



Review

Juvenile neuronal ceroid lipofuscinosis and education[☆]Stephen von Tetzchner^{a,*}, Per Fosse^b, Bengt Elmerskog^b^a Department of Psychology, University of Oslo, P.O. Box 1094 Blindern, N-0317 Oslo, Norway^b Tambartun National Resource Centre for the Visually Impaired, Melhus, Norway

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ABSTRACT

Juvenile neuronal ceroid lipofuscinosis (JNCL) is characterized by severe visual impairment with onset around age 4–8 years, and a developmental course that includes blindness, epilepsy, speech problems, dementia, motor coordination problems, and emotional reactions. There is presently no cure and the disease leads to premature death. There have been few studies of non-medical intervention for individuals with JNCL, probably because of the negative prognosis. The present chapter discusses the education of children and adolescents with JNCL on the basis of current knowledge about the variation in perceptual, cognitive and language abilities through the course of the disease, and the possibilities that exist for supporting coping and learning within and outside the classroom. Adapted and special needs education may contribute significantly to improved learning conditions, better maintenance of skills and less frustration for individuals with JNCL. This article is part of a Special Issue entitled: The Neuronal Ceroid Lipofuscinoses or Batten Disease.

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1. Introduction

The neuronal ceroid lipofuscinosis (NCL) is a group of inherited, progressive neurodegenerative diseases that affect the brain and usually also the retina. Onset varies from infancy to adulthood, and they share the combined characteristics of retinopathy, dementia and epilepsy [1–4]. Categorizations of the NCLs have varied through the years [5,6] and recent classifications include 9–14 genetically distinct disorders [3–5] and 360 different mutations [7]. The juvenile form (JNCL), also called Spielmeier–Vogt disease or Batten disease, is caused by mutations in the CLN3 gene. It is characterized by a severe loss of vision which becomes noticeable around age 4–8 years, with a developmental course that includes blindness, epilepsy, speech problems, cognitive regression, motor coordination problems, and emotional reactions, including anxiety and depression. There is no cure yet and the disease leads to premature death [1,2,8,9].

A considerable number of articles and books have been published on JNCL since Stengel [10] in 1826 described the course of the disease in four siblings in a small Norwegian town, Røros [2,6]. There have been few studies of non-medical intervention for individuals with JNCL in general, probably because of the negative prognosis and the low incidence. Publications with an educational perspective hardly exist. Research publications about cognitive functions rarely mention

any implications the findings may have for education. Medical, psychological and educational professionals seem to have focused more on the disease itself and its devastating consequences for the individual's development, than on the educational challenges and possibilities. The present article discusses the education of children and adolescents with JNCL on the basis of current knowledge about the variation in perceptual, cognitive and language abilities through the course of this and some other diseases with cognitive decline in childhood [11], and possibilities for supporting coping and learning within and outside the classroom. The research literature is limited and the article also includes information from our own and others' practice. The main focus is on the school age and the learning process, and not on the emotional and behavioral reactions that often appear in the course of the disease [12,13], although it is believed that an adapted educational situation will contribute significantly to improved learning conditions, better maintenance of skills, less frustration, and hence less severe emotional reactions for the child or adolescent.

2. Learning and meaning through the life span

It is an inherent characteristic of education that it changes as the students get older. For most children, including children with severe disabilities, education is a cumulative process where the promotion of new knowledge builds on already established competence. Due to their gradual mental and cognitive decline, children and adolescents with JNCL have a learning curve that is very different from children with a normal progression. Technically JNCL is a form of dementia. The use of this term is however controversial in connection with

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children because of the opposing interactive forces of continued progress in a child's development and the counter-developmental effects of on-going or chronic illness [11]. In addition to a deterioration of visual functions, early signs are often related to impaired short term memory [14] and reduced ability to recall what has been said [15]. At the same time, long-term memory allows the children to activate and recall also events that were prior to the loss of vision and cognitive abilities [16]. Logical reasoning and abstract concepts tend to become difficult at an early age [16,17]. Although the auditory system is less affected by the disease than the other sensory systems [18], children with JNCL may have problems understanding the content of what is said. There are also often changes in behavior, difficulties with concentration, and an increasing need for rest periods [9,13,17,19]. It is not clear if and when learning may cease in the sense that new information is no longer coded in memory, retained and retrievable, but experience from practice suggests that individuals in the advanced stages of JNCL may attend to and remember some new events, indicating that some level of learning may still be possible. Also, the gradual stagnation and decline in mental and cognitive development that takes place vary considerably from individual to individual [20,21].

One consequence of the decreasing abilities is that students with JNCL need continuously adapted education. This means that teachers need knowledge about the functional consequences of the disease and the educational strategies that may support learning and coping [18,22]. Like for other students, the individual educational plan should focus on theoretical knowledge and practical skills that the student may enjoy and benefit from in his or her practical and social life. Also students with JNCL learn best when the educational situation is exciting and positively challenging, and when the content is meaningful and matches their skills and interests [23]. Enthusiasm is a key factor in educational success for both the student and the teachers, and will depend on the meaning making process that is implied in all education [24–26]. Further, putting together a functional education plan for a child with JNCL requires the joint efforts of different professions, including teachers, psychologists, occupational therapists and physicians [22].

2.1. Selection, optimization and compensation

Growing up with a disability represents a way of life that in important ways differ from most children in the community. Children with JNCL typically have to find other ways to play than their peer group and select other types of leisure time activities. Some children have few choices for both themselves and the people who are making adaptations to their environment. The differences between the developmental trajectories of children with JNCL and other children, with or without disabilities, are increasing with age.

In the lifespan model of Paul and Margret Baltes [27], development is described as processes of selection, optimization and compensation (SOC). These processes are particularly useful for depicting development in individuals who are losing many core abilities. *Selection* concerns the individual's choice of relevant and achievable goals, while *optimization* is the process of forming and maintaining the means to achieve the goals that are selected. *Compensation* is the use of new strategies to maintain functions and goals that are no longer sustainable in ordinary ways, for example due to physical weakness and functional regression, making it possible to delay some of the losses that may be emerging [28,29]. A particular strength of the model is its ability to explain both processes of growth and processes related to impairment and decreasing abilities. It has mainly been applied to developmental processes in elderly adults, but the model is useful for describing development throughout the lifespan, including early development and development in individuals with negatively progressing conditions [30].

