

Figure 21.4 Dominoes with four different forms of adaptations

dominos can be played. When the draw pile has been depleted, the current player must pass without making a move. The first to place all the tiles is the winner. Playing Dominoes may support the maintenance of number recognition, counting and many other cognitive skills, as well as competitiveness and the motivation to win. Versions designed for people with blindness and low vision are commercially available.

Memory games

Memory games may promote concentration and memory skills. There are many memory games such as apps, tactile games and digital games.

The Sarepta digital memory lotto game (Sarepta Studio, Norway) is a tactile and audio version of the classic memory card game (see also Chapter 19). The purpose of the game is to challenge the memory by linking corresponding sounds, words, pictures and so forth in pairs in categories like animals, nature, transport, humans or geography. It can be used to make the player familiar with areas of special importance or interest.

The *Audio Memory Game* in Sarepta uses a speech synthesizer. There are 16 hidden mixed audio cards numbered from 1 to 16. There are eight identical pairs within the 16 cards, and the players have to find two matching cards. The game will continue to the next player if a player makes an incorrect matching of cards. The players navigate from card to card by pressing a switch, the speech synthesizer will identify each card, for instance by saying «*card number one*» followed by a sound from a *dog barking*. A similar card with a dog barking can be found from for example card number eight. It is important for the student to listen for clues and remember them when other players are doing their trials. The game is over when all the cards have been matched, and the winner is the player with the most matches. The Audio Memory game can be played individually or as a group activity.

The purpose of *Shake Loose a Memory* is to recall memories and share emotional events with others (reminiscing). The game can provide social interactions and stimulate to meaningful discussions. The game consists of a single die and cards. Participants roll the die and receive a corresponding card. Each card starts with a simple instruction, like *keep this card if you have been a scout*. If yes, the student will respond to a question on the card, such as *remember earning any badges?* If the answer is no, the player will be asked to roll the die again or the game will continue to the next player. This game is not about *winning* or providing the *best* answer. It is about maintaining memories and sharing the personal history and good memories with others, in many ways imitating an ordinary conversation. An important consideration with games of this type is the importance of having a group of people playing together who actually share memories and are able to co-construct, and as cognition declines, this consideration assumes greater significance. The game can be adapted to very local, very immediate experiences (e.g., events at school this month, shared TV viewing, shared sport event participation, family activities this month) to accommodate memories. This and similar games can contribute to making classmates more acquainted with the student's home situation and leisure interests, or the family more acquainted with the student's situation at school.

Strategy games

These games require planning and finding solutions, and there are many old favorites, such as *Monopoly* and its numerous spin-off variations based on different themes and places. A quick online search will locate many versions.

Many children find *Connect 4* exciting and entertaining. This game is played by two players and may be played by individuals with low vision or blindness. Each player is given their own plastic discs that can be identified by color or

tactile recognizable patterns, for example black Velcro stickers. The game is about dropping colored discs from the top into a seven-column, six-row vertically suspended grid. The pieces fall straight down, occupying the next available space within the column. The objective of the game is to connect four discs of the same color or tactile pattern next to each other vertically, horizontally, or diagonally before the opponent (see Figure 21.5). The game may contribute to maintaining number and counting skills, tactile discrimination and memory, and the most important goal of all – to have fun. At the simplest level, the game can entail no more than turn-taking between participants in dropping in the discs and enjoying the resulting noise. Even this level of the game promotes active participation and socialization by simplifying the "rules" and thereby making it fun. It is possible to purchase large Connect 4 games for indoor or outdoor use. The large versions are easier to manipulate if the player has problems with fine motor skills. A small hole may be drilled into one set of counters or discs to aid identification.

Pass the bomb is a thrilling game (Figure 21.6). The players sit in a circle while the bomb is passed around the group. The players are asked to think and express a word connected to the *Theme of the Day* when holding the bomb.



Figure 21.5 *Connect 4* with black Velcro for tactile discrimination, red is the winner!

Themes can be things that are found in a supermarket, classroom, bedroom, or related to a special event, for example Christmas or summer holiday. The aim is to not be holding the bomb when it surprisingly goes BOOM! This game may stimulate memory through word-related activities and word identification. It helps improve the ability to recall by encouraging the recollection of memories via verbal interactions or pictures. Old fashioned versions include *hot potato*: no bombs go off, but there is plenty of noise when the player left holding the hot potato screams in dismay!

Noughts and Crosses (also known as Tic-Tac-Toe) is a strategy game that can be composed of a wooden 3D set (see Figure 21.7), or it can be accessed as an app for mobile devices. The players take turns to place their X or O into one of the empty squares in the grid by clicking on it. To win the game, the players must get three of their own symbols in a line horizontally, vertically or diagonally. Noughts and Crosses may help maintaining memory skills, tactile discrimination, strategy use and problem solving. It may also support the maintenance of number and counting skills and fine motor skills. The competitive element appears to enhance the players' enjoyment of the game.



Figure 21.6 The bomb card game

Shut the box is played using dice and a box that has nine levers on hinges, each marked with a numeral and there are one-player and two-player versions (see Figure 21.8). The object of *Shut the box* is to shut all nine levels. One player rolls both dice onto the green baize floor of the box, and shuts the lever(s) according to a number or numbers face up on the rolled dice. If a player reaches a position where the number rolled has already been used to close the corresponding lever, the play passes to the second player. At the start of the game, all levers or tiles are open, showing the numerals 1 to 9. Each player plays by throwing or rolling both dice into the baize. The player adds up the dots on the dice and then "shuts" (closes, covers) one of any combination of open numbers that equals the total number of dots showing on the dice. For example, if the total number of dots is 8, the player may choose any of the following sets of numbers (provided that all the numbers in the set are available to be covered): 8, 7+1, 6+2, 5+3, 5+2+1 and 4+3+1. The player



Figure 21.7 Noughts and Crosses (Tic-Tac-Toe)



Figure 21.8 Shut the box

then rolls the dice again, aiming to shut more numbers. The player continues throwing the dice and shutting numbers until reaching a point at which, given the results produced by the dice, the player cannot shut any more numbers. At that point, the player scores the sum of the numbers that are still uncovered. For example, if the numbers 2, 3, and 5 are still open when the player throws a one, the player's score is 10. Play then passes to the next player. After every player has taken a turn, the player with the lowest score wins. If a player succeeds in closing all the numbers, he or she is said to have "Shut the Box" and wins immediately, and thus the game is over.

In *Build a Beetle* players take turns throwing a die and cannot begin building a beetle until they have thrown a six, which allows them to take a beetle's body (Figure 21.9). A larger tactile die

may be introduced as and when the regression of skills becomes apparent. When a six is thrown, the player has a second turn, and this rule applies throughout the rest of the game whenever a player throws a six. Once the body has been taken, the subsequent parts are selected when the appropriate die value is thrown, as follows: 5 gives the neck, 4 gives the head, 3 gives the tail, and 2 and 1 give a leg. *Build a Beetle* provides tactile stimulation and stimulates number recognition, counting, communication and memory, and helps maintain fine motor skills. The game is competitive and many young people with JNCL find it enjoyable.



Figure 21.9 The Beetle Game with an ordinary and a tactile die

Construction games

Most construction games can be played individually or in collaboration with others. *Meccano*, *Lego* and *Duplo* are examples of constructions games where children and young people are stimulated to construct vehicles, bridges, helicopters or cranes. Many of these games promote children's interests in areas like science, technology, engineering and mathematics.

There are many construction games suitable for individuals without sight. Many structures in the surroundings cannot be perceived as instant holistic features by children who are blind. Structures may be easier to conceptualize through the use of construction games. For instance, building a bridge may make it easier to perceive or understand the structure of a bridge when vision is no longer available. Construction games can be used strategically in education by enabling children and young people with JNCL to act as constructors.

Construction games promote fine motor skills and incorporate different learning styles, such as learning by doing. Learning about physical structures and to imagine them and form a holistic representation without vision might be difficult. Such challenges can be compensated by organizing settings where the child becomes the constructor of a structure (e.g., a spaceship) or a part of the

structure (various projections from the spaceship). Providing the necessary support for the child's construction might facilitate the ability to form an overall concept or idea about the structure based on both earlier visual experiences and present tactile knowledge. By creating different kinds of space craft, the student can build up conceptual knowledge at a categorical level that serves as an overall template, together with the various optional features that might create subcategories. In typical development children learn to construct houses, barns, skyscrapers, and so forth. In so doing, they discover natural categories of "buildings" and elements that may be used to create specific structures.

The picture in Figure 21.10, sent by a parent, shows a young boy with JNCL building a space shuttle with Lego. Lego building has been one of his hobbies for a long time and his skills in mastering Lego are astonishing. The parents and his teacher believe the tactile skills developed through playing with Lego has been a vital asset supporting the boy's performance in reading and writing braille.

Construction games can be used for preparing the child to learn skills within reading, writing and mathematics and the understanding of objectives and

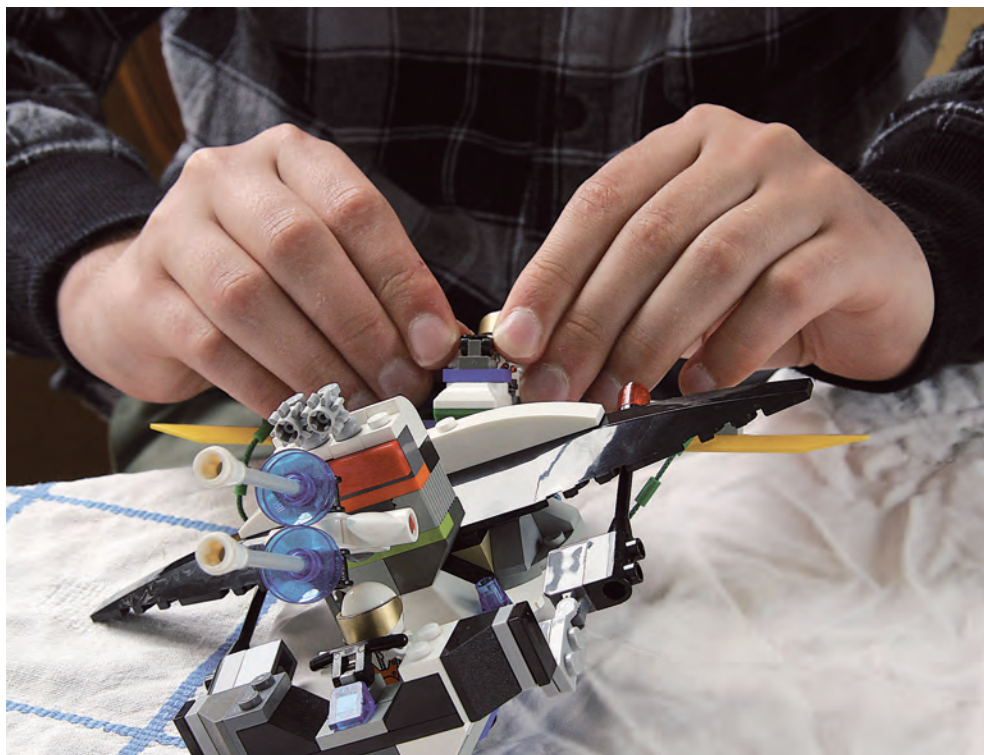


Figure 21.10 A boy with JNCL constructing with his Lego

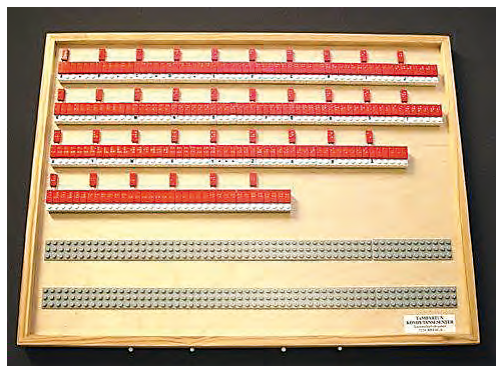


Figure 21.11 Lego braille frame and two examples of bricks with braille letters

structures in the society that otherwise cannot be perceived by other means than through vision. Lego and other construction games have been used as motivators to achieve reading and writing skills in braille. Lego bricks have also been used to teach letters and construct words and sentences (see Figure 21.11). Construction games may support the understanding of shapes and spatial relations, as well as number recognition and counting.

Other games

There are many other games available for young people with visual impairment, like *Chess*, *Othello*, *Parcheesi*, *Checkers*, *Chinese Checkers*, *Racing Crown*, *Tactile Backgammon*, *Snakes and Ladders*, *Rubik's cube with touch marks*, *Sudoku in color and braille* and *Tic-Tac-Toe*, *Slim*. Note that many sports games can be adapted as well (Ardito & Roberts, 2007).

Conclusion

Games are a source of enjoyment and learning. Playing games is important for all human beings, particularly children and youth. Playing games encompasses personal entertainment, socialization and enjoyment – important aspects of the quality of life. This short review shows that it is possible to adapt games of all types to enable children and young people in different phases of JNCL to participate actively and with enjoyment, and how participation can support both development and maintenance of many skills. Games provide mental stimulation and may be considered as therapeutic for people with cognitive disorders. Playing games may in fact contribute to delaying the decline of functions in the zone of developmental maintenance.

In sum, playing games should be an option for most young people with JNCL and might be used as an effective method for achieving learning and preservation of skills in education. An added benefit of playing games in groups is its contribution to building social affiliation and a group identity. Being in a competitive mode may contribute to increased motivation for learning and may enhance patience and skills during performance.

References

- Ardito, M., & Roberts, J. (2007). A catalog of physical activities for visually impaired youth. Bachelor thesis, Worcester Polytechnic Institute
- Celeste, M. (2006). Play behaviors and social interactions of a child who is blind: In theory and practice. *Journal of Visual Impairment and Blindness*, 100, 75–90.
- Giannakopoulos, G., Tatlas, N. A., Giannakopoulos, V., Floros, A., & Katsoulis, P. (2018). Accessible electronic games for blind children and young people. *British Journal of Educational Technology*, 49, 608–619.
- Neugnot-Ceroli, M., Gagner, C., & Beauchamp, M. H. (2015). The use of games in paediatric cognitive intervention: A systematic review. *International Journal of Physical Medicine and Rehabilitation*, 3 (4).
- Rettig, M. (1994). The play of young children with visual impairments: Characteristics and interventions. *Journal of Visual Impairment and Blindness*, 88, 410–420.
- Tzvetkova-Arsova M., & Zappaterra, T. (2017). Play in children with visual impairments. In S. Besio, D. Bulgarelli & V. Stancheva-Popkostadinova (Eds.), *Play development in children with disabilities* (pp. 181–200). Berlin: De Gruyter.

The Social Life of Children and Adolescents with Juvenile Neuronal Ceroid Lipofuscinosis: School and Participation

*Stephen von Tetzchner, Anne Margrethe Andersen,
Anne Hagedorn Hamann and Susan Fugger*

Social relations and participation are core elements in human life. Social participation includes family, peers and the community. Parents and siblings are usually children's first social relations; they have significant influences on the children's development and continue to remain important in adulthood. The mutual influences between children and parents are important but children also engage in interactions with peers in events and activities inside and outside of school and kindergarten. Peers have a prominent place in children's social lives from an early age, and with age peer relations become gradually more important (Chen, French, & Schneider, 2006; Rubin, Bukowski, & Bowker, 2015). Observing and interacting with peers help younger children learn collaboration, conflict management, moral reasoning and other social skills. Among older children and adolescents, peer relations also contribute to the development of identity and a broader understanding of emotions and thinking in other people.

This chapter is about the development and maintenance of peer relations and social participation of children and young people with juvenile neuronal ceroid lipofuscinosis (JNCL). It is a common finding that disabilities will influence children's interaction with peers (Schneider, 2016). The declines in vision, cognition and motor skills of individuals with JNCL will influence their friendships and peer relations but the present study (Appendix A) shows a varied pattern of involvement in peer interactions in this group.

Vertical and horizontal relationships

Children and adolescents engage in vertical and horizontal relationships. In vertical relations, one part has more knowledge and social power than the other, and the relationship is asymmetrical and complementary. Adult-child relationships are typically vertical. Adults have control and children are given protection and

security. Children seek knowledge and help, and adults provide it. Peer relations are usually horizontal; the social power is more equally distributed, although children may have different roles when interacting. One throws the ball, the other catches. One dresses the doll, the other pushes the cart. Peer relations are important because they offer experiences and learning that vertical relationships cannot provide, including both competition and cooperation (Bukowski & Adams, 2005; Hartup, 1999).

Sibling relationships are usually based on some sense of being equals, even though differences in age, development and physical strength may imply differences in status. When one sibling has a disability, this is likely to change the relationship in some ways, and it may become more vertical than the age difference indicates. The relationship is basically the same as for siblings without disabilities but it is characterized by more warmth and care than other sibling relationships (Meltzer & Kramer, 2016; see Chapter 25).

