2
	Young age within the group	31.6
20-29	High IQ	5.4
	Ageing within the group	33.9
30-39	Ageing within group	36.2
40-49	Moderate ID	9.5
	Ageing within group	36.9
50-64	Ageing within group	36.6

Variables in the equation: age, square of age, sex, level of ID, epilepsy, DS, visual and hearing impairment, visual impairment. Interaction terms sex/age and sex/level of ID removed from the final analysis as insignificant.

[1]Coefficient of determination describes the increase in determination of equation by the variable in percentages

From the results it was possible to calculate the proportion of ageing persons with ID in Finland 1997 (Table 11). The number of aged persons with ID will increase fast in next ten years, but the proportion will remain in 0.4% of adult population.

Table 11. Estimated number and proportion of persons with ID in 1997 by age groups, extrapolated1 by the population follow-up study 1963-1997 in Finland.

	Men		*Women*	
Age group	n	%	n	%
40-49	2,777	0.7	2,096	0.5
50-59	1,851	0.6	1,649	0.5
60-69	798	0.4	851	0.3
70-79	415	0.3	670	0.3
80-	170	0.4	330	0.3
Total	**6,011**		**5,596**	

[1]The estimation was calculated from the study population. The number of persons alive represents 9.4% of ID population, so full numbers can be multiplied from them. The age groups for extrapolation were obtained from Statistics in Finland.

12.3 Mortality

Forty-seven percent of the original study population had deceased, yielding a mortality rate 17.7 per 1000 person years (CI 95% 8.4-27.0). In Cox´s regression low level of ID was a significant risk until age 50 years and age itself and visual impairment were significant risks after age 50 (Table 10). In younger ages, epilepsy increased the risk of death 2.7 times in age group under 10 years and visual impairment, respectively, in age group from 50 to 59. The need of care correlated with IQ (Pearson´s correlation 0.653, $p<0.01$); the persons with institutionalised care were more often profoundly disabled and with more associated disorders; and need of institutional care indicated poorer survival in all age groups.

There was excess disease mortality of men with ID under 39 years of age compared with the age-sex stratified general population, but not in the later years (Table 12). In age groups younger than 40 years the relative disease mortality excess was larger for men than women, but not later. Men with ID were in highest risk to die on disease before age 20, but at the same time their risk of accident was only one tenth of the risk of the general population (Table 12). Ageing increased the risk of external causes such as accidents for both sexes with higher risk for women than men (Table 12).

Table 12 . Adjusted risk ratios of death caused by disease and external cause for the persons with intellectual disability compared with the general population by the level of intellectual disability and age groups.

Disease

Age group	2-19		20-39		40-59		60-	
Level of ID	Men	Women	Men	Women	Men	Women	Men	Women
Mild	2.6	1.9	1.6	1.2	1.0	1.1	1.0	1.0
Moderate	1.0	1.0	2.3	1.5	1.1	1.1	1.0	1.0
Severe	2.8	1.8	2.6	1.6	1.2	1.0	1.0	1.0
Profound	3.3	1.9	2.1	1.3	1.1	1.2	1.0	1.0
Total	**2.6**	**1.7**	**2.2**	**1.4**	**1.0**	**1.1**	**1.0**	**1.0**

External causes

Age group	2-19		20-39		40-59		60-	
Level of ID	Men	Women	Men	Women	Men	Women	Men	Women
Mild	0	0	0.5	0.9	1.1	0.6	1.4	2.2
Moderate	1.2	1.3	0.4	0.7	1.1	1.1	0.4	2.1
Severe	0	0	0.4	0.5	0.8	0	0	1.2
Profound	0	0	0.1	0	0.3	2.5	2.3	0
Total	**0.1**	**0.1**	**0.3**	**0.4**	**1.1**	**1.0**	**1.1**	**1.9**

Obduction was performed in 27 % of cases. The annual obduction rate varied greatly from 5 to 54 percent, with an increase in the 1980s. There was no difference between the pattern of ID in obducted and non-obducted people. Men were more often obducted than women were and obducted people were younger. Cases with an external cause were all obducted. In addition, congenital disorders, gastrointestinal causes and neoplasms had higher obduction rates than other categories.

12.3.1 Cause specific mortality

Three most common causes of death were in both sexes cardiac diseases, respiratory diseases and cancer (Table 13). Diseases of digestive system included intestinal obstruction as the primary cause of death in 19 (25%) cases, of which 11 had profound ID, and ulceric perforation in 10 (13%) cases, mostly with mild ID. There were 39 neurological causes of death, of which the most common were epileptic seizures (n=16) and dementia (n=10).

Table 13. Observed (O) and expected (E) deaths of men with intellectual disability with agestandardised relative mortality (RR) compared with the general population aged through 2 to 97.

		O	E	RR	CI 95%	O	E	RR	CI 95%
Age group			*Men*				*Women*		
2-19	Infectious diseases	**3**	1.6	*1.8*	0-11.1	**2**	0.6	3.2	0-3.2
	Cancer	**0**	8.2	*0.0*	0-0.1	**0**	6.7	0.0	0-0.1
	Vascular diseases	**4**	3.9	*1.0*	0-6.6	**0**	1.3	*0.0*	0-0.1
	Respiratory diseases	**14**	2.4	*5.8*	4.4-15.6	**6**	1.4	*4.3*	0.3-4.7
	Digestive system	**2**	0.8	*2.4*	0-4.4	**1**	1.2	*0.8*	0-4.1
	External causes	**2**	16.9	*0.1*	0-0.6	**1**	8.3	*0.1*	0-0.2
	Other causes	**5**	11.0	*0.5*	0-1.4	**9**	6.8	*1.3*	0.2-1.4
20-39	Infectious diseases	**3**	1.2	*2.5*	0-4.7	**6**	1.5	*4.1*	0.6-9.5
	Cancer	**4**	15.9	*0.3*	0-0.6	**2**	22.4	*0.1*	0-0.3
	Vascular diseases	**15**	20.7	*0.7*	0.5-1.8	**15**	12.8	*1.2*	0.6-1.9
	Respiratory diseases	**19**	3.5	*5.4*	2.9-8.0	**11**	3.5	*3.2*	1.1-5.1
	Digestive system	**8**	1.8	*4.3*	1.0-7.2	**6**	1.9	*3.1*	0.4-6.1
	External causes	**14**	52.4	*0.3*	0.2-0.8	**8**	27.1	*0.3*	0-0.3
	Other causes	**22**	27.0	*0.8*	0.6-1.6	**22**	19.8	*1.1*	0.5-1.2
40-59	Infectious diseases	**4**	1.8	*2.2*	0-4.1	**3**	2.0	*1.5*	0-3.8
	Cancer	**17**	30.1	*0.6*	0.1-0.9	**17**	64.3	*0.3*	0-0.8
	Vascular diseases	**54**	73.6	*0.7*	0.6-1.4	**49**	48.4	*1.0*	0.7-1.3
	Respiratory diseases	**32**	5.8	*5.5*	3.5-7.5	**36**	5.8	*6.2*	4.1-8.2
	Digestive system	**9**	2.7	*3.3*	0.7-4.0	**12**	3.8	*3.1*	1.7-7.1
	External causes	**17**	28.5	*0.6*	0.4-1.3	**10**	19.5	*0.5*	0.1-0.6
	Other causes	**18**	19.9	*0.9*	0.5-1.4	**30**	19.5	*1.5*	0.9-2.1
60+	Infectious diseases	**4**	2.3	*1.7*	0-2.9	**6**	2.9	*2.1*	0.3-4.9
	Cancer	**39**	64.7	*0.6*	0.4-0.7	**43**	71.3	*0.6*	0.5-0.9
	Vascular diseases	**110**	144.2	*0.8*	0.5-0.9	**150**	168.3	*0.9*	0.8-1.2
	Respiratory diseases	**63**	23.5	*2.7*	2.8-4.7	**55**	16.8	*3.3*	1.7-3.0
	Digestive system	**18**	5.6	*3.2*	1.0-2.9	**20**	9.4	*2.1*	1.9-5.2
	External causes	**13**	12.3	*1.1*	0.5-2.0	**16**	10.2	*1.6*	0.6-2.0
	Other causes	**23**	16.7	*1.4*	0.4-1.0	**26**	31.3	*0.8*	0.5-1.2

