



UNIVERSITY OF  
STIRLING



# Dementia in children, teenagers and young adults

## *A guide for parents, teachers and care professionals*

Jackie Imrie, Clinical Nurse Specialist, Niemann-Pick Diseases

Elizabeth Jacklin, Clinical Research Nurse, Niemann-Pick Diseases

Toni Mathieson, Development Manager, NPDG (UK)



The Dementia Services  
Development Centre



US Edition, 2009.

The National Niemann-Pick Disease Foundation would like to thank the authors for permitting the modification of their booklet for use in the United States.

## Introduction

Dementia is something that is usually associated with older people. However, children, teenagers and young adults can experience dementia as a result of a number of rare diseases and conditions. Niemann-Pick Disease Type C (NPC) is an example of one of these diseases, and one of its main symptoms is dementia<sup>1</sup>.

Dementia is seen as the loss of mental abilities such as thinking, remembering and reasoning. In most cases, the symptoms develop gradually, often over a period of several years. The early signs, usually memory problems, are normally subtle and may not be immediately obvious.

### Could this be dementia?

**Learning difficulties**

**Unusual behavior**

**Poor concentration**

**Anger**

**Mood swings**

**Confusion**

**Memory loss**

**Asking repetitive questions**

The symptoms of dementia can vary greatly, but they often include one or more of the following:

#### Problems with memory

Normally, recent memories are affected first, with memories of more distant events only affected once the dementia worsens. In children this is often seen as a slowing of learning and an inability to remember things they are taught. If their toys are put away in cupboards, they may not remember where they are. Older children may lose their belongings or, when in school, forget which class to go to next. As the dementia worsens, memory loss may affect memories of recent events so completely that the person appears to be living in the past. The person may forget the names of people they know well, or forget where they live.

<sup>1</sup>Standardized neurological testing demonstrated that most (twelve out of fourteen) patients with NPC disease met the criteria for a diagnosis of dementia at entry into a therapeutic trial (Hinton et al, 2005)

## Problems with speech and language

The person may forget simple words or use the wrong words without noticing. They may repeat what you say (echolalia) and their speech may become slow or slurred and difficult to understand. Their conversation may become simplified, repetitive or irrelevant.

## Confusion

The person may become confused in new surroundings or by new people. They may get puzzled about who or where they are. Children and young adults with NPC may not like change. They may lose track of time and be unsure about what day it is, or even whether it is morning or afternoon. They may not remember that they have just eaten a meal.

## Changes in mood or behavior

The person may become irritable or aggressive. They may laugh inappropriately at something sad. As the dementia worsens they may lose their normal inhibitions and begin to say or do inappropriate or antisocial things. In some cases this can include inappropriate sexual behavior and loss of inhibitions which can put them at risk. They may lose interest in the outside world or their own care, paying little attention to personal hygiene and forgetting to wash or change their clothes.

## Difficulty performing simple tasks

The person may have difficulty doing everyday tasks such as dressing. They may begin a task and then wander away, forgetting what they were doing.

## Problems learning new information, ideas or skills

Although it would be wrong to say that they cannot learn new things, it can become difficult and frequent repetition will be needed.

Some types of dementia can cause less common symptoms including:

- hallucinations
- obsessive or repetitive behavior
- the belief that the person has done or experienced things that never happened (confabulation)
- disturbed sleep, or sleeping in the daytime and being awake at night
- depression



- physical deterioration including loss of appetite, loss of weight and increased vulnerability to infection
- incontinence (loss of bowel or bladder control)

If the dementia becomes severe, it can cause other symptoms including:

- difficulty swallowing
- difficulty changing position or moving from place to place without assistance
- complete loss of short-term and long-term memory

Sometimes, those with dementia are unaware that they have any symptoms, especially symptoms that affect behavior.

## Loss of short-term memory

You may find that a child with dementia begins to lose sense of time quite quickly. They may find it hard to judge how much time has passed because they can't remember what they have done or what they are going to do that day.

Try to keep to a regular routine. Tactful reminders of what day and time it is, and what you are going to do next, may help. Make simple activity lists and use labels such as "Ann's coat" and signs such as "Do you have your money?" where they can see them before leaving home. At school, symbols representing particular lessons can be stuck on timetables and books.

