33 - 19 = 34 - 20 = 14 (adds 1 to each number to make the calculation easier) 6 x 18 = 60 + 48 = 108 (multiplies the 10s and then the 1s) 95 : 5 = 50/5 + 45/5 = 10 + 9 = 19 (splits 95 to make the calculation easier)

For students with visual impairment, it is important to memorize the task while they do the calculations. If the student forgets the task, producing a repetition with synthetic speech or asking the teacher to say it again is more time consuming and less flexible than repeating by looking at the task like the sighted students do. When students read the task in braille, they can check the task independently. However, this requires orienting on the page or on the line and is not quite as easy as throwing a glance. Students with visual impairment thus have to use more of their working memory capacity while calculating. Students with JNCL develop problems with working memory but key skills and a basic understanding of arithmetic may enable them to do calculations within all four arithmetical operations (Åberg, 2001). Frequent repetitions may be required and a short mathematics session every day will probably facilitate learning and give better results than longer sessions once or twice a week.

Learning mathematic skills and maintaining already established skills may contribute to maintaining memory functions. There might be a double gain: memory supports learning of mathematics and learning of mathematics supports memory functions.



Figure 12.3 An abacus

Abacus and calculators

Abacus is a tool of Asian origin for doing arithmetic calculations which has been used in many countries for children with visual impairment, especially during the first school years. Abacus exists in many different designs (an example is shown in Figure 12.3). All four arithmetical operations can be done by using abacus. Using the abacus involves specific procedures which should be learned and remembered. The abacus should be introduced early to children with JNCL.

In the modern digital world, there are a number of assistive technologies which can be used in the education, including calculators and computer-based programs with braille or synthetic speech (see Chapter 19). Calculators with raised keys are usually easier to use for students with visual impairment than touch screens. Some calculators and computer programs can present the tasks in synthetic speech, the answers may be given orally with speech recognition, and the feedback is given with synthetic speech.

Practical use of basic mathematics

Teaching practical use of mathematics, like measuring time and money, is important for students with JNCL. These practical skills are connected with management of everyday situations and can easily be used to teach mathematics at different abstraction levels (Elmerskog & Fosse, 2012). Many everyday situations require use of mathematical skills, such as shopping, cooking, using a calendar with months and dates, following daily time schedules, measuring wood or other materials during art class, counting and finding specific amounts. Skills can be trained and afterwards repeated and maintained by being used in natural or adapted situations.

Every week the group makes a dish in the school kitchen. The group consists of eight students, but the recipes are written for four persons. The task is to solve the problem of rewriting the recipe from four to eight people and make a corresponding shopping list. And what happens when there are two extra joining the group for lunch?

Daily life contains many situations where basic mathematics skills are useful, such as when setting the lunch table for the appropriate number of persons at work place or at home, when filling the right amount of water and coffee into the coffee machine, or when playing Yahtzee (Yatzy) or other games, using dice with raised dots (see Chapter 21). When the learning capacity of individuals with JNCL is declining, they can still maintain and use basic mathematic skills that have

been learned and automatized. Assumptions about activity-specific mathematical knowledge also suggest that mathematics may be taught and maintained in specific activities.

Drama in the education of students with JNCL

«By engaging in experiences within drama and the expressive arts, children and young people will recognize and represent feelings and emotions, both their own and those of others. Drama plays a central role in shaping our sense of our personal, social and cultural identity» (Scottish Government, 2009). This aim of the Curriculum for Excellence (Scottish Government, 2009), to recognize and represent feelings, has particular significance for persons with disabilities as they try to make sense of their place in their environment and maintain connections with others.

With the onset of visual impairment, young people with JNCL are likely to become less aware of eye contact, as well as facial expressions, body posture and gesture. Young people with JNCL will need support to maintain their understanding of non-verbal forms of communication, and drama may play an important role in this process. Drama may offer opportunities for the young persons to use their spoken language and vocalizations to explore the meaning of words, sounds and expressions whilst interacting with others. However, as the condition progresses, they will have increasing difficulty imagining themselves in someone else's environment and it can be a real challenge for them to imagine themselves in a different body and in a different context. They may also find it increasingly difficult to accept that an adult is going into a role and sometimes have difficulty with the difference between reality and pretense. As such it is important that the teacher is sensitive to the young persons' needs and challenges them appropriately. If young persons are struggling to perceive their own world, it may be very confusing to try to eject them from that and imagine themselves in the newly created world within the drama lesson. It is important that young people with JNCL feel safe within the drama structures. A second approach lies in the introduction of pupils to the experience of literature, not simply as a vehicle for teaching communication but to provide access to a "cultural heritage" (Grove & Park 1996; Grove 2012).

Experience from working with young people with JNCL indicates that involvement in drama relies on getting the young people to engage with their emotions, with their heart. If they get completely caught up in their learning, they will be more likely to remember the learning and maintain these skills in communication. Young people with JNCL might require material and tools in other modalities to get this engagement, but with the right support they can participate and benefit from drama throughout their education.

The first step: Finding a good story.

At the Royal Blind School, student engagement in drama has often involved adaptations of many of the classics, including works by Shakespeare (Macbeth, Hamlet, The Tempest), Charles Dickens (Oliver Twist, David Copperfield, Great Expectations, Ebenezer Scrooge), Mark Twain (Prince and the Pauper), Robert Louis Stevenson (Treasure Island), and many others.

Classics are classics because they are great stories with timeless themes that pull the actors and the public in on an emotional level. They can be delivered on the same level of a child's story like «The three little pigs». Why do the three little pigs with a young adult when it is possible to do «Macbeth» or «Great Expectations»? There is no reason that a young person who has JNCL should be doing fairy stories for younger children (although there is nothing wrong with fairy stories in the right context). The key is to find ways to make appropriate material accessible, engaging, fun and educational. And the classics can be adapted for a whole range of needs and abilities (Grove, 2012; Grove & Park, 1996).

For example, «Macbeth» is a story that starts with two friends, Macbeth and King Duncan. There is a war and Macbeth is loyal to Duncan. Duncan rewards him with a new castle and a ceremony. All is well and the relationship has been set – perhaps over three or four drama sessions. Then Lady Macbeth decides they should do something nasty to King Duncan, something that Duncan does not deserve. This has real emotional impact because the story is built on the friendship of Macbeth and Duncan; the students care about them and they are appalled at the injustice. It really gets the students going, they get motivated to speak out and be involved in the drama! And then when Macbeth doesn't get caught but the guards do! The students jump up and down – they are hooked on the story. They can learn all sorts of things. About relationships, about expressing themselves, about emotions, about things not going the way they should. But being involved in the drama, in pretend, means there is a distance, it is not their real life. They can be engaged in a deeply emotional way, but it is not necessarily their emotion. When the drama closes, they come back to themselves and they can discuss how they felt.

Of course, in real life, they will never be involved in a brutal murder with daggers – that is the joy of the story. But they will engage in those related emotions. They can experiment with them and play with them. This is valuable and especially for young persons with JNCL who may be increasingly struggling to manage their own emotions.

Some strategies used in drama class

Hot seat

First of all, a good introduction to drama is the use of the «hot seat». The hot seat is a chair or an area, and once you sit there, you become a character and need to stay in character. This way, students learn about physically going into character and have a very clear cue to start the drama. It is important to remember that for young people with JNCL, there is no eye contact or gesture for beginning the drama as one would with sighted students. This physical change of position works.

The hot seat area can be adapted to any situation and the use of additional props can help the young person get into character. For example, if the person in the hot seat was supposed to be a giant, a ladder could be set up in the hot seat area to give the impression of height and the voice would come from above! Another hot seat might be covered in fur to make it the king's throne or they might just sit on the floor with an old, tattered, smelly coat if they were supposed to be a sad, old homeless woman. The important thing is the physical movement from one area to another, in effect, stepping physically into role and touching the material objects that are related to the role. Remember that young people with JNCL will not be able to recognize standard cues for readiness or acceptance of role play by eye contact, body language or gestures. The physical change to the hot seat acts as the cue instead.

Teacher in role

The teacher can go into role as a character and interact with the class. This way, the teacher can change the direction of the drama, challenge thinking and manage the group within the drama. This keeps true to the drama, without having to be a «teacher» giving instructions.

And depending on the class, the teacher in role can really explore issues and emotions in a grittier way. As long as the students are secure in the convention, they can really enjoy having an encounter with Bill Sykes (from «Oliver Twist») who is threatening, violent and has a foul temper, even if this could induce tears in other circumstances! They can experiment with emotions and empathy and explore how characters react to different situations. Real life issues like bullying or being isolated in society can also be addressed. They can test this out in a completely safe environment.

This leads on to improvisation. Through the «Teacher in Role» exercise, the young people will have been given ideas about appropriate vocabulary and ideas and this can help the young persons have the confidence to take forward the improvisation and use it themselves. In the context of a story, young people with JNCL can be supported to experiment with what it is like to yell and shout and stamp the feet and identify when these emotions are appropriate and how people react. They can explore what is it like to be overcome with anger. What does it feel like to be threatened, frightened, in a situation where something is not right, for example Bill Sykes? How does it feel when someone comes close into your face, invades your space and speaks in a threatening manner? This is pretend, it is fun, it also touches the emotions and it is hugely valuable learning. This can act as a major stress release for a student – especially if they do not really understand their symptoms and cannot articulate how they are feeling.

Freeze frame and thought tap

Drama can focus on emotions, understanding of own and the emotions of others. What do emotions feel like? How do they look? How do other people perceive you? This may help students with JNCL to understand and make connections. Again, the classics are a great tool for exploring these themes. For example, in working on «The Tempest,» a «freeze frame» of facial emotions was created to explore the different characters. Using the cue of a tap on their shoulder, the young person has to speak or vocalize in character. If it is an angry freeze, for example they might say, with conviction «*I hate him!*» or just an angry noise. This may connect the group, give them ideas and reassure them that they are all doing the same work.

In this connection, scripts are sections that are good to work with. In «Macbeth» the script for the Witches scene is particularly engaging: «*When shall we three meet again in thunder lightening or in rain...*» It does not seem to matter whether the students understand the intricacies of the language. The words are poetic and flow, and it is nice to say them. This scene also allows one to build a great atmosphere in the room, using sound effects, music, voice, candlelight as the witches are telling Macbeth he will be King.

Young people with JNCL do not need to have a physical script if this is too much, they can work from a CD or use the support of an assistant prompting lines quietly.

Using materials

The use of tactile material and physical objects within the drama structure is very important to make it a more sensory experience. Connection with a physical symbolic object helps the young person with JNCL relate to a character (e.g., the use of a fur covered chair for a king's throne). It is important to use real objects and props that have meaning and are authentic. Why use a plastic apple when a real one feels and smells so totally different? «Real» props link to real experiences.

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A creative approach to physical movement can also build bridges for communication within the drama. A small working area in some scenarios can give the young person a real experience of being restricted and having no freedom. For example, they can really crawl into a cramped corner to denote a cave or be under thick blankets for an air raid shelter. A total freedom of movement in other situations like being outside in a grassy ditch for a scene on a battlefield gives a very different experience and generates other responses. The use of space is hugely important to help develop and maintain the young persons' spatial awareness and these experiences help them want to keep exploring.

It is also useful to record voices or film in class. This can re-create and remind the young person of the actual physical experiences in which they participated. In a way it is similar to using sound like a photograph to open up a rich set of memories associated with being in that drama. This can be important for them, and also can be a means of assessment and self-evaluation for the young person.

Conclusions

Exploring real and imaginary situations may help young people with JNCL to understand and share their world. If they can get emotionally involved and care about the outcome, they may be more creative and strive to be actively involved in the story, independently or interdependently (see Chapter 16). But the advantage of drama is that the structures enable there to be enough distance to allow the students to be engaged in the feeling without being swept up with the emotion. They can empathize with the characters without getting upset themselves. That is the power of drama!

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Communication and Language in Education and Intervention for Children and Adults with Juvenile Neuronal Ceroid Lipofuscinosis

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Young people with juvenile neuronal ceroid lipofuscinosis (JNCL) will gradually experience a change from speaking effortlessly and fluently to finding speaking to be tiresome and even frustrating (see Chapter 6). It is not possible to stop or prevent the disease from developing. Speech problems eventually will have a major impact on the young person's social life and life situation in general. The promotion of communication and language is therefore always an important aim in the curriculum of students with JNCL. Interventions may contribute to strengthen and maintain language skills, prolong the period with fluent and intelligible speech, and compensate for loss of spoken language. There is a need for developing strategies to prepare for and compensate for the speech decline but there are few studies on language and communication intervention for this group.

The present chapter discusses interventions for maintaining intelligible speech as long as possible and the use of augmentative and alternative communication (AAC) to supplement and compensate for the impairment of speech.

Interventions to strengthen and maintain intelligible speech

The aim of speech and language therapy is to strengthen the child's speech and language skills, and may include preventive measures, diagnosis, education, training, and counseling. Specialist services with speech and language therapy exist in all the countries participating in the project. Interventions related to fluency, and to strengthening and maintaining the functions of breath, voice and articulation, are relevant for students with JNCL, as are interventions related to communication strategies.

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Interventions will not prevent symptoms from developing but clinical experience indicates that decline can, to some extent, be postponed. Gayton (1982) used rhythm strategies to improve speech fluency and intelligibility in two students with JNCL, such as accompanying the speech with clapping the hands or talking in time with a metronome. However, the cognitive problems made it difficult for the students to follow the strategies and they were therefore abandoned. Experiences from Norway suggest that strategies used in stuttering therapy might lead to temporary improvement in fluency and maintenance of speech intelligibility (von Tetzchner, 1993). Structured and systematic training may establish a foundation to meet the upcoming symptoms, and speech and language therapists may give useful advice to parents and schools about training strategies.

In the present survey (see Appendix A) parents were asked about speech and language therapy. Forty-three parents answered that their child had received speech and language therapy (see Figure 13.1), and that speech therapy was first provided at an average age of 9.9 years (SD 5.6). The survey did not ask for details about the speech and language therapy.

The parents were also asked to evaluate the effect of the therapy on a scale from 1 (no impact) to 5 (very high impact). The average evaluation score was 3.2 (SD 1.2, range 1–5). Eighteen respondents (41.9%) answered that the therapy had a high or very high impact, and 11 respondents (25.6%) answered that the effect was moderate. Fourteen parents (32.6%) found little or no effect of the speech and language therapy (Figure 13.2). These findings suggest that a considerable part of the respondents found speech and language therapy useful for their child, and



Figure 13.1 Number of individuals with JNCL who had received speech and language therapy or other communication training



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Figure 13.2 Impact of speech and language therapy (percent of answers) (N=43)

that speech and language therapy for some children and young people with JNCL may be useful for a shorter or longer period.

