The ageing process and the late manifestation of conditions related to the cause of congenitally deafblind adults in Denmark.

Introduction
The Information Centre on Congenital Deafblindness in Denmark conducted a survey that began collecting information in 1999. In this article, Birthe Laustrup, Director of the Information Centre, summarises the main findings of this research. The complete report is available in Danish.

The purpose of the survey
This survey has collected information about the ageing process and late manifestations of the causes of congenital deafblindness.

Who participated?
Everyone, aged 18 and over, diagnosed with congenital deafblindness, in Denmark is included in the survey - 58 individuals in total. 53 of the participants stated that the aetiology of their condition was known. The 5 without a diagnosis were all within the non-rubella group. Two thirds of the group live in residential settings for congenitally deafblind people, and the other third live with a person or people with other impairments.

What did it do?
The survey compared 26 individuals, diagnosed with congenital rubella syndrome (CRS), with 32 individuals with other aetiologies.

Terminology
The statistical data is organised as a "rubella group" and a "non-rubella group". When the term “both groups” is used, variables applicable to both the “rubella group” and the “non-rubella” groups are referred to.

The Summary Findings

Medical and physiological information
The rubella group has more males than the non-rubella group.

Weight
In both groups there is a high incidence of persons born “small for date” (born at term, but with lower birth weight than expected). In both groups there are many individuals who later in life have experienced weight problems. In the non-rubella group, there is typically a problem of undesired weight loss. This contrasts to the rubella group where it is more common to become overweight in the adult years. According to the data collected the problems of weight gain is of a scale that calls for further investigation to identify the causes of this.

Heart
Congenital heart failure primarily occurs in the rubella group. With early surgery, the heart defects pose no problems for the individuals later in life.
Motor development
If we look at the motor functions and physical impairments present at birth, these only occur in the non-rubella group. However, in both groups there are individuals who later in life experience physiological deterioration to such an extent that they need assistive devices for mobility functions. In the non- rubella group this deterioration takes place early in life, and in the rubella group only takes place from age 30 and onwards. This may indicate the presence of a late manifestation of their condition.

In both groups there is a relatively high incidence (15,5%) of muscular deterioration and poor balance in adulthood. This reinforces the requirement that congenitally deafblind persons need treatment from physiotherapists and occupational therapists during adulthood.

Vision and hearing
The majority of the participants, in both groups, have a visual impairment that was diagnosed during the first year of life.
Around two thirds of the population with congenital deafblindness have a progressive vision loss (like glaucoma, retinal detachment, decreasing visual field, and removal of an eye). We can also attribute the generally occurring age related changes in vision to much the same level in both groups. The Danish survey (also) indicates that

- persons with CRS are at high risk of developing late manifestations e.g. glaucoma
- the combination of glaucoma and cataract is specific to the diagnosis of congenital rubella syndrome.
- retinal detachment is also significantly more frequent in the rubella group. A possible trigger factor might be cataract.
- Microptalmus is present in every second individual in the rubella group and in every fourth in the non-rubella group.
- There may be a relationship between microptalmus and later development of glaucoma.

In both groups individuals have severe hearing losses or deafness (the latter is more frequent). “Moderate” hearing losses are only identified in the group non-rubella. Both groups show that the hearing impairments are identified later than the visual impairments.

Individuals in the group non-rubella are generally diagnosed earlier than individuals in the rubella group. This finding may surprise, as the congenital cataract, possibly in combination with low birth weight and heart failure should cause suspicion of a concurrent hearing loss caused by a rubella virus infection.

Hearing aids
According to our information 50 individuals have had a hearing aid at the time they were diagnosed. Today only 16 individuals still wear their hearing aids. Progressive hearing losses are identified in both groups. However, the prevalence of progressive hearing losses may be higher than shown in the data collected, as, unlike progressions in vision loss, the progression in hearing loss will not be immediately detected in observations of changes in behaviour. For this very reason it seems alarming that appropriate examinations are not being undertaken at regular intervals.

