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## Development of an instrument for early detection of dementia in people with Down syndrome

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## **Abstract**

Detection of early signs of dementia in people with Down syndrome could form a basis for early support and for drug treatment. We have developed an Interview instrument and a Test instrument as well as a Frame for diagnosis to combine the findings of interview and test with the demands on diagnosis in ICD-10, DSM-IV and NINC-ADR-DAS. From a large number of tests and interview questions developed here, only those giving the biggest differences between 9 persons in 3 groups of different levels of intellectual disability and estimated dementia were kept. The combination of test and interview is specific, has the potential to detect early signs of dementia, and will be feasible to use in practice.

# Introduction

The longevity in people with Down syndrome (DS) has steadily increased in the western world, over the past 50 years. Presently the average life span is 57 years, which coincides in time (54 years of age) when half of the group has developed DS dementia (Prasher & Krishnan, 1993). According to Prasher and Corbett (1993) the expected lifetime from onset of dementia is on average two years with a maximum life expectancy of five years for people with DS.

There is biological similarity between Alzheimer disease (AD) and DS dementia, due to beta amyloid plaques on the nerve cells. The disease process is developing gradually for 20 years or more (Haxby & Schapiro, 1992). When the persons are in their 5th or 6th decade the development of the symptoms increase more rapidly and the disease gives so grave reductions in the person's abilities that they are designated as having dementia. The same medical treatment that people without intellectual disability with AD receives and will receive in the future should also benefit people with DS if given at an early stage, 30–40 years of age. Nelson, Lott, Touchette, Satz and D'Elia (1995) found a change in DS persons in their middle ages that they related to AD. Not all develop dementia, there are individuals that have died in their 8th decade without signs of dementia, so treatment should not be given to all, diagnosis of the individual is necessary.

It is vital with a correct diagnosis that discriminates between mental disability and dementia; it gives better understanding of the person's behaviour. It is also vital with a correct differential diagnosis to rule out treatable conditions like thyroid deficiency, loss in visual and auditive ability, grief, depression and secondary effects of medication. A diagnosis of dementia among people with DS solely based on Diagnostic Standard Manual fourth edition (DSM-IV) (American Psychiatric Association, 1994), has not been efficient (Oliver, 1999). Aylward, Burt, Lai and Dalton (1997) give suggestions for a diagnosis based on DSM-IV and International

Classification of Diseases (ICD-10) (World Health Organization, 1992). They conclude that presently there is no mental status exam or neuropsychological instrument that can assess the presence of dementia in an individual with intellectual disability based on a single testing. Burt and Aylward (1998) present a test battery, but many of the tests are not designed to be used with mentally retarded persons with dementia as there are not many such tests in the field. A short test, Test for Severe Impairment (Albert & Cohen, 1992) is often used and quick to administer with reasonable reliability (Cosgrave et al., 1998). It can be successfully used with individuals who are severely impaired, but may not be sensitive to the earliest signs of dementia in individuals who are mildly retarded (Burt & Aylward, 1998). Here, we present a modification of the DSM-IV and of the ICD-10 diagnosis models that amalgamates information from a test, with an emphasis on problems with memory and learning that can give an opportunity for indications of dementia after a single testing session, and an interview instrument based on an informant report. The attempt to measure the importance of working memory, cognitive support and interference in learning as indications of dementia at a single testing session we consider to be a new contribution. Persons remembering as little as one item and that accept a test situation can be tested. There are tests in the battery without roof effects. The combination of test and interview is specific, has the ability to detect early signs of dementia and is feasible to use in practice.

## **Purpose of this pilot study**

It is here believed that medical treatment will evolve for persons with AD. The instruments described here are made to detect signs of DS dementia in a sub-clinical phase, in order to give a person access to treatment and to follow him or her through mid-stage dementia.

# Material and Methods

## Subjects

For this pilot investigation, nine persons with DS were chosen. The subjects were on three levels of intellectual disability and three levels of estimated dementia (hereafter called reduction) and they lived in group homes.

(Table 1).

The professional teams at Vuxenhabiliteringen, Uppsala, estimated the person's level of intellectual disability. First persons in three different age groups were singled out by the professional teams, then interviews with informants (see below) were conducted that gave an idea of how the persons fitted into the design. The vision of an individual was determined with a standardised test, LH-test (Hyvärinen, Nasanen & Laurinen, 1980). The Ethical committee of the medical faculty, Uppsala University, ethically approved the study. The subjects' participation was approved by their trustee and by themselves after being informed of the purpose, method, and the hours needed for testing. If they were unable to understand this information, the subjects were observed during testing to see if they disliked the situation, if so the test would have been terminated.

## Interview instrument

The Interview instrument comprised personal data and questions on lability, irritability, apathy and coarsened social behaviour to cover criteria of dementia according to ICD-10 (World Health Organization, 1992).

## Abilities in the Interview instrument

The main part of the Interview instrument was organised into three levels: domains, abilities and aspects on abilities, all considered being sensitive to dementia. There were 22 different abilities in 5 domains to cover criteria of dementia according to American Psychiatric Association, 1994 and an estimation of level of intellectual disability, see below.