Losing one's memories means that the person loses day-to-day connections with some people around them, along with their links to the recent past. The distant past may be mistaken for the present.

It can be puzzling for parents when their child can remember what they did and who they knew years ago, but can't remember the previous day's events. Using a diary with pictures can be a useful way of going over events that have taken place recently. It is difficult for the child to remember what they have done in school and to tell their teachers what has happened at home. Parents can feel out of touch and will need to establish more formal methods of communication with teachers. A home/school diary which can be filled in on a daily basis can be helpful. Again, pictures are very beneficial and an inexpensive digital camera can be usefully employed between home and school.

Loss of short-term memory can lead to confusion and conflict, for instance if the person misplaces objects and blames others. They may also be unable to make sense of some situations.

**"Short-term memory loss is very upsetting. We find ourselves making plans for future events thinking we have given her something to look forward to, only to find that she doesn't remember."**

## Managing short-term memory loss

- Leave everyday objects in familiar, consistent places where they can easily be seen.
- Keep the environment tidy and calm.
- Reinforce a concept by repeating the same ideas in different ways.
- Label all possessions with the child's name; they may not recognize new things as belonging to them.
- A watch with an alarm reminder can be helpful, as can a small, simple digital voice recorder.

Teachers and caregivers who don't understand or recognize the child's short-term memory loss often feel the child is being naughty and just refusing to do tasks. This may lead to the child being reprimanded and then becoming withdrawn.

**“Our son R would get As and Bs [good grades] up until the middle of the 5th grade then it was a fast decline. The teacher would work so hard with R on one maths problem all day and the next day R wouldn't have a clue what she was talking about. The next year R was placed in a special class. He was given work that we knew he could do and he was still given As and Bs. R loved school, he loved his teachers, but when the dementia first showed up we did think he was playing around until R got mad at himself and said ‘What's wrong with me?’ Music did not seem to be a problem. R lost most of his short-term memory.” (NPC)**

## Confusion

**“A was diagnosed at the age of six. One thing I remember was that before the dementia kicked in we could give him laundry or groceries to put away for us. One day on my return from the grocery I asked him to bring a 12-pack of toilet tissue into my room (I would put the rolls away later). When I later went into my room, I couldn't find the pack anywhere! I wondered, how can you hide a 12-pack of toilet tissue? I searched in my bathroom, under my bed, in my wardrobe, yet no luck. I asked A and he couldn't remember. Finally, I found it in his wardrobe. He was obviously confused when given my instructions. I remember once asking him to put away a pair of his socks. He walked into his bedroom, and just stared at the furniture, not sure what he was supposed to do with the socks in his hand.”**



## The need for routine

“For our youngest child, A, the only clear recollection I have of her dementia is when we moved the living room furniture around for Christmas one year. In order to plug in the tree lights, we needed our end table at the opposite end of the room, near a socket. Inside this table was her nebuliser for her daily breathing treatments (she also had cystic fibrosis). Each day, she would go and get her nebuliser cup for either me or my husband to fill with her medicine. After the furniture was moved, she repeatedly went to where this table used to be, even after we repeatedly showed her its new place. As soon as our tree came down, we moved the table back to its original location, yet by this point she no longer understood our instruction to get her cup. I, of course, regret having moved the end table, but our children loved the tree lights. It was a no-win situation.”

## Unusual behavior

If you are caring for a child with dementia, you may sometimes find their behavior confusing, irritating or difficult to manage. This can leave you feeling stressed, irritable or helpless. By learning to understand the meaning behind their actions, you will find it easier to stay calm and deal effectively with the challenges that arise. Try to remember that your child is not being deliberately difficult. Ask yourself whether their behavior is really a problem, or whether you can redirect or ignore it.

“When B was 13 years old, she started pinching everybody – her assistants at school, her teachers, other kids, mom and her home caregiver, Izabela. Sometimes it would really hurt, especially when her nails would need cutting. Well, one day, her home caregiver Izabela and I (mom) took her to a doctor for a check up and she kept pointing to Izabela and said with a big frown on her face – ‘I don’t like her – she pinches me all the time.’ Izabela was so embarrassed and shocked that B should accuse her of that, she exclaimed ‘Oh my gosh – I never pinch you,’ and then all three of us could not hide our laughter. A few minutes before that happened she was giving Izabela lots of kisses and hugs. The doctor explained that sometimes the way they see things can be reversed. While B was actually the one doing the pinching, she envisioned it was Izabela or others pinching her.”