During the 30 year follow-up period, 173 patients with cancer was diagnosed; the expected number was 188 (SIR 0.9, CI 0.8-1.0). The risk of cancer was similar with the general population (Table 14). The level of ID did not have a significant correlation on overall cancer incidence, but women with profound ID seemed to have increased risk of cancer as men had decreased risk, respectively (Table 14). Low IQ increased the risk of neoplasms of nervous system, testicular cancer, oesophagus, gallbladder and tumours of other gastrointestinal organs (mainly omentum) (Study III, Table 3).

12.3.2 External causes of death

Eighty persons (7.8%) died as a result of an external cause, which includes accidental or violent circumstances, suicides. There was no difference in accident frequency between men and women. A risk of accident was lower for persons under 60 years of age than in the general population even when occupational accidents and traffic accidents were considered. For the elderly, women had an increased risk of fatal fracture compared to the general population. Respectively, for men risk of external cause was increased due to the higher suicide rate. Moderate ID and age over 50 years were significant risk factors ($p<0.05$ in logistic regression) for accidents as were the community settings and mental hospital. The fatal fractures were the most common primary cause of death (n=24) caused by either falling or getting hit by a car.

Ten persons of 2,369 had committed a suicide during the follow-up. Six persons who committed a suicide were patients in mental hospitals. Persons divided into two groups, persons in their 30´s and in their 60´s. People with ID seem to committed suicides in

Table 14. **Observed (O) and expected (E) numbers of cancer cases and standardised incidence ratios (SIR) with their 95% confidential intervals among persons with intellectual disability by age and level of intellectual disability.**

	O	E	SIR	95%CI	O	E	SIR	95%CI
Age		*Men*				*Women*		
7-14	0	0.1	0	0-9.6	0	0.1	0	0-9.6
15-29	3	1.4	2.1	0.4-5.2	2	1.3	1.5	0.1-4.4
30-44	7	6.2	1.1	0.4-2.1	9	10.6	0.8	0.4-1.5
45-59	23	21.1	1.1	0.7-1.6	25	27.4	0.9	0.6-1.3
60+	48	65.9	0.7	0.5-0.9*	56	53.5	1.0	0.8-1.3
Total	**81**	**94.8**	**0.8**	**0.7-1.0**	**92**	**93.0**	**1.0**	**0.8-1.2**
Level of ID								
Profound	6	9.0	0.7	0.2-1.4	8	6.2	1.3	0.6-2.5
Severe	10	10.5	0.9	0.5-1.7	8	8.0	1.0	0.4-2.0
Moderate	20	26.6	0.7	0.5-1.1	28	30.2	0.9	0.6-1.3
Mild	45	48.3	0.9	0.7-1.2	47	47.7	1.0	0.7-1.3

*p<0.05

similar frequency in their adult life irrespective of age in both groups. There was no significant difference in suicide rates between women and men in this study, but men with ID had reduced suicide rate of one third of the general suicide rate of men (Table 15). In present study women committed 13.2 suicides per 100,000 and men 19.3 per 100,000, while corresponding figures for general population in 1980 were 13.2 and 52.9 per 100,000, respectively (Statistics Finland 1997).

Drowning was the most common method of suicide. None of the persons was under influence of alcohol at the time of death. Degree of ID was mild in nine cases and moderate in one case (details in Study IV table 2). Aetiology of ID remained unknown in 9 cases and was asphyxia at birth in one case. Two persons had epilepsy and one had suffered convulsions as a child. Three persons were not able to communicate by words. Five persons were hospitalised because of psychiatric disorder and one person was an inpatient because of ID. Of six cases, 4 had psychiatric disorder started in teens and two suffered more from adjustment problems. One person had suffered from sexual abuse. Social support was poor in all the patients, and in four families, family members had mental health problems. All suicide victims with chronic psychiatric disorder had previously attempted a suicide.

12.3.3 Mortality in Down syndrome andepilepsy

There were 85 (42 women and 43 men) deceased persons with DS. The mean age at the time of death was 44.6 years (SD 15.9). They were more vulnerable to infectious diseases (3.5%), respiratory diseases (34.1%) and urogenital diseases (3.5%) than persons without ID in this study. Respectively, less frequent causes of death were neoplasms (1.2%), vascular diseases (30.6%) and accidents and poisonings (5.9%).

There were 131 deceased persons with diagnosed epilepsy in 1962 (64 men and 67 women). They died younger than the total study population with mean age 43.1 years (SD 19.8). Persons with ID and epilepsy were significantly more often profoundly or severely disabled than persons with mild ID. Persons with epilepsy died more often to infectious diseases (3.8%), diseases of nervous system (13.0%), and accidents and poisonings (13.0%) than persons with ID without epilepsy. There was reduced rate in cardiovascular diseases (17.6%) and respiratory diseases (28.2%)

Table 15. Standardised mortality rates (SMR) for people with intellectual disability from the 35-year follow-up study in Finland by sex.

	Population size	O	E	SMR	CI 95%
Men	1,184	6	20.7	30.0*	10.4-63.5
Women	1,185	4	4.3	92.8	15.7-156.0
Total	**2,369**				

*Statistically significant

compared to the general population and persons without epilepsy. Epilepsy increased relative risk of death in all levels of ID (Figure 5) and it was two times higher than in the persons without epilepsy. In school age, epilepsy was a significant risk factor, but in age 29 years onwards epilepsy did not show any significance, perhaps because increased total disease mortality.