Friendship is a horizontal relationship characterized by equality, symmetry and a strong emotional bond. Friendships may have positive influences on children's development: Friends develop social skills and understanding together, talk about themselves and others, and gain insight into moral values, conflict resolution and social and cultural rules (Bagwell & Schmidt, 2011; Dunn, 2004). Children who have friends generally show better development, adaptation and performance at school than children without friends. Friends have more fun together, often engage in more advanced forms of play, and solve school assignments better together than nonfriends (Bukowski, Motzoi, & Meyer, 2009; Hartup & Stevens, 1997; Laursen & Pursell, 2009).

Activities, peer relations and JNCL

Disabilities may influence a child's activities and losing acquired skills may be experienced as dramatic. A negative impact of reduced ability to manage everyday activities has also been found in elderly people with dementia (Barca, Engedal, Laks, & Selbæk, 2011).

Equally important, the loss of skills may influence friendships and other peer relations. Children with visual impairments may be excluded from many activities and tend to have fewer friends and smaller social networks than children without such impairments (Salminen & Karhula, 2014). There are many friendships between children with and without a disability but the prevalence of true reciprocal friendships involving children with severe disabilities is lower than friendships among children with typical development. Peer interaction may depend on adult intervention and "friends" with typical development may sometimes act more as

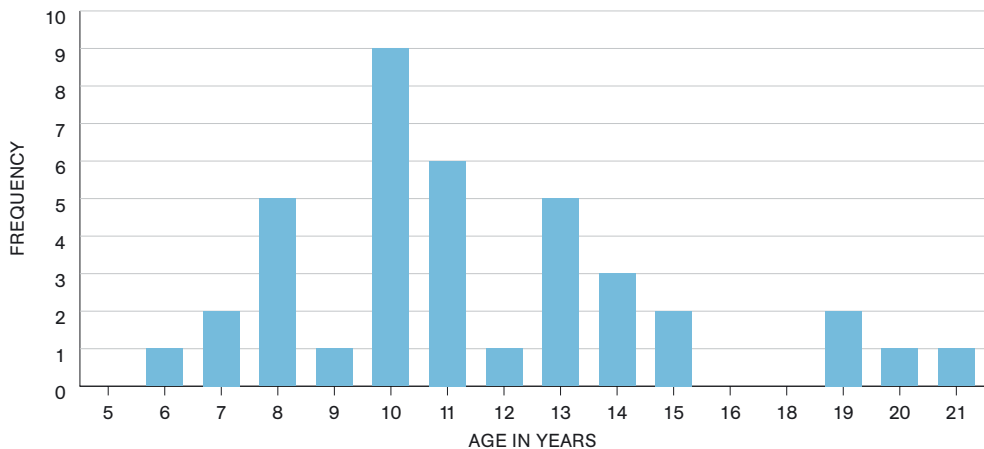


Figure 22.1 Frequency of first observation of loneliness at different ages

helpers than as friends (Guralnick, Neville, Hammond, & Connor, 2007; Rossetti & Keenan, 2018; Sterrett, Shire, & Kasari, 2017).

For children and young people, the school is the most important social environment outside the home. Education and the educational setting are strong contributors to children’s social and emotional development and sense of identity and place in the world (Abdi & Guo, 2008). Children who are blind may attend a mainstream school but find interaction with peers difficult (Opie & Southcott, 2018). Also, intellectual disability tends to make social interaction with peers difficult and to increase the probability of loneliness (Gilmore & Cuskelly, 2014). In the present survey (Appendix A), the first signs of loneliness often appeared in late childhood and early adolescence, but sometimes earlier or in late adolescence or early adulthood (Figure 22.1).

For students with JNCL, the school and classroom may be the only sources of social interaction outside the family, and as the disease progresses social interaction in school may also become limited.

Well, she doesn't really have a social life. To be fair she doesn't have any friends as such outside school. She has got a supportive boyfriend; we do see him on a regular basis, every week.

Because of his disorders he lived at home isolated from other children. The kids didn't want to play with him any longer. From age eight he lived in a residential home during the week and in this way he was in touch with kids having the same problems. They were all the same.

How students with JNCL prosper and function at school will depend on the social environment, the relationships with other students and the degree of adaptive education. Loneliness is inversely related to acceptance and mutuality in the peer social environment (Newson, Mallow, & Watson, 2015). Loneliness is more frequent in young people who are blind, and a lack of mutual friendships will lead to loneliness (Gold, Shaw, & Wolffe, 2010). This quotation is from a young boy with JNCL.

I have become one of the world's most lonely persons, I don't have any friends, and my earlier friends stopped visiting me after I became blind.

Shared activities form the basis of early friendships. When preschoolers with typical development are asked about their friends, they answer that they are friends because they do things together. Participation in joint activities is an important criterion for friendship among older children and adolescents as well. With age, however, perceptions of friendship increasingly involve feelings of perceived closeness and shared emotions. Friends are understanding, loyal and can be trusted. Older children associate friendship to a greater extent with having similar attitudes and values; yet their friendships continue to be based largely on shared enjoyment of the same activities and the same music. In adolescence, descriptions of friendships additionally begin to include shared secrets and feelings, and opportunities for emotional support (Bukowski et al., 2009; Mathur & Berndt, 2006).

In the early years, children with JNCL engage in social interaction in the same ways as their peers, based on joint activities, but quite soon the visual decline makes participation in many activities involving gross motor skills difficult.

That is something that we have seen declining over the last year and a half when she started here in fifth grade. We had some issues last year and it's continuing to be an issue. She does not have very good relations with a lot of her peers.

Childhood is characterized by a high degree of gender-specific activities. Boys participate more in sports and group activities, while girls more often collect in smaller groups and are more preoccupied with intimacy and who is in and outside the clique (Maccoby, 1990). This may also influence the peer interactions of children with JNCL.

His loss of vision made participation in favorite activities like football in the break impossible, and because of this he also lost interaction with his peers. Some of the girls wanted to be with him but he usually ended up in a group with two or three girls, he talked about other things than they were interested in. He was interested in fantasy stories with dramatic events, spaceships and wars, and the girls talked about their own things and only half-heartedly engaged in his play.

Change of school may function well from a narrow educational perspective but may be more problematic for social participation and relationships because it is difficult for a young person with JNCL to make new friends. It is a gradual process, some maintain friendships longer, some lose them in transitions, and this should be a topic when transitions are planned.

The loss of sight was the reason for the change of school. The loss of friends followed. After that the increasing loss of skills made him restless. He became more isolated. In some stage of restlessness it was not possible to have him in the classroom any longer. On the other hand he enjoyed to stay with people, especially when they were in a good mood, when there was action and noise.

It is important to emphasize that the loss of friends is usually not absolute and a few regular friends may be sufficient for a good social life.

Her social life at present is amazing. There are 12 girls on this unit; we have got a boy unit upstairs and a girl unit downstairs, and a mixed unit with youngsters in chairs. She sees and hears them every well five days, four nights a week Monday to Friday. We socialize quite a lot together for birthday parties and other events, as I say going out to the theatre, it's not just the girls, and it's the boys and downstairs. She meets outsiders who come in; we've got young people as volunteers.

John has always had a good social life. And I would say that action and having a social life always made him smile and he loved taking part. He always enjoyed going out on visits or people visiting him. Going to places. And he would also be well visited by the familiar people. Familiar people are always very important but he also liked meeting new people. But he did have a little glint in his eye too as well because he would sometimes see how far he could go too as well in maybe trying a chat up line or something like that.

People with dementia are typically very dependent on other people (Wogn-Henriksen, 2012). Perhaps the most difficult thing about having dementia is the consequences the disease inflicts on the quality of being together with others, not least the closest ones. If a person cannot communicate verbally as well as before, the roles in the family and the relationships with friends change. For everyone, whether they have dementia or not, maintaining close and open contact with those who matter most to them is important for quality of life, as well as security and stability in daily living. The cognitive decline may manifest in many ways. With age, the activity repertoires of students with JNCL gradually get smaller, and their interests diverge from those of peers. As a result, they may be perceived as less interesting communication and interaction partners. Several parents describe this situation.

Peers usually approach and play with Thomas for a limited time, then lose interest when he can't engage in their games. Staff will sometimes endeavor to lead and steer games that can include all but these games are hard to sustain.

It is more and more difficult for him to be with his class. They are growing from him, have other interests and find it difficult to talk with him.

Because interests and activities are an important basis for friendship and peer relations, the interests of children or young persons will also influence their social interaction and participation in general.

His social life really is driven by how he is at the moment, which is that he gets very frustrated and he gets very anxious. So his social life at the moment is just very restricted to being in the lounge with the other young people. Listening to movies. It is very much driven by him at the moment. He does seem to prefer just to be quiet, knowing what's around him and understanding what's around him.

It is not only the range of the person's activities that may be limited, but also the type; some students with JNCL develop special interests that are not shared by peers, and this may exclude them from social interaction. For example, one 14-year-old girl with JNCL had a special interest in Sleeping Beauty and other fairy tales, but the classmates did not share this interest. Her participation in other class activities was reduced and the lack of shared interest made her generally more passive. This example depicts a problem, but also points to a solution: intervention that fine-tune or adapt the special interest may make it more inclusive. For example,

many fourteen-year olds have an interest in mythology. It is a short step from an interest in one fairy tale, to an interest in folk stories, legends and mythology from the world. This example illustrates that a small change may have cascade effects across a teenager's social and psychological development or maintenance.

Other students with JNCL develop special interests and advanced knowledge in areas that others in the class are interested in and which may in fact be relevant for the whole class. They can contribute to classroom activities provided the teachers implement the necessary measures and integrate the special topics of the students with JNCL in the work of the whole class. Music is such an interest that many children and young people share.

Within school, socially she does have a little peer group and at break time, she does have lunch break and morning break, you know, she'll look for someone to maybe go outside with her to get some fresh air and she has friends who are also interested in music and they tend to sit and chat about music. So she does have quite a good quality of social life in that respect.

The reduction in social participation may thus partly be a result of the students with JNCL not participating in the peers' common recreational and leisure time interests. Young people with JNCL may lose contact with the youth culture that exists outside the school and have no part in what their schoolmates are preoccupied with. This can easily become a negative spiral that may lead to

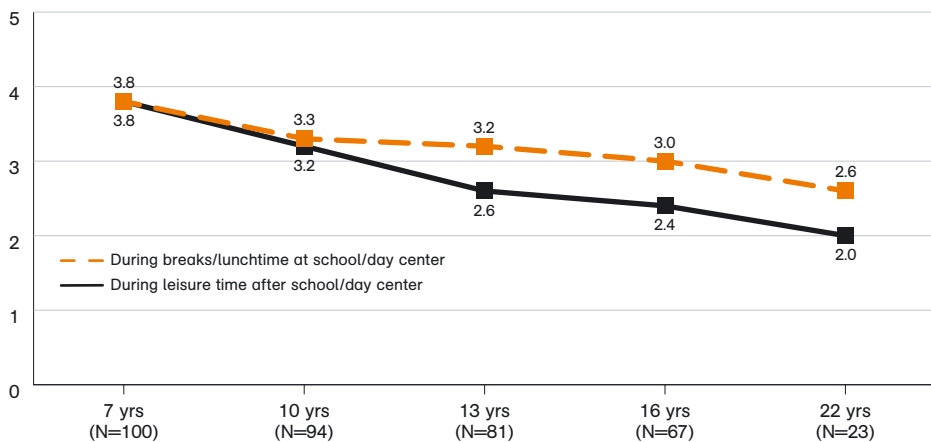


Figure 22.2 Interaction with peers of same age during breaks at school or lunch time at day center, and in leisure time outside school and day center

Scale: 1 = No interaction, 2 = Minimal interaction, 3 = Reasonable interaction, 4 = Pretty good interaction, 5 = Very good interaction

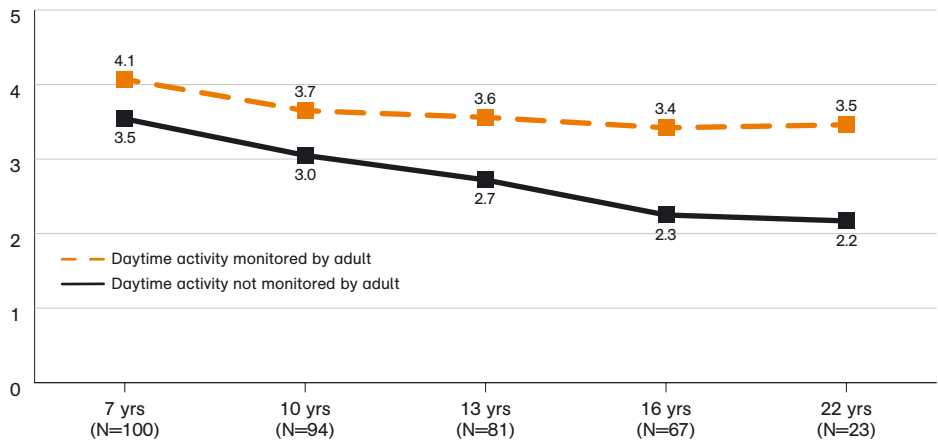


Figure 22.3 Social interaction with peers with and without adult monitoring across ages

Scale: 1 = No interaction with peers, 2 = Minimal interaction with peers, 3 = Reasonable interaction with peers, 4 = Pretty good interaction with peers, 5 = Very good interaction with peers

isolation of the student with JNCL both at school and during leisure time. Schools should therefore give priority to helping students with JNCL to participate in social activities and events. When teachers focus on themes and activities that may engage both the student with JNCL and the schoolmates, this can facilitate interaction between them. This requires that the teachers and other staff at school seek to identify possible shared interests.

The results of the present study (Appendix A) indicate that adult support and adaptation are crucial for social interaction between students with JNCL and their peers. The parents were asked to evaluate social interaction with peers with and without adult monitoring across ages on a scale from 1 (no interaction with peers) to 5 (very good interaction with peers). The importance of school as a social environment is documented by the fact that interaction with peers outside school and day center decreased faster than interaction with peers during breaks at school or the day center (see Figure 22.2).

Furthermore, interaction with peers was on average higher in activities monitored by adults and also the slope or the decline was smaller than in activities without adult monitoring (Figure 22.3)

The role of adults in monitoring social relationships was also mentioned by parents.

All social participation is managed by staff and other adults.

It is important to emphasize that reduced participation was not a result of a lack of motivation. Most of the participants with JNCL in the present project expressed a desire for social participation with peers.

And she would seek people out if she didn't know where they were. If it was particular people she wanted to chat to.

Her social interaction with peers is very limited. She does not approach peers in school and expresses a wish for being together with her boyfriend who attends the same school.

She has this befriender who comes and spends time with her. I think she is been shopping with her and she loves that. So she does arts and crafts with her and then mum takes her to see her friend and Tim so that is quite a good social thing. And she has tea with his family and things; she's quite a part of that as well. So yeah.

Several parents said that their child found the social loss the most difficult and emphasized that this situation should be remedied to the extent possible.

Adolescence is a particularly difficult time for young people with JNCL. The emerging cognitive, speech and motor declines become more severe, while social relations become more intimate and conversations more complex. In addition, adolescence represents new interaction patterns and romantic relations (Dunn, 2004).

Tommy lost his social life when he and his friends entered puberty.

She was able to keep her contacts to peers – two friends from elementary school – until she was 15 or 16 years old. She showed an extraordinary social competence and recognized the needs of others. When she was 18 or 19 it became very difficult and she frantically tried to keep in touch with people of her age. This didn't work out in the end.

The family is the social anchor for children and young people with JNCL, and with age the role of the family becomes even more important.

Ruth has no friendships outside the family.

Helmuth has no friends outside of his own Family. He has one roommate and they are good friends. The roommate has JNCL too.

In general, there is a reduction in peer interaction and social participation but it is still a varied picture that emerges.

She's got a nice group of friends that are very important in her life you know. She's got one boy and he teaches her drumming at lunchtime. She laughs a lot because school is a lot of fun for her. She's in touch with lots of the pupils and ex pupils on her phone at night phoning and emailing people so you know there's a lot of contact outside school and also with pupils that have left. It's quite nice that she is still in contact with them.

Including stories about romantic relationships.

She has a boyfriend who left school last year and the families keep in touch and he lives maybe about 30 miles away so I know that they go and spend weekends together, the families, so she gets to see her boyfriend and spends time with him.

These examples are important because they show that the reduction in peer interaction is not absolute, and some engage in romantic relations. Although peer relations in general were weakened, this was not always the case.

Social adaptation and intervention

The comments cited above suggest that it may matter what kind of support is provided to the young person with JNCL and that it may be possible to make adaptations and give support that contribute to maintaining social relations longer. The fact that it may be difficult to establish new social relations implies that trying to maintain some continuity in existing peer relations should be a main focus. This is also illustrated in the story about the Danish summer school below. When the parents were asked about the interaction with peers (Appendix A), there was a clear developmental pattern.

There were only few pupils in the class and the children and adults knew each other in a familiar environment. Interaction was good but when in a less familiar setting she was shy and liked to observe.

His social relations should be maintained with help from the staff, when he cannot do it himself.

In this example, the peers – with or without support – have found a way to compensate for the person's language decline and maintain interaction.

Social interaction is initiated by peers asking general questions around the person's favorite topic – high school musical or favorite sporting team. No initiation on the part of the person.

Similar strategies might be introduced to classmates by the teachers as declines become noticeable to both teachers and classmates.

