Passive movements of the hip.

that may be caused by an immovable joint. The exercises can be conducted by teachers, parents or assistants with guidance from a physiotherapist or another competent professional. The program should be described in writing and if possible with illustrations, and should also include how long time each exercise should take.

Exercising cardiovascular endurance

The motor impairments related to JNCL may make aerobic training difficult when the person is no longer able to walk or stand. However, aerobic capacity is of great importance and should never be forgotten when planning interventions for individuals with JNCL. The functions of the arms and hands are usually maintained longer than functions supporting walking, posture and balance (see Chapter 7). Exercising the upper body may therefore replace activities requiring the aforementioned skills. For example, a rowing machine is found in most gyms and allows exercise of the upper body without making demands on balance, and the motion resistance can be adapted to the individual's capacity. There are sledges with skies or wheels, and they can be used both in winter and summer. The sledge does not make demands on balance and is moved forward with arms and poles (different poles for winter and summer).

Driving a manual wheelchair may also provide aerobic training. There are special wheelchairs for aerobic exercises, which are lighter than ordinary wheelchairs and can be adapted to individual needs. The flow rate or load in these exercises can be increased by practicing uphill or on soft ground.

Exercises in the water are also a good way of promoting cardiovascular endurance when balance problems make other exercises difficult. The water provides buoyancy and support for balance but the resistance in water also makes exercises and motions a bit harder to perform, thereby making the cardiovascular endurance training more effective. Some parents and staff in the present project commented about participants with JNCL who were able to swim after their ability to walk had been reduced or lost, indicating that exercises in the water may be a good option for them. It is recommended that aerobic training is scheduled four to five times per week, or even more often when possible.

Two stories

This part presents two stories about activities and physical education and intervention. The first is told by a father, and the second by a physiotherapist who started to offer physiotherapy to a young man with JNCL aged 20 years.

The father: Keeping in shape and having fun

This is a story about "Harry", a young man of 28 years. Harry was always active and became interested in sports when he was quite young. For instance, snowboarding was learned at a time when he still could see. His body never forgot it and he was able to practice snowboarding until he was 27 years old. One of his helpers at a winter camp recently related that he met Harry for the first time when he was 12 years old. At that time he was able to snowboard downhill on quite demanding trails a number of times in a row together with an assistant and with a slalom pole between them for steering purposes. At the age of 27, Harry could move five meters with the snowboard with the help of three assistants. Still, it was very clear to a proud young man that he had been snowboarding that day.

In addition to snowboarding, Harry was also a proficient cross-country skier. As long as the tracks are well made and clear he could continue to ski for almost as long as walking was possible.

Harry was also able to follow a normal judo curriculum for seven or eight years but had to give up this sport when he was around 20 years old. Before finishing his judo career, he was awarded a blue belt. Judo is the preferred martial sport for many people who are blind, because body contact to a large extent compensates for the lack of vision.

Swimming has always been a favorite of Harry's, and he is still swimming today. In his younger days, he agreed to work towards a specific goal and he would be awarded a merit badge if he was able to meet the ordinary requirements for obtaining the badge. He had to do 1200 meters breaststroke and 600 meters on his back, float 300 meters, dive to the depth of four meters, swim under water 15 meters, undress in the water and show proficiency in life saving techniques. In addition, he was required to dive from a height of three meters (the toughest challenge). Through enthusiasm and stamina, Harry was able to accomplish all of this over a period of two years. A proud young man at the end of the day!

Wall climbing is another sport that over the years proved to be a good activity for Harry. He participated in this sport on a regular basis until he was 26 years old, and still occasionally climbs when he has an opportunity. Wall climbing can be adapted to the individual, made hard or comparatively easy. Getting to the top of a wall and ringing the bell is satisfying, also for those who watch!

Many individuals with JNCL enjoy horse riding. Harry started riding at the age of eight or nine and still rides once a week. Balance has become a challenge, and his saddle has been modified to accommodate balancing support. Horse riding should now avoid going downhill and he uses the same horse always. Harry and his horse make a fine team!

Finally – Harry enjoys a number of other physical activities. At this stage in his life he is dependent on suitable equipment and good helpers. To assist him in his endeavors, he has a tandem bike with a small motor to help climb the steepest hills. He also has a toboggan or sledge which can be modified to accommodate skis, skates and wheels.

The physiotherapist: Growing with training

The young man was 25 years old when this text was written. He lived in a small rural community offering a lot of activities and physical training. He was involved in activities like hunting, fishing and sports, and participated in parties and fun. He had a good network of friends sharing his interests since he was a young boy. Today, he has his own "company" that is cutting, packing and selling firewood. The company is supported by his parents with participation of friends from the neighborhood. The small rural community has always supported the young man's schooling in the best possible way, and he had excellent teachers from the early school years on.

The young man received the JNCL diagnosis when he was seven years old and he became blind in a very short period. The collaboration between the young man, his teachers and the family always worked well. Today he is living in a staffed apartment. He has had a "normal" life where he has been exposed to similar challenges as his peers in a safe environment, supported by friends, engaged staff and family.

Physical activities and a healthy lifestyle were given priority in the young man's life from the very beginning. However, he found physical exercises rather boring after he became an adult. Sessions of planned physical activities were often dropped in favor of more pleasurable sedentary activities after he left school.

The role of the consulting physiotherapist (here: counselor) was to guide the local staff and a local physiotherapist. Plans were made to change the young man's sedentary lifestyle in close collaboration with himself, the local staff and his family. The overall aim was to improve his physical status and specify appropriate exercises. This process started up with a local workshop in collaboration with the project. Different themes were elaborated and reviewed in this workshop and physical activity and training were given priority together with a few other themes. The staff was taught about the importance of physical fitness for individuals with JNCL, what activities to focus on and how physical training could be integrated into the young man's life.

It was decided that physical training should be a part of the young man's working day. Physical training should also be an important life content during afternoons and weekends. Training programs and corresponding schedules were specified:

- *Hard training three times per week during working hours in a gym close to the young man's workplace.*
- *At least 30 minutes of physical activity every day beyond working hours with low to middle pace of aerobic activities, in accordance with recommendations from the health authorities.*

There were follow-up meetings with the counselor, the staff and the physiotherapist every six months. A tool for fitness review and evaluation was developed, which included measures of muscle strength and movement ratio in joints. The results were used to adapt the young man's exercises to his current situation. New baselines were set every sixth month on the basis of an evaluation of his walking ability.

There was little or no research to be found on JNCL and physical exercise. The counselor borrowed ideas from studies of epilepsy and exercise, dementia and exercise, and from her own studies on cardiovascular exercise (endurance training) and its effect on muscle strength and movability of joints. It is a common myth that physical activity may trigger epilepsy (Brna, Gordon, Woolridge, Dooley, & Wood, 2017) but research shows the opposite: physical activity may actually function as an anti-seizure measure (Häfele, Freitas, da Silva, & Rombaldi, 2017; Pimentel, Tojal, & Morgado, 2015). Research also shows that physical activities may have positive preventive effects on cognition and well-being in elderly people with dementia (see Chapter 5). However, the health effects of physical fitness training are dependent on frequency (how often there are sessions), duration (time length of the sessions) and intensity *(how hard the sessions are). In other words, training has to be carried out frequently, have a minimum duration per day and be executed with some intensity.*

Good routines do not make themselves

Good routines do not make themselves; they must be implemented over time. It is a continuous process where needs are addressed by new staff members and in accordance with the progress of the disease. A training card was developed where the young man, the staff, the family and the counselor could monitor the exercises and the physical status of the young man. The training card became a highly motivating tool for all involved partners. It described his current physical activities, the number of series to be performed, the weight or load to be used in different exercises, and so forth.

Physical training became a good routine in the young man's life. He attended all the training sessions with joy. From having lukewarm feelings for physical exercise, he became very devoted to physical training. Physical exercise became a natural part of his life and today he is in a better physical shape than at the beginning of the program five years ago.

The importance of engagement

The local physiotherapist had over time acquired experience and knowledge about the diagnosis. He was very engaged and his role was to support staff, inspire them, and be available when needed. He was impressed with the staff and reported that training took place in an inclusive setting where the staff was not acting as instructors but as training fellows. The staff and the young man are doing similar activities and this has had a positive effect on the young man's motivation.

Why all this training?

The main aim of training is to delay the emergence of motor problems caused by the disease, such as reduced mobility, forward bending posture, arthritis (with joint stiffness and pain) and shortened muscles. The person-centered question was how the future challenges of the young man could be met in the best possible way. One important goal was to develop and maintain walking. The training built on the assumption that strong muscles, good flexibility in joints and best possible fitness would create reserves to delay future declines in motor skills.

The tool for fitness review and evaluation was used to measure the young man's muscle strength and search for possible problems with arthritis and shortened muscles. A six-minute walking test was included to measure the aerobic capacity. Altogether, the information collected with the instrument provided a comprehensive picture of the young man's physical fitness and possible changes from last measurement, and constituted a basis for evaluating the need for adaptations of existing exercises or for new exercises.

Achievements

The main aims of the physical training were increased aerobic fitness to support the young man's working capacity to meet daily demands and strengthened muscles and flexibility in the joints to support his walking and prevent falls.

- *Aerobic training was performed three times per week with three sessions each day.*
- *Session 1: 4 x 3 minutes running on a treadmill (4 series of 3 minutes) with a two-minute active break (resting by walking). Speed 7 km/h on a treadmill with a climb of six percent.*
- *Session 2: 2 x 6 minutes running on a treadmill with two minutes active break (resting by walking). Speed 6.5–7 km/h on a treadmill with a climb of four percent.*
- *Session 3: 10 minutes running on a treadmill. Speed 11 km/h on a treadmill with a climb of one percent.*

The young man's speed (frequency of steps) and distance have increased during the last eight-month period. For instance, the speed has increased from six to seven km/h in Session 1, and the distance has increased by 50 meters per interval. The young man is therefore able to run 200 meters longer than before in one session with four intervals. The running speed was 10 km/h in Session 3; today the running speed is 11 km/h. The walking test also confirms an improvement in fitness. Today, the young man is able to walk 820 meters in six minutes, compared to 580 meters eight months ago – in spite of the inherent declines of the disease.

The main focus of the muscle training has been quality rather than weight. Too heavy weights would have had a negative influence on the quality of performance. The quality of all exercises has increased and today the young man is working with heavier weights than before.

The young man had an increased inward rotation of the legs, feet and hips when sitting, standing and walking, which is a common problem in individuals with JNCL. This was a possible sign of weakening muscles, which was met by exercising the outward rotating muscles. The young man started with a 20 kilo load, today he is able to work with 35 kilos, and he has progressed from three times ten repetitions to six times ten repetitions. The quality and coordination of the performance involving both legs simultaneously is also a lot better today. Measures of joints and muscles show an improvement on the left side over the last eight months, and also a stabilization of the decline in the right side.

Full stretch and movements in hip, knees and ankles are important for maintaining walking. The young man has bending tendencies in the knee and hip and has therefore made exercises to strengthen the muscles attached to the knees and hips.

In the beginning he was not able to do three times five repetitions with a ten-kilo load when exercising the hip stretching muscles. Today, he does three times ten repetitions with a ten-kilo load and executes the exercises with better quality. Similarly, he was not able to do three times ten repetitions with a ten-kilo load when exercising knee stretching muscles. Today, he is doing three times ten repetitions with a 15-kilo load with better quality. It took only two to three months of training before improvements became noticeable. Running on the treadmill and walking tours in afternoons and weekends were supporting factors on this positive development. The young man's posture has improved as a result of these activities, in combination with exercises for strengthening the muscles in neck, back, shoulders and arms.

Physical exercises have become routine and are very positive activities for the young man. He is very proud of his improvements and of being strong. He has a very competitive attitude, he likes to use heavy loads and test his limits. Today, the young man is requesting training on weekends when he is normally free from training. Physical training has become a meaningful, inspiring and social activity for him.

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16

Coping with Everyday Life

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Marja-Leena Forssas and Tarja Eskonen

After a child has been diagnosed with JNCL and the illness progresses, people surrounding the child may increasingly do things on the child's behalf because they want to protect the child from potentially dangerous situations or to get things done faster than the child can do unassisted. In addition, they may underestimate the child's self-help skills and other abilities. This may over time lead to excessive dependence on help from adults, potentially resulting in learned helplessness and passivity. An alternative approach is to support autonomy and independence when skills decline. Baltes and Baltes' (1990) model of goal selection, optimization and compensation (see Chapter 2) provides a useful framework for the planning process. Independence, with or without assistance and support, is a basis for self-confidence and self-efficacy in children and adolescents and for coping with daily life (Aro, Siiskonen, Niemelä, Peltonen, Stenroos, & Kulmala, 2007; von Tetzchner & Martinsen, 2000).

The progression of vision loss in JNCL can be rapid and the consequences dramatic. There may be a loss of activities, friends, participation and independence. Blindness will restrict the possibilities of acquiring information from the environment. Vision is holistic and immediate, just a short glance may give an overview of what is happening, of the topography and the on-going activities in the near environment. Vision is a core sense behind independence and participation. Additional declines in cognition, communication or motor performances will indeed make independence and participation more complicated, and it is important to meet these challenges with targeted measures to achieve best possible participation and coping in daily life. Compensation for a visual impairment is a matter of learning new behaviors supplemented by making physical adaptations in the environment. The child must learn how to use touch and hearing in a different way and to accumulate and memorize received information to build up a holistic understanding of the environment to facilitate independence and interdependence (see below). Further, compensation is also about making participation in meaningful activities available for persons with JNCL.

Physical adaptation for people with severe visual impairment is about making features of the environment available through hearing and touch. For instance, a green area on the wall is not available through touch or hearing but giving this area a tactile texture will make the area perceptible for persons with blindness. A door that is not noticeable from a distance for a person who is blind, can be rendered perceptible with the addition of an audible signal.

Increasing students' determination, autonomy and independence in coping with life towards adulthood is an overall aim of the school, implying that young people will become able to accomplish daily tasks without help from others. These are indeed important goals in education, but goals of independence will sooner or later entail challenges and difficulties for individuals with JNCL. However, coping with everyday life has a wider perspective than independence, and this is of special significance for persons with JNCL.

Independence and interdependence

The core of the wider perspective on coping is acting as a *driver in one's own life* and not *being a passenger*. Being a driver is an essential element of self-determination and quality of life. There are many ways to show self-determination, such as when children choose to break up with friends outside of school time, refuse to do what they are told to do, and play on their own. These actions may be viewed as declarations of being a driver in their own life. When children do not want to be a part of a group activity, the reason may be a lack of interest or insufficient skills to participate. Alternatively, they may not enjoy the social setting or just want to demonstrate their self-image or role. Such behaviors may also be a way to try to obtain recognition. Protests and refusals may be perceived as annoying by others but are of importance for the personal development of all children.

Experiencing being in the driver's role in their own life is of special importance for children and young people with JNCL. For them it may be a source of motivation for further learning when they experience that it is possible to act as a driver despite reduced independence. Being a driver in one's own life requires self-governed attention and action, which is also a foundation of being active in learning and development. Doing things that are determined by someone else on your behalf does not require similar processes. For instance, it is more effective to learn the route between two places if one is driving the car, compared with being a passive passenger. The concept of being a driver also includes making one's own choices in life. Choice making is likely to increase attention and mental effort even if physical independence is not possible.

Typical development proceeds from *interdependence*, doing things together, to *independence*, doing things by oneself (Greenfield, 1994). In the theory of Vygotsky (1962), interdependence is a characteristic of the zone of proximal development, and there may be various degrees of independence and interdependence in different domains (von Tetzchner, 2019). When a person's developmental path involves decline, the process may go the other way, from independence to interdependence, and interdependence is a characteristic of the zone of developmental maintenance (see Chapter 2).

Independence and interdependence are two basic forms of active participation and coping. From the perspective of being a driver and coping with everyday life, independence means achieving personal goals *without support*, while interdependence means achieving personal goals *with support*. In interdependence, there is mutual reliance between two or more persons who are dependent on each other to achieve the chosen goals. It may be a transactional chain of interactions, where one person's actions trigger actions in the other person, and so on. An interdependent relationship will sooner or later be necessary for persons with JNCL to maintain the driver role in their lives, albeit one that resembles a learner-driver. For people caring for a person with JNCL, interdependence requires a strong and persistent focus on autonomy where they give support but *enable* the person with JNCL to be the driver. Carers may need support in learning to play their part in this new relationship based on interdependence.

Autonomy may be defined as a person's ability to make his own decisions, the ability to act on his own behalf without interference from others; it is about self-determination, not self-reliance (Entwistle, Carter, Cribb, & McCaffery, 2010). Autonomy is recognized as important for achieving good mental health (Taylor, 2005). Intervention that supports autonomy may enhance overall adjustment and life quality. The possibility to behave and act autonomously may make people feel and perform better, while lack of autonomy may lead to loss of interest and motivation, as well as sickness and depression. Development of autonomy may not always be an explicit goal in special education, but it is an overall goal for persons with special needs and in particular for persons with dementia (Agich, 2003). Supporting and maintaining autonomy in persons with JNCL require dignity, respect, understanding and time from people in the surroundings when collaborating.

Their early typical development implies that children with JNCL like their peers are acting as drivers in their own lives. They interact with their peers on an equal footing, display the same motor skills, and participate in ordinary childhood activities. The way from independence to dependence in developmental decline may involve several phases, involving compensations, help and support, and change of goals (Baltes, 1997). In earlier phases of decline, technical aids and

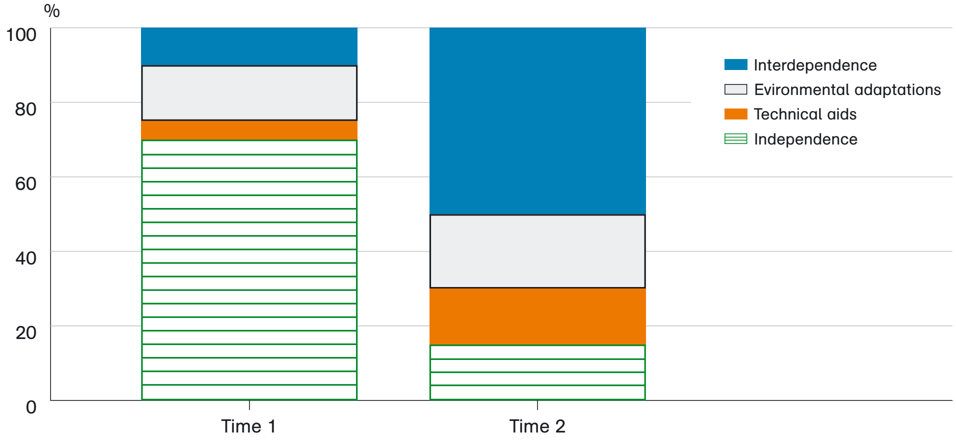


Figure 16.1 Hypothetical development from independence to interdependence, resulting in full participation

environmental adaptation may be enough to maintain everyday functions. Figure 16.1 shows the relative amount of independent functioning, compensation, and help at two points in time. At time 1, the participation of an adolescent boy with JNCL is mainly independent, although with short periods with compensation and help. At time 2, the same adolescent is more affected by the disease. However, he is still participating 100 percent of the time but his participation is dependent on more diverse compensatory measures and especially on collaboration with others through interdependence. Despite the decline, participation is achieved successfully with the help of educational, environmental and other interventions.

In this chapter, the concept of coping is associated with participation in everyday activities. Participation in activities will require the accomplishment of a series of tasks through independence when this is possible and desired, or interdependence when support is required and desired. Allowing for independence and interdependence according to the needs will enhance personal autonomy and coping with everyday life.

Education for coping with everyday life

Orientation, mobility and skills related to activities of daily living are important elements in special needs education for students with visual and cognitive impairments and for achieving independence and interdependence. Goals related to interdependence and autonomy typically become important in education when academic and practical performances are difficult to achieve due to the progressive visual, cognitive and motor declines in children and young people with JNCL. The use of technical aids is also an important element that contributes to the coping process (see Chapter 19).

Orientation and mobility

Severe visual impairment influences orientation and locomotion. Moving around independently requires cognitive resources and is one of the greatest challenges for persons with severe visual impairment, and orientation and mobility are always core elements in the education of children with severe visual impairment (Martinsen, Tellevik, Elmerskog, & Storliløkken, 2007). Understanding the physical environment is essential for all children. Limitations in orientation will influence independence at home and in unfamiliar areas. Teaching of orientation and mobility is therefore crucial for transitions and the child's understanding and knowledge of the respective environments (McConachie, 2018).

There are different definitions of orientation and mobility. Lowenfeldt (1954) defines orientation as the ability to recognize the surroundings and their temporal and spatial relations, and mobility as self-propelled locomotion from one place to the other. Törrönen and Onnela (1999) define orientation as the ability to determine one's position in an environment based on awareness of the body and environment through use of the remaining senses. They claim that the bases of mobility include perception of body and space, control of posture and movements, balance, and basic skills of moving around. Elmerskog, Martinsen, Storliløkken

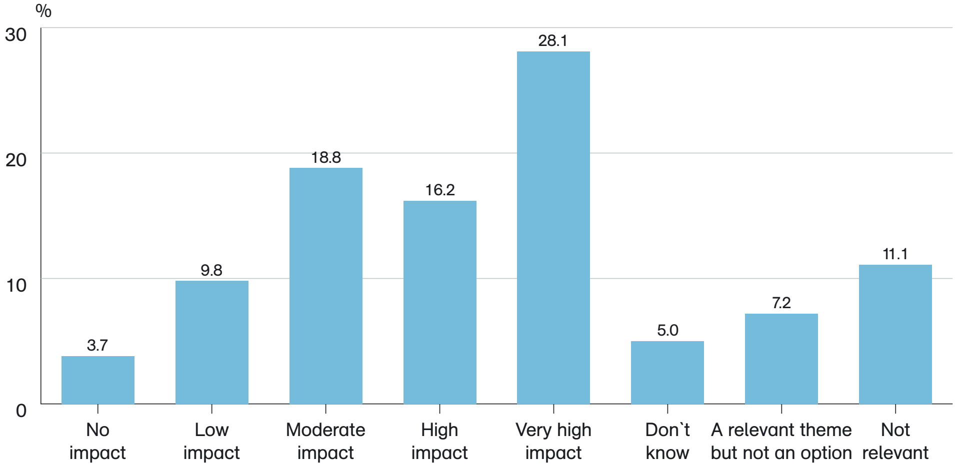


Figure 16.2 Parents' evaluation of impact of orienting and mobility training (percent of answers) (N=109)

and Tellevik (1993) define orientation and mobility as goal-directed activities in relation to objects in space.

Orientation and mobility education may include mobility route learning, use of sighted guides, searching skills and protective techniques, efficient use of the white cane and physical adaptation of the environment.

Parents in the present JNCL project (Appendix A) were asked whether their child had received orientation and mobility training, and if the training had any positive impact on the child's daily life. Of the 109 parents who answered this question, 76.7 percent answered that their child had received this kind of training, 11.1 percent said it had not been relevant, and 7.2 percent had not received training even if it was needed. Five percent said they did not know, but it is not clear if «Don't know» meant that the parents did not know if the child had this form of training or were uncertain if the orientation and mobility training had had any positive impact. Just over 44 percent said that the orientation and mobility training had a high or very high impact on the child's daily life, while 19 percent reported a moderate impact. Nearly 14 percent found that the orientation and mobility training had little or no positive impact on their child. The low impact may indicate that vision was still good or that the training had little effect. The mobility and orientation training was introduced at a mean age of 9.7 years.

There are some technical mobility aids designed for use with children and adults with multiple disabilities (Lancioni, Sigafoos, O'Reilly, & Singh, 2013; Mulloy, Gevarter, Hopkins, Sutherland, & Ramdoss, 2014; Zhou, Parker, Smith, & Griffin-Shirley, 2011), but research on the use of such aids by children and young people with JNCL is lacking.

Activities of daily living

Activities of daily living are generally considered as tasks requiring independence in coping with self-care, home and routine activities that most people do every day without assistance, such as using the toilet, grooming, dressing, feeding and community skills like shopping, home making, and so forth (Giebel, Sutcliffe, & Challis, 2015; Prizer & Zimmerman 2018). A clear definition of activities of daily living training is lacking, but training in such activities is often a core element of special needs education. Trainers and support workers are responsible for adapting the environment to the person's needs. The aim of the training is to facilitate an optimal level of independence in daily life. The level of training and adaptation required depends on the student's needs in coping with daily life. The traditional activities of daily living are in this chapter described as "tasks". Such tasks should be part of a meaningful, real-life activity, be functional and have personal value.

For example, activities like having dinner, playing soccer or visiting friends are meaningful for most people. Each of these activities may comprise several tasks: paying for a ticket is a task within activities like «watching a soccer game» or «travelling on a bus», adding salt is a task in the activity «preparing porridge», and using an electronic communication device with a switch is a task within the activity «socializing with friends.»

Real-life activities will be part of adapted curricula when they are related to the daily life of children and young people with JNCL. It is always a risk that interventions not related to real-life activities reflect wasted effort and time, such as practicing free kicks if the person does not play soccer. A holistic approach emphasizes that training and other interventions should be authentic and have ecological validity for the person (Tellevik & Elmerskog, 2001).