## What do we mean by “unusual behavior?”

- repetitive questioning, actions or movements
- trailing the caregiver around the house
- hiding and losing things
- shouting and screaming
- restlessness
- night-time waking



“There was one night she woke up screaming at the top of her lungs. I’d never heard her scream before. Both her caregiver and I ran into her room to see what had happened. It was the middle of the night. She was pointing to the poster on the wall of two kittens that were so cute. It had been given to her by some teachers in her prior school as she seemed to like looking at that picture on the school wall. She just kept saying ‘They’re laughing at me.’ I said, ‘Who’s laughing at you?’ She pointed to the two kittens on the poster. I asked her if she would like me to take it down. Would that make her okay? She said ‘Yes.’ Then she climbed back into bed as if nothing had happened and smiled before falling back to sleep.”

## How to cope

- Put yourself in your child’s situation. Imagine how they might be feeling and what they might be trying to express.
- Think ahead and plan for situations that could cause problematic behavior.
- Explain the situation to friends and neighbors so they will understand changes in behavior.
- Offer reassurance by touching and holding.
- Distract them with calming activities such as playing their favorite music.
- Make sure you have support for yourself and breaks when you need them.

If you find your child’s behavior really difficult to manage, ask for advice from professionals or other caregivers before you become too stressed.

## Loss of inhibitions

A child with dementia may behave in a way that other people find embarrassing because of their memory loss and general confusion. Try to react calmly.

- If they undress or appear naked in public, they may have forgotten when and where it is appropriate to remove their clothes. Take them somewhere private and check whether they are too hot or are uncomfortable, or whether they want to use the toilet.
- Lifting a skirt or fiddling with flies may be a sign that the child wants to use the toilet.
- If they start to stroke their genitals in public, discourage them tactfully and try to distract their attention. If this behavior is frequent or persistent, consult your primary care provider.
- If they behave rudely – for example, by shouting, swearing or spitting – don't attempt to argue or correct them. Just try to distract them and explain to other people later that their behavior is due to dementia and is not directed at them personally.
- When out and about in the community, it might help to carry a small card with the words, "This person has a medical condition which may cause them to be confused or act inappropriately at times. Please try to be kind!" This can be handed discreetly to strangers when the need arises.

**“L can be indiscreet. She will sometimes enter a room with her trousers undone after going to the toilet ... or on occasions walk about forgetting she has not got a top or bra on.”**

## Ways of helping and strategies for coping

### At home

Try to establish a regular routine. This will help your child feel more secure and make it easier for them to remember what happens during the course of the day.

- Help the child by using visual clues such as labels on doors, cupboards and drawers.
- Try to structure the day so that where possible activities usually happen in the same order.
- If speech is a problem, make use of body language. Simplify sentences and instructions, listen carefully and give plenty of time for the child to respond.
- Leave things that they need in the same place, so they can easily be found.
- Help your child to continue doing things for themselves by using frequent reminders and doing things “with” them rather than “for” them.
- Memory aids can be useful, such as a photo album, diary or electronic devices.
- Give plenty of encouragement. Be patient, and allow your child to do things at their own pace.
- Try to avoid confrontation. Distract, don't argue.

It is often difficult for parents not to try to correct seemingly “bad” behavior. When a child has dementia due to a life-limiting condition, they are unlikely to benefit from a scolding. They simply won't remember and you will be left feeling guilty. It is far better to avoid confrontation if you can, and find creative ways of distracting them that work for you and your child.

Children with dementia may remember things that happened some time ago, but not what happened just a few minutes earlier. Offer simple choices and phrase questions so that they only need a “yes” or “no” answer. Safe, simple activities that involve repetition can be beneficial and fun, such as singing favorite songs, looking at family photographs or reading their favorite book. Start these activities early, while the child can participate and learn. Don’t underestimate the power of laughter and fun!

## At school

Teaching staff should be kept well informed about your child’s needs. The child’s symptoms should be explained to teachers so they appreciate that behavior associated with dementia is different from bad or disobedient behavior.