The concepts of selection, optimization and compensation represent a useful theoretical framework in the present context because they highlight important aspects of the developmental process in individuals

with declining functioning in core areas. A child or adolescent with JNCL will together with parents and professionals select actions and activities that may scaffold "learning in participation". Optimization may include guidance of the adults and peers in the environment, and compensation may include special training as well as adaptation of the education and the classroom.

3. JNCL characteristics of importance for education

It is a commonly held view that education of all students – both normally developing and disabled – should take into account the students' strengths and limitations, as well as possibilities and barriers in the educational environment [31].

3.1. Adaptive and special needs education

Special needs education may be defined as the adoption of educational goals outside the ordinary curriculum, which imply the utilization of strategies not usually applied with the particular student's age group. The extent to which special needs education is necessary will depend on the physical adaptations and the educational strategies applied within the normal curriculum, and the educational support of the student. Note that in this definition, special needs education will always imply a change in curriculum. If the adaptations function to make the ordinary curriculum available to students with disabilities, this is defined as "adaptive education". For instance, learning how to read the clock is a goal within the main stream curriculum. This goal can be adapted by including tactile or auditory clocks and reading quarters of an hour instead of minutes if needed. However, learning the mobility route from the classroom to the schoolyard is a goal within special needs education because it is not described within the ordinary curriculum, and the student with JNCL will not share the training with sighted students in a main stream setting. Education for children and adolescents with JNCL usually includes both adaptive and special needs education.

3.2. Negative progression

A gradual reduction in the ability to learn and maintain knowledge and skills is a defining characteristic of JNCL which has consequences for all aspects of the educational process. The teacher will have to adjust the goals in accordance with the decline in functions, while giving the student challenges that are appropriate and meaningful for him or her in their present cognitive, linguistic and motivational state [16,18,22].

There are considerable individual differences in cognitive functioning between children with JNCL [19,21], and the disease appears to have a more severe course in girls than in boys [32]. One consequence of the negative progression in abilities and the increasing gap between the learning potential of the student with JNCL and the peer group is a growing need for special education through the school years [16,18]. The unavoidable decline in the functioning of students with JNCL is a challenge to the ordinary teacher role, and may represent a considerable strain on the school-home collaboration [16,18,22].

3.3. Vision

The first symptom of JNCL is typically visual loss due to retinal degeneration at 4–10 years of age [33–35]. The children see normally in the early years, and then show decreasing visual acuity and reduced luminance and chromatic contrast sensitivity. They often have large central retinal visual field defects (scotoma), and may compensate this by using para-foveal or para-macular fixation techniques [17,33,34]. This means that the child uses part of the peripheral visual field rather than the central part, for example by fixating above, below or next to the natural foveal fixation point [17].

There is a considerable individual variation in the visual deterioration among children with JNCL which is important in the educational setting [36]. The children can usually utilize their vision actively for orientation and in leisure activities in the early school years, and many can use their residual vision effectively for learning and reading purposes until the age of 9–10 years, in some cases even longer [17].

3.4. Motor skills

Most individuals with JNCL will gradually become motor impaired, and problems with motor function usually become apparent at 14–15 years [20,37]. It is especially the reduction in fine motor skills that influences classroom activities. One consequence of motor coordination problems is that more attentional and other cognitive resources are used on motor performance, which leaves less to problem solving. Studies have shown that motor activity which requires attentional or other cognitive resources may influence problem solving in normally developing children [38]. A Norwegian study investigated blind students' ability to orient themselves with white cane techniques while they were paying attention to their own movements at the same time. The orientation performance was reduced because the movements had not been automatized, which would have reduced the requirement for allocating cognitive resources for monitoring of the movements [39]. It is part of the functional decline in JNCL that earlier automatized actions will gradually require more conscious effort and cognitive resources, and hence reduce the cognitive resources that can be allocated to other tasks. Moreover, some skills that may have to be performed in different ways will require new learning and more cognitive resources. Problems related to cognitive economy may also explain why cognitive assessment has been found to discriminate better between individuals with milder than more severe degrees of motor impairment [19].

There is no absolute relationship between motor impairment and cognitive decline in JNCL. When motor skills are more affected than cognitive skills, a student with JNCL may know what he or she wants to do but be unable to do it. In order to promote active involvement as long as possible, favored activities should be physically adapted or given a slightly different content. For example, a child may be able to do creative art work if the building blocks are larger, or the focus may be directed from swimming activities to alternative bathing activities such as water play [18]. A reduction in fine motor skills may influence Braille reading and make it necessary to change to reading with auditory technology [12,18].

Movement is more than getting from one place to another. The development of self-propelled movement is an important psychological milestone that positively influences the child's relational and emotional life in general [40]. Deterioration of this ability is therefore likely to have a negative influence on development. In childhood and adolescence gross motor skills may influence the range of educational and other experiences of students with JNCL, as well as social participation with peers and others, and physical well-being [18]. Many activities may have to be adapted or replaced in order to maintain social interaction with the classmates and other peers.

3.5. Cognition and language

Problems in cognition and language are sometimes apparent before the visual problems have become notable, but the cognitive decline is usually slow during childhood [41,42]. Some of the reduction observed in intelligence quotients (IQ) will be due to the lack of new learning (especially in the early phases), while some will reflect a loss of earlier acquired knowledge. Seizures are present in most individuals with JNCL, and EEG tends to deteriorate slowly as the illness progresses [43]. On the average, learning difficulties become apparent at the age of 8–9 years, epilepsy at 10–11 years, and speech and language problems at 12–13 years [20]. Lamminranta and associates [44] found that in five years from age six to ten years, the average verbal IQ on the

Wechsler Intelligence Scales for Children (WISC) decreased from 88 to 72, but there were considerable individual differences. Because cognitive assessment of blind children typically is based on verbal scales, it may be difficult to distinguish cognitive from linguistic abilities [45].

3.5.1. Memory

Memory represents the preservation of events that took place a shorter or longer time ago, often many years. Memory is about the individual's past experience and history, but remembering events makes it also to some extent possible to predict the future, thereby creating continuity between past, present and future. Speech, writing and other cultural tools also make it possible to organize and store knowledge in ways that exceed the memory of the individual. Memory is an essential element in all learning, and significant decline of memory and one more cognitive area is a defining characteristic of dementia [11].