A wider perspective on supporting coping in daily life is about making priorities beyond self-care activities, such as play, work or sports activities. Educational interventions, like training new reading techniques or preparing physical adaptations, are identified by making a task analysis of the selected activities. Intervention related to activities of daily living is common in the education of children and young people with visual impairment. In the present JNCL study, parents were asked a) to state whether their child had received this form of training and (b) to evaluate the impact of the training. Of the 106 parents who answered (b), nearly half answered positively (Figure 16.3). However, almost 20 percent had not received training even if they thought it was needed, while 25 percent said it had not been relevant. Nearly eight percent said they did not know,

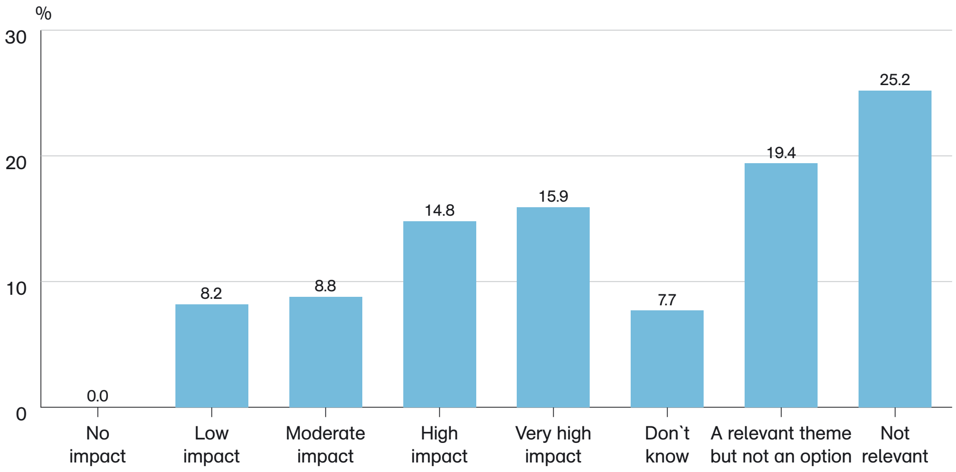


Figure 16.3 Parents' evaluation of impact of training activities of daily life (percent of answers) (N=106)

but it is not clear if «Don't know» meant that the parents did not know if the child had received this form of training or were uncertain if the training had had any impact on their child. Among those who reported an impact of this training (47,7%), 64.4 percent said that the training had a high or very high impact on the child, while about 18 percent reported a moderate impact. About 17 percent reported a low impact on the child from training related to activities of daily living. The mean age for the first training in activities of daily living was 11.1 years.

Training for orientation and mobility or activities of daily living has generally little place in mainstream education but constitutes a central part of special needs education for children and young people with visual impairment. A comparison of Figures 16.2 and 16.3 indicates that the parents considered mobility and orientation more important than activities of daily living. One reason may be that orientation and mobility training is offered to most children with severe visual impairment, independent of the student's cognitive level, whereas training in activities of daily living usually is offered to students with intellectual disability. For instance, in Norway, training in orientation and mobility is a legal right for all people with severe visual impairment, while training in activities of daily living lacks a similar legal status. Training of activities of daily living is usually part of the curriculum for students who are unable to follow the mainstream curriculum. With emerging dementia, activities of daily living should be an integrated part of the education for children and young people with JNCL to maintain independent or interdependent skills.

Decline in independence

The developmental trajectories of children with JNCL differ from those of children with congenital visual impairment or other disabilities. In the early years, children with JNCL are accustomed to managing tasks in their daily life in the same ways as their peers. For instance, the results of the present study indicate that very few children with JNCL have problems with passivity or learned helplessness in early childhood. Despite gradually emerging declines, children with JNCL struggle to be active and maintain the activities of ordinary daily life. Parents and teachers should try to avoid providing care and help when these are not needed, including making unnecessary priorities and life choices on behalf of the child.

The cognitive, visual and motor decline will sooner or later make independence difficult to maintain for individuals with JNCL. Most outdoor and many indoor environments need physical adaptation to make independence or interdependence possible. Physical adaptations include creating topographic patterns like handrails and curbs when needed or marking personal objects in order to make them recognizable by touch or hearing. Adaptations should be useful even in challenging conditions, such as when it is snowy, rainy, or noisy. Training should focus on goals that can be achieved through interdependence with others. This should start early in life for students with JNCL, because they need to learn appropriate strategies and becoming used to seeking help from people in the surroundings to achieve their personal goals. This is a basic requirement behind autonomy, but it requires a close and persistent collaboration with available helpers, including paid support staff, educators, family, and peers. Coping with everyday life will simply not be possible without such support in later phases of the JNCL disease.

Early intervention related to low vision

The length of the period from the first signs of visual loss to blindness varies, from one year up to ten years according to findings in the present study (see Chapter 4). The remaining vision will influence learning and coping with everyday situations, and it is of great importance that needs associated with level of vision are met by appropriate measures. Many complex tasks are easier to learn when the child has some vision. The child can learn by observing others' behavior when performing tasks, for instance how to tie the shoes or soap the hair, and not least, different kinds of social codes. It is therefore important to try to establish and automatize certain complex skills before the onset of blindness, when this is possible.

The period of decreasing vision will entail several practical problems; the child will need more and more time to accomplish routine activities. Carers should be careful not to take over tasks that the child can perform, even if it takes the child longer to complete them. Maintaining realistic expectations to the child despite the declines is important, for instance in connection with potentially challenging tasks such as fetching items, moving around, zipping up the jacket, and so forth. However, sometimes trade-offs must be made. Life is not only about performing daily routine tasks, but also about having fun. Too much time used on undressing may interfere with a forthcoming desired activity like swimming or playing. Finding a balance among these competing needs of daily life is essential.

Most students with JNCL are rather self-sufficient in independence in their early period of low vision. Interventions in this period of life include providing sufficient lighting and better organization of objects and activities, eliminating hazards, creating adequate visual contrast between objects and their background, and offering opportunities and technology for magnification (e.g., placing magnifiers strategically where they are going to be used, without needing to carry them around). Creating a person-centered, user-friendly and accessible environment makes life easier and will strengthen the child's or young person's ability to act as a driver in their own life. The creation of a person-centered and user-friendly environment is often limited to areas frequently used by the child with JNCL. Priorities should be given to classroom, bathrooms, working places, kitchen and/or important pathways used by the child (see Table 16.1).

Table 16.1 Some compensatory interventions for a child with low vision

Proposal (date)	Specific area	Type of intervention needed	Accomplished (date)
31/10-17	Bathroom at home	Additional lightening. Eliminating hazards – always close closets and cupboards	12/11-17
31/10-17	Classroom placing	First row in classroom near teacher's desk	02/12-17
12/11-17	Closet 2 at school	Increase lighting above the mirror	05/12-17
05/12-17	School desk	Gooseneck, swing-arm high-wattage non-glare lamps	10/01-17
10/01-18	Home	Night-lights in bedrooms, bathrooms and hallways to improve safety	26/01-18
10/01-18	Stairway+ entrance at school	Increase lighting above the stairs and antiskid flooring on top and bottom of the stairs to avoid accidents	26/01-18
10/01-18	General at school and home	Organization: Store items near the location where they are used. Put away clothing, cooking utensils, books or CDs immediately after use	Ongoing
26/01-18	Books, CDs and keyboard	Mark books and CDs with color codes, large letters or tactile codes for identification purposes	03/02-18
26/01-18	Keyboard	Mark the keys on the PC keyboard with large letters or tactile dots	03/02-18
26/01-18	Dressing room at school	Children's shoes, packages or other items to be removed from floor to wardrobe	03/02-18
03/02-18	Kitchen and bathroom at school	Install non-skid flooring	10/03-18
03/02-18	Classroom	Install door knobs in good contrast to the door color	10/03-18
03/02-18	General	Contrasts: Place dark-colored items against light background. Example: Place black towels in a white-tiled bathroom	Ongoing
03/02-18	Pathway from school to bus stop	Increase lightening. Hand rail	10/03-18
10/03-18	Playing corner at home	Gooseneck, swing-arm high-wattage non-glare lamps. Magnifying glasses	23/03-18

It is advisable to use appropriate expertise when implementing costly interventions that require knowledge about low vison and low vision technology. However, comments from parents and staff in the present JNCL study (Appendix A) indicate that costly devices in many cases were never used because of both the lack of local competence in how to use the device and the rapid reduction of the child's vision, as illustrated in the following quotations:

> *It was getting more and more difficult for her to see anything at all. At home she had a screen reader and a monocular, but she never actually used them. I don't know but she somehow refused to use these vision aids, and I doubt the school knew how to make best use of this equipment.*
>
> *He was encouraged to read using CCTV, there were sensory stories as well, but it was not followed up as needed. Some appropriate technology was used around at the time – his loss of vision declined fast – I'm not sure it had any important function for him. So probably earlier days of technology but, it's different then to now really.*
>
> *At that time however computers and information technology weren't used at schools yet so I taught her at home to write on a computer.*
>
> *Then, teachers should have some training in how to use the low vision equipment, and the FBA specialist should be able to put in a plan... difficult times.*

The period of low vision can also be used for precautionary interventions, that is, for preparing the child for the forthcoming blindness, such as marking the environment and making the child acquainted with features that can be useful later (see Chapter 12). Adults may choose travelling routes suitable for a child who is blind when moving between places the child is likely to travel later. Outdoor environments may be provided with handrails that later can be used as shorelines. The classroom, bedroom and locker room may be prepared for the child's visual impairment, using tactile marking or placing objects on fixed locations. Interesting texts in braille or Moon letters may prepare the child for later literacy skills, as well as keyboards suitable for tactile writing even if vision-based keyboards are an option in the child's current situation (see Chapter 14). Learning to use technical aids (e.g., a magnifier) can be accomplished through game-like activities that children can enjoy with peers and need not be boring or decontextualized (see Chapter 21). The understanding of the surroundings based on early visual memories may make later coping with daily life easier to maintain after the onset of blindness.

Table 16.2 A step-by-step model to support coping in everyday life

1	Assessment for learning and maintaining activities and making priorities
2	Task analyses of selected activities
3	Task analyses of selected mobility routes
4	Implementation of training and other interventions

Step-by-step support of coping with daily life

Promoting coping in everyday life for children and young people with JNCL includes such things as ensuring participation in desirable activities, providing training in how-to-accomplish tasks associated with those activities, and taking steps to make the physical environment user-friendly for each activity. Barriers and other contextual factors that prevent coping are often found on diverse task levels, such as a lack of physical adaptation and support, problems with transportation, and much more. The ultimate educational goal for individuals with JNCL is that they should be able to participate in desirable activities through independence or interdependence.

Analyzing tasks related to desirable real-life activities may help teachers and service providers identify needs for training, support and physical adaptation. Table 16.2 shows a step-by-step model to support coping in children and young people with JNCL.

Step 1 – Assessment for learning and maintaining activities and making priorities

Mobility routes and activities of daily living should imply participation in meaningful real-life activities. For instance, navigating the route to the activity «play at the playground» is the first and last step or sub-goal for participating in playing. The activity may have more routes, such as the route from the bottom to the top of the slide, or from the slide to the sandpit. The basis for the selection of training mobility routes and activities of daily living is consequently participation in meaningful activities.

Setting priorities for interventions based on real life activities entails a holistic perspective that includes consideration of participation, skills needed, necessary environmental adaptation, support, and other interventions. These factors are analyzed in relation to meaningful contexts. Relating teaching and training to common real-life activities makes it possible to refresh memory and

Table 16.3 Assessment form for activities and making priorities

	Activity	Where	Priority date	Goal	Goal achieved
1	Playing jungle gym	Playground at school		Interdependence	
2	Bathroom activities	At school, ground floor	13/02-18	Independence	05/05-18
3	Bathroom activities	At home	13/02-18	Independence	
4	Eating	Canteen at school		Independence	
5	Arriving at school	Cloakroom at school		Independence	
6	Using smart phone	Anywhere		Independence	
7	Visiting Carl	Home	13/02-18	Interdependence	12/06-18

skills in natural situations and avoid having to schedule training at fixed times that lack a similar degree of contextualization. Supporting learning in real-life activities will in many cases also strengthen the student's motivation (Tellevik & Elmerskog, 2001).

Table 16.3 is an excerpt from an assessment sheet of desired real-life activities of an adolescent with JNCL. This kind of assessment will serve as the basis for prioritizing interventions, as well as for introducing new activities that have not previously been available to the adolescent with JNCL.

Table 16.3 contains seven activities. Three activities were given a priority on February 13th, 2018. Activity number 7 – visiting the friend Carl – is a new activity to improve social interaction. Some goals were defined as independent, others as interdependent, meaning that the activity will require collaboration with a supporter. The goal of activity 2 was achieved on May 5th 2018, and activity 7 on June 12th 2018. Assessing real-life activities as shown in Table 16.3 and prioritizing interventions should be completed as a collaboration between school or institution, parents and – if possible – the person with JNCL.

Steps 2 and 4 – Task analysis of selected activities and implementation of training and other interventions

Most real-life activities consist of several tasks, which often should have a predetermined order. For example, going to the cinema may include «identify a film of interest on Internet», «get money for the ticket», «dress», «walk to the bus stop», «travel by bus», «walk from the bus stop to the cinema», «pay for the ticket», and «find the seat.» All the tasks are parts of the same activity – going to the cinema, but require different skills, actions and help, for instance when searching for information on Internet or walking to the bus stop. It is a chain of tasks that must be performed by the individual – and when needed – assisted by a helper. If the chain of actions or support of tasks is broken at one place it will result in non-participation.

Interventions are defined on the task level and not on the activity level. In the task analysis, the activity is divided into smaller, more manageable components, where skills, barriers in the environment and need for assistance are assessed separately. For instance, to make the environment easier to manage, the coat is marked for identification purposes and should always hang in the same place. Training could be directed at handling the zip or tying the shoes.

The personal need for participation in real-life activities should always be considered stable for persons with JNCL despite their declines. For instance, the need for participation in social settings does not decline because of speech

Table 16.4 Task analysis of bathroom activities for a person with severe cognitive decline

TASK ANALYSES OF BATHROOM ACTIVITIES AT SCHOOL (GROUND FLOOR)				
	Task description	**Physical adaptation**	**Goal**	**Achieved (date)**
1	Walk from classroom to bathroom	Bathroom door marked with wooden block	Interdependence Free guiding	15/05-18
2	Open door turn light on right hand side		Independence	01/06-18
3	Close and lock the door		Independence	01/06-18
4	Locate bathroom sink on left hand side		Independence	01/06-18
5	Mix water to desired temperature	Hot water tap marked with tactile dot	Independence	01/06-18
6	Push soap dispenser 2 times on left hand side	Soap dispenser placed left hand above the sink	Independence	01/06-18
7	Rub hands together		Independence	12/06-18
8	Put hands under the water and rinse soap off		Independence	12/06-18
9	Turn off the water		Independence	01/06-18
10	Dry hands with towel on right hand side	Towel placed right hand side of sink	Independence	18/06-18
11	Locate door, turn light off		Independence	12/06-18
12	Open door walk out of the bathroom		Independence	01/09-18
13	Walk from bathroom to classroom		Interdependence Free guiding	15/05-18

problems, but participation becomes more difficult and requires new training, adaptation and assistance as the disease progresses (see Chapter 13). Task analyses must be revised to adapt tasks, physical adaptation and help in accordance with the various declines. For instance, a red door that used to be identified by vision may require tactile marking after onset of blindness. The cloakroom should be located close to the classroom and a chair should be provided to make it easier to change clothes when mobility and motor problems become more severe.

The result of a well-designed task analysis will be a structured intervention situation, where the same procedures are repeated. At the same time, the detailed level of task analysis must reflect the person's potential. For instance, a severe decline in cognitive and motor skills requires more details compared to less severe cognitive and motor impairments. Table 16.4 shows a rather detailed task analysis for a student with profound cognitive disability. It includes 13 tasks, each reflecting this child's potential and need for training, help and environmental adaptation. A task analysis of the same activity for another child may consist of fewer or more tasks, and greater or lesser amounts of training, help and adaptation.

The goals of the activity shown in table 16.4 are independence or interdependence. The goals of tasks 1 and 13 are movements to and from the bathroom by using free guiding (see Table 16.10). Free guiding means that the child will follow the guide by listening to the sounds produced by the guide in front of him or her. The goal of the remaining tasks is independence. The analysis revealed that the main problem of task 10 was that the towel had not been given a permanent place. Independence was achieved by giving the towel a fixed place. The table shows that all goals had been achieved by September 1st, 2018.

Steps 3 and 4 – Task analyses of selected mobility routes and implementation of training and other interventions

The ability to move around and get from one place to another is essential for all people. The importance of mobility for locomotion is usually not given much attention until it becomes a problem. However, severe visual, motor or cognitive impairment may reduce the individual's possibility to get around if not met with appropriate measures. This requires access to help or learning how to cope with daily locomotion needs. An inability to move from activity to activity, or between places where activities take place, will restrict the person's possibilities for participation.

Orientation and mobility may include different forms of training, such as how to use the white cane, public transport, tactile maps or master routes, as well as how to be guided by sighted helpers, and use of self-protection techniques

(Elmerskog et al., 1993; Jacobsen, 2013; Wiener, Welsh, & Blasch, 2010). In the following section, the focus is on route training because it is of special importance to children and young people with JNCL.

Children with congenital blindness are often given preparatory orientation and mobility training that will lead to later independence and better skills in orientation. They are taught basic concepts such as right and left, improved body awareness, how to move in different terrains, and gross motor functions while walking. The situation for children with JNCL is different. Early orientation and mobility intervention is more about enabling them to travel as they did before the onset of severe visual impairment, that is, with a stronger focus on the child's *life flow* (Csikszentmihalyi, 1997).

Lacking vision places demands on the person's memory and search behaviors. For instance, observing that a particular table has a flat square area, four corners and four legs is done with a single glance by a person who can see. Making the same observation without vision requires investigation or explorative activities using the hands (Fraiberg, 1977; Tellevik, Storliløkken, Martinsen, & Elmerskog, 2007). Information through touch and movements must be accumulated and memorized to build a cognitive representation of the table. To learn new routes also requires movement, searching behavior and memory. Learning new routes based on non-visual cues requires more anticipated and explorative behaviors and makes higher demands on memory than the use of visual cues. The individual must remember what to do at each point along the route to reach the destination. However, when a route is learned and the child or young person master the navigation, less cognitive effort is required (Tellevik et al., 2007). Teaching children with JNCL to move around independently in familiar environments in relation to important activities prior to onset of total blindness will therefore be a goal of high priority. Enabling children with low vision to act as "drivers" when they move around will support their understanding of the surroundings, which in turn supports maintenance of independence and interdependence when vision has been lost.

Teaching orientation

People with and without vision use different strategies in orientation. Perception based on visual orientation may be described as information from a *field*, while perception based on non-visual orientation to a high degree is based on information from a *line* or *tunnel* (Gibson, 1966). To orientate on a *line* or in a *tunnel* means that most information is proximal and limited to the space that can be physically reached with bodily movements.

According to Gibson (1966), visual information tends to be interceptive (the information comes to the person) while tactile or haptic information requires exploration behaviors. Further, in visual orientation the attention is directed forward when moving, in non-visual orientation the attention is often directed sideways. For instance, a sighted person will walk, drive and orientate towards features in the environment, while a pedestrian who is blind will walk forward but orientate along features in the environment, such as walls or curbs.

A child with JNCL who is becoming blind must learn new ways of orientation, such as using fences and curbs. In the early phase, this may cause problems in independent travelling. The child may not be attentive to physical features and structures that are required for independent travelling and must learn to search for accessible and relevant information in the environment. The teacher must be aware of such challenges and guide the child to orientate along physical recognizable features and structures. Directing the child's attention towards features and structures along the path of travel can be made by knocking on the wall or carefully leading the child's hand to walls, fences or objects along the path. The main aim of teaching routes is to guide or direct the child's attention to predefined features along the route when moving forward.

There are two basic types of orientation in route training: *egocentric* and *topographic* (Ferrara & Landau, 2015; Newcombe, Uttal, & Sauter, 2013). Figure 16.4 illustrates the difference between egocentric and topographic orientation. In topographic orientation the person uses objects or features in the environment as cues to determine her own position. The experience of walking on the left side of a river or using a roadmap for orientating are examples of topographic orientation. In egocentric orienteering the person uses her own body as reference point for orientating and determining the position of objects or structures in the environment. People with vision can easily choose to use both egocentric or topographic references in orientation since vision will provide information that one is a part of a stable environment. The situation is different for people with blindness.

Orientation for persons who are blind should be based on egocentric orientation when learning new mobility routes. The

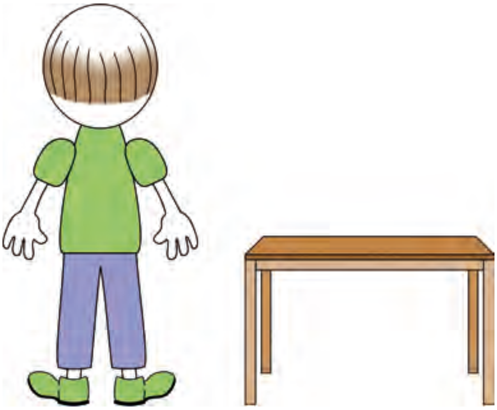


Figure 16.4 Orientation based on egocentric and topographic orientation

The person is observed from behind.
Topographic orientation: *You are standing on the left-hand side of the table.*
Egocentric orientation: *The table is on your right-hand side.*

traveler's body should always be used as the reference point in relation to objects, features and structures along the path. Topographic references may require complex cognitive calculation as shown in the following statement: *I am told that I am standing on the left-hand side of the table. This means that the table is on my right-hand side*. Mixing egocentric and topographic orientation in instructions – which is common among people who are sighted – may make route learning very difficult for persons with JNCL. This example also highlights the complexity of language implied in instruction.

Routes

It is a well-known saying that all roads lead to Rome. People who are sighted may use any road leading to their destination. Learning a mobility route means to learn to master one of these roads. A traveler with JNCL will in most cases keep to the route that she or he has learned when not assisted by a sighted helper.

A mobility route is a pre-defined path from a specific starting point to a specific destination. It can be the long road from home to school, or the short path from the kitchen table to the refrigerator. Learning orientation is learning goal-directed attention towards important features in the surroundings to reach a desired location and activity. The training should be implemented as how-to-do-it or procedural learning for individuals with JNCL. The route should be considered as tasks within the activity it is attached to. For example, walking to and from the playground is the first and the last task in the activity «playing with friends at the playground». When individuals who are blind are navigating routes, the main features are landmarks, shorelines and laps.

Landmarks

A mobility route consists of two or more landmarks, identifying permanent topographical positions within routes, which may function as cues for travel and orientation. Landmarks can be objects such as a posts, curbs, fences or doors, or attributes of objects, for example the sound of a creek or the rough surface of a stone wall.

There are three basic criteria for selection of landmarks. The person must be able to detect and recognize the prominent physical features of the landmark, the landmark must be different from other objects in the area (e.g., a tree in the forest is not a useful landmark), and the landmark must not be moveable but have a permanent location.

Landmarks should have distinct characteristics and be easy to locate, identify, and distinguish from other objects in the environment. Good landmarks are prominent and appear as a change in the surroundings along the route. They must always be available when there is a need to make changes in the direction of travel or indication of a hazard on the way, like traffic or steps. The distance between landmarks needs to be adapted to the attention span of the individual. For instance, the movements between two landmarks should not require more than ten seconds if the attention span – the time the person manages to keep the landmark in mind – is ten seconds. If there are not enough natural landmarks along the route, it may be necessary to create artificial ones, for instance, place a pole where the person is supposed to make a turn, place a carpet in front of the door to identify the classroom, or make a road bump on the sidewalk to indicate a danger of crossing traffic.

Tactile or haptic landmarks may be identified through touch and bodily movement. They may include posts, building corners, the end of a fence, and carpets on the floor, and should be identified and located without too much explorative efforts. These landmarks can be associated with almost any object or feature but cannot be detected from a distance. Objects that are too large are not suitable as tactile or haptic landmarks, because they usually do not function as distinctive points of references for effective orientation. It is important to avoid tactile or haptic landmarks that require too much cognitive effort and searching behavior, such as stretching or bending (Tellevik et al., 2007). Landmarks should be located in a position where they will be detected without unnecessary exploration, that is, on the shoreline that is used in the orientation.

One should try to select tactile/haptic landmarks that can be detected with the foot, hand or white cane, although taking into consideration that landmarks on the ground may disappear under snowy or rainy conditions. Tactile and haptic landmarks are in many cases perceived to be more stable and distinct than auditory landmarks. It is therefore recommended to use tactile/haptic landmarks when planning a new route for children and young people with JNCL who are blind.

Auditory landmarks may be sounds from a fan, a busy road, or a crossing signal on the traffic light. They may also be perceived as an echo from a wall or be related to the acoustics in a room. Objects that produce distinctive sounds when they are touched or hit can sometimes function as landmarks. Most auditory landmarks can be detected from a distance. However, a single sound from a car cannot be re-checked by the listener, and many auditory landmarks are not as permanent as tactile/haptic or visual landmarks.

A child who is blind may gradually change from tactile or haptic to auditory cues of the same landmark (or shoreline). Such a development should be considered a positive development, because it implies that the child has learned a more effective way of orientation, for example to use echo-localization.

Visual landmarks are objects or visual attributes of objects, such as size, shape, or color. Children with JNCL use visual landmarks in early childhood, but visual landmarks must be replaced with auditory or tactile/haptic landmarks after the onset of blindness. From the start, one should try to select landmarks that can be used visually as well as tactile or haptic when planning routes for children with JNCL who have low vision. This may ease the transitions from visual to tactile/haptic cues.

Shorelines

Shorelines give the person continuous information along the way between two landmarks. Whenever possible, landmarks should be connected in pairs by a good shoreline, such as a wall, the edge between asphalt and gravel. There are tactile/haptic, auditory and visual shorelines. Tactile/haptic shorelines are preferred when planning routes for children with JNCL who are blind, because they are perceived as more stable and permanent than auditory shorelines.

It may be necessary to create artificial shorelines when there are no appropriate patterns along the route. For instance, one may build a rail between two landmarks or set up a sound beacon above a door. Further, like landmarks, an outdoor shoreline on the ground may disappear with snow or rain and might be replaced with a hand rail. Figure 16.5 illustrates the design of an artificial shoreline. The crosswalk has been raised, the person with JNCL can follow the raised edge of the crosswalk as a shoreline to allow for safe crossing.

Artificial shorelines do not have to be costly and providing them is rather a matter of finding functional and practical solutions. For instance, connecting a sidewalk and a playground located in an open field with a rope may enable a child with JNCL to play with other children without adult help.

Shorelines can be upright, such as handrails, or incorporated into the path, such as a pattern or surface texture. It is practical to select upright shorelines when possible. They are easy to follow and can be perceived also when there is snow or rain. Moreover, it is easier to follow upright shorelines like handrails with the hands than a pattern on the ground with the feet or white cane (Tellevik et al., 2007).