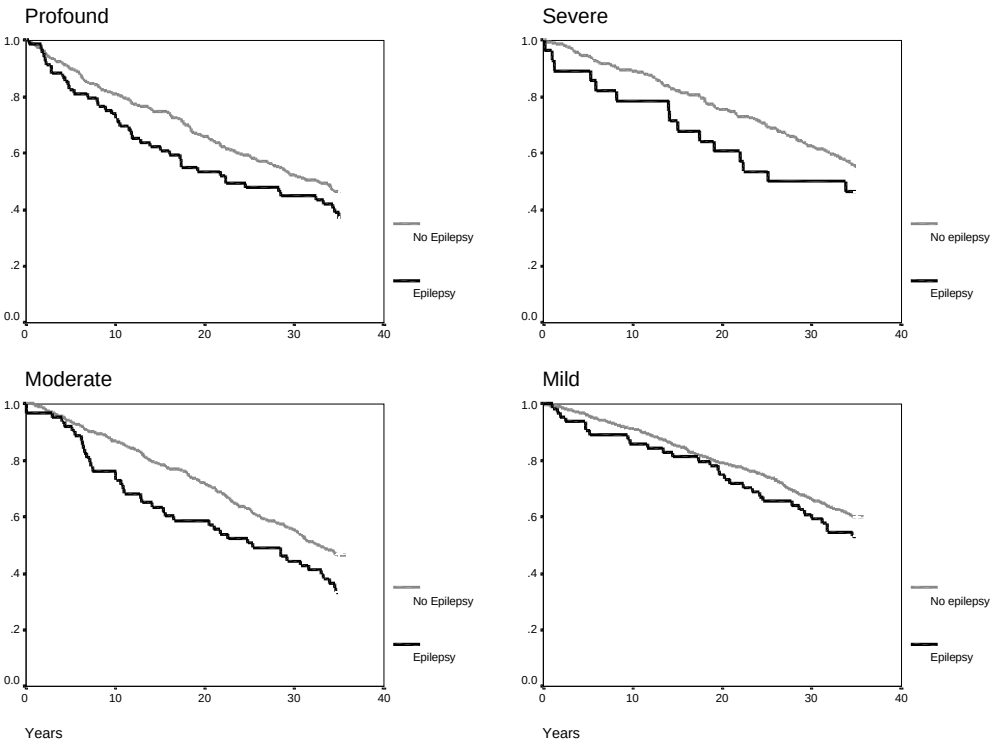

Figure 5. Survival of persons with intellectual disability (ID) with and without epilepsy by level of ID in 35 year follow-up.

13 Methodological discussion

13.1 General aspects

Intellectual disability offers an extensive challenge for researchers. Invention of testing of IQ expanded the possibilities to study the phenomena, but all persons with ID are hard to seek. At the beginning of the epidemiological research it was sought through relatives (genealogical random-test method). Birth registers, started in Germany by Klemperer in 1933 brought conceptions of prevalence and incidence (Göstason 1985). For follow-up purposes it takes time, so period method with cross-sectional samples was introduced with all persons with ID in certain area included in the sample. Studies of ID have many difficulties to encounter: definition, taxonomy, identification, reliability and validity of testing, geographical and social variables affecting the distribution of ID and in addition all other problems of epidemiological methodologies.

13.2 Study design

The study design in 1962 was a cross-sectional, the chosen municipalities represented Finland of that time. The main aim was to study the prevalence of ID between ages 2 and 64 years, to estimate the need of services for persons with ID. The structure was similar with large population based studies later conducted in Finland such as Mini-Finland (Aromaa 1989). It is suitable for prevalence calculations and demographic characterisation of populations, and investigating certain fairly stable characteristics of individuals such as exposures or risk factors to different classified variables. However, causality can not be assessed and in case of rare disorders, a large population is needed to ensure the sufficient number of cases, which is costly (Boyssy and Scott 1993). Moreover, epidemiology of ID is highly dependent on definitions, taxonomy, which have not been consistent (Fryers 1993).

Follows up of cohorts of persons with ID are rare. There are only few population-based cohorts, which were followed longer than 3 years (Stein et al 1976; Baird and Sadovnick 1985; Rantakallio and von 1986; Richardson et al 1986; Emanuelsson 1987; Rantakallio 1987; Goulden et al 1991; Ashwal et al 1994; Katusic et al 1996; Strömme and Valvatne 1998). This study is the longest

follow-up period as far we could ascertain. We used databases to obtain long-term data by data linkages, which saved time and costs, but allows a long follow up period. But it has been stressed, that it exposures to biases from linkage and failures in official records.

At present it would not be possible to repeat the 1962 study, since inclusion with only a suspicion of ID does not fulfil modern ethical research considerations or principles of personal data protection. The testing of large number of persons is also expensive. Nowadays, most samples of persons with ID, base either on registries of persons with ID, institutional samples or area search for persons with ID from educational or public health records, or the birth-cohort studies.

13.3 Definition of intellectual disability

In all epidemiological research, the definition of a phenomenon is the first guide to the results. The debate over defining the term ID has been continuous and there is no global agreement on comprehensive and accurate definition of ID thus far (Detterman 1987). The biological, psychological and social sciences have persisted the belief that individual differences diverge around from underlying type or essence (Gelb 1997). It is evident, that the survival of persons is not dependent only of his individual potentials, but depended upon the environment, community and culture (Fryers 1993). In this study ID is seen through intelligence, but we could use the economical, cultural or area variables. The choice of using definition from health care is biologically deterministic and allows no social interpretation. Historically, the definition of ID is culturally tied to developed world and disciplines of psychology and medicine. As there will not be comprehensive definition of ID for all purposes, we chose the one most often used in the context of epidemiology.

In 1962, the definition of ID lied more on psychological testing of IQ than at present as the social competence is included in the definition of ID. Epidemiological studies are always limited to the definition used and the location and population in which the research is carried out. The limiting values of levels of ID have fluctuated in different studies, especially between mild ID and borderline intellectual functioning. Bearing this in mind, generalisations and comparisons need to be done with caution (Munro 1986). If we use IQ scores and levels of ID as the main defining variable, the rigid nature of IQ testing in contrast to cultural aspects may influence our interpretation (Samelson 1997; Weidman 1997). If we use aetiological factors, such as tuberous sclerosis, we run into difficulties in finding representative patient populations, and moreover, we tend to generalise the features of persons of one aetiological group (Pennington et al 1992). Despite these problems with definition of ID, the epidemiological studies provide important information for further studies and health policy purposes.

13.4 Inclusion criteria in 1962

Inclusion of persons with ID into the study in 1962 had two steps. First, the municipalities had to report all persons suspected to have an ID. Children were recognised mostly by public health nurses in child health clinics. In Finland the network of child health clinics was extensive already in 1962. However, the children with profound ID are more likely to be identified as intellectually disabled than children with mild ID. Teachers are important in recognising mild ID and low schooling percentage has possibly decreased the number of children with mild ID in this study. The low schooling frequency and the differences in municipal systems may have affected the number of persons with ID, the distribution of levels of ID, but unlikely the diagnoses of ID.

In the second phase, persons with suspicion were tested with a set of tests, which were validated in Finland and found to correlate with IQ measure (Ruoppila 1966). Recognising of a person with ID by psychological testing has its problems. Tests were not fully validated for adults, and therefore academic skills were included. However, in the borders of any categories some shift is always possible, for instance, due to investigators misevaluation or persons lack of attention during testing. Testing of IQ of children under school age is not based on lingual-abstract skills; and IQ fluctuation is common during growth. In 1962, there are too few children aged 2 to 5 years according to later studies due these reasons (Hagberg 1983). Moreover, the schooling of persons with ID was less common, one third of persons with ID over seven years, had not been attending the compulsory education in 1962. In his report 1966 Ruoppila estimates, that the error in evaluating the IQ is 2.5%, but reliability of testing was high (Ruoppila 1966).

