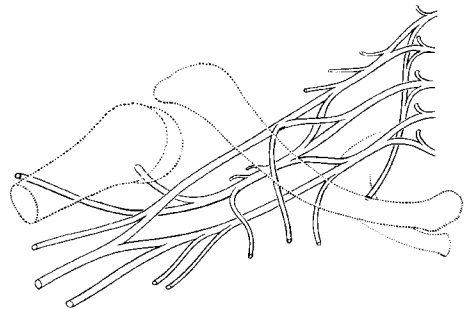


Neuralgic Amyotrophy: idiopathic and hereditary form

UMC St Radboud

Patiënteninformatie



Content

What is Neuralgic Amyotrophy	5
Symptoms	7
Complications	14
Causes	16
Heredity	18
Treatment	21
Personal experiences	27
More information	31
Information for the family doctor and physiotherapist	32
Addresses	

What is Neuralgic Amyotrophy?

Name

The condition that you have is officially termed: '*neuralgic amyotrophy*'. This is also abbreviated as 'NA'. The Latin term literally means: 'lack of muscle growth accompanied by neuralgia.' There are two forms of this condition: the hereditary ('*hereditary neuralgic amyotrophy*' or HNA) and the non-hereditary forms ('*idiopathic neuralgic amyotrophy*' or INA). The latter condition is also called the *Parsonage-Turner syndrome* or *amyotrophic neuralgia of the shoulder*.

Neuralgic amyotrophy is a descriptive term: this tells us what the problem is, but not why or how it is caused. The term also does not specify where in the body the condition occurs. Because, with NA, it is generally the muscles of the shoulder, arm and hand that are affected, the condition used to be termed (and still is by some doctors) differently. For example, '*brachial plexus neuritis*' or '*brachial plexus neuropathy*'. '*Brachial plexus*' refers to the interweaving of the nerves located in the neck at the level of the shoulder blade (see also the picture on the front page of this folder). These nerves are responsible for the 'innervation' of the muscles and the feeling in the shoulders, arms and hands.

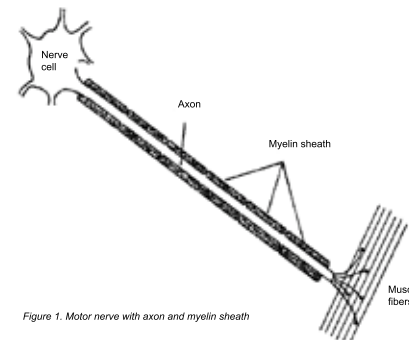


Figure 1. Motor nerve with axon and myelin sheath

Often, it is the muscles and the sensation in shoulders, arms and hands that are affected by this condition. However, in some people, because it also affects the nerves of the vocal chords and diaphragm, the legs or the abdominal muscles, one prefers to have a name that does not exclude these parts of the body. For that reason, we will, from here on out, use the term, '*neuralgic amyotrophy*' or the abbreviation 'NA'.

A nerve disorder

NA is, primarily, a nerve disorder. All complaints and symptoms can be further explained based on this. A nerve fibre can best be compared to an electric wire that conducts electricity. On the inside is the *axon* (copper wire). Surrounding that is an insulating layer (the plastic casing), which we call the *myelin sheath*. In the figure on page 7, there is a drawing of the axon and the myelin sheath. Just as with most electrical wires, the nerves consist of bundles of axons (skeins of copper wire) that are grouped together in a larger casing.

Nerves provide:

- The conductivity of electric stimuli from the brain, via the spinal cord, to the muscles. Thus, the muscles 'know' when and how much they must contract. In medical terms, this is called: '*motor conductivity*'.
- The conductivity of electric stimuli from the skin and the joints, via the spinal cord, back to the brain. This is normally called 'sensation' and it ensures that we are conscious of, for example, whether or not we are being touched or if that touch hurts; how warm or cold something feels and how joints are positioned with respect to each other (whether, for example, we are standing up or sitting down). In medical terms, this is called: '*sensory innervation*'.
- Electrical stimuli from the brain and the spinal cord go to the small blood vessels in the skin and the muscles. Thus, the blood vessels 'know' how much they must open or close. This is very important if, for example, it is very warm: the vessels then open wide and the skin becomes red and warm, so that a great deal of blood can flow through that can cool off 'in the open air'. And conversely: if it is cold, the blood vessels close themselves off in order to prevent chilling. In medical terms, this is called: '*autonomous*' or '*vaso-vegetative innervation*'.