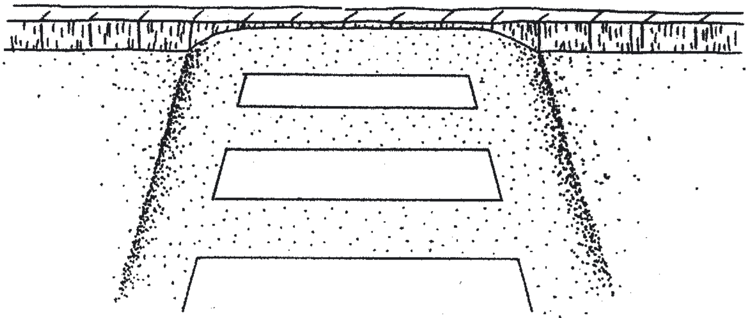


Figure 16.5
Example of physical adaptation of a shore line.

Laps

A mobility route can be divided into laps. A lap is the path between two consecutive landmarks in a route. To achieve independence, the person needs to learn all laps in a route. Learning one or several laps may be sub-goals towards achieving independence or interdependence in the route. The child may move according to interdependence between two landmarks by using free guiding.

There are two types of laps in a route. A *shoreline lap* is a path connected by two consecutive landmarks with a shoreline like a handrail, wall or curb. A *crossing lap* is the path between two consecutive landmarks that are not connected with a shoreline. Crossing laps are generally more difficult to learn than shoreline laps and should be avoided when possible. The first landmark in a crossing lap is often used for taking the right position and indicate the direction of the crossing. The second or last landmark in a crossing lap should be a landmark that intercepts the individual; the landmark should allow a margin of error of direction when crossing.

Learning and teaching laps and routes

Figure 16.6 shows an example of a route between home and school for a child with JNCL. The dotted line is the pathway and X1–X9 are landmarks. The landmarks are features such as beginning and end of a handrail, pillars and doors.

In Figure 16.6, "E" represents a full shoreline-lap with two subsequent landmarks (X3 and X4) connected with a shoreline. The child who is standing at landmark X3 (beginning of the handrail) can locate landmark X4 (end of the handrail) by following the shoreline (the handrail).

The lap from X4 (end of the handrail) to X5 (the edge between tarmac and grass) is a crossing-lap because it does not have a shoreline. The child with JNCL can use the handrail to find the right position for crossing the road. The child must uphold the direction given by the handrail while crossing the road to locate landmark X5. Maintaining correct direction while walking without a shoreline can be demanding for children with JNCL and should be avoided when possible.

Achieving independence in shoreline laps requires that the child or young person learns to locate the next shoreline (which is often attached to the previous landmark), the body's position in relation to the shoreline (left- or right-hand side), and the next landmark (attached to the end of the shoreline). The teacher's task is to direct the child's attention to these three tasks. The child can move independently in the lap when he masters these three tasks.

Achieving independence in crossing laps requires that the child takes the right position at the first landmark of the lap, for example by placing the back toward a wall or a handrail. The next step is to maintain the direction when crossing the open area. The last step is to locate the landmark that ends the crossing lap, for instance, a wall or edge between tarmac and grass. The role of the teacher is to direct the child's attention to these three tasks, and the child will be independent when these tasks are mastered.

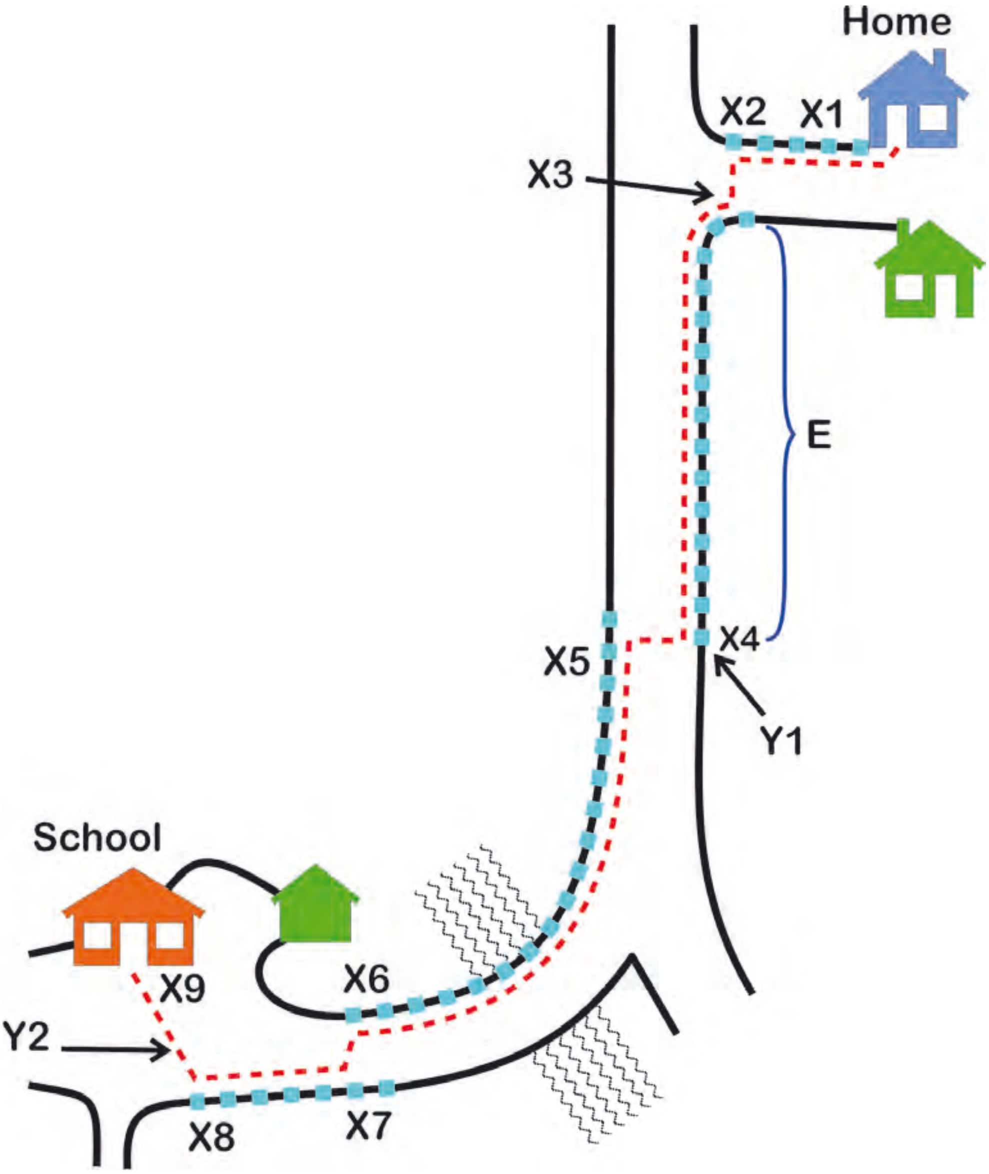


Figure 16.6 A mobility route between home and school for a child with JNCL. The dotted line is the pathway and X1-X9 are landmarks. Y1 and Y2 mark crossing laps and E shows a full shoreline lap (2 landmarks and a shoreline).

Planning routes

It is important to note that a route is a one-way pathway. In most cases it is not possible to use exactly the same landmarks and shorelines when returning to the starting point. The body position in relation to shorelines will always be on the opposite side when going back.

The teacher must always prepare the route prior to the training. This consists of selecting appropriate landmarks and shorelines and making physical adaptations in the environment where needed. An example of a prepared route from the classroom to the locker room is shown in Table 16.5 and the corresponding map in Figure 16.7. The dotted line in the map indicates the pathway. The route has 7 landmarks and 6 laps. Laps number 1, 3, 4 and 6 are shoreline laps while laps number 2 and 5 are crossing laps. The analysis disclosed a need for two physical adaptations, artificial landmarks.

The starting point of the route is the classroom door (landmark 1). By following the wall on the left hand side, the student locates the wall carpet (landmark 2). The wall carpet is a physical adaptation, this location had no features that could be used as a landmark or signal for the next crossing lap. The student takes position at the wall carpet (landmark 2), crosses the corridor and locates the wall on the opposite side (landmark 3). By following the wall on the right side, the student locates the corner (landmark 4), then turns right and follows the wall on the right side to the glass door (landmark 5). The student takes position at the glass door, crosses the corridor and locates the wall on the opposite side (landmark 6), and then follows the wall on the left hand side to locate the hanging rope (landmark 7). Landmark 7 is another physical adaptation, which enables the student to identify his own seat.

Table 16.5 The analysis of a route from the classroom to the locker room

ROUTE FROM CLASSROOM TO LOCKER ROOM			
	Landmark	**Shoreline**	**Physical adaptation**
1	Classroom door	Wall left hand side	
2	Wall carpet	No shoreline (crossing lap)	Wall carpet
3	Wall on opposite side	Wall right hand side	
4	Corridor corner	Wall right hand side	
5	Glass door	No shoreline (crossing lap)	
6	Wall on opposite side	Wall left hand side	
7	Shelf marked with rope		Hanging rope

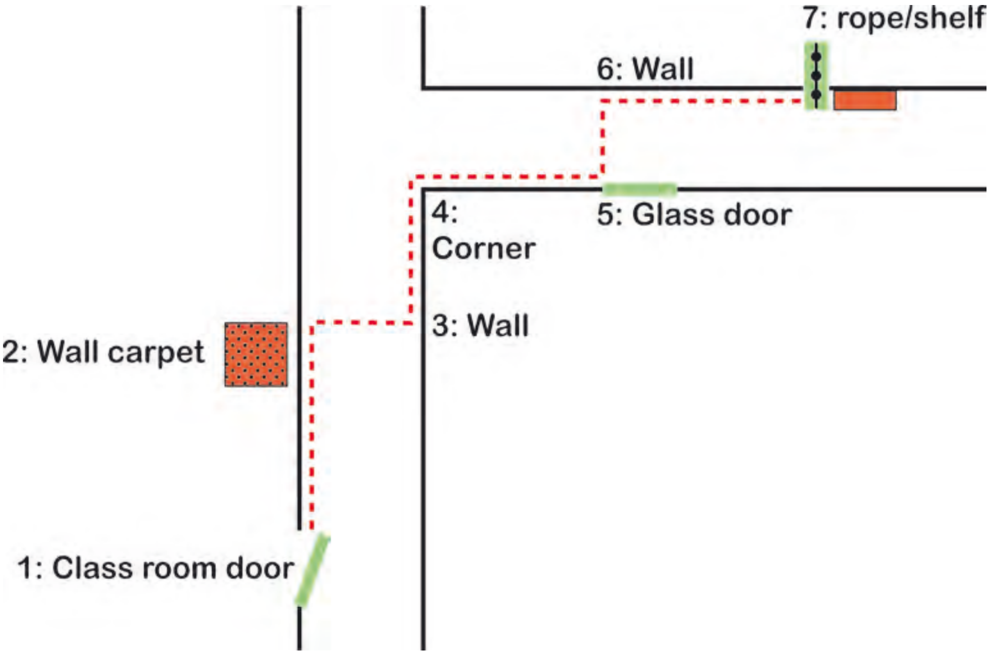
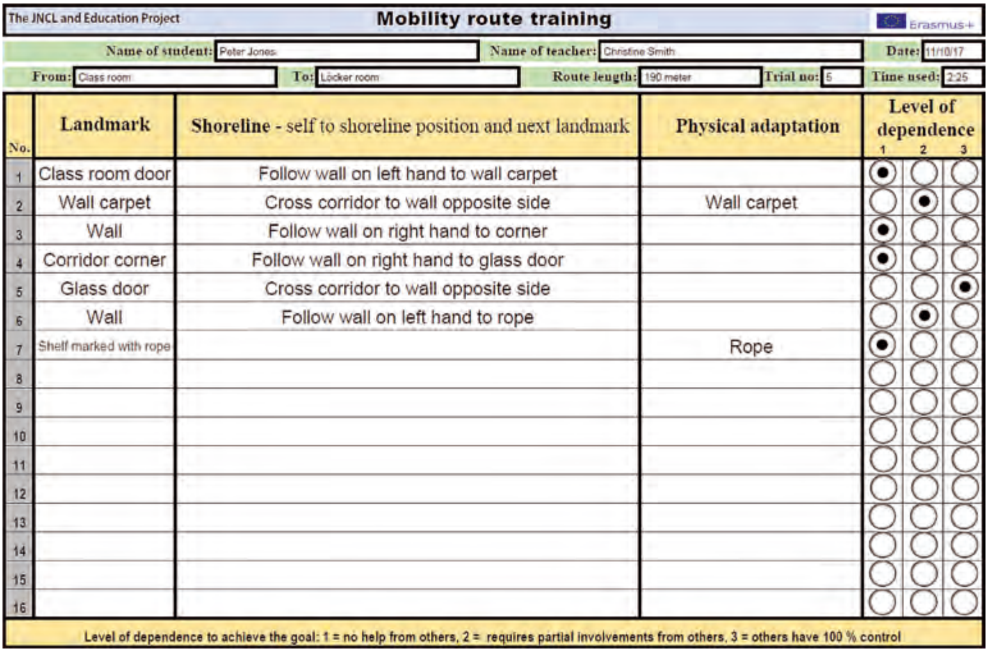


Figure 16.7 Map corresponding to the route in Table 16.5

Route training

Learning a route requires many repetitions of the predefined laps, landmarks and shorelines. The teacher's role is to guide the student's attention towards the pre-defined landmarks and shorelines on the route. The student must learn if the shorelines and landmarks are on the left or right side while moving, according to egocentric orientation. The teacher should not walk in front of the student when training independence, because the student may, without being aware of it, use sounds from the teacher for orientation purposes instead of using the predefined landmarks and shorelines. Unnecessary talking and information about things that are of minor importance for the orientation during the training may interfere with the student's attention towards the landmarks and shorelines. Every lap – from landmark to landmark – should be regarded as a sub-goal in the route training. One can strengthen the student's appreciation of laps or sub-goals by asking the student to stop at each landmark in the beginning of a training period.

Figure 16.8 shows an electronic form that can be used for training and evaluation of route training. The route describes the path from a classroom to the locker room, a distance of 190 meters. All the predefined landmarks are supposed to be signals for actions like «turn right», «cross over,» or «find next shoreline» based on egocentric orientation. The physical adaptations include artificial adaptations for making the orientation easier.



The JNCL and Education Project

Mobility route training

Name of student: Peter Jones **Name of teacher:** Christine Smith **Date:** 11/10/17

From: Class room **To:** Löcker room **Route length:** 190 meter **Trial no:** 5 **Time used:** 2:25

No.	Landmark	Shoreline - self to shoreline position and next landmark	Physical adaptation	Level of dependence		
				1	2	3
1	Class room door	Follow wall on left hand to wall carpet		●	○	○
2	Wall carpet	Cross corridor to wall opposite side	Wall carpet	○	●	○
3	Wall	Follow wall on right hand to corner		●	○	○
4	Corridor corner	Follow wall on right hand to glass door		●	○	○
5	Glass door	Cross corridor to wall opposite side		○	○	●
6	Wall	Follow wall on left hand to rope		○	●	○
7	Shelf marked with rope		Rope	●	○	○
8				○	○	○
9				○	○	○
10				○	○	○
11				○	○	○
12				○	○	○
13				○	○	○
14				○	○	○
15				○	○	○
16				○	○	○

Level of dependence to achieve the goal: 1 = no help from others, 2 = requires partial involvements from others, 3 = others have 100 % control

Figure 16.8 Form used for route training

The columns *Level of dependence* in Figure 16.8 is an evaluation of the student's current orientation level on each lap. Level 1 indicates that the student can find the next landmark without help from the teacher. Level 2 indicates that the teacher must intervene in this lap, for instance by guiding the student to follow the shoreline or to locate the next landmark. Level 3 means that the teacher must take full control of the student's orientation. The figure shows that the student with JNCL has achieved full independence in lap 1, 3 and 4, partial independence in lap 2 and 6, and needs full support in lap 5. The evaluation of lap 5 may indicate a need for further adaptation, for instance putting a carpet across the corridor which will change the present crossing lap into a shoreline lap.

A well-described route form is important when two or more teachers are training the same student. Route forms make it easier for staff to have the same expectations to the student and contribute to ensuring that they use the same laps, landmarks and shorelines.

Level of independence will over time be reduced for young people with JNCL. They will need more help when moving to desired activities. The earlier learned mobility routes may still be of importance despite less independence and more interdependence and dependence. Established mobility routes can be used for creating anticipation and maintaining personal overview and control over what is happening. For instance, landmarks like a door with a bell or a floor with a thick carpet can make the student aware: *I am now on my way to the kitchen and lunch*. In later phases of the disease, use of fixed routes throughout the day is one method of supporting and maintaining the person's ability to understand what is going to happen next.

Guiding persons with JNCL

Sighted guides will become a necessity for persons with JNCL when the visual, motor and cognitive problems become more severe. Guiding is typically needed in unfamiliar environments, crowded places and places with certain hazards, or when there is a need to move fast. However, guiding should be avoided when not needed and it is an overall aim that students with JNCL should be as active as possible when they are guided. Active guiding means to use free guiding (see Figure 16.10) or verbal guiding, instructing students what to do rather than leading them with physical contact. Active guiding also implies letting the students open and close doors or pulling the chair out when they are sitting down.

The guiding grip

The person should hold the sighted guide as shown in Figure 16.9 when guiding by holding is needed. Children can hold the guide's wrist or fingers. The sighted guide should always walk a step ahead of the person with JNCL. There are several techniques that can be used when guiding by holding is needed (see Elmerskog et al., 1993; Jacobson, 2013; Wiener et al., 2010).

Figure 16.9 The guiding grip

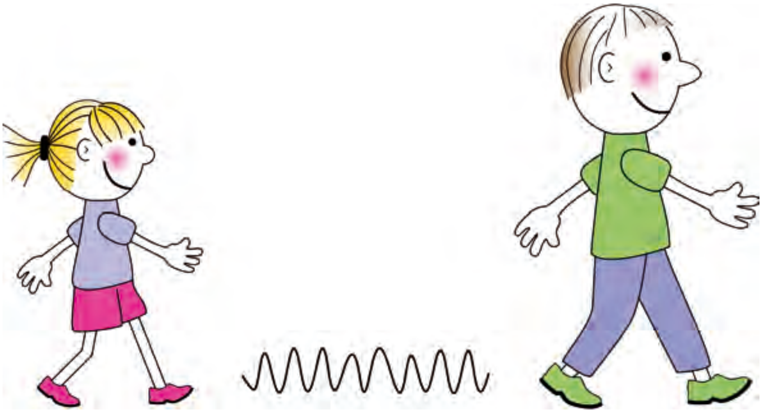


Figure 16.10
Free guiding

It is a risk that children with JNCL become overwhelmed by offers to be guided by holding just after the onset of the severe visual impairment. Adults and peers in the surroundings may think that they are supporting the child in best possible way when offering guiding. However, too much guiding may rather be a disservice, because the child may become accustomed to being transported or being a «passenger» when moving from place to place. The provision of guiding must be managed in a balanced way.

Free guiding

Free guiding should be given priority when guiding is needed for children with JNCL. Figure 16.10 shows an adult guiding a child with JNCL. There is no physical contact between them, the child uses sounds from the guide as cues for her orientation. The guide may make sounds with the feet, by talking to the child or by holding a device that makes sounds. Guidance from a distance requires more attentional resources from the student, compared to guiding by holding.

Free guiding may be a step towards later independence. It is therefore recommended to use designed routes when free guiding is used, because the child may memorize laps, landmarks, shorelines and turns on the way to the destination.

Guiding wheelchair users

Using designed routes may also be important for a person with JNCL using a wheelchair when moving from place to place. Perceiving 90 degrees swings at the same location every day and passing the same uneven and rough surfaces, carpets or doorsteps may establish a foundation for later orientation. Also changes in speed can be used for orientation. For instance, a pronounced 90 degrees turn combined with a door still may be recognized as the entrance to the school canteen. Guiding a person with JNCL in a wheelchair should always be combined with verbal information about where to go, features in the surroundings, and that the destination is reached. A person with JNCL in a wheelchair should always be placed close to known and recognizable physical features and objects, such as behind a known desk or beside a familiar wall while sitting still.

Suggestions from parents and staff

Comments from parents and staff in the present JNCL and education study (Appendix A) included several suggestions that may be useful for enabling children and adults with JNCL to cope in daily life. Some of these were related to self-care and grooming. Containers with soap or shampoo can be marked with braille, Moon, a rubber band or small object taped to the container. Identification will be easier with different sizes and shapes of bottles and tubes. It is easier to get a suitable amount of soap with a pump bottle, or to press out the right amount of toothpaste by squeezing it directly into the mouth from personal tubes.


Figure 16.11 Systematic organization of clothes

Other suggestions were related to dressing and undressing. Clothes should be easy to handle for the young person when the disease progresses. For instance, shoes with Velcro fastening may maintain independence when shoes with laces become difficult to handle. Short moments of training in natural situations may provide a basis for automatizing skills and developing fine motor skills such as using a zip, buttons, or laces. Putting the clothes in a certain order when undressing makes dressing easier (see Figure 16.11). Parents should take the prevailing youth styles, dress codes and the child or young person's own voice into account when choosing or purchasing clothing. A mother told that her daughter was obsessed with having a hair braid, a style that was very popular prior to the daughter's visual impairment. The fact that this hair fashion had faded was never communicated to the daughter, and her obsession with hair braids disappeared when she was told about the new hair trends.

It is advantageous to practice skills related to eating and drinking from early childhood. The child can take part in preparing food and learn hand-over-hand to chop vegetables or butter a slice of bread. Pouring drinks can be learned with practice, as well as laying the table, taking food from the kitchen and carrying it to the table, or taking the dishes to the dish washer after dinner. Such skills can be accomplished independently or through interdependence. As skills decline, eating becomes more difficult. Physical adaptation may include putting a supplementary edge on the plate, using thicker cutlery, or putting an anti-slip pad under the plate.

The disease may make swallowing more difficult, and with age feeding the person with JNCL may be needed. In such cases the person should be told with speech or touch when the food is just about to enter the mouth. Severe problems with swallowing may require chopped or liquidized food. Drinking may become easier if the liquid is thickened.

Many parents mention that the use of money is an inspiring activity for many children with JNCL. A wallet with money or a card and paying for purchased items in the shop can be used for building up an understanding of money values and mathematics in general. A coin holder can be used for storing money, which also makes the differentiation of coins easier.

Parents in the present study reported sleeping problems for two thirds of the participants with JNCL, at a mean age of onset of 11 years. The need for rest and sleep will increase but it is important to plan for activity and participation, and to a lesser extent for rest and sleep, although considering the need when it becomes comprehensive. Sleeping problems can be met by medication, but the establishment of best possible daily rhythm is as important as for most people. The daily rhythm should be regular and contain relatively few changes. Quiet routine activities before bedtime, like listening to an audio book or soft music, may make it easier for the person to fall asleep. The best possible daily rhythm is a matter of being energized and engaged in appropriate activities through personal involvement.

Parents and staff reported that nightmares, confusion in connection with sleeping and waking up were increasing with age. Some emphasized the importance of immediate contact with helpers or other adults after waking for stopping further escalation of confusion and desperation. However, falling asleep during daytime is much more common when not being engaged or involved in activities.

Some persons with JNCL may take instant naps while being involved in activities. According to parents and staff in the present study, long-lasting activities should be avoided when possible for persons with short attention span; variation and short-lasting activities are of help to maintain spirit and engagement.

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17

Music Education and Music in Education

Adam Ockelford and Rebecca Atkinson

This chapter is about the role of music in the education of children and young people with juvenile neuronal ceroid lipofuscinosis (JNCL). The survey presented in this book (Appendix A) is the first systematic study ever to be undertaken in this area, although anecdotal accounts of parents, and reports of teachers and music therapists, have in the past suggested that music may potentially have an under-utilized role in the lives of individuals with juvenile neural ceroide lipofuscinosis (JNCL) (Bills, Johnston, Wilhelm, & Leslie, 1998; Gylfason, Jóhannsdóttir, & Einarsdóttir, 2006; Heikkinen & Railosvuo, 2006; Olsén & Laine, 2006).

Music, whether heard or performed, is something that is experienced through the body and in the mind, and research has shown that the brain can continue to respond to music even when there is damage or degeneration (Baird & Samson, 2015). While music uses certain specialized areas in the brain, there are close connections too with the neurological resources devoted to language, movement, memory and emotion, and it has been suggested that the existence of these connections may be central to the efficacy of music interventions (Woods et al., 2005).

Main results from the survey

Parents were asked about their child's use of music and what impact the music had had on the life of the child. Similar questions were asked to professionals working with children and young people with JNCL. The informants were asked at what age the children began to engage with music, and for how long this engagement continued. There were questions too concerning music lessons and music therapy sessions, and about the potential power of music to facilitate social contact, to support communication, to provide comfort, to aid learning and understanding, and to offer stimulation and relaxation.

Most of the children and young people in the present study were said to be interested in music, whether as listeners or performers, or as an accompaniment to dance or a topic of conversation. In the survey, 87.4 percent of the parents said that music had or once had a high or very high impact on the life of their child. While the importance of many activities was declining with age, the percentage who found music important increased from around 36 percent in the age bracket 7–9, to around 50 percent at ages 13–15 and 16–21, and nearly 80 percent among those above the age of 22 (Figure 17.1). These substantial changes from childhood to adolescence and then young adulthood are probably a reflection of the fact that the enjoyment of music does not rely on intellectual capacity or motor abilities. Similarly, the impact of audio books, another receptive activity, was said to increase gradually from less than ten percent at 7–9 years to over 50 percent above the age of 22 (see Chapter 14).

> *Susan always had her music with her after she started to go blind. Up to the end music was a big part of her life.*

The active use of music, such as performing and dancing, was most prominent in childhood and early adolescence. Singing was said to be at its best (and on a par with sighted peers) around the age of 12.