Bullying also happens, maybe due to a lack of knowledge about the disease and its consequences, and it may be necessary to interfere in the peer environment.

The person's peers are generally accepting and tolerant however the first year of high school was very difficult. Some of the peers were very intolerant and would actively tease and try to cause anxiety in the person. This resulted in a new set of classmates for year eight which has been a huge improvement.

In school the breaks are important social events and school may make special arrangements to utilize the breaks as inclusive windows.

When Martha with JNCL began to lose her vision, the school established a system of break friends. Peers who wanted to spend some breaks with Martha put their name on a list. This became quite popular among the girls and they decided themselves that they wanted to be on the list. In the higher grades, boys in the class also asked to be on the list. Martha was very happy with this arrangement and asked every day who should be together with her in the breaks.

In addition, peer interactions may disclose issues that should be addressed in the education.

Martha's speech was not affected but she was not always aware of what was acceptable to talk about. The classmates were sometimes embarrassed when she talked about her own body and other private and intimate things. Observing this, the teacher initiated special sessions with Martha about advisable topics for conversations with break-friends.

Martha's story may also demonstrate the importance of peer continuity. It would probably have been much more difficult to establish a system of break friends if she had been new in the class and the classmates had not known her from the first grades.

The Danish summer school

As self-initiated and independent social participation gradually become difficult, the importance of organized activities increases. This part describes a Danish initiative to bring children and young people with JNCL together in shared activities and opportunities for social networking. Here is an invitation to our summer school experience!

One morning at the summer school: come on in!

Bacon and eggs are ready, together with coffee, juice and lots of Danish pastries. Of course, there is also oatmeal, hot and freshly cooked, standing steaming and ready to eat. The oatmeal is a special request from Ole. At home he eats oatmeal every morning, except for Sunday, when Ole, together with the family, eats bread from the baker. This is what makes Sundays special.

At all the tables everyone is eating. There are special plates where the edge is high and the food therefore stays on the plate even if the person is blind and has trouble with the fine motor skills. But first and foremost, this is a place with a great atmosphere; the room is filled with laughter, talk, lively debates about whether there will be an evening fire today or tomorrow, and each new entrant is showered with «good morning» greetings from all over the room.

During the meal, Peder stands up. Peder is Robert's assistant, or "supporter" as they are called in this assembly. And support is a good term; it means being the eyes, hands, body and memory for a child or a young person with JNCL. When Peder takes his place with a firm and safe hand on Robert's shoulder, this means that Peder and Robert are in charge of morning gymnastics. Or as Peder says: «Now, Robert wants to do morning gymnastics with you». Robert sits next to his chair with a big smile.

Anyone who can stand must get up, and then there will be lifting shoulders, shaking bodies, clapping, and all the hands are directed upwards as if trying to reach the ceiling, but nobody is that tall. A final highlight of the gymnastics comes when everyone is jumping on the spot making a good sound that is getting louder and louder as they stand there jumping. Phew! It is hard work at the morning gym!

Afterwards someone describes the weather and today's program. The task of recording "Today's sound" is allocated to one person and after dinner everybody tries to guess which sound was recorded during the day's activities. Then we are ready for today's. The first activity of the day is song and storytelling in the fireplace room. That is how a new day starts at the summer school.

JNCL Summer School

The NCL Team (see Chapters 9 and 24) organizes a one-week school event every summer for all children and adolescents in Denmark with JNCL. It is located at the Pindstrup Center in Djursland where there is ample space and a disability-friendly interior. Food and snacks are served throughout the day. There is a good atmosphere and helpful staff who will find scissors, paper and musical instruments from the stores when somebody has good ideas to be tested. The engagement and welcoming attitude of the staff at Pindstrup has a positive influence on the quality of the summer school. The rooms are distributed in the best possible way and special requests for food and drink are taken into account.

When the chef returned

For several years there was a chef at Pindstrup with a very special approach to the children and young people. He managed to create an atmosphere of peace and warmth. The chef could remember the children and young people from year to year, and it was clear that his presence became part of the summer school, activating both memories and energy. In 2016, for economic reasons, the chef had to leave and the food was delivered from the outside. This new arrangement was worrying, because it resulted in the loss of the former warm, supportive atmosphere for the children and young people.

However, the chef chose to visit the summer school in 2016, and his visit instantly created the good atmosphere and elicited many lovely memories. For the children and young people, and for supporters and consultants, it became an afternoon with a lot of smiles, and the chef reached all the children and young people with greetings and special remarks.

The summer school theme

The summer school is characterized by the pleasure of meeting new and old friends, learning and enjoying life. Every year, the summer school program is built around a special theme, such as "X-factor", "witches and trolls", "Nordic mythology", "Circus" and "Christmas". The basis for selecting a theme is that everyone in the group should be able to relate to it and contribute in accordance with their level of functioning. For example, everybody prepared for the great summer Christmas party, where everyone participated in singing and dancing with dresses and gifts.

Song and music

The summer school is characterized by song and music, which stimulate everyone's amusements and memories. A flexible Summer School Songbook has been prepared and has become something of a fond tradition. Old favorites are mixed with new songs, which are added as each new cohort of children adds songs to the collection. The songbook is much used throughout the summer school week and there is a special summer school song that is sung many times during the week. There is a great tradition of history reading and storytelling, and the program always includes afternoons with storytelling and songs where many express a wish for their favorite songs and stories from before and now, such as H. C Andersen's fairytales and stories from Nordic mythology, tales about Prop and Berta. All who can are singing. Most of the children and young people have favorite songs and they can sing or mime these even after their speech has disappeared.

Networking

A guiding principle of the summer school is the importance of children and young people being an active part of a social network. The summer school is a place where children can develop and sustain social connections; they get to know each other partly from prior summer schools, and also from regular activities sponsored by the Danish NCL Family Association. This network is often the only social network outside the immediate family circle that children and young people with JNCL maintain throughout their lives.

The support personell (supporters) of the children and young people with JNCL also form a network where experiences and best practices are shared. The summer school gives the support staff a lot of experiences in working with these children and adolescents, and for them the summer school represents a very useful «practice course».

Structure, predictability and the pedagogical foundation

Fundamental characteristics of the summer school are structure and predictability. There are sounds, pictures and many repetitions. The individual child or young person influences the program and how it is implemented. An important role of the support staff is to "translate" and interpret the wishes and tales of the children and young people. All children and young people have one or two supporters at the summer school and they are part of the long-term planning of the summer school as shown in the following excerpt of the invitation letter.

This letter to you as supporters is written because we from the NCL Team want to prepare the summer school in collaboration with you.

The role of the support staff in the summer school is, like in all the other network activities for children and young people with JNCL, to help to strengthen the fellowship among the participants including both the children and young people and the accompanying support staff. The summer school will in addition provide opportunities for learning a lot from each other.

One of the things we agreed to during the evaluation of last year's summer school was not to make the program too extensive and compressed. There must be room for inspiration and improvisation, creating good ideas on the spot and then pursue them, so the program for the summer school of this year is not so detailed. We want to allow room for the supporters and children to influence the content of the days – of course, based on the selected theme of the year. In that way, spontaneity and creativity will be the leading star of the forthcoming summer school and everyone are hereby invited to contribute to making the summer school amazing. The details of the individual days will be decided at our joint evening meetings.

We encourage you to prepare for summer school by talking to the children about the theme of the year (Christmas) and how they want it to be. How should the Christmas party be prepared, who would you like to be – Pyrus, Kandis or?

The Pindstrup Center has good facilities, so there will be ample opportunity to prepare this year's theme. The creative and practical preparations will take place in the workshop and on the first day, where we will delegate tasks and responsibilities, such as who will be responsible for each workshop.

You may think about everything from costumes to songs, tales, stories, and bring the things and props you might need. If you have a guitar or another instrument, please bring it – you do not need to be perfect on the instrument, we need every voice and tone! There is also a piano on the site. Bring songs that you sing in everyday life. We would like to learn new songs. And of course, there are also the songs in the Summer School Songbook!

Every evening there will be a gathering where we talk about the day and plan for the following days with support from the NCL Team.

We would like to talk with you about your role as a support person and how you can fill this role in the best way – that is, how you can contribute and strengthen the fellowship between all participants. As part of this, it is important that supporters who have experiences from previous summer schools share their knowledge and experiences with new supporters at the summer school.

Support staff at work

Like all other pedagogical work within the field of JNCL it is essential to consider the developmental decline, which sets its mark on the summer school program and the expectations of the support staff. The days have an overall structure and predictability, but the daily content is organized in a way that both welcomes and positively challenges the child and the young person with JNCL.

The Team and support staff work to create links between the new children and the group of slightly older children and adolescents. The supporters play an important role in this work, where traditions and practices are shared. Along the way, based on interests, there are reminiscences from former summer schools with songs, stories and repetitions.

The importance of the summer school for the participants with JNCL, for the support people, and for the NCL Team

The summer school offers meetings with old friends where memories are kept alive and old skills are refreshed. In the surveys and interviews with parents and staff (see Appendix A), tours and special activities were often mentioned as important events that their child with JNCL remembered particularly well. Two parents considered these events as one of the three most important activities or interests in the life of their child.

Working as a supporter for a child or young person with JNCL can be difficult but also rewarding with opportunities for learning. Children and young people with JNCL with a need for closeness and security make it necessary to work with professional values and reflections in collaboration with parents, other staff groups and the NCL Team (see Chapter 8). Supporters may experience many dilemmas in their daily work with children and young people with JNCL. The working conditions are not always consistent. It is important to establish support groups to encourage cooperation between members with different professional backgrounds. An important issue is how one "translates" and interprets the world to a child or a young person with dementia. Another issue is how to ensure that the selected goals and interventions reflect the true interests of the child and contribute to learning and good experiences.

The summer school emphasizes networking and learning in a community; that is, the summer school environment facilitates a transfer of important knowledge and best practice through the efforts of the supporters, who together create a good week for the children and young people with JNCL. The individual

supporters can act as role models for others in the network, ensuring that the children and young people with JNCL are in the center.

For the members of the NCL Team, the summer school week with children, young people and support staff provides a good opportunity to gain insight into the situation of each child and young person with JNCL. As a whole, the summer schools have an important function for the NCL Team, in relation to both the children and young people with JNCL and the continuous collaboration with the support staff who participates and receives guidance and supervision from the Team throughout the year.

Summer school in perspective

There are children and adolescents who have attended the summer school every year since they were diagnosed with JNCL. There are also children and young people whose parents have avoided the summer school and instead tried to bring the child into other social contexts, such as the summer school for children who are blind or have low vision.

There are many concerns for parents when deciding if their child should be a part of a community with other children with JNCL who have more pronounced declines and needs in later phases of the disease. There are no "correct" answers to these questions, families will differ in how and when they want their child to be introduced to the community of children and adolescents with JNCL.

At a recent parent meeting, the topics of the summer school and the children who did not attend the summer school were discussed. Several parents were curious, and also concerned. The NCL Team was asked to show the film that was recorded every year at the summer school to the parents who did not want to send their young children to the summer school. The parent of a 12-year-old said: *If you show the movie, everyone will understand how important it is for the child to attend the summer school as early as possible.*

Several of the video recordings from Summer Schools show young people without expressive speech "singing" when familiar songs were played. The young people were sitting and holding hands with friends from previous summer schools many years back. The young people had big smiles, despite serious losses of everyday skills and lack of control over body and facial expressions. Professionals have to learn to interpret what the children and the young persons are showing despite illness, declines and dementia. Participation and fellowship are important parts in the quality of life.

References

- Abdi, A. A., & Guo, S. (2008). Education and social development. In A. A. Abdi & S. Guo (Eds.), *Education and social development – Global issues and analyses* (pp. 1–12). Rotterdam, Netherlands: Sense Publishers.
- Bagwell, C. L., & Schmidt, M. E. (2011). The friendship quality of overtly and relationally victimized children. *Merrill-Palmer Quarterly*, 57, 158–185.
- Barca, M. L., Engedal, K., Laks, J., & Selbæk, G. (2011). Quality of life among elderly patients with dementia in institutions. *Dementia and Geriatric Cognitive Disorders*, 31, 435–442.
- Bukowski, W. M., & Adams, R. (2005). Peer relationships and psychopathology: Markers, moderators, mediators, mechanisms, and meanings. *Journal of Clinical Child and Adolescent Psychology*, 34, 3–10.
- Bukowski, W. M., Motzoi, C., & Meyer, F. (2009). Friendship as process, function, and outcome. In K. H. Rubin, W. M. Bukowski & B. Laursen (Eds), *Handbook of peer interactions, relationships, and groups* (pp. 217–231). New York, NY: Guilford Press.
- Chen, X., French, D., & Schneider, B. H. (2006). Culture and peer relationships. In X. Chen, D. C. French & B. H. Schneider (Eds), *Peer relationships in a cultural context* (pp. 3–20). Cambridge: Cambridge University Press.
- Dunn, J. (2004). *Children's friendships: The beginnings of intimacy*. Oxford: Blackwell.
- Gilmore, L., & Cuskelly, M. (2014). Vulnerability to loneliness in people with intellectual disability: An explanatory model. *Journal of Policy and Practice in Intellectual Disabilities*, 11 (3), 192–199.
- Gold, D., Shaw, A., & Wolffe, K. (2010). The social lives of Canadian youths with visual impairments. *Journal of Visual Impairment and Blindness*, 104, 431–443.
- Guralnick, M. J., Neville, B., Hammond, M. A., & Connor, R. T. (2007). The friendships of young children with developmental delays: A longitudinal analysis. *Journal of Applied Developmental Psychology*, 28, 64–79.
- Hartup, W. W. (1999). Peer experience and its developmental significance. In M. Bennett (Ed.), *Developmental psychology: Achievements and prospects* (pp. 106–125). London, UK: Psychology Press.
- Hartup, W. W., & Stevens, N. (1997). Friendship and adaptation in the life course. *Psychological Bulletin*, 121, 355–370.
- Laursen, B., & Pursell, G. (2009). Conflict in peer relationships. In K. H. Rubin, W. M. Bulowski & B. Laursen (Eds), *The handbook of peer interactions, relationship and group* (pp. 267–286). New York, NY: Guilford Press.
- Maccoby, E. E. (1990). Gender and relations: A developmental account. *American Psychologist*, 45, 513–520.
- Mathur, R., & Berndt, T. J. (2006). Relations of friends' activities to friendship quality. *Journal of Early Adolescence*, 26, 365–388.
- Meltzer, A., & Kramer, J. (2016). Siblinghood through disability studies perspectives: Diversifying discourse and knowledge about siblings with and without disabilities. *Disability and Society*, 31, 17–32.
- Newsom, D., Mallow, J., & Watson, J. (2015) Loneliness in school age children: An integrative review of quantitative studies. *Psychology and Education – An Interdisciplinary Journal*, 50, 32–41.

- Opie, J. L., & Southcott, J. (2018). Inclusion for a student with vision impairment: «They accept me, like, as in I am there, but they just won't talk to me». *The Qualitative Report*, 23, 1889–1904.
- Rossetti, Z., & Keenan, J. (2018). The nature of friendship between students with and without severe disabilities. *Remedial and Special Education*, 39, 195–210.
- Rubin, K. H., Bukowski, W., & Bowker, J. C. (2015). Children in peer groups. In R. M. Lerner, M. H. Bornstein & T. Leventhal (Eds.), *Handbook of child psychology and developmental science, Volume 4: Ecological settings and processes* (pp. 175–222). New York, NY: Wiley.
- Salminen, A. L., & Karhula, M. E. (2014). Young persons with visual impairment: Challenges of participation. *Scandinavian Journal of Occupational Therapy*, 21, 267–276.
- Schneider, B. H. (2016). *Childhood friendships and peer relations: Friends and enemies, Second edition*. Abingdon, UK: Routledge.
- Sterrett, K., Shire, S., & Kasari, C. (2017). Peer relationships among children with ASD: Interventions targeting social acceptance, friendships, and peer networks. *International Review of Research in Developmental Disabilities*, 52, 37–74.
- Wogn-Henriksen, K. (2012). «Du må ... skape deg et liv»: En kvalitativ studie om å oppleve og leve med demens basert på intervjuer med en gruppe personer med tidlig debuterende Alzheimers sykdom («You must ... make yourself a life»: A qualitative study on experiencing and living with dementia based on interviews with a group of persons who have early onset Alzheimer disease). Dissertation, Department of Psychology, Norwegian University of Science and Technology, Trondheim, Norway.

23

Transition to Adult Living

Svein Rokne and Bengt Elmerskog

Passivity, if life takes longer breaks and loses its fluency, if development and maintenance of skills of independence and interdependence are not given enough attention, and if participation in a socially inclusive environment is lacking, should be considered as threats for individuals with JNCL. These areas, often associated with the life flow, must be given special attention in situations of life transitions, and in particular to the transition to adult living.

