

General Information on Educational Management for Young People with Niemann-Pick Type C

The following information is aimed at teachers working with young people who suffer from Niemann-Pick Disease Type C (the abbreviation NPC will be used hereafter in the text). Only a brief reference will be made to the medical aspects of NPC to illustrate the progression of the disease. Readers should familiarize themselves with the medical aspects of the disease in order to help them understand the different types of Niemann-Pick Disease and the varying age of onset of Type C.

This article will restrict itself to the Type C where a young person experiences a healthy life until primary school age and then shows a gradual decline in both mental and physical functions.

For teachers, it is important to know the age of onset and to remember that, prior to the diagnosis, the child attended a mainstream school and had a healthy life. As with some other neurologically degenerative conditions, i.e., Juvenile Batten's Disease, it is probable that the long-term memory of young people with NPC remains with them until the final stages of the disease although the level of functioning deteriorates. Unlike children born with severe brain damage, children with NPC have a reference point of "normality" that stays with them while other faculties deteriorate. Changes in educational placement, i.e., from a mainstream school to a special school or class for children with severe learning difficulties may cause the child distress, frustration, and loss of self-esteem. The young person may be distressed and confused by the inappropriate behavior of other pupils and the lack of peer group language.

The pattern of deterioration in children with NPC is specific to the individual, but there are stages which can be identified. In the initial stages there are learning difficulties with the child beginning to experience some problem in concentration and short-term memory recall. Speech may begin to appear slurred and the child may struggle to find the appropriate words to communicate. Motor coordination may appear slightly impaired. The child will experience difficulty with upward and downward eye movements (vertical supranuclear gaze palsy) which will make copying from the blackboard difficult.

In the middle stages these difficulties become progressively worse. There is unsteadiness of gait, clumsiness, problems with walking (ataxia), slurred, irregular speech (dysarthria) leading to eventual loss of speech and difficulty swallowing (dysphagia), sudden loss of muscle tone which may lead to falls (cataplexy), difficulty in posturing of limbs (dystonia), tremors accompanying movement and sometimes epileptic seizures which can be difficult to control. The type and frequency of the seizures will be specific to the child.

The final stage, after several years, will involve more and more nursing care with educationalists offering a stimulating program to maintain general awareness.

The Middle Stage

The following section will concentrate on the role of teachers working with young people in the middle stage. Knowledge of the medical aspects of the disease, together with gathering firsthand observations of the pattern of the disease specific to the young person, will help staff modify, where necessary, the curriculum and management strategies. The fact that a child with NPC begins formal education in a local

primary school and has experienced everything socially and educationally within that peer group should never be forgotten by teachers. While in the middle stages, the child may continue to attend a local school with additional support or may transfer to a special unit within a local mainstream school. The type of school a young person with NPC attends will often depend on the resources within the local educational authority. A new school placement which bears little resemblance to the young person's previous school environment and expectations of school life may cause distress and resentment.

Modifications to the Curriculum

The most significant change will be in the written curriculum. As writing skills deteriorate, some children will find it easier to use a word processor or to move from a written curriculum to oral communication using tape recorders. Some young people with NPC may not want staff to scribe for them as this will serve to emphasize their disability. The child may still attend classes in English, Geography, etc. But when it comes to recording their work, they will do it orally with support staff.

Recording personal interests and activities can be useful for both staff and the child. A diary dictated by the young person to a member of staff about visits, news events, family occasions, television programs, etc., will prove useful when the child's speech eventually deteriorates. The diaries can be used to prompt the young person with a key word or phrase and then s/he can recall recent events without further prompts. Ideally such a diary should travel around with the young person at school and go home with them so that parents can contribute. This can help the young person with NPC to join in discussions about out-of-school activities, with their support teacher using the diary to give the minimum of prompting to enable the student to recall. As a speech deteriorates, teachers and parents can pick out key words in what the young person is trying to say and refer to the diary so that they can understand the significance of what is being communicated.

The use of camera and video to record the young person's work in progress, special visits, friends, family and staff can also prompt discussion with the young person, who may be able to recall from memory specific events of life. The earlier the visual, written and taped recordings of a young person's life begins, the more effectively communications can be maintained with them. Photographs will remind everyone that throughout the various stages of NPC, life remains positive, enjoyable, and meaningful.