> *I think that song and use of the voice is an instrument. He sang a lot up to the time when his voice began to break. The decline of skills started to accelerate during puberty.*

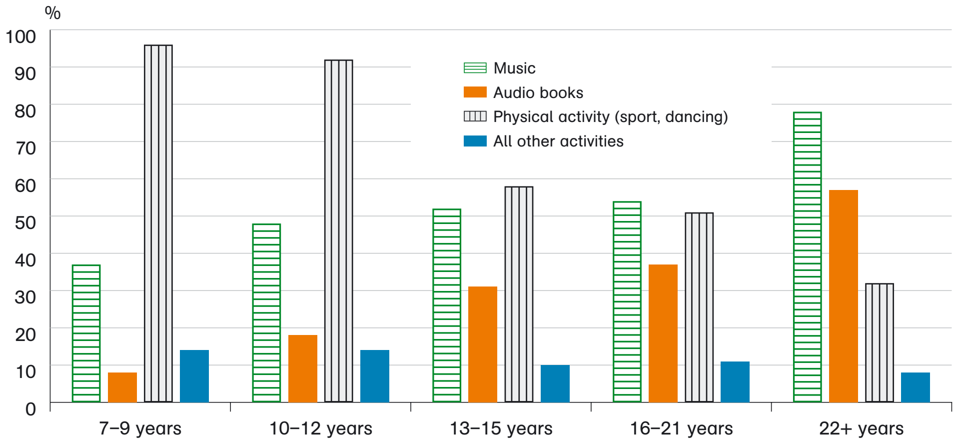

Figure 17.1 Percent of participants for whom music, audio books, physical activties and all other activities were important at different age levels

> *She used to have a very nice singing voice, but that has disappeared as the disease has progressed. She still enjoys singing, although it is usually just the last word of each line.*

Around half of the children and young people with JNCL played, or had once played, an instrument, and some had tried two or more instruments. The most popular instruments were the keyboard (including the piano), drums (or other percussion) and the guitar.

> *Drums and drum kit. Started to play around 6–7 years of age. Achieved best skills (the peak) around 12–13 years of age. Played in a band organized because of him, he continued doing this up to 20 years of age. He was also offered special training (one-to-one).*

Most children started playing instruments around the age of eight (some earlier), which continued for around five years. Around two thirds (65.5%) did not get beyond the level of beginners, while the rest were described as performing at a similar level to or better than their peers.

> *Since the age of 4–5 years (7 now), he wanted to play and own every instrument. He took drum lessons for eight months and guitar for two months when he was six and seven. He is neither technically trained nor proficient, but he has great rhythm and musicality. He has played or plays the following: drums, cowbell, cymbals, triangle, washboard, harmonica, recorder, spoons, guitar (acoustic and electric), clarinet, trumpet, violin and keyboard.*

It seems that the children started to engage more with music as their eyesight deteriorated, but generally stopped when they could no longer read print music notation. Only one adolescent was reported to have attempted to learn to read music in braille. Around 20 percent were reported to make up (or to have made up) their own music. This occurred largely between the ages of 11 and 15.

Around one third of the parents (32.1%) said that their child had received music therapy. The median age for starting was 14, and the mean duration of the music therapy was eight years. Within this group, around 90 percent of parents and 80 percent of professionals reported that music therapy had (or once had) a high or very high impact – primarily as a source of stimulation and comfort.

The potential function of music for children and young people with JNCL

Sixty-nine parents commented on the potential role of music in support of wider functioning and wellbeing. Many said music helped their child to sustain communication when words alone no longer functioned, that music was a medium through which the child could convey feelings when other channels of emotional expression were occluded. Parents also reported that music reawakened memories that otherwise appeared to be lost, and how music helped their child to maintain a sense of wellbeing in an increasingly confusing and frightening inner world.

This section discusses the findings from the survey in relation to these issues, compares them with those from other research, and makes suggestions for musical activities and strategies that may be useful for families and for professionals working with individuals who have JNCL or other comparable disabling conditions.

Feelings

Just under half of the respondents identified music as being important in promoting individuals' wellbeing, assisting with emotional regulation and expression, and relaxation. This is particularly important for individuals with JNCL since they experience multiple losses – in skills, friendship and the availability of activities in which they can participate – which can cause anxiety, uncertainty and anger.

> *Music was very important to her. She liked music for relaxation, as well as music for singing along (which was functioning for an astonishingly long time). Music also evoked strong emotions, grief as well as joyfulness. Music therapy at school was one of her favorite subjects. She always liked music while being together with others, at school, with the family or on organized vacations. Music brought about a high level of wellbeing.*

> *She played her instruments, guitar and drums, and she enjoyed singing. Soft music was used to calm her down the last years she was alive (to prevent anxiety and restlessness).*

To manage such feelings, one parent described how «music could change a child's mood and make it better», sometimes through bringing back good memories. Another parent reported how playing the drums helped her child maintain his self-confidence: *music has been, and still is, his greatest passion*. Similarly, a professional noted that music could both produce *a high level of excitement* as well as having *a calming effect during critical situations*.

Using music to create a feeling of calm can be particularly important at bedtime: one family used it when their daughter was going to sleep *to create a feeling of unity*, acting as Ockelford (1998) puts it, as an auditory "security blanket." A number of parents and staff commented on how music was used to distract or as a diversion during moments of personal care, to reduce anxiety in everyday activities like travelling in the car, and to help with pain management. Whatever the circumstances, *preferred* music was said to be the key: Bills and associates (1998, p. 14) reflect that children with JNCL often have «favorite music … that they seem to hang onto and will listen to over and over.»

Olivia's story

Olivia is 15 years old and lives in the UK with her mother, father and two brothers at home. She attends a school for children with special educational needs, to which she moved just over a year ago from a mainstream secondary school. Olivia is chatty and sociable, and becomes animated when talking to others about her passions, preferences and experiences. She loves music, and among her favorite pieces are songs from Disney, Westlife, Britney Spears and Rihanna.

Olivia says that she enjoys being at school, and is «always happy». However, when she first moved to her new school, this was not the case, and staff reported that Olivia suffered a good deal of anxiety, clutching at the walls of the corridors, apparently terrified of being in unfamiliar surroundings. It was suggested that music could be used to help her relax as she got used to her new school environment, and weekly music therapy sessions were arranged with a clinician at school. Staff commented how these sessions lifted her mood.

Olivia.

Olivia has significant difficulties with speech. She talks very quickly, often mis-accentuating words or stammering, and repeating things in an effort to be understood. However, when Olivia is singing along to her favorite music – for example, in music therapy sessions – her capacity to use words is transformed. She sings in time, with linguistic clarity and precise articulation. She maintains the contours of melodies, though not the precise pitches of each note. But the important thing is that she is able to produce language that others can understand.

Language and communication

As the disease progresses, children and young people with JNCL increasingly face challenges in the production of language: speech increases in speed, with rapid stuttering; articulation is impaired and words are mispronounced (see Chapter 6). Eventually, the speech may become incomprehensible. However, the capacity to sing appears to remain intact – sometimes for years – beyond the point at which speech is difficult or impossible to understand, and this was reflected in a number of the observations made by parents and professionals. For example, a child was reported to speak more clearly when singing what she wanted to say, and another recognized that the *singing of texts is better than speaking*.

> *Music was very important – she enjoyed listening and singing. Long after her speech went she was still able to sing or mouth the words to "Happy birthday to you" for example.*

One young person was said to really enjoy listening to her favorite artists and songs throughout the day. She also went to concerts. Singing clearly brought her pleasure, and in her singing the *lyrics came out clearly, even though her speech was so little, stuttering and slow*.

Why should this be the case? Research extending back over several decades has suggested that in songs, music and language may be encoded together in the brain (e.g., Morrongiello & Rose, 1990; Serafine, Crowder, & Repp, 1984), findings that have found support in more recent neuroscientific work (e.g., Schön et al., 2010). According to Patel (2007) elements of linguistic and musical processing may share some resources in the brain, while nonetheless having areas that are discrete. Hence one may speculate that in conditions such as JNCL, when the "direct" route to speech production becomes damaged, an "indirect" route remains open, through singing. The "shape" or "contour" of a song's melody (its ups and downs) and its rhythm (pattern of accents over time) may open the door to song production. The effect is that the natural prosody of language – its "musical" qualities – are exaggerated as strings of words are produced, but the sense is clear.

Despite this, little evidence emerged from the present survey that music was being used systematically to support, enhance or maintain speech, although one child was reported to be taking singing lessons to stimulate speech production. However, work in the UK, developed in the 1980s at Linden Lodge School in London, with children with visual impairment and additional disabilities, including

Figure 17.2 Songs for social expressions

children with JNCL, pioneered the use of "micro-songs" – key words and phrases that were set to short melodies (Ockelford, 1994) (see Figure 17.2). These were taught to children in the course of music sessions as part of social games, and children were subsequently encouraged to transfer them to real life situations to be used functionally. In some cases the teachers supported the production of words by children who were in the early stages of speech, or had acquired articulatory difficulties through brain injury or disease. In other cases, the micro-songs served as a proxy for words – for children who were unable to make speech sounds but could nonetheless vocalize. Here, the rhythm and, in some cases, the contour of the melody, was all that was required for the meaning to be clear. Key factors in the success of this use of music were that all those involved in the education and care of the children concerned were aware of the songs and their potential meanings, and that teachers and others were prepared to employ them consistently themselves, to model and reinforce their use (Ockelford, 1996). Here, longer songs incorporated key words and phrases that consistently used the same melodic shape and rhythm – characteristic motifs that could be extracted for deployment in everyday situations (see Figure 17.2)

There are many anecdotal accounts that young people with JNCL – like other young people – enjoy singing and music, but there is hardly any research on the potential impact of music therapy and education in this group. Phase I of a new UK research project termed *Music in Neurodegenerative Disease* (MIND) is examining the potential efficacy of music to support communication and thereby enhance the life of young people with JNCL.[1] The project includes the creation of a library of 39 potential "micro-songs" and Figure 17.3 shows how these are organized conceptually. This scheme is currently being piloted with children and

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[1] Sponsored by the Baily Thomas Charitable Fund, and supported by The Amber Trust (www.ambertrust.org), and the subject of doctoral research by the second author.

young people with JNCL. It is anticipated that each person will use only a small selection of micro-songs to help with communication, and will start with the core. New songs will almost certainly need to be developed for individuals, based on preferences and needs.

The approach set out here is similar to techniques used in an intervention called Neurologic Music Therapy. These include Melodic Intonation Therapy (MIT) and Music Speech Stimulation (MUSTIM) (summarized in Thaut, 1999). MIT aims to facilitate spontaneous speech through sung and chanted melodies that resemble speech intonation patterns. The step-by-step process should be used over an extended period of time, and it may be helpful for family members as well

Figure 17.3 Map of "micro-songs" used in *All Join In 2*

as music therapists to adopt the approach to encourage expressive communication. MUSTIM uses musical materials such as songs, rhymes and chants to encourage language production through the completion of learned familiar song lyrics, association of words with familiar tunes, or musical phrases. For example, spontaneous completion of familiar sentences is stimulated through familiar tunes or obvious melodic phrases. It can be used as a follow-up to MIT, as an attempt to increase the number of functional spoken utterances that the individual is able to produce. These techniques are widely used by speech and language therapists and neurologic music therapists when working with individuals who have various forms of neurological impairments (see for example www.chilternmusictherapy.co.uk).

Music and movement

Music is related to movement in two important ways, both of which pose challenges and afford opportunities for children and young people with JNCL. First, hearing music naturally stimulates movement; indeed, some cultures do not separate the notion of "music" from the concept of "dance" (Lewis, 2013). Second, movement (of some kind) is invariably required to make music, whether through singing, playing an instrument or using specialist technology.

The close connection between music and movement can be used to assist those with JNCL, for whom physical activity of any kind becomes increasingly difficult as the disease progresses. For example, one parent noted that *walking … succeeds when she is listening to her favorite music and singing along* and another parent commented that music helped her son to be more coordinated and aware of his body. Although formal research into the capacity of music to function in this way for young people with JNCL has yet to be undertaken, it seems likely that some of the specialist techniques of Neurologic Music Therapy can be of benefit. For example, rhythmic auditory stimulation (RAS) draws on the brain's ability to entrain to the underlying rhythmic regularities that most music exhibits – enabling people to move the body in time with the beat of the music.

A number of songs from *All Join In!* (Ockelford, 1996) were designed to encourage and offer a framework for movement in blind children with other disabilities, and these have been extended in the MIND project. They include music to walk, rock, stretch and relax to. The titles of these songs appear in the upper left quadrant of Figure 17.3.

Producing music and sound is a sensorimotor experience that can be both felt and heard, and one that requires co-ordination and control when using musical instruments (Ockelford, 1998). For example strumming a guitar or pressing the keys of a piano demands finger dexterity and strength, while beating drums and cymbals requires large upper or lower body movements to create sounds. Vocalizing, singing or blowing instruments regulates breathing and encourages people to exercise breath control and strengthen their respiratory system. Whatever movement that is required, the driving force is intrinsic motivation: the will to move the hand derives from the desire to make a certain sound.

The challenge for professionals working with young people with JNCL is how to offer them access to music-making as physical coordination and strength diminish. One option that is being explored in the MIND project is to use MIDI-based technology through which switches and beams can be used to control sounds generated by means of a computer (see Chapter 19). This means that even the tiniest movement can be used to trigger a whole orchestra of instrumental possibilities. In terms of research, currently the most relevant in-depth study pertains to Abby, a young woman in the late stages of juvenile Tay-Sachs disease (Ockelford, 2012). In the nature of its progressive mental, physical and sensory decline, Tay-Sachs is very similar to JNCL – indeed, the two were often confounded in the past (Walkley, Siegel, & Dobrenis, 1995).

Abby's story

Abby's illness had started to take effect when she was two years old. By the age of seven, she had lost the ability to speak, although she was still able to sing the words in songs for some time afterwards. The challenge of Abby's music teacher in the last stages of her illness, when she was unable to vocalize or grasp a conventional musical instrument in order to manipulate it, was how to enable her to engage proactively in her class music sessions. He tried a technological approach, using the OptiMusic® system. This works by emitting beams of light, which, when broken at different points by a reflective surface, are detected by a sensor, and cause corresponding MIDI signals to be sent to a digital library of sounds. The reflective surface was provided by an ultra-lightweight plastic paddle, which, with some preliminary assistance, Abby could grasp with her right hand. It initially rested on her lap, and the beam was calibrated so that a movement of only two or three centimeters was sufficient to trigger a note. The system was set up so that Abby had a melodically pleasing series of pitches available to her (C, E and G). The music teacher led the supporting staff in improvising a song that used Abby's notes, and provided a keyboard accompaniment. The results were remarkable. Having been unable to produce music herself for a number of years, in only a short space of time, Abby showed that she was indeed able to participate in music-making, and produced sounds with increasing vigor as the song progressed over a couple of minutes. Most surprising of all, she could not only keep in time with the beat, but she was able to imitate motifs from the tune that the staff sang (her achievements can be seen at www.soundsofintent.org).

Music to mark time and provide information about activities, places and people

In the present survey, a number of parents and professionals mentioned that music was used throughout the day to mark out key events, and to provide emotional succor at potentially difficult times. For example, one young person with JNCL had *set songs for going to the toilet and brushing her teeth*. Another invariably listened to music in the morning before school, to ease the transition.

> *Music is used every day. He has special songs to fall asleep to; different songs have been used in different situations (pee song, wake up song, be together song).*

Beyond this group, work with children who are blind and have learning difficulties over the last three decades has shown how music can be used systematically to structure the environment using auditory support, such as adding clearly defined and consistent sounds to rooms and other areas to compensate for the lack of visual information available to the individual (Ockelford, 1998, 2008). For example, brushing past a particular set of wind-chimes may indicate entry to a pupil's classroom; the same relaxing music may always be played on entry to the multisensory room; and a tuneful door-chime may be used to remind the individual that he or she has come home. At the same time, it is important to reduce "auditory clutter" that may function as distraction: classrooms can be very noisy places! Sometimes, moving to a smaller, quieter space may facilitate the pupils' concentration.

Key activities may be initiated using sounding objects: a small bell for music, for example, a larger one for gym, or the rattle of a chain for an outdoor play area. Such objects can be used to make "auditory timetables", so that students can be helped to anticipate a morning's activities, and can reflect on them once they are past – perhaps by placing the objects in a series of boxes whose lids are closed, in order to indicate completion. The identity of people too can be enhanced through the use of different sound-makers such as jangly bracelets, as well as micro-songs (see Figure 17.3).

Music to encourage socialization – structured and informal

Engaging with music, whether as a listener or active participant, offers an interest that all young people can share with their family and friends. Developing strong musical preferences in adolescence, and sharing these with peers, is a normal part of growing up and developing an identity that is at once unique and indicates belonging to a group of like-minded peers. The comments in the survey suggest that this is just as true for young people with JNCL as it is for young people without disabilities. For example:

> *My daughter has loved music all her life and still does. Singing was very important to her and sharing music with friends; she used to want to record music on cassettes to give to her girlfriends … it was a social interaction too.*

From an educational perspective, one professional observed:

> *Through the morning gatherings with music every day at school, the child has shown development both socially with other pupils and has been able to communicate.*

However, the parents cautioned about possible dangers. One parent emphasized how music could be misused, as a substitute for social contact, which had a detrimental effect on the care of her son with JNCL.

> *Music has been something he has been good at since he was a child, and it gives him a lot of pleasure. But it can be abused by the carers in the sense that they think they can just play him a CD instead of spending time and communicating with him. He likes being talked to by the carers, even if he doesn't verbally respond. It's a really big challenge with a lot of unskilled workers.*

Some proprietary music technology programs also offer an effective way of engaging some individuals in music-based projects with their friends or peers, although using these independently in the absence of vision can be difficult. Nonetheless, when working as a group with sighted peers, young people with JNCL may be able to choose and build up loops, beats and sounds, and contribute to song-writing activities by simply indicating their choices through words, vocalizations or gestures. Other forms of song-writing are possible too, in which the song words are changed to relate to specific interests or topics that are relevant to the individual with JNCL.

Music can also be used in more formal educational and therapeutic contexts to facilitate social contact, scaffold interaction, promote turn-taking and listening to others, and to promote participation. Examples are outlined in Figure 17.4.

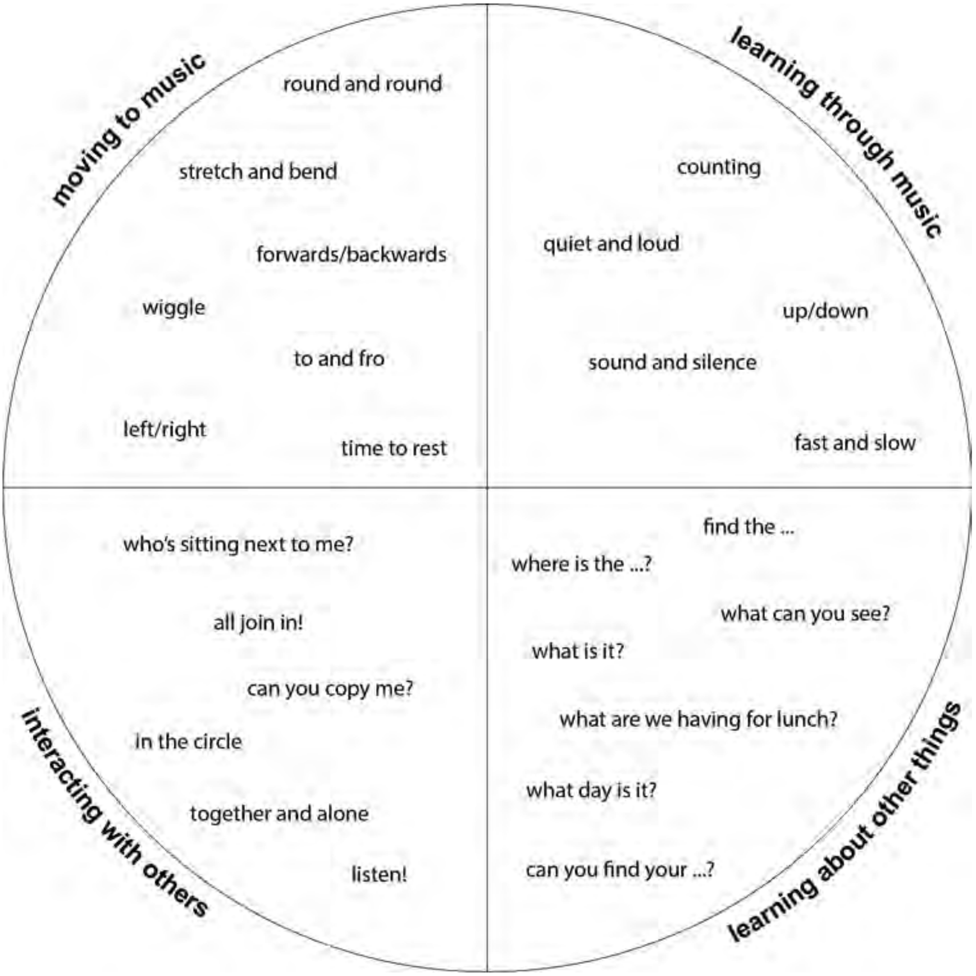

Figure 17.4 Map of songs to encourage wider development, understanding and wellbeing used in *All Join In 2*

Laura's story

Laura is 18 years old and lives on a supported campus in the UK, which is specifically set up to support young adults with JNCL. However, she regularly sees her family for trips, overnight stays and when attending church together.

Laura has a passion for music and enjoys listening to her vast music collection in her spare time. She particularly enjoys Disney songs, Avril Lavigne and Leona Lewis. She has music therapy sessions each week, and brings with her a large folder full of music for the music therapist to sing and play with her. As with many other young adults with JNCL, Laura's working memory is poor, and she often asks the same questions repeatedly in order to confirm events that have happened recently or may happen in the near future. Her long-term memory appears to be more intact, and in therapy sessions Laura has an almost errorless memory for singing familiar songs, and for melodies that she used to play on the piano as a child. She will even correct the music therapist regarding the exact number of choruses in each song.

Laura.

In order to sustain and nourish these memories, the music sessions focus on using both familiar and improvised music. During one session, Laura was taking great delight in beating on a large drum to the words "a crash of drums" and softly sweeping the wind chimes to represent "a flash of light" during a rendition of the song *Any dream will do* from *Joseph and his amazing technicolor dreamcoat*. Through this interactive activity she was creative, playfully painting the song with musical sounds. This led Laura to think about how music and instruments can portray other stories, discussing what sound and music could be used to depict the characters and actions within them.

The music therapy sessions provided Laura with a motivating context in which to rehearse skills that might help in her everyday life. By engaging in activities that involved choosing instruments, sounds and songs, Laura was encouraged to maintain her ability to choose and to communicate these choices – allowing her voice to be heard. Through song-writing activities, Laura was given the opportunity to create new materials, involving verbal comprehension, planning, organizing and problem-solving. Put simply, music was the mainstay of Laura's life.

Music and memory

Music can fulfil a special function in helping to revive memories. For example, one professional reported that *familiar pieces of music … often sparked a memory of events from the past,* while another observed that *well known songs promoted his memory, comforted him and made him feel fine*. A third professional noted that:

> *Music is one of her greatest interests, and she remembers several lyrics because she has sung them so many times … the music helps with her memory.*

A parent noted that listening to music helped the daughter to remember:

> *If she went to that particular concert, she would remember what the group were wearing or doing during a particular song. She will also remember what we did before the concert or any funny things that happened. This is really important for us as parents, as it helps not only with her communication but also her long-term memory.*

Several parents mentioned that among young people with JNCL the songs from childhood seemed to be enjoyed and remembered.

> *Music was the most important activity in his life. He listened to music every day, played his drums and sang his favorite songs. His music taste changed later years towards children's songs.*

Those supporting young people with JNCL can ensure that the music associated with pleasant and important memories can be retained by putting together "banks" of songs and other pieces, together with notes on when they were heard, and other details of the occasion, to help prompt recall (see Chapter 13). In this way, music can serve as a "bridge", as one parent put it, to a past reality, in the final stages of the disease.

Concluding comments

For individuals with severe visual impairment, hearing is the main compensating sense. Although most children and young people listen to music in many situations, the lack of vision can mean that music is even more important for children and young people with JNCL. There is, however, limited research on the educational use of music and song with this group, and it is not safe to assume that all the findings from research with other groups can be transferred to children and young people with JNCL. It should also be noted that research on music-based interventions for elderly people with dementia suggest positive effects of music in some areas, but results are mixed (Koger, Chapin, & Brotons, 1999; Raglio et al., 2015; van der Steen et al., 2018; Vink, Bruisnma. & Scholten, 2003). The present survey demonstrates the importance that parents and professionals attribute to music activities for children and young people with JNCL. This emphasizes the need for more research in this area.

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18

Musical Action to Support Children and Young People with Juvenile Neuronal Ceroid Lipofuscinosis

Jochen Lippe Holstein

> *As I enter the classroom, once again Marion sits completely limp in her wheelchair. School days have become very stressful for her, so she often lies down on the couch in the classroom. In the music room, I play her a new piece of music. The improvising soprano saxophone seems to remind Marion of the ducks at home, because she is particular happy about this sound. Together we dance to the music whilst standing. The dance, in which I let myself be completely guided by Marion, is getting wilder: She jumps and skips with all her strength, whereby she completely exhausts herself. We sit down again – now I give her a duck pipe which she uses to respond to the soprano saxophone. This dialog is also becoming increasingly fierce. To regain relaxation, we let the ducks fly away (becoming quieter as they fly away) and listen to quieter music whilst sitting, in which the saxophone also plays softly and slowly; we sway to the rhythm of music.*

Marion loves music. My experiences with her are an example of the possibilities to support children, adolescents and adults with juvenile neuronal ceroid lipofuscinosis (JNCL) through «musical action». This chapter discusses how joint action within the music context with a degenerative disease may be an appropriate and meaningful support for development and maintenance.

Musical action

Human beings are characterized by having complex cognitive abilities. They are able to memorize experiences of the past, find relations and link them with other experiences. These experiences result from acts of purposeful activity and from managing the everyday world. Through these actions, human beings acquire knowledge about the world (Piaget, 1952), and the ability to take action is thus the basis for the identification ability. Conversely, due to earlier experiences, it is possible to develop a differentiated ability to act. It is possible to observe the abilities of the individual and define goals that he or she may be able to achieve. These abilities are here divided into the domains of perception, movement, expression and communication. However, they are closely related, and influence and depend on each other (Amrhein, 1993).

Musical action means the arranging and the experiencing of musical contexts: playing, listening and responding to music. The everyday subject music is both a cause and an aim of perception, movement, expression and communication. Music addresses these four abilities in a special way, and it may become a common and wonderful subject for communication.