Participation in various kinds of activities is mandatory for inclusion in the community and quality of life for all. However, there is a prominent risk that such goals are unobtainable for individuals with JNCL if their special needs are not considered and sufficiently met by appropriate measures. Involvement in *interactive participation* implies that individuals with JNCL can achieve personal goals when needs for personal assistance and physical adaptations are met by appropriate interventions. Interactive participation is closely related to personal *autonomy* (see Chapters 11 and 16). It supports personal engagement and applies to the personal rights of influencing one's own life, making one's own choices, having a meaningful work, having feelings of self-worth, being around people one likes, and having opportunities to enjoy experiences associated with learning and social and cultural living (Oakley & Marsden, 1984). Interactive participation is for instance used in theater plays where the audience is engaged in being an active part of scenarios (Skye, Wagenschutz, Steiger, & Kumagai, 2014). Interactive participation is an important strategy for empowering the individual, in particular in situations where the person's voice and interests should be articulated.

Interactive learning engages learners by challenging them to actively explore a material or situation. Through real-world applications, the individual makes meaningful connections between learning and its relevance for real-life situations. It reinvigorates the learning scene; one-way speeches or lectures are changed into

dialogues, discussions and collaboration. The learner and staff become partners in the journey of knowledge acquisition. It is found to be an effective way of learning, not only for people with learning disabilities, but also within higher education (Krusche, Seitz, Börstler, & Bruegge, 2017; Stanford University School of Medicine, 2018). Interactive learning can take many different forms. Individuals may strengthen their critical thinking and problem-solving skills using a holistic approach to learning in real-life situations. Interactive learning can thus have a place across the curriculum in a holistic manner. The close and timely interaction will promote immediate and specific feedback on comprehension (Stanford University School of Medicine, 2018). Interactive participation and learning are for individuals with JNCL closely related to interdependence where tasks are performed in collaboration with others (see Chapter 16).

Focus on participation in activities

A focus on participation in activities and participation-based learning has many advantages (see Chapter 11 and 16). Participation is an important basis for learning and development; children and adolescents learn from participating in activities – most knowledge and skills are in fact based on participation in real-life activities. For instance, children learn a lot through participation and exploration in preschool years. The learning is implicit and incidental, not based on predefined goals or curricula. This type of learning is valid for all humans, regardless of age and functioning, it is more about making participation possible.

The JNCL and education project (see Appendix A) has defined five areas of participation in activities to be considered when planning and implementing an optimal adult living: domestic activities, work activities, physical activities, leisure time activities, and vacation and excursion activities.

Participation in domestic activities is related to engagement in household activities, such as washing clothes, doing the dishes, cleaning, preparing food, and in domestic activities outside the home, for example shopping groceries and using the mobile telephone.

Participation in work activities is related to engagement in activities that give the young adult a social role in society or within a smaller group of peers. A working activity is associated with a clear and defined responsibility. This could include all activities undertaken in a vocational center or a sheltered workshop, such as metalwork, woodwork or pottery making where the products are available for sale either within or outside the vocational center or sheltered workshop. Participation in work activities usually take up a large part of the time spent outside the residence.

Participation in physical activities is related to engagement in activities that promote physical health and wellbeing. These are of special importance for individuals with JNCL and should take place on a daily basis. Physical activities may overlap with leisure activities, as when they reflect personal interests, but they should be defined as a separate category to ensure that they take place on a regular basis in order to sustain physical health and wellbeing. Physical activities can take place as organized activities (e.g., attending a sports club every Thursday) or as non-organized activities (a walk when the weather is fine).

Participation in leisure time activities is related to engagement in activities taking place outside work, school and domestic life, often during afternoons and weekends. Leisure time activities are strongly associated with personal interests and may be related to the person's social needs (e.g., visiting friends or playing cards) and cultural needs (e.g., playing the piano, attending a football match or visiting the church). This category also includes certain self-entertainment needs such as playing a computer game, reading or listening to audio books, playing an

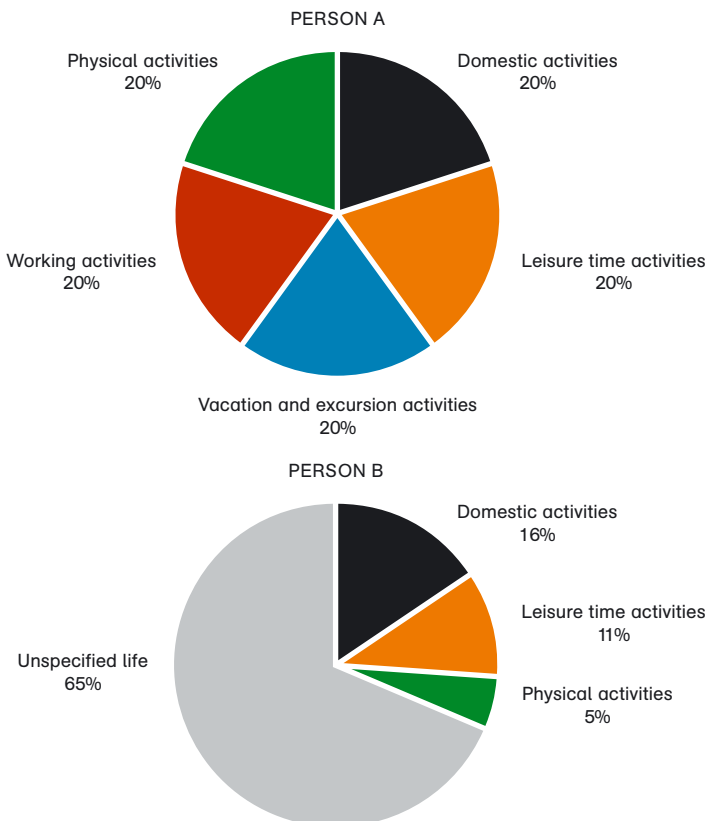


Figure 23.1 An overview of the life situation of two adults with JNCL

instrument or listening to music. Leisure time activities can be organized (e.g., participation in a book club) or unorganized (e.g., spontaneous activities such as listening to a CD to achieve relaxation).

Participation in vacation and excursion activities is related to engagement in activities, happenings or events beyond the daily routine. Vacation and excursion activities are strongly associated with personal interests that break the usual routine. A vacation activity can be a holiday trip to the mountains, a tour to a festival or visiting a museum in London. Vacation and excursion activities are considered as highlights taking place on an infrequent basis.

It is assumed that a reasonable balance between these activity areas will promote life quality and participation in the community. Participation in the different spheres of activities may function as an index of quality of life.

Figure 23.1 shows different life situations for two young adults with JNCL. Person A has a well-balanced life situation, while Person B has a limited life situation. The size of the sector named "unspecified life" seems worrying for Person B. A focus on participation in the different spheres of activities constitutes a fundamental basis to achieve personal goals associated with interactive participation and learning, in particular at the time of transition to adult living.

Participation when interdependence is required

Independence will be reduced as individuals with JNCL get older (see Chapters 3–7, 16). There is always a risk that participation will be threatened when abilities and functions decline. Gray and Hollingsworth (1999) developed a model for describing the relations between participation in activities and performance in activities. Table 23.1 illustrates four scenarios for the relations between ability, performance and participation in particular activities or tasks for one individual. *Scenario 1* is a rather normal situation where participation and independence are correlated. *Scenario 2* is a situation that may have been caused by poor planning in connection with the transition to adult living for a person with JNCL. For instance, the person was a devoted and skilled person in using the computer the last year in school. However, the person became excluded from this activity in the new residential home due to a lack of equipment or staff competence related to using the specialized equipment. *Scenario 3* may be positive for an individual with JNCL. The person is not able to do much but is provided with enough support to participate through interdependence. *Scenario 4* might be a situation where low performance automatically led to non-participation caused by contextual factors, such as the lack of support and help.

Participation in activities may over time become limited or take so much effort that the person with JNCL will not find it worthwhile to initiate participation in

Table 23.1 Four scenarios related to activity and participation (after Gray and Hollingsworth, 1999)

	Ability to perform an activity	Inability to perform an activity
Participates	1 The person has the capacity to do things and does participate	3 The person cannot do things but participates
Does not participate	2 The person has the capacity to do things but does not participate	4 The person has no capacity to do things and does not participate

some activities. The risk is however that non-performance automatically leads to non-participation caused by lack of support or other contextual factors. Scenario 1 and 3 in Table 23.1 can consequently be considered as positive goals for young adults with JNCL (see also Chapter 11).

Persons with JNCL are affected by declines that may threaten participation. The anticipation of these declines requires that the staff is introduced to and taught about the Team Model (see figure 23.2) which may help them to achieve continued participation when abilities decline in persons with JNCL. It is the way that the activity is organized that governs the situation. It is necessary to acknowledge that persons with JNCL at all times may participate according to their abilities, achieve personal goals and experience pride in spite of progressing declines in performance. The person with JNCL and a support person should work

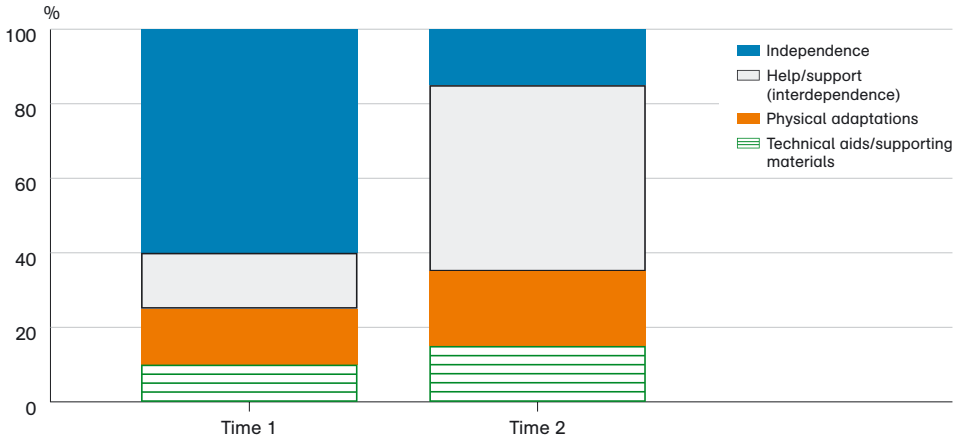


Figure 23.2 The Team Model or The Interdependence Model describing 100 percent participation at two times

together as a team, although the relative contribution from the person with JNCL and the supporter will change over time (see Figure 23.3).

The base line at Time 1 in Figure 23.2 shows that the child with JNCL performs 60 percent of the task while the supporter contributes 15 percent. Ten percent of the task is supported by technical aids and 15 percent by physical adaptations of the environment. At Time 2 the person is contributing only 15 percent of the task. However, the level of participation, perceived by the individual with JNCL, is 100 percent at both times. The participation level at Time 2 is based on more support, more physical adaptations and additional supporting materials compared with Time 1. (The two scenarios are illustrated in Figure 23.3.) It is thus important that the people who are assigning resources understand the necessity of establishing the Team Model when working with individuals with JNCL. The access to individual support or other adaptations will sooner or later constitute the difference between participation and non-participation for persons in this group. The Team Model can be applied to any situation, for instance at home or at work, as the parent story below illustrates.

Roger is genetically predisposed in more than one way; from his father he has inherited an interest in fly fishing and has found profound pleasure in catching representatives of the aquatic domain on flies tied by himself. He learned the basic techniques of fly tying quite early, while he still



Figure 23.3 Participation through a) independence and b) interdependence

had some residual sight. When seeing became gradually more difficult, he appointed his father to the post of assistant fly tier, and the flies were produced through a joint effort.

Fly tying is a process requiring fine motor skills and the ability to consider and make decisions related to type and size of hook, tying pattern, choice of color scheme, hackle type, ribbing, wing type and more. These elements were discussed before starting the tying process at the tying vice. Salmon flies have always been central to Roger's fly tying, particularly since he had been able to try out the results of his efforts in the salmon rivers during the summer, often successfully!

When Roger was 20 years old, the idea to take the fly tying one step further materialized. Why not use the salmon flies as a basis for production of earrings? Roger's father was appointed as Roger's business manager and 5 percent of sales were considered a fair remuneration for this work (determined after tough negotiations). This resulted in a planning phase, where "Team Roger" had to consider and decide upon several factors related to production, buying, marketing, sales work, logistics, accounting, invoicing and managing receivables. A company was established: Roger's Products.

The process from hatching the idea to making the first sale (and collecting the money) was incredibly exciting and educational for Roger and his father. Standard patterns needed to be developed, fly-tying components had to be purchased (involving several visits to various pro shops for selecting the raw material), and solutions for clasping the product to the ear (not a trivial point) needed to be developed. In addition, a marketing plan was conceived, and Team Roger had a discussion whether to base the business on a one-tier or two-tier sales model. Also, a price strategy needed to be established, and then the actual marketing and sales work started.

An invoice is produced for each delivery, and Roger's father ensures that these are being paid and that the money thus accruing is deposited into Roger's account. Rest assured that Roger follows this part closely!

During the two to three years of production since the inception of the business, Roger's abilities have been somewhat reduced, meaning that more of the physical labor is carried out by the second participant of the Team. However, Roger owns the business, the production process and the sales efforts, and participates all the way. There has been no diminishing of pride each time Roger's Products produces and sells a new set of earrings to a satisfied customer.

From adolescence to adulthood

Reaching adulthood is an important milestone in anybody's life, also in the life of a person afflicted with JNCL. For individuals with JNCL, the transition period between adolescence and adulthood is of particular importance. The quality of their adult life will depend on several factors discussed in other chapters of this book and on how the transition period is planned and carried out.

What does it mean to be an adult? And more specifically, what does it mean to be an adult with JNCL? Most encyclopedias define an adult as a fully grown and developed person, mature and sensible – not childish. In the legal sense, an adult is a person who has attained the age of majority (or maturity) and is therefore regarded as independent, self-sufficient and responsible.

The first part of this definition is problematic as far as persons with JNCL are concerned. The progressive nature of dementia means that the definition is only partly applicable. In the legal sense the definition applies as long as the person has not been declared legally incompetent. In the present context, none of the definitions are of any concern. The goal is to examine the process of transition and recommend strategies which may contribute to the person's quality of life as an adult, which here means a person with JNCL who has finished secondary school, entered a place of work or is engaged in other suitable occupation, and who has moved out of the family home.

All the interventions, education, and training during the school years were intended to contribute significantly to learning and quality of life during these years, as well as to prepare the person with JNCL and adapt the environment to future adult living. A transition to adult life often means that the person with JNCL moves to a new environment, an apartment or as a resident in a sheltered house and community of peers. The person should in addition participate in some meaningful daytime work-related occupation, which is located outside the residence. These are important aspects of adult living.

Several considerations must be made before, during and after the transition period. This chapter is based on findings from the present study (Appendix A), including some recommendations to achieve *best practice* in times of transitions to adult living. Core factors behind successful transitions are a functional responsibility group (see Chapter 11), service providers' competence and attitudes, and available resources. There are vital differences between countries in terms of culture, how society is organized or how services for persons with disabilities are provided. However, the transition should always be person-centered, independent on the overall system or cultural attitudes. Individuals with JNCL share the disease but in all other aspects they differ as much as other people.

Findings from the JNCL project

The present study did not find any research or documentation related to adult living among people with JNCL that go beyond the obvious medical needs. Quality of life encompasses areas beyond medical health, and includes needs in relation to social life, life content, learning and maintaining skills and development. The findings of the present study (Appendix A) give a better understanding of the transition period from school to adult living and the factors that may contribute positively or negatively to the adult living for persons with JNCL beyond their medical needs.

There was a consensus among the parents that local competence had been lacking, not only in relation to JNCL or early onset of dementia, but also to meeting the challenges related to teaching a person who is blind. Particularly problematic was the comprehensive change of staff and external counseling when the person reached legal age, which in most countries is 18 years.

A majority of the parents of persons who had reached the age of 22 were reasonably satisfied with their collaboration with the local community, but fewer with local education counselors and, perhaps surprisingly, educational counselors in resource centers. These responses can indicate a lack of sufficient competence or of access to competent services.

Interest in physical activities was low from 17 years onwards, although parents seemed to agree that this is an important area for their child (see Chapters 7 and 15). The results show a significant reduction in physical activities as the person gets older. One of the reasons may be that while physical activity is emphasized as an important part of the school curriculum, it becomes more difficult to implement physical activities in the family home or in residences for adults with disabilities. The lack of physical activity can also be due to a lack of knowledge about how to achieve a healthy lifestyle when the person is blind and has reduced motor functioning.

When evaluating the various school subjects and their impact on life as an adult, the parents considered music, music therapy, physical education and physiotherapy as most important. Literacy, handicraft, cooking, kitchen work, social training, occupational therapy and social gathering were considered as having a positive impact but were given somewhat lower scores. Mathematics, social studies, nature studies and foreign languages were given low impact scores by the parents. A large majority (90%) found that augmentative and alternative communication training had little or no impact on their child's life after the conclusion of school but only a small number of persons had received this form of training (see Chapter 13).

Encouragement of special interests or hobbies and the integration of these into adult living had been lacking according to parents. Early establishment of skills and a continuous maintenance of these skills seem to give better chances for these skills to be maintained into adulthood. Examples from the present study are playing an instrument, horseback riding, swimming, music and dancing. Computer skills and literacy are also mentioned as long-lasting skills – reading and writing probably had a positive influence on the preservation of cognitive skills. Many parental comments suggested that having a suitable daytime occupation after school helped to maintain skills for a longer period.