Working memory is a system that individuals utilize when they try to remember something in a short time or are working with a problem, as when children are looking for a piece that fits into the puzzle. It has limited storage and processing capacity; the content is what the person is aware of, and it is quickly reduced – in a matter of seconds – if it is not refreshed. Working memory is important for learning. It processes information from the outside world and from long-term memory, and helps to bind together new information and what is already stored [46,47].

Children with JNCL seem to develop particular deficits in working memory which limit their ability to learn new things [16]. On the average, children with JNCL score lower on Digit span than the other verbal subscales of the WISC-R [19,44]. Working memory is assumed to be important for the acquisition of language and of reading and writing [48,49], and reduced working memory capacity has been suggested to make reading acquisition difficult for students with JNCL [14]. Also children with Duchenne Motor Disorder (DMD) show impairment of working memory but their problems are specifically related to verbal material [50]. Although both conditions are characterized by severe deterioration and premature death, cognitive development is different in DMD and JNCL.

Long-term memory includes storage of both short and long duration, and it is considered to have unlimited capacity. However, usage of what is stored depends on an ability to retrieve information when needed. Moreover, long-term memory is divided into systems with somewhat different functions. *Procedural memory* is about how something is done, and comprises a variety of skills, including the ability to form habits and learn skills. *Declarative memory* includes everything that can be expressed explicitly or be “declared” as an image or a statement, objects, persons, names, places, events, and so on [51,52]. *Episodic memory* is the personal part of declarative memory, the individual's life story [53].

The long-term memory of students with JNCL is supposed to remain relatively intact [16] and may thus be a resource in their education. Similar to people with other degenerative diseases affecting learning and memory (e.g., Alzheimer disease), individuals with JNCL appear to retain distant events better than memories of more recent events. They seem to preserve many aspects of what they have experienced and learned, and may recognize songs and events they are being told about [54]. This may be possible to utilize in many classroom activities. However, memories may represent static or slowly deteriorating knowledge rather than a learning ability that can be applied to new tasks.

3.5.2. Language and communication

In most individuals with JNCL, language and communication skills will deteriorate, but the deterioration will not always follow the same course and speed of progression. There are large individual differences, with some individuals showing the first signs of language problems in the early school age, others may not show such difficulties until well

into their teens. In rare cases, the speech disappears completely but poor intelligibility due to articulatory apraxia is a more common problem than a total lack of speech [45,54]. There are very few detailed descriptions of language and language change in individuals with JNCL. Some of the changes that have been observed are increased dysfluency and a greater prevalence of omissions of inflections at the end of the word, splitting of words, word-finding problems, incorrect or idiosyncratic use of words without showing awareness of it, odd constructions like “confirmation ingredients” and “world champion dancing dress”, syntactic errors and problems initiating conversations [41,45,55].

The problems students with JNCL experience in learning are probably a result of an interaction between cognitive disorders, speech and language disorders, and contextual difficulties caused by the visual impairment. The language problems may be related to the impairments observed in working memory functions in individuals with JNCL, as reduced function in the working memory is considered a core factor in developmental language disorders [56]. Their cognitive problems may also lead to difficulties in perceiving situational cues that contribute to activate relevant words. Word finding problems are common in language disorders [57,58]. For some people with the JNCL, word finding problems may be most apparent, while dysfluency of speech is the greatest problem for others. As the stuttering or cluttering progresses, the speech may become unintelligible even to those who know the person well [45,54].

Also language comprehension deteriorates, but there is usually a considerable gap between comprehension and expression of speech, which may make communication aids an important tool in intervention [59,60]. In fact, vocabulary may continue to increase when other cognitive functions have started to decline [19]. Both comprehension and production seem to be influenced by the familiarity of the situation, and as the disease progresses, the individual's comprehension becomes increasingly dependent on being in a situation that is familiar and does not contain too many disturbing elements [18]. This means that it is important that the classroom represents a stable environment and that teaching is appropriately situated. The cognitive problems may lead to fragmented experiences, which in turn may make language comprehension fragmented and lacking in coherence.

The written language represents a problematic area. Some children with JNCL have been observed to write short sentences, have a tendency to just list subjects, show an increase in the use of “also”, use “I” and “me”, and never “we” and “us”, split words without hyphens, and have new problems with spelling, for example beginning to confuse single and double consonants in words that were mastered before. However, some individuals may be able to use written language after their speech has become unintelligible [61].

For blind people, proximal social interaction is mainly based on language use. Many children with JNCL are rather talkative, and may experience comfort from conversations about past experiences [16]. In spite of the children's language problems, language is important for their social closeness and well-being.

There have been some attempts to improve expressive communication through traditional speech therapy [62,63]. It has also been shown that for individuals with JNCL who use words and word combinations in an idiosyncratic manner, personal dictionaries that describe the idiosyncratic meanings and contextual relations may make it possible for them to maintain a higher level of communication. The deterioration of communication and language skills is a major source of frustration and despair, and may be an important cause for the behavior problems sometimes seen in individuals with JNCL. In spite of this, clinical rating scales and recommendations for care in the later phases of JNCL do not include language and communication competence. The promotion of communication and language should always be an important target in the curriculum.

4. Education

The education of children and adolescents with JNCL will depend on the knowledge of the educational system, as well as at the general traditions in mainstream and special education. There are schools and institutions with considerable experience in working with this group, and some of these have published guidelines for teachers based on general clinical experience [12,18]. There are few studies based on systematic observations, and besides a few case studies there are hardly any studies about the efficacy of the educational practices [22].

The two foundations of education is the choice of educational goals and the adaptations. The teaching and learning activity should be adapted to avoid problems and utilize strengths. This may for example mean giving immediate clues that does not tax working memory, and a short time between instruction and performance.

As individuals with JNCL get older, the focus changes from increasing knowledge and acquiring new competence to maintaining already established skills. Evidence from other diagnostic groups suggests that dementia makes learning more difficult, but not impossible [64–67]. Learning requires more time, efforts, assistance and adaptation. There is evidence that systematic educational efforts may slow down the effects of dementia in elderly people, for example by giving them practical tasks that require skills that are manageable for them [68]. Studies have shown that people with memory impairment may remain more independent and maintain better performance of everyday tasks through 20 minute daily reading [68]. Similar strategies may be attempted with children and adolescents with JNCL, for example by applying objects and sound recordings that activates memories in the student [69,70].