In this chapter, several aspects of musical experience are discussed. Some neurophysiological and psychological assumptions are elaborated. This distinction is purely for methodological reasons, because music is always experienced individually *and* in a context – «music as a whole».

Music as sensory-motor experience

Music is not solely an acoustic phenomenon: perceptual information arrives through the auditory and tactile senses of the experiencer. For example, the stimulation obtained from an orchestra playing is experienced simultaneously through the sensory cells («hair cells») in the cochlea, within the ear, and through sensory receptors on the skin activated by the air pressure from sound waves generated (vibration) from musical instruments. Music can therefore be both felt and heard. Someone playing music also receives additional haptic information from the musical instrument itself (e.g., holding a mouthpiece against the lips, or plucking or bowing strings).

From a neurophysiological perspective, perception is not limited to the sensory afferent of environmental stimuli. An efferent motor order is often the result, which is carried out either deliberately or involuntarily, as a reflex or automatic response. It appears to be a close link between sensation and motor activity (see Gibson, 1979; Liberman and Mattingly, 1985; Guenther & Vladusich, 2012). In addition to the heteroceptive perception (on the outside), the human organism also has a proprioceptive perception, a sense related to self-awareness, such as balance or sense of movement (the vestibular and tactile-kinesthetic sense). Heteroceptive hearing and proprioceptive self-awareness are related structurally and functionally through anatomical proximity.

Whilst music motivates movement, it moves humans in two ways: The person feels the tactile-kinesthetic vibrations and the movements of his body parts, the vestibular and the spatial variation in his body, and is moved emotionally!

Moreover, a noise or sound can only be produced with movements. Thus, to make music, targeted and coordinated movement is necessary. Music speaks to the human organism in a diverse sensory and motor activity interrelated manner, very unlike other contexts of actions or action objects. Music is perceived as sensory-motor activity in action.

Music as personality of action

With music, I here mean music that originates from humans and where material is consciously brought to make the sounds which are used for the subjective view or subjective expression. In each case, the phenomenon of music is malleable, since it is produced and perceived by humans and thereby structured. Emotional content is perceived as sensory-motor as well as cognitively detectable structures, object-like features can be illustrated, associations are activated, music challenges the person to make a statement. When the individual has visual impairment, differentiated acoustic perception provides important information about the environment. The distinction of objects as sound sources is also facilitated by the recognition of (cognitive detectable) structures, which also facilitates spatial orientation. The sound quality provides further information about the nature of the sound-making object. These qualities can be used consciously to express something specific in a music-making process. The malleability of music gives the person opportunity to express himself through musical activity, to express emotions, illustrate thoughts and thereby visualize and process. An emotional expression can be put into a piece of music, or it can be experienced subjectively within a piece of music.

Music as an independent form of communication

Musical action can be understood as a communicative process, because between the music interaction partners, defined and definable symbols are purposefully replaced, similar to language.

However, a special feature of musical communication is that there exists a possibility for the person to develop his own symbols, or even intersubjectively to define symbols with instrumental or vocal sounds. As a result, a relationship can be established between the music performers, something can be expressed or described through music (Reinecke, 1982).

In contrast to verbal communication, the focus is often a more idiosyncratic expression with musical and non-verbal communication: There is no right or wrong information, although culture-bound evaluation criteria may be created in regard to embodiment and performance shape. In musical action, communication can take place on a very basic level. For example the music therapist may take over the rhythm of the other's breathing, start to make contact and thereby put forth an equal level. The music therapist may change the rhythm playfully, insert a new aspect in the relationship, to which the partner can consciously respond (stay at his own pace or acclimatize himself to the new rhythm). Musical dialogues can be highly differentially designed, if both players adjust to each other, respect common rules and can use differentiated skills and abilities. In any case, through musical action the participants create call-and-response sequences in which an action (e.g., an utterance or rhythm) of one participant evokes a related reaction (e.g., imitation of variation of that utterance of rhythm) from another participant. If both participants refer to the same rules, musical action can result in an expectable communication – or a surprise is deliberately presented by consciously violating the expectations. In any case, both people are actively involved in designing a communicative action.

Unlike linguistic communication, a simultaneous polyphony can even be desirable and consciously used to express something condensed or parallel. Musical communication therefore has usages other than verbal communication. It can approximate and mimic phonemes – music can however deliberately use the differences to supplement the verbal action.

In a group context, musical action provides a very diverse communication frame for the fellow players. They can focus their attention, perception and movement on a common content, in order to play, listen and experience music, and coordinate their movements in such a way that a uniform process can arise. It can be a wonderful experience when a whole group of people is so closely related and involved in the same process!

Nurture despite degenerative disease

The aim of educational activity is to support in the individual a sense of differentiation of skills and abilities. Strictly speaking, this means the differentiation of each ability in action, that is, the ability to act purposefully in a specific context. Jetter (1987) defines the ability to act as «The ability in the given situation and for the anticipated living conditions, the greatest possible solidary use of natural and social conditions for the purpose of a multiplicity of experiences indulgency, according to the physical and mental needs and the ability to aesthetic-communicative shaping of the life situation» (p. 79). Jetter refers to the work of Piaget (1952), who described development as a dynamic connection between the individual and the environment as follows: Every human organism strives to live in harmony with its environment. The young child forms structures of action schemes, in order to adapt to the surrounding conditions (accommodation), and then again in order to assimilate the environment to his own structures (assimilation). In the young child, these two adaptation processes run as simple activities of the actual need of satisfaction. With further development, conscious, planned action shapes the more complex connections between the individual and the environment. The changing conditions of the environment (in Jetter, these are the «natural and social conditions») are perceived and taken into account. Likewise, the structures of the developing individual change continuously. When a child or young person has JNCL, the structural change involves a reduction or loss of already acquired possibilities for action (see also Chapter 2). The degenerative disease is thus to be understood as a component of the «given and expected living conditions» (see Jetter, 1987) of persons with JNCL. Their change is also development, as adjustments need to be made, but they also need support. By motivating pupils with JNCL in their subjective and actual reality – that is, their ever-changing capacity for action – the teacher encourages purposeful activity.

Music as an occasion and goal for common action

As explained above, growth is achieved by appropriately addressing the abilities of the individual within a context, which he recognizes as valuable. Music presents itself for shared action, since it can be experienced in different and diverse ways. This ensures that musical action, even under changed and further changing conditions, in particular with the limitation and loss of abilities, is a valuable mind context over a long period of time. Between the educator and the student, a private music or a private musical understanding can arise. The structures can only be recognized and understood by them, as they reflect their personal relationship. Changes in the musical capacity for action therefore do not have any hindering effect on the quality of the mutual musical life. For example, in spite of limited motor possibilities in the mutual sense of the time, music can be played together, interdependently (see Chapter 16). The pupil may play only every fourth note while the teacher continues to play the complete piece, but they play together and the pupil is actively involved and experiences himself as effective and with equal entitlement.

If the orientation to objective assessment criteria within a musical context (playing the piano correctly) represents an excessive demand or a motivation obstacle (especially when playing well-known pieces of music), the rich range of musical trends and styles can be relied upon at any time, allowing each person to choose the music which appeals to him. In musical improvisation, for example, subjective assessment criteria can be implemented without the impression that the result is a makeshift product or something provisional. Musical improvisation is therefore particularly appropriate, because subjective changes in the game that result from the disease-induced change in the ability to act can be sensibly absorbed.

Promoting the general capacity to act through musical action

Humans receive knowledge about the world through the active experiencing of parts of this world. In this case, music is such a part, which can be experienced through action: «In acting [that is, through the purposeful use of the capacity for movement, expression, perception, and communication], the subject reveals that the 'outer' reality music is accessible and understandable, and so on. At the same time, the inner 'subjective musical reality' – the ability to move, express, perceive and communicate – is developed»(Amrhein, 1993, p. 571). With regard to Jetter's (1987) definition of ability to act, music can be regarded as a «natural and social condition», because it is experienced as an acoustic and tactile phenomenon, as a component of nature, and, on the other hand, as an eminently shaped component of culture. The notion of «experience of multiplicity» as the purpose of capacity for action has already been discussed. Jetter emphasizes the physical and spiritual need of this experience and that the «ability to aesthetically and communicatively shaping of the life situation» is evidently promoted through musical action.

Positive influence of the psychological experience through musical action

The damage to the nervous system in JNCL (see Chapter 3) does not lead only to impairment in motor and cognitive domains. Children and adolescents with JNCL also notice accompanying changes that occur. For example, they experience multiple losses, such as the loss of already learned or acquired abilities and mastery of activities, as well as loss of social contacts and ties. They also experience the changing responses from the environment towards them, such as disillusion, worry, anxiety, anger, perplexity, and constant observation and classification. Since they recognize the cause of these changes in themselves, they also often feel responsible for the reactions of their surroundings. Moreover, they become massively unsettled when information about their future is lacking or unclear, and fears arise.

It is often observed that taboo subjects, such as illness and death, are avoided or not dealt with by everyone involved – the young persons with JNCL as well as significant people in their environment. Since there is never the opportunity to talk about such serious topics in everyday life, young people's concerns (e.g., fears, anxiety, uncertainties, misunderstandings) arising from these topics always remain diffuse and are never openly addressed. If at a later stage, the ability to speak or use language are lost, this situation is further intensified, because the young people's unaddressed concerns remain and are compounded because it is now more difficult to address them openly.

Therefore, it is necessary to create a framework within which crisis topics can be dealt with. Conditions for this are a trusting relationship and an appropriate medium of expression. Musical action allows an active symbolic examination of one's own thoughts and fears, especially when the medium of language is increasingly impaired. The aim is to address such topics and prevent or take away the paralyzing hopelessness and menace, by showing confidence and confirmation in the present moment, which can contribute to a positive evaluation of the young person's identity and development.

How to make music together

First of all, I would like to emphasize that every offer of action must be in a context that is meaningful to the pupil and within the pupil's subjective reality, so that he or she can use their current competences. Depending upon the severity and extent of the disorder, the body and its tools can be recruited to make sound, for example in rhythmic breathing or knocking games (anticipations and reactions in the form of imitation or variation), or in imitation of noises or operations, thus allowing for whole sound stories to occur. When participants are singing songs, they can invent sensory or rhythm-supporting movements.

The voice is the person's most obvious medium of expression, and it is usually used for speaking and singing. In speech, the content is relayed by means of words which are intended to represent what is rationally detectable in a conventionalized manner (e.g., in statements, questions, or commands). However, the spoken statement can be modified by paralinguistic means (prosody, patterns of intonation, modulation of loudness and pitch), and non-linguistic means of expression, (e.g., kinesics including gesture and facial expression; proxemics, such as spatial closeness; and chronemics, including the relative timing of communication). When meaning is conveyed through the medium of singing, the non-verbal aspects employed are of special importance. The sounds of the voice (i.e., patterns of intonation, modulation of loudness and pitch) can be used to express, for example the personal significance of events, objects and people, by using them phonetically. Conventional and homemade instruments lend themselves to taking musical actions, such as exploration and experimentation. It is also possible for participants to perform the same reaction and imitation games as in the breathing and knocking games, and also to create a rhythmic accompaniment of songs or other music pieces. For this purpose either drums or other rhythm instruments or (bass) tones are useful for harmonic-rhythmic song accompaniment.

Listening to music motivates music-related movements. These movements can be used to represent parameters of the musical process – including structure, dynamics, tempo and pauses. The vibrations of large gongs or basins or of bass instruments are perceived as pleasant and relaxing, if they are embedded in that kind of context, for example telling a story or fairy tale. The perception of these vibrations usually stimulates responsive movements. These movements are experienced as a meaningful activity within this situation. It is quite pleasant and meaningful to just listen to music. This also gives the opportunity to be «completely within the music» or «completely by oneself.»

Examples of musical action

The next section presents situations of shared musical action with two young people who have JNCL, «Klaus» and «Marion». Their current competences, the related goals and the teaching situations are described and discussed.

Klaus

Klaus is 14 years old and attends a class in the department for multiple or severely disabled children in the National Education Center for the Blind. He is a very lively, daring and communicative boy. By now, most of the time, Klaus makes a well-balanced impression since he feels very comfortable in his class. Some days he is rather obstinate and more confrontational than usual. When I first met Klaus some years ago, I noticed that he was tense and displayed a certain aggressiveness. I put this down to the fact that he could not accept his new situation. He had recently been moved from the mainstream part of the school to the part for people with multiple disabilities. In his new class, he had little opportunity to communicate his knowledge or even to compete with other pupils, since there were no equal conversational partners among the other pupils in the class. A constant, self-produced performance pressure could not be satisfied. After a further change of class to the current stage of the lower classes, more favorable conditions were created for Klaus. He was able to establish appropriate contacts with peers who were at a comparable intellectual and emotional level.

Klaus is inquisitive, but he also likes to share his knowledge with others. For every topic addressed, Klaus first tells me what he knows, and it is often quite extensive. Klaus knows something about everything, and he can make connections from anything to topics that are familiar to him. However, like many adolescents with JNCL, Klaus has significant difficulties with speech. He talks very fast and blustering, deletes or reduces words and syllables, repeats himself frequently, and sometimes does not find the right words. Klaus can orientate himself and can generally walk without any external support. However, when he walks, a knee flexion is noticeable, which may be observed in most young people with JNCL. Occasionally Klaus has a problem keeping himself on his feet. Increasing knee flexion when he walks appears to compromise his balance. In many situations, his muscle tone is increased and his movements appear tense. He often sits with raised shoulders and crossed legs, and his hands clasped under his thighs. To demonstrate power and compare himself to others are important to him. Klaus has good fine-motor skills and is motivated to use his hands.

Support goals

Helping Klaus preserve his self-image is a priority when one is working with him. To do this one must ask numerous questions: How does Klaus see himself in his development? What goals or ideals does he pursue and why? Why does Klaus claim that he can see, although this is actually not true? One goal consistent with preservation of self-image might be to look for possibilities of physical and psychological relaxation for him, since in the long term both will be of increasing importance. In order to be able to adequately counter possible instances of restlessness, it is of great benefit to know the individual relaxation preferences or framework conditions.

In addition to the goals mentioned above, which primarily aim at his mental well-being, is a goal to support him in maintaining a certain range of movements. Movement can be for fun or as a means of pursuing a goal. However, maintaining gross and fine-motor functionality, whether for mobility or participation in activities has special relevance to the later need for care.

The individual sessions with Klaus are designed in such a way that either a range of materials or the general framework of the treatment remains the same for an extended period of time. The pre-setting of action topics or action material initially opens up a common scope, which can then be designed individually.

A story as an occasion for musical movement action

Reading a story aloud functions as an opener for presenting topics and tasks for individual sessions. Today, a story that stretched over half a year ends with a festival of sports activities. «The butterfly workshop» by Gioconda Belli (1994) is about the origin of many species, and to celebrate the creation of the butterflies, a feast is arranged with self-invented program themes, including «Dance of the Butterflies», «Tug of the Spiders», «Boxing Match of the Kangaroos» and «Soccer

Game of the Moles.» Activities included in the last two program themes are described below.

In «Boxing Match of the Kangaroos,» a boxing match is symbolically represented by beating the fists or using mallets on a timpani (kettledrum). Klaus and I are sitting opposite each other. At first Klaus plays (punches) with his hands: quietly and slowly or loudly and fast, and then I change strategically, mimicking him. The roles of being initiator and responder change equally. The relationship to a real boxing match and, above all, the idea of kangaroos boxing, has a motivating and stimulating effect. Klaus tries out different techniques – on the drum ring (box ring) and on his own chin, in order to experience the effect of a drum beat.

«Football game of the moles» is played in a self-made carton football field table. We both try to flip or shoot a marble with our hands in the opponent's goal. The game is characterized by rapid changes between fast and quiet phases. Klaus locates the sphere partially by using his hearing, partially with searching hand movements. The «football lingo» is of great importance to Klaus. The relevant accompanying comments, shouts, and so forth, recreate a real football game.

Search for expression

One day, before the start of an individual session, I was sitting at the piano and improvised a little before Klaus came to the music room. He asked me if I could play something sad. I did. Klaus, at that time very restless and hyper in his mood, listened quietly at first, then he told me about deaths in his circle of relatives and acquaintances. He asked what happens after death, and how or what death is at all. Then I was reading some sections from the «Letter to a child with cancer» by Elisabeth Kübler-Ross (1979). The illustrative examples (seasons, day and night of Earth) seem to be understandable to Klaus. He questioned the examples and showed thereby that they made sense to him. Klaus himself does not use music to express sadness; for him, making music is something powerful and joyful. However, he listens to melancholic music and therein finds the desired expression of his feelings.

Marion

Marion is 16 years old and goes to middle school. She attends school irregularly because of frequent seizures and a growing demand for rest. Marion has long been dependent on the wheelchair for mobility. With maximum support she can walk small stretches or dance or rock to music, which she likes very much. Her posture is JNCL-typical, with forward inclination. Without a backrest or other support, she does not have sufficient support for sitting, and sitting unaided is hardly possible. Marion is stiff in the joints and each movement is met by counter-pressure. Activities are often exertive, even lifting her head requires a great deal of effort. Therefore, after most movements she returns quickly to her flexed posture. In her classroom is a sofa on which she spends more and more of her time, as sitting a long time has become very demanding. Marion frequently appears tired or absent-minded.

Marion's comprehension of words has developed in accordance with her age. She understands every message and request, even ironic remarks. Based on her reactions, it is evident that she follows the topics and forms her own opinions. However, her expressive language is less functional. Marion speaks very quietly and gently, and seems to lack the air to speak. It happens often that the air is not sufficient for her to reach the end of a sentence, so that the last words are incomprehensibly weak or not pronounced.

Although Marion is very quiet and reserved, and hardly able to express herself verbally, it is apparent that she enjoys being with other people and experiencing things that interest her. When asked if she wants to do this or that, she often reacts with a shrug, but sometimes she very enthusiastically agrees to a suggested activity. Especially when she is part of a group and involved, her joy can be observed. Marion likes to accept certain offers. Likewise, she is very fond of being «part of the scene,» she is happy when many people around her do something that she can be part of, usually meaning listen to.

Support goals

In the work with Marion, the focus is her ability to move, in order to maintain her expressiveness and scope for action. Through the active participation in situations that are meaningful and interesting to her, many movements and forms of communication may become activated. Since Marion cannot express all her ideas in speech, debates in the form of conversations are not appropriate for her situation. Under certain circumstances, topics which she would like to address may be torn, because of her restricted ability to speak, and this could lead to frustration. That is why the sessions are used to develop our own communication symbols, by means of which an inter-subjective expressive possibility is created.

Musical participation history

One day, I had prepared a participation story for Marion. The content of the story was built on Marion's interests and preferences, so that she could make a connection to the storyline. Through the following activities, she was given participation opportunities for action: different animal sounds were produced, the story ended with a joint celebration of animals and dwarves, which Marion and I use for dancing. Marion enjoys dancing very much, she is able to stand when support is provided.

The individual sequences – how the animals are represented with sound, how the animals react to certain events – are made so that Marion can take part independently. Marion replies to me according to her possibilities in the form of imitation or variation. Of particular importance for Marion is the representation of the ducks. Her parents are breeding ducks at home, which is why Marion has an exact idea of how these animals sound for example when they are excited. We use a duckling pipe to produce the typical quack-cackle sound. However, it contributes only a sound representation: Marion represents a duck with the whole body. She expresses «duck» on the basis of many duck experiences. Marion, who usually is very quiet, speaks gently and sits in a bent position, raises herself to fill her lungs so as to give the ducks a voice.

Improvisation around annoyance

When I picked up Marion from her classroom one day, she was sitting listlessly in her wheelchair, punching without interruption – but also without power – on a pillow filled with sand. She reacted to my greeting, but continued with the treatment of the pillow. In the music room, I asked her if she is annoyed. She answered «yes.» In the further conversation, I did not find out what had made her so annoyed, but we had chosen «annoyance» as the topic of the session. The sand sack was treated in a variety of ways, such as stroking it like a beloved guinea pig, and beating it like it was something dangerous, and soon drums were drilled. In the meantime, Marion had freed herself up by playing. She sat upright in the wheelchair, with raised head and a concentrated and content facial expression, enjoying the volume that she caused. We took turns bashing and stroking, and soon the drumming developed into a musical act. We took turns with drum whirls and signals – we made music together.

Conclusions

Musical action is a pedagogical principle that provides suggestions for the education of children and young people with JNCL. The stories presented here illustrate the ways in which pupils' interest areas and abilities may be used to establish joint musical action. The beauty of the music, and the use of its formality are clearly visible in the representation of the animals and their activities in the stories. However, even in search of expression and in improvisation, the capacity for personal expression, including the expression of annoyance, was only possible because it could be found in the music elements and transferred into the children's own musical actions. What the two pupils really felt was malleable and can only be observed by an outsider by looking at their actions. The fact remains however, that the two pupils could find expressions of sadness or annoyance in the music. Marion expresses her anger through these musical structures. The exchange between the pupils and me did not only take place at the vocal verbal level. In the boxing match, we spoke mostly with music accompaniment, after that, the game was reversed, thus accompanying music-playing. Musical parameters (e.g., dynamics or tempo) determined the course of the action, not grammatical, syntactic or other linguistic parameters. Likewise, a chaotic and two-part drumming session represented Marion's improvisation of anger excellently, perhaps even more appropriately, with greater social acceptability than would be possible to express through language. The sensorimotor experience of music in musical action is always recognizable. This is most evident in scenes where dynamics and tempo are experienced physically as tension and relaxation, such as in boxing or experiencing the duck. However, even in the quiet football game, the rolling of the marble was sensory-motor experience.

In musical action there exist possibilities for targeting therapeutic goals by exploiting earlier experiences that the child recognizes. This therapeutic strategy is motivated by the observation that pupils perceive the musical action in itself and recognize its value. The encouraging – and probably also motivating – common factor in musical action is that different people orient themselves towards a goal, influence each other in their actions, and thereby enrich each other.

A final aspect of musical action of great importance is that musical experience is possible even when the neurodegenerative process reduces motor action possibilities!

Note

This is a revised version of an article in «JNCL. The living situation of blind children and adolescents with an incurable degenerative disease» from 2001.

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19

Technology in Education

Mohammed Beghdadi

Technological advances have resulted in an extended use of technology in many areas, including education (Craft, 2012) and rehabilitation (Cook & Polgar, 2008; Scherer & Glueckauf, 2005). Technology is used to support and maintain sensory functioning (Edwards, 2007), motor skills (Henderson, Skelton, & Rosenbaum, 2008), and cognitive functioning (Carey, Friedman, & Bryen, 2005) in children as well as older people. The aim of introducing technology in special education is to adapt the teaching to the needs of each student. Adapted teaching does not necessarily imply a particular way of teaching, but rather a situation that is characterized by certain qualities that make students and teachers develop together. The teachers put themselves in the students' situation in order to acquire information that will improve the classroom situation for students with special needs (Buli-Holmberg & Ekeberg, 2008). The development of the students' competence, coping, self-determination, co-operation and appropriate work habits is crucial for successful learning.

The present chapter gives an overview of assistive devices and tools that may be useful in teaching students with juvenile neuronal ceroid lipofuscinosis (JNCL). The devices and educational tools presented here may be useful for students with JNCL but most of them are not designed with this group in mind. Children and adolescents with JNCL have generally had very limited access to computers and teaching aids that are tailored to their particular needs. The combination of visual, motor, language, and cognitive impairments and challenges in social interaction makes it difficult to find an appropriate way to cater for their needs. There is a need for more devices and tools designed for students with JNCL, and these may also be useful for other people with disabilities.

The tools presented here have two basic purposes: to compensate for declines and other educational needs and to support participation. Tools, including electronic braille displays (see Chapter 14) and devices with voice recognition and production, can serve to compensate for declines in visual, cognitive, language and motor functions. The compensation thus provided can in turn provide these students with support in participating in educational and other activities at school, at home and with peers outside school, thereby stimulating them to maintain previous patterns of participation, and possibly encouraging them to maintain and extend those patterns even in the presence of decline. The use of technological tools in education may enhance the students' confidence and self-esteem, and function as help to better inclusion through independence and interdependence (see Chapter 16), and thereby to increasing the students' benefit of the teaching. The use of technology may also contribute to increase motivation, engagement and achievement efforts in areas where the students need support, training and practice. It is the experience of the present author that the use of technology and innovation in education may contribute positively to an interesting school day for the students with JNCL and give people in their environment a better understanding of the students' opportunities and abilities, for example when the student is using the specialized computer program Sarepta (see below). Technology is also used in games (see Chapter 21) and quizzes (see Chapter 20). Some of these technologies may also have a positive impact on adult life after finishing school.

Aims, possibilities and challenges in using technology

One of the main aims of using assistive technology for persons with severe visual impairment, including JNCL, is to make information available through touch and hearing (Freitas & Kouroupetroglou, 2008; Petty & Frieden, 2012). Assistive technology programs can be used to translate spoken language into digitized text, turn spoken commands into actions or magnify the text in a word processor, web browser, email programs or other applications. Assistive technology has contributed to removing many barriers for persons with disabilities within education and employment. Possibilities and goals within these areas are also valid for persons with JNCL. Students with JNCL can for instance complete their homework, do research, take tests, and read books along with their sighted classmates, thanks to advances in technology.

Assistive technology tailored for people who are blind makes it possible to organize and handle files and records containing text, music, audio-recorded memories or almost anything that can be stored in the electronic world. Assistive technology can also be used to support inclusion. Students with JNCL can for instance share the same classroom activities as their peers by using technology where visual information is transferred to sound or tactile information (see Chapter 14). Further, some programs and devices may be used in some situations to compensate for loss of speech, memory and mobility. Last but not least, use of assistive technology can provide entertainment and pleasure at all ages for persons with JNCL within and beyond education (see Chapter 23).

Assistive technology for persons with special needs is often associated with possibilities for independence. This is a mandatory goal also for children with JNCL when independence is still achievable. Independent use of assistive technology will however require understanding and skills related to the operation of electronic devices, and this will require a certain degree of memory and motor functions.