Asking staff the same questions yielded the same results as far as music, physical education and physiotherapy are concerned, while their impact scores for reading and writing were very low – they found that these subjects had little impact on life after school for the participants with JNCL. Comments point to a feeling of defeat and, perhaps, little understanding of the necessity of continuous stimulation to maintain skills. A descending curve of abilities should not be used as a reason to terminate initiatives to measures that may maintain skills in young adults with JNCL.

A large majority of the parents (86%) agreed with the statement that the school had done reasonably well in preparing the individual with JNCL for adult living. Sixty-five percent of the parents agreed that the school did well in transferring important information to the person's new environments (home and day center), and a majority (68%) also agreed that the home or day center had been reasonably well prepared. A majority (68%) agreed that the collaboration between parents, the school and staff in day centers or homes had been successful, whereas 59 percent were satisfied with the role external counselors had played in this process. However, some parents expressed dissatisfactions with the transition period as shown in the following quotation:

It is important to emphasize the transition to adult living for the school. For us nearly two years have passed since end of school and not much is in place.

Seventeen of the fifty responding parents reported that they were offered a sheltered housing or similar after the conclusion of school and 39 of the persons with JNCL had been offered to join a day center or similar. It seems that housing remains a difficult issue.

Although their child's transition to adulthood and living as an adult had been, and remained, quite challenging, several parents reported satisfaction with the process as well as with the period following the transition. Some parents expressed satisfactions with highly motivated service providers, who did their work with the right kind of attitude and often beyond their formal responsibilities.

The preparatory phase

A long planning period may be required to find a home and an occupation for a person with JNCL, depending on several factors. The transition period is the time from initiating the transition project (at least two years before school ends) to the actual milestone: Starting work-related activities and possibly moving to a new home. The strategies, activities and tasks in the transition period should be described in detail in the long-term Habilitation Plan developed by a Responsibility Group (see Chapter 11). The plan should list tasks, responsibilities and success criteria with associated dates. The process should be monitored through frequent evaluations, at least once every third month during the transition period. The local government may assist in this and the municipality usually has a planning period

Table 23.2 Topics that may be included in the Habilitation Plan

Topics, needs and functionalities for the future	
Network	Family, friends, care givers
Daytime activity	Work, activity center, other
Leisure	Holidays, daily activities, sports, hobbies, areas of special interest
Preparatory training	Training at current school to prepare the individual for adult life in the forthcoming home, day center or work? How? By whom?
Living preferences	Physical layout, own room, adaptations needed, the neighborhood
Aids	Computer equipment, wheelchair, other aids.
Communication and language	How does the person communicate? Tools to aid communication (manual signs, communication aid, computer, other)?
Behavior, social life	Resources, strengths, vulnerabilities, wellbeing, play, mood, frustration, aggression, withdrawal, sleep pattern, depression, anxiety
Nutrition	Dietary requirements, meals, appetite, food and fluid intake
Motor skills, disabilities	Mobility, gross and fine motor functions, technical aids
Support	Support needed (individual or shared)
Health	Diagnosis, medication, epilepsy, teeth condition, bowel function, sight, hearing, allergic reactions, contingency plans, etc.
Economy	Legal rights, social security, other
Aid measures	Educational and other services provisions, scope of services rendered, other useful information
Staff competence	Competence building. How, when, and by whom?

of several years. The findings of the present study emphasize the importance of starting this work early.

In addition to, and as an integral part of the Habilitation Plan, it will be of great help to have a written narrative of the personal history of the individual with JNCL. This written narrative – «The book of (the person's name)» – is regularly updated. This is a valuable tool for guiding service providers and staff to a shared frame of reference. The Responsibility Group should consist of individuals who are able and willing to carry out the plan. One member of the group should act as the group's coordinator and have the additional responsibility of organizing meetings, setting the agenda of the meetings, and monitoring the work progress in general. This also includes measuring results compared to the goals of the plan and evaluating the overall strategy. The Habilitation Plan includes functions and needs in all areas of life (Table 23.2).

The future home

A good beginning would be to design the "perfect" future home for the person with JNCL; considerations should include the physical location and the quality and quantity of service provisions needed in the new home. Designing a new home two or more years into the future is challenging. The person with JNCL may at the onset of the transition period be functioning quite well and with some degree of independence when performing daily tasks. However, there is a need to consider needs that are likely to appear years ahead and design the optimal home with those in mind. The local government may assist the parents (and other family members) in designing and finding the new home.

There are several possible strategies for identifying the new home. One strategy is to apply to an existing house in which a group of young adults with disabilities have their own facilities surrounding common areas such as community kitchen and drawing room(s). The new home should be adapted to the needs of persons with visual and physical impairments. For instance, it should not have physical barriers such as steps or dangerous indoor and outdoor environments. Young adults with JNCL may also be sensitive to neighbors with challenging behaviors and environments with high noise levels. The rationale for moving into a community is usually to create opportunities for social interaction with peers. The current study shows however that persons with JNCL over time tend to spend more time with caregivers and staff and less in social interaction with peers. Loss of friends is an unfortunate fact of life for most individuals with JNCL (see Chapter 22), and planning should take this into consideration.

Another strategy is to move into an apartment which can also accommodate one or more staff as the need arises. However, finding a suitable apartment can

be difficult and costly. A third strategy may be to rebuild the family's house to accommodate a separate home for the individual with JNCL. Proximity to parents, siblings and other family members may have benefits as well as disadvantages, for the person as well as for the family. Finally, in some countries there are special institutions that cater to the needs of individuals with JNCL, such as Heather House in the U. K. and the Bartimeus Foundation in the Netherlands.

It is not always possible to find a home that matches the needs of the individual. The result may be that the person with JNCL continues living with the parents for a shorter or longer period after finishing school. However, if a suitable home is found, it must be furnished with care and on the basis of the person's anticipated physical and mental condition at the time of moving.

Moving out of the family home should be a gradual process, where the person moves in steps, beginning with shorter day visits accompanied by parents, which are gradually expanded into overnight visits. Comments from parents and staff in the present study emphasize the importance of preparing the young adult with JNCL for the transition by talking about it, and when possible – organizing many visits to the future home. The young adult will become familiar with the layout of the apartment or rooms and may become acquainted with the other residents (if any) and the staff before moving there. Such an approach reflects that moving is a process and may make the move more interesting and feasible for the person with JNCL.

In some European countries, young adults with JNCL receive a pension for persons with disabilities from age 18 years and onwards. Together with other forms of economic support from the government (basic and extended economic compensatory benefit, living cost subsidy), the young adults will usually be able to fend for themselves from an economic point of view. In some countries, purchase of living facilities, if necessary and desired, may be subsidized through low-interest loans and grants from the government.

The future occupation

Finding a suitable occupation at a sheltered workshop or day center after leaving school is of extreme importance. The last years at school should include vocational and work-related activities to facilitate the transition to adult life. One or two days should be used to train the person in occupational activities similar to those that may be provided after the young person leaves school. Consequently, the identification of an appropriate occupation should also be discussed early in the transition process. In the present study (Appendix A), some parents reported that the student with JNCL had been offered opportunities to work one or two days per week at their future adapted workplace as a part of their Individual Educational

Plan (IEP, see Chapter 11). According to the reports, such educational planning measures made the transition much easier.

Although blindness and cognitive and physical declines may limit the availability of job-related activities, no efforts should be spared in securing a suitable occupation. Comments in the present survey and interviews indicate that getting up in the morning and going to a different location for work-related activities seemed to have significant therapeutic effects, including heightened self-esteem, particularly if a small remuneration was attached to the work. In addition, the understanding of The Team or Interdependence Model (see Figure 23.2) will simplify the process of finding a suitable occupation. Team members should consider what the person may be able to do or produce when he or she is collaborating with a support person. The concept of supported employment relies on the concept of interdependence.

The challenge, of course, is finding a meaningful occupation for the person. Ideally, the person has skills and preferences that can function as a basis for working activities. It will be useful to arrange meetings with representatives from the local government and work places early in the process. Most municipalities have sheltered work places catering to the needs of young adults with disabilities, and many activities in such places can be tailored to fit the needs of a person with JNCL. Meetings with representatives from these work places may create an understanding of the needs, that is, the anticipated progressive decline in abilities and how a work place can cater for such needs with an approach based on interdependence (see Chapter 16).

Staff and service provision

When planning the transition to adult life, it is necessary to determine what resources are necessary to adequately cover the person's needs once he or she moves to a separate home and starts the working life. The course of the disease, in particular the fact that epilepsy may become a severe problem, calls for proximity to staff around the clock. As soon as adequate housing has been found, an application must be presented to the local government for necessary staff at the living facilities. How this is organized in practical terms may vary considerably, not only from country to country, but also within countries and communities.

Service provision for young adults who need continuous staff attendance is covered by several laws. However, because of the broad leeway often given to how the laws are interpreted, the process will not always lead to good solutions. Often resolving the problems resulting from deficits in the process entails hard work for the parents with or without assistance from resource centers or parent organizations. For example, although the legal process typically recognizes the

need for continuous medical care, it often fails to recognize the resources needed to maintain an optimal quality of life. In view of the progressive nature of JNCL, the human resources required for maintaining the goals of interdependence and autonomy (see Chapter 11 and 16) will gradually increase, and planning must take this into account.

Local governments often have preferences as to how services should be organized. Most local governments in Norway recognize User-Managed Personal Assistance as a way of organizing services. Under this scheme, the user, in this case an individual with JNCL, is *de facto* the employer and must hire personnel to provide services within an economic frame decided by the local government. In theory, this gives some degree of quality control, but in general, user-managed personal assistance represents extensive administrative work for the parents. Most often, however, services are provided by the local government. The advantage is a reduction in the administrative burden of the parents, but the disadvantage is the potential risk for compromises in the quality of the services provided to the person with JNCL.

If the parents wish to continue being the guardians for the individual with JNCL, it is a good idea to issue an authorization from the person with JNCL to one or both parents, authorizing them to act on the person's behalf in all matters, economic and otherwise. The authorization signature (made with or without help) should be confirmed by two witnesses. If the parents do not wish to have this role after the person reaches legal age (18 years in most countries), they should apply for a guardian to safeguard the person's rights.

Educating staff

When an appropriate home for the person has been found and the time to move is getting closer, the future staff should be educated and informed to avoid unnecessary problems in the moving process. An appropriate working culture should be encouraged from the very beginning. Parents and staff in the present study (Appendix A) had found the use of person-centered work-shops to be a good method to educate staff who will be in direct interaction with the person with JNCL. The workshops should focus on possibilities instead of limitations. Getting to know the person rather than the disease should be the main focus of the workshops. Education of staff is an ongoing process, for instance built on the stepwise method presented below. The first work-shop may be based on *The book of (name of the person)* (see Chapter 11), preferably presented by the parents, aiming to make the staff comfortable with their roles in supporting and communicating with the person. An early open dialogue between parents and staff will contribute to building a foundation for further collaboration and competence building.

Life as an adult

The quality of life as an adult depends on many factors. Early established skills and interests can often be maintained in a longer perspective and thus contribute to a richer life for the young adult. The implementation and quality of the preparatory phase is another important factor, including the establishment and maintenance of good relations between the family, staff and the person with JNCL.

Teamwork

An optimal adult living for a person with JNCL will be based on the coadjutant equal partners: parents, staff, representatives from the local government and counselors with knowledge about JNCL. The team must find a good and effective way of working together, based on mutual trust and efficient communication. The meetings can be held as workshops where all participants are given the possibility to learn, share knowledge and promote ideas. Minutes should always be kept from these meetings and agreed actions should be clearly spelled out.

The team may have chosen a place to live with a (small) number of peers occupying their own facilities around a common kitchen and a drawing room, and with staff in accordance with the person's needs and as determined by the local government. The team should have agreed on the important principles or framework for the work. The principles agreed by the team should emphasize maintenance through daily or weekly repetitions of acquired skills and knowledge. Staff turnover should be minimized, and an effort to maintain a core staff group responsible for the person should be made so that the person's daily life routines can remain as stable and predictable as possible. High staff turnover is a reality in the field, and a plan for continuous training should be in place for new staff. Likewise, a system should be in place for providing substitute staff members with the required basic knowledge when needed. Daily physical and mental exercises and challenges should be managed in accordance with the goals defined by the team. The daily routines should be followed by all staff members, even in times of worry and pronounced health problems.

A method for supporting adult living

A method for establishing the best possible adult living for persons with JNCL has emerged from the quantitative and qualitative results of the present study. The method does not focus on the medical needs but on the needs associated with content of life. The establishment and maintenance of rich and varied life content are essential factors that will contribute to the quality of life and general

health of persons with JNCL. There is an emphasis on daily cognitive and physical activity and learning and maintenance of skills from early morning to bedtime. The method includes several tools.

Tool 1 – The book about (name of the person)

A continuously updated written narrative of the person's history is an integral part of the present method. The content of *The book about (name of the person)* includes historical and present facts about the person with JNCL and an overview of important topics, areas of interest and preferences. Having access to this book makes it easier for others, specifically supporters, to understand the state of the person at any given time and support an active life style. *The Book of (name of the person)* is a central document for service providers and mandatory reading for all staff working with the individual with JNCL. Provision of time for supporters to read the book might be one of the overlooked planning resources described earlier. The book is a valuable tool for new personnel and regular reading may serve to refresh the memory of those who already know the person. The book is introduced as part of the preparatory phase (see above) and remains a part of the Habilitation Plan. The aim is that the book should contribute to promoting an understanding of the person with JNCL as a unique individual with a personality and history despite current and future declines (see Table 23.3). It should help establish a shared frame of reference between the person and the staff, and between staff members, thereby facilitating communication. The book should not only make it easier for staff to maintain focus and engagement, but also provide them with a way to enrich their own work days by enabling them to experience success doing their job. It is easy to overlook the daily challenges staff may face, not just from doing their duties, but also from external pressure and demands from family and institutions.

Tool 2 – The Annual Wheel

Every year there are important events and activities taking place at regular or irregular times, such as vacations or birthdays. *The Annual Wheel* is used to identify such events and activities. Participation in vacation and excursion activities is related to engagement in activities, happenings or events beyond the daily routines. Vacation and excursion activities are strongly associated with personal interests that break the usual humdrum routine. A vacation can be a holiday tour to the mountains, an excursion can be a tour to a festival or museum in London. These vacations and excursions are typically highlights in the ordinary time flow, taking place infrequently. Figure 23.4 shows an Annual Wheel for a young adult with JNCL. *The Book of (name of the person)* and the parents should be consulted when constructing an Annual Wheel.

Table 23.3 Possible contents of The book about (name of the person)

Topics, needs and functionalities for the future	
1 – Why this book?	A brief introduction to (name) and his or her special situation and what his or her condition means in terms of necessary requirements and desired skills in daily interaction with him or her. A list of the most important elements contributing to the person's quality of life can be included here. Why is it necessary to have this book, what does it include, who is it for and how should it be used?
2 – Who is (name)?	A brief life history
3 – (Name's) routines	Daily and weekly routines
4 – To lose one's sight	What are the consequences of blindness? How does (name) cope with this situation and how can service providers help. Writing/reading with Braille or Moon. Computer tools, daily living aids, mobility issues.
5 – Medical conditions	Emphasis on the here and now and on symptoms rather than the diagnosis. Medication and how it is administered. Reference to contingency plans.
6 – Communication	How does (name) communicate? Any differences between expressive communication and comprehension?
7 – What (name) is concerned about	Skills and learning, hobbies
8 – Interests	All areas of interest and how best to stimulate the person through active use of these. Most individuals with JNCL are interested in music, and early learning for example of piano playing, drums or other instruments can be maintained for a very long time.
9 – Important persons in (name's) life	Family members, important friends, others
10 – Important places for (name)	All important places and why they are important. Home, work place, places of important activities, travel destinations. Can provide common ground for interaction and communication.
11 – Important activities	Activities contributing to the person's quality of life, summer and winter activities, programmed activities as well as activities of a more ad hoc nature.
12 – Aids	For example wheelchair, tandem bicycle, winter and summer sports equipment, kitchen aids, the white cane, telephone, MP3 players, and camera.
13 – Calendar	All important regular and irregular events, such as birthdays, vacation plans, or work schedule
14 – Food and drink	What the person likes to eat and drink for breakfast, lunch or dinner, and special dietary needs
15 – Personal hygiene and need for assistance	What the person can do with or without assistance. Routines for morning and evening; special needs in the shower, special toilet requirement, clothing
16 – List of names	All important names, telephone numbers, mail addresses, who does what
Addenda	For example, general information about JNCL, information about epilepsy and medication, or medical contingency plans

The Annual Wheel Important events throughout the year not appearing on regular basis (birthdays, vacation etc.)			
Name of Person:		Erik Jenssen	Year: 2018
Specification of activity or event:		From:	To:
January			
	Dad's birthday	10/1/18	10/1/18
	Weekend with Mum and Dad	19/1/18	21/1/18
	Winter camp - Hurdal Center	22/1/18	27/1/18
February			
	Weekend with Mum and Dad	23/2/18	25/2/18
	Participation in Roeros Festival		
March			
	Sister Ida's birthday	21/3/18	21/3/18
	Participation in Melhus masquerade ball	23/3/18	25/3/18
	Weekend with Mum and Dad	24/8/18	26/8/18
April			
	Easter vacation (not specified yet)	9/4/18	15/4/18
	Weekend with Mum and Dad	27/4/18	29/4/18
<i>and so on.....</i>			

Figure 23.4 An Annual Wheel

Tool 3 – The Activity Bank

The activity bank contains activities related to the five areas of participation in an optimal adult life described earlier: (1) domestic activities, (2) work activities, (3) physical activities (4) leisure time activities, and (5) vacation and excursion activities. The activity bank is supposed to be used for activities that may change from day to day but which are of special importance to the person with JNCL. The aim is that the young adult with JNCL should be engaged in all the activities in the activity bank on a regular or irregular basis. Figure 23.5 shows the activity bank for a young adult with JNCL.