When knowledge and skills are reduced, children and adolescents with JNCL become gradually more dependent on the surroundings, require more assistance and support in everyday tasks, and although they may have helpers around most of the time, participation in society may become sparse. In order for education to promote participation, it should be considered what forms of participation that are possible with compensatory measures. With this form of selection [27], educational activities are chosen with regard to whether they support participation in meaningful activities [18,71]. The result may be different from focusing on skills and knowledge only.

Research also suggests that social settings are most efficient for learning. Humans are social beings and teaching may function best when several individuals work together [72]. Collaborative learning has also shown positive results [73]. Inclusive education will often establish good social settings, especially when the schoolmates know the student with JNCL before the symptoms became apparent. They may include morning gatherings, teamwork, breaks, and practical-esthetic subjects such as woodwork and music. Research indicates that mental activities like attending music, help to orientation and refreshing memories improve cognition and mood in people with dementia and delay the fading of memories [74–76].

4.1. Planning the education

The individual education plan represents an important tool for planning education for students with JNCL and other disabilities [77]. Such plans may vary considerably but they usually include an overview of the student's earlier educational activities and how they functioned, of current educational activities, and of activities that are likely to be implemented within a shorter or longer time horizon. The individual education plan should be made as soon as possible after a child has been diagnosed. There is no reason to wait, even if the situation is challenging for the parents. Working with the plan will give them a focus and possibilities for discussing positive actions in a situation that otherwise may be overwhelming. The education plan will ensure that the equipment needed in the education is ordered and gives motivation and direction to teaching and learning.

Parents of children with JNCL will always have a central role in the education planning, and the quality of the parent–school relationship may be decisive for the child's or adolescent's education and well-being [78,79]. Communication and exchange of ideas and knowledge with the parents should therefore always have a high priority in education planning for students with JNCL.

Even if JNCL represents a severe dementia disease, the individual education plan should focus on how selection and compensation may support learning, rather on decline that will be coming. It should specify possible developments that may enable personal development, and how one may meet potential barriers that the disease may create. This means that the individual education plan will have a preventive function.

4.2. Assessment

For all children with disabilities, the outcome of the educational process is uncertain. Even standards for typical development within diagnostic groups tend to be lacking (but see [80,81]). In order to support selection, optimization and compensation in relation to the student's goals, it is necessary to have sufficient knowledge about the student's strengths, weaknesses, needs, interests and wishes. The differences between various areas of development make a broad assessment necessary, avoiding for example drawing conclusions about cognition on the basis of motor ability [19]. They also underline the role of sensitive teaching as part of a continuous assessment and the need for educational adaptation. Parents are also an important source of information. Other sources of information are ordinary assessment, observations and documentation of the earlier education, as well as computerized approaches [82].

In Norway, the visual functioning of children with JNCL is assessed as soon as the diagnosis is confirmed. This is an assessment, in which acuity levels for distance and near, degrees of para-foveal or para-macular viewing, levels of luminance contrast sensitivity, visual orientation skills, lighting needs and magnification needs during reading are recorded [18,83].

For education purposes, there is a need for regular assessment of the visual functioning of students with JNCL. These do not have to be advanced evaluations of visual functions in an eye clinic, as this might direct the student's attention to his or her loss of vision. It is sufficient that the teacher, with the support of vision specialists, observes the child in different activities, evaluates functions and needs, and make the necessary adaptations in the environment or in the educational means or strategies, without making too much fuss about it.

4.3. Selecting educational goals

Selection of goals is a core element of making an individual education plan. The plan defines which areas of learning that are giving priority, and which areas that are lower on the list. Selecting goals means to select activities. For example, tactile learning becomes more important when the sight disappears, and the plan should identify activities where the student can use the hands for exploration and creation. Change of school and other transitions are always important elements in the plan. The short-term plans are more detailed than those that are farther away in time. In addition to giving priority to some activities over others, individual education plans contain strategies for optimizing the fulfillment of the educational goal, as well as a list of possible compensatory measures. Because the actual course of learning (and of the disease) in an individual with JNCL is not known beforehand, it is necessary that the plan contains a variety of measures that the teacher can choose from when it becomes clear how the student's learning proceeds. It is crucial that the current and future teaching needs of the individual and the individual education plan are in harmony.

The goals specified in the individual education plan for a student with JNCL have to be realistic and achievable, concrete and well

described [18]. If goals are too generally formulated, teachers who have limited experience with JNCL may not be able to create functional activities from the goals. At the same time, it is important to describe goals that are dynamic and adjustable, allowing a diversity of developmental paths. Dynamic goals may imply re-selection and compensation. When considering goals, the time it takes for the student to perform an activity or learn something will become more and more important with age. This raises the questions of what is necessary to learn, how the knowledge is going to be used, and how the achieved skills can be applied in everyday life. For example, as the disease progresses, there may be fewer traditional academic activities and activities of daily living gradually may become a larger part of the curriculum, including going to the canteen and “boring” activities like dressing and undressing [16]. It is an overall aim that the student remains as independent as possible in such activities, even if they take a disproportionate amount of time. When a student does not achieve or maintain academic goals because of the course of the disease, this should not be perceived as failure, because the activity implied in the attempt may have been positive for the student and this may be more important than the final achievement [84].

The individual education plan is a tool for ensuring an appropriate amount of cognitive and physical activity. Experiences from New Zealand and Norway suggest that the individual education plans for students with JNCL tend to have a large proportion of goals related to entertainment and enjoyment, and rather few related to the promotion of learning [18,22]. Moreover, although the student may have an increasing need for rest, overestimating this need may lead to an unnecessary increase in segregated time during school hours [18]. An imbalance between activity and rest may also imply an imbalance between social involvement and isolation. The main focus of the individual education plan should be activity and involvement, not rest, and a sensitive teacher should cater for rest on a day-to-day basis when needed.