## Out and about

- Meeting other children and getting out and about will enable the child with dementia to maintain social skills longer. It can also help to counteract the apathy and withdrawal so common in dementia. However, remember that the child will need plenty of individual attention at social gatherings or on outings.
- Do not negotiate an outing or ask, “Are you ready to go out?” Instead, use statements such as, “Here’s your coat,” “Let’s go,” and “We’re getting into the car now.”
- Reassure the child.
- Plan your route as carefully as possible, paying attention to parking, toilets, elevators and other details.
- Leave plenty of time so you do not have to rush.
- Bring items, such as snacks or books, to occupy the child if you need to wait somewhere.
- Encourage the child to take pride in their appearance so that they feel more confident. Helping them to get dressed up before they go out or before visitors come can make it more of an occasion.
- Look for activities that are stimulating but that don’t involve too many challenges or choices. Children with dementia can find it difficult to process too many options.
- It is important to maintain a sense of humor, so look for activities that you will both find entertaining. Having a good laugh will do you both good!
- Dementia often affects a child’s concentration, meaning they can’t focus on what they are doing for very long. They may need to do activities in short bursts.

## Further reading and reference

Mace NL and Rabins Peter, *The 36-Hour Day: A Family Guide to Caring for Persons with Alzheimer Disease, Related Dementing Illnesses, and Memory Loss in Later Life*, Wellness Central (2006)

*The cognitive phenotypes of Niemann-Pick type C disease: neuropsychological characteristics of patients at baseline in a clinical trial with oral miglustat* (poster), Veronica Hinton, Darleen Vecchio, Helena Prady, Ed Wraith and Marc Patterson (2005)