In the follow-up study in 1998 professor Ruoppila, who conducted the psychological testing in 1962, evaluated the reliability and validity of 1962 inclusion criteria (Ruoppila 2000). There was one person out of 253 re-tested persons misdiagnosed with ID (Ruoppila 2000). Persons included in the study in 1962 fulfilled the definition of ID, but among persons, who were excluded from this study with borderline or normal IQ, there are most likely persons with mild ID with good social competence. He evaluated the number of these dropouts from service records. Of them about 8% had used the services for persons with ID during the follow-up period (Ruoppila 2000). Of course, persons with the diagnosis or suspicion of ID are more likely to get labelled as intellectually disabled and for instance in rural communities this may have caused underdiagnosing of ID. The inclusion criteria used in 1962 excluded persons with ID rather than included persons without ID (Strömme 2000a). This may overemphasise the differences between the ID population and the general population, especially in the mild ID.

13.5 The prevalence of intellectual disability

In 1962 participation of a large variety of municipal officials was compulsory.

Extensive network of public health care nurses and social workers can be held as competent in identifying persons with suspicion of ID. Reporting a suspicion of ID varied, so some persons could have been left out from the first phase. The structure of municipal services and selective immigration from rural areas varied in Finland. Rural communities integrated persons with ID better, and schooling of persons with ID was also less common in these areas, resulting perhaps less frequent reports of suspected persons with mild ID. It has been also found, that low social class predisposes to mild ID (Strömme 2000b), which could increase the prevalence in rural areas in 1960s in Finland. However, the regional distribution of prevalence of ID followed the preconceptions with lower prevalence in the urban area and higher in the rural area. Prevalence varied also by age as expected.

The prevalence of ID and the distribution of its levels in the 1962 study are comparable with other subsequent population studies from same era (Balakrishnan and Wolf 1976; Slavica et al 1995). Males outnumber women in younger age groups relative to other Finnish studies (Rantakallio and von Wendt 1986; Louhiala 1995). However, the prevalence of profound ID is lower than in later studies (McDonald and MacKay 1996; Fernell 1998), but similar with the studies from same era (Rantakallio and von Wendt 1986). Mortality in profound ID was higher prior to 1962 than at present (Miller 1978; Chaney 1979).

The prevalence of persons with mild ID in this study, 2.97‰, is most likely too low. In older age groups prevalence is more reliable (Ruoppila 1966; Roeleveld 1997). The main reasons are the cut off age at 2 and the low number of children. Rantakallio and von Wendt reported prevalence of mild ID 5.6‰ at age 14 from Northern Finland (Rantakallio and von Wendt 1986). The present study did not include children less than 2 years, and so the figures are not fully comparable, but the prevalence of persons with mild ID between 10 and 14 years of age in 1962 was 6.6‰ (Amnell 1966). As the mortality of persons with ID was high throughout the life, the prevalence would not have greatly exceeded the present prevalence in 1962. If we include those persons, who were excluded in 1962, but later found intellectually disabled, the prevalence of mild ID would rise from 2.97‰ to 3.19‰ (Ruoppila 2000).

13.6 Medical examinations

Psychological tests were chosen as inclusion criteria and medical and social examination followed them. Some progressive diseases may have been missed because of this. Medical examination was contributory to the psychological testing and its quality is not adequate for analysing the quantity of risks during the follow-up period. Most aetiological and diagnostic information relayed on information from parents, caregivers or patient records (34% of cases) (Amnell 1966). This decreased the reliability of aetiologies and associated disorders. The prevalence of them is comparable to studies of same era (Amnell 1966), but low prevalence reveals more the poor diagnostic possibilities in 1960s than their reliability. In this study we used DS, epilepsy, CP, and hearing and visual impairments as background variables for

life expectancy, but the results describe direction of the additional risk, but do not quantify the magnitude of association with disorders and comorbidity. Also the mortality of persons had been higher and more selective in previous decades producing the healthy survivor effect with low frequency of associated disorders (Record and Smith 1955; Carter 1958; Collmann and Stoller 1963; Primrose 1966). Therefore the risk of adults with ID with associated disorder may be underestimated compared to the present. However, these risk factors can not be passed. In lower levels of IQ they are important predictors of survival, but in higher IQ groups other diseases overshadow them.

13.7 Identification

Identification process was complex, since in Finland, the personal identification number was not commissioned until in 1st of January in 1966. Therefore the identification process was dependent on the names and dates of birth. Personal identification number was not needed for vital status and cause of death. Since we had the original forms, the names and dates of birth of parents of persons with ID included in the study were also used in identification, if needed. It is possible that a person might have had an error in name or date of birth causing non-identification, but not a false identification. Retention rate was 97.4% and can be considered high.

13.8 Databases

The databases linked with registries have two major problems: the linkage of the study population to the registry and inaccuracies in the registry. The linkage of the study databases was done with individual identification numbers, which have been proven to be most reliable (Boussy and Scott 1993; Teppo et al 1994). For vital status and for death certificate name and date of birth were sufficient. The identity of deceased persons was rechecked during the coding of causes of death.

The mortality statistics rely on death certificates, which are based on either autopsy or clinical evaluation. It has been reported that even in just having used the main categories of ICD-9, the main cause of death was categorised wrong in 22% of cases (Cameron and McGoogan 1981a; Cameron and McGoogan 1981b). Furthermore, it appears that in the majority of the most common causes of death grouped according to ICD (neoplasms, cerebrovascular and digestive), social class gradients would be steeper if mortality data were based on pathologists' rather than clinicians' diagnoses (Samphier et al 1988). The inaccuracy of death certificates could overemphasise respiratory mortality of persons with ID as respiratory diseases are thought to be the most common cause of death in this subpopulation.

The validity and reliability of suicide statistics has also been under debate (Rockett and Thomas 1999; Lindeman et al 1995; O'Carroll 1989). There is general agreement that suicides are likely to be undercounted both for structural and for sociocultural reasons. Some true suicides are in fact certified as other than suicide. The sensitivity

with which coroners and medical examiners certify true suicides has reported to vary from approximately 55% to 99% (O´Carroll 1989). When official statistics are interpreted with a degree of caution and an understanding of the source and direction of biases likely to affect the published rates, however, it seems unlikely that major conclusions based on these statistics will be in error. In this study we examined all undetermined deaths and found that in half of them could have been determined as suicide, when patient documents were studied. In conclusion this could mean, that suicide statistics of persons with ID undercounted suicide more than in the general population. This might lead to lower estimates in our study than expected, but no reference study existed so far.

Studying the death certificates during this study confirms the fluctuation in primary causes of death given in the death certificate, since author did not always agree with the interpretation of primary course of death, which was used in the study. For instance, epilepsy was listed as contributory cause of death rather than the primary cause, the initiator of process by definition (WHO 1975), in situations, where suffocation or fall followed the epileptic seizure. Also changes in diagnostic practice was seen during 35 years, for instance in 1960s the ischemic heart disease was coded as congestive heart failure and in 1970s similar course of disease was listed rather as a coronary infarct, but the main ICD-9 class remained.

