

A project funded by



**The Juvenile Neuronal Ceroid Lipofuscinosis and
Education Project (2014 - 2017)**

**ASPECTS OF LEARNING FOR INDIVIDUALS WITH THE
JUVENILE NEURONAL CEROID LIPOFUSCINOSIS (JNCL) OR
THE CLN 3 DISEASE**

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INTRODUCTION

1.a ABOUT THIS PAPER

This paper addresses some findings and experiences from the Juvenile Neuronal Ceroid Lipofuscinosis and Education Project (2014 - 2017). Juvenile Neuronal Ceroid Lipofuscinosis (JNCL) is also called CLN3 disease, Spielmeyer-Vogt Syndrome and Batten Disease. JNCL is used in this paper.

Developmental patterns of JNCL in relation to principles for pedagogical interventions are discussed in this paper. It is believed that pedagogical interventions can contribute to an improved quality of life for individuals with JNCL.

This paper is a co-operative venture between Statped Heimdal in Norway, University of Oslo in Norway, The Norwegian NCL Family Organisation, The Danish NCL Family Organisation, The Vision Center Refsnæs in Denmark, The Bildungszentrum für Blinde Sehbehinderte Hamburg in Germany, The Batten Disease Family Association in UK, The WESC Foundation in England, The Royal Blind Asylum and School, Edinburgh in Scotland, NCL-Gruppe Deutschland in Germany, The Valteri Centre for Learning and Consulting, Onerva in Finland and The Finnish Federation of the Visually Impaired.

1.b JUVENILE NEURONAL CEROID LIPOFUSCINOSIS (JNCL)

i. Diagnostic characteristics

Neuronal Ceroid Lipofuscinosis (the NCLs) is a group of rare and progressive neurological disorders with clinical coherence. The name refers to the fat compounds ceroid and lipofuscin, which can be seen in the cells of individuals with NCL. JNCL is the most common type of the NCL diseases. Children with JNCL are healthy in early age until the first symptoms of the disease are evident. JNCL is an autosomal, recessive, hereditary disease. This means that the disease affects both genders, and the disease is inherited from healthy parents (carriers) (Elmerskog and Fosse, 2012; Helland et al. 2016).

JNCL develops slowly and causes blindness, epilepsy, learning difficulties, mental stagnation, language difficulties, motor difficulties and eventually dependency on a wheelchair. Emotional reactions are common. Individuals with JNCL eventually lose their ability to speak. The complications that occur lead to dependence on care/nursing at a young age and a shortened life expectancy - most individuals pass away between the ages of 20 to 30. There is currently no medical treatment for the disease (Mole et al., 2011; Helland et al., 2016).

The development of symptoms of JNCL is very individual both in terms of life expectancy and the age when different types of symptoms will or may occur. In most cases, the visual impairment leads to identification of the correct diagnosis, often between five and nine years of age. Most children with JNCL have had normal development prior to this time.

ii. Occurrence

Most countries do not have reliable statistics regarding the number of individuals with JNCL. There are 35 individuals with JNCL in Norway, in Denmark 25 individuals, and in Sweden approximately 30 individuals in 2017. The prevalence varies between countries: in Norway it is approximately 1.5 children per 40.000 new-borns (Elmerskog and Fosse, 2012).

iii. Stress on families

Having a child with JNCL is very stressful for the family. The majority of parents do not give their child all the information about the diagnosis. Parents want to protect the child from learning about shortened life expectancy and future development of symptoms. This often results in complicated life situations. The fear of people accidentally revealing information about the disease makes parents consider carefully who needs to know about the diagnosis. These same considerations apply to siblings.

1. JNCL – A CHILDHOOD DEMENTIA DISEASE

2.a DEVELOPMENT PERSPECTIVES

The project emphasizes childhood dementia as an important perspective to consider in pedagogical work with individuals with JNCL. Dementia is traditionally linked to the elderly population. Because of this, the project has experienced some resistance when using the term in relation to children. Some also argue that it is unethical to discuss learning and development when the child – eventually – will be affected by loss. This dilemma can lead to a lack of development expectations and a resistance to planning for future situations and needs (von Tetzchner et al., 2013).

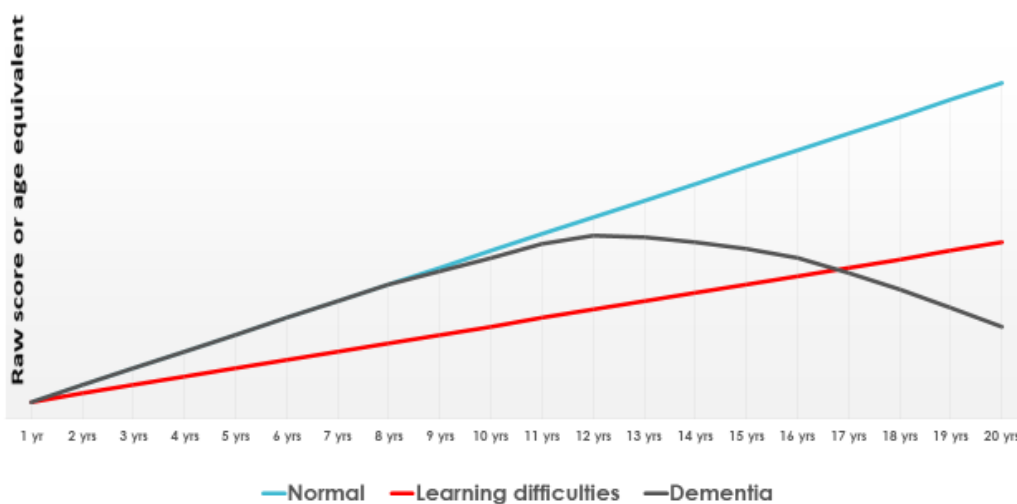
The term childhood dementia must have a pedagogical content to benefit the affected individual. Childhood dementia is providing us with a number of “*windows of opportunity*”. It is possible to ask - *what is possible when?* The term “*windows of opportunity*” emphasizes the need to implement appropriate interventions at the right time, and in addition, the duration of times for successful interventions are limited. What is not done today may be impossible to achieve tomorrow due to the progressive development of dementia.

There is a significant difference between dementia in elderly people and dementia in individuals with JNCL. Dementia development in the elderly population comes gradually and unexpectedly. However, for children with JNCL we know that dementia will develop eventually. It is therefore possible - through pedagogical interventions - to prepare the child for the forthcoming dementia development, and see dementia in connection with other forthcoming symptoms. In addition, children's brains are particularly receptive to learning and development through its unique plasticity. This also applies to children with JNCL. Plasticity is a prominent feature of the nervous system at birth and is still very pronounced in childhood. After adolescence, the plasticity of the brain decreases (Jensen, 2016).

2.b DIFFERENT DEVELOPMENTAL PATTERNS IN CHILDREN

Figure 1 is a simplified version of a graphic representation describing three types of development in children: the normal development of children, the development of children with learning difficulties, and the development of children with dementia (After Shapiro and Klein, 1994).

Figure 1. Different developmental patterns in children
Different development patterns in children



The blue line in figure 1 refers to the normal development of children. The red line refers to the development of children with congenital learning difficulties. The red line is steadily rising, but at a slower pace when compared with the normal population. The development and learning of these groups shows constant progress through childhood and adolescent years. The black line refers to the development of children with dementia or JNCL. This group of children has a normal development in early life. The development eventually slows down and comes to a stop, followed by decline later in life.

2. THE PROJECT

3.a OBJECTIVES

There is a danger that the comprehensive health problems caused by JNCL can give predominant focuses on the shortened life expectancy or forthcoming losses. This may result in situations where educationalist, social workers and others are unable to act in the best interest of the affected individual. The intention of the project is to reverse this perspective - *what can be done despite all difficulties?* The project emphasizes learning perspectives and interventions focusing on possibilities in spite of all difficulties.

The JNCL and Education Project is, as far as we know, the most comprehensive pedagogical project about JNCL that has been implemented. The project considers learning and development in a lifelong perspective. The aim of the project is to build new non-medical knowledge about the target group, and accordingly develop new pedagogical strategies, new teaching methods and relevant teaching tools.

3.b ORGANISATION, PROJECT PARTNERS AND PUBLISHING

Seven countries participated in the project - Norway, Finland, Scotland, England, Germany, Denmark and the United States. Five of these countries received financial support from the EU (Erasmus +). Denmark and the United States participated in the project without financial support from the EU. Other financial contributors included various organisations in all participating countries - family organisations, universities, special needs/specialist schools and organisations providing counselling services. The project management consisted of a JNCL parent representative, a manager from a national service provider for individuals with JNCL and an educationalist in JNCL from each participating country. The project manager was Bengt Elmerskog from Statped in Norway.

Among the project publications are the following:

- An interactive assessment and intervention tool named as EDO (Educational Development Observation) for making the services offered to the target group better and more efficient
- An information video about JNCL
- Various types of educational forms that can be used for planning, implementation and evaluation of actions
- An ICT-based tool (quiz) to promote the target person's knowledge acquisition
- A textbook based on the research results will be published in 2018.

All publications are in English, and many are available in Norwegian, Finnish, German and Danish.

3.c TARGET GROUPS

The project's primary target group is individuals with JNCL. Secondary target groups are parents/guardians, pedagogical personnel (teachers, residential staff, etc.) with daily responsibility for the target group, and organisations such as higher education.

3.d PROCEDURE

The project was divided into three phases. Phase 1 (2014-15) included planning, literature search and preparation for the research. Phase 2 (2015-16) included data collection and analysis of incoming data, as well as the development of relevant educational tools. The last phase (Phase 3, 2016-17) includes documentation of research results, production of educational tools and publication/dissemination of results.