Only 11 participants had received communication training other than speech and language therapy (see Figure 13.1), and this was provided for the first time at an average age of 12 years (SD 6.1). Five respondents answered that the training had a high or very high impact, and two that the impact was moderate, while four found little or no impact of the training. The number was small and the evaluation varied but seemed to indicate that communication training other than speech and language therapy may be useful for some children and young people with JNCL.

Augmentative and alternative communication (AAC)

Augmentative and alternative communication (AAC) is an umbrella term that includes diverse communication modes other than speech for individuals with impairments of language and communication, including manual signs, graphic symbols and tactile communication systems. During the last decades, AAC has become widely used for children and adults with severe speech or language disorders. AAC can be an alternative or a supplement to speech. The function of AAC is to support communication and language skills in general and should preferably be available throughout the day and in all situations. Today, children and adults with different diagnoses use AAC (von Tetzchner & Martinsen, 2000).

Unaided and aided communication

AAC can be divided into two broad groups: unaided and aided communication. Unaided forms of communication are *produced* by the user. There is no need for any device but unaided forms of communication require some degree of motor ability. They may include vocalizations, facial expressions, gestures, manual sign systems and sign language. Communication partners must be able to interpret and understand the communicative expressions, and the message is often produced as a collaboration between the AAC user and the communication partner (Clarke & Wilkinson, 2009; Hörmeyer & Renner, 2013; Solomon-Rice & Soto, 2011).

Aided forms of communication are *selected* by the user. They imply some kind of device, such as a folder, a board, a book, or an electronic device with synthetic speech output, usually with graphic symbols or letters and words (von Tetzchner & Martinsen, 2000). Aided communication thus depends on the availability of a communication aid, and availability may be limited in many situations (von Tetzchner et al., 2018). Because most communication aids contain pictures and graphic symbols, they usually require some degree of vision but it is possible to adapt them to a tactile recognition or auditory scanning. For children with JNCL, the selection of expressions should be as simple as possible, so the expressions can be used for communication also in later phases of the disease with dementia.

Objects may function as *tangible* or *tactile* symbols (and visual for children with residual vision), often called "objects of reference" (objects used in a symbolic manner to refer to something). It is important that it is not the function of the object itself that determines how it is used but the category it represents, of people, animals, things, activities, and so on (McLarty, 1997; Park, 1995, 1997). For some they are a step on the way towards more advanced symbol learning and literacy. Objects of reference may be an alternative to graphic symbols. Braille can also be used in aided communication (Park, 1997; Rowland & Schweigert, 2000).

Both aided and unaided communication can function as alternatives to speech or to augment speech. Families and professionals need to work closely together when choosing an AAC mode. If possible, an interdisciplinary team should be involved in the decision-making process, as well as in planning, implementation and evaluation of the intervention. The communication form or forms that are chosen should be based on an assessment of the individual's strengths and weaknesses. To make the environment communicatively accessible, the communication partners, too, need training (von Tetzchner & Stadskleiv, 2016). If a child or young person with JNCL learns manual signs because speech is slurred and difficult to understand, the family, staff and peers must be taught the same sign vocabulary. Different forms of AAC can be combined, both with each other and of course with speech.

User groups

Various forms of AAC have been developed for persons with limited spoken language and are used with children and adults with a variety of diagnoses, in particular with motor disorders, intellectual disabilities, severe language disorders and autism spectrum disorders. Users may have age-appropriate comprehension and problems with expression only, or problems with both receptive and expressive language. Some children with severe and multiple disabilities need AAC to develop any communication skills. There are three main groups of AAC users (von Tetzchner & Martinsen, 2000).

- 1) The expressive group is characterized by a significant gap between comprehension and production of spoken language. Many in this group have motor impairments but also children with Down syndrome may belong to this group.
- 2) The language support group may need AAC temporarily to support the development of spoken language, or as a lifelong supplement to speech.
- The alternative language group includes children and adults with little or no comprehension and production of spoken language. AAC becomes the main communication form for both them and their communication partners.

People who use AAC differ considerably in language comprehension and this has implications for the intervention. «For persons with good comprehension, developing conversation skills is the most important educational goal. Lack of language skills is not the cause of their limitations (...), but lack of capability to use what they understand» (von Tetzchner & Martinsen, 2000, p. 277).

When AAC is part of the intervention, communication partners must know the person's repertoire of communication expressions and strategies. AAC may affect communication fluency by slowing down the communication pace, and it is important to give the child or adult enough time to participate in the dialogue. Manual signs, objects of reference or other aided expressions may be used in unusual and idiosyncratic ways (see also Chapter 6). In an inclusive environment, it is essential that all potential communication partners, adults and peers, become familiar with the special elements in conversations involving AAC. Otherwise, the AAC user may become restricted to communicating only with a few persons, mainly adults. Measures should thus be taken to ensure communication with all relevant persons in the environment, including peers.

AAC and JNCL

In early childhood, children with JNCL have typical language development but as the disease progresses, many gradually become difficult to understand even for those who know them well. In the later phases of the disease, comprehension also becomes more affected by the cognitive problems (see Chapter 6). The main reason for offering AAC to children and young people with JNCL is their increasing gap between comprehension and production, even if comprehension also tends to decline.

AAC for children with severe visual impairment must be adapted to compensate for the vision impairment. AAC may include tactile symbols, gestures, manual signs, object of reference, communication boards and books, and electronic devices. Most communication aids are designed for sighted children or adults, and have graphic symbols (e.g., PCS, Pictograms, Blissymbols), photographs or drawings, but it is possible to equip electronic communication aids with tactile symbols or a braille keyboard and synthetic speech (Goldware & Silver, 1998; Rowland & Schweigert, 2000). Information about AAC for children who are visually impaired is available at resource centers or special schools for students with visual impairment. In Norway, *Tactile symbols and plans* provide information about tactile adaptations of communication aids (Grini & Aasen, 2013). *Tangible symbol systems primer* (Rowland & Schweigert, undated) is available for free download.

A literature search revealed almost no descriptions of the AAC for children or adults with JNCL, the case history below is an exception.

A case history

A 17-year-old girl was provided with an electronic communication aid with speech output when her speech was beginning to become unintelligible (von Tetzchner, 1992). When the communication aid was introduced, she had good comprehension of spoken language and a large vocabulary. Her speech was intelligible to her family, friends and teachers, and many of her utterances were comprehensible for people who did not know her as well. At that time, she only occasionally experienced communication breakdowns and misunderstandings, but because the ability to learn was declining, it was necessary to start then, before she really needed the aid. The overall intervention goal was to help the girl maintain social interaction and conversations for as long as possible. This goal was the basis of all intervention strategies.

Hello	l want something	Hungry	I	Person	Help	Finished	Goodbye
l want to say something	Try to guess	Thirsty	Before	After	Music	Radio	When
I must talk this way	More	Tired	In	Out	Read	Weave	Stupid
Yes	Almost yes	Again	Wrong	Don't know	Wait	Almost no	No

Figure 13.3 The girl's first vocabulary

Aid and vocabulary

Because the girl was blind, she was provided with an electronic aid with speech output. When she pressed a square on the overlay, the device produced the word. At that time, Norwegian synthetic speech was not well developed, so digitized speech was used. The vocabulary was recorded using the speech of a young girl of the same age and using the same dialect. In Norway, technical aids are provided free by the State, and the communication aid was borrowed from the local technical aid center. The girl had a large vocabulary, which she would not be able to express through use of an aid with a limited vocabulary, because she was unable to learn navigation to select a large number of lexical items. Therefore, the intervention strategy was not to give her a means for expressing all her needs, interests and ideas, but to use the communication aid to guide the conversational partner to infer what she wanted to say. Some of the words were chosen to gain attention, some to define conversational domains, and some to direct the inferences of the partner. To help her navigate, some of the words were marked tactually on the communication aid.

The initial vocabulary is shown in Figure 13.3. Because of her good language skills, it was believed that the girl could learn 32 words and phrases, and possibly more later. However, even a vocabulary of 32 items proved too difficult. It was difficult for her to find what she wanted to say and in collaboration with the girl, the vocabulary was reduced to nine items by omitting words and combining topics into one utterance.

The selection of vocabulary was an unhurried process where she participated actively together with parents and teachers. She was a member of the project team, and it was an important motivation for her that the project may be of help to others with the same disease as herself.

Use of the aid

When the girl got the communication aid, she did not need it to make herself understood in conversations, but it still had a positive impact on both her communication skills and her life situation. She immediately took a liking to it, named it "Talking Lady", and demonstrated it willingly to family members, friends and helpers. She had great expectations with regard to what it could do for her.

The girl experienced some communication difficulties before she got Talking Lady. Sometimes she was misunderstood or not understood at all, and the conversation broke down. However, conversation failure was too sensitive for her to discuss and she refused to talk about it. Talking Lady represented a means for her to manage her difficulties and made it possible for her to talk about her problems, and to discuss how to cope with them. This influenced the interactions positively and improved the communication without the girl actually using the device for communication. The inability to talk about the communication problems had been a significant stress on the family, and by catalyzing openness and communication, the device led to a better family situation. According to her family, the positive effects had already given more benefits than the efforts invested.

With time the girl's speech became more difficult to understand. Sometimes even her mother was not sure whether she said yes or no. Such situations were resolved by the mother asking more yes-no questions, and the Talking Lady was never really used for communication. But it remained a psychological support.

AAC use among children and adults with JNCL

In addition to speech and visual problems, support of expressive language needs to take into account decline in motor and cognitive functioning. Different symptoms become noticeable at different ages, and there is considerable individual variation (see Figure 1.1 in Chapter 1). The fact that speech problems often become noticeable before other motor problems may imply that communicating by using hands and arms might be maintained longer than speech.

When the parents in the present project were asked about AAC or special communication strategies, eleven parents mentioned that they and the child used some kind of alternative means in communication. One family mentioned that their daughter with JNCL used to squeeze their hand to say "yes" and "no", another child clapped the hands to express herself, although it was not always clear to the family what this meant. A third young man would nod and smile or frown to indicate "yes" and "no". A young woman coughed when she needed to go to the toilet. Some families used assisted auditory scanning, they listed several items and waited for a yes or no. A few families had tried electronic devices with synthetic speech output, but the results were rarely positive.

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They tried some communication programs on the iPad, but they were all visual and she was blind, so they were fun for the teachers but not useful for my daughter.

However, both awareness and use of AAC and support with alternative means seem to be increasing. When the parents were asked about AAC intervention, twelve parents mentioned using specific gestures or manual signs, six mentioned objects of reference, eight mentioned electronic devices (including switch-operated devices), and eleven parents mentioned "other strategies". Most of the participants were still in the process of learning, and for some the training had been abandoned. Some mentioned that AAC might be introduced as a precautionary measure. One parent commented:

Alternative communication methods should be provided for youths with JNCL (and children) earlier (age 10–15), as they still have a good opportunity to learn and adapt to new things.

In summary, the results of the present survey indicate that most young people with JNCL develop severe problems with expressive speech. Only a small number of children, adolescents and adults with JNCL have been given opportunity to learn alternative communication strategies, but such strategies seemed to become more common in this group. There is also a greater focus on trying out these strategies, like the manual sign project below. AAC training can be provided by speech and language therapists or by other professionals.

Manual signs

Manual signs and gestures may be useful when the ability to speak declines. Manual signs are rarely used in interventions for people who are blind, partly because of their inability to learn through visual observation. However, teaching methods can be adapted to compensate for the missing visual sense, for example using signs in the tactile modality (Downing & Eichinger, 1990). Manual signs may be taught to students with JNCL (and the other students in their class), with the aim of establishing a communicative competence that may be useful later in life if speech becomes unintelligible.

Manual signing as AAC does not mean a national sign language with full sentences and correct grammar, but rather signs from a manual sign system like Signing Exact English, Signalong, Irish Láhm or Signed Norwegian, representing familiar objects, persons, activities or events. This usage of signs is called key-word signing or sign supported speech (Budiyanto, Sheehy, & Khofidotur, 2018; Glacken et al., 2018; Grove & Woll, 2017). In many countries, there are websites with sign dictionaries, which include videos of signs. These websites are useful for parents and teachers when they are choosing signs. In addition, there are many apps available for mobile phones or tablets that can provide the sign for a word immediately (in various sign languages or manual sign systems). Most people who provide support for individuals with JNCL are not familiar with manual signs. Having an app handy can help them see how the sign is produced (via a video) so they can then instruct the child. Pictures of signs in books can be hard to follow, and books are not always available.

As the disease progresses, language ability gradually declines, expressive speech more than comprehension (see Chapter 6). The communication partners will speak when communicating with the young person with JNCL. They do not need to use signs while speaking, because the child with JNCL will not be able to see the signs. However, successful communication will depend on their understanding of the child's signs. Signs may not only be learned in special sessions – the partners may show the student with JNCL new signs or help him or her articulate known or new signs in natural situations.

Alternative methods with signing on the body include *Tactile Signing for Sensory Learners* (Tassels) (Woodall & Charnock, 2017) or *Canaan Barrie on-body signs,* developed by Mary Lee and Lindi MacVillians at the Royal Blind School in Edinburgh (von Eichwald, 2015). Manual signs articulated on the body may be easier to learn for some students, because they are designed for people with a visual impairment and additional learning needs.

Manual signs may be easier to articulate than speech. The individual uses the hands and body and does not depend upon the availability of a communication aid. As motor problems in hands and arms appear relatively late in most persons with JNCL (see Chapter 7), manual signs could prolong the period of successful communication. Manual signs may have the same flexibility and diversity as spoken languages. It is possible to create idiosyncratic "home signs" when needed and the signs can be adapted to the motor skills of the person (Grove, 1990; Rudd, Grove & Pring, 2007). Signs can both augment and substitute for spoken language and can be combined with communication aids or other forms of AAC.

During recent years, projects on manual signs have been conducted in the Nordic countries for a small number of children with JNCL. The following is a short description of a Norwegian project with four participants with JNCL.

A project on manual signs

The aim of the project was to investigate if students with JNCL could learn manual signs and use them spontaneously for communication. The project was organized by Statped (the national Norwegian agency for special education support) and took place from September to December 2014. The project started with a two-day course for families and staff to enable the schools and families to start teaching manual signs to the students. The course included information about communication problems for individuals with JNCL, use of AAC and how to teach signs to students who are not able to imitate by using vision, and where to find sign dictionaries on the Internet. The families and staff chose a sign repertoire for each student and decided what signs should be taught first.

Methods

Four students, aged 8, 14, 17 and 19 years, participated in the project (the names below are not their real names). Information about sign use was collected with the help of three registration forms that had been reviewed during the course, and a short interview with staff and/or families. The forms were completed by parents and/or staff daily. Educational counselors from Statped visited the participants during the project period. They were also available for families and staff on email and telephone during the three-month project period.

Results

At the closing time of the project, information from the registration and interviews indicated that the students had learned to perform a varied number of signs and could produce the signs when asked to do so. This was a step in the learning process towards communication. The students had also used signs on their own initiative a few times to ask for or talk about something. This suggests an emergent understanding of signing as communication. One student, Arne, initiated communication by using signs several times. During a visit from the resource center, another student, Christer, spontaneously signed DRINK to tell that he wanted something to drink.