4.4. The student as the center of a system

The individual education plan will always include structure and organization, describing the arrangements and adaptations of the classroom, the teaching and the diverse everyday situations at school according to the student's level of functioning [18]. However, the description of organizational structure should extend beyond the student's own activity and include the whole school as both a social and an educational system. This system includes the school's physical and social organization. As the disease progresses, students with JNCL will fall behind their peers in most subjects and may spend much of their time in a smaller classroom, often shared with 2–3 students with other disabilities. This classroom should have a location near the rest of the class and with optimal access to other parts of the school and for other students to visit, for example allowing classmates and other peers to visit it easily in breaks.

Some students with JNCL have a strong need for predictability. They are vulnerable to unexpected changes in the activities of the day schedule, staff and the rhythm of the school as a whole [18]. The individual education plan should specify such needs, as well as how the school as a whole will adapt to these needs.

Teachers of students with JNCL will usually experience that many aspects of their teaching take more time compared to the past, in particular the teaching of new skills. Also established educational activities may gradually take longer time [18]. This does not mean that these activities should be abandoned immediately, although this may be necessary later. The focus should rather be on how certain forms of compensation may decrease the time usage without losing active participation. It is also important that the teacher takes into account that activities that take more time also require more effort.

Good educational practice is based on teachers having appropriate expectations to the student and seeing the possibilities which may

require commitment and initiatives from the teacher or assistant. Participation stopped because of inactivity may pose a severe threat for students with JNCL. Over time they may lose the ability to influence their situation. This represents an important educational and ethical dilemma [85,86]. The aim is that participation should lead to activity, learning and maintenance. Reducing participation will also reduce the most common bases for learning and stimulation. Maintaining educational and social participation should be the first working rule for a teacher who becomes responsible for the education of a child with JNCL.

5. Educational phases

The education of students with JNCL will go through phases where the student will have gradually lower vision and abilities to learn. In positive terms of what the student is able to do, these phases may be considered as “learning windows” and hence opportunities for teaching particular skills. The development of educational strategies will be from utilizing the residual vision of the student to having to rely on auditory and tactile means [18].

5.1. The early school years

In the early school years, the cognitive problems of students with JNCL may not be very pronounced, and they may be able to follow some of the ordinary curriculum [18,20]. There is a gradual deterioration of visual functioning but in the first 1–3 school years it is usually possible to use visual teaching strategies. Importantly, in this phase, the student may learn skills like using a knife and fork and tying the shoe laces through visual observation. From an educational perspective, it is a strength that students with JNCL have had normal eyesight for some years and are able to “think” visually. For instance the student will be able to associate visual experiences with later tactile or auditory stimuli. The residual sight may also be utilized to optimize learning of mathematics and many other complex academic skills as this may be easier for the student to grasp with the use of the visual sense [18]. This means that teachers should facilitate visual learning as long as it is possible, and monitor the student’s visual functioning carefully in order to know when it is necessary to introduce strategies based on other modalities.

Experience shows that much of what is learned in this period may be maintained for a considerable length of time. The selection of early educational goals are therefore of particular significance. Some skills are “pivotal” in the sense that they lie a foundation for later learning and functioning, for example spatial orienting without sight and reading. Such skills should be given priority.

5.1.1. Mobility and orientation

All children need strategies for orienting themselves and moving around. Children with severe visual impairment will have to learn to use tactile/haptic and auditory landmarks and guidelines when orienting, sometimes with the help of assistive technology [87]. Adaptation of the environment will also imply having a relatively stable physical environment, allowing the child to use established knowledge about the environment and build up a cognitive map of non-visual cues [88–90].

5.1.2. Reading and writing

Reading – text access – can be visual, tactile (Braille) or auditory with the use of text-to-speech, that is, the text is read aloud with synthetic speech [87]. Studies show that many individuals with visual impairment combine ordinary print, Braille and auditory text access [91–93]. Also students with JNCL may be utilizing all three modalities for accessing text in a relatively short period of time, typically from second to fourth grade. During this period there is usually a shift from primarily visual to tactile and auditory text access.

It has been suggested that deficits in working memory make it difficult or impossible for children with JNCL to learn to read [14]. However, this view is not supported by research which indicates that many students with JNCL benefit from active reading with Braille [21,94]. In the years 1995 to 2011, records show that 23 of the 39 students with JNCL in Norway could read and write Braille. Seven of the children used a Braille keyboard only for writing and one child wrote touch on an ordinary keyboard. Eight students were unable to read visually or tactually, and accessed text with devices with synthetic speech output (personal communication Mohammed Beghdadi, 2012). Independent of mode, it is important that all children with JNCL are given opportunity to experience reading in one way or another [18]. Implementation of reading may both utilize existing interests and contribute to creating new interests, like in the case below.

Peter was diagnosed with JNCL when he was in the second grade. After the diagnosis, the school had visits from a visual resource centre about three times a year. An additional teacher was allocated to the class. One of the early measures was to arrange a Braille course for Peter's teachers and family. A little later, the other children in the class were also introduced to Braille. In the beginning of the third grade, Braille training had become part of Peter's schedule. Together with his father, Peter wrote small histories about Tintin, Lego, Harry Potter, Robinson Crusoe, Star wars, Saturn and places the family had visited in Braille, which became part his ABC.

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Technical solutions contribute to making adaptation flexible [87]. If the students’ visual functioning allows identification of letters, words or pictures, they are provided with magnification systems like reading television systems or computerized systems. Some textbooks have large print that allows reading without magnifying systems. The teacher can print books in different font sizes and thus change easily from one book to another when the visual function deteriorates. This may allow children with JNCL to read at home without technical support, in the same way as their sighted peers.

Some children with reduced vision like to vary between different ways of accessing text while they are still having the visual means to do so [93]. However, starting with Braille exercises while they can still use their residual vision may be problematic to accept for children with JNCL and their parents. The children’s motivation for learning Braille may be low as long as their vision functions adequately and it is most important that they learn the phoneme-grapheme correspondence – the reading code – and this may be done visually, with or without enlarging equipment. On the other hand, early learning of Braille reading may be important as the emerging decline in cognitive impairment may cause both perceptual and learning problems later in life. In inclusive settings, one may approach this by introducing all the children in the class to Braille. The earlier different techniques for reading are acquired, the better the prospects are for using alternative “learning modes” later. This is also the idea underlying the design of “Sarepta”, a multimedia computer program for communication and learning which may be introduced early because it may be useful for all blind students. Sarepta may be used also when motor and cognitive skills are more limited [96,97].