With regard to hearing and vision losses the survey records that the awareness of the risk of progressive sensory impairments is lacking in the environments where congenitally deafblind adults live. The survey also indicates that there is more awareness of the possibilities of decrease in vision. This is because every second individual is regularly taken for eye examinations, and only one in five gets the hearing checked.
Hormonal conditions
Problems with metabolism are a known late manifestation of Down syndrome, and have been mentioned in foreign studies as a late manifestation of congenital rubella syndrome as well. None of the participants in the Danish survey reported any diagnoses with regard to metabolic problems, so it may seem not to be a matter of late manifestation. Nevertheless, we must state from the data collected that 10 out of the 20 individuals in the rubella group are overweight and in the same 10 people between 2 and 6 symptoms of low metabolism (e.g. tiredness, sadness, increase in emotional instability, passivity, less stamina and weight gain) are reported.

Foreign studies demonstrate that CRS entails a major risk of developing diabetes. In our survey we identified a few cases of diabetes (3), in the rubella group only. Whether this finding is a statistical coincidence or it actually expresses a late manifestation cannot be judged from the present study.

Growth retardation has been mentioned as a specific problem in persons born with CRS. We have identified cases of growth retardation in both groups, but the prevalence is clearly higher in the rubella group.

Neuropsychological conditions
Epilepsy, according to our information occurs with almost identical frequency in both groups, but the age of diagnosis is different.

In the non-rubella group the epilepsy is diagnosed at an early age and must be considered a part of the clinical picture. The reports of individuals having epilepsy in the rubella group confirm the assumption that epilepsy is a late manifestation - likely to appear in the teenage years.

In our survey we have also asked for information on changes in neuropsychological phenomena like attention, concentration and memory. Here we find distinct differences in the two groups, as decreased ability in these three functions is remarkably more frequent in the rubella group. Our survey, which comprises all aetiologies present in our country among people with congenital deafblindness, shows that reductions in memory and attention are late manifestations significant to the diagnosis of CRS. A number of individuals in the non-rubella group also report increasing problems with concentration, thus the difference between the two groups is less significant.

Development of behaviour problems
In our survey we have asked for information on changes in behaviour in 11 different areas. They are: self stimulation, motor agitation, self abusive behaviour, aggression, tolerance to changes in routine, levels of confusion, restlessness, increased impulsivity, initiative, ability to start an activity, difficulties in changing from one activity to another.

In six of the areas there is a dominant score from individuals with CRS.
• Self-stimulating behaviour, motor agitation and increased self-abusive behaviour are three types of behaviour which primarily are identified in the rubella group. This tendency seems to confirm the hypotheses of a late manifestation with respect to an increase in these types of behaviour. There is a significant difference between the two groups.
• Increased aggression, less tolerance to changes in routineregus and increased level of confusion are three types of behaviour which have previously been considered as late manifestations of CRS. These behaviours are relatively frequent in both groups, however, with the difference that aggression and less tolerance to changes in routines are much
more significant in the rubella group, which could indicate a connection with the original diagnosis.
There is also the possible explanation that these three types of behaviour express general deprivation in the congenitally deafblind individuals, or are related to early ageing processes. We know from gerontology that the behaviours mentioned may also be observed in the normal ageing process.

- Increased restlessness and increased impulsivity are described as late manifestations of CRS in previous foreign studies. Our study cannot confirm this. According to our data increased restlessness and increased impulsivity is reported in both groups, but not extensively. These behaviours are also known from gerontology studies.

- Less initiative, a fall in ability to start activity, as well as more difficulties in changing from one activity to another are also behaviours which previous studies have characterised as late manifestations of CRS. However, none of these types of behaviour seem to be characteristic of any of the two groups in our study. Despite a small minority of the rubella group showing these behaviours our data cannot confirm previous assumptions. As these behaviours are also described in gerontology, they may be indicators of early ageing processes.

Among people who work with congenitally deafblind people it has often been discussed whether these types of problem behaviour could be explained by environmental factors. We have therefore tried to identify possible common features of each person and their environment when the focused behaviours occur. None of them seems to depend on whether the setting is deafblind specific or not. Nor can we state a correlation with the individual's general level of functioning or ability to communicate.