Technology-based information can be operated simultaneously or sequentially. Visual information allows simultaneous actions where segments are presented in a two- or three-dimensional hierarchy or structure. The user with sight can select an option by ticking the desired segment presented in the structure, a glance on the structure is enough to identify the correct option. To transform such structures to tactile or audio-based information requires a more complex strategy. First, pictures on a screen can only be transferred as time-consuming narratives. Second, tactile and audio-based information requires sequentially-based or consecutive actions where the user follows a string or succession to identify the correct option. This step-by-step strategy requires time and memory: the user must remember the purpose of the string, and previous actions to enhance the next actions. Sequentially based

Figure 19.1 Rolltalk Designer

Rolltalk Designer is a tool for communication programs for persons with disabilities. The program has a large amount of finished content, both symbolic and textual.

information in assistive technologies is a special challenge for persons with JNCL when memory problems are evident, a finding emerging from the experiences recorded in the current project. This memory demand might be a main reason why electronic communication aids (see Figure 19.1) are not used to a higher extent by persons with JNCL. In addition, there are significant demands on attention because someone using the technology must be able to filter out background noise, remain focused on the task and so forth (see Chapter 6).

Assistive technology is used in a range of areas and for a variety of purposes, such as performing daily tasks and activities, supporting social participation, monitoring health and improving safety; however it is not without limitations: Assistive devices can never replace human contact and interaction for persons with JNCL. Skills and functions will decline for individuals with JNCL (see Chapters 3–7) and independence will sooner or later not be obtainable. When that point is reached, *independent* use gives way to *interdependent* use, allowing assistive technology to continue to have an important and positive impact on life for persons with JNCL even when independence is difficult to achieve. Objectives related to sharing memories and pleasure, and compensation for loss of speech and other functions can be achieved by using interdependence strategies where assistive technology is used with assistance (see Chapters 16 and 23). The long-term goal of using assistive technology for persons with JNCL should consequently be less about its facilitation of independence, and more about its supportive role for facilitating participation and interaction through interdependence strategies.

Compensation through use of audio devices in teaching

Assistive technology can support functioning in the zone of developmental maintenance (see Chapter 2 and 11). As the disease progresses, children and young people with JNCL can compensate loss of vision with information received from touch and hearing. The present project (see Appendix A) found that the tactile sense tended to weaken earlier than the auditory sense (see Chapters 1 and 14). It is therefore important to consider assistive technology and teaching methods based on hearing.

The use of audio and video media in education has grown considerably in recent years, although not always with success. Constraining factors are lack of technical standards, technological expertise in schools, and technological and educational guidance and supervision (Craft, 2012). However, for students with visual impairments in general and students with JNCL in particular, the use of audio provides opportunities for improved learning and participation.

In early phases of JNCL, the use of audio for presenting texts should usually function as a supplement rather than as a substitute for reading and writing (see Chapter 14). Reading and writing are pivotal skills for further learning and development. In the present study (see Appendix A), some students with JNCL had learned to read and write braille or Moon, others only managed to write in the tactile mode, while some never managed to utilize tactile texts. With the support of a speech synthesizer this last group could nevertheless have access to written texts through listening. Moreover, hearing will sooner and later become the most important sense for participation and learning in all individuals with JNCL, and assistive technologies with speech synthesizers should therefore be considered as soon as possible after they have received the diagnosis.

The present study shows the importance of making sound recordings of early situations of special importance for the student, such as conversations, interviews, stories, and excursions at school, as well as presentations, jokes and expressions that the students used in different situations. Audio recordings can be used to preserve memories for the future, as in the following narrative from a teacher:

> *The student had used the program "Sarepta" for several years. As part of the pedagogy, the staff at her school had documented the student's school day by recording a lot of activities and school assignments. For many years, the parents recorded family trips, travels, birthdays and special adolescent events like the Christian confirmation on video and audio. She had a unique, extensive and comprehensive audio bank from her childhood and adolescence years, with material she produced at school, such as interviews, music and dance videos, workshops and school graduation celebration. When the student became an adult and her illness had reached the last stage, listening to these recordings was one of the few activities she could take part in. She responded to what was being said, expressed emotions and sang along to her old childhood songs. The recordings had now become a medium and a tool for communicating with her closest family and friends. The mother stated that this was the best activity they had together, where they could communicate and reminisce.*

Audio can function as an inclusive medium. Collective listening to sport, radio plays, audio books, poems or music gives students opportunities for sharing experiences with others. However, it is important to be selective in what to save for the future, and how audio recordings are saved and stored for the future, how the files are edited, labeled and categorized. An overwhelming number of audio recordings, which are not labeled or categorized in a manageable way, will prevent future use of important recordings.

Compensation through use of speech recognition

Mainstream technologies such as Voice Assistants (for example Apple Siri, Microsoft Cortana, Google Assistant, and Amazon Echo) are today used in the mainstream classrooms and many other everyday settings (Jana, 2009; Shanmugapriya, Gayathri, & Deepa, 2016). Speech recognition technologies, speech-to-text, can recognize spoken words. The user talks into a microphone and words are transformed into a text file. Some of these technologies need to be *trained* to recognize a specific voice (Anusuya & Katti, 2009).

Speech recognition and speech-to-text programs have a number of applications for users with and without disabilities. These technologies can promote independence and autonomy and improve the individual's participation in the society (Shadiev, Hwang, Chen, & Huang, 2014; Silver-Pacuilla & Fleischman, 2006). The present study shows that some individuals with JNCL are benefiting from using these technologies, for instance when accessing and commanding the devices to do different things. Speech recognition requires however a minimum level of intelligibility and fluency in speech and cannot be used by persons with comprehensive speech problems. The usefulness of speech recognition for some individuals with JNCL should not be underestimated, as the following story illustrates:

> *A young man with JNCL was blind and used several electronic devices. He used speech recognition (Apple Siri) to perform daily tasks which he had not been able to do earlier. He was an active user of a website where he shared thoughts about music and activities. He managed voice calls and messages independently by using speech recognition and making commands like «Call James» or – «Send a message to Yvonne, I will visit you this evening». The young man was also able to store reminders, including date, time and activity. The young man found information on Internet using speech recognition, he simply asked questions like «What is the capital of Germany?» Speech recognition made it possible for the young man to access emails, calculators, alarm clocks and games, to make notes and to do his favorite activity – listen to music.*

Assistive devices to support inclusion

How successfully students with JNCL prosper and function at school will largely depend on the social environment and the relationships with other students (see Chapter 22). The use of assistive devices can play a significant role in supporting peer relations in the class. Some assistive devices are advanced multimedia tools for children and adolescents with communication difficulties. Many computer programs and devices may be regarded as socio-educational tools that give students new social opportunities, especially programs with a low entry level, such as social media, games, chats and music. Comments from parents and staff in the present study also show that many students with JNCL had become ardent users of Facebook and YouTube. Some assistive devices may be used for maintaining interests (and skills) and social participation when functions decline. The following story is about a young adult with JNCL who was a gifted piano virtuoso:

> *Using the music program Magix, a young adult with JNCL put self-produced music videos on YouTube. He was the acting composer and the music was produced and put on YouTube with assistance applying the Team Model (see Chapter 23). The person himself, his parents and friends, and general users of YouTube had great pleasure in watching and listening to the music videos. The composer with JNCL has received many remarkable feedbacks and comments on his productions from far and near.*

Assistive devices to promote motivation

Student motivation is of great concern in education. Some students continue to work on an activity or assignment despite great difficulties, whereas others give up very quickly. Students' motivation is a main driving force in all learning activities and it also supports their self-efficacy (Bandura, 1997, 2006). Students' motivation influences their academic achievements, and what they learn and achieve will in turn influence their motivation. «In other words, it is not the actual learning situation itself that is central, but the students' own apprehension of themselves as learners» (Sølvberg, 2003, p. 17).

Some programs and applications presented in this chapter may function as motivation-generating activities for persons with JNCL and help them to accomplish many everyday tasks. Some programs have been developed in collaboration with the parents, school staff and housing staff with the intention to support work on school subjects and strengthen the students' confidence and self-esteem through achievement.

Opportunities for repetitions is a basic strategy to consolidate learning, in particular in individuals with memory problems (see Chapter 5; Cull, 2000). Quiz games may be used for achieving over-learning, based on necessary repetitions of content, alternative answers and responses. Comments from parents and staff in the present project indicate that Quiz games in addition were perceived as fun by many of the students with JNCL, and quizzes were used to support motivation, learning and inclusion with peers (see Chapter 20).

Some tools that may be useful in education of students with JNCL

Although technology can provide enrichment to the learning process, deploying technology does not necessarily lead to better learning for anyone, regardless of whether they have a disability or not. Success will depend on a match between the technology and the user, the purpose of the technology in accomplishing a learning objective, and the choice of the most appropriate tools for the task (Cook & Polgar, 2008). Nevertheless, technology can help make learning interesting and it might be essential in making learning accessible. There is still a lack of educational software adapted for students with disabilities, but there is a growing range of mainstream technology, both hardware and software, that can serve useful functions for people with diverse characteristics and needs. In the following sections, a diverse range of tools will be presented. Some tools can serve many functions, and it is therefore difficult to categorize these tools. However, tools that are important to the student should be included in the student's curricula and IEP (see Chapter 11).

Many special utility programs for use by teachers have been developed. These programs enable teachers to create applications that suit a particular student's needs. Examples of such applications are Rolltalk Designer (see Figure 19.1; https://www.abilia.com), Grid (https://thinksmartbox.com), Book Creator (https://app.bookcreator.com/), Tobii Communicator (http://www.tobiidynavox.no/), Clicker (https://www.cricksoft.com/uk), and Sarepta (https://www.regionorebrolan.se/sprida).

Different software programs and apps may require different computer skills. Findings from the present study (Appendix A) suggest that some are easy to use while other programs require technical knowledge and practice. These applications often give instant feedback on answers, and some programs can be adapted to the student's level of functioning.

Tools for supporting communication

Students with severe expressive communication impairments have difficulty communicating with peers and adults and may benefit from using assistive technology to support communication. Many factors may contribute to communication difficulties: a decrease in speech intelligibility from motor dysfunction, a decrease in ability to put one's ideas into words, and decreased memory for topics one might want to share. The tools listed below can contribute in very different ways to providing support for communication. Thus, it is important to consider each tool from the perspective of how it might help a given student communicate with others. One must reflect on the communicative demands and the challenges faced by both the communicator and the specific communication partners.

A range of low- to high-technology solutions are available, including communication displays with objects, communication boards and books, switches with speech output, communication devices with synthetic speech and computer programs for communication (see Chapter 13). There are communication devices in the form of software, apps or analogue tools. Programs are often structured as pages, with each page presenting a specific field through pictures, audio, text or symbols. The student uses the structure that exists in these programs, selecting specific fields to activate one or more functions, such as reading a message, activating an MP3 player, writing letters, words or phrases, sending email or SMS, or activating spoken text or a calendar.

Rolltalk, *Tobii Communicator* and *Sarepta* can be used to make applications for communication, concept training, home assignments and educational materials. These tools come with examples, templates and prefabricated subprograms. The programs can be operated in different ways, with switches, joysticks, foot or head control, or adapted keyboards. These tools can be used for communicating through digitized or synthetic speech. The programs provide the student with bimodal choices and the student confirms or rejects messages, statements or options. The student may for example use a keyboard or a switch or produce vocalization to confirm an option, for example:

Level 1. *I want to <u>drink</u> / eat / go to the toilet*
Level 2. *<u>cold</u> / hot / warm*
Level 3 *water / juice / <u>milk</u>*
Level 4. *from a glass / <u>using a straw</u>*

A digitized or synthetic voice will confirm the student's choices. In the example above, the use of the hierarchical structure inherent in the program enabled the student to express «I want to drink – cold – milk – using a straw» through a sequence of choices. Using these tools without vision may be difficult, they make demands on the working memory capacity, because expressing the wish is based on a series of consecutive actions. There is thus a need to develop communication tools that are more tailored to the abilities and challenges of persons with JNCL.

Sarepta

In 1994, Statped (http://www.statped.no) in Norway initiated a collaborative project with Sprida Communication Center (https://www.regionorebrolan.se/sprida#) and Ekeskolan (https://www.spsm.se/om-oss/english/) in Örebro in Sweden to develop a computer program for students with JNCL. The first program was ground-breaking in many ways. It was among the first to use digital audio, images and synthetic speech. The program was first used by students with JNCL in Norway and Sweden and has since then gone through a long process of development in close cooperation with students, teachers and parents in several countries. The program was named Sarepta (Ragnarsson & Björk, 2009).

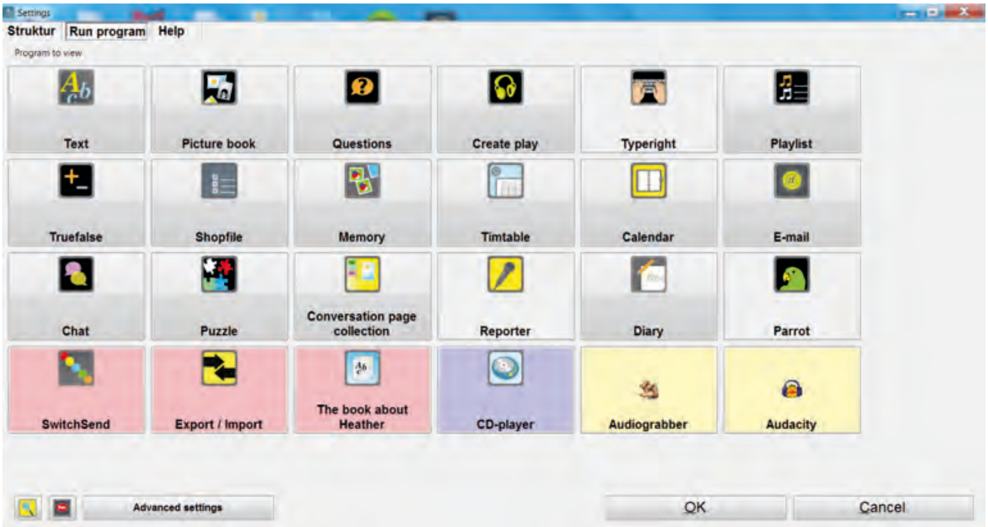


Figure 19.2 Modules in Sarepta

Sarepta is available in Danish, Dutch, English, Finnish, German, Norwegian, Sami and Swedish.

Sarepta is a shell program that makes it possible to tailor solutions for students who lack educational materials in school (Tengs & Torgersen, 2012). Teachers and residential staff fill the program with relevant educational material, which the students can subsequently make use of. The program is a multimedia tool with possibilities to mix audio files, video files and text files. Sarepta has been under continuous development and considerable efforts has been put into motivation-generating activities with new technological solutions, such as integrated digital and synthetic speech.

Sarepta consists of several modules, which give access to digital memory banks, texts, a media player, shopping lists, emails, a diary, picture books, playlists, memory games, quizzes, timetable, the book about me, logbooks, and so forth (see Chapters 20 and 23). The modules provide interactive exercises with feedback on assignments. Direct feedback is important for students with JNCL. The program includes a multitude of tasks and different ways of working, both individually and in groups with peers. The navigation of the program is simple and does not require literacy skills. Students have access to the modules and folders through bimodal choices (yes or no).

All the modules in Sarepta can be controlled by a simplified system with up to five switches (see section below). These five switches can be used to control external applications such as *Point braille, ABC* or other games (see Chapter 21).

Digital books

Digital books can be created with several computer programs and apps such as *Book Creator, Picture book in Sarepta, Phonto, Pictello, Album App* and *Clicker*. Some are developed specifically for people with visual impairments but not all of them are suitable for individuals with JNCL. These programs allow students to present themselves, their activities, interests, surroundings and background. The students can build a memory bank and store self-produced files with text, sound, film or photo. The following story illustrates how digital books may be used to encourage learning, motivation and inclusion:

> *"John" had a vocabulary below average and difficulty reading long words. Together with the teacher, he recorded sounds and made a series of digital images from a visit to the zoo. After the trip, John wrote a text describing his experiences on the trip. The audio recordings were used as sound illustrations, the supporting text was recorded digitally together with other students and edited with help from the teacher.*

Figure 19.3 Picture book «The Elephant»

In the Picture Book, a student can write about a specific subject, such as this book about the elephant, which includes detailed information about elephants, physical traits, eating habits and geographical distribution, as well as stories and myths about the elephant as «the elephant never forgets». The student can later study the information and revise previous findings.

Digital books can also contain facts and information about school subjects and topics, such as «The elephant» with texts, images and sounds (Figure 19.3).

It is also possible to create digital books about the student's specific interests or hobbies. If the student is interested in auto racing, one can scan images and download sound tracks, for example from a Formula 1 race, and then add information about car producers, countries of origin, names of drivers, number of employees to expand the student's knowledge within his or her personal area of interest.

The Book About Me

The *Book About Me* (a module in Sarepta) is a written narrative of the person's life history (further described in Chapter 23). The *Book About Me* can be created as a digital multimedia platform with texts, audio recordings, photographs and films (like digital books above), for collecting and presenting information about the person. It can contain facts about the person, important events, daily activities, school attendance, projects, prizes and appointments, family, friends, neighbors, special interests, hobbies, and a map of all the places where the person has lived or visited.

Figure 19.4 The Diary module in Sarepta

The diary module in Sarepta is filled with information, events, activities, hobbies, adventures, experiences and topics selected from the person's personal experiences and interests. The structure of the program makes it easy for both students and staff to attach written materials, video clips, photographs, audio recordings and multimedia displays.

The Diary

The digital diary (also a module in Sarepta) is a program based on the standard calendar as shown in Figure 19.4 (see also The Yearly Wheel in Chapter 23). It can be operated independently using a braille keyboard with individualized switches (Tash, joystick, concept keyboard, etc.) or with assistance (interdependence, see Chapter 16).

The diary provides a range of possibilities, such as composing daily narratives or using preassigned categories and topics. It includes user-friendly editing functions like delete, save, undo and print located on a toolbar. It is possible to create a workbook, a logbook or a hobby book, where the student can store favorite movies, eBooks and music. A search function makes it easy to find special topics, dates or events. The diary can be used as a basis for conversations and social interactions with parents, relatives, friends or staff. The book may become an important part of the *Book about (the name of the person)* (see chapter 23).

The use of the digital diary may support students' development and maintenance of literacy skills, communication and ability to express wishes, thoughts, needs, feelings and experiences. Another user-friendly diary app is *Day One* (https://dayone.me/), which is simple to use and its content can be synchronized with *iCloud* or *Dropbox*.

Digital conversation sets

The idea behind the programs like *Talking Mats* (https://www.talkingmats.com/), *Notes* (https://www.get-notes.com/) and *Proloquo2go* (https://www.assistiveware.com/) is to support individuals with expressive communication problems. These programs can help individuals with JNCL express opinions related to selected themes. In *Talk* and similar tools, all statements can be answered with yes or no. Phrases that can help meet the identified needs of the person are recorded by teachers or others who know the person well. The programs can be operated by switches where the person listens and selects a phrase or option. Supporters can assist the person with JNCL by scrolling through the various options (see partner-assisted auditory scanning, Chapter 13).

Digital Logbook

A digital logbook may be used for communication between staff members and staff and parents (see Chapter 23). A log book can be used to record events of special importance, for example to describe happenings in the classroom or to communicate problems associated with epilepsy.

Social Media

Social media play an increasingly important role for youths in the modern society, and are important for social contact and communication (Mehraj, Bhat, & Mehraj, 2014). Some social media can be adapted to meet the abilities and challenges of individuals with JNCL. Comments from parents and staff in the present study (Appendix A) indicate that some individuals were ardent users of Internet resources like Facebook, Instagram, YouTube, Chatting and Snapchat with support from parents and staff. Social media make it possible for people with JNCL to keep in touch with acquaintances, friends and family and to share activities with their peers.

Some persons with JNCL have created their own profile on Facebook, with access for their friends and family. These persons keep in touch with friends with or without the use of writing. Written materials on Facebook can be read with a braille screen reader or synthetic speech. The person can get help to write comments or updates of any kind.

Instagram consists mainly of photographs and videos and may therefore be less relevant for persons with JNCL, although it may be used with interdependence where somebody describes the visual content (see Chapter 16).

Smalltalk and chatting play an important role in youth interactions, and since the 1990s, some persons with JNCL have used Chatting, a specially designed program which also includes synthetic speech output (https://www.

regionorebrolan.se/sprida). The chat groups are relatively shielded and accessible only to members of the group. Chatting has functions such as automatic saving and printing in print or braille. It may be useful in education, for example with quizzes and discussions around special themes, as the following story illustrates:

> *A girl with JNCL organized fund-raising activities when she was a child. Two times a year she organized a bazaar to collect money for a particular child in Africa, and made her bazaar known through the social media. This activity occupied the girl throughout the year and she involved family, friends and people in the neighborhood. She was the head of the organizing team, collected items, asked for support from the local bank, wrote letters requesting sponsorship, managed the bazaar with support from others and finally sent the money to the child in Africa. The girl's motto was «There are a lot of children who have a worse situation than me!»*

Skype, Viber and *S-Chat* are other examples of social media adapted for persons with visual impairment. Being engaged in social media may support literacy skills and contribute to social well-being.

Taking pictures

Photography is not usually a special interest of children who are blind, but there are exceptions. The story about this young man with JNCL illustrates that a positive and open mind can lead to astonishing results by using creativity:

> *"Jack" was very interested in taking photos whilst he still had some vision left and kept this interest after he became blind. He took photos together with his family and it was a very exciting activity for the boy. After a few years, the parents started to carefully study these photos and discovered that many of them were good and special. Sometimes Jack would miss the motive and the photos would have a new content and other artistic dimensions. He challenged himself to learn photography using other senses – being more attentive to sounds and tactile qualities. For example, by listening to someone talking he could more or less assess the distance and height he needed to aim for. The family had great fun when watching and discussing these photos and lots of humor was triggered by the pictures.*
>
> *The parents presented the photos to the well-known Norwegian photographer, Morten Krogvold, who was so impressed with Jack´s artistic pictures that he set up an exhibition with Jack's photos at the Nordic Light-festival in the Norwegian city Kristiansund. The exhibition was a huge success and received positive reviews in the media. It was a great experience for Jack and his family. His story is now spread on social media.*

Jack's story shows that early interests – in this case photography – can be maintained with support even if a decline – here blindness – gives the activity a new and unconventional dimension.

Email and SMS

Most people send and receive emails and SMSs, and these tools can also be used by persons with JNCL for keeping contact with peers, family and society. The use of email and SMS may contribute to maintaining friendships when physical meetings with friends are limited. The following example shows that email correspondence can strengthen participation in society:

> *A few years ago, a young man with JNCL corresponded with family and friends across the country via email. He had an assignment to make his own electronic newspaper about football which was distributed via email. He surfed regularly on the websites of Manchester United (football club in UK) and Rosenborg (football club in Norway) to catch the latest news of these clubs. He also made interviews with his favorite players in these clubs, which were published in his newspaper. The paper became very popular.*

Speech recognition may be of great help when young people with JNCL use email and SMS when their speech is intelligible. However, *interdependence* or the *Team Model* will be needed when speech becomes unintelligible (see Chapters 16 and 23).

Many assistive technology programs have integrated email systems with automated storage features. The person with JNCL can record their own voice or speak directly in the email and send the recordings as digital files. Email systems can be operated by switches or keyboard. Comments from parents and staff in the present project indicate that active use of email was motivating and contributed to the maintenance of writing and reading.

Multimedia equipment and text editors

With multimedia editor programs, persons with JNCL can combine files with audio, text, images and video. It is possible to create individual material which enables students to adapt their learning efforts to their own learning progression and regression. The possibility of using sound, switch control, screen magnifier programs and braille display makes digital solutions available to persons with JNCL.

The Braille Cell

The Braille Cell (https://www.regionorebrolan.se/sprida) is a computer program with audio support designed for teaching braille at an early stage. It may support the student's understanding of the braille cell structure by using a regular keyboard or a braille keyboard (see Chapter 14). The program produces letter sounds or spoken words as the braille cells are produced (Figure 19.5). The program includes, in addition, an interactive part named *The Adventure*, with several premade adventures. The student can work with these adventures to facilitate literacy skills. They can also create their own adventures in *The Adventure*. The Braille Cell can be used in parallel with applications such as *Braille Hunt* (http://www.textax.se/) and materials for *Flexiboard* (https://www.abilia.com).

Alternative keyboards

Alternative keyboards, such as *Flexiboard* (see Figure 19.6), can be adapted to a person's special needs and may enable students with JNCL to use the computer. The board has several layers of tactile material which the student can identify through touch. It is possible to add recorded speech or other sounds. Flexiboard has many other applications for leisure activities and schoolwork. For example, the student can be given an overlay with the map of Finland. By touching different areas of the overlay, the student will be given information about different places, for example *Helsinki is the capital of Finland and has 620.000 inhabitants*. Map use can be expanded to include topics for conversation, including telling others where one has traveled, or where friends and family live, and then inviting others to expand on those ideas. Flexiboard can support learning in all school subjects, for instance in biology or music. It is possible to create a multimedia presentation by using Flexiboard, for example include speech, music and videos in one presentation.

Figure 19.5 The Braille Cell

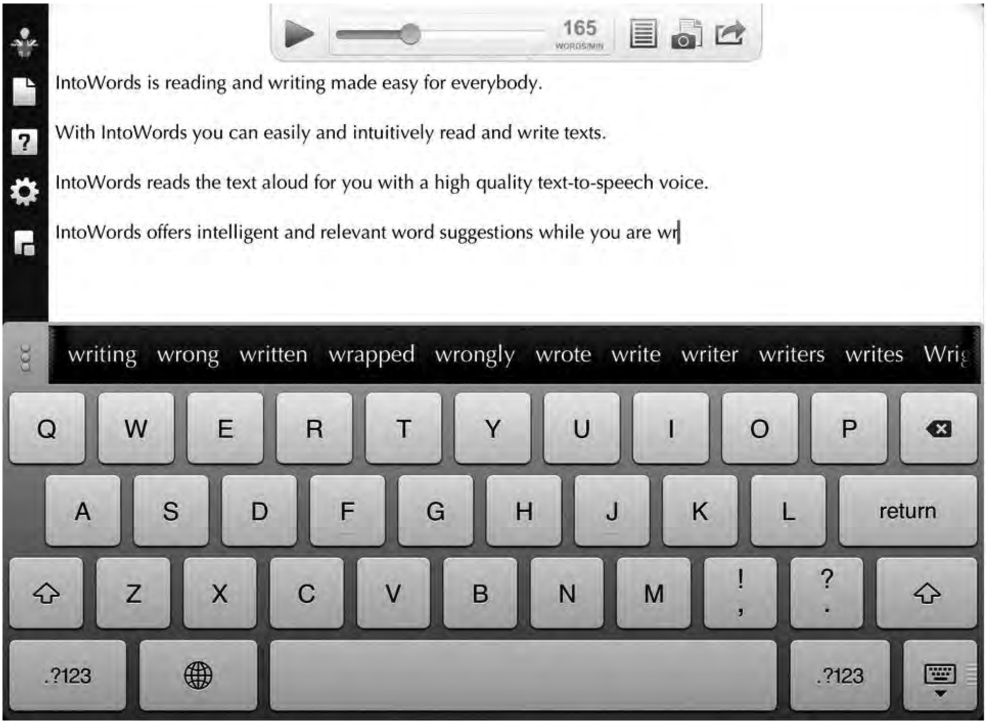
The user gets audio feedback as the braille cell is built.

