

Respiratory (chest) problems

Chest infections are common in those affected by NP-C, however they usually respond to antibiotics. Sometimes more than one course of treatment may be needed before recovery. Additional problems can be caused by the aspiration of food – this is when food, drinks or even saliva ‘goes down the wrong way’ leading to an episode of coughing and spluttering. This can cause an inflammatory response and/or an infection which may lead to a chest infection or pneumonia. Repeated aspirations, however small, can result in long-term damage to the lungs. As the disease progresses, the person’s cough reflex may become weakened, so that when aspiration occurs they can struggle to clear the lungs properly, creating further problems.

What may happen

In the latter stages of the disease, the person may experience repeated chest infections, bronchitis and pneumonia. These infections often need treating with antibiotics and oxygen, and therefore the person may require a hospital stay. If these infections recur then it is very important to discuss the possible treatment options with your healthcare team. See the End-of-life choices booklet for further information and advice about emergency care plans.

What you can do

Most parents and carers become adept at recognising the signs of a pending chest infection and GPs may prescribe a dry powder antibiotic to prepare at home should an infection flare up. You will still need to inform your healthcare team, but treatment can often start sooner.

Your physiotherapist can suggest exercise techniques which assist breathing and these may help to maximise lung capacity. Posture is also important, for instance, sitting in a well supported semi-reclined position will give the chest freedom to expand and



contract. If breathing becomes laboured, try leaning the person slightly forward, again making sure they are well supported. At night, sleeping in a semi-reclined position may be more comfortable; your occupational therapist can advise regarding the use of mattress elevators or adjustable beds. Keeping the room well ventilated can also help, as long as the temperature is comfortable. You may also want to consider use of a humidifier, or a vaporiser.

You can also monitor the person's ability to swallow and eat, and tell your healthcare team if you have any concerns. As the disease progresses you may need to mash or puree food, thicken liquids, or consider tube feeding. Each of these options can be discussed with the NPDG (UK) clinical nurse specialist or your local healthcare team.

How we managed ... one family's story

As the disease progressed, our son suffered more chest infections, twice having to go into hospital for intravenous antibiotics. The first time I heard the word pneumonia I was scared. However, it was treatable and we went home with a nebuliser and a set of exercises – 'chest percussion' – to help keep his chest clear.

Who can help?

Your consultant, GP, community nurse or the NPDG (UK) clinical nurse specialist will be able to provide advice about medication. A speech and language therapist can help you to assess swallowing, and a physiotherapist can show you a number of exercises that will help to keep the chest clear.

This leaflet forms part of a resource pack published by the Niemann-Pick Disease Group (UK). It is intended to be read in conjunction with the other parts of the pack. If you do not have the other documents in the pack or you would like further information, please contact us at the email address below.

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