Table 13.1 shows that the results varied a lot between the children. Arne was the youngest and learned a substantial number of manual signs during the project period. He and his family attended a one-week seminar, where they received guidance from a resource center for sign language. This was a major support for Arne and his family, and the center gave much more input and assistance than the other families and staff had access to.

Since the project period ended, Arne has continued learning signs and is signing quite a lot. He signs fluently as part of his communication along with speech. He finds it helpful, especially after he experienced episodes with severe

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Table 13.1 Manual signs taught to the participants in the Norwegian sign project

Arne 8 years	VOLCANO, STAR WARS, SPACESHIP, GALAXY, PLANET, PIRATE, SWORD, PISTOL, LIGHT SABER, WALKIE TALKIE, HANDCUFFS, POLICE, THIEF, PRISON, SPY, ZOO, TORCH, REFLECTIVE VEST, BIKE HELMET, LEGO, DUPLO, HEADPHONES, AUDIO BOOK, DAISY PLAYER, RAT, SVEIN (NAME), SEWAGE, JAMES BOND, SKI, PLAY CARDS, PLAY, RAIN, SNOW, ICE, LAVA, MONDAY, TUESDAY, WEDNESDAY, THURSDAY, FRIDAY, SATURDAY, SUNDAY, PLATE, KNIFE, FORK, SPOON, BREAD, BUTTER, SUGAR, LIVER PASTE, JAM, GRAPES, ORANGE, APPLE, BANANA, CUCUMBER, HAM, TEA, COFFEE, EGG, MILK, COCO, CHOCOLATE, JUICE, SODA, STRAWBERRY JAM, BLUEBERRY JAM, RASPBERRY JAM, CHEESE, BROWN CHEESE, WHITE CHEESE, RED, ORANGE, WHITE, YELLOW, BLUE, PURPLE, PINK, GREEN, BLACK, BROWN, DARK BROWN ETC, LIGHT BROWN ETC, HOUSE, STONE, SUN, TREE, MOUNTAIN, HEAVEN, RIVER, TUNNEL, GRAFFITI, WINDOW, DOOR, HEART, TOMBSTONE, CHURCH, HELLO, GOODBYE, THANKS FOR FOOD, HUNGRY, THIRSTY, GLASS, CUP, DRINK, TEASPOON, GOOD NIGHT, GOOD MORNING, BOOK, WATER JUG, COFFEE POT, CAT, DOG, TIGER, LION, SNAKE, COW, RIDE, SHEEP, GOAT, TROUSERS, SWEATER, JACKET, SHOES, SOCKS, HAT, FEATHER, GLASSES, TOGETHER WITH SISTER/BROTHER, MAYBE, TODAY, YESTERDAY (PAST), TOMORROW (FUTURE), MOTHER, FATHER, CHILD, TEACHER, TO TEACH, TO LEARN, FLOUR, ICING SUGAR, GLAZE, CAKE, MELT, RUBBER SPATULA, PLASTIC, SHEET, WINDOW, ONE, TWO, THREE, FOUR, FIVE, SIX, SEVEN, EIGHT, NINE, TEN, ELEVEN – TWENTY, HUNDRED, THOUSAND, MILLION, YATZY, CLOWN, EYE , NOSE, MOUTH, TOMATO, SALT, KETCHUP, THE WHOLE ALPHABET, GARDERMOEN, BERGEN, TROMSØ, OSLO, FILM, CD.
Bente 14 years	HELLO, GOOD MORNING, YES, NO, MUM, DAD, MUSIC, GOODBYE, PAIN, COFFEE, HOME, SUN, SIBLING, TEACHER.
Christer 17 years	FOOD, DRINK, YES, NO
David 19 years	YES, NO, QUIET, WAIT, WHAT, STOP.

stuttering that made communication difficult for him. Another student stopped learning signs shortly after the project period ended, because of declines in memory and fine motor skills. One student had learned a few signs before the project started and expanded his sign repertoire with three signs during the project period. For one of the students there is no information about signing after the project ended.

Factors related to signing

The reasoning behind introducing manual signs for children and young people with JNCL was the possibility that signing might reduce the communication problems related to stuttering, articulation problems and general speech decline (see Chapter 6). Articulating manual signs is less complex than articulating words,

and this may contribute to maintaining motor skills related to signing longer than the motor movements of speech. This was demonstrated by Arne.

It is conceivable that signing may reduce the word finding problems typical of dementia, but it is not obvious that it is easier to recall a manual sign than a spoken word. In addition, memory for newly learned manual signs might not be as robust as memory for spoken words acquired much earlier in development. The parents observed that Arne used signs when he had difficulties remembering the words. This might indicate that manual signs somehow is easier to recall or that he has two ways of expressing something and can choose the other way when the first does not work. Having alternative modes may thus facilitate communication through cross-modal stimulation (von Tetzchner & Martinsen, 2000). These are important issues to investigate further.

Factors of importance for AAC intervention

Many factors may determine whether AAC intervention will lead to positive learning results, including administrative and educational practices, and political and ideological attitudes and regulations.

Daily learning and practicing opportunities

Learning depends on practice, and AAC should be used during the whole day. Learning will be delayed if AAC is only used with the teacher who is responsible for the training. After intervention has started, the use of AAC should be encouraged in different situations and with different communication partners, within and outside educational settings. Some forms of AAC, for example manual signing or using a technical communication device, will require some formal teaching. A brief formal teaching session should be scheduled for every day, preferably early in the teaching program. Such daily repetition may be necessary to ensure progress and maintenance. Communication partners should have the training necessary to be able to give help and guidance when needed. Opportunities for communication should always be emphasized and periods when acquired communication skills cannot be understood should be prevented.

To ensure that AAC becomes a natural and functional part of the child's communication, the child should take part in a variety of situations where signing or other forms of AAC can be used in a natural way. Such situations can be at school with teachers and peers, or at home with family. Preferably, the young person with JNCL should be able to use AAC in all common situations. Because young people with JNCL often use short utterances (speech and AAC) in everyday

situations and conversations, communication partners must be able to recognize the person's repertoire of manual signs or other AAC modes, know how these are used by the person, and infer their potential meaning in the context. This applies to family members, staff and peers.

Barriers in the learning environment

The pathological processes involved in JNCL gradually reduce the ability to learn. However, these processes are not the only factors that may influence learning negatively. There can also be barriers in the learning environment. Schools might be reluctant to implement new educational strategies, especially when these strategies deviate from the usual guidelines of the curriculum, such as teaching manual signs to children who are blind. Schools can also focus too much on their own lack of competence. AAC may be seen as a difficult domain for teachers if the school has no prior experience with AAC. This can create resistance from staff and prevent the search for possibilities and competence. Sometimes there are disagreements concerning responsibility, whether an intervention is a matter for the educational or health authorities, or a specialized service outside the school (for a thorough discussion, see Goldbart & Marshall, 2004).

Other barriers concern structures and systems, for example related to resource allocation, time needed for staff to attend courses or get external guidance. Lack of communication structures for information transfer within the school or institution and rigid staff schedules can be barriers. In one of the schools participating in the Norwegian manual sign project, only one assistant was involved in the intervention. The assistant was very competent, but her schedule only allowed signing sessions twice a week. This was probably too little for an optimal learning process.

It is important to identify and overcome barriers in the learning environment. Teaching AAC is not really very complex or time consuming. Solutions should be sought to secure the student's learning and development as fast as possible. Students with JNCL should not have to wait for clarifications while the disease continues progressing. It is usually possible to find solutions if there is willingness to change and discussions are unrestricted and creative. Support from specialist services can be an important factor to help getting started.

Learning for the future

For young children with JNCL, the need for AAC will usually lie many years ahead. At the same time, it is important to start teaching early in order to utilize the child's best learning capacity. Resource centers are sometimes asked about the motivation behind AAC intervention when the child or adolescent still speaks well. Enhanced and precautionary learning are basic principles when preparing AAC intervention for students with JNCL (see Chapter 12). All students engage in learning for the future. The knowledge and skills acquired by children and adolescents constitute a foundation for a successful life in society. To ensure learning, the goals and teaching methods applied are adapted to the students' age and capacities. Students are not usually aware of why they should learn the specific skills and knowledge presented in the classroom, but they learn anyway. Learning for the future is a basis for all students.

Inclusion

The principle of inclusion is important in all the countries participating in the present project. It is an overall aim included in the legislation of many countries that education should take place in a community of peers. Learning in a social setting may in itself be a strong motivator (see Chapter 12). At the same time, students have different needs and abilities. The learning environment must be adapted to give students learning opportunities, irrespective of their abilities and challenges. This implies that the objective and methods will vary among students with special needs, but ideally, this should not compromise their social affiliation.

Communication is important for participation, and declining communication skills can make inclusion difficult. This is the motivation for implementing measures that may strengthen the communication skills of students with JNCL. These measures are likely to make demands on both the child with JNCL, his or her peers and the school. When a student with JNCL is learning AAC, the class or group also need to learn AAC to maintain his or her participation and communication with the peer community. This is illustrated in the story below.

A young boy with JNCL, "John", was communicating with a combination of speech and manual signs. His signing was fluent and functional, and it was obvious that signing was very beneficial for his communication. It was however, not always easy for communication partners to understand what John wanted to say. He attended a school where all the students in his group had learned key-word signing. The boy's teachers had a saying that illustrated their view on the learning environment of his group: "A life necessity for John is also good for the other students and completely harmless for everybody".

The teachers' saying is a good and simple guideline for inclusion.

Selecting AAC vocabulary

When choosing AAC vocabulary the individual's preferences, interests and needs must be in focus. The words that are selected should be meaningful and useful for him. Individual preferences include words for special interests and favorites, activities he often wants to do, and activities he often wants to talk about.

Talking about emotional events is important. The vocabulary should enable the person to communicate about current and earlier events, as well as other important life events, and express emotions like happiness, pride, grief and fear. For some individuals, emotional events can be preferred and recurrent topics of conversation. Vocabulary should also include activities that happen routinely, daily or weekly. The person may for example want to ask whether he is going to do a particular activity, when it will happen or who will be joining him.

Play and games

For children in primary school, play and games often give extra motivation for activities and schoolwork. A board game developed in Norway, «Eltho Tactile», consists of a board, dice and activity cards. The teacher decides what tasks should be asked for on the activity cards, for example «Show the sign for Star Wars», «Sign the names of your siblings» or «Show the sign for your favorite activity». The tasks can be written in braille if the student has the necessary reading skills, or in print for the staff to read aloud. The potential for individual adaptations is considerable.



Figure 13.4 Cards with braille and print used for teaching manual signs

In the Norwegian sign project, Arne's family developed several kinds of material for teaching and maintaining signs. The parents made cards with words written in braille and print (see Figure 13.4). The cards had holes and were strung on a rope, and the rope was hanging across the room. When Arne followed the rope, every now and then there would be a card showing a word and his task was to sign the corresponding sign. The parents would count the number of correct signs: «You remembered 16 signs out of 20!» or it could also be turned into a competition: «You remembered 16 signs and I only remembered 14!» Cards that were mastered were marked with a tactile sticker to help both Arne and the family to keep track of the signs he had learned.

These kinds of activities can be used together with others, for example family, friends, teachers or classmates (see also play and games in Chapter 21).

AAC and transitions

Transitions often represent challenges for inclusion and participation. Children and young people with JNCL may attend a new class and meet new classmates. Staff also changes. Young adults go through the transition from being a student to attending a sheltered workshop or a day center (see Chapter 23). For many there is also a transition from living with the parents to moving into a residential home with services and support from staff. In adulthood, speech is often severely affected by the disease and difficult to understand for peers and staff who are not familiar with the person's communication and history. When preparing a major transition, the individual's needs and wishes should be the foundation of the planning process. New communication partners will need information about the person's history, interest and preferences, and about present communication skills, including AAC. There is a considerable risk that an individual with JNCL may be underestimated if the new staff are not aware of the gap between the person's comprehension and production of speech, and his sometimes idiosyncratic way of using words and alternative expressions.

Some useful tools to promote successful communication

As communication becomes more difficult for the individual with JNCL, several tools may be useful. Some of the tools have technical equivalents, for example computer programs or electronic communication devices (see Chapter 19).

Partner-assisted auditory scanning

This technique utilizes comprehension in supporting expressive language and may be useful in supporting communication in persons who have severe motor and speech impairments (Clarke & Price, 2012). The technique supports memory by presenting some alternative responses when the person with JNCL has difficulties finding the words or the expressive means are very limited. It may be useful for persons with severe visual impairment because it relies on the auditory sense.

In partner-assisted scanning, the communication partner presents a sequence of utterances, one of which the person might want to say (see procedure in Table 13.2). The utterances might be «I want to talk about something», «I want something to eat», «I want something to drink», «I want to do something», or «I want to tell you how I feel». All the categories are presented and then repeated one by one with pauses to wait for a possible response from the person. When the person has chosen a category, a sequence of alternatives within the category may be presented in the same way – first the whole sequence and then the alternatives one by one with pauses. The number of utterances presented can be adapted to the cognitive level of the person.

Comments from several parents in the present project indicated that they had used partner-assisted scanning with their child when speech was becoming difficult to understand, and some had developed homemade systems based on the same principle.

Tabl	e 13.2	Routine	for	partner-assisted	scanning
					()

• Ensure that the person is well-positioned and comfortable.
• Get the student's attention by saying his/her name.
• Tell the student what you are going to do: "I'm going to give you (3, 4 or 5) choices. First listen carefully to all the choices".
• List the choices slowly and clearly, for example: "musicbookvideosomething else".
• Make sure you still have the person's attention.
• Say: "[Name], tell me when you hear the one you want".
• List the choices in the same order as before, pausing slightly between each of them.
Watch and wait for the person to give a response.
• Repeat the person's response, for example: "You said".
 If the response is unclear, tell the person you are having difficulty reading the response and repeat the sequence again.

I decided that the best strategy was auditory scanning, where I would say individual items in a list and wait for her to respond yes or no.

When it became difficult for him to explain/remember what he wanted to say, we parents developed a 'Question system' to define the subject and afterwards continue questioning about that specific subject.

I was responsible for training her in auditory scanning.

We use a question technique to find out what she wants to talk about, because otherwise we cannot understand what she says.

For persons with JNCL, the only prerequisite for using partner-assisted scanning is the ability to express "yes" and "no". This can be spoken words or vocalizations, gestures, signs or body posture. If it has become difficult for the person to indicate "yes" and "no" the communication partners need to be observant and sensitive. It may be a very subtle movement indicating a positive or negative response, as described in these examples.

He would nod his head or give us a big smile when we gave him the desired words or requests or he would frown or tighten his brow if we were off the mark. Later we would give him only two choices at a time so he could use "yes" or "no" to make his desired choices.