Data is based on three comprehensive surveys and interviews. Parents of children with JNCL completed survey 1, bereaved parents survey 2, and pedagogical personnel survey 3. The comprehensive and detailed surveys in Norwegian, German, English, Finnish and Danish could be answered electronically or on paper formats. 192 surveys were answered according to the following:

- Survey 1 and 2 (answers from parents): Norway 18, Finland 24, Germany 33, UK 6, Denmark 16, and USA 14.
- Survey 3 (answers from personnel): Norway 12, Finland 40, Germany 11, UK 7, Denmark 7 and USA 4.
- 55 interviews were conducted by an interview guide. The interviews involved parents and professionals in all project countries. The interviews were transcribed and translated into English.

The data collection was approved by REC (Regional Committees for Medical and Health Research Ethics) in 2014.

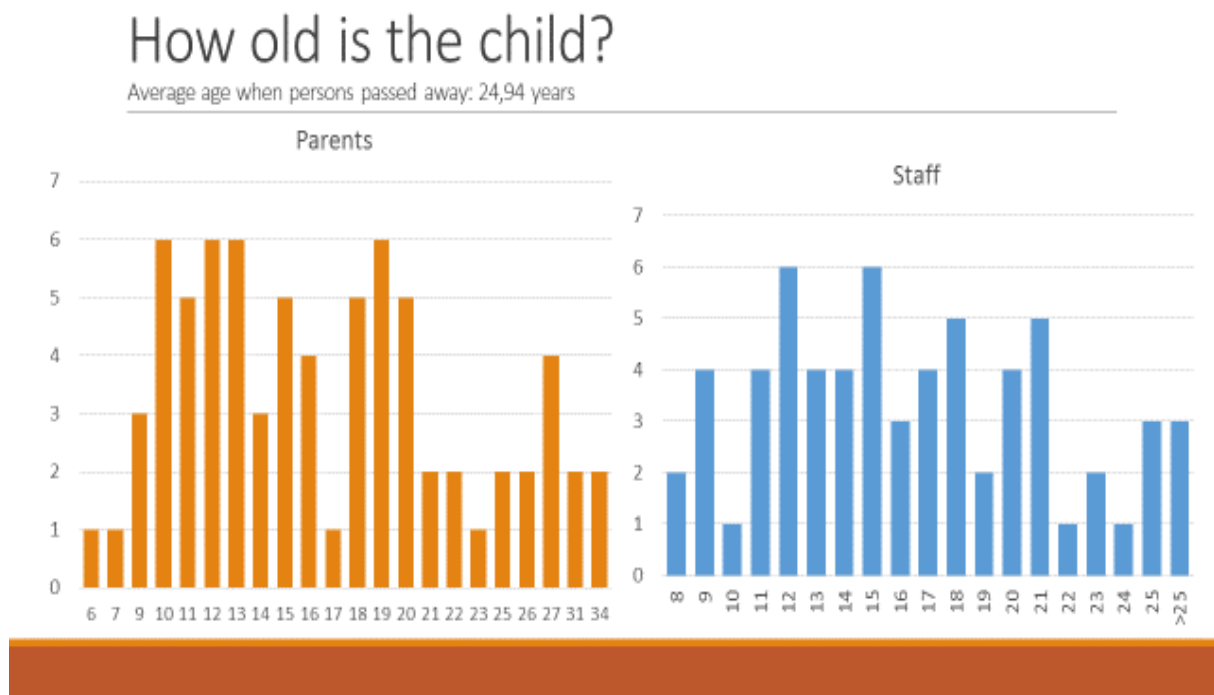
3. RESULTS

4.a ABOUT THE PROJECT SAMPLE

i. The age distribution

Figure 2 gives an overview of the age distribution of the project sample.

Figure 2. Age of the participants



The brown bars show the number of participants from survey 1 completed by parents. The blue bars show the number of participants from survey 3 completed by personnel. Participants who have passed away are not included in figure 2 (survey 2 from bereaved parents). These individuals achieved a mean age of 24.9 years. The gender distribution from survey 1 and 2 were 49 males and 62 females and the gender distribution from survey 3 were 37 males and 36 females.

ii. Types of schools

Figure 3 shows what types of schools the project sample attended in relation to grade.

Figure 3. Different types of schools in relation to grade

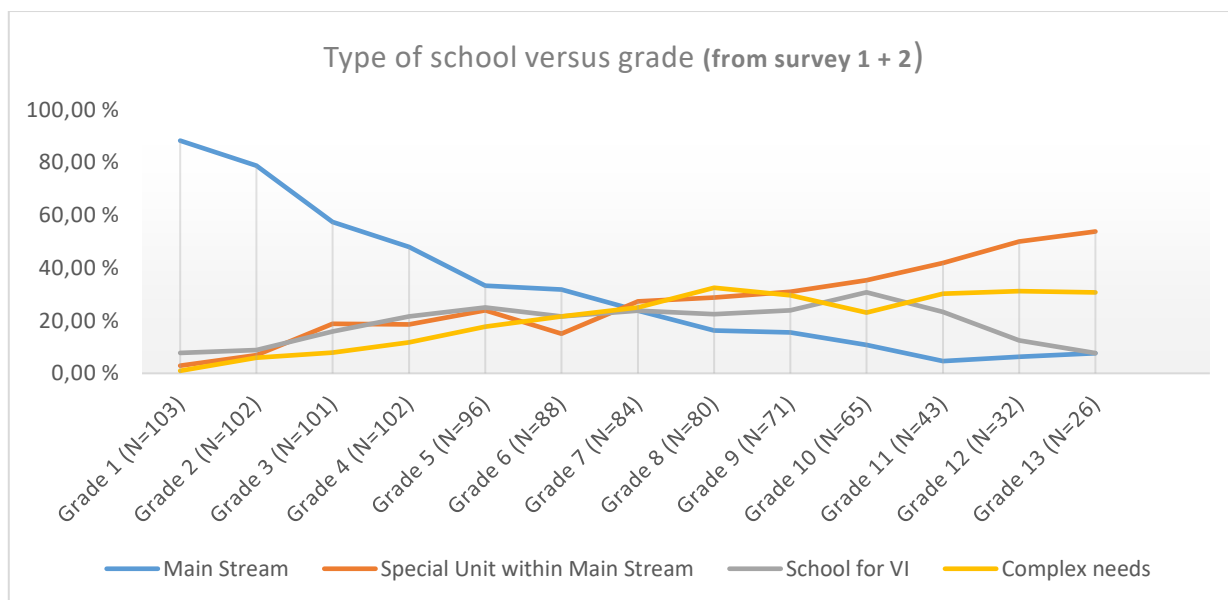


Figure 3 shows the relative distribution of type of school from grade 1 to grade 13. The following categories of education were used: mainstream school, special unit within mainstream school, special/specialist school for visual impairment and special/specialist school for children and young people with learning difficulties/complex needs. Note that special schools are very rare in Denmark, Finland and Norway.

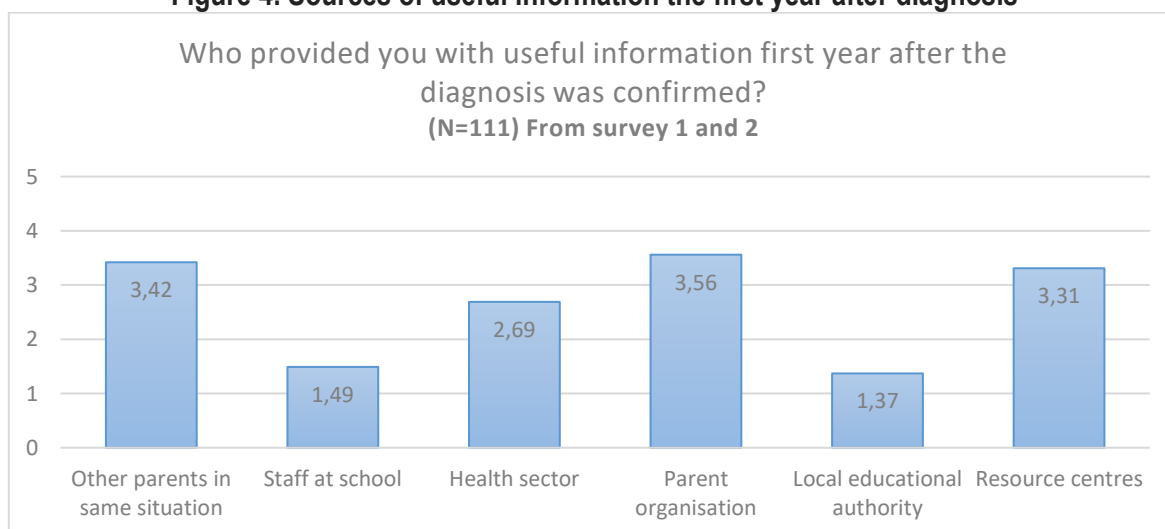
Many children in the project sample started attending mainstream schools with a regular or adapted curriculum up to grades 5 and 6. Approximately 90% of the sample did not attend mainstream classes at grade 13. Some students attended special/specialist schools for visual impairment in UK, USA and Germany from grade 3 and 4 up to grade 10. These students later attended schools for learning difficulties/complex needs. Both positive and negative aspects of this transition were described in several parental interviews.