**Psychological state and psycho-socially conditioned factors**
In total the study investigated 11 types of behaviour: emotional withdrawal, increased separation anxiety at social withdrawal, frequent waves of sobs and increased passivity, less energy, depression, emotional instability, lowered frustration threshold, sleeping disorders, sleeps only a little and circadian problems

Two of these are reported as low frequency for both groups. Five of the remaining nine are recorded with moderate frequency in the behaviours scored, with much the same level in the two groups. Four variables have scores in both groups, but are clearly higher for the rubella group.

- Emotional withdrawal and increased separation anxiety are, according to the findings in our study, not a particular problem among congenitally deafblind adults.
- If, however, social withdrawal, more frequent waves of sobs and increased passivity, are behaviours that are seen in one in five congenitally deafblind persons, with nearly the same occurrence in both groups. As these behaviours also are known in gerontology, they may be tokens of early ageing.
- Less energy seems to be a problem to every third congenitally deafblind person in both groups. We cannot find a clear explanation to this. Could it be the lifelong deprivation caused by the deafblindness? Is this a sign of early ageing? Could the explanation be something so simple as too little exercise?
The risk of developing a psychiatric disease is relatively high in both groups. The only difference between the groups, which can be stated, is that the disease is diagnosed at an earlier age in the non-rubella group.

- Depression is the most frequent diagnoses in both groups. Is this caused by a permanent state of deprivation or is it due to early ageing?

- Emotional instability occurs more frequently in the rubella group than in the non-rubella group. The survey reports it in every second individual with CRS and one in four of the deafblind persons with other aetiologies.

- The same is reflected in the findings related to a lowered frustration threshold; again the problem is present in every second individual with CRS and in one in four in the non-rubella group. Is this evidence of late manifestation in the rubella group? Or equally is it a symptom of early ageing?

- Sleeping disorders, sleeps only a little and circadian problems reveals a significant difference between the two groups. Every second individual in the rubella group experiences considerable sleeping disorders, whilst in the non-rubella group we speak of one in six. The absence of residual vision affects the individuals in the rubella group most. In this group the data indicates a significant correlation between early infection during gestation and severe sleeping disorders later in life.

The individuals among whom we have rated all these problem behaviours constitute a representative section of the congenitally rubella group of the deafblind adult population in Denmark that lives in sheltered homes.

Viewed from the level of functioning the group studied is also highly representative, apart from those individuals who manifested sleeping disorders. They are basically persons who, to a certain extent, are able to communicate in a linguistic way with their environments.

Concluding comments
The changes in behaviour observed in the rubella group that are significantly different from the non-rubella group might indicate rubella-specific late manifestation of the syndrome. Changes of neurological or physiological character take place in some of the organs of these individuals, which result in the onset of problems at somatic, behavioural and psychosocial levels in adulthood. Some indications may point to earlier ageing in the rubella group than in other people.

When facing changes in behaviour we must question whether internal factors, external factors or a combination of both are the trigger of the changes observed. At first it must be clarified whether the cause could be an unknown somatic condition, a neurological condition or decrease in hearing and/or vision. Attention must be paid to the sudden or gradual character of the changes observed. A neurological and psychological assessment is also important, and the individuals' interaction with their environments must be observed and analysed as a part of the total evaluation of the persons' situation.

We recommend annual health checks for all persons with congenital rubella syndrome. In Appendix 2 of the full report there is a guiding checklist, which may be presented to the physician examining the person with CRS. Also, we again stress the importance of regular appointments for examinations of hearing and vision. In general, there is a need for far more attention to be paid to the risk of deterioration in the two main distance senses for the deafblind population as a whole.
If we look at how quickly changes take place in the two groups studied with respect to the variables scored, it appears that within the two-year period of our survey the positives occur almost exclusively in the rubella group (reported in Appendix 3 of the full report). If this is a valid tendency we can estimate that within five years the differences between the two groups compared will be even more important.

**Future work**

The Information Centre on Congenital Deafblindness recommends that this survey should be followed up by a quantitative study of the same group of individuals starting no later than 2006. Until then, qualitatively oriented interviews may contribute to the development of the existing knowledge in identifying useful details from selected participants. Experience-based development is still needed in this very important area and there is evidence to suggest that there is a clear advantage when this work is combined with medical research. Systematic information of this kind would be highly valuable to the clinical work in our field and considerably contribute to an increased quality of life for the individual with congenital deafblindness.