There is no need for every aspect of the school curriculum to be changed immediately. Physical education may be an enjoyable subject and a young person should continue accessing the gym, working at their own level. As mobility and movement begin to stiffen and coordination becomes jerky, physical therapy exercises may be incorporated into the physical education program but carried out in the gym as part of general exercising. Cookery can continue, but the student may prefer to remain seated throughout the session and support should be given, i.e., to locate equipment and assist with utensils, etc., if required. Mathematics may become more practically based for daily living skills, e.g., working with money, weighing and measuring.

Resources should be age appropriate regardless of the level of functioning. Observing 14-year olds threading beads or using inset form boards only compounds their handicaps. The use of age appropriate resources implies respect for a person's years and membership of their peer group.

When fine motor coordination deteriorates, handling small objects requiring a firm pincer grip may be frustrating. The young person still needs to be encouraged to use both of their hands. Art is an excellent

subject to motivate and enable the person to experience achievement by using both hands, e.g., using bold materials and working in 3D.

Speech therapists, physical therapists, and occupational therapists will all have a role in providing support to the child, the family and professionals. All of the above therapists should be involved with the family in the early stages of the diagnosis, so that parents and teachers can learn specific strategies and access equipment designed to maintain the child's independence for as long as possible.

Speech Therapy

When speech begins to slur, the therapist can provide a program of lip, tongue, and jaw exercises aimed at reminding the young person to remain aware of how to make sounds.

In group situations, the young person may not be heard clearly or the speed of conversation moves too quickly for them to respond. Small group sessions for oral communications may help the young person to relax by knowing that they do not have to compete within a large group. In a smaller group it will be easier for the listeners to hear and understand what is being said. Staff will need to give the young person time to respond; they must never feel "rushed."

Speech will eventually be lost. It is important to realize that the loss of speech can cause great suffering. Speech is very important in many ways. We are social animals and contact of minds is essential.

- We need to share thoughts, ideas and feelings.
- We need to question and inquire.
- We need to communicate everyday trivia and to give instructions to each other.
- We need to communicate to others the kind of person we are.

Young people with NPC will eventually have difficulty with most of these. Although speech will eventually be lost, it is important to try to preserve as much of a student's remaining speech for as long as possible. Specialist help with music therapy can provide an excellent opportunity to encourage speech, as well as an opportunity for creativity in making music. Young people with NPC can often remember and sing the words to songs they learned in the past. Thus singing is particularly enjoyable for them while, at the same time, encouraging them to use speech.

Many of the muscles involved in speech are the same as those involved in swallowing, so preserving speech is important for physical health as well as intellectual health. Eating and swallowing will become difficult and the speech therapist will advise parents and professionals about appropriate exercises.

Physical Therapy

The physical therapist can offer advice and programs designed to maintain suppleness in joints and muscles by stretching exercises. When mobility deteriorates, the young person will rely more and more on a wheelchair. It is important that the young person does not spend unnecessarily long periods of time in a wheelchair. Always give them the option of walking wherever possible, even if it is just out of the classroom and into the corridor to a wheelchair. The young person should not sit in the wheelchair when being taught but sit alongside peers on ordinary chairs so that they do not feel physically and socially isolated.

Occupational Therapy

The occupational therapist will advise on seating, special utensils and resources to help with personal care in eating, toileting and bathing.

Changes in Educational Placement

A time comes when a young person with NPC begins to feel under pressure by constant contact with a mainstream peer group who are all progressing. They will, from time to time, remember their earlier childhood when they were able bodied and experienced success through achievement and progress. A combination of the above experiences together with increased epileptic seizures, mobility problems, and increased support required for their physical and personal needs, may result in the young person transferring to a special school or college which can accommodate any additional needs. Transfer to another educational establishment should take place while in the middle stage of NPC so that the young person can re-establish themselves with new peers and staff.

Transferring a young person in the final stages of NPC will add to their feelings of isolation, because their short-term memory will have deteriorated to the extent that the person will be unable to build up new relationships. The staff coming into contact with a young person in the final stages of NPC will not have had the experience of knowing that person when they were able bodied. It may be difficult for staff in this situation to understand that the young person they are working with has a reference point of “normality” in their long-term memory.

It is important that documents, diaries, and videos go with the young person to give as smooth a transfer as possible.