The *Book of (name of the person)* and the parents should be consulted when constructing the person’s activity bank. It is a vital source of information for staff both in the residence and at the working place, for instance for promoting sufficient physical activities to achieve the best possible physical fitness or for promoting or maintaining domestic and cultural skills.

Tool 4 – The Week Plan

The *Week Plan* is a week schedule showing the structure of the routine activities taking place every day (Figure 23.6). Some of the activities in the Week Plan are however open or not predefined and should be filled with activities from the *Activity Bank*. These activities can change from day to day according to the person’s wishes or needs, for instance in accordance with the goal of staying physically and cognitively fit or to maintain skills and interests in music.

Participation and Activity Bank				
Name of Person: Erik Jensen		Year: 2018		
A. Leisure time activities	B. Physical activities	C. Vacation and excursion activities	D. Domestic activities	E. Work related activities
1 Play Cards	1 Walking tour	1 Concerts	1 Go shopping	1 Sort screws and nuts
2 Play Board Games	2 Physiotherapy program	2 Café	2 Set the table	2 Polish cups and plates
3 Play Piano	3 Weight-lifting program	3 Restaurant	3 Cook simple food	3 Knitting machine
4 PC games	4 Visiting the gym	4 Fishing	4 Cut the vegetables	4 Make fire briquets
5 Listen to Audio Book	5 Swimming	5 Visit Parents	5 Empty the garbage bin	5 Empty paper bins
6 Listen to Music	6 Aerobic program	6 Visit sister Ida	6 Clear the bedroom	6 Fill firewood sacks
7 Make Music	7 Tandem cycling	7 Picnic	7 Do the dishes	7 Transport firewood to store
8 Singing	8 Mountaineering wall	8 Visit Grandfather	8 Make the box lunch	8 Sell firewood at market
9 Play the drums	9 Ski sledge	9 Winter camps	9 Bring the post	9 Prepare lunch to fellows
10 Play the harmonica	10 Wheelchair exercises	10 Summer camps	10 Make shopping plans	10 Make candlesticks
11 Call family and friends	11 Relaxation exercises	11 NCL peer meetings	11 Make coffee/tea to others	11 Parcel candlesticks
12 Check/write E-mails	12 Articulation exercises	12 Visit children's museum	12 Fetch firewood	12 Fill paper - copying machine
13 Socializing with neighbors	13 Stretching exercises in bed	13 Vacation trip to Spain	13 Dry the dishes	13 Label the products for sale
And so on.....				

Figure 23.5 An Activity Bank

Tool 5 – Registration of Participation

In the present study (Appendix A), many parents commented that they were barely aware of what was happening to the person with JNCL in the new home or at work. Some parents expressed frustration that the level of participation was lower than the expectations, as in the following quotation:

They said they were going to visit the indoor swimming pool on a weekly basis, but this has not happened so far.

Making daily registrations of events and activities is a means to avoid misunderstandings between parents and staff, and to support the aim of participation. Such registrations do not have to require much effort from the staff

Weekly plan							
Time	Monday	Tuesday	Wednesday	Thursday	Friday	Sat.	Sunday
07.00	Wake up, medication	Wake up, medication	Wake up, medication	Wake up, medication	Wake up, medication	Wake up, medication	Wake up, medication
07.10	Hygiene, dressing	Hygiene, dressing	Hygiene, dressing	Hygiene, dressing	Hygiene, dressing	Hygiene, dressing	Hygiene, dressing
07.45	Breakfast	Breakfast	Breakfast	Breakfast	Breakfast	Breakfast	Breakfast
08.15	Dressing for work	Dressing for work	Dressing for work	Dressing for work	Dressing for work	Dressing for work	Dressing for work
08.30	Transport to work	Transport to work	Transport to work	Transport to work	Transport to work	Transport to work	Transport to work
14.45	Return from work	Return from work	Return from work	Return from work	Return from work	Return from work	Return from work
15.00	Dinner	Dinner	Dinner	Dinner	Dinner	Dinner	Dinner
15.30	Domestic activities (act. bank)	Domestic activities (act. bank)	Domestic activities (act. bank)	Domestic activities (act. bank)	Domestic activities (act. bank)	Domestic activities (act. bank)	Domestic activities (act. bank)
16.30	Physical activity (act. bank)	Physical activity (act. bank)	Physical activity (act. bank)	Physical activity (act. bank)	Physical activity (act. bank)	Physical activity (act. bank)	Physical activity (act. bank)
19.30	Supper	Supper	Supper	Supper	Supper	Supper	Supper
20.00	Leisure time (act. bank)	Leisure time (act. bank)	Leisure time (act. bank)	Leisure time (act. bank)	Leisure time (act. bank)	Leisure time (act. bank)	Leisure time (act. bank)
20.45	Undress and hygiene	Undress and hygiene	Undress and hygiene	Undress and hygiene	Undress and hygiene	Undress and hygiene	Undress and hygiene
21.30	Good night	Good night	Good night	Good night	Good night	Good night	Good night

Figure 23.6 A Week Plan

Registration of Participation in Activities

Name: Erik Jenssen

Month and Year: October 2018

No.	Activities	Date:	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	29	30	31
1	Play Cards		✓						✓		✓	✓					✓		✓				✓	✓			✓	✓	✓	✓	✓	✓	
2	Play Board Games					✓			✓		✓		✓	✓	✓						✓		✓	✓			✓	✓	✓	✓	✓	✓	
3	Play Piano																		✓						✓							✓	
4	PC games				✓		✓		✓		✓						✓				✓	✓					✓					✓	
5	Listen to Music		✓			✓																		✓				✓	✓			✓	
6	Check/write E-mails								✓		✓						✓							✓					✓	✓		✓	
7	Walking tour								✓		✓				✓									✓								✓	
8	Physiotherapy program											✓				✓				✓			✓		✓							✓	
9	Visiting the gym		✓			✓			✓	✓											✓		✓						✓			✓	
10	Mountaineering wall						✓				✓																		✓			✓	
11	Relaxation exercises				✓		✓										✓										✓						
12	Stretching exercises in bed					✓							✓				✓						✓		✓								
13	Concerts								✓		✓						✓					✓	✓	✓					✓	✓	✓	✓	
14	Café							✓		✓	✓					✓	✓			✓		✓		✓				✓				✓	
15	Visit sister Ida		✓						✓		✓														✓		✓	✓	✓				✓
16	Visit Parents				✓	✓						✓					✓				✓				✓		✓						✓
17	Winter camps					✓	✓					✓											✓	✓		✓		✓	✓				✓
18	Set the table								✓															✓						✓	✓		✓
19	Cut the vegetables										✓						✓		✓	✓			✓		✓								✓
20	Empty the garbage bin				✓	✓																✓	✓		✓	✓							✓
21	Go shopping			✓				✓		✓		✓			✓									✓		✓		✓					✓
22	Sort screws and nuts		✓					✓		✓	✓	✓	✓				✓						✓		✓	✓		✓					✓
23	Knitting machine		✓																														✓
24	Empty paper bins					✓						✓									✓	✓	✓						✓	✓			✓
25	Make candlesticks		✓				✓			✓		✓					✓					✓	✓										✓
26	Fill paper - copying machine		✓			✓		✓	✓	✓		✓									✓	✓	✓		✓	✓							✓
27	initials (maximum 2 letters):	AG	ØE	ØE	ØE	ØE	ØE	SR	SR	SR	ØE	SR	ST	ST	ST	SR	AG	ØE	AG	ØE	AG	AG	AG	SR	ØE	ST	ST	ST	AG	AG	AG	SR	AG

Figure 23.7 Form for registration of participation in activities

and should be based on the quality of life issues collected from the *Activity Bank* and the *Annual Wheel*. Electronic registration may make the registration easy to maintain (Figure 23.7).

Figure 23.7 shows a registration form dated October 2018. Similar registration forms can be used for evaluations, for instance to explain why certain activities were not taking place according to plan. It may also be used for communication between parents and staff. Comments in the present study indicate that some staff members favor certain activities, and that the personality or interests of staff members will influence the daily programs of young adults with JNCL. This may not be wrong, but it is important that such patterns are made explicit and discussed. Daily registration of activities and participation may help to disclose such patterns.

Tool 6 – The Log Book

Residential staff, staff at work and parents should be required to post daily entries into a log book. The log book should contain information that is not covered by other forms of registration, for instance information about health, sleeping patterns, or unexpected changes in the daily program. The log book follows the person between residence, work and the family home, and is updated on a daily basis.

The local government may have an intranet for all their service providers and staff, where exceptional situations are noted, and to which parents may have (limited) access. Despite this, parents find that there is still a need for a handwritten log book. It is a personal, less formal and often more useful instrument, according to the parents. The log book can also be used to promote targeted dialogues and communication with the person with JNCL, for instance discussions about the events or happenings that took place last weekend with friends or the family.

Conclusions

It is possible for individuals with JNCL to lead a full life after reaching adulthood. However, given the circumstances, it is necessary to start planning the transition to adult life early, and to involve educational authorities and service providers in this project from the very beginning. Assistance and advice should be solicited from competent sources, which certainly will include parent organizations, local and regional authorities, and national competence centers where these exist. Maintaining and upholding the best possible quality of life for the young adult with JNCL require tight and effective collaboration between family, staff and the person with JNCL. There should be regular meetings between the family and the staff at the work place and in the residence.

The Habilitation Plan should be used for planning and implementing the transition to adult life. It should have a strong focus on participation in activities. It should be supplemented with tools like the Book about (name of the person), the Annual Wheel, the Activity Bank and the Week Plan. Daily registrations of what the young adult with JNCL has participated in may promote a good working culture among staff members and contribute to an optimal quality of life for the young adult with JNCL. In addition, parents find the informal Log Book to be an excellent tool to maintain effective communication between the family, service providers and staff.

References

- Bricout, J. C., & Gray, D. B. (2006). Community receptivity: The ecology of disabled persons' participation in the physical, political and social environments. *Scandinavian Journal of Disability Research*, 8, 1–21.
- Gray, D. B., & Hollingsworth, H. (1999). Development of new measurement tools for studying participation in context. Presented at American Public Health Association Annual Meeting, Chicago, Illinois, November, 1999.

- Krusche, S., Seitz, A., Börstler, J., & Bruegge, B. (2017). Interactive learning: Increasing student participation through shorter exercise cycles. In *Proceedings of the Nineteenth Australasian Computing Education Conference, January 31st to February 3rd 2017* (pp. 17–26). New York: Association for Computing Machinery.
- Magasi, S., Wong, A., Gray, D. B., Hammel, J., Baum, C., Wang, C. C., & Heinemann, A. W. (2015). Theoretical foundations for the measurement of environmental factors and their impact on participation among people with disabilities. *Archives of Physical Medicine and Rehabilitation*, 96, 569–577.
- Oakley, P., & Marsden, D. (1984). *Approaches to participation in rural development*. Geneva, CH: International Labour Office.
- Skye, E. P., Wagenschutz, H., Steiger, J. A., & Kumagai, A. K. (2014). Use of interactive theater and role play to develop medical students' skills in breaking bad news. *Journal of Cancer Education*, 29, 704–708.
- Stanford University School of Medicine (2018). Interactive learning. Downloaded from <http://med.stanford.edu/smili/interactive-learning/>
- Stark, S., Hollingsworth, H. H., Morgan, K. A., & Gray, D. B. (2007). Development of a measure of receptivity of the physical environment. *Disability and Rehabilitation*, 29, 123–137.

24

The Diagnostic Process and Parental Needs and Support

*Svein Rokne, Bengt Elmerskog, Merete Staureby, Susan Fugger,
Anne Hamann and Susanne Joensen*

Diagnosis implies the identification of a pathological condition. For parents it can be a process of shock and trauma when the diagnosis of their child is severe (Holliday, Stanley, Fodstad, & Minshaw, 2016). The family is vulnerable around the time they receive the diagnosis. Most children with a recent diagnosis of JNCL are not aware of their diagnosis, but the responses to questionnaires and interviews in the present project (see Appendix A) indicate that the children with JNCL and their siblings were affected by the new situation, by the changes in behavior and the strange events taking place. It is therefore important that there is support from competent professionals and, when possible, also from parent organizations or individual parents who have been through the same situation. It is vital that the process and the support given are perceived as supportive and useful by the family.

A typical story

The following story by a parent illustrates a diagnostic process that worked quite well.

Our son, who has JNCL, is 29 years old when this is written. We received this diagnosis some twenty years ago and have thus had ample time to think about the way we received these news and what happened during the process of medical investigation leading up to that moment in time.

We discovered quite early that something was wrong with his eyesight, and glasses were prescribed. They did not seem to help much and I remember quite clearly the day on a beach somewhere in Greece when our son, then eight years old, called our names, apparently not being able to locate us five meters away.

On further investigation at the hospital, his eyes were examined and blood samples taken. The blood sample disappeared and he was given the diagnosis of Retinitis Pigmentosa. Apparently this is (or was) often the "first" diagnosis given in these cases. Although this was a shock in itself, we insisted that blood samples be taken again. After several new examinations we were asked to a meeting where we were told the bad news. It must be said that this took place in adequate surroundings and with people (an ophthalmologist and a psychiatrist) who understood their jobs. We were subsequently able to have the diagnosis confirmed by DNA analysis through the good offices of Pirkko Santavuori (1933–2004) in Finland.

Parents are just as different as their children; some become extremely withdrawn and try to come to grips with their new situation on their own, while others seek help. Again fortunately, and unknown to us, there existed an organization which had been entrusted with a national responsibility to support the education of children and young people with this diagnosis and their families. Tambartun Resource Center immediately took charge of a number of practical matters which needed to be dealt with in an orderly fashion and which we, as parents, had no way of taking care of on our own, being at the bottom of the pit, as it were. We did, however, consult the Internet, but this did not give us much comfort under the circumstances.

Tambartun arranged meetings with the local school, they wrote the necessary applications for various technical aids, and we were persuaded that our son now needed to learn braille and that he had certain rights in terms of extra school time to learn this new language. We were also told that there existed a parent organization where we would find other parents in the same situation as ourselves. It must be said that in those days, this institution interpreted their mandate in the broadest possible terms, rendering a number of services not necessarily exclusively within the pedagogical area.

After a couple of years on our own, we finally contacted the parent organization and were able to discuss our situation with peers. This was a very positive experience, and membership in this organization has given us both much in emotional and practical terms over the years.

In retrospect it is clear that we were luckier than many others in our situation. In those days there were no clear routines, and in particular how the diagnosis was conveyed varied considerably. Finally, in 2011, the parent organization in collaboration with resource centers and medical institutions was able to create guidelines and a clear routine to be followed for all new cases. Much had happened in the meantime, of course, including the establishment of a national medical resource center. It has the responsibility

for children and adults with an NCL disease throughout Norway and for coordinating a group of institutions to support all people with NCL and their families. Information to this effect has been distributed to ophthalmologists and others in the medical field throughout Norway.

Requirements for guidelines for the diagnostic process

Several countries have developed guidelines for the diagnostic process, that is, the process from when an NCL disease is suspected until the diagnosis is determined and the family is in a stable situation. Guidelines that follow the requirements for good practice can go a long way towards providing a safe environment for new parents.

Good practice guidelines for the diagnostic process imply that there is a cooperative effort across service providers representing several disciplines, including medicine, education and social work. The guidelines spell out clearly the areas of responsibility and how communication should take place between the various parties.

The guidelines should define four phases: 1) Suspected NCL diagnosis, 2) Determination of the diagnosis, 3) Provision of information about the diagnosis, and 4) Immediate support phase. The guidelines should spell out clearly who is responsible, precisely what needs to be done and how it should be done. The aim of the guidelines is to ensure consistent best practice in relaying the diagnosis to the family and the immediate follow-up for the family. This should include the presence of representatives from the parent organization when possible, who will talk with the parents when and if it is desired by the parents.

The immediate support phase is completed when a functioning municipal support service for the family has been established and a Responsibility Group (see Chapter 11) for establishing and maintaining the individual plan or habilitation plan is in place.

Guidelines following good practice principles have been established (and are being followed) in several countries, including Denmark and Norway. The guidelines may be obtained from the national parent organizations or from the centers that are responsible for providing services to children and adults with NCL, and may serve as a template for providing good practice locally.

The guidelines practiced in Denmark today are presented below. It should be noted, however, that consideration should be given to local conditions when preparing guidelines in other countries.