5.2. The later school years

In the later school years, the need for regular activity re-selections and re-schedule of the school day is likely to increase. It will be important to ensure that the activities are interesting and positively challenging [18].

In secondary school, children with JNCL often develop the need for a clear structure in the teaching situations [18]. “Structuring” may be defined as a sequential organization of activities and tasks, and is an important feature of interventions for people with intellectual disability, dementia and other forms of cognitive deficits. A structured school

situation may enable students with JNCL to gain overview and some control over their school day, while more improvised school days without a clear structure may lead to more confusion and more fluctuations in mood. The use of tactile or auditory daily and weekly plans will make it easier to build an understanding of the school day and week [18].

As the students get older, although the course of the disease varies, maintaining skills usually becomes a major focus [18]. Maintenance can be enhanced through repetitions with some variety, and the students' knowledge about their personal experiences may be maintained through frequent memory refreshments or "reminiscence therapy" [98]. The education plan will need to specify learning goals and maintenance goals, and how these can be achieved, for example how often repetitions and refreshments of distant and recent events should be initiated by the teacher.

5.3. Transitions

Education undergoes major transitions approximately every third year. In most countries, the four major transitions in an ordinary school career are from preschool to school (primary school), from grade six to grade seven (starting lower secondary school), from grade 10 to grade 11 (entering upper secondary school), and from grade 13 to university college or a job. These transitions usually involve change of teachers, locations, classmates and support services. Some changes may also imply new rights to special education services.

Transitions may be less pronounced when students attend special schools which tend to cover the full school range, and sometimes also preschool. In Norway and many other countries, children with visual impairments attend ordinary schools, although they may spend a considerable part of the time in smaller group settings. Because of the need for continuous educational adaptation, transitions for students with JNCL may be complex and end up in chaos if not well planned [18]. There is a risk of these students' educational life "taking a longer break" caused by lack of planning, or that the students will meet a misunderstood form for caretaking without an educational focus or learning goals. It is therefore necessary that these students have a transition plan included in their individual education plan.

The main objective of transition plans is to achieve the best possible educational flow, and prepare the next school for meeting the student with JNCL in the best possible manner [18]. It is usually attempted to make "soft" transitions, where the student's current school somehow intertwines with the next school. Important aspects of transition plans are to secure the long-term goals, enable new planners in charge of education to see the longer lines, and to prevent unnecessary discontinuity. Transition plans normally have a wider perspective than educational plans because they also consider life outside school.

6. Some educational strategies

There is still a dearth of knowledge about the education of children and adolescents with JNCL. There is a need to develop new strategies and to gain systematic experience with the use of already established and new strategies, in order to make better selections, optimize methods for reaching educational goals, and developing new ways to compensate functions that slowly are deteriorating.

6.1. Use project work as educational strategy

The project method implies selecting a topic and building different forms of learning around the selected topic. This approach has been successful with students with JNCL [18]. The selection of topic will be based on the student's motivation and interests, for example an interest in rocks, popular music or, like in this case, football.

Frank was an enthusiastic football player and a member of the local team in primary school. When his sight began to fail, his role as a

player was gently and gradually transferred to other club tasks, such as administering materials and supporting players with services. Frank's big interest for football was used to motivate him to learn to read and write Braille and use computers. Over time Frank became a rather capable writer and user of computers, which was followed up in secondary school. When his school started a popular electronic newspaper that was published on Internet twice a month, Frank started a career as a sports journalist. His work included interviews with football celebrities, analyses of different teams, and reports from matches. He was also given the opportunity to act as a sports journalist after completing secondary school, and this became his adapted job. He was assisted by a full-time employed helper and his parents helped him fulfil the assignments. With this help, it was possible for him to visit football matches and players, and to write the interviews and other experiences on the computer. Frank gradually lost the ability to speak and to write, but with assistance from the helpers, it was still possible to engage him in work with the electronic paper. Frank was involved in planning whom to interview, what matches to visit, and how to evaluate different events. He typed the reports from such events together with his helpers. Frank's early interest for soccer enriched his life, and functioned as a gateway for learning.

Projects may incorporate theoretical and practical tasks that facilitate learning, for example of mathematics, foreign languages, science, the use of computers and history. The structure of the project may support communication and collaboration, as well as independence, for example by investigating what tasks one student can do alone, and what tasks that require collaborative efforts. This means that projects may also include social and inclusive goals, where the student collaborates with one or a group of classmates.

Teacher experience indicates that the project format may be an effective educational tool. The project is based on the student's interest and he or she usually focuses with enthusiasm on the project goal, while the teacher uses the process to promote the learning goals that have been selected for the student. There is however a risk that the motivational basis of a project might make the teacher more oriented towards entertainment than teaching. In order to avoid this, the teacher should design a project plan that describes the specific learning goals, and how these goals will be achieved through the teaching in the project.

6.2. Utilize prior visual experience

Children with JNCL show normal visual development and grow up with visual experiences in the first years of life. It is important to utilize these experiences when the student's learning depends primarily on other senses than vision, and to refresh the memories of visual conceptual knowledge. This may be done by talking about visual and non-visual qualities of things and events in a constructive and "conceptual" manner, "translating" from the visual to other modalities, relating qualities to each other and constructing new ideas on the basis of combinations and expansions. This can be done in a number of ways, for instance by starting early using the multi-media programs (see Section 6.7).

6.3. Facilitate exploration

Exploration learning involves thematic integrated-subject instruction and active participation in a project-based authentic learning process [99]. The real community is used for learning through play, theme immersion, activity-based learning and related philosophies to structure an environment that allows students to learn through their own discoveries.

Exploration also implies the use of established knowledge, combining it with new knowledge and other forms of established knowledge. For students with JNCL, exploration may function as an activation of

prior knowledge and thereby refresh their memories of earlier experiences. This may range from identifying simple tactual or auditory sensations to investigating whole or parts of complex events, which the teacher may help to integrate.