Pressing the hand once: "yes", pressing the hand twice: "no".

To be sure all communication partners know how a person expresses "yes" and "no" when the expressions are atypical or idiosyncratic, it is practical to have a photo or written description of these expressions. Partners may also benefit from pre-made category sets made by professionals, that can guide strategic use of the «20 questions» technique. Partners sometimes panic in the moment, if the child needs to communicate urgently, and give the child random options. When given a predetermined set of categories to present to the child, systematically, partners can be more confident and helpful playing their role. The child with JNCL benefits from the presentation of categories that are relevant, and not randomly determined by the partner in the urgency of the moment. Additional memory support is provided through the use of the fixed order because it benefits expectation and recognition.



Figure 13.5 Objects of reference used on an electronic device with speech output

Objects of reference and time support

Objects of reference can be used to support choice making, memory, structuring of the day, orientation and communication (McLarty, 1997; Park, 1995, 1997). It is important that the objects that can be attributed a symbolic function are introduced at an early stage, so that the young person becomes familiar with using them. The objects can for example represent people, animals, objects, locations, activities or events. They can be whole objects, parts of objects or miniatures. Objects of reference can also be attached to electronic communication devices with speech output (Figure 13.5), enabling the person to express something, make comments and social greetings even if he is unable to speak with his own voice.

It is vital that the person can understand what is happening during the day and, when possible, make choices about what to do and when. In the early stages of the disease, the person may be able to use timetables in braille or Moon. Linking objects of reference to the text on the timetable may pave the way for using them when the person can no longer read braille or Moon (see Chapter 14).

When persons with JNCL reach the stage when they find it difficult to remember the timetable of a full day, the time table can be simplified by giving cards with a tactile symbol or an object of reference for only the upcoming and the next activity, for example for physical education while saying «Now physical education» and the symbol or object for lunch while saying «then lunch». If considered useful there can be a "finished box" to put tactile symbol cards and objects for activities that have been finished. There is considerable literature on this strategy and it can be accessed readily online by using the key words such as *calendar box* or *anticipation box*.

Chat book

Chat books enable the person to remember what he has done recently, and to communicate about these events and activities. They are also known as *remnant books* (Marvin & Privratsky, 1999), or *conversation books*. Whenever the person does something memorable or experiences something interesting or amusing, it can be made into a story. The story can be written in a book with a souvenir or memento attached, enabling the person to choose which story he wants to talk about (see Figure 13.6). In this way, the person may remember events and activities and share information about things that have happened. The chat book needs to be updated regularly. The person knows that his communication partner has read the story and therefore will understand the conversation, even if the person's speech is not easily understood. A parent described the use of a chat book.



Figure 13.6 A chat book

Key ring with laminated sheets that describe something about her. For example five red sheets about school, five white sheets about the family. Every sheet gives some information about her and by using the ring, you can ask her further questions about the subjects.

At a later stage, the book may be supplemented with audio recordings, which the person can listen to, or even be recorded on to a computer or an electronic communication device, enabling him to share the stories independently.

Memory/experience book

A memory book is another way to enable the person to activate memories and communicate with people about things that they have done and which are interesting and meaningful to them. A memory book can contain stories about the person's life and include recordings made over time of activities, friends, music, and so forth (Figure 13.7). This is a more permanent and long-term record of the person's life. The book should be created jointly by everyone who knows the person well, particularly family and friends.



Figure 13.7 Music page of a memory book

Communication passport

The communication passport is usually a small booklet with the most important information about the person (Millar, 1998). This may be a useful tool for people who are not familiar with the person, for example hospital staff or support staff, to quickly learn some relevant biographical information, like the things that the person is interested in and the most effective ways to communicate with him. It describes how the person shows emotional states and physical needs, for example sad, happy, tired, hungry, and thirsty or need to go to the toilet. It is written in a simple, easily accessible style in the first person and will help people to understand and relate to the person rather than his problems or disability.

The passport needs to be updated regularly, so that it contains current, relevant information about the person's needs, abilities and interests. Photos can be included to illustrate how to support the person or to provide communication partners with topics of conversation.

Community request cards

Community request cards are a simple way of helping persons communicate more effectively in the wider community with people who are not familiar with their communication support needs (Figure 13.8). People supporting the person use all available communication methods to find out what he wants to say in a particular

situation, for example what he wants to order in a café or make an appointment at the hairdressers. The support person then writes short note outlining this а information which the person with JNCL gives to the person he wants to communicate with. This enables members of the public to know exactly what the person wants even if they do not understand his speech or other forms of communication. It makes the person the center of the interaction, rather than the support worker or family member, and the communication partner is more likely to address the person with JNCL directly in return.



Figure 13.8 Community request cards

Prioritizing communication

The findings from the present project show the importance of giving priority to communication and starting early. It is not possible to predict the individual course of the disease, but young people with JNCL will develop problems with expressive speech sooner or later. AAC should therefore be considered. The learning capacity is best early in life, before speech problems start developing. Teaching AAC therefore should start early and preferably before speech starts to decline or has become severely impaired. At the same time, there are large individual differences and some young people with JNCL probably will be able to learn AAC in adolescence or even adulthood, as illustrated in these parents' story about their son:

We began asking for evaluation of a communication device when he was 12 on the advice of a Batten researcher. Unfortunately, the school did not get on the ball and get anything done until he was around age 16. He is 18 now and really just starting to use the device.

Some staff members also mentioned that they had tried alternative means of communication:

When speech and facial expressions were lost, we tried to invent dialogues hoping to create the ideas, which she wanted. That was only possible because we knew her well. It was nearly impossible to invent new signs etc., because she could not keep them in mind. Only when she knew them from before we could use them.

I have always thought it was a pity for her having been deprived of different aids and initiatives – because many educational measures were offered when it was too late to be of any use for her – because it gets more and more difficult to learn new things as the disease progresses.

The need for future research

Both the findings from the present project and clinical experience indicate that augmenting communication for individuals with JNCL can be important for sustaining their life quality. The ultimate goal is to give persons with JNCL some extra years of fluent communication when possible. However, there is a need for more systematic knowledge. The present JNCL project is just the beginning of this process. Conducting new research and collecting clinical experience to obtain more information is necessary for learning more about augmenting communication for children and adults with JNCL, and thereby improving their life quality.

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Teaching Reading and Writing to Students with Juvenile Neuronal Ceroid Lipofuscinosis

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Literacy is a basic element of modern cultures and teaching of reading and writing is a core element in education. Literacy allows information seeking and is a tool for collective memory and history construction, as well as for self-expression. Literacy is also important in communication and when using technical aids and computer programs. It is an aim that all children should be given opportunities to reach their individual learning potential, independent of their abilities and disabilities. There are teaching guides and teaching materials for braille in many countries, developed for children with visual impairment. For children with juvenile neuronal ceroid lipofuscinosis (JNCL), visual impairment and dementia will influence their ability to maintain reading, writing and mastery of literacy throughout childhood and adulthood, but their desire for self-expression may be maintained and expression through writing may remain a central activity even in adulthood.

Teaching of literacy

The function of written language is to reflect the spoken language (de Saussure, 1974; Liberman, 1999). Thus, teaching reading means helping the student understand the relation between spoken language and the characters representing the alphabet. This main aspect of literacy teaching is the same regardless of the modality of the characters to be taught, whether they are print, braille or Morse.

Literacy instruction includes two main approaches. *Skill-oriented instruction* is described as a phonological part-to-whole approach (Wormsley & D'Andrea, 1997) typically starting with speech sounds and letters, followed by spelling to construct words, or an initial whole-word approach which starts with words that are then analyzed into spelling. The teaching is generally the same, there is no individual choice, and reading materials tend to be prefabricated. *Meaning-oriented*

instruction is a whole-to-part approach, starting with meaningful sentences or text that is subsequently broken down into words and letters. The material consists of stories and key words from the children's own experiences, knowledge and interests, and focuses on both whole word and letter recognition. The reading material is often produced through a collaboration between teachers and students (Wormsley & D´Andrea, 1997; Wormsley, 2016). Many teachers use a combination of the two approaches, to accommodate the students´ individual needs.

Literacy learning is often described as a stage-like process. *Emergent reading* is the first stage. The child understands that spoken words correspond to written words and subsequently that words are made up of letters and letter clusters that correspond to speech sounds. *Beginning reading* is the second stage. The child learns elementary-level reading skills and understands the contents of sentences and short and simple texts. Finally, *advanced reading* implies reading for inquiry and knowledge building (Kamei-Hannan & Ricci, 2015). Teaching literacy is therefore not only about teaching the code (Swenson, 2016; Wormsley & D'Andrea, 1997).

Modes of reading

Reading can be visual (ordinary print, alphabetic or logographic) or tactile (braille or Moon). Access to texts can also be auditory, such as when text is read aloud by another person or by text-to-speech programs with synthetic speech. Studies show that many individuals with visual impairment vary between tactile reading, visual reading and auditory text access (Vik, 2008; Vik & Fellenius, 2007). For a short period, students with JNCL may be able to use all three modes, but for many, the visual reading mode becomes inaccessible relatively early. When a child has received a JNCL diagnosis, it is assumed that the future reading mode will be tactile and that auditory text access will be very important. Listening to audio books remains a favorite activity for many adolescents and adults with JNCL (see also Chapter 19).

There are two main tactile reading systems in use by students with JNCL: Moon and braille. For a period of approximately 200 years, Moon and braille were both used, but about 50 years ago, the use of Moon subsided. One important reason was the lack of functional writing tools for Moon relative to those available for braille, which had the slate and stylus and later the Perkins Brailler (see Figure 14.5 below). Today, the problems related to writing Moon letters are solved because Moon letters and text can be printed using an *embosser*. However, today braille is used all over the world while Moon is mainly used in the UK and alongside with braille.

Moon

Moon is a tactile alphabet and the shape of many Moon letters are similar to printed capital letters or parts of capital letters (see Figure 14.1). Because of the strong resemblance to the print alphabet, Moon seems easier to learn than braille for peers and people in the person's environments. The Royal National Institute of Blind People (RNIB) published a report based on interviews with teachers and other professionals working with Moon (Cryer, Home, & Wilkins, 2011). The report suggests that Moon is easier to learn than braille. Moon letters are easy to identify with touch, they have a strong resemblance to the print alphabet, and are easy for sighted people to learn (peers, parents, teachers etc.), and the use of Moon can therefore lead to more inclusion than braille. In the UK, Moon is

recommended (a) for persons who used to have normal vision but who have lost their eyesight and therefore need to learn a tactile reading mode, and (b) for students who for some reason have problems learning braille.

In the UK, Moon is chosen for some students with JNCL because most children with JNCL have achieved some print reading and writing skills before they start to lose their vision. In the present project (see Appendix A), some of the students did not become good braille readers. It is possible that Moon might have been a better alternative for some of them, as suggested by these parents' comments.

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Figure 14.1. The Moon code

My daughter would do well in spelling tests up until 8, I seem to remember words like elephant and started to be less able to retain the spellings from memory. Braille was ok at first. She learned the alphabet and she started grade 2 braille but then started to forget even when enlarged and the VI teacher was quite disparaging (before diagnosis). It was a distressing time. I had researched and found Moon font and attended a conference and Moon base and it was agreed to try it and it was very successful and my daughter really enjoyed having her own language and the symbols being similar to the words she had learned. Reads in Moon, was taught this over a period of a school year before sight had completely gone. Moon was chosen over braille as the VI teacher wanted to have something in place for when fine motor skills started to decline. Reads well in Moon although is a little slower and reads shorter books.

In the early stages of Moon it was very successful but lack of resources meant that books were mainly homemade ... the font size needed to be bigger than standard Moon font. My daughter really enjoyed Moon.

Braille

Braille is a system of raised dots. The dots in a braille cell are numbered from 1 to 6. The left column consists of the dots 1, 2, and 3 with 1 at the top, 2 in the middle and 3 at the bottom. The right column has dot number 4 at the top, 5 in the middle and 6 at the bottom. Braille letters consist of different dot combinations, for example the letter *m* consists of dots 1, 3 and 4 (see Figure 14.2).



Figure 14.2 The braille cell

Braille is usually read by moving the fingers on both hands from left to right while touching the

braille letters lightly and smoothly with the fingertips. To facilitate writing braille on a computer the braille cell is extended to 8 dots to make more dot combinations possible. Computer-based reading and writing tools have made it possible for braille readers to navigate on the internet and have access to synthetic speech that reads the text on the screen. Braille is the tactile alphabet that is most commonly used all over the world.

Uncontracted and contracted braille

Uncontracted braille corresponds to visual writing as all the letters in words are written, for example the word *happy* in Figure 14.3. Figure 14.3 also shows an example of a sentence written in contracted braille. Contractions are abbreviations, such as writing the letter combination *th* with one instead of two braille cells. Also, *in* is written with one braille cell in contracted braille. The word *thin* consequently is written with two braille cells in contracted braille, one braille cell for *th* and another braille cell for *in*. The word *braille* is written with three braille cells as shown in figure 14.3. There are several levels of contracted braille. Braille readers will usually achieve a higher pace when reading contracted braille.

School beginners start with uncontracted braille as this is considered easier to learn. There are considerable differences between countries concerning how early they start with contracted braille. In Norway, contracted braille is introduced through separate courses and switching to contracted braille is a question of individual choice and personal preference. Braille texts on paper are usually written in uncontracted braille, irrespective of the student's age. Consequentially, in Norway the majority of braille readers read uncontracted braille. However, when reading braille on a computer display, students can choose between uncontracted and contracted braille. The policy in UK and some other countries is different. Children start learning contracted braille during primary school and from an early age, most teaching materials and texts are produced in contracted braille in these countries.

Teaching braille

«A common definition of literacy is the ability to read and write at such a level as to be able to meet daily living needs» (Argyropoulos & Martos, 2006, p. 676). National curricula describe literacy skills as pivotal and a foundation for learning in general. There is an international interest in making braille literacy as accessible

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Figure 14.3. Letters, words and sentences written with braille code

and unambiguous as possible to facilitate literacy learning for braille readers worldwide (Cryer et al., 2013). Tactile reading is considered more difficult to learn than visual reading and represents bigger challenges for memory. Sighted children meet written words and texts in their environment to a much higher extent than children who are blind and are thus incidentally exposed to varied learning opportunities every day (McCall, 1999). To educate teachers and facilitate better learning opportunities for pupils with blindness, several countries have defined special learning areas for children with severe visual impairment. An example is the Expanded Core Curriculum (ECC) as described by Lohmeier (2009), which promotes tactile reading and writing as core components.

Traditionally, teaching of braille is mostly based on a parts-to-whole approach. This is due to the fact that tactile readers are not able to perceive whole written words or sentences simultaneously, such as sighted students do (Millar, 1997). During reading, the fingers of the tactile reader will encounter one letter at a time. In skill-oriented instruction the child learns the smallest elements first, that is, the letters, and builds upon these fundamental skills to develop more sophisticated literacy skills. This teaching method is the traditional method for teaching literacy both for visual and tactile reading and writing (Millar, 1997; Wormsley & D'Andrea, 1997).