History and familiarity

The first patients with NA were described around 1850 but it was not until around 30 years later that a German physician realised that there was also a hereditary form of the condition. In 1948, the British doctors, Parsonage and Turner, described 136 patients with NA. This description was so striking that the disorder carried their names for a very long time. Nonetheless, NA has remained a largely unknown disease for many physicians. Because of this, this disorder was not always recognized. For the approximately 750 NA patients at the University Medical Center St. Radboud, it took an average of seven months before they were correctly diagnosed. In the first instance, the doctor generally thought it was stress or strain, bursitis in the shoulder or a hernia in the neck. The minimum estimate is that, in the Netherlands, around 500-600 people contract the disease every year. But because the disease is so poorly diagnosed, it is possible that NA occurs much more often than is generally thought. It is estimated that 1 in 10 NA patients have the hereditary form of the disease.

Symptoms

Origin of symptoms

For reasons that are still unknown to us, a number of *axons* 'suddenly' becomes inflamed and breaks down, either partially or completely. The isolation layer surrounding it appears to be unaffected. For comparison: If an electrical wire breaks, little or no electricity can pass through it. The same applies to the nerve with this disorder: It passes on few to no stimuli.

If the nerve becomes damaged, functions can no longer be carried out and symptoms develop in sensation, muscle control and the opening and closing of the small blood vessels in the skin. These symptoms consist of pain, loss of muscle strength, disorders in sensation and sometimes disorders in the blood circulation in the skin.

Affected nerves

The most common is pain and failure of the shoulder, arm and/or hand. Alongside that, the nerves can be affected that go to:

- The upper and lower legs and feet (in around 10% of the patients).
- The muscles of the diaphragm or the vocal cords (in around 5%). In the case of a paralysis of the diaphragm, patients often become extremely short of breath if they lie flat or bend over forwards. Sleep is also often disturbed.
- The skin and muscles of the abdomen (seldom).
- The muscles of the face which causes an acute paralysis of one half of the face (occurs very rarely).
- The ear and organ of balance which causes acute deafness in one ear or extremely severe dizziness as if spinning in circles (occurs extremely rarely).
- One half of the tongue (occurs extremely rarely).

By 'affected' we mean loss of strength in the muscle as well as pain and disorders in sensation or the blood circulation of the skin. These can occur together, but also separately from each other.

Pain

A nerve reacts to damage, first of all, with pain. Nerves pass on their own pain to the brain by means of small fibres. This pain is generally extremely severe, sharp or nagging, penetrating and intense.

When NA patients assign a grade to their pain, in which 0 is no pain and 10 is the most severe pain imaginable, most NA patients grade their pain at 7 or higher in the beginning. The peculiar thing about pain caused by damage to the nerve itself is that someone often feels the pain at a different place than where the nerve is damaged. They feel it, namely, in that part of the body the nerve goes to. Perhaps you recognise this from, for example, the nerve pain caused by a herniated disc in the lower back: the problem is in the back, but the pain is generally in the leg because that is where the pinched nerves go to.

The pain in an NA attack seems to pass through three phases:

Continuous pain

At the beginning of an attack, there is continuous, extremely severe nerve pain that often seems to become even worse in the evening and at night. Most people can then hardly sleep and have to sit up or walk around, holding their arm. Even extremely strong painkillers often hardly help at all. This pain can last anywhere from a couple of hours to several months; the average is three weeks. Then, the

pain generally subsides to a grade of 5 or less. We think that this initial pain is because the nerves are inflamed (see also chapter 6: Causes).

Pain when moving

After the period of continuous pain in the beginning, there is generally a phase in which the shoulder or arm does not hurt anymore when resting but, with specific movements or positions, sudden sharp shooting pains can occur. This pain subsides a bit within a couple of hours. Most likely, this pain is the result of a hypersensitivity of the damaged nerve to stretching or pressure. It is slightly comparable to a nerve in a tooth that is exposed in a cavity: if you touch the tooth, you get a shooting pain in it. Generally, this hypersensitivity of the nerves with NA subsides gradually after a few months. In some people, however, the nerves remain sensitive to stretching and pressure for a very long time. This can be because scar tissue has developed so that the nerves lose some of their elasticity and cannot move as easily in their sheath. If the arm is moved, this pulls more than usual on the bundle of nerve fibres and causes irritation or a shooting pain.