4.b COMPETENCE BUILDING

i. Sources of information about JNCL

The infrequency and complexity of JNCL are often highlighted as the two main challenges for the educational system and the local education authority/municipality when supporting the children and their families with good, targeted services. The parents were asked who provided useful information in the first year after the JNCL diagnosis was received (Figure 4)

Figure 4. Sources of useful information the first year after diagnosis



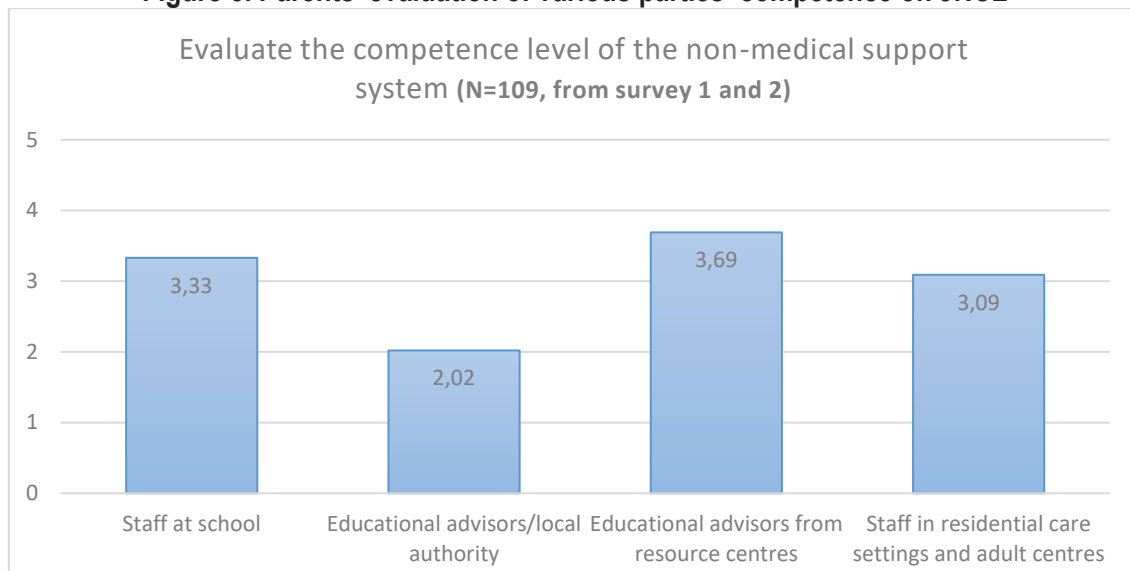
Explanations: Level 1 = no contribution, 2 = some contribution, 3 = moderate contribution, 4 = large contribution and 5 = very large contribution

Other parents in the same situation and parent organisations were often considered to be the most important sources for information, followed by resource centres and health institutions. Schools and local counselling services received the lowest scores as useful information sources.

ii. Competence level in different service providers throughout life

The parents were asked to consider the various parties' pedagogical competence in relation to JNCL, not only during the first year after diagnosis but throughout life. The responses are shown in Figure 5.

Figure 5. Parents' evaluation of various parties' competence on JNCL



Explanations: Level 1 = No competence, 2 = Low competence, 3 = Moderate competence, 4 = High competence and 5 = High competence

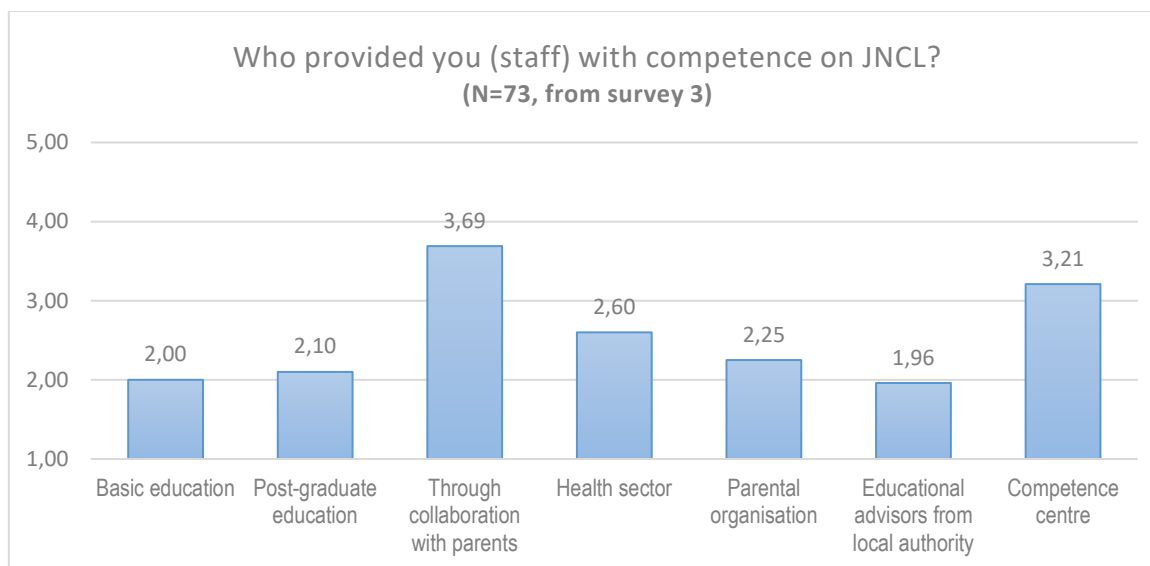
Figure 5 shows that resource centres received the highest score followed by personnel from schools and personnel in residential care settings and adult centres. Local educational counselling services received the lowest score. This picture was confirmed by many parental comments. Here are some examples:

- *My best partners and advisors have been the parent association, the resource centre and the family doctor*
- *The municipality always was too late in relation to the disease development. No help was received from there...*
- *The municipality has never been a supporter. We are suffering from this today because things were done too late. We – the parents – have always carried the burden ourselves.*
- *The family doctor, physiotherapists and occupational therapists + the municipality has no expertise in this area. The so-called teamwork is not working, no coordination. The parents have always cleared the way in collaboration with the resource centre.*
- *Educational counsellors from the resource centre have the competence needed.*
- *The municipality has very little competence. The “enthusiasts” in resource centres are the driving force, but this is very vulnerable.*

iii. Competence building in personnel

Pedagogical personnel responsible for children, adolescents and young adults with JNCL were also asked who contributed to their competence building in JNCL. The answers are shown in Figure 6.

Figure 6. Sources of competence building of personnel



Explanations: Level 1 = No contribution, 2 = some contribution, 3 = moderate contribution, 4 = large contribution and 5 = very large contribution

Pedagogical personnel consider collaboration with parents to be the most important source for building their competence, followed by resource centres. The local advisors/counsellors were not considered to be important sources for competence building. Examples of some comments from personnel:

- *Close collaboration with parents has been vital for my competence development and this has reduced the vulnerability for NN and the staff. The parents have always participated in meetings and answered questions from the staff. In the beginning we had meetings every week, today one meeting every third week, but extra meetings when needed.*
- *The resource centre has provided good counselling on the disease, regarding the person's needs, and they have provided competence building in how to use the data programme "Sarepta".*
- *Close collaboration with the counsellor from the resource centre when regarding impact of interventions has contributed to our competence building.*
- *The resource centre has held several workshops for the staff team.*
- *The municipality [the municipality's educational advisors] has not provided any educational counselling. They do not know the pupil or anything about the disease, and they rarely attend meetings.*

Similar quotes were received from all participating countries. Parents and personnel often consider competent, enthusiastic, individual professionals as the important competence providers, and not the services or systems. This situation is described as very vulnerable - *what will happen if the individual source is no longer available?*

4.C SOME PEDAGOGICAL ASPECTS ON THE JNCL DISEASE

i. Symptoms

JNCL is a complex disease with many different symptoms. Figure 7 shows the average age in the project sample when different symptoms were registered for the first time.

Figure 7. Average age for symptomatic symptoms (n = 111)

Onset of symptoms - mean age in years (N=111, from survey 1 and 2)