1. Receptive abilities; the person's understanding of spoken language and of symbols.
2. Expressive abilities; the person's ability to make himself or herself understood, to explain what happens, to name objects, the person's writing, and ability in sign language. Reduced verbal output was considered as an early sign of dementia (Lai & Williams, 1989).
3. Temporal understanding; the person's understanding of time schedules, his or her ability to tell the time on the watch, and to follow a program on TV. Temporal disorientation was considered as an early sign of dementia (Lai & Williams, 1989).
4. Activities of daily living (ADL) were considered to be abilities in the realm of room cognition. They were: the person's ability to learn to find his or her way, the size of the area in which he or she moves freely, his or her drawing ability, ability to lay the table, his or her eating manners, hygiene, and ability to dress (Evenhuis, 1990).
5. Memory and Executive function; the person's ability to remember what he or she had eaten the last days, the person's ability to remember what is essential to him or her, his or her power of initiative, ability to plan, and to remember his or her plans. They were modified from Lezak (1995). Memory impairment is a major characteristic of dementia (American Psychiatric Association, 1994).

### **Items covering aspects of every ability**

There was a need to know more about changes in every ability, the following aspects are intended to shed light on how learning works in every ability and how the person perceives himself or herself in every ability and on vitality in every ability.

To quantify the abilities they were measured on a 6-level. From the following aspects it was questioned whether the person had changed:

1. First a global question on change in ability.

2. Then about the person's ability to learn in the ability.
3. To adapt to changes concerning the ability.
4. The level of needed support in the ability. To learn, capacity to adapt to changes and level of needed support are here considered to be aspects of learning. Reduced capacity to learn is a diagnostic feature of dementia (American Psychiatric Association, 1994).
5. The person's insight in his or her level of functioning in the ability (aspect of insight). Persons in early dementia do not have insight in their reductions and that is of clinical importance according to Knight and Godfrey (1995).
6. The person's perception of being inferior to others due to low performance in the ability (aspect of depression).
7. His or her mental presence performing the ability, his or her pleasure in life, liveliness, apathy and slowness performing in the ability (aspects of vitality).

Later three aspects were excluded from further analysis: Insight in level of functioning as the informants reported almost no insight on behalf of the persons. Depression as perception of being inferior to others was excluded because the subjects were not reported to feel inferior, maybe because of lack of insight or because the symptoms of depression in mentally retarded persons are more often irritation. The aspect apathy was excluded, because the informants for the persons in the Possible Reduction group gave few answers. A hypothesis is that informants might have felt a chill: a person they like might have Down syndrome dementia, and so the informant mentally turned away from answering the question. The question may be important but difficult to handle.

In the dementia of Gunnel Boman the first signs were occasional lapses, moments of reduced ability that came more often and lasted longer (Boman, 1990). When the informant described deterioration of ability, it was possible to draw a diagram illustrating the time between two lapses, including the level of ability and how it is reduced over time and what triggers the lapses. The informants were also asked for comments throughout the interview.

## **Estimating levels of intellectual disability**

The person's level of mental retardation has been estimated by criteria of Kylén (1974) on communicative abilities, understanding and using symbols, temporal and spatial ability. Kebbon, et al. (1992) developed scales on temporal and spatial ability and the scales have been further developed here.

Level of intellectual disability was estimated for the abilities to understand symbols, to make oneself understood with words, the person's ability to write, to tell the time on the watch, to understand time schedules, to learn to find his or her way and the scale for the size of the area in which he or she moves freely. 6-level scales were used.

## **Test Instrument**

The Test instrument was made to measure memory, learning and other cognitive abilities sensitive to early signs of dementia.

## **Construction**

Construction is a memory test in the form of stacking rings to make a pyramid (test material: Mula, IKEA, Sweden). The test procedure was that the person with DS on his own stick rebuilt the test leader's stacked rings, to form a pyramid. Either with the test leader's stacked rings out of sight as a test of memory, or, if the person did not manage, with them in sight as a task of perception and spatial ability. The person's learning was interfered in four ways, looking at cards, repeating words, clapping hands and a 30 second pause in silence. Ageing persons are sensitive to interference between learning and recall (Mayes, 1995), and so are persons in dementia. Rebuilding the stacked rings upside down tested spatial working memory. During the testing of stacking rings it was observed how wrong solutions were solved. The test has the ability to test persons from super able to severely disabled. There is a possibility to avert floor effects using the test material to observe the person's constructive play with it and his or her interaction with the test leader.

## **Verbal tests using objects known and named**

Aphasia, agnosia and apraxi were measured in a pre-test called Naming. The test person is asked to name, point at and show the use of 30 everyday objects. Objects successfully named by the person were used in the following tests:

1. Objects To Be Remembered, a learning test with the everyday objects, immediate recall and 15 seconds delay, testing visual and auditory input and vocal and motor output. Two forms were tried, with few and all objects on the table.
2. Auditive Learning with words for objects known from the test Naming. Auditive Learning tested short-term memory, delayed recall and working memory. Repeating backwards tested verbal working memory.
3. Visual Learning was tried as an alternative to Objects To Be Remembered with the same objects but using vocal and motor inputs and outputs simultaneously, with 30 seconds delayed recall and with working memory with the objects visible on presentation but not on recall.
4. Supported Learning, a test of the person's ability to profit from cognitive support. The supporting conditions used here are to include the words to be remembered in a tale, to learn them in similar sounding pairs and to learn them in conceptually similar pairs.
5. Sensing Items Inside A Bag tested agnosia.
6. Where Did I Put It, a memory test where the test person is requested to remember in which of several envelopes he or she puts items.