# Useful contacts

<p><b>National Niemann-Pick Disease Foundation, Inc.</b> Nadine M. Hill, Director of Family Services 401 Madison Avenue, Suite B Post Office Box 49 Fort Atkinson, WI 53538 Toll Free: (877) 287-3672 Phone: (920) 563-0930 Fax: (920) 563-0931 E-mail: <a href="mailto:nhill@nnpdf.org">nhill@nnpdf.org</a> Web Site: <a href="http://www.nnpdf.org">www.nnpdf.org</a></p>	<p><b>NNPDF Coordinator of Education, Referral and Advocacy</b> Cate Walsh Vockley, MS, CGC Medical Genetics Children's Hospital of Pittsburgh Children's Hospital Drive 45th Street and Penn Avenue Pittsburgh, PA 15201 Phone: (412) 692-7349 Fax: (412) 692-7073 E-mail: <a href="mailto:catherine.walshvockley@chp.edu">catherine.walshvockley@chp.edu</a></p>	<p><b>Canadian Chapter – National Niemann-Pick Disease Foundation</b> Nadine M. Hill, Director of Family Services 401 Madison Avenue, Suite B Post Office Box 49 Fort Atkinson, WI 53538 Toll Free: (877) 287-3672 Phone: (920) 563-0930 Fax: (920) 563-0931 E-mail: <a href="mailto:tammy@nnpdf.ca">tammy@nnpdf.ca</a> Web Site: <a href="http://www.nnpdf.org">www.nnpdf.org</a></p>
<p><b>Niemann-Pick Disease, Type C - Medical</b> Marc Patterson, MD Mayo Clinic, Department of Neurology 200 1st Street SW Rochester, MN 55905 Phone: (507) 284-9974 E-mail: <a href="mailto:patterson.marc@mayo.edu">patterson.marc@mayo.edu</a></p>	<p><b>NPC Natural History Study</b> Director: Forbes "Denny" Porter, MD Phone: (301) 435-4432 E-mail: <a href="mailto:fdporter@mail.nih.gov">fdporter@mail.nih.gov</a> Coordinator: Nicole Yanjanin, RN Phone: (301) 594-1765 E-mail: <a href="mailto:nyanjanin@mail.nih.gov">nyanjanin@mail.nih.gov</a></p>	<p><b>Genetic Alliance, Inc.</b> 4301 Connecticut Avenue, NW Suite 404 Washington, DC 20008-2369 Phone: (202) 966-5557 Fax: (202) 966-8553 E-mail: <a href="mailto:info@geneticalliance.org">info@geneticalliance.org</a> Web Site: <a href="http://www.geneticalliance.org">www.geneticalliance.org</a></p>
<p><b>Batten Disease Support and Research Association</b> 166 Humphries Dr. Reynoldsburg, OH 43068 Toll Free: (800) 448-4570 E-mail: <a href="mailto:bdstra1@bdstra.org">bdstra1@bdstra.org</a> Web Site: <a href="http://www.bdstra.org">www.bdstra.org</a></p>	<p><b>National Gaucher Foundation</b> 2227 Idlewood Road, Suite 12 Tucker, GA 30084 Toll Free: (800) 504-3189 Fax: (770) 934-2911 E-mail: <a href="mailto:rhonda@gaucherdisease.org">rhonda@gaucherdisease.org</a> Web Site: <a href="http://www.gaucherdisease.org">www.gaucherdisease.org</a></p>	<p><b>National MPS Society</b> P.O. Box 14686 Durham, NC 27709-4686 Toll Free: (877) MPS-1001 Phone: (919) 806-0101 Fax: (919) 806-2055 Web Site: <a href="http://www.mpssociety.org">www.mpssociety.org</a></p>
<p><b>National Organization for Rare Disorders</b> 55 Kenosia Avenue PO Box 1968 Danbury, CT 06813-1968 Toll Free: (800) 999-6673 (voicemail only) Phone (203) 744-0100 TDD: (203) 797-9590 Fax: (203) 798-2291 Web Site: <a href="http://www.rarediseases.org">www.rarediseases.org</a></p>	<p><b>National Tay-Sachs &amp; Allied Diseases Assoc., Inc.</b> 2001 Beacon Street, Suite 204 Boston, MA 02135 Toll Free: (800) 906-8723 Phone: (617) 277-4463 E-mail: <a href="mailto:info@ntsad.org">info@ntsad.org</a> Web Site: <a href="http://www.ntsad.org">www.ntsad.org</a></p>	<p><b>Alzheimer's Association</b> 24/7 Helpline Toll Free: (800) 272-3900 TDD: (866) 403-3073 E-mail: <a href="mailto:info@alz.org">info@alz.org</a> Web site: <a href="http://www.alz.org">http://www.alz.org</a> <b>National office</b> 919 N. Michigan Ave., Suite 1100 Chicago, IL 60601-7633 Phone: (312) 335-8700 TDD: (312) 335-5886 Fax: (866) 699-1246</p>
<p><b>DASN International</b> Dementia Advocacy and Support Network Web Site: <a href="http://www.dasninternational.org/">http://www.dasninternational.org/</a></p>	<p><b>Alzheimer's Foundation of America (AFA)</b> 1250 24th Street, NW, Suite 300 Washington, DC 20037 Toll Free: (866) 232-8484 Phone: (202) 466-0590 E-mail: <a href="mailto:speschin@alzfdn.org">speschin@alzfdn.org</a> Web Site: <a href="http://www.alzfdn.org">http://www.alzfdn.org</a></p>	<p><b>FamilyPracticeNotebook.com</b> Dementia resources Web Site: <a href="http://www.fpnotebook.com/Neuro/Cognitive/DmmtRsrcs.htm">http://www.fpnotebook.com/Neuro/Cognitive/DmmtRsrcs.htm</a></p>

## Acknowledgment

The authors would like to thank the parents and families who contributed to this booklet



# **Dementia in children, teenagers and young adults**

## *A guide for parents, teachers and care professionals*

Jackie Imrie, Clinical Nurse Specialist, Niemann-Pick Diseases

Elizabeth Jacklin, Clinical Research Nurse, Niemann-Pick Diseases

Toni Mathieson, Development Manager, NPDG (UK)

Published by: The Dementia Services  
Development Centre, Iris Murdoch Building,  
University of Stirling, Stirling, FK9 4LA

t +44 (0)1786 467740  
f +44 (0)1786 466846  
e [dementia@stir.ac.uk](mailto:dementia@stir.ac.uk)  
w [www.dementia.stir.ac.uk](http://www.dementia.stir.ac.uk)

The University of Stirling is a charity registered in Scotland, number SC 011159