Since the 1940s different teaching methods based on a whole-to-parts meaning-oriented instruction have been developed. These methods were not considered applicable for teaching braille because of braille readers' problems perceiving larger entities like letter combinations or words simultaneously. However, since the 1990s whole-to-parts methods have been adapted to suit braille readers. An example is I-M-ABLE (see below) that was developed specifically for at-risk students who found learning braille difficult and showed a slower learning pace than expected for their age (Wormsley, 2016). Slow learning pace can be due to for example language disorders, dyslexia, intellectual disability or attention deficit disorders.

Literacy teaching should suit the individual needs of the student in the best possible way. «The answer [to teaching reading] is not in the method; it is in the teacher. It has been repeatedly established that the best instruction results when combinations of methods are orchestrated by a teacher who decides what to do in light of children's needs» (Duffy & Hoffman, 1999, p. 11). Reading pace – number of words read correctly per minute – is often used to evaluate students' reading skills. Braille reading is usually slower than print reading, but the reading pace varies a lot among braille readers. Many achieve functional reading skills even if the reading pace is slow. Children develop a fast reading pace when they engage in varied, motivating and enjoyable reading (Swenson, 2016).

Braille courses for students, families and staff – an example from Finland

In many countries, the educational authorities offer braille courses to students, teachers and parents. In Finland, a five-day course for parents is organized by The Finnish Federation of the Visually Impaired (FFVI) twice a year. The aim is that the course participants should become familiar with braille as well as pre-braille skills. FFVI offers separate braille courses for professionals.

In Finland, braille teaching for children with visual impairment follows a three-step procedure. In the first step, the braille material is produced with extra spacing between letters, words and lines. When the child has achieved a smooth touch to the braille letters and shows progress in reading, teaching proceeds to the second step. The extra spacing between letters and words is removed, but extra space between lines is kept to make it easier for the child to follow the lines. In the third step, the extra space between lines is also removed. Each child proceeds individually through the steps, according to his or her skills and abilities.

The resource center Valteri, Center for Learning and Consulting, Onerva-unit in Jyväskylä, organizes courses for students with visual impairment in preschool age and school age. A one-week support course for students who are blind always includes different subjects and techniques to support education and functioning in everyday life, and braille training is always included in these courses.

Reading and writing for children with JNCL

Early literacy learning for children with JNCL will be influenced by two main factors related to the disease. First, visual decline and subsequent blindness will gradually compromise visual reading and make tactile reading the only reading mode available (see Chapter 4). Second, dementia will eventually influence all learning, also literacy learning. Complex skills like reading and writing are particularly vulnerable. However, problems with memory and learning typically become apparent well after the child has started school but there is considerable variation in age of onset (see Chapter 5). Also, reduced skills in the hands and arms will affect tactile reading and writing skills, but for the majority of persons with JNCL this occurs quite late in school age. In the present study, the mean age for onset of these kinds of motor problems was 15.5 years (see Chapter 7). At this age, many children with JNCL who had relevant training have been tactile readers for several years.

There is a relationship between reading and working memory, but there is not agreement between researchers with regard to how working memory influences reading and reading disorders (Kendeou, van den Broek, Helder, & Karlsson, 2014;

Nouwens, Groen, & Verhoeven, 2017; Pelegrina, Capodieci, Carretti, & Cornoldi, 2015; Pimperton & Nation, 2014; Van Dyke, Johns, & Kukona, 2014). Some authors have suggested that braille places greater demands on working memory than print (Daneman, 1988) and others that problems with working memory capacity will make it very difficult or impossible for students with JNCL to learn to read and write (Kristiansen, 1988). However, research findings contradict the claim that it is very difficult or impossible for children with JNCL to learn to read and write. A review of the reading and writing skills of 39 students with JNCL who received support from Tambartun resource center (Statped) in the period from 1995 to 2011 reported that 23 of the 39 students could read and write braille at some level, but skills varied considerably among the students. Seven of the 39 students could write but not read braille. These students used braille keyboards and displays, except for one student who touch-typed on an ordinary keyboard. The remaining nine of the 39 students were unable to read and write braille (personal communication, Mohammed Beghdadi, 2012). These findings indicate that a majority of this sample of students with JNCL benefited from tactile reading and writing, and thus support the claim that students with JNCL should be given opportunity to learn braille, although for some, learning Moon might be a better option.

Also, findings from the present survey (see Appendix A) contradict the view that students with JNCL are unable to learn to read and write. The parents were asked to indicate their child's best level of reading competence in different age periods, that is, whether the child was a) reading letters, b) reading words, c) reading short texts, or d) reading books. Both reading print and reading braille were assessed. Seventeen participants of a total of 107 (15.9%) were reading books in braille during at least one of the age periods. Two of the 17 children had good braille reading skills before the age of seven, because they had started receiving braille instruction very early. Another 21 persons (21.6%) were able or had been able to read short texts. These findings show that about one-third of the students in the project developed quite good braille reading skills. We note that some of the children in the project were rather young, and it is possible that some of them might have the capability to learn to read books or short texts when they are older.

The age of the participants ranged from 6 to 34 years. Figure 14.4 show the percentage of the participants who had been able to read and write a text in print or braille in different age periods. The results show that print was most used in the early phases and peaked around age 8–10. At this age, 40 percent of the children read text in print, after that the percentage dropped. Braille reading and writing peaked at 14–17 years and then started to drop. Nine participants between the age of seven and 13 years were reading in both print and braille. The typical picture is thus that most of the younger participants read either print or braille.



Figure 14.4 Percentage of individuals who could read and write texts in braille and print in different age periods

Note: The number of participants (N) varies due to the age distribution. Each age span has two Ns: The first is for reading, the second for writing.

Braille reading skills were further developed and maintained after the ability to read and write texts in print was lost. Figure 14.4 also shows however that ability to read short texts or books in braille did not last. Even those who developed excellent reading and writing skills eventually lost these skills as the disease progressed. The length of the period with preserved braille reading varied considerably. None of the 23 participants described by parents who had reached the age of 23 by the time of the survey practiced reading or writing, but two participants aged 21 and 22 were still reading books in braille. One person, who was only described by staff and therefore not included in Figure 14.4, was still reading at the age of 23. The findings from the present project further indicate that many adults still took pleasure in listening to audio books (see Chapter 17). Thus, the ability to enjoy the content of texts and books can be preserved through auditory access.

It seems likely that experience with reading print will facilitate braille learning. For a child who has learned to read printed letters, learning braille demands practicing the already learned reading skills with the addition of using a new modality. However, many factors may influence braille learning and it is not clear how the skills associated with reading print through a visual mode are best transferred to using a tactile reading mode. The present study indicates that children who became blind before the age of ten tended to become the best braille readers, although there were a few exceptions. Several parents mentioned that early braille learning was due to early onset of blindness: With his rapid loss of eyesight, we began braille along with large print access. He did better with braille and really enjoyed learning it. He spent hours typing on his brailler, learning spelling words and reading his books. He was very proud of the fact he could do braille. When older he'd get frustrated with not remembering cells, the teacher just went back to what he could do and the simpler words. For him, braille was the best thing he did, he read books in the dark way after bedtime many a night.

During preschool (age 5–6) he was already interested in letters and numbers, his visual impairment increased and he suddenly lost his interest in these. Starting his second year at the school for the blind, he started learning braille and he could read like a little world champion. He also learned shorthand braille and mastered it. At the age of 20–21 years his reading competence decreased and stopped about age 22 years.

"Arthur" was blind at age six years. After that he read braille. Soon after getting blind, Arthur received teaching in braille and he was skilled in braille. However in the final stage of life, it became more difficult.

Several parents however, described late onset of braille teaching because the school was giving priority to residual vision and print reading as long as possible, such as these parents:

He stopped reading normal print, as even the screen-reading device wasn't sufficient in its magnification. Learned braille for only a short time, as his mental deterioration happened very quickly. Nevertheless, he went on practicing on type writer.

Gone blind at the age of about eight. Cognitive skills were too low from 8–10 years of age to learn braille.

Stopped reading because of cognitive decline and complete loss of sight.

Because of her blindness she cannot read normal print any longer. While it was still possible, they did not work with braille at the school for the blind. Eventually it was too late.

She wasn't able to read black print at the age of ten, neither with technical aids. Her cognitive capacities were no longer sufficient to enable her to learn braille.

Staff members also commented on braille reading:

Could read print well until JNCL symptoms at about age eight. Print was enlarged but by age 13, she could no longer read even enlarged print. Was taught grade 1 braille but progression of JNCL prevented consolidation of braille learning due to deterioration of memory.

Even in this area there was considerable variation. A few students who became blind in their teens still became good braille readers. The tendency however was that early introduction to braille appears to provide the best chance of becoming a good braille reader, irrespective of residual vision or reading skills in print. It is not possible to infer from the results whether print reading skills facilitate braille learning, but as long as the child enjoys reading print, this activity should be encouraged together with the provision of braille instruction.

The question has been raised whether students with JNCL should advance to learning how to use contracted braille. This progression would entail learning more braille symbols and abbreviations (see Figure 14.3) and learning to read words written in different ways from what they had first learned, that is, through uncontracted braille. One parent commented:

Our son began braille instruction in first grade, was fluent reading contracted braille by fourth grade and loved reading volumes of books until he was around 17. At that point fine motor coordination was declining and short passages and much younger level of texts were his favorites. He stopped reading braille at the age of 20–21.

This child had started to learn braille in first grade and became a very good reader. Literacy learning from young age might be one reason accounting for fluent reading through both uncontracted and contracted braille. Because contracted braille presents additional cognitive challenges, many children with JNCL will likely have problems learning it. Thus, for them uncontracted braille will be the most efficient way of reading. Learning uncontracted braille constitutes, in itself, a success for a child with JNCL. Teachers and families should take into account individual learning potential and motivation when deciding about which path to follow; both paths begin with uncontracted braille and the choice is then either to remain with that system, or progress to contracted braille.

Braille education for students with JNCL

In the present project, most of the participants had been offered braille instruction. However, seventeen percent of the participants did not get this opportunity. Failure to offer the opportunity of learning a tactile reading mode might be driven by parents' and teachers' low expectations for learning capacity in individuals with JNCL:

The parent thinks that the student should not use energy on something that would decline anyway. There were enough defeats.

Because of learning disorders, the professionals recommended not to start teaching braille at all. After she lost her vision all reading is impossible.

The special school would not offer braille.

Moreover, comments indicate that some schools did not focus on literacy for students with JNCL, like this teacher:

No, that [braille] is not something we are focusing on. Because those who know more about the disease have told us that she will never become a braille reader.

In an interview, a parent seemed to express regret with the decision not to teach braille:

She would have been able to learn braille, I actually think so. If we had started at once I actually think she could have learned braille.

Staff and parents alike described problems with access to professionals with braille competence when the child was not attending a special school or unit for students with visual impairments. Changes in staff resulted in loss of competence, and this usually happened several times over the school years. Transition to a new school or new counseling services could also result in long periods with no braille competence in the staff. One parent commented:

Changing teachers has led to several very long periods with no braille teaching. Knowledge and skills are lost as a result of teachers lacking competence.

A staff working with a young girl with JNCL commented:

The teacher [at the previous school] meant that they could not [learn braille]. The teachers had no competence in braille. So I said neither do I, but then you have to get somebody else come and help her.

Some parents and staff in the present project describe students with JNCL who had received braille training but who nevertheless did not learn to read or write braille and so eventually the teaching was terminated. Comments from parents indicate that this abandonment of braille instruction was related to the students' cognitive problems or lack of motivation.

He was never interested in reading.

His intellectual stagnation made it impossible for him to learn braille.

She learned to read just like most kids but at age eight she started to go blind and the school tried to work with braille. However, she never really took to braille so it made it hard to work with her. She just couldn't get it and make sense of the braille.

Dementia is one likely cause for the problems with learning braille. If learning is difficult and progress slow, problems with motivation are also likely to occur. Another factor is that some children in the general population have persistent problems learning literacy. It seems reasonable to assume that some children with JNCL have similar problems and that these problems are not specific to braille. The last quotation above illustrates the case of a child who had successfully learned to read through the visual mode (i.e., print) and experienced reading problems only with the tactile mode (i.e., braille). This case suggests the possibility of modalityspecific literacy learning difficulty.

Braille literacy and impact on life

In spite of difficulties and decline in reading and writing skills, for some of the participants in the present study, literacy skills have played a crucial role in their lives, as evident in this quote from an interview with the mother of young woman with JNCL:

I think maybe the most important for her was that she took pleasure in writing when she learned writing braille. And it meant so much to her that she could write down her thoughts on paper. That was also important. She expressed it [the thoughts] that way, both pleasures (...) and when she was angry we heard her go upstairs and then we heard the Perkins brailler. But she wrote, maybe you know that, that she published a poetry collection. She wrote those things before she was 18 years old.

A staff member working with a young man recounted:

Yes, well, when he came here at the age of 23, he read braille. So he read books himself, sat by the computer.

Then [*at the age of 23*] *braille books were sent every month from the national organization for blind people.*

Today he does not read braille. That is, it happens, we have cards with braille, he plays cards, and even that is getting difficult, and that is only one letter per card.

In summary, the results from the present project show a great variety in braille reading skills among students with JNCL, which in part may be due to differences in age when teaching of braille was initiated. Some of the participants became proficient readers and some did not progress beyond letter- or singleword recognition. The participants' interest in reading and writing also varied considerably. Some took great pleasure in reading or writing, and some did not. Some were never given the chance to become literate as braille teaching was never offered. Some of the quotations above are striking in that they speak to a strong liking for self-expression of one's own thoughts, ideas, feelings through the medium of written language, an interest that was maintained over many years. Braille made this self-expression possible.

Strategies in teaching reading and writing skills

Reading and writing are both technical and functional skills. Technical skills are about recognizing letters and words, producing letters and spelling words. Functional skills concern the use of reading and writing for a purpose, to do something with words, such as writing a story, doing school work, reading a text to find specific information, or reading the day schedule. It is important to ensure that the child's reading and writing become functional from the very beginning. Children with visual impairment should encounter tactile writing everywhere in the environment, just as sighted peers experience visual writing.

Reading and writing braille

Teaching braille usually starts with teaching letter and speech-sound correspondence, and print-braille correspondences if the child has already learned to read print, and then proceeds with words and sentences. Teaching starts with the letters that are easiest to recognize and discriminate. Many children with JNCL develop an interest in reading early and find reading rewarding because it gives access to literature and information. However, some children find learning to read and write very difficult. Their progress is slow, they feel unsuccessful and may lose the motivation to learn to read and write.