Danish guidelines when a child is found to have JNCL

In Denmark, the municipalities are responsible for services for people with additional support needs, including persons with disabilities or a rare disease. The social welfare system allows families to choose either to keep their children or young adults with JNCL at home, or in an apartment or sheltered house provided by the municipality. In both cases, the young adults with JNCL will get the help and support needed, and they receive a social pension from the age of 18. At present, most young adults with JNCL in Denmark have their own apartment connected to their parents' home. For support and care, the municipality provides support and care, and teachers and nursing staff come to the home as well as to the school(s) or activity center.

The Danish NCL Team (The Team) is organized and financed by the government, and is part of a national institution for the visually impaired. The support and advice provided by The Team *supplement* the services provided by the municipality.

Guidelines for relaying information about the diagnosis

These guidelines are a highly valued tool for The Team, as they are covering maybe the most important part of The Team's work. The guidelines outline best practice(s) in supporting parents in a situation of severe shock after having received the message that their child has JNCL.

When a child has been diagnosed with JNCL at the Medical Genetics Laboratory of the Danish Kennedy Center or a local hospital, the coordinator or the medical consultant of The Team will be contacted in order to find a time and place for a meeting with the parents. It is very important that The Team is contacted *before* the doctor informs the parents of the diagnosis. The Team will contribute to arranging this meeting in the best possible way. The doctor is advised, in advance of the meeting, to inform the parents that they are called in for a serious conversation and advise them not to bring the child or any siblings.

Two members of The Team – preferably one pedagogical consultant and a social counselor – will be at the hospital on the day the parents receive information about their child's diagnosis, and will join the meeting after the doctor has informed the parents. The Team will have a short preliminary meeting with the doctor, in preparation of the doctor's meeting with the parents. The purpose of The Team's presence is to offer help in a chaotic situation and inform the parents of The Team's existence and that assistance from The Team is available from this moment.

According to the guidelines, the doctor informs the parents of the child's diagnosis, and then the two members of The Team are invited to join the meeting. At the meeting, the two representatives of The Team tell the parents about The Team, that the function of The Team is to support the family in different ways. In addition to a brief presentation of the work of The Team, and equally important, the parents are informed about NCL Danmark (The Danish Parent Association) and that the two parent members of The Team are prepared to meet parents if and when the parents wish to do so.

The Team offers to visit the family as soon as possible, preferably the next day. A declaration of consent form is brought to one of the first meetings with the parents, allowing The Team to inform the family's municipality, school, and other services when needed. The Team offers to drive the parents home in the parents' own car after the diagnosis meeting.

Support from the professionals in The Team

The parents will be shocked after the diagnosis meeting, and it is therefore of importance that they are supported without being overwhelmed. Over time, The Team will talk with the parents about present needs, who they should involve, and future needs. These conversations take place in a way and at a pace that the parents can cope with. The situation of the family has changed dramatically, but the child's life situation at this time is not changed as a result of the diagnosis.

The Team offers to participate in a meeting with relatives and friends of the family to inform them about JNCL and answer the many questions that arise from people close to the family. The parents decide if and when they want to have a meeting, and who they want to include.

The parents often wish to see the medical consultant of The Team, meaning the professor and chief physician at the Center of Rare Diseases, where all the children with JNCL receive continuous follow-up. At this stage, the medical consultant can give insights into the disease, whereas later with the progression of the disease, the medical consultant will be closely involved with the parents in relation to the medical treatment of symptoms and other medical issues.

As soon as the parents are ready, the coordinator and the pedagogical consultant of The Team will contact the local municipality in order to inform the school, kindergarten, etc. of the diagnosis, and of the present and future needs of the child. The Team recommends the creation of a network of professionals of the municipality: social workers, teachers, pedagogues, visual consultants, and so forth. Both The Team and the parents participate in these meetings.

Support from NCL Denmark

With the parents' approval, all the members of The Team are informed that a new child has received a JNCL diagnosis. It varies how early the parents want to meet other parents. However, the message from the parents in The Team is clear: «Do not hesitate to contact us».

The leader of NCL Denmark (Danish NCL Family Association) is also informed that a child has been newly diagnosed with JNCL (see Chapter 26). Also the other members of the Association are informed, possibly without mentioning names, if the parents give their consent. At the diagnosis meeting, the parents are informed about the Association and its activities and services for children and parents. They are also told that one purpose of meeting NCL Denmark is to learn from other parents in same situation, that is, from parental experiences based on «learning by doing». These procedures have generally been highly valued by parents of children with JNCL in Denmark.

How are parents supported?

Parents have considerable needs for support in addition to medical support. The family needs to know how to cope with the needs of their child but also teachers and local authorities need information as soon as possible after the child has been diagnosed. The competence needed is in most countries represented by parent organizations and competence centers. Usually, neither the teachers nor the local government have previous knowledge of JNCL.

The parents in the present project (Appendix A) were asked to consider who gave useful information during the first year after the diagnosis was confirmed. Figure 24.1 shows that parents found that other parents and parent organizations gave most useful information, together with competence centers. In addition, books, articles and the Internet were also considered good sources of information. Parents in other studies have also mentioned the Internet (Cozart et al., 2017). The health sector was expected to be an important source of useful information but the results for the present project may indicate that information from the health sector was not useful in helping parents to meet the challenges of daily life. Medical information about JNCL is important but tends to be more about declines than what can be done to support the child with JNCL in the best possible way. The municipality or other local authorities, such as educational or psychological agencies and schools, were regarded as giving little useful information (Figure 24.1).

Typical comments from parents in the questionnaires or interviews are that local doctors, physiotherapists and the municipality do not have sufficient

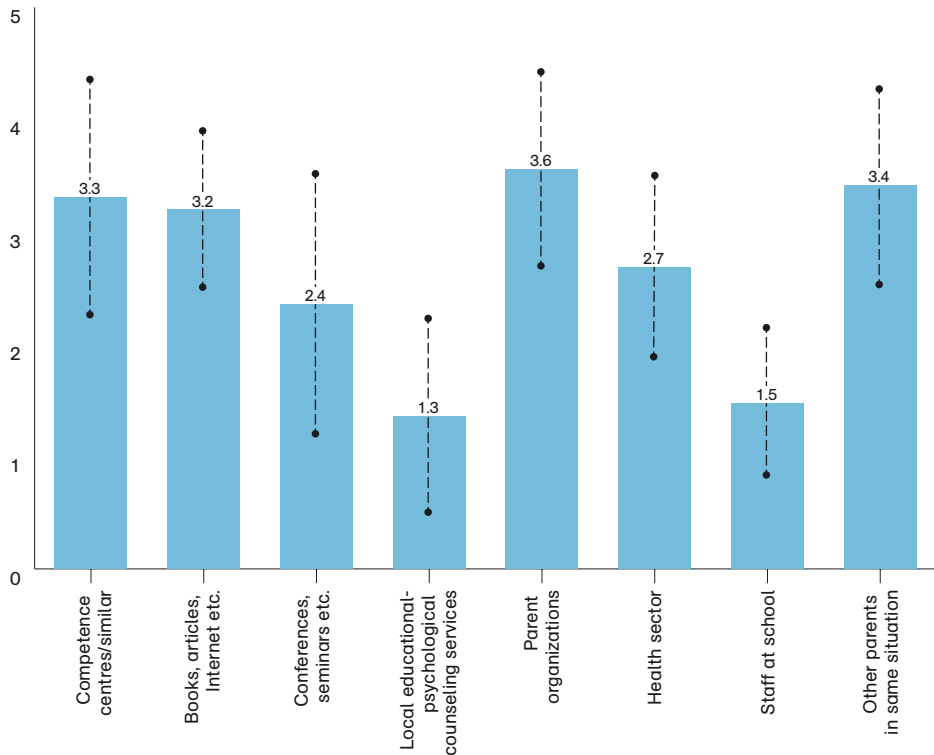


Figure 24.1 Parents' evaluations of information from different sources; average scores on a scale from 1 (no contribution) to 5 (very high contribution), and standard deviations

knowledge about JNCL to provide good services. The contact between the family and the health sector tended to be rather limited in the early phase of the disease. Many parents said that they had to "clear the road themselves," sometimes in collaboration with resource centers and parent organizations. Specialist organizations such as BDSRA in the USA, Onerva in Finland, BDFa in UK, The NCL Team in Denmark, BZBS in Hamburg and Statped in Norway were mentioned as giving useful information. Some parents emphasized that competence and usefulness of information provided were related to individuals in the system rather than the system itself. In fact, some parents considered local authorities more as a burden than as a support.

It is often stated that parents of children who have recently received a severe diagnosis are not receptive to information related to education, development and learning. However, comments in the questionnaires and interviews in the present project emphasized the following urgent need. – «What can I do to help my child in best possible way?» There is always a risk that preconceived assumptions may

preclude proper action by professionals, for example they may give less information because they believe the parents are not ready for it. Schools and other institutions should provide services in accordance with the needs of the individuals. The results from the survey emphasize the importance of implementing tailored initiatives as soon as possible after the diagnosis has been established.

The Danish system illustrates that it is possible to establish an efficient system for managing early interventions and immediate measures. The Team, with different professionals and parent representatives, has been given a strong mandate to contact and follow up families when a child is diagnosed with JNCL and support authorities and institutions in the municipality when needed. The Team is given resources to support all such families in Denmark. The fact that The Team consists of professionals from the health sector as well as other services makes it possible to act fast when needed. Feedback from the Danish families indicates that they are satisfied with the support they receive from The Team.

One important finding in the present project is that specialists within the health services and specialized educational services, as well as parent organizations, are the key sources of useful information for the families and schools. These key representatives can be found in all the countries participating in the project, however, they vary in availability and in how they can coordinate their support. All the Scandinavian countries have a registration system and a service structure which ensure that all children with JNCL and their schools sooner or later will be contacted by the support systems. The systems are different in the USA, UK and Germany. These differences may be related to variation in population and geography but also in national laws and regulations.

Denmark has developed a model for effective accessibility. Also Finland and Norway have established systems where families and schools sooner or later will be offered competent support. However, the coordination or collaboration between the health services, educational services and social services in these countries is not always as efficient as could be desired.

The immediate support phase

Some parents may not be able to act appropriately just after the child has been diagnosed with JNCL. Life may be filled with grief, shock and trauma. This is a phase where parents might need a support system that can act proactively on their behalf. The main issue in the immediate support phase is the child's schooling. A well-functioning school is an effective means of supporting the families.

From the child's perspective, the school situation is not different immediately after the JNCL diagnosis is established, provided the education accommodates the child's daily needs at school. However, there is an immediate need for *building competence* in staff, related to childhood dementia and especially to precautionary learning and hastened learning (see Chapter 12). Parents may not be able to be involved in such interventions at the beginning, but it is necessary that parents are informed about the actions that are taken to promote the best possible development in a difficult situation. At this time, it is also important to establish a close collaboration between the school and institutions that have the required competence about education for children and young people with JNCL. This should be handled as a matter of urgency – there is no time for waiting. The questionnaire comments and interviews of the present project show that many parents highly appreciated the presence of early proactive actions taken on the child's and the family's behalf until the family situation had become more stable. However, they also show that many parents felt very lonely in the beginning and lacked the immediate support from the community and the education system. In the words of one parent:

There were a lot of people involved in the beginning which was very overwhelming. I don't really know how school was supported. I think the school tried to find information. I did very often mention the BDFA but that was never taken up. Maybe the RNIB was contacted.

This is no criticism really of the school. They couldn't – they didn't really allow my child to go out into the playground and play with the other children. Because focus, obviously health and safety and – and I understand it. They – they wanted to make sure she was safe. So that became hard and difficult for them – and us.

I felt I couldn't trust the teachers and their competence. In my opinion, the teachers were not able to deal with the illness and did not have sufficient expertise. They covered a basic program, but it was me who supplied my daughter with additional materials. For instance, I prepared workbooks or exercises from the early beginning.

This short comment emphasizes the need for both information and action, for parent support as well as dialogue and participation. The function of the guidelines is to provide a foundation for this.

References

- Cozart, E. J., Augustine, E. F., Mink, J. W., Thatcher, A. R., & Adams, H. R. (2017). Batten disease and parents: Marital quality, support, and communication. *Journal of the International Child Neurology Association*, 17, 114.
- Helland, I. B., Barlinn, J., Åbyholm, G., Paus, T., & Rokne, S. (2016). *Guide to symptomatic treatment of neuronal ceroid lipofuscinosis*. Oslo: Norsk Spielmeier-Vogt Forening and Oslo University Hospital.
- Holliday, E. L., Stanley, H. C., Fodstad, J. C., & Minshawi, N. F. (2016). Stress and satisfaction in the diagnostic process. In J. L. Matson (Ed.), *Handbook of assessment and diagnosis of autism spectrum disorder* (pp. 137–155). Cham, Switzerland: Springer.
- Labbé, E. E., Lopez, I., Murphy, L., & O'Brien, C. (2002). Optimism and psychosocial functioning in caring for children with Battens and other neurological diseases. *Psychological Reports*, 90, 1129–1135.
- NCL Danmark (The Danish NCL Family Association). <http://dsvf.dk>

Being the Brother or Sister of a Child with Juvenile Neuronal Ceroid Lipofuscinosis

*Minna Sarola, Egil Rian, Linn Sophie Rian,
Charlotte Holmen and Ida Holmen*

The family is a social system, where the members influence one another. Children spend much time with their siblings, often more than with their parents (Howe & Recchia, 2014). Sibling relationships represent both social resources and challenges, and the family's emotional climate affects the relationship between siblings (McHale et al., 2012). Sibling relationships are usually based on some sense of being equals, even though age differences can lead to a number of differences in status and physical strength. When one sibling has a disability, this may change the relationship in some ways (Meltzer & Kramer, 2016). Children with severe disabilities often become "younger siblings", regardless of their actual age ranking in the family. In many areas, the relationship is the same as in siblings without disabilities but characterized by more warmth and care than other sibling relationships (von Tetzchner, 2019). For severely disabled children, who may have difficulties engaging in ordinary peer relations, siblings typically have a very special role (Petalas et al., 2015).

This chapter presents experiences of being a sibling to a child or young person with juvenile neuronal ceroid lipofuscinosis (JNCL). It includes results from a study where siblings of six individuals with JNCL were interviewed (Sarola, 2016), a sister's and a mother's account of the sibling relationship, and a father's interview with his daughter whose brother had JNCL.

Six families' views on sibling relations

A child's illness or disability is always hard for parents and when a family member falls ill, it affects the whole family (Mäenpää, 2014; Seligman & Darling, 2007). It may absorb many of the parents' resources and influence the life of the entire family.

When he was at home there were a lot of things to take care of, like feeding him and helping him to go to the toilet. It made everyday life much harder but we survived pretty well.

We had ramps installed in our house and everything you need when you use a wheelchair.

When a child in the family is diagnosed with JNCL, it therefore affects the life of the siblings. Anxiety, fear, uncertainty and mourning of functional loss become a part of everyday life (Kaulio & Svennevig, 2008). Much of the time and energy of the parents may be focused on the child who is ill, and the siblings may get less attention. In particular when the child with JNCL experiences health problems or needs hospitalization, the parents' thoughts may be pre-occupied with the treatment of the child. This inevitably affects the life of the siblings (Kulomäki, 1984). Parents and professionals therefore need to be mindful of the siblings and their need for support (Sarola, 2016).

Siblings may also feel that after their brother or sister has been diagnosed with JNCL, there are greater expectations on their own shoulders. This can cause considerable pressure, both at school and at home (Kulomäki, 1984). Sometimes siblings of children who are ill feel the need to protect their parents from any extra stress, and therefore hide their grief (Kaulio & Svennevig, 2008). Older siblings may take on a caregiver role and take great responsibility for the sibling who is ill, as well as for everyday household tasks. Moreover, siblings may feel that this is the only way to get their parents' attention and acceptance (Kulomäki, 1984). However, under a surface of strength and kindness there may be hidden feelings of anxiety or depression (Kaulio & Svennevig, 2008). One of the siblings in the interview study described the situation:

One year after the diagnosis, it started to show in me. I was depressed and I suffered from an eating disorder. It happened because I hid my own problems and tried to be as good as I could be to make my parents' life a bit easier. At school I only got top marks.

The interviews confirmed that the illness of the child was a permanent part of the everyday life of the siblings. In fact, the life of the family was to a large extent determined by the nature of the illness of the child. When an illness is progressive like JNCL, the family has to adapt to new situations continuously. All the siblings mentioned that the parents gave more time to the child with JNCL, and that they had to some extent concealed their own problems and worries to make their parents' life a bit easier. The siblings also found it necessary to take

more responsibility for themselves than a child of their age would normally do. Adults should always remember that there may be more going on than the sibling might show.

Disease evokes feelings

The attention given to a child who is ill may cause envy in siblings (Kulomäki, 1984). A child's disability can raise a variety of emotions in siblings, such as anger, joy, confusion, fear, love and pride (Finnish Association for Parents of Children with Visual Impairments, 2015). Siblings may also feel guilty because they are healthy while their sibling is ill (Batten Disease Family Association, 2015), as this sibling explains:

Sure I thought about why I get to be healthy while my sister is ill. When you see your sister suffering, it makes you wonder what right you have to be healthy.