For those who do not need objects to understand, we developed an alternative to test the person's learning with digits and numbers, on auditive short-term memory, delayed recall and working memory.

## **Indications of reduction at a single testing session**

We want an indicator of dementia and an indicator of a sub clinical phase of dementia. Short-term memory measured as immediate verbal memory tend to show little change

with ageing but is sensitive to dementia (Haxby & Schapiro, 1992), so decline in short-term memory can function as an indicator of dementia. Also extremely low short-term memory with a person known to function in the moderate or mild levels of intellectual disability should be a sign of a need to check if the person is in a dementing process. As an indicator of a sub clinical phase of dementia we looked for a comparison between short-term memory and three conditions of learning more sensitive to dementia.

1. As known a person in dementia is easily distracted. When a person's learning is interfered, or recall is delayed, the non-demented person learns by repetition, but for the person with dementia the memory deteriorates.
2. Baddeley, Bressi, Della Sala, Logie and Spinnler (1991) found AD patients impaired when working concurrently on two working memory tasks, as an aspect of executive function. Here, one working memory task is tried, as an adequate level of difficulty for an intellectually disabled person.
3. Hippocampus is vital for learning and is according to Schapiro and Rapoport (1988) affected in DS dementia. This suggests the possibility to increase the sensitivity of learning by separating impressive stimuli and the mode of expressing the answer as either vocal or motor, as it increases the vulnerability of the learning process. This is done for a verbal test, Objects To Be Remembered.

## **Tests of cognitive abilities**

The other tests of cognitive abilities were:

1. Understanding Pictures
2. Simplified Arithmetic
3. Telling Time On A Clock (corresponds to Tell the time on the watch in the Interview Instrument)
4. Ability To Estimate Time Normally Used, in various activities
5. Understanding Causation In What Happens First And After
6. Drawing Ability

7. Proper Prepositions, by placing objects in relation to a box using the proper prepositions
8. Copying Pictures With And Without The Original, to see if the objects were kept touching each other
9. Agnosia—What Did You Draw
10. Word Fluency
11. Routine Decisions
12. Understanding Of Reverse Order, and
13. To Arrange A Coffee Break.

## **Questions to the test person on his or her long-term memory**

1. Questions on the person's biography
2. Memory of the last days
3. Past and present friends and staff in the apartment and the occupational setting
4. Semantic memory, and
5. Prospective memory, by remembering promised activities in the breaks during testing.

## **Data Collection Procedure**

### **Persons interviewed**

The trustee chose the person to be interviewed (informant) as the one closest to the subject. Most of the informants were staff in the subject's home that had known the subject for many years.

### **Test procedure**

The Test Instrument was divided in four tests. The subject was tested 1.5 to 2 hours each time with a pause every 30–45 minutes. At the first occasion there was an introduction with the person with DS.

## Analytical frame for diagnosis

The data from the Interview instrument and the information from the Test instrument needed to be put into a conceptual framework to form a tentative diagnosis. The models for diagnosis available are the DSM-IV, the ICD-10 and NINCDS & ADRDA (McKhann, G. et al., 1984) Criteria for Alzheimer's Disease

The National Institute of Neurological and Communicative Disorders and Stroke and the Alzheimer's Disease and Related Disorders Associations (NINCDS & ADRDA) in 1984 (see *McKhann, G. et al., 1984*)

the NINCDS-ADRDA. All three include cognitive deficits, DSM-IV and ICD-10 specify symptoms of memory deficits, short term memory and deteriorating learning ability as necessary. The NINCDS-ADRDA contribute levels of certainty in the diagnosis, possible and probable dementia. The ICD-10 is including personality traits, irritability, apathy, lability and coarsened social behaviour with signs in either of them pointing to a diagnosis of dementia. The DSM-IV diagnosis has its strength in describing cognitive functions, deficits in one of aphasia (impressive and expressive), agnosia, apraxia or executive function. The criteria were adjusted to fit the living conditions of persons with mental retardation. Executive function was estimated as ability to stick to personal priorities, initiative, planning capacity and ability to remember one's planning.

In order to define reduction as dementia there should be gradual onset of reductions in abilities lasting more than 6 months, here it is estimated with a retrospective measure. There should also be an estimate of pre morbid and present personality functioning. There should be absence of or treated thyroid deficiency, grief, secondary effects of medication, visual and hearing loss, and depression. According to DSM-IV demands for the normal population, the cognitive deficits cause significant impairment in social or occupational functioning, here the deficits should cause changes in what the person does in his or her occupation and cause increases in the support he or she receives in his or her daily living. The Interview instrument can be used retrospectively, even though that may not be so reliable.

# Results and Discussion

## Interview Instrument

### Subscales dementia, depression and common personality changes

The difference between the groups with and without reductions was biggest for questions on reduced work performance, memory disturbance, spontaneous crying, irritability, slowness, loss of judgement, and lack of energy.

### The aspects in the Interview instrument and the 6-level scale

To practically apply an instrument it is important that there are limits assigning an individual's test result as belonging to one group. There are clear limits between the groups No Reduction, Possible Reduction and Reduction in the matrix (Table 2) for the first five aspects.