Most students with visual impairment start learning to write braille on a Perkins brailler, which has six keys, a space bar, a line spacer and a backspace (Figure 14.5). The six main keys correspond to the six dots of the braille cell. Paper is inserted into the top of the brailler and once the keys are pressed, raised dots appear on the paper. During writing the six writing fingers are permanently placed on the six keys and there is no need for searching to locate letters.

There are also braille keyboards for computers (see Figure 14.6). When writing on a computer the student can receive continuous feedback with synthetic speech, which makes it easy to discover and correct spelling mistakes. The keyboard in Figure 14.6 has a tactile display of 40 braille cells so the student can read the last written words with the fingers.

Children who have started to write on a Perkins brailler later usually switch to a braille keyboard and synthetic speech. Pressing the keys on a Perkins brailler requires more strength than pressing the keys on a braille computer



Figure 14.5 A Perkins brailler

keyboard. Using a Perkins brailler thus has positive effects for finger coordination and strength.

Screen readers read the information on the computer screen in synthetic speech. The user utilizes keyboard commands for navigation instead of a mouse. A screen reader can be set to read all text on the screen, including menus and dialogue boxes. Selection of a screen reader may depend on the

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Figure 14.6 A braille keyboard with a small reading display

operating system of the computer, funding and individual preference. The user can choose synthetic speech with a familiar dialect and the output speed of the screen reader, and decide which punctuations in the text will be read out loud.

Touch typing

When writing fluently on a Perkins brailler or braille keyboard many students with visual impairment go on to learn touch typing on an ordinary QWERTY keyboard to increase the speed of writing. In the present project, one of the participants had learned and used touch typing.

"Jane" was fluent in reading braille and read complex language (the whole Harry Potter series, Romeo and Juliet) in braille until she was 15 years old, then lost the skill. She could produce braille, but never as well as she could read. She could type by touch, but never as well as she could read. She listened to audio books and her comprehension was good until around age 14–15. Then she started to regress, preferring books for younger children and more repetition.

The story of Jane above illustrates that some students with JNCL can learn touch typing. However, the decline in cognitive and fine motor skills will for most students compromise learning a new way of writing that is completely different from the way they already have learned. Although there might be exceptions, in general students with JNCL might benefit most from continuing to write on a Perkins brailler or a braille keyboard.

Writing with Lego blocks

This is a writing tool created especially for young students with visual impairment. It consists of a Lego board and Lego blocks with braille letters (Figure 14.7). The board has six Lego blocks with each letter (29 letters in the Norwegian alphabet) and six punctuation signs, for example full stop and exclamation mark. The student can write words and sentences by placing Lego blocks with braille letters on two lines.



Figure 14.7 Lego board with braille letters and two lines for writing words

Reading and fingertip sensitivity

The fingertips are used when reading braille and finger-tip sensitivity is therefore important for perceiving braille cells and dots when moving the fingers across the paper. The same kind of tactile sensitivity is not required for braille writing. Writing braille is therefore considered easier to learn than reading braille, although there are individual differences even here.

Some students with JNCL never learn tactile reading but may still learn braille writing. In

fact, writing may become a person's favorite activity even when his reading skills are poor. When a person can write but is unable to read braille, it is important to ensure that the person receives reading help or has access to a computer with synthetic speech output. Lack of equipment or support may reduce the continued use of literacy skills and hinder access to an engaging activity, as indicated by the staff member's comment about the young woman below.

She became an expert on that [writing braille], she never read braille because she did not have enough motivation, but she wrote and took pleasure in writing throughout school.

I think she could have managed this [to write] even longer, but then she stopped going to school (...) and it became difficult to follow up because residential staff did not have enough competence to use the computer.

Single word reading and writing

Functional use of reading and writing skills is possible even if the person reads or writes only single words. Single-word reading or writing may be used for labeling books and teaching materials, contents of shelves and drawers, electrical equipment for cooking, *on* and *off* on light and other switches, calendars and daily or weekly schedules, or making recipes, and thus contribute to independence and orientating in the environment (Wormsley, 2003).

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If a student can recognize only a few single letters, this skill may be exploited to provide more functionality. For example, people routinely use abbreviations (even single letters) as codes in text messaging to save time and effort. By pairing a single letter with an associated word, preferably a word beginning with that letter, the word's referent can be identified. For example, the braille letter *s* can indicate the activity "swimming" on the child's schedule. In some activities, single letters can be more practical than complete words because of limited writing space, such as labels on a deck of cards or other game equipment (see Chapter 21).

The I-M-ABLE method for teaching braille

I-M-ABLE stands for «individualized meaning-centered approach to braille literacy education» and is developed for emergent and beginning braille readers who have problems learning braille the traditional way and need a more individualized approach for learning tactile literacy (Wormsley, 2016). This approach was inspired by meaning-centered literacy teaching for sighted students and adapted for tactile reading on the basis of Wormsley's own experience as a braille teacher and counselor.

An important element in Wormsley's method is using words and texts that give an immediate sense of meaning for the individual, that is, based on individual experiences and interests. Wormsley claims that the students' emotional evaluation of the reading material can be of great importance. She recommends a highly individualized reading vocabulary and the production of individual teaching materials. There are no ready-made materials and the teacher creates the teaching materials after assessing the student's interests and experiences.

I-M-ABLE can be used as an alternative or a supplement to other teaching strategies. The list below contains some of the main elements in early literacy instruction from I-M-ABLE. Note that some of the elements on the list are used in both skill-centered and meaning-centered approaches.

- Initial assessment of the student's reading and writing skills and attitudes toward reading and writing.
- Collecting information about the student's interests.
- Creating a braille-rich environment: Wherever there is print, there should be braille.
- Frequent instruction. Emergent and beginning braille readers should have 1–2 hours of training every day.
- Identifying key vocabulary words and phrases. Words and phrases must be emotionally laden and promote the student's engagement in

reading activities. The key vocabulary is the basis for choosing words for the first reading instruction.

- Using brailled flash cards for the first words and phrases.
- Selecting first words and phrases that have features that make them easy to recognize and to discriminate, such as short words and long words, or different initial letters.
- Teaching tracking of finger movements to promote fluent reading and prevent unnecessary stopping and scrubbing up and down on the letters to identify them.
- Not asking the students to identify words that they do not yet easily recognize. In the early stages of reading instruction, this will probably lead to scrubbing. Tell the students what word they are going to find on the card and ask them to find the word and read it.
- Teaching reading some key words first, and then introduce the first letter.
- Making sure the teaching materials are motivating and based on individual preferences.
- Using word games as a supplement.
- Keeping continuous records of words and letters learned.

Writing stories based on own experiences and preferences

Instead of or as a supplement to ordinary textbooks, students can write their own stories. The contents and complexity will vary but the stories should be based on the student's own interests and preferences. For emergent writers, texts should be very simple and if possible contain many repetitions of words, such as in this example:

I like pizza I like birthdays I like Harry Potter

Repetitions may function as formulas (Tomasello, 2003) and make the story easy to read. The child may learn the story by heart and have a feeling of reading it, even if the reading skills are not good enough for spelling and reading the words. Moreover, motivation for reading one's own texts is often higher than for reading ready-made stories and texts. Motivation is very important for emergent learners, and the importance of motivation for people with JNCL has been amply demonstrated in the quotations presented earlier in the chapter.

Creating a braille-rich environment

Most students with JNCL attend mainstream classes during their first years in school. In some countries there are no or very few special schools for visually impaired children and other special needs. In such countries, many students with JNCL attend mainstream schools throughout the school years. The teachers will rely on courses and guidance from resource centers to build the necessary competence. However, when counselors visit such schools, there is often no sign of braille in the classroom, except on the desk of the student with visual impairment. There is a lot of printed materials on walls and in shelves and drawers for sighted students, but not for the braille readers who are exposed to written language only while sitting at their desk.

Braille instruction sometimes takes place in a separate room to minimize distractions for the student, and to avoid disturbance to other students from the synthetic speech or noise from the Perkins brailler. Thus, the classroom situations experienced by the braille reader and the classmates might be very different, and they may unintentionally signal that braille is not something classmates should engage in. Creating a braille-rich environment is therefore one measure to enhance inclusion in the mainstream classroom (see Table 14.1). Braille should be present in every part of the classroom, not only around the desk of the braille reader.

The classmates will also learn something about braille when braille letters and words are present in the classroom and elsewhere in the school. Braille becomes less special and more available to all students. In fact, introducing braille to the sighted classmates is the best way to ensure inclusion of the braille reader. Experience from mainstream classrooms shows that sighted students often find braille fascinating and learn braille with great enthusiasm. It is necessary that peers get some knowledge and basic skills related to braille to understand and appreciate the work of their braille-reading classmate. With rather simple means, braille can become a natural part of the school environment. Peers will become aware of the similarities between alphabets, independent of modality, so there will be learning gains for everybody.

Table 14.1 Some elements in a braille-rich environment

- Place a braille alphabet on the wall next to the print alphabet
- Write labels on the students' belongings in print and braille
- Label the contents of cabinets and shelves in print and braille
- Write messages between home and school in both print and braille
- · Write lists and recipes used in the class in both print and braille
- Write song texts and other materials used for the whole class in both print and braille

Introducing braille to the classmates does not have to be time consuming. Much can be done with a couple of lessons on braille instruction, and integrating braille into some other lessons, and it may have a substantial effect on inclusion.

Simmons (1994) describes a successful approach to braille teaching in the mainstream classroom. She worked as a teacher for students with visual impairments in an elementary school, and some students in the school were braille users. However, she observed negative attitudes to braille, especially among the students who were blind. They often avoided using braille in class, such as this girl: «I saw her [a beginning braille reader] begin to develop some subtle, negative attitudes toward braille and decided I needed to act as a resource to inform and encourage developing social skills and positive attitudes toward braille». Simons therefore started to teach braille to the sighted students in collaboration with the class teacher. She gave the sighted students access to braille and tactile materials, including a Perkins brailler. The sighted students became very interested in braille and all the negative attitudes evaporated. Simons organized a voluntary braille class, which quickly grew from three students to two classes with 12 students in each. Over a period of three years, approximately 125 sighted students attended the braille classes, which were organized every day during lunch recess or just after school. Finally, Simons organized a Braille Club where sighted students and students who were blind worked together on projects with themes varying from Eastern cultures to social etiquette or Abacus. The students who were blind thrived in the positive attention from the classmates and gained confidence and status. Braille became a natural and integrated part of school life for all the students.

A positive attitude to braille learning and braille materials and tools is a prerequisite for successful braille teaching. If a student is working alone with tools that the classmates do not understand or show interest in, inclusion is likely to be seriously compromised.

Creative writing and fan fiction

As writing skills become more advanced, students will be able to write with more elaboration and complexity. Creative writing and fan fiction are examples of genres that have been important for some persons with JNCL in Norway.

Creative writing

Creative writing is based on the writer's imagination. It may include different genres, such as poetry, fiction or screenplays. The creative writing process may involve developing characters and settings for the story, elaborating the story plot and so

forth. Sometimes creative writers are writing only for their own pleasure. At other times, they may want to make the texts available to others. Then the writers must have the skills necessary for making the story interesting for potential readers.

When using creative writing in education, the individual's motivation for text production is the main success factor. Teaching of orthography will be second to encouraging the creative process and the students' wish to express their thoughts and feelings. In the final stage of the writing process, the teacher can ensure that the text becomes comprehensible with a minimum of focus on spelling mistakes and other needs for correction.

The feedback from a teacher, parent or classmate is important. The teacher's feedback should be adapted to the student's writing skills and take the form of a dialogue between the student and the teacher. The feedback should motivate the student to continue working with the text, and comments and questions should be offered during the writing process and not after the student considers the text to be finished. To ensure that the student understands the feedback, the teacher's questions and comments should be specific and directly to the point. The adult should be an interested and enthusiastic supporter in the writing process, and ideally, the student should be looking forward to the feedback.

Students will often enjoy writing more when the text is related to their personal interests and favorite activities. One of the genres within creative writing is fan fiction.

Fan fiction

Fan fiction is typically written by persons who admire a specific novel, movie, television series, and so forth. Most fan fiction writers are inspired by genres like science fiction and fantasy, such as the Star Wars films or the books about Harry Potter or Lord of the rings. Their focus may for example be on the story characters, the story plot, or the setting or world where the story takes place.

It is practical for the writer to locate the story in an environment described by another author, where the setting and the characters already have been developed. It saves a lot of time and work and enables the fan fiction writer to concentrate on designing a plot and creating an interesting story.

Quite a lot of people engage in writing fan fiction. Fan fiction mostly addresses a dedicated group of readers who are familiar with the literature from which the fan fiction is derived. Fan fiction enthusiasts can communicate with other fans on a variety of web sites. They can share their manuscripts and ask for reviews from other fan fiction enthusiasts. Fan fiction is rarely published, although there are some examples of published fan fiction stories. However, this requires knowledge, careful consideration and caution regarding copyright.

Three creative writers with JNCL

Early in a child's life there is no way of knowing whether writing can become an important base for future activities. It will depend on the individual's personality, interests, strengths and opportunities. This section describes the work of three Norwegian adolescents with JNCL for whom literacy has been very important. It includes short presentations of two young authors and their books, and a more extensive presentation of a young author written by his father.

Åshild – A poet writing about her own life situation

The book *Thanks for life* (in Norwegian: Takk for livet) contains poems that Åshild wrote during her teenage years (Figure 14.8). The poems describe her thoughts and reactions to losing her vision and having a shortened lifespan due to JNCL. Åshild's faith in God had a strong impact on her thinking and coping, and in many of the poems she asks God for answers and reassurance in a difficult life situation. Åshild passed away some years ago.



Figure 14.8 A poetry collection



Figure 14.9 A fantasy story

Ruben – Author of fantasy stories

The book *The live watermelons* (Norwegian: De levende vannmelonene) is about thousands of watermelons trying to destroy and take control of a city (Figure 14.9). The watermelons are stopped by the armed forces during the attempted conquest. There is a great fight between watermelons and soldiers, and as the book review stated: «You certainly don't want to miss it!».

Braille writing from a parent's perspective

This story, written by a father, integrates his son's braille writing in the wider life story of a young person with JNCL, with diagnosis, education, support services, and ordinary and not so ordinary events as part of a daily life.

When I got the message that our son had Spielmeyer-Vogt disease (JNCL), it felt like walking down a hill towards a dark tunnel. I imagined myself in a spiral without the circle being closed.

Tambartun Educational Centre

We met with Tambartun, which is a state educational center (Statped) responsible for facilitating the teaching of children who lose sight – children who become blind. In life, sometimes you get a hint about something that is wise. When you are young, someone hints that she is in love with you. As an adult maybe this could be a tip about an important book, a piece of music or a bargain.

At Tambartun, a wise woman said that «maybe it is a good idea for him to learn braille while he still is using vision». That became my token.