Chronic pain

Many NA patients develop a kind of chronic muscle pain in the area of the (partially) paralyzed muscles and the places where they are attached to the bone or a joint. In particular, muscles that must actually be tensed all day long (for example, the muscles that keep the shoulder blade in place on the torso) are sensitive for this. People also have pain in the non-affected muscles that are overburdened because they must take on all the work of the partially paralyzed muscles. In practice, this often causes pain in the area between the neck and the back of the head, around the shoulder blade and under the armpit on the torso. This chronic, nagging pain is often very uncomfortable and can hardly be treated with medication or rest. What helps the most is finding an adequate balance between activities and rest, with the help of treatment by an occupational therapist and a physical therapist working together. These therapists can provide you with an explanation of what is going on. They can show you in the practice what it is you are doing, and how that is reinforcing the symptoms. By learning to move differently, you can gradually decrease the symptoms. Alongside of that, your physiotherapist will teach you how you can keep the pattern of movement in the affected shoulder

or arm as fluid, supple and normal as possible. Thus, you can make sure that the weakened muscles are not burdened too much or too little and your joints and muscles remain supple and flexible. Your occupational therapist can help you translate these insights into a balance between what the arm and shoulder can sustain and the tasks that must be done in daily life and in work.

The pain with NA is, unfortunately, often difficult to manage with medication alone. For that reason, pain control with this disorder is often a question of trial and error and the combination of various treatments, such as physiotherapy, medication and well-balanced movements. You only know by trying what methods of pain control work for you. For advice concerning pain medication and other therapies, consult your specialist.

Loss of muscle strength

If the damage to the nerve is severe, you will not only have pain but also other symptoms because the normal nerve functions can no longer be carried out sufficiently. The most prominent symptom is generally the loss of strength in the muscles. This can vary from a slight loss of strength which is hardly noticeable to a complete paralysis of specific muscles.

The loss of strength occurs because the stimuli that the nerve generally passes on are now barely – or not at all – passed on at the place where the nerve is damaged. There is nothing wrong with the muscle itself, it simply is not receiving the signal that tells it when and how much it must contract. In medical terms, this is called *denervation (loss of nerves)*: If the muscles are not called upon to contract for awhile, they decrease in size and weight. You notice this as certain muscles get thinner so that the bone is sometimes more visible. In medical terms, this is called *atrophy* of the muscles.

In daily life, people generally do not notice that they are losing muscle strength until they have lost around 30% of their maximum strength. The strength that lies between 70% and the maximum of 100% is, as it were, 'extra' in case heavy exertion must unexpectedly be made.

With neuralgic amyotrophy, the strength in the affected muscles is often decreased to (much) less than 50% of the maximum. It is often not even possible

anymore to carry – or lift – the weight of the arm itself, let alone an extra weight (for example, a purse or bag). It also becomes difficult to maintain movement or postures. It is sometimes possible to make a specific movement once (such as extending the arm or putting something in the cabinet above your head) but it is not possible to do that a number of times or to keep doing it for a specific period of time. Both the loss of strength and the difficulty with maintaining movement are serious impediments for NA patients in their daily work, sports or activities at home.

Disorders in sensation

Moreover, the stimuli that come back from the skin and the joints are also no longer passed on completely. Because of this, areas develop on the skin that feel numb or 'different.' 'Different' can mean that a tingling develops but also that an area of the skin feels extremely unpleasant or painful when touched. It sometimes feels like the affected arm or hand is thick and swollen or cold while, on the outside, there is nothing noticeable. This is because the affected nerves pass on, as it were, the wrong sensation signals to the brain. Something that would ordinarily feel 'normal' is now passed on as 'pain' or 'cold'.

The medical terms for 'numb,' tingling' and unpleasant' feeling when the skin is touched are *hypesthesia*, *paresthesia*, and *dysesthesia*, respectively. The sensation in the joints (*proprioception*) is almost never affected with NA.

Disorders in the skin blood flow

Disorders in the skin's blood flow are generally the less conspicuous symptoms with nerve damage. Very locally – generally in the hands – the skin can become redder, slightly purple or spotted, and skin, hair and nails can start to grow more quickly. The thermostat of the skin is, as it were, no longer completely well adjusted. Certain places on the body can, therefore, no longer react adequately to cold by closing off the capillaries of the skin so that the heat is kept in the body. Thus, the hands and feet, in particular, cool off more than they actually should and you feel this as 'too cold'.