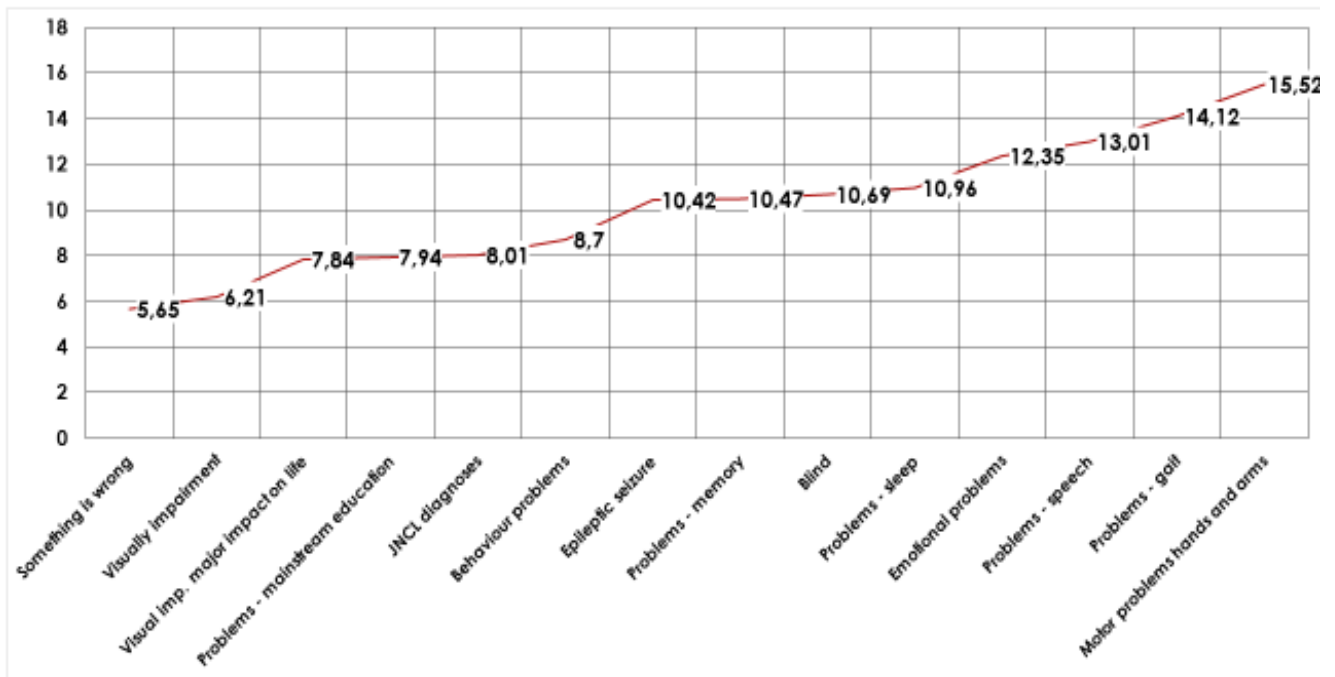


Figure 7 shows that individuals with JNCL are affected by a series of declines and losses: visual function, communication, cognition, behaviour, motor performance etc. The figure shows the project sample's average age for onset of various symptoms. It is important to note that the individual order of losses/declines vary from child to child. An example: some children had problems with speech before motor difficulties were apparent, while other children had these problems in the opposite order. Some symptoms are not registered at all in some individuals, e.g. emotional problems. A relatively fixed pattern, both in time of onset and order, is the occurrence of visual impairment. Visual impairment is confirmed at a young age and is one of the earliest symptoms of the disease.

The complex picture of different symptoms in the sample indicates a need for different areas of competence. Individuals with JNCL are mostly organised under the "faculty of visual impairment" regarding pedagogical services probably because problems with vision lead to the diagnosis. The project organisation confirms this picture - only professionals representing the field of visual impairment were members of the project's working/steering committee. The children's loss of vision is consequently often met by required competence, while other symptoms and needs are not necessarily met with the corresponding expertise. Experience from the project shows that services for children with JNCL need to be supplemented by interventions that traditionally fall outside the field of visual impairment.

ii. Visual impairment

Age when visual impairment occurred

Visual impairment usually affects children with JNCL from 6 to 9 years of age (von Tetzchner et al., 2015). The parents were asked about the child's age when visual impairment was confirmed, when visual impairment had an evident impact on daily living, and when the child became blind. The responses are shown in Figure 8.

Figure 8. Average age for visual impairment

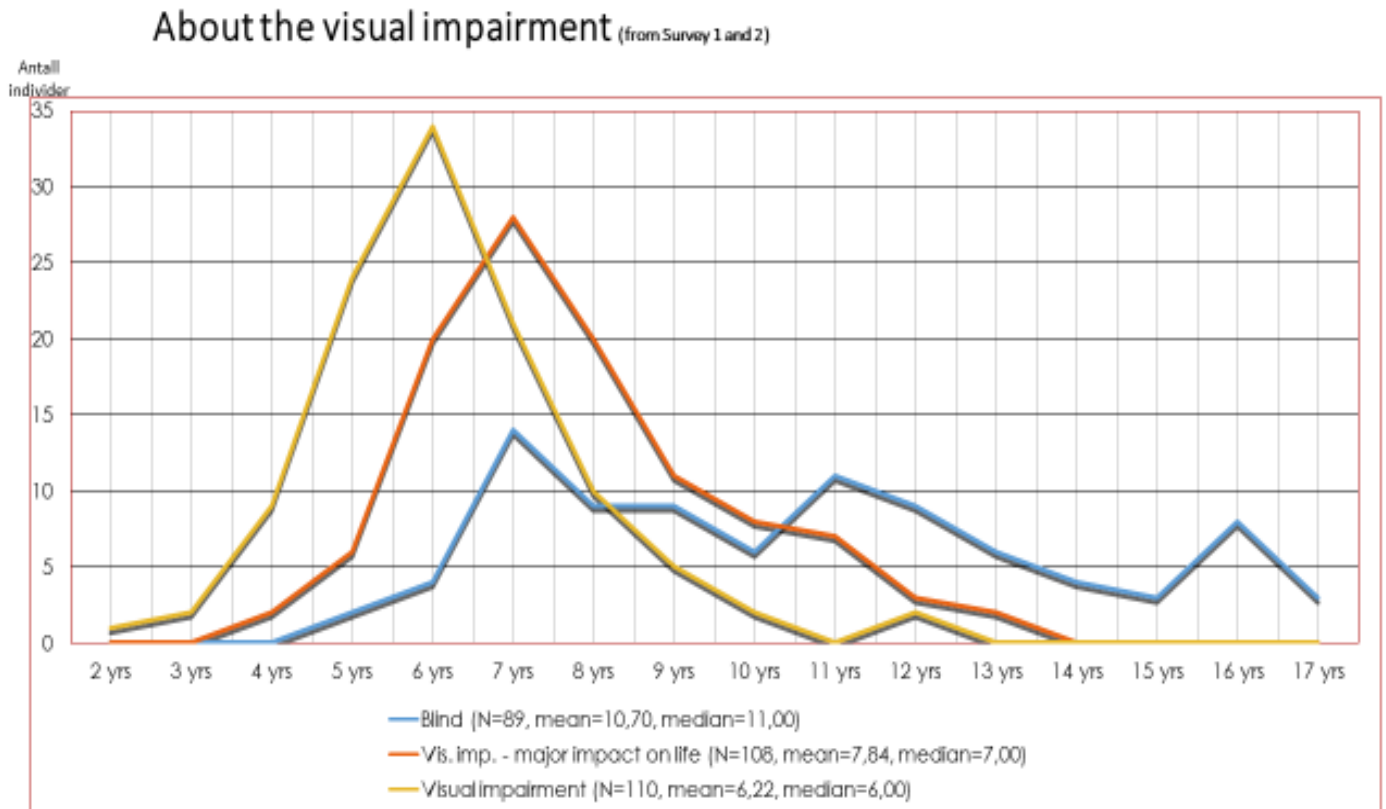
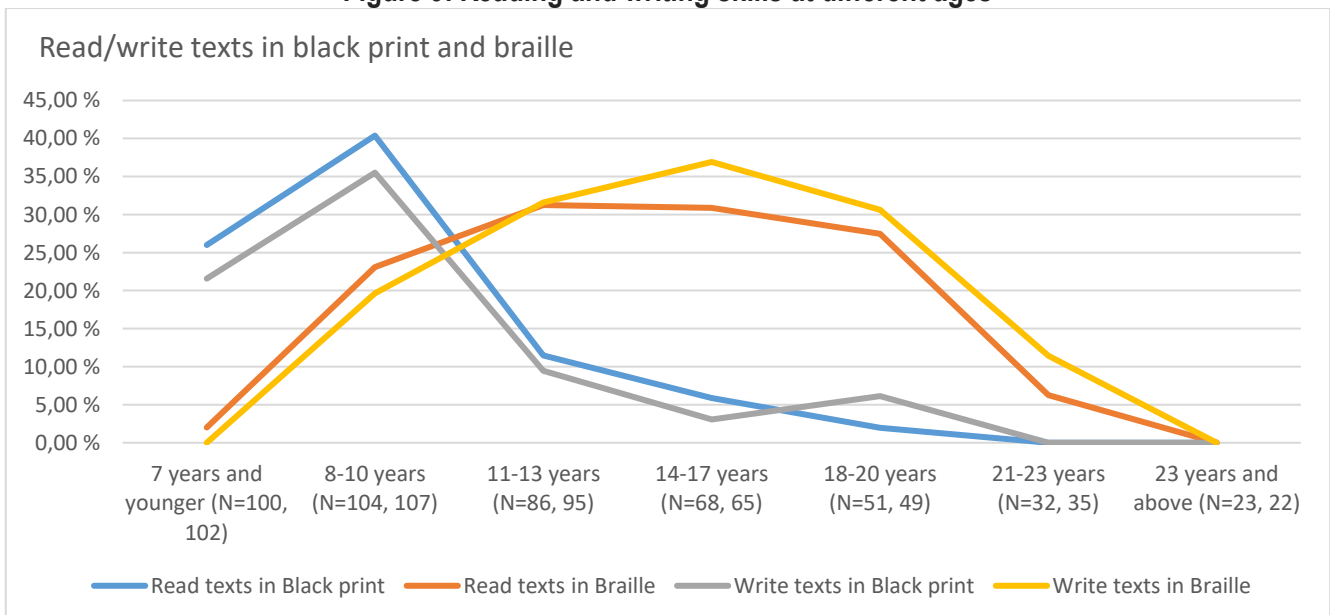


Figure 8 shows that the project sample became visually impaired at the average age of 6.2 years (yellow line); the visual impairment had a major impact on everyday life at the average age of 7.8 years, and the average age when blindness occurred was 10.7 years (blue line). This shows that blindness often occurs significantly late. The category blindness also shows a larger variance in age of occurrence than the other categories. The project regard this as important when it regarded in a pedagogical perspective.

Reading and writing

The parents were asked about the children's reading and writing skills at different age levels. Figure 9 shows the results.

Figure 9. Reading and writing skills at different ages



Skills in reading and writing texts in black print were at their best between 8 to 10 years of age on average; about 40% of the project sample achieved this skill. About 25% achieved the same level by using braille in the same age group. The best skills in reading and writing braille texts were found between 11 and 20 years of age: about 30% to 35 % of the project sample achieved this skill.

The average skills in writing braille were somewhat higher than the braille reading skills after the age of 11 years. More importantly, the group retained relatively good skills in braille for a longer period of time when compared with skills for reading and writing black print. The results indicate that braille skills had a greater impact on life as a whole.