On the other hand, the cancer registration system in Finland is virtually complete (Teppo et al. 1994) and the computerised record linkages based on identity numbers are precise (Pukkala 1992). Though there is some regional variation of cancer within Finland, it has a only slight effect on incidence rates (Pukkala 1992).

13.9 Statistical methods

Since the study from 1962 was a nation representative sample, we were able to use same methods than in the studies carried in the general population. It possible to stratify the general population for every person entering the interval period together with the number of those whom died during the interval and for deceased persons to calculate the age-adjusted relative mortality rates. Specific cancer types were calculated from age and sex matched general population for sexes, all levels of ID and DS. This increased the both comparability and reliability of the results of this study.

14 Discussion of results

14.1 Life expectancy and mortality

Persons with mild ID shared a life expectancy similar to that of the general population, which is longer than previously reported (Dupont et al 1987). In younger age groups, the expected life span was slightly higher than in the general population. The life expectancy was not, most likely, greatly influenced by lower prevalence of mild ID. If more persons with ID had found, they would have been mostly children with mild ID or few old persons living in rural areas. However, persons with profound ID have a significantly decreased life expectancy in all age groups, with few individuals reaching old age. This is connected with the serious neurological deficits and more severe associated disorders, common in these groups (Kelleher and Mulcahy 1985), which have been found to be the strongest predictors of high mortality and low life expectancy (Plioplys et al 1998). Some researchers have interpreted the outcome measures of these neurological deficits, such as non-mobility and tube feeding, to be the strongest predictors of mortality (Eyman et al 1993a), but they actually reflect the severity of neurological deficits and associated disorders than are independent risk factors themselves. The problem with using outcome measures rather than considering aetiologies behind them, the lack of lifelong predictability is poor, since neurological deficits can progress or new skills may develop. Although disregarding the level of ID may simplify calculations, it must be included in studies on life expectancy.

Overall mortality in this study was comparable with service registry based study of O´Brien and Zacharia in 1998, which had similar age range (17.7 versus 16.6) (O´Brien and Zacharia 1998). It is from 1990s and it might be, that if the present study would have been carried out now, mortality would be lower in this kind population based sample with more persons with mild ID than in the service

based sample with more profoundly disabled persons. The mortality of external causes in this study was only one fifth of the previously reported from U.S., but rate of external deaths is higher there for the general population as well (Strauss et al 1998b).

The level of ID has had a great importance in evaluating the cause specific mortality. In this study, when the degree of ID changed from profound to mild, the disease mortality profile moved towards that of the general population, yet some differences remained. There are no reference studies for this finding. Accidents and other external causes, which are considered as avoidable mortality, are concentrated in persons with mild ID, who more often live in community-based residents, which has been found by other studies as well (Strauss 1998b). Mortality pattern of persons with mild ID might have shifted even more toward the general population, if inclusion criteria would have been more permissive. This would have increased the number of deaths for external causes. The normalisation concept (Wolfensberger 1972) changed the living environments of people with ID, which resulted in narrowing the difference between this group and the general population. Studies that compare the mortality of people with ID in institutions and in community settings have found a higher mortality in the community than in the institution (Staruss et al 1998a). However, avoidable mortality is more common outside the institutions, and the deinstitutionalisation following the normalisation of persons with ID has increased the risk of dying from accidents rather than increasing total mortality. Normalisation of living conditions will evidently to some extent normalise the cause-specific mortality risk. In the future, the debate over normalisation and its consequences to mortality should include an analysis of cause-specific mortality for all levels of ID. It should also include the suicide rate of persons with ID, which mainly affects persons with mild ID.

Respiratory mortality, mainly caused by pneumonia, was expectedly high and in particular linked with a low degree of ID (Chaney et al 1979), whereas in the general population, respiratory diseases tend to accumulate in old age. As the risk remained at least three-fold throughout the lives of both sexes, ID appears to contain a greater risk for respiratory death than warranted by neurological damage. There could also be a bias, since respiratory diseases are known as the most common cause of death in ID population, are therefore frequently, and perhaps mistakenly, used in clinical evaluation of cause death in all levels of ID.

14.2 Cancer

This is the first population based cohort study of ID and cancer ever published. The overall risk of cancer among persons with ID was similar to the risk in the general population, with a few exceptions. Unexpectedly Down syndrome did not carry an excess risk of cancer for the adults, contrasting with previous findings (Hasle et al. 2000). However young adults with ID had an increased risk of cancer and results suggest that the risk continue to be high during early adulthood, in line with earlier findings (Al-Saleem et al. 1998; Dexeus et al. 1988; Dieckmann et al. 1997).

People with ID smoke less than the general population. The risk of lung cancer was zero among persons with profound and severe ID, and close to average risk among persons with moderate and mild ID. Low smoking might also explain the significantly low incidence of urinary cancers. There were moderate, but significant excess in some cancer types, such as gallbladder, oesophagus and undefined gastrointestinal cancers. Persons with ID had a two-fold risk of neoplasms of the thyroid gland. This may be connected with the higher prevalence thyroid gland featuring malfunctions in ID (LaFranchi 1999). Similar, those with profound and severe ID had 3.5-fold risk of neoplasms in the nervous system. Malformations of the brain, neurological deficits and progressive destruction of neuronal networks disturbing defence mechanisms against cell damage might attribute to this increased risk (Schull and Otake 1999).

Neoplasms of the gallbladder were significantly increased among those with profound and severe ID. Two thirds of those with ID use regular medication (Hand 1994) with substances affecting the liver enzyme function, and nearly half of them are overweight (Bell and Bhate 1992) with increased risk of fatty liver. Gallstones increase the risk of gallbladder cancer (Chow et al. 1999). Pains produced by gallstones are often indefinable and sometimes remain underdiagnosed, or misdiagnosed and untreated, which heightens the risk of gallbladder cancer. The assessment of pain may be difficult (Brandt and Rosen 1998; Martinez-Cue et al. 1999) as poor cognitive function disturbs the expression of pain and the non-verbal determination of pain in persons with profound or severe ID depends on the caregiver's interpretation (McGrath et al. 1998). In addition indefinite nature of gastric pains may therefore increase the risk of neoplasms in the gallbladder and in the oesophagus too.

We found no difference in the incidence of breast cancer between women with ID and the general population. The additional risk factors for breast cancer in the general population are high dietary fat, alcohol consumption, low physical activity and hormone replacement therapy with only oestrogen (Cooper et al. 1999). Women with ID have protective factors such as low oestrogen levels (Calson and Wilson 1996) and low alcohol consumption, but also predisposing factors such as obesity and low physical activity and low frequency of breast screening. (Cowie and Fletcher 1998). Our study showed gynaecological cancer risk estimates, which are accord with general knowledge of risks for these cancers. Low sexual activity (Wingfield et al. 1994), low oestrogen levels (Calson and Wilson 1996) and short menstrual life (Schupf et al. 1997) reduce risk of cervical cancer, but overweight (Bell and Bhate 1992), poor hygiene (Wingfield et al. 1994) and low frequency of child births (Calson and Wilson 1996) increase risk of cancers of the corpus uteri and ovaries. Women with ID need same health promotion procedures with the general population such as cervical smears and mammography. The very low incidence of prostate cancer suggests that screening and other types of diagnostic activity may not be extended to persons with ID. The nature of ID may complicate the task of diagnosing cancer in these individuals. As those with ID have equal or elevated risk of developing neoplasms, cancer prevention needs to be directed at them, too.