Respite as Part of the Timetable

There will be periods when a young person’s behavior appears to be irrational and unpredictable. These periods may appear isolated events or become more frequent.

Temporary changes in behavior can be triggered by a person’s own awareness of their loss of independence or by an inability to find the right words to express their feelings. The student may forget what happened an hour earlier but remember in detail events of previous years. Recalling events from long-term memory may become extremely significant.

A student who requires food cut into small pieces to help swallowing may, at times, resent staff doing this and want to be totally independent in eating a meal. The student may be remembering an earlier childhood when they ate independently and feel indignant about people offering assistance. Staff will need to be sensitive and not confront the student with their present disabilities.

The discrimination between background noise and conversation directed personally to the young person may become confusing. Noisy environments will add to the students’ difficulty if they begin to lose the discrimination between background noise and direct interaction. Staff may need to assess specific environments that may trigger a student’s confusion and modify them with calmer, controlled environments, e.g., not being in contact with certain peers or spending time with a smaller group of peers/staff.

In contrast, a young person with NPC, whose long-term memory is rooted in the mainstream of life, may resent being placed with a group of students who function at a similar level, but who were born with severe learning difficulties. Inappropriate noises and challenging behavior will distress the student as their own long-term memory reminds them of social norms.

There may be times when a student is aware of losing control and this manifests itself in feelings of real fear. The student may appear to panic and seek reassurance from parents or staff whom they know well. The feelings of fear may last for a few moments or even several days.

A young person may suddenly resent being supported when walking and insist on walking unaided. The person will be remembering when they walked independently and may be annoyed by the constant contact of staff offering assistance. The provision of a “safe” environment, enabling a student to continue to experience the dignity of independent movement, and the sensitivity of the staff encouraging them to walk unaided helps alleviate the problem, e.g., when in a swimming pool, water at waist level will often provide enough support to enable the young person to safely walk and play independently.

These are a few examples of behavior that can be interpreted as irrational by the onlooker, but perceived as rational to the person with NPC.

Parent/Teacher Support

From the onset of NPC, teachers already involved with the young person will need to share information, concerns and decision-making with the parents. NPC is an extremely rare condition, and it is likely that both teachers and parents will learn together about the progression of the disease and the day-to-day management of supporting the young person. Any change in resources will need to be discussed with the parents to ensure that the timing of introducing adaptations, either in the curriculum or in resources, reflects a comprehensive assessment. The use of a home/school diary, as previously mentioned, is another way of establishing and maintaining parent/teacher communications.

There are support organizations throughout the world dealing with Niemann-Pick disease including the [National Niemann-Pick Disease Foundation](#) in the U.S., and [others outside of the U.S.](#) Many of these organizations were established and are managed by parents for the benefit of families and professionals caring for young people with NPD.

Staff Support

Teachers involved with young people suffering from NPC will need to develop, within their own establishments, a support system to help each other deal with the emotional feelings often experienced when working with people who have a terminal condition.

Staff support may be informal and spontaneous or it could be structured; for example, a regular time set aside for staff to meet and share experiences and concerns.

Conclusion

The progression of NPC will affect each young person differently. Medical information will help those caring for young people with NPC to understand the progression of the disease. However, the individual's

motivation, interests, happiness, and self-esteem will need to be encompassed in an education which MUST continue to be positive and meaningful for the student in order to ensure their quality of life.

After the initial onset and diagnosis, deterioration continues over several years bringing with it changes to the student's level of functioning which will require constant and sensitive changes in teaching.

Towards the start of the final stages, deterioration accelerates. Speech and walking skills will be lost and cognitive skills at a primary level deteriorate. The student may become confused about where they are and who they are with. They may have problems maintaining the "here and now" and events in their long-term memory may dominate their thoughts. The students will need sensitive and positive responses by staff.

In the final stages of NPC, education will begin to move to the periphery of a student's life and nursing care will be a priority. However, the knowledge which teachers and support staff have acquired over the years about what motivates the student will help with the development of an environment which can continue to offer basic stimulation to the student.

The knowledge that young people with NPC may retain an awareness of their immediate environment and long-term memory into the final stages of the disease should ensure that all caretakers continue to communicate with them with respect and a positive attitude.

***Note:** This article was written in the United Kingdom and some specifics may not apply to schools in the United States. However, the general concepts apply no matter where one goes to school.*

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