The faster growth of skin, hair and nails and the 'different' feeling of cold and warmth together are, in medical terms, *vasovegetative*, *sympathetic* or *autonomous disorders of the skin*. Generally, with NA, these vegetative disorders are

most severe in the hand and forearm. In the case of NA, they are only in those parts of the body where the nerves to the muscles and skin are also affected.

Attacks

The symptoms with neuralgic amyotrophy occur in attacks. Generally, there is first an extremely severe pain in the shoulder or arm and, a few hours to days later, a number of muscles in that arm become partially or completely paralyzed. The pain lasts, on average, three weeks, but can also be gone in a day or continue for six weeks. The pain is often the most severe at night. In the non-hereditary form, three fourths of the patients have only one attack and in one fourth of the patients, the attacks reoccur once more. In people with the hereditary form ('HNA') the chance of recurrence is greater and three fourths of the patients suffer from recurring attacks.

The place, the severity and the duration of the pain, as well as the paralysis, can differ greatly per patient. In the mildest form, someone can, for example, have a pain in his shoulder and forearm for a few hours and then difficulty with bending the tips of the thumbs and index fingers for a few months. On the other end of the spectrum, the attack can also be extremely severe with pain symptoms that last a long time, a great loss of muscle strength in both arms and difficulty breathing when lying flat, so that – even after five years – recovery is still not complete. There are all types of forms between these two extremes. Your neurologist can determine if you have had a mild or more severe attack. The personal stories of a few patients in this folder do not, therefore, mean that your NA will also look like theirs.

Recovery

The recovery after an NA attack depends upon how severe and extensive the damage to the nerve fibres (*axons*) was during the attack. Most NA patients recover over the course of one to two years to around 70-90% of the level of before the attack. This seems pretty good, but it is, however, no longer the 100% that someone was used to. In practice, this difference often leads to residual symptoms. If there were several attacks in the same arm, then the recovery after each subsequent attack is consistently a bit less.

Your body always tries to repair the damaged nerves, but this goes very slowly. There are two ways in which the affected nerves can repair themselves:

Taking over tasks

If, within a nerve bundle, no more than two thirds of the *axons* are damaged, then the remaining *axons* will take over the work of the damaged cables within three to four months. This mechanism is called: '*collateral reinnervation*'. This repair mechanism works well because the muscle can then again be completely controlled. However, this is at the expense of some *load endurance*. That means that the muscle can certainly provide maximum strength once but cannot maintain it well or keep it up for long. In daily life, patients often notice that, after a time, they can, in principle, carry out all of the movements with the arm, but that the arm becomes heavy and tired after being used for awhile and they must stop and rest before they can continue with what they were doing.

Nerve growth

If, within the nerve bundle, more than two thirds of the *axons* are damaged, the remaining *axons* will no longer be able to take over all of the work of the damaged fibres. After a few months, there is still part of the muscle that is not yet connected to the nerve and which, therefore, has no strength. If this happens, the body still has two ways to repair the nerve whereby, from the tip of the damage, new *axons* in the neural tube grow towards the muscle. This process is called '*proximal reinnervation*' and is extremely slow: less than one centimetre per week. If, for example, the nerve that services the muscles in the thumb is damaged at the level of the armpit, it can take anywhere between eight months and four years before the new nerve fibres reach the thumb muscles. This is why the recovery of the partially paralyzed muscles with NA takes so long. That means that, even after two years, improvements in muscle strength can still occur, especially with the muscles that are the furthest away. Recovery, however, is generally not a complete then.

If the nerve has been affected a number of times, the chance of recovery decreases. Also, nerves are not able to tolerate everything: If they become even more damaged, they will be 'broken' for good. In practice, this means that the function of, for example, the arm after an initial period of painful loss of function will still

recover for the most part, but that, after a second or third time, will not recover as completely and, ultimately, will not be able to recover at all. The loss of strength, but also the disorders in sensation and blood supply to the skin, then become permanent.

There is no known medication that can stimulate the recovery of nerves. As a warning against all types of ads on the internet: vitamin B6, in high doses, can even damage the nerves. In addition, electrical stimulation of the muscle in order to strengthen the muscle mass is advised against because this will likely hinder the regeneration of the nerve by the body. However, with NA there are no objections to the milder electrical currents that are used in the so-called TENS treatment (see chapter 9).