14.3 Suicide

In this study we found 10 suicides and the SMR rate of suicide for persons with ID is descriptive. Suicide is unusual among the persons with ID, rate being one third of that in the general population. A similar ratio has been found in only study of suicide rate in ID from California (Strauss et al 1998b). Annually this counts for 3-4 suicides of persons with ID in Finland. On the other hand, suicide rate reported here, may underestimate the number of suicides, since the suicide of a person with ID causes anxiety and therefore is not recognised, but classified as an accident or undetermined death.

There are also differences in suicidal methods and sex ratio between those with ID and the general population. Methods used were categorised as passive, requiring no skills to acquire. Women committed a suicide nearly as often as men did. However, the risk factors for suicide are similar with the general population. Although in ID individuals, mental disorders are very difficult to diagnose and treat, which complicates the preventative work, threats to commit suicide still need to be taken seriously. In the present study eight out of ten suicide victims were in contact with mental health services at the time of death. This accord with the established fact, that person at risk of suicide tends to be those persons suffering from severe psychiatric problems. Most of our cases had suffered from untreated depression, though their symptoms were not always similar to depression symptoms in the general population. The social support was also minimal in all cases, diminishing the possibility of coping with anxiety and changes. During the study period institutionalised care was more common than at present, but still individuals suffering severe mental problems are still often managed as inpatients.

Six out of ten persons had previously attempted suicide before using a similar method for their final act. Obsessive-compulsory disorders are common in ID, but in 1962 none of these persons had any compulsive symptoms, such as self-injury, which would indicate additional risk of suicide. Abuse increases the risk of suicide in the general population (Birhammer et al. 1996). Persons with ID are vulnerable to abuse, which is encountered in all residential settings (Furey 1994). Communication problems - a possible additional risk of suicide - appeared in one third of persons. People with ID are more dependent on others. Ageing and loss of parents or siblings are confusing to person with ID, causing anxiety and depression. The suicide victims in this study were rather old, which may be connected to changes in their health and social environment; in four cases there was an emotional loss before the final act. High dependence on other people, communication problems, loss of family members and emotional loneliness may thus be risk factors for suicides among people with ID. Those working with people with ID need to be aware of any psychiatric disorders underlying the behavioural disturbances in order to recognise severe depression, emotional instability and lack of will to live, possibly leading to either attempted or completed suicide.

14.4 The effect of age in life expectancy and mortality

Children with profound and severe ID are more vulnerable to disease than children of normal intelligence (Plioplys et al 1998). In this study, the relative risk of death by disease was high until the age of 30, revealing the physiological fragility of children and adolescents with severe or profound ID. They have more congenital malformations than normal children (Grech and Gatt 1999), leading to a higher mortality; however, in this study, a lower rate was found than in the previous studies, because the small proportion of children with ID (Similä et al 1986). This indicates the assumption of a healthy survivor effect (i.e. persons who survive over a longer period are healthier because of selective mortality) in the older age groups of persons with ID suggested earlier by Havemann (Havemann1989). This had lead to longer life expectancy of survived and larger number of adult persons with ID, although the incidence of ID is not likely to increase. In Finland, the number of aged persons with ID will increase rapidly in next ten years.

Ageing naturally increases the risk of disease, and in this study, this risk was similar to that of the general population after the age of 40 years. This also supports the concept of a healthy survivor effect. However, some differences did exist in the cause specific mortality in this population compared with the general population. Respiratory diseases were the most common cause of death, the risk three-fold throughout the life span, whereas in the general population the risk is cumulative by age.

Ageing will lead to losses of parents or siblings and more likely to health problems. These may be confusing to persons with ID causing anxiety and depression increasing the risk of institutionalising. In this study persons committing suicide were rather old, which may be connected to influence of changes in health and social environment.

Problems of ageing will influence the health care needs of this population in the resent future. Persons with ID may have their special needs for certain diseases or symptoms, but ageing process is similar with the general population (Havemann 1989; Hogg 1993). Whether to integrate geriatric services for ID with the general population is not yet clear, but information on epidemiology of diseases among ageing persons with ID is needed for the basis of this process.

14.5 The effect of sex in life expectancy and mortality

In all levels of ID, the survival between the sexes differed less than in the general population. This finding has not been reported before. More men with ID are born, but the survival difference remained small even when sex was controlled for in the analysis. This phenomenon was first described by Carter in 1958 (Carter 1958) and confirmed in several subsequent studies (Primrose 1966; Forssman and Åkesson 1970; Strauss et al 1998b), and it is not syndrome-specific (Crichton et al 1995).

The difference in the sex distribution at birth is related to sex-linked genetic factors affecting the development of the central. Just as our knowledge of neurobiology is fast increasing, we are likely to find more syndromes with malfunctions in the network of neuronal synapses (Johnston and Harum 1999). This was also seen in this study, as survival of women stayed lower than survival of men until age 20. ID phenotypes are more often X-linked, so men outnumber women in genotypes associated with ID (Crow and Tolmie 1998), and genetic factors are magnified in consanguineous marriages (Fernell 1998). The genetic background of ID is three-fold in persons with profound and severe ID compared with mild ID, where only one-fifth of aetiologies are suggested to be genetic (Crow and Tolmie 1998). It has also been noted, that social factors make ID more conspicuous in males as they tend to more often have behavioural problems and require a longer time to acquire verbal skills (Howe et al 1998; Wilkinson 1999).

Women continue to form the majority in the ID population 25 years later than in the general population due to the different initial population structure and the survival, in which mild ID seems to be protective factor in the early decades of male life. The cause-specific mortality between sexes differed less than in the general population. The three most common diseases being the primary causes of death in this population were cardiac diseases, respiratory diseases and neoplasms for both sexes. There was an equally low risk of cardiac disease for both sexes, indicating more similar risk profiles of cardiac disease for the sexes than in the general population. Protective factors, such as non-smoking and lower blood pressure (van Schrojenstein Lantman-de Valk et al 1997), and predisposing factors, such as obesity (Rimmer et al 1994), have been reported. External causes were equally common for both sexes unlike in the general population, where men die on external causes three times more often than women (Statistics in Finland 1997). The observation that children with ID, especially boys, were exposed less to domestic accidents than their age mates of normal intelligence suggests that ID children are more protected by their parents and caregivers, which increases their life-expectancy. In the general Finnish population, men commit suicide three times more often than women (Öhberg et al 1995). In this cohort women with ID committed suicide nearly as often as men and the rate was same with the women in of the general population, as men with ID had significantly reduced rate. The similarity of external causes was the main contributory factor to similarity in the survival patterns of both sexes.

14.6 The effect of aetiology and associated disorders in life expectancy and mortality

DS was a risk factor for premature death in middle age, since the mortality rate for this population rises sharply after age 30, with very few surviving to the old age consistent with other studies (Fryers 1986; Baird and Sadovnick 1987; Eyman

et al 1990). Epilepsy is common in this population, increasing mortality in the first two decades of life, as has been reported previously (Forssman and Åkesson 1970; Sillanpää et al 1998). Hearing impairment also had a negative impact on life expectancy, which may be due to the severity of neurological damage rather than its being an independent risk factor. A more detailed evaluation would be needed to assess the effect of different aetiologies on survival and life expectancy.