Many children in the project sample achieved simple reading and writing skills in braille, e.g. reading single words (not texts). They also benefitted from reading and writing by e.g. being able to read their day schedule or to play cards marked with braille. These children are not included in figure 9.

Does learning of braille become easier for children if they first break the reading code by visual reading? This correlation is not found in the project data. A positive correlation, however, between braille skills and the age when braille was introduced is found. This result indicates that the cognitive capacity is of importance for learning skills in braille. It indicates that it is beneficial to start learning tactile reading and writing as soon as possible after the diagnosis is known, even if the child has vision to enable visual reading and writing.

Some children with JNCL in UK are offered the tactile literacy code "Moon" as an alternative to braille. It is claimed that Moon is easier to learn, and that skills are easier to maintain. Moon is partly based on the Latin alphabet and therefore it is easier to learn for parents and peers. The major disadvantage of Moon is poor availability of materials and equipment, even within UK.

Reading and writing in braille was of great importance to many individuals in the project sample. Some of them had become "authors" writing fantasy books and poetry. Some individuals over the age of 20 had braille writing as a special area of personal interest, others were active users of e-mail (requires certain skills in braille), and some loved playing card games because they could read braille-marked cards. The following quotation from a parent illustrates the importance of early learning of braille:

"With his rapid loss of eyesight, we started braille along with large print access. He did better in braille and really enjoyed learning it. He spent hours typing on his brailier, learning spelling words and reading books. He was very proud of the fact he could do braille. For him, braille was the best thing he did. He read books in the dark way after bedtime many nights."

Some individuals in the project sample never learned to read and write print or braille due to an early onset of dementia. The project results also indicate differences between countries: some countries emphasize braille teaching more than others. The results include quotations from parents having received questionable recommendations not to expose the child to learning challenges such as reading and writing. Such recommendations had come from both doctors and educators referring to future declines caused by the diagnosis.

Visual decline and dementia development

Nothing in the project's data material indicates that there is a concurrence in time between the development of the dementia and the visual decline. The dementia developed early and fast for some children while visual functions persisted, and these symptoms occurred in the opposite order for other children. This issue is of pedagogical interest. Today, it is possible to maintain visual reading and writing with severe visual impairment by using technical aids. Letters can be enlarged to accommodate the visual impairment. The teacher not receiving guidance will not, in general, consider tactile reading or writing as long as the student's vision allows visual reading and writing. When visual reading and writing become impossible, due to blindness, it may be too late to learn complicated skills like braille because of the development of the dementia. The following quotations from the parents exemplify this:

- *She was not able to read black print at the age of 10, not even with technical aids. Her cognitive abilities were no longer sufficient to enable her to learn braille.*
- *The person was not motivated in braille because the person had sight for a long time and she was trying hard to cope with CCTV to find out the words. After getting blind, they started with audio books.*

- Because of her blindness, she cannot read normal print anymore. It was too late to learn braille.

iii. Loss of communication skills

Decline in intelligible speech

Individuals with JNCL experience increasing difficulties with speech intelligibility. Figure 10 shows the intelligible speech decline over age (from survey 1 and 2)

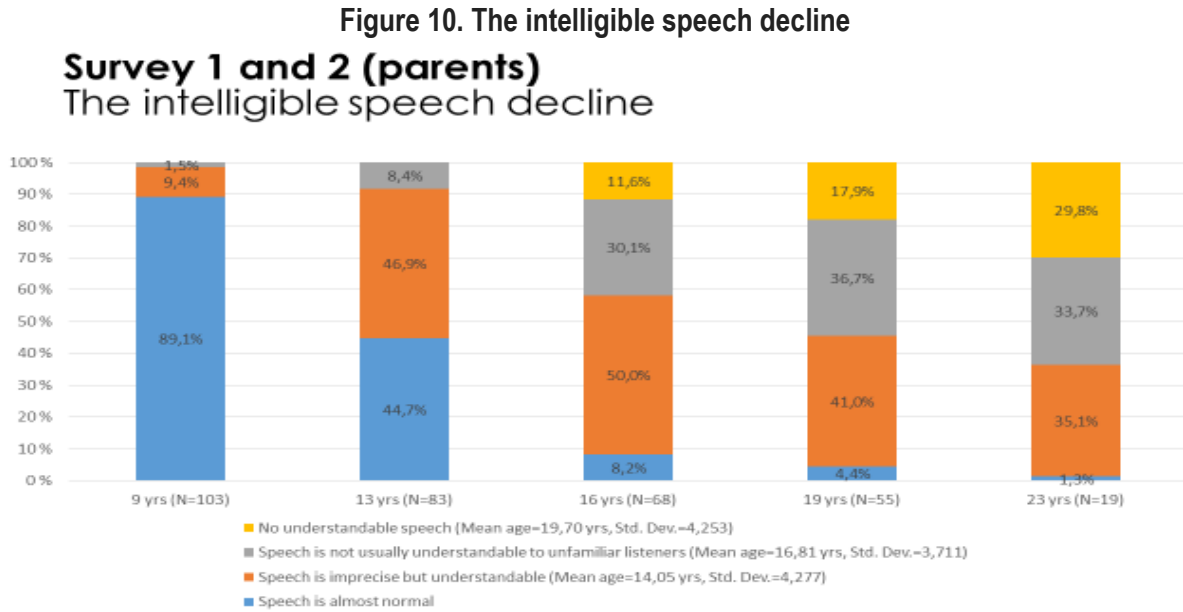
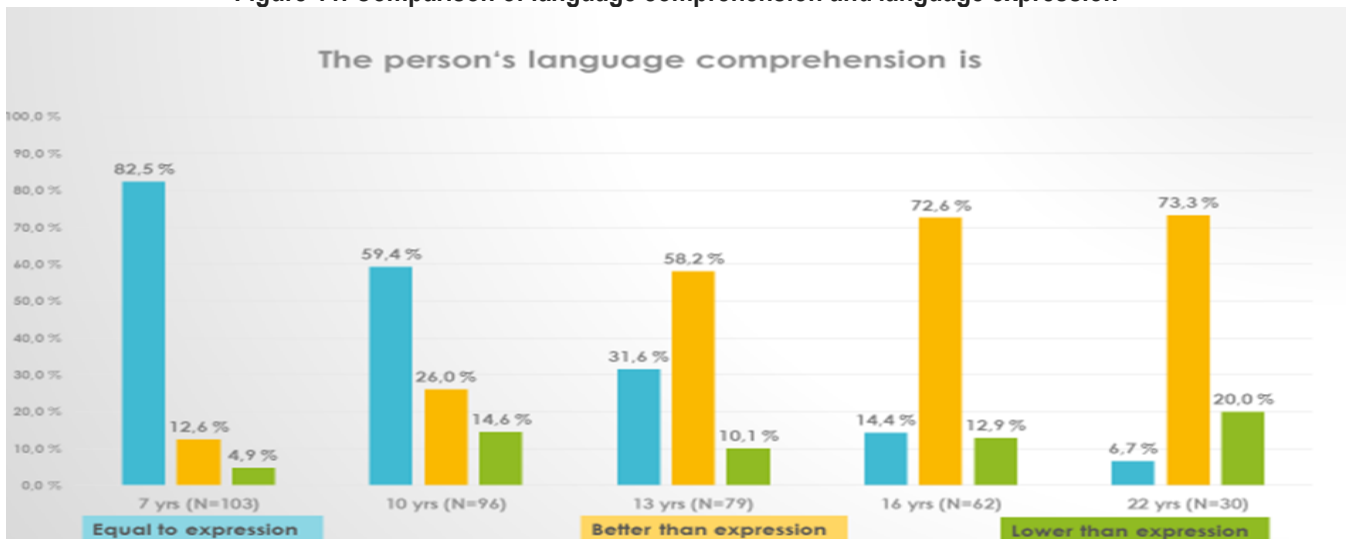


Figure 10 shows that more than 50% of the project sample had major difficulties with speech intelligibility at 19 years of age; some individuals experienced this situation before 13 years of age. The project results show that the speech problems with highest prevalence in the sample was word-finding, followed by mumbling and stuttering. Most individuals with JNCL eventually have combinations of different speech problems, and the situation will eventually become very complex for most individuals. Many parents considered communication problems as the biggest challenge for their children. The personnel also considered communication to be a highly complex area.

Use of speech versus comprehension of speech

The parents were asked to compare the child’s language comprehension with the child’s expressive language. Figure 11 shows a comparison of expressive language and language comprehension at different ages.

Figure 11. Comparison of language comprehension and language expression



The blue bars show that children's language comprehension and ability in language expression were considered equal, the yellow bars show that language comprehension was considered to be better than the children's ability to express themselves, and the green bars show that the children's expressive language was considered to be better than their language comprehension. These results indicate that maintenance of language comprehension is better through life than maintenance of language expression. The project believes this imbalance may cause stress, frustrations and a reduced quality of life for the individual.