Table 2

The aspects are arranged in the order they discriminate between the groups with different levels of reduction. There are no reductions in the No Reduction group, 11 reductions in the Possible Reduction group and for the Reduction group 51 reductions and no improvement. A fruitful hypothesis is that the matrix gives an opportunity to assign an individual's value to a diagnostic group, independent of his or her level of mental retardation. Cross validation and a larger number of individuals are necessary before this can be taken as a fact.

The 6-level scales registered changes on all levels of retardation, more for profoundly retarded (20.5) than for moderately (14) or mildly retarded (11.5). The informants should be asked on the person's level of performance on good and bad days as this better expresses the reality of a person, than present performance and performance two years ago. Retrospective reports tend to be unreliable. Therefore the

demand of the DSM-IV diagnosis on reductions lasting more than 6 months should be accounted for by interviewing on two occasions.

## **The diagram**

The diagram was used when the informant described deterioration of an ability. There were two parallel types of reduction, first that abilities deteriorated slowly over long periods of time, second that performance deteriorated during short periods from ten minutes to a day or during long periods from a week to a month. The periods of bad performance were related to change of staff, disliked activities, increased demands, new surroundings and the like, reducing the person's motivation rather than his or her ability. The periods of bad performance should be sensitive to treatment with positive reinforcement and to social reinforces. One person dismissed from the study probably had a psychotic process giving dramatic bad episodes. The informants should in a screening be asked if there are occasional drops or lapses of function and if so, be asked to do the diagram. The possibility to find these early changes probably depend on staff education, here the informants said there probably had been lapses but they had passed unnoticed.

## **Effects of reduction versus mental disability**

The instrument is meant to be specifically sensitive for persons with reductions. Therefore there should be no difference of performance between the groups of different mental disability, but large differences between the groups of different levels of reduction. The groups differed substantially in terms of reduction as expected, but while persons with moderate and severe intellectual disability performed equally, the mild mental disability group performed much better than the others. This may be explained by the fact that a person with mild mental disabilities should learn new abilities easier and therefore more often improve than more disabled persons.

## **Abilities in the Interview Instrument**

All results were added for the aspects Change, Learning, Need of support, Mental presence and Slowness, to see the differences between the groups No reduction, Possible reduction and Reduction for the 22 abilities. We drew the conclusion that the abilities differentiate in the desired way with more reductions for the persons supposed to be in dementia, except for questions on cognitions of time and space. The unsuccessful abilities are those not being trained or experienced in the group homes. The successful abilities probably guide the informants' thoughts to situations they have experienced with the person; they are to be kept for a new shorter interview instrument (Table 3).

### Table 3

The size of change on the 6-level scales are small, underlining the impression of slow reductions, except for Initiative. It is of value to describe the size of the reduction for abilities in all domains, therefore also some not so sensitive 6-level scales are kept.

In addition to the questions in Table 3, the ability To follow a program on TV will be incorporated to have an ability on attention and Remembering dinners the last days to have an ability on memory. In a new instrument a question on near memory will be added, "how many objects can the person remember when walking from one room to another to get them?" The abilities The area in which he or she moves freely, Writing, Understanding schedules and Tell the time on a watch will appear only with 6-level scales to give an estimate of level of intellectual disability. To learn to find one's way will also be used for intellectual disability estimations.

On Learning the informants were first asked if they had tried to teach the person new skills the last year, if they had not, the question on the person's ability to learn was not asked. The consequence is attrition on the question, this attrition is large in the group Possible reduction while the groups No reduction and Reduction are lower and on the same level. The explanation can be that persons in the Possible reduction group experience new learning as a burden, it is difficult, not stimulating, and they forget (possibly due to sub-clinical dementia), so their staffs stop training them on new skills. The ranking of the individuals on this attrition is found in Table 4.

## **Test Instrument**

### **Introduction and conclusion of the test**

The first test opportunity contained mutual presentations, orientations to person, room and time, and questions around the testing. The persons did not know why we wanted to test them. They agreed on being tested if it did not take too long, did not interfere with their interests and could be of use to others in the future.

### **General impression**

On all the tests on short-term memory (direct recall in Construction, Objects To Be Remembered, Visual Memory and Auditive Memory) the persons in the group Reduction remembered few items with direct recall. This confirms the results by Schapiro and Rapoport (1988), that short-term memory is reduced in dementia.

### **Construction**

The result was largely as expected with the learning of the No Reduction group improved or not changed when disturbed, the Possible Reduction group either unchanged or lowered and the Reduction group lowered. Construction is probably a memory test that is not so dependent on intelligence since moderately intellectually disabled persons performed as well as mildly disabled. Two adaptations in the memory test can be made for a person with a severe intellectual disability: First, place the sticks close together if he or she needs to look between the test leader's stick and his or her own to remember (if the two sticks are placed too close together it will rather be a task of perception and of motor ability). Second, observe the test persons spontaneous interaction with the test material and his or her ability to imitate the test leader.

### **Verbal tests using objects known and named**

The test Objects To Be Remembered was given in four different forms, the person was shown or told what he or she should remember, and he or she answered by pointing or telling. The differences in performance were on the expressive side, a vocal answer being easier than pointing. Differences were obtained with the most competent and least demented, the demented could only respond with none or one word, a floor effect. Testing different inputs and outputs can probably be used to determine learning options and detecting difficulties. Two forms were tried, with few and all objects on the table with no definitive advantage with few objects, but the test leader then felt less uneasy about the understanding of the persons tested.