There are other factors that also play a role in how fast and how fully patients can recover. For most of the muscles, one can say that they function well in daily life once they have regained 70% of their former strength. However, some muscles must truly be nearly 100% recovered before they can function normally again. The muscle that, when it fails, causes a protruding shoulder blade, (the serratus anterior) is the most important example of this. Because this muscle needs its maximum strength and endurance in order to work well, it sometimes seems as if recovery of the nerve to this muscle takes much longer than the recovery of the other nerves. That is probably not the case. It simply takes longer before it makes a difference to you.

Complications

In spite of the fact that there is recovery of strength, more than half of NA patients develop persisting pain symptoms. This pain is generally the result of overtaxing both the weakened muscles and the muscles that must compensate for them. Through the pain and the decreased strength, NA patients are often hampered in carrying out such daily activities as household tasks or work. The most important constraints are often in the difficulty with reaching and lifting. However, also non-strenuous repetitive movement (such as typing or ironing) or sitting in one position for a long time can induce and maintain this type of pain symptoms. If NA patients, nonetheless, continue to carry out their normal activities, this often hap-

pens unconsciously by much compensating and overtaxing. This means that the movements are no longer done in a normal way but by using other muscles or by tightening the entire body. This way of moving often costs much more energy than the normal way and leads more quickly to overtaxing and chronic muscular pain. Sometimes, the shoulder moves in a totally different way than normal so that, consequently, new pain symptoms develop.

In the normal situation, most people do not even think about how their shoulder moves. Someone generally only thinks about what he or she wants to do or pick up with the hand. What the body must regulate beforehand with respect to the shoulder in order to be able to use the hand securely and steadily is completely unconscious. Because of this, NA patients often have no idea which muscles are weakened – or they are not even aware of muscle weakness – but only know that the arm is very difficult to move and that movement cannot be maintained for long. Patients are also often not aware that there is a compensating pattern of movement and what the long-term consequences of this can be. They notice the consequences over time because of the (extra) pain, loss of strength and/or fatigue it causes. Lack of information about the consequences and the course of NA can lead to the feeling that the symptoms are ‘unmanageable’.

The shutting down or partial paralysis of a number of muscles in the shoulder, arm and/or hand does not only have consequences for strength. Also, the position of the shoulder, arm, wrist or fingers can change considerably. If the muscles that keep the shoulder blade in place shut down, what often occurs is that, through the weight of the arm, the shoulder shifts a bit downwards and to the side. This then creates problems if the arm is lifted because the space in the shoulder joint, between the ball of the upper arm and the shoulder, is smaller. Thus, the tendons surrounding the shoulder joint can become pinched during certain movements (*secondary impingement*) and that is painful. If the shoulder blade droops, the muscles that are connected to it – starting in the neck and the back – become overstretched. This is also often painful, certainly in the long term. This is especially because muscles cannot take it if they are stretched out and must still contract.

If certain muscles in the shoulder blade are extremely paralyzed, it is even possible that the ball of the upper arm can no longer be held in place well in the shoulder joint. When that is the case, a partial dislocation of the shoulder occurs; in medical terms this is called a *subluxation*. This often causes discomfort or pain when lying on the shoulder, but the arm is not completely dislocated.

Causes

The precise cause of NA is not yet known. It is a combination of a number of factors. The attacks themselves are probably caused by an error in the *immune system*. In NA patients, the immune system sometimes accidentally produces antibodies that not only attack bacteria and viruses from the outside but also cause an inflammation of the nerves. It is, therefore, not true that the immune system in NA does not work well. It actually works too well and sometimes too enthusiastically, so that, by accident, antibodies are produced that attack parts of one's own body, in this case the nerves.

Most likely, the nerves, when constructed in people with NA, are more sensitive for this type of damage. This is even more pronounced in people who have the hereditary form of the disease. They also have more attacks throughout their lives. What constitutes the increased sensitivity is still unknown. A change in the hereditary material alone is not sufficient to cause the attacks. There are events that can elicit the pain and loss of strength:

- Infections (in particular, infections of the nose and throat, for example, a cold or flu).
- Pregnancy and birth (in particular, an attack with pain can occur a few hours to days after a delivery).
- Heavy exertion or stress on the shoulder and arms.
- Stress, both physical and psychological.
- Vaccinations and treatments with blood products or *immunotherapy*.
- Surgery and anaesthesia.
- An accident or injury in the shoulder or arm.
- Change in the weather, generally from warm to cold or wet weather.