Language and communication training in education

The parents were asked if loss of communicative skills was met by pedagogical interventions

Figure 12. Age when the target person received communication training (n = 74)

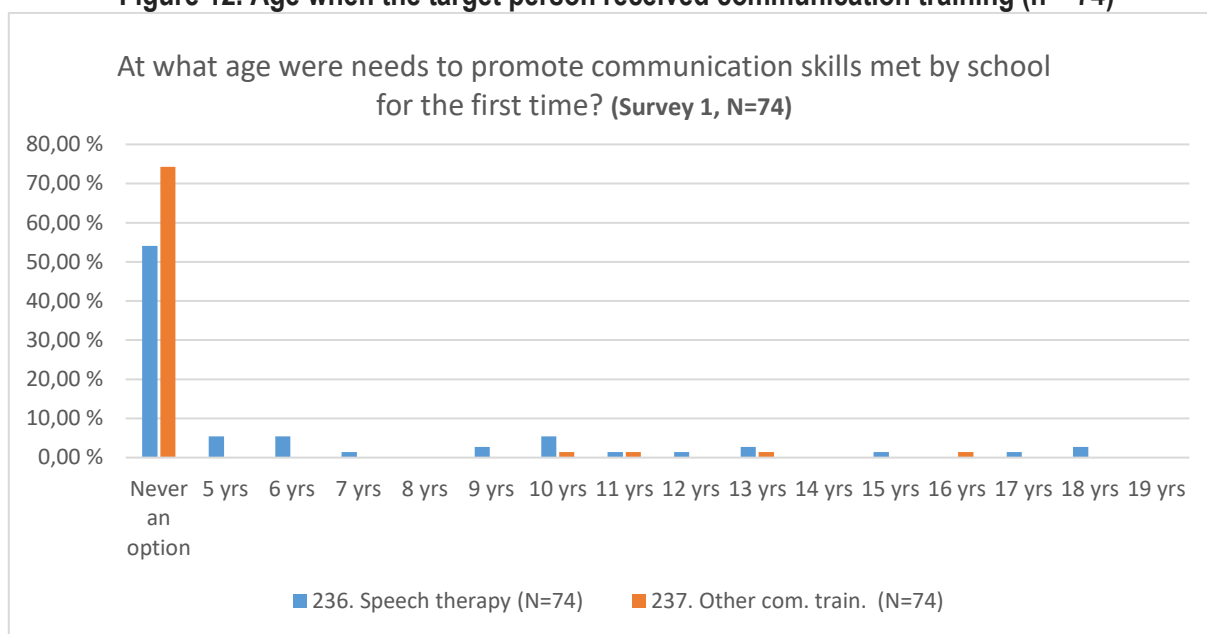


Figure 12 illustrates that more than 50% of the individuals were never offered speech therapy, and 75% were never offered other forms of communication training. The picture is representative of all project-participating countries, but the results indicate that some special schools, e.g. in Scotland, have a stronger focus on these areas than other institutions.

Use of manual signs

10 individuals with JNCL from 8 to 19 years of age in Norway and Denmark received training in using manual signs as Alternative and Augmentative Communication (AAC) for language expression during the project period. This was time-limited sub-project. It started with a two-day seminar where teachers and parents identified a sign repertoire based on the individual's interests, preferences and daily activities. The seminar presented appropriate teaching methods and some tools, and registration forms for learning outcomes. Two of the children withdrew from the project of different reasons. Some children changed school and personnel during the period, which affected the continuity of the programme.

The transition from showing signs on instructions to using signs in natural situations was difficult to achieve in the short project period. However, two children started to use some few manual signs in natural situations during the project period.

There were big differences in how many signs the children learned. The number of learned signs varied from four signs and upwards. The youngest student in the group, 8 years old, learned most signs. This student learned about 200 signs during a three months period. This student started to use signs in natural situations shortly after the conclusion of the project. His manual signing eventually solved some of his communication problems caused by stuttering.

It is today difficult to conclude if signing skills will persist over time, but the results are so far promising. The project found that many obstacles for achieving good results were related to barriers in the environment such as collaboration problems, the timetable, or a lacking focus on transferring established skills to natural situations. The Swedish film “Att gå bredvid” (to walk beside), is a film showing a young man with JNCL using manual signing for expressive communication (<https://www.spsm.se/kurser--aktiviteter/nyheter/nyheter/en-sallsynt-film-om-en-sallsynt-diagnos/>, 2016)

iv. A transition project for a young man with JNCL

Introduction

An individual project, related to the transition from a student to adult situation for a young man with JNCL, was conducted in cooperation with parents, pedagogical personnel in the municipality and a Norwegian resource centre between 2014 and 2016. The young man had just moved from school to a residential home, and started a job five days a week.

The personnel were offered competence building in JNCL, and guidance in how to cope with declines in function. *The Book about Me*, written by the parents, was the core and reference book for building competence in the personnel group. The book described the young man’s personality and history, his interests and skills now and earlier, his social life with family and friends, health-related needs and key routines in everyday life.

Objectives and special considerations

The main goal of the project was to establish the best possible quality of life for the young man. The young man had many skills and specific interests at the time when the transition took place, compared to others with JNCL of the same age. The main objective of the project was to maintain the young man’s skills, interests and participation through collaboration between the young man and his personnel.

The figure and model shown in figure 13 was used for competence building of the personnel. D. B. Gray and H. Hollingsworth model describes four different pedagogical scenarios. The model is adapted for pedagogical work with individuals with JNCL. The model was used to address alternative ways of describing daily scenarios and or defining alternative targets:

Figure 13. The performance and participation model

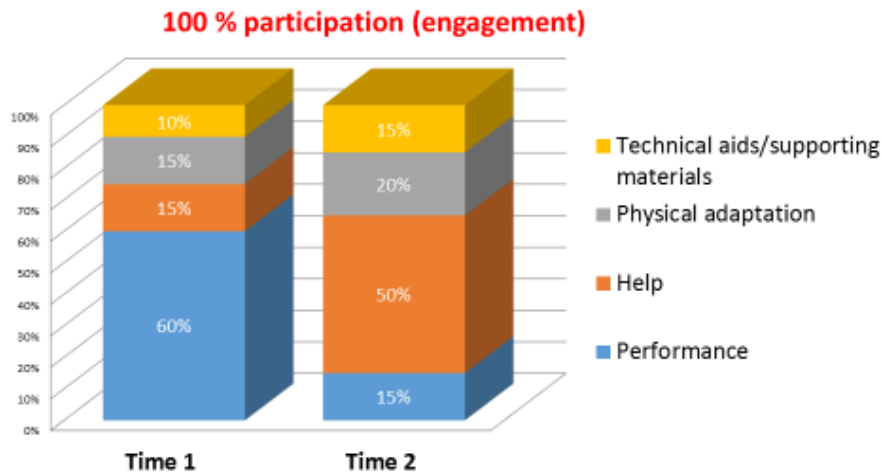
	Ability to do activity	Inability to do activity
Participation	1. Assessment shows ability to do, combined with participation	3. Assessment shows no ability to do, combined with participation
No participation	2. Assessment shows ability to do, combined with no participation	4. Assessment shows no ability to do, combined with no participation

Figure 13 outlines four scenarios where ability or performance is seen in relation to participation. In an ordinary pedagogical context scenario 1 will be the obvious target scenario, i.e. the individual participates and performs. However, scenario 3 may also be a target scenario for individuals with JNCL. At a certain stage of the disease, it will be unreasonable to expect that a person with JNCL can retain performance in a given life situation, but participation can still be possible based on factors such as “active” presence, commitment and engagement.

Figure 14 is showing that loss of individual capacity is replaced by a successive increasing degree of assistance and/or other adaptations. The figure is illustrating a specific collaborative way of working named as the Team Model. The team model was implemented in the project.

Figure 14. The team model

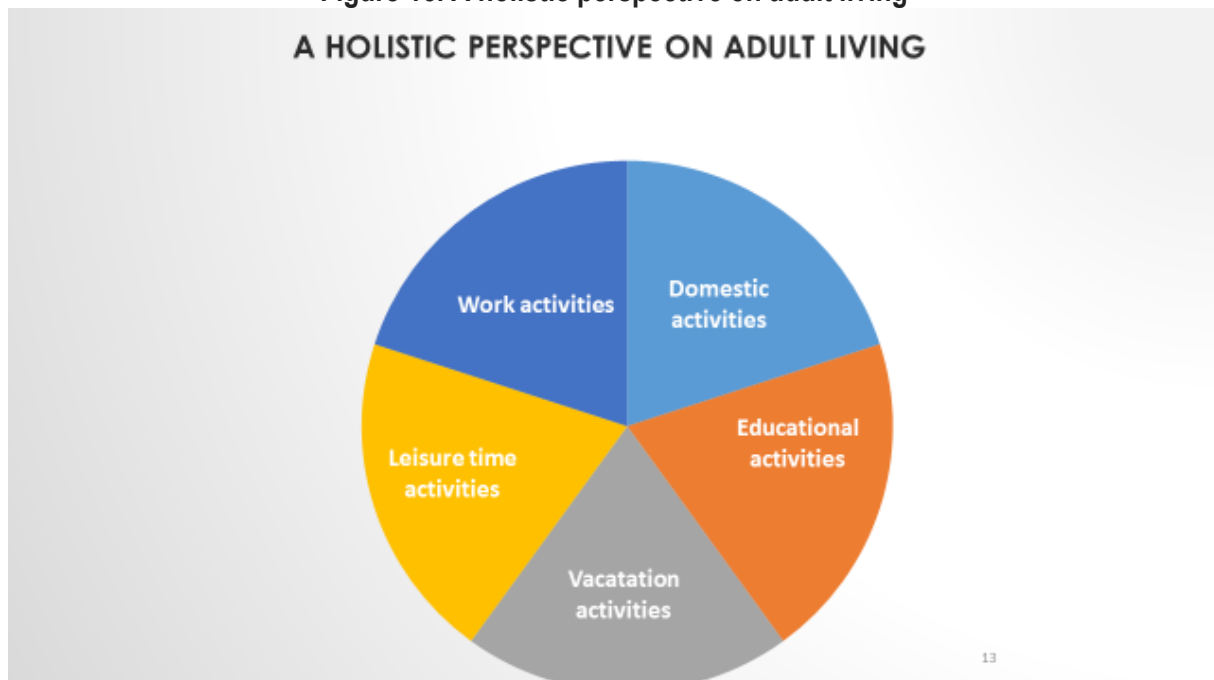
The team-model



The left bar in figure 14 shows that the young man was participating 100% in a life situation at time 1. Participation was largely based on his ability to perform. Eventually the young man's ability to perform diminished as a result of the progression of the disease. However, the level of participation was maintained at 100% because the decline in performance was supported by personal assistance and other forms of interventions.