The test Visual Memory is intended as an alternative where the person is shown the same objects as in Objects To Be Remembered. Here, the test leader and test person both point and speak. The response forms are immediate recall, recall after a 30 second pause, relearning for those who failed delayed recall, and working memory with a pre-test. A non-demented person is supposed to use the pause for repetition but the person with dementia will probably be disturbed. The differences between direct recall and delayed recall is bigger here and with Auditive Memory (30 seconds delay) than with Objects To Be Remembered (15 seconds delay). It could be that a pause of 15 seconds is too short to erase the memory. Relearning gives an uneven result compared to direct recall and is perceived as redoing the same thing over and over. The pre-test on working memory should be given to all persons that will do the working memory test in order to give the test of working memory a better capacity to differentiate on lower levels.

A valuable variant is Supported Learning; as the persons were only able to remember few objects it was easier to assess the difference between the groups on not profiting from support rather than in disturbing the learning. To support the learning helped all the persons. Those not in a process of reduction profited more from the support, which increased the difference between the groups. Person 2 was by chance tested twice on parts of supported learning and the performance varied with only one item, here her best performance was recorded. Her total strength of memory varied less

than in what category she remembered more or less. Supported learning can in the future be used with short-term memory as the one side to compare the more sensitive conditions with, as the support functions also for persons in early dementia.

What differences appear if the objects are seen both at presentation and recall, covered at recall or only mentioned and not seen? What happens when the person is tired? This was tried with person 5 on Supported Learning. Out of 12 objects she remembered on average 10 when they were seen at presentation and recall, 7 objects when covered at recall, 4.5 when they were only mentioned, and 3.2 objects when she was tired. The conclusions are that the test should not be given when the person is tired and it can be made easier for those who score close to the “floor”.

Where Did I Put It was also a functional test of learning, more lengthy though than Objects To Be Remembered and has therefore been excluded. Sensing Things In A Bag was a functional test of agnosia. When the person cannot name an object he or she should be asked what it is used for. If he or she then manages to remember the name of the object, he or she has not problems with agnosia but with retrieving the memory.

## **Supplementary cognitive tests**

The tests most sensitive to change were: Word Fluency, Telling Time On A Clock, Understanding Pictures, Simplified Arithmetic and Proper Prepositions. The pre-tests on understanding reverse order were sensitive to dementia, but not level of intellectual disability for a person with mild or moderate intellectual disability. These are saved for the shorter form of the Test Instrument. The ranking of individuals on cognitive tests are given in Table 4. As Understanding Pictures and the pre-test Naming do not differentiate between individuals on this level of reductions (person 7 was not tested), they can be kept to measure reductions later in a process of dementia. The difficulties of the test persons on the test Routine Decisions were big, may be much younger persons, 10–20 years of age, could be tested.

Copying Pictures With And Without The Original, to see if the objects were kept touching each other, might have been more sensitive to level of intellectual disability than desirable, as person 2, with an excellent memory and moderate level of intellectual disability, did not do so well.

## **Summary of Test Instrument**

The tests using objects named in the pre-test Naming and tests of cognitive abilities such as Understanding Pictures, Simplified Arithmetic, and Proper Prepositions have ecological validity, and can be used when planning treatment as they tap important everyday abilities. Tests on verbal and spatial learning, distractibility, working memory, supported learning, and understanding of time and numbers were tested and found useful for finding early signs of a process of dementia. Understanding Of Pictures is a short comprehensive test that can be used later in a process of dementia.

## **Analytical frame for diagnosis**

### **Different measures of memory from the Interview and Test instrument**

The rankings on different measures of memory should overlap and there is considerable overlap. The ranking of individuals on memory in the Test instrument follow the ranking of individuals on the cognitive tests in the Test instrument. For memory on the Test instrument there is an interaction between reduction and level of intellectual disability, as for other cognitive tests. An advantage with questions to the persons themselves was that it gave greater variation than any other measure, from almost 50 words to 1. The severely retarded persons' memories were often not verbal in character but of recognition of persons and places, so they had difficulty reporting verbally. There are roof effects in the Interview instrument on memory. The question to the informants

was whether the person with DS remembers dinners the last days. The test on the other hand gives a quantity of performance, to remember many things gives credit.

## **Rankings of individuals on diagnostic tools**

To evaluate different systems for diagnosis the individuals' rankings were compared (Table 4). In the DSM-IV criteria for dementia, agnosia is not included, as agnosia is measured in the Test instrument and can only be obtained by testing twice.

Table 4

The design and the first five rankings have the informants as source. The near perfect match between the design and the rankings of aspects in Table 2 and DSM-IV adds to their accountability.

ICD-10 is significantly correlated to both design and age and thus seems to be a sound measure. The cognitive tests, memory from the Test instrument and Objects To Be Remembered are significantly correlated to age. The conditions that Indications of reduction at a single testing session was intended to measure, they seem to measure better.

The fact that the personality questions from ICD-10, cognitive tests, memory from the Test instrument and Objects To Be Remembered are significantly correlated to age, can indicate that they give us an early hint of a developing DS dementia.