6.4. Maintain active participation in learning

This reflects Dewey's slogan "learning by doing" [100], adding a social context. Teaching and learning are complementary processes of an essentially social nature. Activity may in itself promote learning. Participation in educational activities with others will support learning. To be present in a situation means that there will be a potential for learning as opposed to a situation "without presence". There is a considerable risk that the lack of action by many students with JNCL will result in "no-presence" situations, because the inactivity may be perceived as having little meaning by the teacher. It is part of the teacher's job to create engagement, but the teacher may need support to perceive the student's potential to be engaged in the situation. Otherwise the result may be more inactivity and less learning [18].

6.5. Refresh memories

Refreshment of memories is the repetition of earlier experienced events or presentation of objects or story elements related to the event [101]. Research shows that refreshing memories by activating and elaborating old memories may improve cognition and mood in people with dementia and delay the fading of memories [98,102]. Memories may be refreshed by reviewing events and supporting self-activation of memories, that is, by giving clues that activate certain memories [103]. This may be through conversation about the past combined with old cues, that is, natural sounds, songs, voices, speech and objects that are well-known from the past of the person. They may also be refreshed by telling the student histories that they finish or at least acknowledge during the story telling. Active registration of memories may therefore be an effective strategy when teaching new facts and skills [96]. It is therefore important that teachers search for and register the student's knowledge.

6.6. Support communication

Communication is a prerequisite for education, the teacher's teaching and guidance of the students' active or passive learning process are dependent on the communication processes. Good communicative strategies in education may be essential for overcoming the student's comprehension and production problems. This strategy also builds on studies that show that receptive vocabulary may continue to expand when other cognitive skills have started to decline [19]. Although complex utterances may be difficult to comprehend for students with JNCL, using language to provide information may help the students orient themselves, and thereby influence the whole educational situation.

6.7. Utilize tools that support functions

There are many educational and technical tools that may be of help, in and out of the classroom, and today, most students with disabilities are using various forms of computer-based equipment in education. Electronic magnifying devices are used during reading, but may also be used for studying the details of objects and pictures. Such devices should be introduced early and be available during the early teaching of literacy and exploration [87]. The equipment that is used by students with visual impairment in general is often adequate also for students with JNCL, at least in the early stages of the disease. However, the combination of visual, motor, communicative and cognitive declines in students with JNCL may make it difficult to find adequate use of such equipment in the later stages.

When the student with JNCL needs auditory and tactual access to text and exploration, this may be introduced together with the Sarepta program [104]. This program (formerly called "Structure") consists of 15 modules which are operated with the function keys on a standard keyboard or with switches, depending on the student's motor functioning. All modules have speech synthesis, but can also store digitized speech and other sounds. For instance, recordings of the student's voice and interesting events can be stored and used in different modules. Most of these modules may be applied together or independently of each other. The teacher may use Sarepta to create modules with different contents and functionalities, including various projects and assignments for the student. He may for example create a project where the student learn about wild animals, using text, audio recordings of animal sounds, drawings, animations and video [18,104].

The overall objective of Sarepta is to give children and adolescents with JNCL an accessible tool for collaboration, learning, communication and pleasure. The teachers and parents will regularly fill Sarepta with content that is relevant for the student's abilities and life situation, including general academic and practical information, as well as activities of interest to and events experienced by the student. Moreover, the parents, teachers and staff can later rediscover the stored sequences and fragments from the student's life in the form of multimedia text, sound recordings, photographs, video clips and storytelling [18,97].

Sarepta has become a much used tool in education and social interaction with children and adolescents with JNCL in the Nordic countries. In a Norwegian survey of 40 individuals with JNCL, twenty-one used Sarepta on a regular, more or less daily basis, and most of the others had used it at an earlier time [105]. In the age group below ten years, two of the four students used the program, while 14 of the 21 students aged 10–19 years used the program. In the group above 20 years, five of the 15 adults used the program. The text module was the most used by all 21 users of the program, followed by the picture and play modules (18 users). The high utilization of the text module may reflect the importance of this module of the program in literacy activities for educational purposes and in leisure time. The reasons for the lesser use in the adult group may be that they had left school and therefore did not need it for educational assignments, had difficulties using the program due to deteriorating abilities, or lacked motivation. However, the interviews indicated that usage of the program depended on the presence of an engaged person. The housing staff working with the older group may not have had this enthusiasm, lacking both computer competence, knowledge about the program's possibilities and motivation for learning to use the program [105].

Studies show that there is a high turnover of staff in social services, including day centers and sheltered housing [106]. For individuals with JNCL, staff changes may represent a form of transition but relevant knowledge and experience are often not satisfactorily transferred when the staff is changing. One result may be that useful available tools are not being used.

6.8. Support meaning making

A main functional goal of education is to promote individual knowledge and skills that enable participation in societal life, including school, work, leisure time activities and social life in general, as illustrated in the case history below.

Anders had an early interest in outdoor activities. The forest was of great importance to him, his family and his friends. Anders had some problems with learning to write and read Braille, but did well in mathematics, social studies and practical subjects like woodwork. He was very independent and his school made inclusion a primary goal. The class spent many hours in the forest and Anders' home,

where different kind of projects related to forestry, hunting, farming, etcetera were introduced to develop his proficiency in mathematics, English and biology, as well as everyday skills. Projects were for example related to building forest cabins, farming and cutting Christmas trees. The close cooperation between the family and the school contributed to Anders becoming a very popular fellow. When Anders decided to join the regional forest school, an upper secondary boarding school, the company "Gourmet firewood" was established. Especially adapted equipment was purchased, including a cutting machine that was controlled with a joystick which Anders could operate himself. Anders and his best friends began to sell the firewood in the region and the company became a commercial success. Anders eventually had to have easier tasks than cutting the wood, but his interest for the company remained, illustrated by this remark from his younger sister: "Is it not possible to talk about other things than firewood in this house?".

Anders showed an exceptional education path for a student with JNCL. It should be noted that his educational (and commercial) success was also a result of open-minded and creative teachers, and a devoted and resourceful family.

6.9. Build on intrinsic motivation

Meaningful activities are intrinsically motivating. In education it is a common strategy to use rewards of different kinds. Such external forms of motivation may function to keep the student in an activity that he or she is not motivated for. However, a lack of intrinsic motivation may reflect that the activity is lacking in meaning for the student [107]. "Meaning" does not imply that the activity is funny or without effort. Even traditional academic skills with high demands can be intrinsically motivating, such as reading and writing for Mary in the case history below.