The beginning

We started with braille training at school before the summer break at the end of the second year. Matias is seven years old. Matias learns most of the letters from the teacher. He uses a mechanical braille machine diligently every day but for short periods. The teaching strengthens the use of muscle memory, like a piano exercise. When one plays a musical instrument, one knows that this is important.

The surprise

Matias has consistently made stories that have been printed and delivered to school. Just before summer comes the surprise. He has kept a big secret. In secret, the stories are sent to Tambartun. There, the texts are made with braille lettering, and they have made a book. This book is titled «Different *Stories» and has both black writing and braille. The book becomes his reading book in fourth grade.*

Introduction to braille and aids we used in the beginning We had a 6-egg cardboard package inserted with ping-pong balls to illustrate the braille cell. We had a board with Lego – each marked with dots, like braille letters. We had a small piece of wood that showed the braille cell and a braille typewriter that prints letters on thick paper.

Autumn

A little later, in the autumn, we got computer equipment with a keyboard and a program called "Sarepta". The computer program has speech synthesis that reads letters of words and sentences. The text appears on the screen for viewing. There is a braille keyboard and a display with 80 braille cells for reading braille below. On a separate board are five buttons or keys for quick navigation. We get going.

At home, we try to write every day, preferably at the same time, with established routines. A ritual between Matias and dad is emerging. A calm, intimate gathering one-to-one, like learning to play a musical instrument. It is all about communication and little about performance. We manage five to ten minutes at a time before he is tired. We have a fixed time every day after dinner, early evening. Sometimes we only turn on the machine and then turn it off again. Other times we get some words written.

We have simple writing tasks. We try to practice the keys. It is hard to remember a sentence while remembering the letters. Matias is able to remember simple words, but not more. Dad remembers what Matias is saying and helps with the writing. The reward is that the speech synthesis reads the word and the sentence.

Eventually this develops into a conversation, where I interview Matias when he tells. When a sentence is good, I catch the sentence and he writes. During this period, I will correct the orthography without paying any attention to it.

I see progress, I see frustration, I see happiness, and I see the feeling of mastering. I expect a faster progress. It takes a much longer time. We stay focused. At the same time, he is losing his sight gradually.

Our method

We try to do the same every night. Have as regular environment as possible. Sometimes he does not get tired. Sometimes we last for a long time.

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Now he asks: are we not going to write? Earlier he could not remember long words, he is doing well now. Now he can complete sentences. Reading skills increase as he reads his own texts.

At school A story is read in class, it gives self-esteem. He writes a poem.

First example text:

The stupid troll and the fox by Matias 12 years old

Once upon a time, there was a troll who had two hearts, but one of them was false. He boasted about it to a bear, and he said the bear would not be able to find it. However, the bear set off to look for it. He met an old woman who had her nose stuck in a stump. The old woman said: «Can you help me, I have had no food for a hundred years». «Well, if you have had no food for a hundred years then you can wait for a hundred years more», he said and laughed.

At home

It is easier to get started with the writing at home when there is something self-perceived to be told. Matias reflects on books we read in the evenings, or he uses stuff from movies and from fairy tales. Dad is easily bored, so he has to read something for Matias that he likes too. We read Harry Potter, Gulliver's Travels, Robinson Crusoe and Captain Nemo, Twenty Thousand Leagues Under the Sea and The Mysterious Island. We read The Three Musketeers. We read Les Miserables, and much more.

Learning

We work one-to-one. I am his supporter. I am organizing and correcting text. It is not so important to write correctly. Speech synthesis is the reward when the sentence is read. The teaching should not lead to any qualification. That is not the goal. Learning is a way to participate in society. We learn about things. About science, about history, about Christianity, religion, philosophy and philosophy of life. We learn math and we learn physics. In order to participate in the community around you, you must understand what others are talking about.

Learning = *Participation in society. Learning* = *not necessarily a qualification for a profession.*

He calls himself an author

Matias has read all the books about Harry Potter, Hermione Granger and Ron Weasley. Afterwards he is upset. There are no more books. He decides to write a continuation of the books about what happened to the children of Harry and Ginny, Ron and Hermione. It is called "fan fiction".

He is engaged. Coming home from school, goes up to his room and writes. Together we correct the texts.

We engage his little brother to make drawings.

Father is the main text proof reader. Mother is given the role as publisher consultant and editor. She checks whether the texts are consistent. She checks that the texts have facts, relationships with people, and that the stories are in a logical development.



Illustration to one of Matias' stories

Second example text:

In the streets of London, a sailor walks on the cobblestones. He walks towards the hiring office.

«I have always enjoyed cemeteries. For some, death is an end, for me it's a beginning. I wanted a job. It was given to another person. He is going to be killed.»

Chapter 1. Outside the Sea

«In this destiny, I write Captain Landen Snow, my ship Murovania is sailing to Cuba and the sugar plantations.» Unexpectedly somebody knocked on the door. «Come in!» Said Landen with a tired voice. Through the door of the door came a tall man with a sailor hat, an oilskin mackintosh and long boots. «Captain, I have a message for you. We have sailed straight into a shear. We could not stop when we heard the foghorn.»

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Third example text:

Fairytale three frogs

Once upon a time there were three frogs. They lived in a swamp. They had a good time there. A troll too lived in the swamp. The troll was a moody lazy toad, ugly as a tree and grumpy as an old woman. The frogs were in fact three princes who had been bewitched after they had eaten fungi. The only thing which could make them princes again was an egg cracked by a dragon high up in a large mountain far up in the sky.

Ashlad from the Trysil wood in Alvdal came to search for a girlfriend. He met the three frogs. They told him that they had been bewitched and that the only thing which could save them, was an egg cracked by a dragon. Ashlad went to search for the dragon. After a while he met an old man. The man asked whether he could please have some food. I have not eaten in a hundred days. Yes, but I only have an oatcake, and a drop of flat beer.

Status today

Matias is completely blind. He will have his 23-year birthday in the summer. He walks with his stick and a companion. He is hard to understand when he talks.

Matias writes a lexicon about Harry Potter from A to Z. The letters A to D he wrote himself. Now mom is a consultant. Together they go online to find facts. They have an intimate gathering. Now the mother does the typing. Matias participates with his knowledge. It is nice for Mom and son, the moments when they write together.

Grandmother is also engaged to correct texts with short stories he has written. When Matias visits grandmother, they read the text together. This makes



Self-portrait

Portrait of a man in the future

communication much easier. The time they have together means a lot to grandmother. She likes that they are engaged in something he makes.

Why strive to learn?

Matias learned to write braille by slowly developing in a safe environment, spending time exercising and practicing. Learning goals that his father thought would be achieved in two to three weeks took maybe half a year. However, as the time went by, learning became fast, the texts became better and longer. I have been his supporter from the start. After a while, I just needed to be nearby, outside his writing desk. He managed the rest himself.

As told, learning is a way to participate in society. Our goal is not a qualification for a profession. Learning gives the feeling of mastering something. It is important for participation to be able to share things with others.

For Matias, it has meant a lot to be able to express himself in writing. He has been able to express his creative abilities for a long time. He has a feeling of mastering; he has become a happy and harmonious boy. For a long time it was both reading and writing. Now he is most keen on writing the stories.

It has helped us communicating with him now that his speech is harder to understand.

Giving priority to literacy

Literacy should have high priority for all children, also for children with JNCL. For children with JNCL early intervention is important for literacy development. Teaching of braille for children with JNCL should start early (see Chapter 12).

Precautionary learning

At the time of diagnosis, many children with JNCL are still able to read print and have no immediate need for braille. Teachers should all the same start braille training to make the best possible foundation for future literacy. Otherwise, cognitive decline may make learning braille very difficult or even impossible. Braille teaching should start early to prepare for future needs.

Hastened learning

Reading and writing are complex skills and should therefore be given high priority when the diagnosis of JNCL has been confirmed. With age, learning will proceed at a gradually slower pace. To utilize the child's best learning capacity, there should be formal and informal teaching situations every day and acquired literacy skills should be useful and functional elements in the student's everyday life.

The learning environment

Some causes of poor literacy skills might be found in the educational systems. It is necessary to evaluate whether the services and the individual adaptations are sufficient, or if more should be done. It may be easy to attribute all lack of progress to the student or the disease. One should rather ask if more could be done to compensate for the loss of sight and cognitive decline, and if the teaching strategies are suited to the child's needs. One should also ask if braille teaching is starting early enough, that is, before the onset of dementia or while dementia still is in a mild phase, if the teaching of literacy is given sufficient priority and if the student is provided with varied reading and writing opportunities every school day. Comments from a staff member demonstrate the need for asking about these issues:

Earlier he received teaching in braille. He did however not benefit from this teaching because braille was very difficult for him to learn. Maybe it was presented too late.

One of the questions in the present survey asked about the usefulness of different school subjects in situations outside or after school. Some of the parents' comments concerned reading and writing.

Writing/reading: When school ended, his vision and fine motor skills had deteriorated to a degree where he was no longer able to use them. All the same they were of value when he still had these abilities.

She remembered very well the things that were important to her well past the age of 20. (...) She mastered typing on a computer with a speech synthesizer and wrote even at age 22, but at that age she lost the ability to form words by writing. When I told her letters she was able to write them on her computer.

Writing, reading and music were important and made her life rich. She enjoyed writing stories and poems already in primary school. The poems (...) often came in periods when she was frustrated by her life situation. I collected and took good care of her poems. They were her products, something she had created.

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Physical Education and Activity for Individuals with Juvenile Neuronal Ceroid Lipofuscinosis

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otor and physical activities are important for promoting health for people of all ages, and regular physical activity is one of the most important Lactivities people can do for sustaining best possible health (Donnelly et al., 2016; Janssen & LeBlanc, 2010; World Health Organization, 2018). The course of juvenile neuronal ceroid lipofuscinosis (JNCL) involves motor impairments, which are likely to influence both somatic health and social participation. Persons with JNCL usually show problems with posture, gait and motor coordination resembling symptoms of Parkinson disease. Hypokinesia and rigidity of muscles and joints lead to difficulty performing routine activities, such as moving around, exploring the surroundings, walking to school or shopping. Similarly, fine motor skills may deteriorate, resulting in difficulty dressing, switching on a computer, packing a school bag, making a sandwich or tying shoelaces. The gross motor problems generally become noticeable around the age of 10-to-12 years, but there is significant variation (see Chapter 7). The aim of physical intervention is to prevent and postpone the problems, and intervention must be based on knowledge about motor disabilities in general and JNCL in particular (Jeremiassen, 2016). The present chapter discusses the importance to individuals with JNCL of physical activity in general, participation in activities involving physical actions, and physiotherapy and other interventions targeting physical engagement in activities of daily living.

Physical activity and JNCL

In their early years, most children with JNCL have similar interests and participate in the same activities as their peers and show normal physical development. By the age of six, they have learned many basic skills, such as walking, running and jumping. Similarly to other children, they view themselves in terms of their potential for action, such as being or becoming football players, swimmers, climbers, or skiers. The situation changes when the visual loss starts to affect their ability to move around freely. Participation in activities like football, tree climbing or running becomes difficult when vision is severely reduced or lost. Still, many individuals with JNCL maintain an interest in physical activities like sports, dancing and outdoor life into adolescence, even if the activity level has declined considerably. The visual impairment will however affect the child or young person's agility, speed and balance, and usually result in a more sedentary lifestyle characterized by less participation in activities involving motor skills. There is therefore a need for support and adaptation to compensate for physical inactivity caused by visual impairment (see Chapter 4 and 7). The decline in motor performance will aggravate the situation and make further development of motor skills more difficult. Stooped posture and poor balance are common in individuals with JNCL from adolescence, and many rely on wheelchairs for mobility from late teens.

In general, motor impairments and a sedentary lifestyle will have a negative influence on physical health, cognition and academic performance, whereas physical activity may actually delay cognitive decline and protect against mood disorders (see Chapter 7). Anxiety and depression are common in adolescence, and young people with JNCL are more vulnerable to these disorders than their peers (see Chapter 27). Establishing and maintaining a healthy physical lifestyle is an important element of interventions to support positive development, learning and well-being. However, the need for a healthy physical lifestyle in children, adolescents and young adults with JNCL seems to receive little attention from those who are responsible for planning and has been neglected in research.

Physical activities among individuals with JNCL

The parents in the present survey (Appendix A) were asked how often their children had been engaged in physical activities involving gross motor actions at different ages. Figure 15.1 shows a steep decline between age seven and age 23 in the percentage of participants engaged in daily physical activities, from 74 to 26 percent. The results suggest that engagement in daily physical activities are replaced by weekly physical activities. In addition, at age 23, quite a large percentage of participants were no longer engaged in any type of physical activity. Considering the potential positive impact of physical activity, the descending curve in Figure 15.1 appears worrying.

On might accept the reported decline in physical activities as a natural consequence of blindness, dementia and motor impairment, but this stance



13 yrs (N=83)

16 yrs (N=66)

23 yrs (N=23)

Figure 15.1 Reported frequency of physical activity at different age levels

10 yrs (N=101)

20 10 0

7 yrs (N=106)

might lead to a lack of initiative on the part of professionals. Instead one might try to identify and eliminate shortcomings in the environment by asking basic questions, such as what could be done to compensate for the lack of football participation when severe visual impairment makes playing impossible, or to compensate for difficulties in walking to school when gross motor problems become evident. Many barriers in the environment can be met by compensatory measures (see Chapter 16). Compensatory measures may include adaptation of current activities, introduction of new activities, use of technical aids, provision of support or adaptation of the physical surroundings.

The results of the present study show that physical inactivity is not inevitable for everyone. Some young adults with JNCL participate in one or more exercise programs daily, whereas some of their peers are not participating in any kind of physical exercise. Such differences may be a reflection of not only differences inherent among individuals with JNCL but also in their environment. The parents in the present study were asked to indicate who took the most responsibility for ensuring that their child with JNCL was engaged in physical activities: the person with JNCL (self-governed), the family (family-governed), the school or residence staff, or collaboration between the family, the school and the residence. Parents reported that, for seven-year olds, the responsibility was split between the children themselves (36%) and collaboration between school and family (48%). Self-governed activities were reduced to three percent at age 13. At this age, participation in physical activitives was mainly family-governed for 26 percent of the participants and a result of collaboration between the school and the family for 55 percent. At 16 years the pattern had changed again: upholding a physical
lifestyle had become family-governed for 59 percent and a collaboration with others for 14 percent. This trend was strengthened for participants who were 22 years old.

The results indicate that many individuals with JNCL were engaged in physical activities on their own initiative when they were younger. However, owing to the visual impairment and emerging cognitive and motor declines the children and young persons with JNCL became more and more dependent on others in order to be engaged in physical activities. The results emphasize the importance of the collaboration between the family and the school for maintaining the young person's participation in physical activities throughout the school years. However, they also indicate that the school could have a stronger role in ensuring a physically active lifestyle for older students with JNCL. In adulthood, physical activity depends more on the residential staff. The declining curve in Figure 15.1 may indicate a need for more support and adaptations with increasing age, to support maintenance of a best possible physical lifestyle.