The following question was addressed in the project - *what is a good and stimulating life for the young man?*

Figure 15. A holistic perspective on adult living



The project group in collaboration with the young man identified a large number of desirable activities in each sector shown in figure 15. The activities chosen were based on the young man's abilities, his history, interests and his preferences. The identification of activities were always considered under the framework of the team model – *what*

can the young man participate in when he is assisted by a helper? Two "participation banks" were developed, one for his residential home and one for his workplace.

Results

The young man's participation in activities was recorded daily in combination with his sense of well-being and his performance.

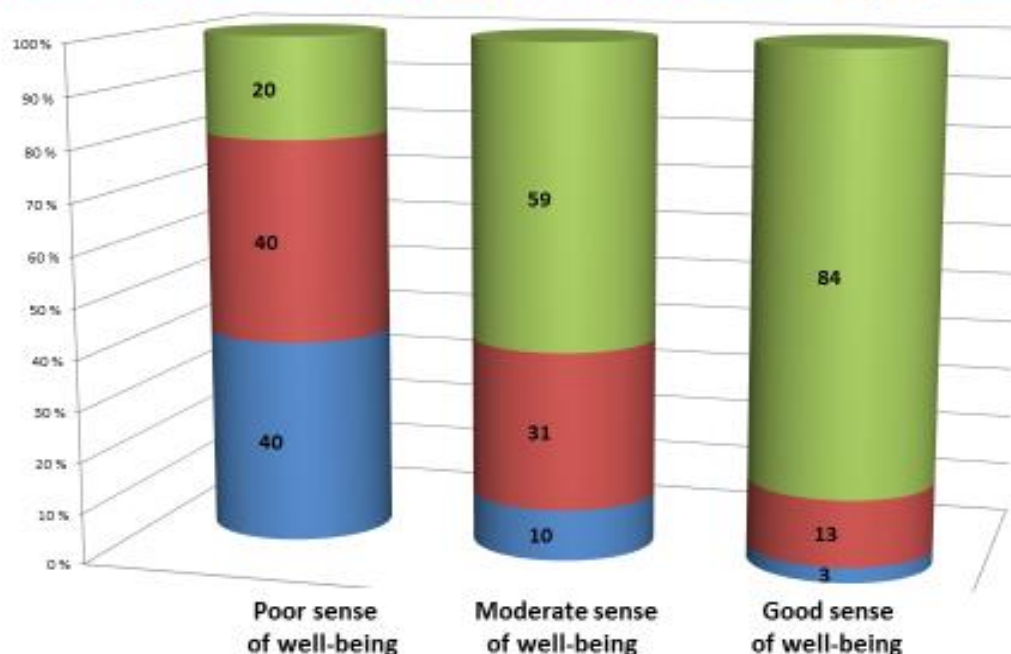
The significance of the sense of well-being

Figure 16 is based on 297 daily registrations and shows the coherence between the sense of well-being and the level of performance.

Figure 16. The relation between sense of well-being and performance level

Sense of well-being versus performance in activities (relative distribution)

Green colour = good performance, Red colour = moderate colour, Blue colour = poor performance



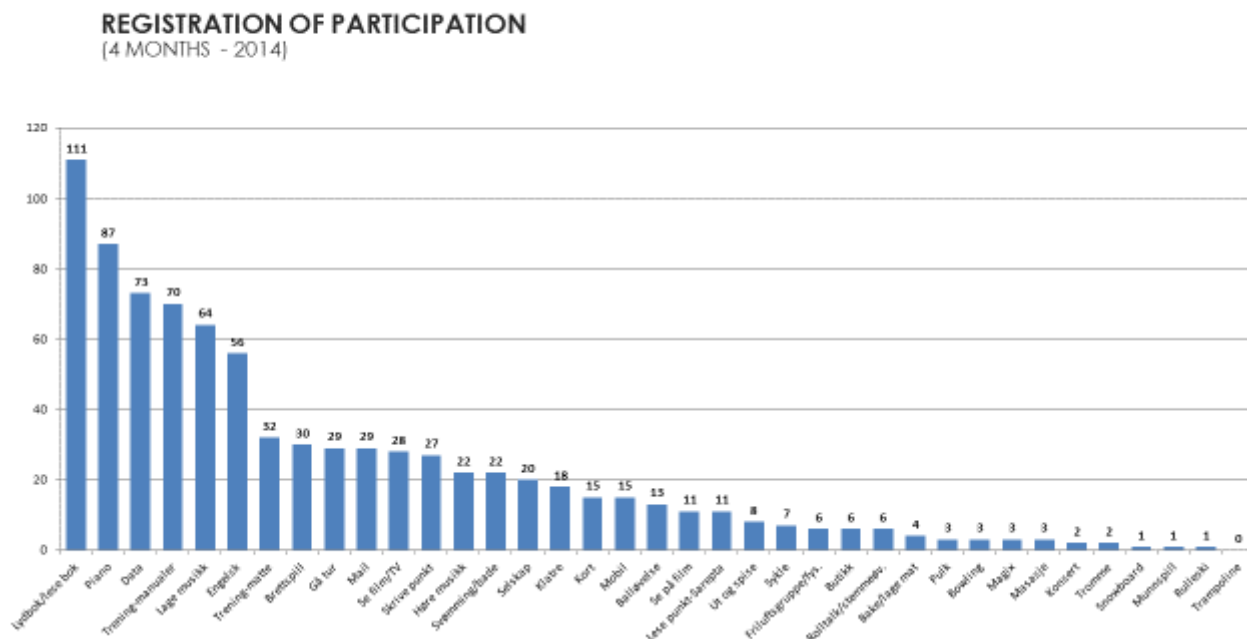
Registrations showed 100 days with poor sense of well-being, 100 days of moderate sense of well-being and 97 days of good sense of well-being.

Figure 16 shows a clear correlation between sense of well-being and performance. However, the personnel stated that they had different expectations of performance according to the young man's sense of well-being; it is therefore difficult to say whether the level of performance was caused by the personnel's expectations or by the young man's loss of performance.

Participation in activities

Figure 17 shows the young man's participation in activities chosen from his participation bank in his residential home for a four-month long registration period in 2014

Figure 17. Participation in various activities



The young man participated in 839 recreational activities during the four-month period in 2014: that gives an average of 7.0 activities per day. The most frequent activities were 1) Reading audio books, 2) Playing the piano and 3) Undertaking computer activities. These are indoor activities but the need for physical activity was also well catered for. He was involved in physical exercises 131 times during the period, on average more than once per day, activities the young man really loved.

The disease symptoms became more and more pronounced during the period from 2014 to 2016. The decline in motor performance and speech was particularly noticeable, while mood and cognition remained at a relatively good level.

A comparison was made between the participation level in a four-month period in 2014 and the same four-month period in 2016. The young man participated in 7.0 activities per day in 2014 and in 6.9 activities per day in 2016. A few activities terminated during the three years, for example using a climbing wall, while other activities like sitski (equipment for cross-country skiing while sitting) was introduced with success. The proportion of physical activity was as high in 2016 as in 2014, but the activities were adapted to accommodate the physical decline.

Conclusions

During the three-year period it was possible to maintain the participation level despite the declines in motor performance and communication. There were high fluctuations in the performance level from day to day depending on the sense of well-being, but this had little impact on the level of participation. Some personnel were able to maintain a higher degree of participation for the young man than others. The amount of help provided also varied in the personnel group. The parents and the personnel concluded that it was possible to maintain a high level of life quality despite all the declines.

4. CENTRAL PROJECT EXPERIENCES

5.a INTRODUCTION

Today it is not possible to stop the progression of JNCL, but it is possible to compensate the effects of some declines with pedagogical interventions. Personnel working with individuals with JNCL need continuous pedagogical input to understand this. JNCL is a rare and complex disease. It is unreasonable to expect personnel in the community to have sufficient knowledge to provide the individual and their family with targeted pedagogical interventions without guidance.

Interventions for individuals with JNCL must be based on perspectives of childhood dementia and corresponding windows of opportunity. The project results indicate that a targeted pedagogical agenda, based on windows of opportunity, will increase the individual's quality of life.

5.b EDUCATIONAL PRINCIPLES AND STRATEGIES

The project has defined some central principles and strategies for pedagogical work with individuals with JNCL. The pedagogical principles are:

- Regarding visual decline: *Decline can be compensated by learning, adaptations, technical aids*
- Regarding cognitive decline: *Decline can be compensated by learning, daily stimulation, repetitions, and adaptations*
- Regarding motor decline: *Decline can be compensated by physical activity, technical aids, and adaptations*
- Regarding communication decline: *Decline can be compensated by learning, technical aids, adaptations*
- Regarding declines in independence: *Decline can be compensated by learning, adaptations, technical aids, and a successive expanding focus on autonomy*

The project has identified five learning strategies related to childhood dementia: 1) Hastened learning, 2) Proactive learning, 3) Skill-based learning and stimulation, 4) Participation-based learning and stimulation and 5) Life-flow.

Hastened learning is about early learning of complex skills and the establishment of personal competence and interests that may be difficult to learn or achieve later in life. Experiences show the importance of building capacity at a young age. This applies to complex skills such as reading and writing, the use of ICT etc. and the early development of life-long interests such as music and sports.