## **Conclusions concerning the assessments of persons**

According to the ICD-10 criteria of personality deterioration for dementia, one of emotional lability, irritability, apathy, or coarsened social behaviour should have deteriorated within the two last years. Person 7 meets close to all 4 criteria, person 9 meets 2 and person 8 no criterion. Person 5 in the possible reduction group meets 2 criteria. To be regarded as in dementia they should also have deteriorated memory. Person 7 most certainly will be regarded as in dementia, ranking low on memory in the test and Objects To Be Remembered. Person 9 is also in a process of deterioration,

meeting the criterion of reduced memory in the Interview instrument. What is said of persons 7 and 9 is confirmed by DSM-IV. According to DSM-IV person 8 is in dementia and he is among the three lowest on memory in the test. To assign a person a diagnosis following DSM-IV when ICD-10 is not applicable should be correct, considering that early signs of dementia could be in either personality or abilities, but always also with memory and learning. Person 2 has no memory deficit in the Test instrument and is here not regarded as having dementia. Concerning person 6 there is no guidance in table 4, he ranks 6 on the aspects, DSM-IV and 5 on ICD-10 and could not be tested. No one else is close to a diagnosis of dementia. It is noteworthy however that those closest to persons 4 and 5 have stopped teaching them new skills.

## **Concluding remarks and future use of instruments**

This report illustrates the possibility to design functional instruments for detection of reduction in a sub-clinical phase. By detecting signs of dementia in people with DS 20-40 years before a clinical dementia, treatment could be possible (Edman & Wallin, 1998). The medical treatment that people with AD without mental retardation receives, for a better quality of life, should also benefit people with DS if given in the sub clinical phase. It adds to the validity of the content of Interview and Test instruments that it can be used with DSM-IV and ICD-10.

The Test instrument can be used in a very short version for repeated screenings, with one fifth of the questions from the tests of cognitive abilities and using one instead of four disturbing conditions in the verbal and spatial tests. In clinical use there is a need for a verbal and a spatial method for indications of dementia at a single testing session. Client groups with varying aetiology and diagnosis can be evaluated with the Test instrument in its original shape. Different parts can be used to illuminate different needs of the individual: susceptibility to disturbance, range of working

memory, various cognitive abilities etc. The instruments can be used to further illuminate what dementia is.

The 6-level scale in the Interview instrument gives a format to reductions, not only the hard core abilities, but also the widening bad episodes. The diagram in an informant's report gives the frequency, duration and intensity of the bad episodes. The abilities are slowly declining over a long time period, months or years; during that long period there are many short episodes of very reduced performance lasting from minutes to several days. Analysis of the good and bad episodes gives an understanding of the expression of the process of dementia. The episodes of reduced performance generally come with low motivation, high demands, and changes in the environment, and the frequency of the bad episodes increases with time.

The widening gap between good and bad episodes may have the potential to become an alternative way of describing the dementing process. The first lapses of function is a sub-clinical sign of a dementing process, which could be found by well-educated observers. If the indications of dementia at a single interview and testing session were tried out, 6 months would not have to pass before a diagnosis could be assigned, and thus the time between diagnosis and treatment would be shortened.

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Table 1. Description of subjects.

Degrees of reduction	<u>Levels of retardation</u>		
	Mildly retarded	Moderately retarded	Severely retarded
No reduction	1. M age 42	2. F age 26	3. M age 54
Possible reduction	4. F age 33	5. F age 34	6. M age 55
Reduction	7. M age 56	8. M age 53	9. M age 55

M = male, F = female

Table 2. Individuals' values on the aspects.

Aspects	<u>Groups</u>								
	<u>No reduction</u>			<u>Possible reduction</u>			<u>Reduction</u>		
	1	2	3	4	5	6	7	8	9
Change	8	12	4	-3	-4	-1	-8	-7	-14
Support	4	3	5	-2	-4	-3	-8	-12	-14
Learning	10	4	4	-1	-1	-2	-4	-8	-5
Mental presence	3	0	3	0	-1	-5	-14	-19	-10
Slowing	0	0	0	-1	-1	-5	-8	-20	-17
Joy of life	4	0	-5	-1	0	0	-4	-19	-20
Adjustment	-1	1	2	-1	-1	-2	-1	-6	-8
Liveliness	1	0	-4	-1	-2	-5	-2	-18	-9
Sums of aspects	29	20	9	-10	-14	-23	-49	-109	-97
Ranking	1	2	3	4	5	6	7	9	8

Table 3. Abilities differentiating most between groups and kept for a shorter Interview instrument.

Abilities	<u>Aspects</u>					6- level scale <sup>c</sup>
	Change	Support	Learning	Mental presence	Slowness	
Understanding spoken language	a, b	B				3
To make oneself understood	a, b	B	b			4
To learn to find one's way	a, b	B		b	b	3.5
To lay the table	a, b	B	b	b	b	6
Eating manners		a, b	b			4
Hygiene	a, b	a, b		a, b	b	2.5
Dressing	a, b				b	1.5
Initiative	A	B	b		b	11

<sup>a</sup> Questions with biggest difference between the No Reduction group and the Possible reduction group.

<sup>b</sup> Questions with biggest differences between the No reduction group and the Reduction group.

<sup>c</sup> Differences between the No Reduction group and the Possible Reduction group. Maximum value is 18.

Table 4. Rankings of individuals.