Figure 19.6 The Flexiboard

The flexiboard offers more possibilities than the traditional keyboard.

Aids to support writing

Some aids support the process of composing one's ideas in print. These are typically apps or programs, not hardware (see Batorowicz, Missiuna, & Pollock, 2012). Many such programs, including *Ghotit* (https://www.ghotit.com/) and *Lindys* (https://lingit.no/produkter/), are designed to help students with reading and writing difficulties (MacArthur, 2009). They are mainly used by students with dyslexia, and some students with JNCL can benefit from using these programs, especially in primary school. For example, if a student can write the first letters, the program can predict a number of words that begin with these letters. The student can then select the right option. This function is common in text messaging. Two types of support are provided: writing might be quicker with fewer keystrokes, and it can compensate for difficulties in spelling and memory. The combination of spelling support, a built-in dictionary, word prediction, multimodal input, screen



IntoWords is reading and writing made easy for everybody.

With IntoWords you can easily and intuitively read and write texts.

IntoWords reads the text aloud for you with a high quality text-to-speech voice.

IntoWords offers intelligent and relevant word suggestions while you are wr

Figure 19.7 IntoWords is a reading and writing support tool

IntoWords is a digital tool for reading texts aloud and compensating for reading and writing difficulties. It uses word prediction, helping the student by suggesting the next word.

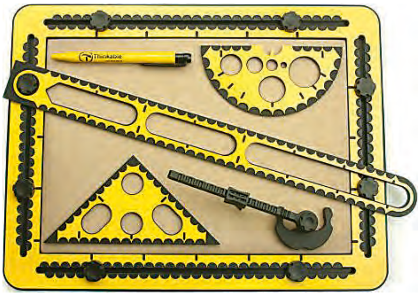


Figure 19.8 TactiPad with drawing tool

TactiPad is a versatile drawing board, which enables a visually impaired person to make a tactile drawings by firmly pressing down on the foil with a pen; the lines will raise instantly, forming a tactile image.

reader and synthetic speech output makes *Textpilot (https://lingit.no)* useful for working with texts.

The app *IntoWords* (https://www.mv-nordic.com/en/products/intowords) also provide writing and reading support (Figure 19.7). *Clicker Grids* (https://www.cricksoft.com/) may help students learn the relationship between written and spoken words. The synthetic speech feedback allows students to review and take control of their own writing (Genlott & Grönlund, 2013).

The app *WordWav* converts text into an audio file which can be read aloud on different audio players. This makes WordWav a useful tool for some students with JNCL.

The *TactiPad*

The TactiPad is a versatile drawing board, which enables persons with visual impairment to produce tactile drawings (Figure 19.8). These drawings are made on a thin sheet of plastic foil on top of a layer of rubber. By firmly pressing down on the foil, the lines will rise instantly, forming a tactile image (https://irie-at.com/product/tactipad-drawing-tablet/). The technology is similar to swell paper (see below). The drawings might be used to support the student's own written texts, or substitute for them. One can also communicate ideas through drawings.

Digital recorders

Digital tools, such as digital cameras and voice recorders, are useful in education. During visits to museums, Christmas workshops or outdoor fieldwork, teachers and students can work together to record the activities, for example by taking photographs or making video and audio recordings. These creative products may allow the student to relive the activity, refresh memory and create a basis for conversations, discussions and school assignments.

Students can use an ordinary audio recorder or apps to record television or radio programs, speech or other sounds in their surroundings. Students may for example record their thoughts and opinions about events during the day, which can be included in their diary.

The use of a digital recorder within and outside the classroom may facilitate learning and motivation. The *Reporter* module in *Sarepta* (see above) functions as a sound recorder that can be used when interviewing people. This module is a simple digital recording app which includes speech synthesis. Recording can be controlled by two switches (on and off) or with F4 and F5 keys on a standard keyboard. The files are automatically saved in the audio bank in MP3 or WAV format, and recording time is limited only by the size of the hard disk of the computer. Recorded audio files can be deleted or edited by numerous media programs, such as Microsoft's *Sound Editor* or *WavePad*.

The student's library

With the help of programs such as Audacity (https://www.audacityteam.org), students with JNCL can build up a library of self-produced texts, audio recordings, audio books, radio programs, and so forth.

Music

Music is a source of pleasure and of special importance for individuals with JNCL according to the present study (see Chapters 17 and 18). Active participation in singing, playing an instrument and listening to music together with others, provides social and cognitive stimulation and facilitates inclusion. Comments from parents and staff in the present study indicated that music is a life-long interest for many persons with JNCL (see Chapter 17).

There are a number of computer programmes and music technology products which parents or staff can utilise to engage children and young people with JNCL in music activities.

Programmes on the iPad such as *Echostring, Drumbeats* and *Sound Prism* have been shown to be successful for individuals with some range of movement in their hands or arms, and for whom conventional instruments are too difficult to play. One parent commented on her son's enjoyment of *Drumbeats*.

> *It has all the instruments that he likes. He loves the ocean drum and when you tip the iPad, it makes the same sound. This is brilliant because he cannot really hold the real ocean drum as well anymore, but the iPad he can still grip and tilt.*

These interactive programmes allow children and young people to make sounds easily and help develop an awareness of how they can use small movements in their hands to make interactive sounds. However, they are too limited for children and young people who have the cognitive ability to engage in more complex music activities, such as song-writing or music improvisation. For this group, programmes such as *Garageband* and *Launchpad* may be used in school and at home, to encourage children to execute decision making, planning, and enjoyment in making their own music.

The *BEAMZ* unit, an interactive laser-beam console, has been trialled with individuals who have a good awareness of the cause and effect of movement, but who have limited gross motor skill in the upper or lower body. Children and young people with a higher cognitive ability have navigated their way around the console using their fingertips to determine where the lasers are situated and how sounds can be manipulated by movement. They have also recorded pieces of music and listened to these being played back to them.

Makey Makey is a system which can turn everyday objects into touchpads and combine them with the internet to play games, music or create switches.

Audio books

Audio books represent an important compensatory medium for people with severe visual impairment or reading and writing difficulties (Saine, Lerkkanen, Ahonen, Tolvanen, & Lyytinen, 2011). Listening to audio books supports social participation by supplying topics for conversation. It is also a meaningful leisure activity, as reported by parents in the present study (Appendix A). Many books and radio programs are available as audio books. Audio books give people with JNCL access to a large selection of fiction, nonfiction, academic books and newspapers. Audio books are also available in many libraries. In addition, there are many applications for purchasing and downloading audio books on internet, such as *Storytel* (https://www.storytel.no/), *E-book* (https://www.ebooks.com/), and *Audio Book* (https://www.audible.co.uk/). The national associations for persons with visual impairment produce audio books in order to provide equal access to literature and information. These services are free and nationwide in many countries.

Electronic tools for daily living

Time management is important in modern societies, but understanding time and reading *digital clocks* can be difficult for many persons with JNCL. There are watches with synthetic speech that can be adapted to individual needs. It is possible

Figure 19.9 – A QR code

QR codes can store any kind of information. They can be used to identify personal objects like books or CDs, or to download digital versions of audio books on Internet.

to add personal messages to some of these watches. *Cell phones* can be used in many different ways and may be adapted to the student's abilities and needs. They may provide support for time, planning, structure and memory. The calendar on the cell phone can show planned activities and alert the person when an activity is starting or ending. *SmartVision* (http://www.sightandsound.co.uk/) is a smart phone that is designed specifically for people with visual impairment. This type of phone is suitable for children and adults with JNCL. *SmartVision* combines a physical keypad, a full voice interface, and a touchscreen. It can be organized according to personal preferences and is user friendly, with a large display, large icons, four font sizes, tactile buttons for numeric keys and navigating, voice commands and speech-to-text facilities. It can receive and send text and audio messages.

A digital schedule provides the same features as a normal calendar but may have a range of further possibilities. The schedule can be supplemented with personal information and text, images and audio. For instance, a gym program can be specified in the digital schedule and provide information about the plan for the next day. A digital schedule (and other devices) can be used for making shopping list, check list and so on.

A QR code (quick response code) is a matrix barcode, which can contain comprehensive information about an object, for example in supermarkets (Figure 19.9). The QR codes are generated online and can be read by QR scanners on the cell phone, and apps that utilize QR technology are readily available for desktop and mobile applications. QR codes can be useful in education as they can store all kinds of information, such as week plans, shopping lists, mathematics and biology. QR codes can be used to identify personal objects like books or CDs, or to download digital versions of audio books from Internet.

Pocket-sized GPS navigation devices with voice recognition and synthetic voice output, like *Captain mobility, Breeze* and *Blind Square* are practical orientation and navigation aids for persons with visual impairment (Zegarra & Farcy, 2013). These and similar GPS-based tools are able to provide information about current location, the streets at the next junction, places of special interest, shops, restaurants, pharmacies, and more. Some of these aids have information about buses, trams, trains, subways and other forms of public transportation. For instance, when the user clicks the *Where am I* button, the device gives information about the street name of the current location and the travel direction. It is possible to add personal points of interest into some of the devices. It is not known if any of these technologies are used by persons with JNCL, but they might be perceived as fun and exciting by some individuals with JNCL.

Maps and atlases can be useful when talking about places, or to find the name of a town or country. There are various map applications to choose from, including *Google Maps* and *Google Earth*. It is possible to use speech input to navigate and synthetic speech output with these maps.

The *Mobile Reader* (https://acrobat.adobe.com/), *TapTapSee* https://taptapseeapp.com/) and similar apps installed in cell phones have scanner, speech and reading functions. The scanner function makes it possible to take pictures of documents like bills, menus and banknotes. The image is transformed to a spoken description which can be saved on the phone. It is not known if such devices are presently used actively by individuals with JNCL.

The PenFriend (https://www.rnib.org.uk/) is an audio labelling system for individuals with visual impairment. The system consists of a digital pen and labels which contain readable barcodes. The pen has a scanning and memory function, a microphone and speaker system. Each label can be combined with an audio recording. PenFriend can be used for cooking instructions, dietary information or the date when something was placed in the freezer. It can also be used to keep track of personal items like CDs, DVDs and books.

Assistive technologies such as the *Tactile Image Maker* (http://www.perkinselearning.org/) make touch-based drawings on special swell paper. The student can draw, print or photocopy pictures onto a swell paper and pass it through a swell machine. The heat from the swell machine makes the illustration readable with touch (see ttp://piaf-tactile.com/producing-tactile-graphics/).

Similar assistive technologies allow persons with JNCL to make drawings and design or read simple graphics, maps and similar. This type of technologies may be useful for persons with JNCL, both in education and leisure activities, and may promote inclusion when persons with JNCL are taking part in classroom activities with peers.

Technologies like *Loc-Dots* (https://www.independentliving.com/) consist of raised dots to be used for tactile marking. The dots are simple to use and can be placed where needed, for instance for marking of switches, the computer keyboard, books and tape recorders. Such technologies may simplify daily living for individuals with JNCL.

There are several computer programs and apps available for doing crossword puzzles, memory games, board games, chess, lotto games or card games. Some of these games may be used to enhance learning and pleasure for children with

JNCL. Examples are *Scrabble Braille, Monopoly Braille, Arcade* and *Digital Memory Lotto* (https://shop.rnib.org.uk/) (see Chapter 21).

Persons with JNCL who have severe motor impairment are often dependent on switches to control computers, communication devices, cell phone and other devices. It is important that students with JNCL are introduced to such switches at an age when their learning capacity is at its best, according to the principles of enhanced and proactive learning (see Chapters 11 and 12). Moreover, care should be taken in switch selection, so that the properties of the switches match the sensory and cognitive capabilities and preferences of the switch user. Switch technology now accommodates a very diverse range of users (Gibson et al., 2013)

Careful consideration must be given to the quality and quantity of the feedback a switch affords. For example, many switches make a clicking sound to notify the user that a "click" has been activated. Switches have a range of tactile properties to cater to tactile preferences, in the case of people who have atypical tactile sensitivity, or tactile needs, as in the case of someone who requires considerable tactile feedback. Vibrating switches, for example, provide constant feedback during the entire time the switch is activated. Over the course of the period of decline that accompanies JNCL, there will be a need for re-evaluating the type of switches used and the provision of individual support.

Recommended computer equipment and other aids

Recommendations for assistive technologies for persons with JNCL should always be based on an assessment of needs, functions and possibilities (see Chapter 10). Assessments must be made by professionals who have sufficient competence related to JNCL and assistive technology. Assistive technologies are often expensive, and comments from parents and staff in the present study indicate that many of the devices that were provided never had a positive impact on the learning and well-being of the person. The reasons were often a lack of local knowledge about the devices and an over- or underestimation of the functional use of the device.

A core device for a person with JNCL is a laptop computer with large storage capacity. Files with audio, images and video require extensive memory capacity. A good sound card is mandatory, and appropriate switches or other input devices for operating the electronic equipment. Braille keyboard and display are often needed and a Flexiboard may serve as an adaptable keyboard and mouse. High-quality audio equipment plays an essential role for persons with JNCL, including a high-quality microphone. The device should have high quality speech synthesis, for several languages if the person is living in a multilingual environment or learning a foreign language in school. A portable multi-player with optical character recognition (OCR) and text-to-speech for scanning books and documents is also a basic equipment for individuals with JNCL. The devices should have a numeric keypad and a dictionary with possibilities for doing calculation and making memory notes. The device should be able to play audio books and music files from internal or external memory devices and should be supplied with an earphone that turns off the external speaker when connected, to avoid disturbing others.

A digital camera may be used for educational and social purposes. A braille printer, located where it does not disturb others, is essential for braille users. A color detector is easy to use, and has a wide range of useful applications, from checking the color of clothes to the ripeness of fruit.

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20

Quiz Games for Building and Maintaining Knowledge

Mohammed Beghdadi and Bengt Elmerskog

Quizzes are multiple-choice games or mind games that are becoming more common in education in many countries. Quizzes are usually scored in points and many quizzes are designed to determine a winner, a single person or group. Many quiz platforms are available online, such as Kahoot and Triviaplaza, and these can provide ideas and resources for teachers and students, when there is a need for quizzes with an educational focus. Quizzes lend themselves to many formats allowing teachers to use them as a class activity or to structure materials for students to use in self-study to repeat reviewed material. Teachers sometimes encourage students to design quizzes themselves as a review strategy with a competitive twist.

Quiz in education

In school, the quiz has a long tradition. Asking everybody in the class specific questions is a standard teacher activity, as well as using tests with multiple choice answers or questions that require a short answer to evaluate the knowledge of the students (McDermott, Agarwal, D'Antonio, Roediger, & McDaniel, 2014). Most quizzes in education are developed locally to support predetermined teaching goals. Many quizzes are programs that can be performed on a computer, tablet or smartphone, but few such quiz programs are adapted to meet the special needs of students with visual impairment and cognitive disorders.

Using quizzes can function as an alternative or complementary method to achieve and support different educational purposes or teaching goals (Brusilovsky, Sosnovsky, & Shcherbinina, 2004). A quiz can provide multimodal input and repetition that may contribute to improved learning. They can be used as a variation from the traditional forms of teaching and learning. Many students perceive quizzes as fun, and the questions may stimulate and promote new interests. The teacher can use quizzes to assess the students' knowledge of a particular subject or theme, or the students' ongoing growth in knowledge, abilities and skills. Students may use quizzes to test themselves and get feedback on their own learning process. For instance, a quiz may be used to check the comprehension level of a type of mathematical exercise in a class. Quizzes are often presumed to be less formal, more short and concise, and easier to organize than other forms of written tests and examinations.

Quizzes can be used to *refresh* the content of one or more lectures. The students attend a lecture and take a short quiz on the lecture content afterwards. Such use of quizzes can also function as a student response system that can be used to guide the teaching (Melero, Hernández-Leo, & Blat, 2014). It can measure what and how students are thinking and address the needs immediately in class. Quizzes are also used to check students' prior knowledge, probe their current understanding, and uncover student misconceptions. They can provide feedback to the teachers about their students' understanding and to the students about their own comprehension.

Some schools schedule a daily or weekly quiz for the purpose of having the students review recent lessons before attending the next class. Further, quizzes are used to check if the students have done their homework according to the plan. Using a "pop quiz" means that the teacher gives it without warning: students are not given time to prepare for the task, and so they are taken by surprise.

Learning facts and making personal reflections

Quizzes are most appropriate for learning facts, for instance within geography and history, or learning by heart, for instance the multiplication table. Quizzes are less appropriate for learning things that require certain insights, appreciations, judgments or reflections. However, further elaboration around questions presented in a quiz game may emphasize further insights. It is often motivating for the student to compare answers with others while waiting for the results. Such peer collaboration may contribute to further insight and to the students becoming aware of different aspects of the presented tasks. This may stimulate curiosity and learning because it may lead to discussions involving personal experiences and emotional engagement. The teacher can in addition use the interest caused by the quiz to present further elaboration and explanation. The quiz may be used strategically as a door opener or a starting point for deeper understanding and further capacity and competence building in predefined domains.

Refreshment and consolidation of learning

Retrieval practice is a common teaching strategy where the teacher introduces actions and activities to consolidate the student's knowledge (Chang, 2015). The student is practicing exercises to activate what is already learned as opposed to acquiring new knowledge. The act of retrieval, or calling information to mind, will refresh the memory and make forgetting less likely to occur (Morey & Cowan, 2018). Retrieval practice is a powerful educational strategy to consolidate knowledge and improve the effect of learning (Uner & Roedigeret, 2018).

Some authors recommend use of quizzes as vehicles for providing repeated retrieval practice (Karpicke, Butler, & Roediger, 2009; Karpicke & Bauernschmidt, 2011). Learning is typically identified with acquisition, encoding or construction of new knowledge, while retrieval is considered a means for accessing knowledge, rather than a process that contributes to new learning. The process of consolidating new knowledge in a strategic way is not always emphasized in education. However, some authors suggest that retrieval is a key process for both understanding what has been learned and for promoting learning, yet retrieval is not granted the central role it deserves. Research, particularly that of Karpicke and colleagues, shows that closed-book repeated quizzes have better impact on long-lasting learning than open-book questions. Existing educational practices can easily be converted into retrieval-based learning activities according to the authors, for instance by answering questions by taking repeated quizzes. Classroom quizzing is one effective method among other methods for effective implementation of retrieval practices (Agarwal, Karpicke, Kang, Roediger, & McDermott, 2008). The importance of retrieval practices holds true even in preschool populations. During the preschool years, children are learning vocabulary, but some children have difficulty relative to their peers and require support. Activities that provide retrieval practices are thus valuable because even preschoolers learn vocabulary better when they are given such opportunities (Fritz, Morris, Nolan, & Singleton, 2007). This research might provide insight into ways that can promote enhanced retention of knowledge in children with JNCL in the face of decline.

Quizzes and teaching

Quizzes can be used by teachers to build up a positive atmosphere in the classroom, for example to reduce stress. Quizzes are often perceived by students as fun and relaxing if they are used in a balanced way; they can create feelings of pleasure and joy. Quizzes are rather informal compared to other forms of evaluation. They may have a competitive element that creates excitement among most children and young people. The provision of multiple-choice answers will in addition give the student a fair chance to appraise appropriate answers by ticking the most probable alternative. Multiple-choice answers have the advantage of reducing some of the memory retrieval load: the correct answer is available, and it is up to the child to appraise the options, not generate them.

A quiz with multiple-choice options usually requires less time to administer than a test with written responses. The teacher does not need to interpret answers because students are graded purely on their selections. Moreover, clarity of presentation is not an issue when using an online or electronic quiz and poor handwriting will not influence the results, although the child or young person may accidentally mark the wrong box. An additional advantage is that the evaluation of a quiz makes it easy for students to acknowledge their own knowledge or lack of knowledge in a rather distinct way. Quiz tests can be strong predictors of overall student performance compared with other types of evaluations, but the complexity of the material and other factors may influence the learning effects of quiz tests (Adesope, Trevisan, & Sundararajan, 2017; van Gog & Sweller, 2015).

Quizzes for people with dementia

Quiz games are often used beyond education, in connection with festivity, celebration or other types of social meetings to create a good atmosphere. A quiz can easily be adapted to facilitate and adjust the content in accordance with individual needs and abilities, for instance for individuals with learning difficulties. Quizzes are used for elderly people with the diagnoses of dementia (Elder Care, 2016). Quizzes and similar games, like crossword and puzzles, are common activities among elderly people with or without dementia, since such games provide pleasant social settings and cognitive stimulation at the same time.

According to cognitive stimulation theory, quizzes are considered to be "brain games" that may prevent or at least delay the loss of knowledge and skills in connection with the progression of dementia (Dormann, 2016; Livingston et al., 2017). Quiz games may be a useful way for elderly people to exercise their mind. Questions about various subjects will get their mind working and stimulate parts of the brain that otherwise may not be so much used. Elderly people with dementia are found to do rather well in quiz games compared to many other cognitive games, partly because it is easier to recognize correct options than to recall information (see Chapter 5). Using quizzes on a regular basis may contribute to maintaining old memories of special importance for the person (i.e., reminiscing). Studies of reminiscence therapy suggest benefits to adults with dementia in the form of mood and cognition improvement, and benefits to caregivers in the form of lower strain (Cotelli, Manenti, & Zanetti, 2012; Woods, Spector, Jones, Orrell, & Davies, 2005), especially when related to the life history and personal experiences of the individual (Subramaniam & Woods, 2012).

A strength of the use of quizzes for people with dementia is the immediate response to the answers that are selected (Balota, Duchek, Sergent-Marshall, & Roediger, 2006). Immediate responses require lesser demands on the capacity of the short-term memory. Keeping the mind stimulated is even more important in younger people with dementia disorders, and doing quizzes, puzzles or crosswords that invigorate the brain are highly recommended by experts. For instance, quizzes that incorporate numeracy can be useful for keeping those skills alive according to Alzheimer's Society in UK (2018).

Quiz use among children and young people with JNCL

In the present JNCL study (see Appendix A), quiz games were often mentioned by parents and staff as an activity of special interest for the participants with JNCL. Moreover, the interest for doing quizzes was described as a life-long interest for some individuals with JNCL. Both school teachers and staff in residential homes for young adults with JNCL said they were using quizzes as strategic tools to enhance learning and help the individual maintain memories and skills. Many parents reported using quizzes as a joyful leisure time activity to promote fun and well-being. The quotations below from parents and staff suggest that quizzes may have a special role in the life of children and young persons with JNCL:

> *He has always loved participating in quiz games. He makes his own music on YouTube, we make quizzes about music, for instance, who composed this song? These games are used for building up his self-confidence, he becomes very proud; playing quiz games is also a way to see if he is able to accomplish same tasks as previous years.*

> *So like, for example, we were doing time and math, they focused on the calendar and we worked on the months of the year and we started off with how many months there are and – and we gave her the little rhyme that goes «30 days has November» and she brailled that out and then we had the little quiz with all the months and she had to say right she'd find, she'd take the month January for example, how many days does January have?*

> *And she wouldn't know so we went through the rhyme again and that and she would work it out and put the right – find the right number and put it on the calendar, there was maybe a choice of two numbers.*
>
> *We have all types of games, board games for the blind or self-made games. Sometimes we modify original games and adjust them to her mental progress. She prefers quizzes, she just loves it. She also loves the Tiptoi books. Her interest in games grew over time, games have been very helpful.*
>
> *We use themes in life when preparing quizzes. You could talk about topics which concern and move them, more in the direction of life planning – with different levels. Some kids with JNCL could complete tasks by writing braille and they enjoyed it. This was just their competence level. With other kids we had to do a quiz game: «What's the capital of … and which river passes through …?» Then they answered and were very proud and happy. That was helpful. Or we utilized their knowledge of foreign languages. «Which language do you speak? What is 'good morning' in French?» and so on, that's all possible if it accords with the interest or knowledge the kids have.*
>
> *Our child is very eager to learn. She learns a lot by doing quiz games (for children, Tipp Toi etc.) about general knowledge but especially about nature, environment and animals. She is very creative and again she emphasizes that she has an advantage over blind-born children.*
>
> *We conclude every school week by doing a group quiz. It is a perfect way of concluding and recapitulating the week.*
>
> *The most popular activities in the leisure time club are swimming, bowling and doing quizzes. Her performance level in doing quizzes is impressive.*

The comments show that quizzes are used in the countries that participated in the present project to promote learning and maintain skills and knowledge, as well as to promote joy and excitement in children and young people with JNCL. However, both parents and staff expressed the view that quizzes could be used more, both for teaching and cognitive stimulation, and for stimulation and joy beyond school.

Quiz Tool for persons with JNCL

As part of the present JNCL and education project (Appendix A), a simple and automatically-controlled quiz program, the Quiz Tool, which has the necessary level of repetition of content, alternative answers and immediate responses was developed for children and young people with JNCL. This part of the project received extra funding from The Norwegian Directorate for Education and Training and the Norwegian NCL Family Association. The motivation behind the project was that a quiz program represents an alternative method for contributing to achieving new learning, to refreshment and conciliation of knowledge, and to maintaining cognitive skills and knowledge in children and young persons with JNCL. The Quiz Tool may be used by people with or without vision. The instructions can be read through vision (print), the tactile modality (braille), or hearing (speech synthesis).