Physical education and physiotherapy

The parents were further asked about the impact on daily life of the physical education their child had received at school. Figure 15.2 shows that about 77 percent of the parents found that the physical education had a high or very high impact on their child's daily life. Nearly 17 percent reported a moderate impact and about seven percent found little or no impact of the physical education.



Figure 15.2 The percentage of parents giving each score for the impact of physical education on daily living (N=103)



Figure 15.3 The percentage of parents giving each score for the impact of physiotherapy on daily living (N=109)

The parents were also asked if their child had been given physiotherapy and if the physiotherapy had had any positive impact on the child in daily life. Seventythree percent of the participants had received physiotherapy, eight percent had not received physiotherapy despite a need, and for 15 percent physiotherapy had not been considered relevant (Figure 15.3). For the participants who had received physiotherapy, eighty-five percent reported a high or very high impact on their child's daily life. Eleven percent found the impact moderate and about four percent that it had little or no impact.

Physical activity in school and physiotherapy were thus evaluated as having a positive impact on the daily life of the participants. However, only a minority of the children and young people with JNCL were provided with extra physical training and some parents reported that their child had been denied physical education at school because of the disabilities.

Observations or registrations of declines and disorders are often used as the criterion for the provision of services and resources. Physiotherapy was mainly introduced as a result of physical decline. The fact that only thirty-four percent of the 7- to 10-year-olds received physiotherapy suggests that gatekeepers of services might not have viewed physiotherapy as a preventive or proactive measure for a majority of the children with JNCL in this age group. Some parents reported that they had to fight the system to obtain services, as reflected in the following comment:

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Physiotherapy was finally provided, after many years of fighting, when our daughter lost the ability to walk.

Further, some parents reported that physiotherapy was terminated when the authorities or the physiotherapist could not see any evidence of positive effects of the physiotherapy:

We wanted more physiotherapy for our child, but the services were terminated when she was 18 years of age. They claimed it had no positive effect for our child

Policy barriers, such as dismissal from services when progress cannot be documented, reflect a model of service delivery that is not in keeping with the contemporary views on habilitation reflected in International Classification of Functioning, Disability and Health (Castro & Palikara, 2018). The implications of a failure to consider preservation of existing functionality and prevention of unnecessary decline are serious for people with chronic or progressive conditions.

Physiotherapy was sometimes provided at school and terminated after the young person with JNCL left school:

Physiotherapy and occupational therapy were stopped when our daughter left the educational system. Today she is 22 years old.

Moreover, the provision of physiotherapy seems to be related to how the effects are measured. Relating the provision of physiotherapy to observed developmental gains is not appropriate for individuals with JNCL. It is often difficult to measure effects of proactive and precautionary measures. The provision of physiotherapy and extra physical training may rather be related to aims of maintaining the physical condition, such as delaying or reducing ongoing decline or supporting engagement in activities in daily life. It may also be related to support of the general health and physical and mental well-being of the person. It is therefore unfortunate that these interventions are terminated when the individual is not showing the expected progress. Delaying a forthcoming deterioration is outside the traditional aim of interventions for children. There are thus urgent needs to build insights and knowledge about JNCL and similar diseases in organizations dealing with such services.

The positive effects of participating in physical activity were evident not only from the parents' evaluations in the present study, but also from the participants' own interest in such activities. The results indicate that physical activities were a main interest for most of the younger participants, an interest that sometimes lasted into adulthood. Ninety-six percent of the seven-year-olds were interested in physical activities and 51 percent of the 16-year-olds. Moreover, thirty-two percent had physical activities as a main interest at 22 years (note the difference between having an interest and actual doing the activity of interest). The interest seemed to decline over age for individuals with JNCL but this is also true for the general population (Sallis, Prochaska, & Taylor, 2000). Many persons without disabilities become less interested in physical activities with age and physical activities are replaced by other interests. However, in spite of the considerable difficulties related to their developmental decline, some of the participants in the present study maintained their interest in physical activities. The following quotation from a parent of a child with JNCL illustrates that participation in physical activities may have many perspectives:

Being involved in physical activities provides my son with a sense of mastery, pride and having a meaningful life.

In the present project, the most common interests involving gross motor activity for individuals with JNCL were riding, swimming, dancing, football and outdoor life. An active physical lifestyle may promote social interaction when such activities are shared with others.

Some factors related to physical activity in individuals with JNCL

Physical activities may be organized, such as doing sports or physical education, or attending a gym club, or non-organized, such as playing with friends, walking to school or climbing trees. The total amount of organized and non-organized physical activities is an index of the individual's level of physical activity (Van Duyn, McCrae, Wingrove, Henderson, Boyd, & Kagawa-Singer, 2007).

The health benefits of physical activities depend on the duration (the amount of time spent in one activity session), the intensity (rate of energy expenditure per time unit) and the frequency (the number of exercises per time unit) of the activity. The physical status is measured in different ways, including muscular strength, endurance and oxygen uptake. The endurance is dependent on the heart, lungs, blood flow and blood vessels (Tremblay, Colley, Saunders, Healy, & Owen, 2010).

The flexibility of movements and range of motion of joints also influence the physical status (Jeremiassen, 2016). The joint's flexibility or motion range is normally good in children but tends to decrease with age or when particular movements are not repeated frequently. The length of a muscle and tendons restricts the range of a motion, as stiffness in joints caused by other reasons. Severe forms of stiffness – joint contractures – may cause great pain and immobility. For persons with physical disabilities, it is particularly important to maintain flexibility in joints that are seldom used.

Many countries offer recommendations for physical activity and it is usually recommended that 5- to 17-year-olds should accumulate at least 60 minutes of moderate-to-vigorous physical activity daily (Janssen & LeBlanc, 2010). More than 60 minutes of physical activity may provide additional health benefits. The World Health Organization (2010) recommends that most of the daily physical activity should be aerobic exercises that make one perspire and breathe harder, which elevate the heart rate, resulting in quicker and more efficient delivery of oxygen throughout the body. Aerobic exercises include walking, swimming, running, skiing or dancing for at least ten minutes at a time. Maintaining a physically healthy lifestyle requires paying attention to the duration, intensity and frequency of exercise activities. The overall principle behind physical fitness is simple: the body becomes a mirror of the lifestyle and motor capacity can only be built through motor activity.

Motivation and interest for participating in physical activities are often established at an early age and may have long-lasting effects. Education and in particular physical education may contribute to establishing such interests in young children, but it is important that the activities are associated with excitement and having fun (Wuest & Bucher, 1999). The results presented above show that individuals with JNCL can sustain interest and engagement in physical activities, even when impairments become more severe. It is recommended that the school, in close collaboration with parents, takes the initiative as early as possible to promote interest in physical activities in children and young people with JNCL. Many exciting activities are accessible even after the onset of a severe visual impairment. Judo, swimming, goal ball, outdoor activities, gymnastics, wrestling, dancing, cross country skiing, tandem biking are activities that do not require vision or much orientation (see chapter 16).

Physical activities for individuals with JNCL

Motor skills vary among individuals with JNCL. Some of the motor skills learned and automatized in early life are maintained past adolescence and into adulthood. Some persons with JNCL have maintained motor skills by practicing advanced activities, such as downhill skiing, skating, fishing and hunting into emergent adulthood, despite their visual, motor and cognitive impairments. Maintaining



Participation in active outdoor life may continue into adult age.

participation in such activities may depend on personal motivation but also on sufficient adaptation and support (see Chapters 16 and 23). To create an early foundation for an active physical lifestyle is a core issue in habilitation for children with JNCL.

Children and young people with JNCL may need support to develop and maintain fine and gross motor skills and to develop a healthy lifestyle. This area should be a core element in the adapted or special curriculum and the Individual Educational Plan (see Chapter 11). The Educational Development Observation Tool (see Chapter 10) includes observations of motor performance in various situations. The results of the assessment may indicate a need for a physical activity bank where activities are listed and timetabled (see Chapter 23). It is an overall aim that the activities should take place in social and inclusive settings, at school and at home or in the residence. Early motor interventions may have positive lifelong consequences for the individual (Jeremiassen, 2016; Wuest & Bucher, 1999).

An overview of physical activities that children and young people with typical development are involved in, in terms of content, extent and frequency, may constitute a basis for planning of physical activities for individuals with JNCL. Such an overview might show that peers usually participate in soccer training twice a week, physical education at school three times a week and scout activities

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Saturday. They might every walk or bike to school on a daily basis, visit the playground every second day, go shopping once a week and help the family with gardening every weekend. Such activities constitute the physical lifestyle or baseline for children in general and they contribute to the children's motor development. Not all of these activities are suited for children with INCL, but they represent a way to explore activities that may be selected for further consideration. For instance, if children with a typical developmental course walk or run 50 kilometers per week and same-



Exploring objects with the fingers.



Constructing with Lego.

age children with JNCL move five kilometers per week, this may indicate a need for intervention. The concern is to compensate for loss of activities caused by the impairments and to provide support and activities. This will require adaptation as well as organizational measures, for example extra hours in physical education or support to enable active participation in the local swimming, judo or riding club. Walking to school or doing chores (e.g., fetching the milk or disposing of the garbage) and other routine activities at school and home will usually require planning and organization.

Fine motor skills are important for children with JNCL. For example, such skills are needed in reading and writing braille or Moon, exploring objects and tying the shoelaces. The hands and fingers become the individual's "eyes" when vision is lost. Examples of fine motor activities are construction play with Lego, doll play, playing cards (braille marked when needed), and everyday activities like cooking and dressing.

Playing the piano or other instruments are also good exercises for developing fine motor skills. Shooting may coordinate fine motor skills and sound location. Children and young people with JNCL may engage in such activities at a frequent and regular basis (Jeremiassen, 2016).



Shooting with the help of audio signals.

Movement restrictions and stretching

Stretching means that muscles are deliberately flexed or stretched in order to maintain or improve the range of motion. It can be performed individually or together with a partner. Stretching will increase the elasticity of the joints and improve the muscle tone, which may help maintain posture and reduce motor decline (O'Sullivan & Portney, 2014). The introduction of stretching exercises is recommended for young children with JNCL to avoid slow developing or non-observable decline in range of motion. Stretching exercises for hips, knees, wrist and shoulders should be prioritized.

Jeremiassen (2016) emphasizes the need for regular assessment of joints and muscles from an early age in children with JNCL. The main aim of the assessment is to register possible ongoing restrictions and limitations in movements. She describes three levels of movement restrictions: (1) full movements (no restrictions), (2) moderate movement restrictions and (3) severe movement restrictions. Restrictions can be caused by motor impairment but may also result from a sedentary lifestyle where joints and muscles are not used on a frequent basis. Most movements restrictions are a result of both, and there can be evidence of a downward spiral where a lower activity level contributes to decreased functionality, which in turn further limits activity. Planning is essential to avoid or reduce this spiral.

Inactivity or inability to use certain movements and joints may result in shortened muscle fibers, increased reflex activities and spasticity of the muscles involved. A severe condition may inflict pain and have other unpleasant consequences for the individual. It is important to be vigilant when evaluating hip and knee movements, since restrictions on these joints may result in problems with walking, caused by an inward rotation of legs or other joint anomalies. Beginning at early age, there should be regular examinations for the following joint functions: (a) elbows: degree of flexion, tension, and inward/outward rotation; (b) wrists: degree of flexion and tension; (c) fingers: extensions; (d) shoulders: ability to elevate and rotate arm; (e) knees: flexion and tension; (f) hips: stretching and inward/outward hip rotation; and (g) ankles: flexion and stretching the ankles. Movement restrictions on Level 2 (moderate movement restriction) and Level 3 (severe movement restriction) should be met by appropriate measures to maintain or improve the flexibility in joints and muscles. Measures may include exercises requiring active and passive movements. Difficulty achieving a full range of motion can be observed at a relatively early stage of the JNCL disease, especially in weight-bearing joints, such as hips, knees and ankles. Activities that will preserve optimal range of motion and mobility are therefore important in early stages of the disease, including activities requiring passive movements, and participation in all-round activities that require all the muscle power available.

The problem with balance and body posture is partly caused by restrictions in the joints and muscles (Jeremiassen, 2016). The joints become stiffer with aging and it may only be possible to execute movements at a slower pace. Preventive interventions should be introduced to avoid an accelerated development of stiffened joints and movements and maintain the best possible posture. Severe forms of stiffened joints and muscles will entail difficulties in undertaking certain body positions, such as stretching out when resting, often combined with pain and discomfort for the individual. A physiotherapist or JNCL counselor should be contacted when such problems are observed. In the present project (Appendix A), several parents and professionals commented that participants with JNCL had been experiencing pain when they went to bed because they were unable to stretch their legs in a lying position. Such problems can to a large extent be avoided with appropriate interventions.

It is important to observe the joints and muscles that are used for posture, walking and movements of the arms and hands. Joints and muscles in the neck, back, hip and knees are important for maintaining posture and walking. The joints and muscles in the arms, hands and fingers are important for daily activities like dressing, reading braille or playing games. Any signs of limitations in these joints and muscles should therefore be met with interventions. The joints should be flexed and stretched and the muscles strengthened or maintained on a frequent basis.

Active movements

Active movements are executed by the person and may have positive effects on the exercised joints and muscles and contribute to maintaining joint mobility and muscle strength. Exercises with active movements can be a part of a training program and executed independently or with support from helpers. The exercises will not improve the movement range or prolong the muscles or tendons, but they can be used to maintain the current movement range.

Active movement exercises can be organized as ordinary body-building programs in a gym or in the classroom or living room. Both the exercises and the results should be registered, documenting both positive changes and challenges. Records that show improvement may increase motivation both in the person and the involved staff. However, a declining curve should never be used as a reason for stopping the exercises, but instead for adapting them. Decline is inherent in the disease, so the aim of the exercises it to slow down the decline as much as possible.



Active movements.

Passive movements

Some movements may be difficult to execute for individuals with JNCL, particularly in the later stages of the disease. Contractures will result from a failure to exercise joints and the corresponding muscles (see Chapter 7). The joints may become immovable if not met by appropriate preventative measures, including a regime of passive range of motion exercises (Jeremiassen, 2016). Passive range of movements implies that a helper is performing the movements of the joints and muscles while the person remains passive. Passive movements can contribute to maintaining and extending the movement range in a joint and are especially important when a joint has become stiffened due to inactivity, such as when a person with JNCL is unable to exercise the joint and muscles. However, passive movements should be made by trained professionals or appropriately trained family members or others. since they may entail pain and discomfort.

Exercises with passive motions can be used with most joints (e.g., the wrist, elbow, ankle, fingers or hip). They should be performed in pleasant settings, such as when the person is listening to music or an audio book, and should be organized as a routine, for example twice a day or as needed. The aim of passive exercises is to prevent both the restrictions on movements or improve mobility, and the pain