	<u>Person</u>									<u>Design</u> <sup>b</sup>		<u>Age</u> <sup>c</sup>	
	1	2	3	4	5	6 <sup>a</sup>	7	8	9	r <sub>s</sub>	p	r <sub>s</sub>	p
Sums of aspects (from Table 2)	1	2	3	4	5	6	7	9	8	0.98	0.000002	0.58	0.1
DSM-IV, except memory	1	2	3	4	5	6	7	8	9	1		0.63	0.07
ICD-10 personality	1	1	5	3	7	5	8	3	8	0.71	0.03	0.72	0.02
Being taught new skills, from most to least	2	4	5	8	9	1	6	2	6	0.13	0.75	0.18	0.64
Memory, Interview instrument	6	1	2	2	2	6	2	8	9	0.59	0.09	0.49	0.17
Memory, Test instrument	1	3	8	1	4		6	5	7	0.54	0.17	0.77	0.03
Cognitive tests	1	3	8	2	4		7	6	5	0.5	0.21	0.71	0.047
Reductions on a single testing session	3	1	7	4	5		8	6	2	0.26	0.53	0.57	0.14
Objects To Be Remembered	4	1	7	5	2		8	2	6	0.26	0.53	0.79	0.02
Age, from youngest to oldest	4	1	6	2	3	7	9	5	7	0.63	0.07		
Age, person 6 excluded	4	1	6	2	3		8	5	7	0.57	0.14		

<sup>a</sup> Person 6 could not be tested due to profound level of intellectual disability.

<sup>b</sup> Ranking correlated to design, Spearman rank.

<sup>c</sup> Ranking correlated to age, Spearman rank.

**INTERVIEW INSTRUMENT** Date: Interview on:

Interviewer: Interviewee: Relation:

**To know before testing:** Correct pair of glasses, Yes No, Visual handicap:

Reduction of audition, Yes, No Auditive handicap:

Personality: His or her greatest ability:

His or her greatest need now according to staff:

His or her greatest need according to the person:

His or her greatest difficulty:

If he or she has reductions in ability, does he or she know it? Is it a change from before?

Does he or she feel bad or worse than others? (depression) Is it a change from before?

Present losses of close ones/housing/occupation/other:

Has the person changed occupation or housing because of reduced personal capacity or independence (criterion B i DSM-IV)?

**CHANGES IN PERSONALITY:**

Compare the persons performance now and a year ago.

- 1. Emotional lability,** improved\_\_\_\_, unchanged\_\_\_\_, deteriorated, debut \_\_\_\_.

2. Bad sleep, improved\_\_\_, unchanged\_\_\_, deteriorated,  
debut\_\_\_.

3. Fear, spontaneous crying, increased hostility, improved\_\_\_, unchanged\_\_\_,  
deteriorated,  
debut\_\_\_.

4. Worry, anxiety, improved\_\_\_, unchanged\_\_\_, deteriorated,  
debut\_\_\_.

5. Restless/overactive improved\_\_\_, unchanged\_\_\_, deteriorated,  
debut\_\_\_.

**6. Irritability,** improved\_\_\_, unchanged\_\_\_, deteriorated,  
debut\_\_\_.

7. Does the person harm himself, improved\_\_\_, unchanged\_\_\_, deteriorated,  
debut\_\_\_.

Does the irritability reduce the persons social and occupational function:

**8. ABILITY TO TAKE INITIATIVE, THE PERSON'S DESIRE AND DETERMINATION:**

**When you think of the person's desire, determination and ability to do different things, has it changed?** Improved\_\_\_, Unchanged\_\_\_, Deteriorated,

**Support:** Does he or she need more, the same or less support than last year?

If there is a change, what is it, in what way, when did it begin:

The person's performance when it is good **0** and when it is worse **X**.

<b>6</b>	<b>5</b>	<b>4</b>	<b>3</b>	<b>2</b>	<b>1</b>
The person has the same desire and will to do things as before.	The person has desire to do different things and gets started but gives up easier than before	The person has desire to do different things, gets started but gives up with a hint of difficulty.	The person has in part lost desire and there is a hint of goal-directed behaviour. When staff gets him or her started he or she thinks the activity is fun.	The person has lost desire and shows no hint at getting started. Can proceed in an activity with continuous support.	The person has lost both will and desire, no participation despite support.

**Learning new skills:** Has the person more/the same/less ability to take initiative than last year?

**Vitality in the situation:** More/the same/less mentally present when he or she takes initiative than the year before? : More/the same/less slow when he or she takes initiative than the year before?

Did the reduction start suddenly or gradually?

Is a reduction of such magnitude that he or she is less independent? More difficulty to function in a social context?