15 Conclusions

1. The life expectancy is greatly influenced by the level of ID. Sex difference is smaller than in the general population, perhaps due to similar environmental risks for both sexes.

2. Persons with mild ID have same life expectancy with the general population. Profound ID decreases the life expectancy to one third of the life expectancy of the general population.

3. Improved life expectancy has changed the cause-specific mortality toward the patterns of the general population, because the number deaths of aged persons with ID have increased and environments of persons with ID have started to normalise.

4. Three most common causes of death are cardiac diseases, respiratory diseases and cancer. Respiratory diseases are three times more common and than in the general population. Cancer incidence in similar with the general population, consisting of a few types of cancer with low incidence (lung, prostate) and those with high incidence (gallbladder).

5. Main difference in cause specific mortality in comparison to the general population is the low frequency of external causes of death. Suicide rate is one third of that of the general population. However, aged persons with ID are vulnerable to fractures and their risk of external death is equal with the general population.

6. Persons with ID live longer and healthier life than before. This increases the number of aged persons with ID, but as the general population is fast ageing as well, their proportion will remain the same, 0.4%.

16 Implication for healthcare and future research

Epidemiological research serves an important role providing information on populations for planning services in particular geographic locations. It is essential in planning and prioritising the need for services of diagnostic, treatment, prevention and future research (Munro 1986).

It is evident that the life span of persons with ID has prolonged during the last three decades rapidly. This has created a new subpopulation: ageing persons with ID. In Finland the growth in a number of aged persons will increase fast in next ten years both in ID population and in the general population and need geriatric services is growing. The proportion of ID population is expected to remain constant. The normalisation of persons with ID would emphasise integrated services in the health care system. However, the specific health risks of this population, which can be seen in cause-specific mortality, would imply retaining some special structures to support and study the health issues in this population. If we totally integrate the services for persons with ID with the general population, we may loose the expertise in ID. The knowledge of normal ageing in ID population is yet poor and just starting to grow. The ageing process is likely to be similar for all people, and the knowledge of ageing in the general population must be utilised for benefit of ID population.

The need of health promotion and prevention was recognised in this study. As persons with ID move from institutions into the community, they are exposed to the similar environmental risks, including shortage of health care resources, as the general population. If we intend to decrease the avoidable mortality in this group, we need to focus specially on accident risks, on preventing infections and cardiac diseases, and include the persons with ID into health promotion in the community. At least in Finland, where health care is unlikely to receive addi-tional funding and workloads are expanding rapidly, we are

forced to choose. Fortunately, preventative work is seen to be profitable and we need to include people with ID and their unique needs in this process.

Ageing of persons with ID is becoming an urgent research question. The specific differences of normal and pathological ageing of persons with ID are under research. Studies of the pathogenesis of Alzheimer's disease in DS and ID will also benefit the ageing studies in the general population. Many functional disabilities appearing in the old age are similar with impairments in ID. There is a long tradition of multidisciplinary studies with psychologists and sociologists, which would benefit the collaborative research in geriatrics for all disciplines.

The epidemiology of ID is in constant move. We need to repeat and improve the epidemiological studies with comparable methods, which are used in the population studies. Though a small number of persons with certain rare syndromes, produce unreliable epidemiological data, but important clinical information. We are running into difficulty of defining ID thoroughly, but in this context aetiology of cognitive impairment will be more important, since diagnostic abilities are improving. Therefore ID provides endless opportunities for fascinating studies.

17 Summary

A population based, nation representative cohort of persons with ID was followed 35 years. Sufficient data was received for 97.4% of the original study population in 1997. The study cohort included 2,309 persons 1,165 (50.4%) men and 1,144 (49.6%) women with ID producing a total of 60,969 person years. The mean follow-up time was 26.9 years (SD 10.9). At the end the follow-up period 50.6% of the original study population was alive. The mean age of persons alive was 54.7 years (SD 11.9) and of deceased 57.7 years (SD 18.8), respectively. Life expectancy, cause specific mortality and cancer incidences of the cohort were compared with a sex- and age matched general population.

The life expectancy had a positive correlation with IQ. For persons with mild ID, the life expectancy did not differ from that of the general population during the first three decades of life. In the moderate ID group, the life expectancy decreased slightly and in the severe and profound ID group, it decreased substantially to 20% for all age groups.

The mortality rate was 17.7 per 1,000 person years (CI 95% 8.4-27.0). Comorbidity decreased survival as expected. Low level of ID, epilepsy, hearing impairment and DS were associated with increased mortality, but not in all age groups. There was excess disease mortality of persons with ID under 30 years of age compared with the general population, but not in the later years. Three most common causes of death were cardiovascular diseases ans respiratory diseases and cancer for both sexes. The relative disease mortality was higher for men than for women until age 40 years. Men with ID were at highest risk to die on disease before age 20, but at the same time their risk of accident was only one tenth of the risk of the general population. Ageing increased the accident risk for both sexes with higher risk for women than men.

173 patients with cancer were diagnosed; the expected number was 188 (SIR 0.9, CI 0.8-1.0). Persons with ID had non-significant increased overall risk of cancer between ages 15 and 29

for both sexes, but later the overall risk of cancer was similar with the general population. The level of ID did not have a significant correlation on overall cancer incidence, but women with profound ID had increased risk of cancer compared to men. Low IQ increased the risk of neoplasms of nervous system, testicular cancer, oesophagus, gallbladder and tumours of other gastrointestinal organs.

Ten persons committed a suicide during the follow-up. Women committed 12.4 suicides per 100 000 and men 15.5 per 100,000, while corresponding figures for general population in 1980 were 13.2 and 52.9 per 100 000. People with ID seemed to commit suicides in similar frequency in their adult life irrespective of age but with passive methods.

The life expectancy of persons with ID has improved, though is depended on the level of ID. The cause specific mortality of people with ID differs from the general population. The improved life expectancy and changes in the cause specific mortality in this study can be interpreted as a result of improved health care and social support and normalisation.

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The quality of mercy is not strain'd,
It droppeth as the gentle rain from heaven
Upon the place beneath: it is twice blest;
It blesseth him that gives and him that takes:

Act 4, Scene 1
William Shakespeare: The Merchant of Venice

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Let me play the fool:
With mirth and laughter let old wrinkles come,
And let my liver rather heat with wine
Than my heart cool with mortifying groans.

William Shakespeare:
The Merchant of Venice
Act 1, Scene 1

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19 Appendix I

General information

The Pienois-Suomi- Study (Finland in Miniature) was planned as a cross-sectional study between 1959-1960 and conducted in 1962 by the Bureau of Social Research of the Ministry of Social Affairs. 57 municipalities were selected by socio-economical range within the country to represent Finland. They were chosen in by their population, financial classification, social expenditure and occupational structure. There were 416,973 persons, which was 9.4% of the total population. There were three areas of research: medical, psychological and social studies.