Mary belonged to an academic family with high expectations to education and the concept of "learning for life". Mary lost her sight rather early and was introduced to creative writing in order to speed up the learning of reading and writing Braille. The main idea behind creative writing is that the students write about things that engage them. With Mary, it was established through a close collaboration between the family and the school. Every day, Mary was given interesting writing assignments that were followed up at home. In a rather short time, she became an excellent Braille reader and writer. Her ability to write turned into a life-long devotion, there were no days without reading and writing, and finally she became an author. She wrote her own stories, summarized long books into shorter ones, and wrote emails and letters to friends and relatives. At the end of secondary school she was still an active writer and reader, even if it took more time and effort than before. Her early project on creative writing had a much bigger impact on her life than anyone could have foreseen.

If at all possible, rather than using extrinsic rewards, teachers should search for activities that are intrinsically motivating for the student.

6.10. Support student narratives

Using narratives in education of students with JNCL may support their meaning making and contribute to coherence in their activities and psychological life space [108]. Narrative is a way of thinking and organizing especially social aspects of the world [26]. Narration may be fictional or personal, and children learn to talk about the past in general before they start making their own personal life story or narrative [26]. Supporting fictional narratives may support motivation and creativity in the classroom, and a fictional narrative may be the core of a project, like in this example.

"The boy who would find the heart" was inspired by a movie called "Chasing the kidney stone" where the main character shrinks himself with a chemistry set and crawls into his grandfather's mouth. Peter's version is about a boy who wants to find the heart in the body. He first told the story spontaneously on a long drive. When he later wrote it down, the teacher asked questions about the size of the shrunk boy, what happened when he came into the mouth, how he came further down into the stomach, whether bile acid has colour, whether there is a kind of music in the heart, and how the brain sends messages in the body.

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Sharing experiences of the past with parents and others is an important basis for autobiographical memory. Children do this through conversations with parents and other adults about earlier events, and the foundation of autobiographical memory is made through internalization of such conversation [26,109]. Through childhood and adolescence, there is increasing coherence and details in the life story [110,111].

Because students with JNCL are able to recognize or recall earlier events, personal narratives may have a pivotal role in the educational activities in late school age. The teacher may present cues that help the adolescent activate earlier (episodic) memories and use them as a basis for teaching general knowledge, building on the relations between personal and general knowledge.

6.11. Inform instead of asking (or controlling)

When people have cognitive problems, it is usual to ask them many questions, which implicitly may function to document that they are demented. One core aspect of dementia is susceptibility to confusion and reduced ability to understand what elements in the situation are relevant for the context and the questions that are asked. Being asked about something and failing to find the answer is likely to increase confusion rather than reduce it. The answers to such questions may indicate that the person is more demented than is the case under better circumstances and should therefore be avoided before a clear context and basis for the question has been established. This means informing the person with JNCL about the situation and the events or what kind of information that is sought instead of asking questions. This may provide the person with the cues that are necessary for understanding the situation and thereby increase the potential for knowing and learning. This strategy, that also may include interpretations of events and situations, is extremely important as it may contribute to maintaining positive social interaction and a feeling of coping, and reduce confusion, stress and frustration.

6.12. Case histories

There is little documentation of the role of education and learning for children and adolescents with JNCL. The case histories presented here illustrate that education may have a significant impact on the life situation for individuals with JNCL. Although it should be emphasized that they may not be representative for students with JNCL in general they do demonstrate possibilities for individuals in this group. One common factor in these histories seems to be that the persons with JNCL established strong interests early in life, interests that could be used in education to promote the development of knowledge and skills. The students' education was characterized by continuity, with each school knowing and building on the work of the former school. There was close collaboration between families, schools and multidisciplinary teams, and the resources were provided that were necessary for continuity in the long-term and goal-oriented education. The important role of the parents in the present case histories cannot be overestimated; they were the "experts" of their children and the final guarantors for flow and continuity. All of these students started their adult life with a rucksack and agenda full of activities, interests

and skills. They had a personal style and image which were respected by the people who were taking over the responsibilities for their adult life. This means that their sheltered workplace and new home took each individual's weaknesses, strengths and life history into account.

However, there is clearly a need for more histories about students with JNCL with typical as well as exceptional educational paths.

7. Teaching support competence

The education of children with JNCL requires specialized and multidisciplinary knowledge about the disease, as well as its implications for teaching and learning that most ordinary teachers and many special teachers do not have [22]. In fact, most professionals may be totally unprepared for getting a student with a terminal disease [112]. The stress related to observing decline in a student with JNCL may lead to severe burnout and strong feelings of failure in teachers, and one teacher should therefore never have the sole responsibility for a student with JNCL. Building up the teachers' competence may be the best way to cope with the stressful situation. It is easier for teachers to manage stress when they know what may happen and what they should do when it does happen. An important task of a multidisciplinary JNCL team is therefore to guide and support teachers and give them the knowledge and skills they need.

8. The future

There has recently been a greater focus on the processes involved in educating children and adolescents with JNCL, especially after the first conference on JNCL and education [113]. There appears to be many useful approaches but there is a lack of evidence in relation to the educational strategies that are currently in use. Case histories represent an early phase in the scientific evidence-gathering process and this approach is still useful as there have been few case stories about individuals with JNCL that include details about observational assessment and interventions related to education. In addition, there may be traditional basic research related to the characteristics of JNCL and educational experiments, provided they can be done in an ethically acceptable and non-intrusive manner. This should be made by universities or in collaboration with universities in order to ensure appropriate theoretical and methodological competence.

There is still very limited knowledge about the typical learning paths and variation within this group, indicating a need for larger observational studies. This may imply a need to develop a standard set of longitudinal observations, which could be collated in data bases, in order to avoid self-fulfilling stereotypes, and more varied observations and descriptions of atypical developmental paths should be encouraged. It would be a natural task for national or regional competence centers for JNCL to register and systematize educational strategies and teacher experiences within their geographical area. International collaborative research efforts may compare the development of students with JNCL in different educational (e.g., segregated and inclusive) and cultural settings.

Research priorities may be many but the central role of language and communication in teaching and learning, and for most other aspects of life, means that researchers may give priority to developing evidence-based interventions that may slow the deterioration of language, compensate for unintelligible speech and maintain communication skills longer.

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