Pro-active learning is about skills not needed today but in the future. This is a particularly important strategy at a young age. It is about preparing the child for future symptom developments, for example in communication. Project experiences show that basic knowledge of JNCL is necessary when negotiating for resources to implement pro-active learning, for something that works well today but will be a challenge in future.

Skill-based learning is about achieving predefined goals, often related to curricula-based learning goals for students. This is a particularly important strategy in young children with JNCL. Examples of pre-defined skill-based learning are skills in reading and writing, mathematics, skills within activities of Daily Life etc. Skill-based learning is associated with explicit learning, see below.

Participation-based learning is about learning through engagement in everyday life activities to achieve development and maintenance of undefined or curricula-based learning goals. This is an important learning strategy throughout life, but in particular for individuals with severe dementia when it is difficult to pre-define learning goals. Participation-based learning is linked to implicit learning, see below.

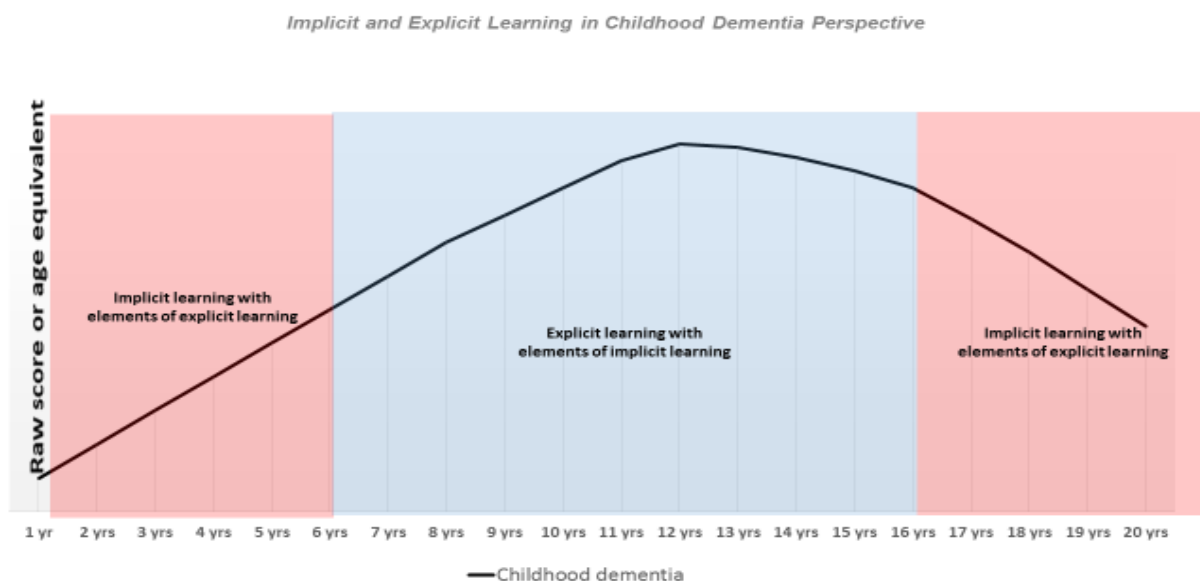
Life fluency is about maintaining continuity of desirable interests, activities, social and community participation, maintenance of skills etc. throughout life. This strategy is, in particular, important in times of transition such as change of school, the change from being a student to adult living etc. Life-flow means that planning and interventions must ensure continuation of the person's history and background.

5.c EXPLICIT AND IMPLICIT LEARNING

Hastened Learning, Proactive Learning and Skill-Based Learning are often most important in an early dementia phase, before the loss of cognitive functions has become too prominent. These strategies require explicit, target- defined learning. Participation-based learning is based on implicit learning, which is an intuitive form of learning. Participation-based learning is often taking place during shared activities with helpers. It becomes more important when explicit learning becomes more difficult. The focus on Life Flow becomes more important during the severe dementia phase. Major changes in life content will require new learning and personal adaptations, which in this phase should be avoided when possible.

Figure 18 shows an illustration of the variations in explicit and implicit learning in a life-long perspective for individuals with JNCL.

Figure 18. Explicit and implicit learning in a life-long perspective



The figure shows that children during early childhood learn and develop through implicit learning (the pink field on the left in the figure). Explicit learning becomes more pronounced when children attend school (the blue field in the figure). This period will also include important implicit learning, e.g. learning of social norms and unwritten cultural codes through participating in the community. At some point, it becomes more difficult to predefine learning goals for individuals with JNCL because of the progressive development of dementia (the pink field on the right in the figure). Facilitating implicit learning through participation and involvement in everyday life situations is vital at this stage. Traditional school-based learning of knowledge and skills is essentially explicit, while participation-based learning and stimulation are often implicit.

5.d THE EDO TOOL (EDUCATION DEVELOPMENT OBSERVATION)

The different strategies can be linked to different phases – windows of opportunity – in the development of the disease. What is important is that the right interventions are implemented at the right times. The large individual variation in the course of disease in individuals with JNCL means that learning capacity and skill levels cannot be predicted by biological age. The project product EDO (Education Development Observation) is a tool that can be used to define good pedagogical practice at different stages of an individual's life. EDO has defined the following pedagogical diagnosis-specific core areas: (1) Vision, (2) Communication, (3) Writing and Reading, (4) Social Life, (5) Gross motor performance, (6) Fine Motor performance, (7) Physical Activity, (8) Behaviour, anxiety and mood, (9) Attention and memory, (10) Autonomy, (11) Interests and equipment/aids. EDO can be used once a year in a lifelong pedagogical perspective; inside, outside and beyond school, and in particular in connection with major transitions.

5. EPILOGUE

Personnel working with individuals with JNCL need systematic information and knowledge about the individual and about the disease. Such information is necessary to maximise the individual's potential at all times. Working with children, adolescents and young adults with JNCL also requires an interdisciplinary approach to address the individual's many needs. This means close collaboration between health service, advisory services, school or adult services, the person with JNCL and parents. JNCL is so complex and so rare that it requires special insights of all professionals involved. Project results have shown that good service provision for individuals with JNCL depends on relevant and available services.

It is always important to consider possible barriers in the environment when planning for pedagogical interventions. When intervention does not work, it can be the result of the system that was unable to initiate appropriate measures at the right time.

Children, adolescents and young adults with JNCL constitute a very small group in society. Some countries do not have a complete overview of the JNCL population, and this has influenced the sample of the project.

There are differences between project participating countries concerning how services are organised. This has influenced the project results. The families that have contributed to data collection are families who are known by service providers or have been active members of a family organisation.

6. ACKNOWLEDGEMENTS

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LITERATURE

- Elmerskog, B. & Fosse, P. (red.). (2012). *Lære for livet, en innføring i pedagogikk ved Juvenile Nevronal Ceroid Lipofuscinoce* (JNCL). Oslo: Akademica forlag.
- Gray, D. B. & Hollingsworth, H. (1999). *Development of a new measurement tool for studying participation in context*. American Public Health Association Annual Meeting, Chicago, Illinois.
- Helland, I., Barlind, J., Åbyholm, G., Paus, T. & Rokne, S. (2016). Veileder for symptombehandling ved Nevronal Ceroid Lipofuscinoce. Publikasjonen kan bestilles ved www.nsvf.org
- Hempel, C. (2010). Hentet fra 20.09.2017 fra <http://addiandcassi.com/dementia-in-children-and-teens-when-kids-brains-regress-like-the-elderly/>
- Jensen, J. (2016). Hentet 15.10.2017 fra <https://sml.snl.no/plastisitet>
- Mole, S., Williams R. E. & Goebel, H H. (2011). *The Neuronal Ceroid Lipofuscinoses* (Batten Disease). Oxford, UK.: Oxford University Press.
- Shapiro, E.G. & Klein, K. A. (1994). *Dementia in childhood: Issues in neuropsychological Assessment with application to the natural history and treatment of neurodegenerative storage diseases*. I: Tramonata, M. G. & Hooper, S. R. (Eds.) *Advances in Child Neuropsychology*, vol. II. New York: Springer, 119-171.
- SPSM (2016) Att gå bredvid” <https://www.spsm.se/kurser--aktiviteter/nyheter/nyheter/en-sallsynt-film-om-en-sallsynt-diagnos/>
- v. Tetzchner, S. (1993). *Utviklingen hos mennesker med Spielmeier-Vogt-syndromet med spesiell vekt på språklige og kognitive funksjoner*. Melhus: Tambartun kompetansesenter
- v. Tetzchner, S., Fosse, P. & Elmerskog, B. (2013). Juvenile neuronal ceroid lipofuscinosis and education. *Biochimica et Biophysica Acta (BBA) - Molecular Basis of Disease*, Volume 1832, Issue 11, 2013, 1894-1905, ISSN 0925-4439, <https://doi.org/10.1016/j.bbadis.2013.02.017>. (<http://www.sciencedirect.com/science/article/pii/S0925443913000628>)
- v. Tetzchner, S., Elmerskog, B., Tøssebro, A.-G. og Rokne, S. Ikke publiserte resultater fra det pågående prosjektet.
- United Nations (2006). *Convention on the Rights of Persons with Disabilities*. Hentet 07.02.2012 fra <http://www.un.org/disabilities/default.asp?id=259>
- Unesco (1972/2005). *Learning to do. Values for learning and working together in a globalized world. An integrated approach to incorporating values education in technical and vocational education and training*. Hentet 18.03.2012 fra http://www.unevoc.net/fileadmin/user_upload/pubs/Learningtodo.pdf