A pilot- study was conducted in one municipality of the selected municipalities and based on pilot study, the age range was selected to be from 2 years to 64 years of age. The data collection of psychological and medical data took place from April 9th 1962 to January 12th 1963.

The fieldwork was performed in teams composed mostly of two psychologists and one physician. The examinations were performed in Communal Health Centres, where Public Health nurses brought the subjects. Their parents or other persons having them in their custody usually accompanied persons. All examinations were free of charge for subjects. The number of reported and examined persons by the are presented in Table 1.

There were altogether 12 psychologists, 4 Masters of Art degree and 8 Bachelor of Art degree, performing the psychological tests. The medical team was formed of 6 Licentiates in Medicine and 2 Bachelors of Medicine (non of the physicians was specialised in neurology, since the speciality of neurology was introduced in Finland in late 1960s and part of psychiatry at the time of the study). Before the fields work both psychologists and physicians were given special instructions and detailed written guidelines.

Table 1. **Reported persons for examination in 1962 for Pienois-Suomi- study, proportion per thousand in area and number of examined provinces of Finland**

Province	Reported	*‰ of whole population*	Study population[1]
Uusimaa (Southern Coast)	359	*7.74*	356
Turku ja Pori			
(South western Finland)	312	*6.48*	304
Ahvenanmaa (Åland)	69	*6.56*	68
Häme (Lake Region)	434	*10.81*	381
Kymi (Lake Region)	225	*7.85*	215
Mikkeli (Finnish hill county)	252	*13.52*	247
Pohjois-Karjala			
(Finnish hill county)	116	*12.22*	111
Kuopio (Finnish hill county)	199	*10.56*	199
Keski-Suomi			
(Finnish hill county)	353	*11.91*	349
Vaasa (Plain Ostrobothnia)	584	*8.94*	573
Oulu (Plain Ostrobothnia)	908	*11.28*	842
Lappi (Lapland)	202	*9.79*	191
Total	**4,013**	**9.62**	**3,832**

[1]Number of examined person by provivinces.

Psychological tests

A persons was defined as intellectually disabled if ones performance measured by KTK Perfor-mance Scale was more than two SD´s below the level of the mean scores of ones age group. The definition thus covers both a social criterions, being suspects of ID, and a psychological criterion. The test was selected for elimination tests, because it found with high correlation with general intelli-gence. The impairment of maturation, learning or social adjustment was only evaluated, because there were no suitable measures for adaptive behaviour at the time. As the KTK performance scale was used as inclusion tests, it was, however, supplied by the use of two vocabulary tests, the school achieve-ment tests for reading, writing, arithmetic's and rating. The ratings have covered the understanding of speech, verbal expression, emotional stability and the need or necessity of institutional care and special instruction. The time spent on testing was not reported.

One of the aims of the psychological part were to estimate the reliability of prevalence tests. Ruoppila claims, that though the performance battery worked and was able to find persons with ID accurately than the verbal tests before. The reliability of test battery was good.

There was lower prevalence if ID found in school age group. This may have reflected rare schooling of persons with ID prohibiting them to be recognised as intellectually disabled. Ruoppila says, that the main emphasis in the interpretation of the results has been directed towards the social criterion and its selective influence, because the frequency figures of mental retardation (as a synonym to ID) established in this study, fell significantly below expected on the basis of the normal distribution curve. In the IQ>65, the social competence may cover ID.

This effects most likely the prevalence of mild ID. Though the schooling was not common for persons with ID in previous decades, the rural community adjusted persons with mild ID better than more demanding urban community. Therefore some adult persons with mild ID living in the rural community were not reported to the study. Another factor influencing the prevalence is the high mortality of persons with ID. If the life expectancy in general was less than 20 years in previous three decades, the number of adult persons with ID was obviously lower than expected by the standard distribution. Altogether, prevalence of mild ID was likely to be too low.

Medical examinations

Medical examinations, defined by the study form, were performed as clinical evaluations assisted by the Public Health nurses. The form resembles the clinical evaluation form used in the clinic of neurology. Patient records were utilised when possible, but aetiological information based mostly on information from parents, caregivers or nurses. Clinical evaluation of functional systems (e.g. respiratory, circulatory, neurological) were evaluated. The classification of diseases was done by 7th revision of International Classification of Diseases (WHO 1955). Examinations took from 30 minutes to 90 minutes.

The need of care was evaluated and based on based on clinical psychological evaluation together with medical examination and social inquiry. It was divided in four groups: need of permanent institutional care, institutional care or highly supported home care, supported living at home and independent living. The medical part based on clinical examination and persons abilities to recognise the clinical finding issues always a source for error. Yet, the clinical status and anamnesis is still the base of diagnosing diseases. There were only 8 physicians, which is

Table 2. Contents of the medical form used in the Pienois-Suomi- study in 1962.

Divisions	Contents		
Cephalic indexes			
Clinical findings:	Respiratory	Urogenital	Neurological
	Circulatory	Digestive	Speech
Family history			
Pregnancy and delivery			
Child development schedule			
Diseases in childhood			
Main diagnoses	Skin manifestations	Neurological	Psychiatric symptoms
	Cranial and spinal	General finding	Other symptoms
	Down Syndrome		
Associated disorders	Sensory	Epilepsy	Neuromotor
	Other		
Self-help skills	Motorfunction	Toileting	Self-injury
	Eating	Sleep	Mysophilia
	Dressing up	Aggressiveness	

Table 3. Classification of variables in 1962 for medical history and percentage of findings

		Classification (percentage)
	Visus	Not available
		Normal
		Possible amblyopia (10.5%)
		Blindness (0.9%)
	Hearing	Not available
		Normal
		Possible hearing defect (6..3%)
		Deaf (1.2%)
	Epilepsy	No epilepsy (89.4%)
		GM
		PM
		F
		M
		X
		Not reliable information
History of convulsions		No
		Yes (70.6%)
Cerebral palsy		No
		Yes: 9 classes defined by region (9%)

a low number considering the number of examinations performed. Although all were not fully qualified, they had training prior the examinations and the study form was utilised. There was also a possibility to consult senior researchers.

The reliability of the findings of the clinical evaluation was comparable with studies conducted at the same period of time. The frequency of DS was 1 per 1000 and higher in the previous studies. Amnell suggests that, the decreased mortality may have increased the prevalence of DS. Neurological diseases were reported in higher frequency (12.8%) than before. Epilepsy was found from 10.6% and in the studies

Table 4 . Classification of variables in 1962 for need of care.

Need of care	
Institution	Minimal senso-motor functioning, no verbal communication, low self-help skills
Institution or Semi-independent	Weak motor functioning, minimal verbal communication, fairly low self-help skills
Semi-independent	Fairly good motor functioning and verbal communication, poor social skills, good self-help skills
Independent	Good social and communicative skills, good self-help skills

from same period figures were 9.3% and 7.6% in the ID populations. However, although Amnell was confident with the clinical findings, the aetiological findings were in most cases unreliable. Modern laboratory and imaging technics have improved finding the aetiologies of ID and also associated disorders. The prevalence of different diseases reveal the situation in 1960s and therefore some diseases or disorders are used as risk factors for mortality in the beginning of the follow up period, but not as explanatory variables.

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