The way parents respond to the disease will also influence the reactions of siblings (Kulomäki, 1984; Trachtenberg & Batshaw, 1997). The child who is ill is a large part of a sibling's life and the situation may make them more open minded and also affect their identity formation. Siblings may wonder what their life would be like if the child did not have a disability and how different it would be if the child was not ill (Kulomäki, 1984). Allowing all kinds of feelings and talking openly within the family may help siblings adapt to an extraordinarily difficult situation (Kulomäki, 1984).

I felt my parents' sadness even though they tried to cover it up.

At times I was ashamed of my brother even though I thought it was wrong to feel that way. But as a child, illness sparked all kinds of emotions.

I'm not bitter towards my parents even though most of their energies went on my sisters, but still I feel that I was little bit of an outsider in the family.

All the interviewees felt that it was natural for a sibling to experience a wide variety of feelings. They also said that it was difficult to witness their parents' grief. The interviewees felt that it was important for a sibling to feel that all possible emotions are natural and permitted. They also considered it very important to have opportunities to discuss the emotions that the disease had evoked in them, either with their parents or with another adult.

The need for information

All the interviewees expressed the view that siblings have a right to know about the diagnosis. They all felt strongly that factual and age-appropriate information had helped them understand the disease and its symptoms. The siblings interviewed also pointed out that children are very sensitive and notice if their parents are not coping. There is an abundance of inaccurate information easily accessible on the Internet. All the interviewees said that a lack of knowledge or incorrect knowledge is much more harmful than factual and reliable information about the diagnosis.

When you have enough information, you are able to understand that the symptoms are just part of the disease and that makes it easier to handle.

I was told about the disease little by little. I was around ten years old when I heard where it would lead in the end.

The fact that JNCL is a hereditary condition also raised some important questions and the siblings had just as many thoughts and questions as the other members of the family (Norio Centre, 2015). Factual and age-appropriate information is important when the illness of a child raises questions. Siblings need to know why the child who is ill needs so much attention and care (Kulomäki, 1984). A rule of thumb is to answer any questions that the sibling is asking about the child's illness honestly (Vehmanen & Vesa, 2012). However, it should be borne in mind that a sibling may not know how to ask all the relevant questions. For example, if the parents do not talk about JNCL but only about visual impairment, a sibling may not be able to ask questions about JNCL, and may therefore depend on other accessible but misleading information (Parisaari, 2015). One of the siblings in the study expressed this view.

If you sort of know in advance that the ability to speak and move are going to get worse you are able to better appreciate the times when those skills are still there, and you don't feel like you wish you had known it was going to get this bad so you could have enjoyed the moments when she was less affected.

Children under school age are often open-minded towards diversity, and at this age a sibling's disability does not necessarily raise any particular thoughts. In general, school-age children begin to reflect on these matters: Why is my sibling different? Am I different? Why do some people have disabilities? What causes disabilities? At this point, when the questions are topical, it is essential that the

relevant information is to hand. Incorrect perceptions can cause anxiety and fear. For example, a sibling may believe that he has caused the child's disability if he does not have correct information about it (Vehmanen, 2012). Moreover, it is important for siblings' self-esteem and development that they know how to tell their friends about the disability of their brother or sister (Vehmanen, 2012).

Peer support

All interviewees said that support from peers was vital. Some of them had experienced peer support while others wished that they had received more. They all said that it was a relief and empowering to meet others in the same situation. It was comforting for them to realize that there were other families in the world experiencing a similar situation.

A peer group can share common traits such as age, gender, culture, religion or ethnicity, shared experience, sexual identity, health or education (Rubin et al., 2015), and many support programs focus on peer relations (Western Australian Centre for Health Promotion Research, 2015). The idea of peer support is to give and receive help by sharing experiences (Inclusion Finland KVTL, 2015; Repper, 2013). Peer support and professional help are two different things. Sometimes a peer can offer the best help to another person (Kinanen, 2009; Repper, 2013).

I would have liked to have met other families in the same situation. Sometimes it felt like we were the one and only family like this and nobody else could understand what it was like and how we felt.

The interviewees said that meeting other families was also important because they saw other children who were at different stages of JNCL, even if it was hard to see children where the disease had progressed further than in the child in their own family. This was important because seeing how the disease would progress helped them prepare and adapt to the idea of their brother or sister getting worse. The interviewees said that it made them appreciate the time when the child's general condition and mobility were still good. The siblings said that seeing other families with a child at a different stage of the disease progression evoked feelings and questions which had been important to talk about with an adult. Seeing how other families had managed their situation gave them comfort and hope for the future.

I remember that it was frightening at first to see how the disease had progressed in children of other families. On the other hand, it was very important because in the future your own sibling's disease would progress too, and seeing others was a way to prepare yourself for it.

In conclusion, the results of the interview study indicate that adequate information and peer support have significant value to the sibling's ability to live happily as part of a family where a child has JNCL. Kinnunen (2006) found that peer support has a positive effect on parents of children with special needs. Peer support is seen as hugely beneficial when, for example, dealing with the feelings that a child's special needs evoke in parents (Kinnunen, 2006). Kolehmainen and Pulkkinen (2009) found that sisters of those with an intellectual disability felt that the peer support that they had received was empowering and useful, and those who had not had peer support felt the lack of it. Their comments support the findings of this study.

Reflections of a sister and a mother

This subchapter consists of two independent narratives about a young man, Nils aged 25 years, with JNCL. The first is written by the young man's sister, Ida, when she was 23 years old. The second is written by the young man's mother.

Being the Sister of my Brother

It is not easy growing up as the sibling of a child with special needs, but with great challenges come great rewards. I could not have asked for a better family and I am forever grateful for the joy my brother gives me and for the love my parents radiate towards me. But it was a bit harder for the blond, shy, little girl to understand the great strain that had been put on her parents when she was just a child. I was three years old and my brother Nils was five when he was diagnosed with JNCL. At that moment, the dynamics of our whole family changed.

The different adaptations and coping mechanisms I developed during those early years have become a large part of who I am today. My parents' time and energy were, by necessity, divided unequally between me and my brother. I adapted to the fact that my brother always took the bigger piece of the cake, so I convinced myself that the smaller piece was good enough for me. I felt my parents' grief, despair, and stress, so I tried as hard as I could to make them happy, and to unload their stress. I understood that my parents had enough to worry about, so I stayed out of trouble and became a perfectionist and peacemaker. In all of this "rightfulness", I lost myself. I did not realize this until I moved away from home, and found myself at the age of nineteen with no clue about my own needs and wants, not even what I preferred to eat for dinner. I had a hard time making decisions, even with no-one else to consider in the equation. I only had to focus on my own desires and this paralyzed me. I was so used to choosing whatever was best in a situation by



Ida and Nils 20 years ago.

calculating all the different variables that I forgot to pay attention to what I wanted. I guess knowing what you want is also a part of growing up, so it is not only my role in the family that got me to that point. I still have some problems separating what I want and what I should want, but as I get older and more confident, I am also learning more about my own needs and desires.

It is important that the sibling of a person with special needs feels seen and heard. It is easy for an adult to forget that the three-year-old sitting on their lap and smiling also is feeling what is happening to the family. Even though it is totally understandable that the person who is ill gets more attention, it is important to be aware of the impact of this on the sibling. I have always felt the enormous love my parents have for me, and I know that they have done their best for both of us, and that is all they can do. I am lucky and truly thankful for the childhood I have had. Lucky in the sense that my family is resourceful and we have been able to create a good life from what has been given to us. Luck should not be a variable in the debate of outcomes. All families should be given the possibilities we have, and everyone should know their rights.

I have now lived in the United States for a couple of years, and the hardest part is being away from my family. We are very close and a great team together, so it is difficult for me to feel whole without them. I feel both beautiful joy and dark pain every time I come home after several months away. The beautiful love



Ida and Nils today.

we have for one other flourishes, but the severity of the disease hits me when I can see more clearly that my brother is slowly getting worse each time I am away.

The hardest thing for me to face is that in the future my eccentric food-loving brother will not be able to eat normally, my curious question-machine will not be able to talk, my active nature-loving adventurer will not be able to walk, and I will not grow old with my hero by my side.

At the same time, the ability to eat, talk, walk or live can be taken away from any person at any time. The difference is that my family has already been given a kind of guarantee that it will happen to one of us. I do not want to say that we are lucky that this has happened to us. Absolutely not. But I want to say that we are lucky in the great love our situation has brought us. Love that touches more people around us than we know.

I am glad that I did not hear the whole truth about JNCL until I

was eighteen. It would have been hard to understand the fact that my brother, most likely, will not always be by my side. It is still hard to understand. I chose not to do any research about the disease the first years after I heard its name. I was, and still am, confident that my brother has a new form of JNCL, so I figured I would not bury myself in the dark truth if there is a chance that it does not apply to him. Since we have been advised to protect my brother from knowing the outcome, we have chosen to keep the severity of the disease from our community. Growing up with a big family secret also shapes you. None of my friends really knew what I was going through and how serious my situation actually was.

When I look back, it was hard growing up faster than most of my peers, but I think it has only made me stronger. I am good at keeping secrets and have

become a trustworthy person. I also feel that I have developed a stronger sense of self and what really matters in life. I still remember scenarios from high school where some of the girls were devastated because they got the wrong color phone for Christmas. A feeling of annoyance surged inside me when they could not see the insignificance of such a problem.

Today, the three-year-old me has become twenty-three years old, and I am extremely proud of each member of my family. I am convinced that we would not be filled with so much love and strength without the challenges we have been through. We will experience great grief and sorrow in our lives but we will have each other, and that is all that matters. To be the sister of my brother is the best thing in the world, and I cannot wait to move back home.

Siblings – A mother's reflections

It has not always been easy to be the little sister of Nils. He always demanded a lot of attention from me and his father, and he needed a lot of help. Our daughter Ida retreated and withdrew in most situations, patiently waiting for her turn. Nils dominated my life and his father's life; he was loud and Ida was silent, and he would shout if not receiving enough attention. Ida has never complained, but we observed that she often felt sad. It is difficult to put things into words when so young.

Nils is very proud of his little sister Ida. He has always considered her clever and beautiful. He boasts and brags about her whenever he is given the chance. His loud bragging and boasting about her were at times a bit embarrassing for Ida, but she has never been uncomfortable about being his sister. She was always proud of him and included him in social gatherings, when having friends home or having a party. That he may have looked at bit different, or said weird things, never embarrassed her. She always defended and supported him, irrespective of his appearance and behavior.

Her experience as Nils' sister has affected Ida and her personality. She always defends those with special needs or those who are different in any way. Everyone has the right to be met with dignity. Ida gets angry and despairs when less fortunate people are treated badly, unfairly or without respect. She never judges other people. Her relationship with Nils has made her an open, understanding and helpful person.

Ida was three years old and Nils five when we received the diagnosis at the hospital. Ida was staying with her grandmother and I have always wondered what she thought, felt or understood when we returned home with the diagnosis. She certainly understood that something was wrong. We were sad but we tried our best to keep our spirits up in front of her.

Our lives changed totally after we received Nils' diagnosis. Work and other things in life that had been important before suddenly became insignificant. Other questions took over: Is there really no treatment? Do we just accept the situation? Cancer is treated with chemotherapy and radiation. Can this treatment be used for Nils? Shall we sell everything we own and move abroad to find someone who can help us? Where can we find help? We spent many hours on the Internet looking for answers but there were none to find. What we found and read online was terrible and depressing and it just pushed us further down.

At first we felt panic. We must allow Nils to experience as much as possible before he becomes blind, dependent on a wheelchair and loses his speech. We must travel and allow Nils to see and experience as much as possible before he becomes blind. We must allow him to learn as much as possible before it is too late. But eventually we calmed down. We realized the most important thing for Nils is to feel comfortable with friends and family in his home environment. He needs predictability, security, love and the same challenges as other children his age. He must learn to cope with daily challenges and he must continue to learn new things. Our primary goal was to enable Nils to have the best possible quality of life.

But what about Ida in this chaos? From the very beginning she was a shy and quiet child who did not demand anything. Nils took up much of our daily life. Ida had a more hidden role at home. She quickly became Nils' helper, showing great care and sympathy. Being caring and sympathetic is good in itself, but it worried me that she took too much responsibility. I was afraid that her adaptive role in the family and her selflessness went far beyond her own needs.

I have felt bad because I spent so much time on Nils and less time on Ida. There was always a need for complex planning and organization with Nils. This is a dilemma that I am still living with 20 years later.

Another dilemma I have had through the years concerns Nils "getting everything he is pointing at". Ida has hardly asked for anything and I sometimes feel she has been "forgotten". I remember thinking «it is now we have the opportunity to do things together. In a few years Nils may become so disabled that it is too late. This is our time of opportunity». At the same time I was thinking «we have more time with Ida, it is no hurry». But I soon realized it is wrong to think that way. We live here and now, regardless of any disease, and we have to make the best use of time for us all. Wasted time will not be given back and quality time with Ida is important now and in the future. We are always keen to give a lot of praise and positive feedback to Nils and Ida. I think this is very important, regardless of the situation and despite their abilities.

Early on we focused on establishing an "open house" for Ida and Nils, and their friends were always welcome. There have been lots of indoor and outdoor activities taking place. At times it has been tiring, but we are grateful that we have

been able to give priority to this. I am sure the open home has benefitted both Nils and Ida. They are both very sociable with wide social networks. Nils is now living in his own flat with staff funded by the community. Many of the staff were childhood friends of Ida and Nils, and we can see that those early relationships have a positive effect on their work with Nils today. Their efforts are based more on friendship than paid employment.

The fact that Nils is blind and has epilepsy is of course a difficult issue. However, these challenges are easier to deal with than the ongoing cognitive decline. The prospect of our son and Ida's brother having dementia at a young age and eventually losing his speech is tough for us. Nils does not know about his disease. He is aware of his blindness, epilepsy and that he sometimes has problems finding the right words.

Ida was eighteen years old when she was told the full truth about the disease and what is going to happen to Nils in the future. We took the decision to wait until she was eighteen mainly because we wanted her to have a childhood as normal as possible, without too much worrying and sorrow too early in her life.

What could I have done differently as a mother? I have certainly made mistakes, but at the same time it is important to remember that I have also done a lot of good things. Either way, myself and my husband are only human. I think we have done what was possible in our situation.

Ida has learned a lot from having a brother with JNCL, a learning experience few other siblings will experience. Today, she is strong and wise with a great perspective on human life and she has great empathy and respect for people with special needs. These are good qualities for the rest of her life.

Perspectives on siblings

This account is written by a father and his daughter about their son and brother with JNCL. The young man, Henning, passed away some years ago. It is based on an interview, where the father asks his daughter about her experiences as a sister to her brother with JNCL.

Introduction

A story must start somewhere. This family story started in the winter of 1983 and 1984. At the time we thought we were a completely ordinary family of four: mother Vigdis, father Egil, six-year-old big brother Henning, and little sister Linn Sophie who was barely two years old. An ordinary family with good prospects. Most things happened as we expected in life. We – the adults – had both found

a partner we were happy with, we lived in a nice house, and we had good jobs and nice friends. In addition, we had two beautiful and healthy children. Our lives progressed as we had wished; things looked really promising, with no major problems. Of course.

But our family had reached a turning point. During the winter of 1983–1984, it was discovered that Henning had some vision problems. We thought these problems would be corrected with a pair of glasses or something similar. Henning's vision was assessed by an ophthalmologist. When we returned to the doctor, nothing turned out as we had hoped or expected. The doctor was very worried and immediately referred Henning to hospital for further assessment. We were told that something could be seriously wrong with Henning. In April the same year, Henning was given a diagnosis that hugely affected the life of our entire family. Of course, we had never heard of Spielmeier-Vogt disease or JNCL before. The information the doctor gave us about Henning's future shocked us deeply. Nevertheless, life had to go on. We have tried to cope with the situation the best we can, despite all the challenges we have experienced over 30 years. It has been a very long journey for us all.

Henning lived a long life, considering the typical outcome of the disease: he was 38 years and three days old when he passed away. We do not know anyone else with JNCL who has reached this age. When we, the three remaining members of the family, are sitting together reflecting on our history and where life has led us, we have many memories. Many good and many difficult memories. The sadness of losing Henning still marks our lives, but we are also very grateful for everything we received through Henning and the resulting close family bond. Many have commented that we became a very close and intimate family. A good friend referred to us as the "Rian Club" and we identify closely with this name.

We – the remaining family members – have our mutual experiences and history, but we also each have our own, individual story. What we remember and what we have forgotten are individual matters. The same story can be told in different ways, based on how we process memories. The same story can also be shaped differently, based on individual differences in interests and emotions, and what is of importance to the individual. This is the story of little sister Linn Sophie.

The story of little sister Linn Sophie

FATHER: Linn Sophie, you were not even two years old when Henning got his diagnosis. This means that we – your parents – had to reflect on your behalf on the first challenging years of your life. Even if you did not understand what was happening to Henning and our lives, we could see that you clearly noticed certain things and family challenges. You noticed the change