**Debut:**

**Comments:**

**Part 2. Learning capacity with and without interference. Results in Interview instrument, Vision, cataracts, eye-glasses, to lay the table, hygiene, to get dressed, remembering the last days dinners:**

**CONSTRUCTION: Form 1.** Show, don't speak unless it is indicated in this text. Two sticks on the table each with their rings in front of them. The test leader puts one ring on his stick, the test person is expected to do the same on his. If the person doesn't do as expected, try it again, say: "Do as I do." **Scoring:**

- 0 point** The person does not manage to follow the instruction to put one ring on his stick in two attempts.
- 1 point** One ring, **big green**, the same as on the model stick, by the test person's stick. If the person succeeds on 1 point but not 2, go to form 2, Matching test.
- 2 points** One ring of each colour by the test person's stick. Put **one small yellow** on the test leaders stick 2 points. If the person succeeds on 2 points but not 3, go to form 3, Perception of colour test.
- 3 points** All 8 rings to a stick in front of the test person's stick, **blue ring**. If the person succeeds on 3 points but not 4, go to form 4, Perception colour and size test.
- 4 points** As a memory task: One ring of each colour in front of the test person's stick. **orange** Ttest leader: get eye contact with the test person. **Say:** "This time I am going to take away my stick, when you start working on yours." The test person may study the model until he or she starts looking at and collecting his rings. The model is taken away and the test leaders remaining rings are covered, 4 points). If the person succeeds on 4 points but not 5, go to form 5, Perception of colour and size test.
- 5 points** Same procedure but with all rings to a stick, (5 points). **Little green** Proceed if the person failed and managed to correct his or hers faulty solution. **Stop** if the person fails twice. If the person succeeds on 5 points, go to form 6, Perception colour and size test.

**CONSTRUCTION: WRONG SOLUTIONS. Do it parallel to each form.** Note the number of wrongs the person makes. Does nothing = 6 wrongs. Puts rings on both sticks/the wrong stick in spite of verbal explanation=1 wrong **and** concrete guidance=1point more. Despite one ring on the test leaders stick the person puts more on his own =1 wrong, puts all rings =2 wrongs. Sums of wrongs.

6 wrong    4 wrong    3 wrong    2 f wrong    1 wrong    0 wrong  
**1 point.    2 point.    3 point.    4 point.    5 point.    6 point**

**CONSTRUCTION: To correct a wrong solution, an executive ability. Done parallel to each form.** When you replaced the model, observe the person's behaviour, does he compare his to the model? Does he see if he made his right or wrong? Make notes here also for test items on interference in Form 2, not for working memory.

- 1 point.** Can't see his solution was wrong.
- 2 point.** He can see his solution was wrong, but makes no attempt to correct it.
- 3 point.** He can see his solution was wrong, attempts to correct it bit does not succeed.
- 4 point.** Succeeds in correcting one wrong solution or had only one.
- 5 point.** Succeeds in correcting his wrong solutions.
- 6 point.** Has no wrong solutions.

**CONSTRUCTION: Form 6.** If the person managed 5 p. on form 1. Always put smaller rings on top of bigger, 'pyramid shape'. Rings as in form 3, one of each colour, big yellow, big blue, dark blue, little green, orange and red top. **The model is taken away.** Cover the the test leaders remaining rings. **Stop** if the person fails twice.

**1 point.** Puts one ring, as model. Big yellow/.

**2 point.** Puts två rings, as the model. Pyramidshape Dark blue/orange.

**3 point.** Puts tre rings. Pyramidshape. Big yellow/big blue/dark blue.

**4 point.** Four rings. Pyramidshape. Big blue/dark blue/little green/orange.

**5 point.** Five rings. Pyramidshape. Big yellow/big blue/little blue/little green/orange.

**6 point.** Repeat five rings. Pyramidshape. Big blue/dark blue/little green/orange/red knob.

Ready? At 1-3 points, to Motor interference form 2. At 4-6 points, try Form 7.

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***I point at things, I want you to point at the same***

*Results in Interview instrument, To follow a program on TV. Remembering dinners the last days:*

**OBJECTS TO BE REMEMBERED.**

Let the objects the person could name in Naming stay on the table. **THE**

**OBJECTS CONTOURS ARE DRAWN ON A SHEET OF PAPER, 32**

**COMMON THINGS:**

*Book, pencil, cap, necklace, scissor, washing-up brush, glass, pencil, comb, spoon, teaspoon, knife, eraser, button, fork, hanger, toothbrush, dish-cloth, jug, lid of a sauce-pan, screw driver, hammer, tube of tooth-paste, screw, shoe-laces, matches brush, box with thumbtack, battery, bell for a cycle, telephone receiver, pocket diary.*

## Direct recall.

Point at an object. If the person does not point at the same, repeat the instruction and try three times with other objects. If it works, proceed to pointing at **two** other objects, then **three** etc. 1point per word.

0 point: *Does not remember.*

1 point. *book,*

2 points. *a hammer, pencil,*

3 points *a tube of tooth-paste (common), cap, necklace,*

4 points *a screw, scissor, a washing-up brush, glass,*

5 points *matches, brush, shoelaces, button, comb.*

6 points *spoon, teaspoon, knife, fork, a box with thumbtack, battery*

7 points *eraser, hanger, toothbrush, a dish-cloth, a bell for a cycle, a telephone receiver, a pocket diary.*

## Delayed recall 30 seconds

Each time you do the task above, make a 30 second pause and ask him or her on the same objects again.

<b>0 point,</b>	<b>1 point,</b>	<b>2 point,</b>	<b>3 point,</b>	<b>4 point,</b>	<b>5 point,</b>	<b>6 point,</b>	<b>7 point</b>
Doesn't remember	1 object	2 objects	3 objects	4 objects	5 objects	6 objects	7 objects

**Did the person guess:** Remembers in total\_\_\_\_, those that are correct\_\_\_\_, the proportion of correct answers 0,\_\_\_\_

The difference; delayed recall minus direct recall =+/-\_\_\_\_