The Quiz Tool can be used to create a variety of quiz categories, for example of school subjects such as geography or history, activities like playing or sports, or events such as birthdays and holidays (Figure 20.1). This can help the student with JNCL to get an overview of important events and activities. The Quiz Tool is flexible and enables teachers to teach subjects with questions tailored to each student's needs, desires and abilities. The tool can be used for most subjects

Adjective

capitals in Europa

Cars

Food

Fotball england

Figure 20.1 The Quiz Tool with different quiz categories

The desired quiz game will open by ticking the category. The selected category, for example «Adjective» can be read or heard with the speech output of the computer.

Table 20.1 Examples of questions used with the Quiz Tool

1. Standard quiz questions
Question: Which city organizes the annual Academy Awards, also known as the Oscars?
A: New York B: Los Angeles C: Chicago
Question: When was Michael Jackson born?
A: 29. august 1958 B: 29. august 1956 C: 29. august 1960
2. Incomplete sentence
Question: Norway's annual international Fish Convention is organized in:
A: Oslo B: Bergen C: Trondheim
3. Grammatical error
Question: Which sentence is correct?
A: I saw a football match yesterday B: I saw football match yesterday C: I see a football match yesterday
4. Quiz questions requiring different focuses
Question: The town Tromsø in Norway
A: is located in Nordland county B: has more than 250 000 residents C: is described as Paris of the Scandinavia
5. The Quiz Tool can be used with daily activities such as cooking
Question: If salmon ceviche is on the menu – how is the salmon prepared?
A: Marinated in red pepper and fried B: Marinated in lime C: Prepared in a plastic bag and put in hot water (60 degrees).
Question: In which of these dishes is there a lot of salt?
A: Caviar B: Cheese C: Fruit salad
~~**6. Avoid using negative expressions such as never, not etc.**~~
~~**Question: Which of these countries is not located in Europe?**~~
~~A: Spain~~ ~~B: Germany~~ ~~C: Syria~~

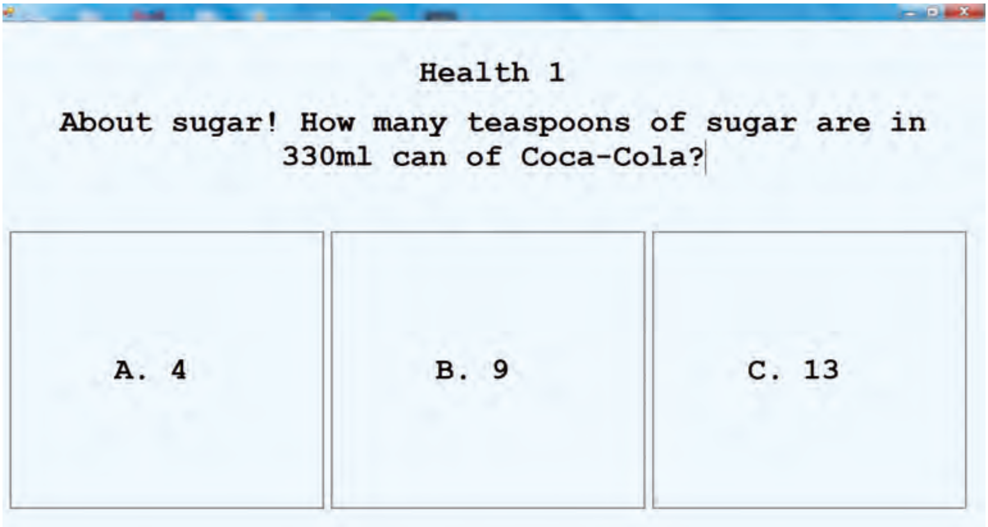
and school grades to promote motivation and inclusion. Table 20.1 shows some examples of questions and answers in the Quiz Tool.

More material can be added to the program by supplying more personalized information or questions of relevance to current school topics, and by giving feedback and comments related to the student's answers. The difficulty level of multiple-choice questions can be adapted to the student's cognitive level and knowledge.

The Quiz Tool can also be used for self-study, for example as an obligatory home assignment or a daily routine. It is possible to repeat the same quiz many times, and comments from parents and staff indicate that such repetitions may promote learning and feelings of mastery. The Quiz Tool may also be used for retrieval of materials that the student learned some time ago.

If possible, the quiz questions should be related to the mainstream curriculum. This goal will become difficult to achieve for students with JNCL as the disease progresses. A moderate or severe dementia requires that the quiz is modified to be appropriate for the individual person. A person-centered quiz requires good knowledge of the person with JNCL and his or her life. In any case, constructing a quiz based on the person's life, desires and experiences, requires a close collaboration with parents.

In the present project, some young persons with JNCL often selected the last presented alternative when given three alternatives in the Quiz Tool (see Figure 20.2). The decline in working memory capacity may have made it difficult

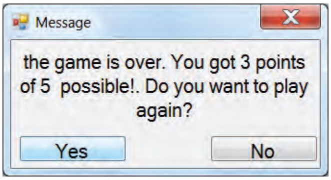


Health 1

About sugar! How many teaspoons of sugar are in 330ml can of Coca-Cola?

Figure 20.2 The Quiz Tool – a quiz question with three possible answers

The question and the answers can be repeated as many times as the player wants.



the game is over. You got 3 points of 5 possible!. Do you want to play again?

Figure 20.3 The Quiz Tool – end of the game

At the end of the game, the number of points is displayed and the player is asked if he wants to do the same quiz again. The information is presented in print, braille and by speech synthesis.

for them to remember all three alternatives. In spite of such difficulties, the quiz organizers (parents and staff) were never in doubt about the usefulness of the program. They said that the persons with JNCL usually showed great enjoyment when playing quizzes, just as they had done when they were younger. Playing quiz games raised their engagement and mood, and according to staff and parents, they were also more awake. Participating in enjoyable activities is an important element for quality of life and playing quiz games seems to provide a good social setting. Ensuring that some of the last quiz response alternatives are correct will promote feelings of mastery and pride, according to the parents and staff. At the end of each quiz, the players are asked if they want to do the same quiz again or leave it (Figure 20.3).

Observations indicate that playing quiz, in addition to supporting cognitive functions, also may be used to promote social learning in children and young people with JNCL. A specially constructed quiz can be a helpful element in handling or discussing difficult topics and situations, such as bullying, violence, democracy, or children's rights. It is not known to what extent quizzes are used to support teaching of such concepts, but they are an integrated part of the mainstream curriculum in many countries, and should also be taught to children and young people with JNCL. Maybe a tailored quiz about society and social behavior can contribute towards a better understanding of the importance of appropriate behavior for students with JNCL. Some teachers reported that some of the questions they included in the Quiz Tool had inspired the class, including the students with JNCL, to show good sportsmanship.

The Quiz Tool is available in English and Norwegian and can easily be translated into other languages.

Self-made quizzes

The Quiz Tool may be part of the teaching repertoire but it can also be used by children and young people with JNCL to construct their own quizzes, with their own choice of topics, questions and multiple-choice answers. There are some automatic quiz generators on the Internet but they are based on factual information found at Internet, and quizzes generated from them are therefore less tailored to the needs of individual students, which is a necessity for young persons with JNCL (see Samuelsen, 2016).

It is possible to construct a quiz with or without the ability to see, but basic knowledge about operating the Quiz Tool is necessary. Reports from the present study indicate that a few young persons with JNCL were deeply engaged in creating their own quizzes and presenting them to peers and adults when ready. The parents further reported that building and creating quizzes resulted in strong positive feelings of ownership. The process of making their own quizzes stimulated the children and young people to investigate particular knowledge domains, such as sports, music or film. Parents and staff said that these investigations increased interest and motivation, which in turn lead to further study of the topic or domain. Self-made quizzes are good examples of self-driven education for these students. Scoring and announcing the results of the quizzes when they were played, and dispersing the prizes, were very exciting experiences for the students with JNCL.

It is possible to create a bank of self-made quizzes for later use in school or when meeting friends after hours. Self-made quizzes can also be shared with other students or friends through internet or email. The present study learned about a young man with JNCL who was very devoted to developing, sharing, and exchanging quizzes with others.

The Quiz Tool and collaboration

Observations from teachers in the Quiz Tool project indicate that doing quizzes in the class or in a group will motivate learners with JNCL to participate, be active and collaborate with peers, rather than working alone. Many students with JNCL preferred to work in teams and discuss solutions to quiz questions before answering. All the students in the class seemed inspired by the quizzes and tried to achieve a common group response in a positive and inclusive way. In some cases the questions and the answer alternatives elicited long discussions, and the exchange of ideas between the students provided some golden moments of implicit and incidental learning. The use of the Quiz Tool in the classroom thus seemed to contribute positively to the teachers' efforts to build positive attitudes and teamwork in the class.

The special interests and quizzes of a young man with JNCL

Many of the students who used the Quiz Tool appeared highly motivated and clever despite their severe disabilities. A young man with JNCL was a big fan of the books about Harry Potter written by the British author Joanne "Jo" Rowling.

He had read all the books and had become an expert on Harry Potter. This great interest made him compose his own stories about Harry Potter in braille with incredible imagination. His publications attracted public attention and were exhibited in a Norwegian library. The young man learned to make his own quizzes using the Quiz Tool and became a very devoted user of the Quiz Tool. He has constructed and dispersed several quizzes based on Harry Potter books for friends and peers with and without vision. The young man is also an expert on the films and stories about Star Wars invented by the American author and director George Walton Lucas. For some time, he has been busy making quizzes based on the stories from Star Wars.

Conclusions

Quiz games are commonly used in education and for pleasure. They can utilize different modalities and may be adapted to players with different abilities and disabilities, including blindness and dementia. Research on dementia and experiences with elderly people indicate that quizzes may stimulate cognitive function and well-being. The findings of the present study, using the computer-based Quiz Tool, show that persons with JNCL can take part in quizzes both individually and in groups. The quiz seems to stimulate their motivation, learning and the maintenance of skills and knowledge. Quiz games make repetitions and over-learning possible if used in a strategic way within and beyond education.

The visual, tactile and auditory options of the Quiz Tool inspire collaboration and inclusion in mainstream settings because sighted students and students with visual impairment can play together on an equal footing. The Quiz Tool can further be used to assess the students' knowledge in different domains, and to find out whether the student is ready for the next level or rather may need more help in the zone of developmental maintenance (see Chapter 2). In this way it can help the teacher to collect information for evaluation purposes in a non-threatening manner. Observations further showed that the Quiz Tool can be used to steer the students' attention towards specific concepts or subjects, and to promote inclusion and pleasure in social settings.

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21

Games

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A game is a structured form of play for enjoyment and pleasure, but games may also stimulate learning and cognitive functioning. In education, games may give positive challenges, improve performance, or maintain skills and knowledge. Games can be competitive, like chess, or non-competitive, such as role play. There are group games and games that are played individually. Both individual and group games may be experienced as exciting and motivating. Leisure time activities include games of which there is a great variety.

Children with JNCL may participate in group games on par with their peers when the games are constructed or adapted to facilitate inclusion of players who are blind (see Ardito & Roberts, 2007; Tzvetkova-Arsova & Zappaterra, 2018). Consider a deck of cards. The individual card can be marked in a corner with braille characters or Moon letters (see Chapter 14) according to a system where the suit (Hearts, Spades, Clubs and Diamonds) is written with the corresponding letter and the denomination is similarly marked just underneath (with letters for Ace, King, Queen and Jacks, and a number for the other cards). Pre-marked cards may be obtained from a number of sources, including national organizations supporting people who are blind or have low vision, or from commercial vendors.

Dice are important elements of many games and can be constructed with embossed numbers to be read by people who are blind. Dice are used in Yahtzee and in board games like Ludo and Monopoly. Dice may serve as useful educational tools in addition to their primary use. Using dice provides cognitive exercising, particularly in games where two dice are used as in Monopoly or in Yahtzee where decisions must be made based on the face value of five or six dice at a throw. The tactile properties of texture and Velcro can be used to differentiate between game pieces. Attaching magnetic tape to boards and game pieces can reduce frustration arising from pieces being knocked out of position. The board games themselves can be marked in such a way that children and young people with JNCL may be able to participate with little or no intervention from an assistant, at least initially. There are many resources readily available on the internet (e.g., http://www.pathstoliteracy.org/strategies/tips-adapt-games-children-vision-impairments).

Research indicates that intellectual and motor exercises might help people with dementia maintaining skills and abilities (see Chapter 5 and 7). Games provide mental stimulation and may function therapeutic and slow down mental decline in people with cognitive disorders, including playing strategic games, assembling puzzles, playing board games, chess or card games. Playing games may also support communication, literacy, and social interaction. Games that earlier were enjoyed by the young person with JNCL are usually a good starting point. Many games can be adapted to meet the changing needs of the young person as the decline in skills becomes more apparent. New games can also be introduced, but it is important to analyze the complexity and abilities needed.

Over time, it will gradually become more difficult for young people with JNCL to read the boards, the cards or the dice. Thus, there is a shift from *independent play* to *interdependent play* (see Chapter 16) when the degree of assistance when playing increases, but the feeling of participation (and indeed the gloating when winning) does not diminish even as these skills deteriorate. Verbal cueing and verbal orientating assistance become necessary and dialogue becomes a (pleasurable) part of the game.

Experiences of parents and staff

In the present project (see Appendix A), both parents and staff mentioned the use of games for education and leisure. A counselor and teacher expressed the following:

> *Throughout my 25 years of experience working with students who have JNCL I have experienced that games are very popular activities for students with JNCL. Playing games is stimulating their cognitive function and reducing anxiety, maintaining relationships leading to more enjoyable moments. I have also seen that students are concentrated, aware and conscious – playing games can actually help them retrieving memories.*

Popular games need to be modified in line with the progression of different declines for individuals with JNCL. Such modifications may entail more structured settings, more support from fellow players, or making key information available through touch and hearing. Modifications of games may support further engagement for games, as described by these parents:

> *We modify the original games and adjust them to her mental progress. She plays very often but she has always preferred quizzes. Her interest in games grew over time.*

The comments from parents and staff show that the amusement of participation in competitive activities is preserved throughout life for individuals with JNCL. A few parents have even reported that the children's interest for playing games grew with age. Winning games or the excitement of the possibilities of winning, is expressed by parents as a thrilling experience throughout life. The zest for competition somehow becomes a part of their child's personality, as described in the following two quotations:

> *He's really into the games on the iPad like Battleship and Plants Versus Zombies. He plays it and he talks about it whenever there is a listener.*

> *He loves to play Sponge Bob and video games, but he loves to talk about it even more.*

However, staff and parents see games as more than just an activity where one can win, lose or beat another person or group; ordinary daily activities like shopping, walking to school, or reading can be made into a game. The following teacher is using shopping to teach mathematics:

> *Learning about money and mathematics was done through playing shopping and handling money. We used reading games to learn how to use a magnifying glass and other technical aids.*

The extent and quality of a child's engagement in an activity is a matter of how the activity is perceived by the child and how the exercise is presented to the child. An activity may appear boring unless steps are taken to make it more like a game that is enjoyable and meaningful to the child. Games may thus serve an additional purpose beyond the actual playing. Many parents and staff members emphasize that strategically adapting potentially "boring" activities into ones that have the properties of games is an effective way of enhancing learning and maintaining knowledge. Participating in activities attached to educational themes, objects or topics, was perceived as playing games by the participants with JNCL, sometimes resulting in astonishing and positive side effects on learning:

> *A calendar box was introduced to him to push the button to communicate when he wanted to eat or read, but he looked at it as more of a game. He learned braille by doing this. The same with the Go-Talk – a speech augmentation device – he learned how to use it through playing.*

> *She learned a lot about nature via the game «Der Natur auf der Spur»* («On the trail of nature»). *It was about nature, it was very informative. The game included pictures of trees and various questions and so on and she learned it all. But it was not her hobby. She preferred reading and listening to music or to cassettes.*

> *We thought this game would be done only once, not a long-term enjoyable activity. We play with a large collection of porcelain cats. Our daughter's role was to name the cats, touch and explore them, to describe their size, length, thickness, type of tail, placement of the ears, and so on. She always asks for this exercise or game. I can even ask her «What colors do the cats have?» and she can manage doing that as well. It is remarkable and very pleasing that she is able to devote herself to these cats for such a long period of time. This game is good memory training, my daughter is not aware that I'm actually working with her, and that's the wonderful thing about it.*

Games have been used to achieve different kinds of skills and knowledge like in this case:

> *In the game "Performing magic" something is placed under a cloth and our son must say what it is by exploring it in a tactile manner. For instance, he learned the difference between metal and plastic by doing this. Similar methods were used to distinguish between different sounds and so on. We do all these things and they may not only work well, but he has a lot of fun doing such exercises.*

A teacher of children with JNCL emphasizes the use of quiz games to achieve learning:

> *Quiz is just a wonderful way of combining pleasure with learning. «What's the capital of ... and which river passes through ...?» Then they answered and were very proud and happy. That was helpful. Or we utilized their knowledge of foreign languages: «Which language do you speak?» or «What is good morning in French?» and so on, that's all possible if it accords with the interest or knowledge of the kids.*

The use of Quiz is described further in Chapter 20.

Some parents emphasized the potential of using games to achieve learning and participation in schools. One parent mentioned games from Royal National Institute of Blind People:

> *There are some games in the school like dominos and different RNIB games marked with braille. I'd like to think [laughs] that there might be someone that could play a game with him, but it does not happen, you know – adults could facilitate but – I don't know. I don't think they understand the potential in using games as a motivator for learning and socializing.*

An educational counselor expressed the following:

> *I have experienced that the disease can cause, unfortunately, individuals with JNCL to withdraw from everyday relationships and activities with friends and family. Playing games is one of many ways of maintaining and building up such relationships.*

Modifications and engagement

The visual impairment is the first of several barriers to participating in games. Most games require vision. There are games adapted to the needs of people with blindness and sources of ideas for how to modify them or for commercially available adapted equivalents, but they are not generally known. It is always a danger that declines will lead to non-participation in playing games if not met by appropriate measures like in this case:

> *She liked playing cards – she knew how to play cards. But it was difficult when she got blind. She would have been interested in it. This interest was replaced by listening to audio books.*

The example illustrates many dimensions of the compounding problem: loss of participation opportunities may lead to losses in other domains, such as socialization, communication, emotional engagement and expression, and so forth. Although listening to audio books can be pleasurable and educational, and may provide topics to discuss with others, it becomes a passive activity if not shared with others. Passivity has been noted as a concern in adolescents with JNCL (see Chapter 4).

Participation opportunities in play are also lost because of other factors than deterioration of vision. The decline in communication and onset of childhood dementia may make situations with game playing rather complex, in particular when the child or young person has problems in remembering what happened last or when the overview of the playing activities is lost. Some parents and teachers have the experience that participating in one-to-one games is easier for individuals with JNCL when communication problems and childhood dementia become more apparent. Moreover, many parents and teachers emphasize emotional engagement by those participating as a key factor to achieving pleasure and entertainment in activities:

> *Playing games works when people enjoy the game as well.... yeah. For him at the moment the thing that engages him most is his physiotherapy session. He's most alert and engaged, he almost considers the physio as a game to play. I know we just felt really strongly that they (the helpers) needed to be engaged, you know, physically and mentally engaged, no matter where they were at.*

These parents' comments emphasize the fact that support is more than practical help; in fact, the staff's emotional engagement may be decisive for how a person with JNCL functions in the zone of developmental maintenance (see Chapter 2).

A variety of games

Many games are already accessible either because of their inherent properties, or because there are commercial versions appearing on the market. It is possible to make existing activities and products accessible to children and young people who are blind by adapting materials and rules (e.g., the complexity of a game) so that participation continues to be possible even in the face of declining vision and cognition. The following sections provide general ideas about the kinds of adaptations that are possible, together with explanations of how these modifications may support ongoing participation in game activities.

There is a rapidly increasing pool of online resources that can provide more ideas, and greater detail about how to adapt specific materials, where to obtain ready-made products, and how to integrate the materials and activities into activities of daily living, education, and leisure. Many popular games are now available as physical packages or as apps for mobile phones, tablets, and other computerized media. Large screen television and computer monitors, particularly those with touch screens, can increase the size and luminance of popular games, rendering them more accessible than they would be if presented on a tiny screen.

Online searches can include key words for games for specific populations (e.g., people with dementia, intellectual impairment, or visual impairment) in addition to games according to skill sets (e.g., strategy, memory, words, numbers), and the medium (real or virtual on an app). National support organizations maintain extensive online lists of resources and can serve as good starting points for searches.

Board games

Creating words in board games may promote learning in reading and writing, and such games are available for persons with visual impairment. *Scrabble*© (Hasbro) can be shared with sighted peers. Scrabble with braille works best for advanced braille users in collaboration with sighted helpers. It is possible to purchase versions equipped with everything needed for individuals with JNCL: braille tiles, tactile game board and braille and audio instructions. The game board is pasted

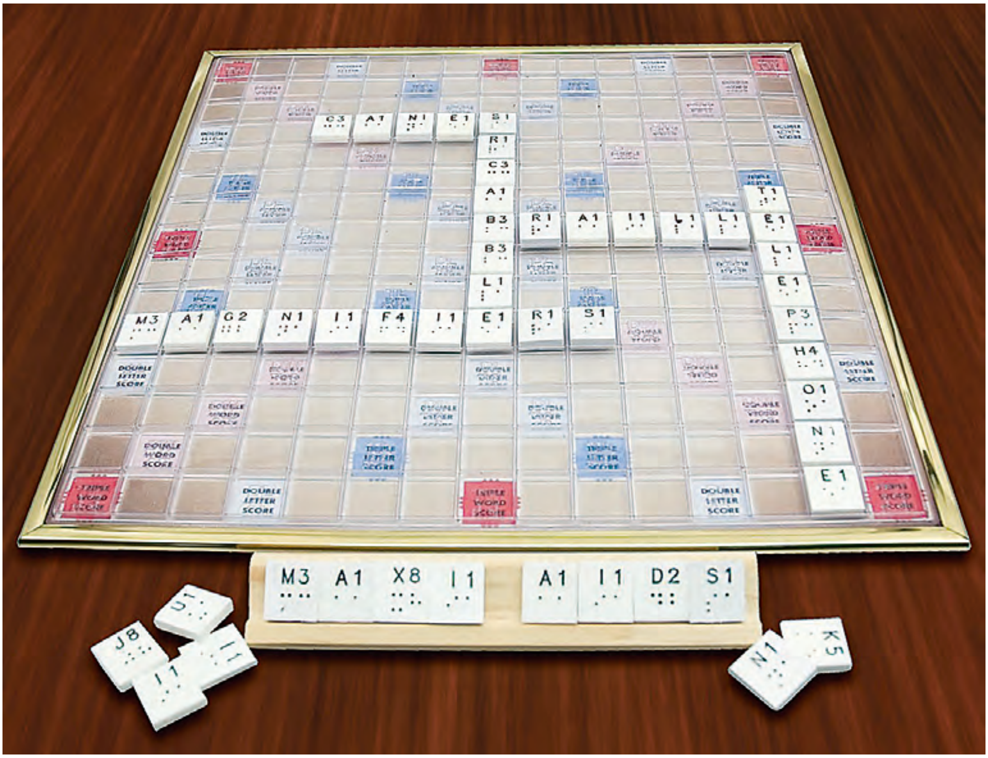

Figure 21.1 Scrabble with braille

onto a wood backing and covered with a clear hard plastic film that is ridged along each cell and acts as frames in which a player places a tile. Each cell has printed information and letter tiles with raised braille markings.

The *Scrabble* word game shown in Figure 21.1 is rather advanced. There are simpler versions of word games, such as the *Bananagrams©*. The Bananagrams set consists of 144 pieces in braille and print on a white background. It is easier to master because the young person with JNCL can form words in collaboration with family and friends. The words are readable for players with full sight, low vision and blindness. The manufacturer supplies a version for people with low vision *(Bananagrams Big Letter)*.

Choice Words© (MindWare) is a rather simple but funny game that involves auditory recall by asking students to identify words, terms, titles, and phrases that involve the root word. For example, the root word *cut* may stimulate recall of phrases that include it, such as *cut rate, cut throat, haircut, paper cut, cold cut, short cut, cutting edge, cutting board, cutting room floor, cut-and-paste, cut-and-run*, or *cut it out*!

There is a commercial braille version of *Monopoly©* with dice and cards in large print fonts and braille *(Braille and Low Vision Monopoly)*.

Blindfold Sudoku is for players with sight and visual impairment and available as an app (iOS). It comes with five Sudoku 9-by-9 puzzles – easy, medium and difficult – for audio play and is controlled through iPhone gestures. Students solve the puzzle by listening and imagining the puzzle layout in their mind.

A tactile-based adaptation of *Nine Men's Morris*, a traditional strategy game, consists of a grid with twenty-four intersections or points. Each player has nine pieces or "men" that usually are colored black or white. Players try to form "mills" – three of their own men lined horizontally or vertically – which allow a player to remove a man from the opponent. A player wins by reducing the opponent to two pieces (when it is no longer possible to form a mill and thus winning is impossible), or by leaving the opponent without a legal move.

Follow your Nose is a game that encourages exploration and discovery through the sense of smell. It stimulates both the olfactory sense and cognition while it elicits reminiscences. There are about 30 distinct aroma diffusers in tamper-resistant flasks which the players match to the corresponding images. The images must be adapted to tactile or auditory use to make the game accessible for persons who are blind.

Matching games

In these games the players match an object to a word in print or braille, a photo card or similar. Good games to play may include guessing games with a box, sack or tray containing familiar objects that the players must identify. These may

Figure 21.2 Game tray with matching pairs

include everyday objects which can be named, classified or sorted. This is a useful game to support key vocabulary. Figure 21.2 is an example of a matching game.

Figure 21.3 shows a game where the players are matching a card with a braille or printed word to a corresponding object. Games that stimulate mental capacity through word-related activities, such as word identification and odd one out, help support and maintain word identification and memory.

Dominoes is a family of matching games that easily can be modified for players with low vision, blindness and cognitive disorders (see Figure 21.4). The dominoes can be played with the tiles placed face-down and mixed. Each player selects seven domino tiles which are placed in front of the player but hidden from the opponents. The remaining tiles constitute a drawing pile. The player with the highest double (six-six) starts the game by placing the double on the table. During each turn, the player selects a domino tile from his pile that matches at least one side of one of the domino tiles on the table. A new domino tile is picked from the draw pile if none of the players'

Figure 21.3 Game where words are matched with corresponding objects