However:

- Not every incident causes pain and loss of strength with every patient. This can vary greatly per person.
- Such an event does not elicit pain and loss of strength every time. Very often, one can only say afterwards whether or not an unusual or striking occurrence preceded the symptoms.
- This list is not complete. Perhaps you yourself have noticed symptoms that can also be elicited by other events. If that is the case, would you please let us know what these events are?

The mechanical load of the arm and shoulder also play a role in the chance of having an NA attack. In the group of ± 750 patients with NA who are being treated in the UMC St. Radboud, there are noticeably more athletic people and people who do heavy physical work than in the general population. The belief is that doing heavier work – or more work than average – with the arm, places more stress and strain on the nerves of the plexus. Thus, the barrier between the nerve and bloodstream is less tightly shut and the nerve becomes more vulnerable to damage by the immune system.

Thus, a number of things must go wrong at the same time before there is an NA attack. Unfortunately, no advice can, as yet, be given as to what patients must do or not do in order to decrease (or increase) the chance of such an attack. Moreover, it cannot be predicted whether or not someone will get an attack (or have another one). And even though some attacks occur right after an infection, operation or delivery, there are no known or necessary special measures for such a situation. NA patients are, therefore, advised to get the necessary vaccinations because the chance that you will become ill while travelling is probably much greater than the chance that an NA attack will occur after a vaccination. Moreover, there are no special measures that your midwife or gynaecologist need to take during the delivery. It does not make any difference if you delivery naturally or via a caesarean section and an epidural is not any more or less safe than other forms of pain control. It could be a good idea to let your midwife, gynaecologist or surgeon know that you have NA and that there is a small chance that you could

have an attack after the delivery or operation. They are then well informed and can quickly set up good treatment.

Heredity

The hereditary form of NA occurs in several members of the same family and is passed on from one generation to the next. If there is no one else in your family who has complaints or symptoms that are in keeping with NA, then you can assume that you do not have this hereditary form. This chapter, then, does not apply to you.

Dominant heredity

What *heredity* means is that characteristics of people in one generation are passed on to the following generation. Characteristics can be both physical (for example, the colour of the eyes) or psychological (one's personality). The passing on of characteristics occurs by means of the genetic material in the cells. This genetic material is called 'DNA'. The DNA is rolled up in the cells and packaged in the *chromosomes*. A piece of hereditary material that passes on one characteristic is called a gene (plural: *genes*).

Every child receives, upon conception, two versions of each gene: one from the father and one from the mother. The father and the mother have, in turn, also received two versions from their father and mother.

At present, characteristics, also hereditary characteristics, can be passed on in one of two ways. For one way, you only need the gene from one parent. What the gene of the other parent does then is not important. This type of heredity is called, in medical terms, *dominant*.

For the other way, the two *genes* from each parent are necessary. If they are not both present, the characteristic will not be passed on. This form of heredity is called *recessive*.

HNA is a *dominant* condition. Thus, you only need a piece of hereditary material from one of your parents which has a change in it that other people do not have. This change is likely to be the reason that the nerves have an increased sensitivity to damage.

We know that HNA is *dominant* by the way in which the condition is passed on in families. For example, in no single family that we know of are both parents of a patient affected.

Prognosis for HNA

The fact that this condition is *dominant has consequences for the chance that your children will also get the same condition*. Statistically, that chance is 50%. That means that, *in theory, half of the children in a family will inherit the condition from their parents*. You must remember here that it is *only in theory that it will be exactly half*. Theories about these types of chances are always based on large groups of people. For you and your family, this can, in practice, turn out very differently. Sometimes, none of the children will come down with the condition, sometimes they all will. It is also not the case that if the first child does not have the condition, the following child will have a greater chance of getting it, or vice versa. You can compare the chance of inheriting it with tossing a coin for heads or tails. If you get heads, your child will not have the condition, if it is tails, he or she does have the chance to get the condition. And, for every child, you must, as it were, toss again.

In summary, it is not possible to predict in any way what the chance of getting this condition will be in your personal case.

Physical characteristics

People who suffer from the hereditary form of NA (HNA) sometimes have a striking appearance. These changes in the appearance are, in medical terms, called 'mild *dysmorphic* characteristics'. This includes such physical characteristics as: close, deep-set eyes, a 'Chinese' shape to the eyes, unusual shape of the ear, a narrow or non-symmetrical (both halves not the same) face, a narrow mouth, a cleft palate or uvula and wide-set teeth.

In the rest of the body, the following characteristics may be found at birth: webbed fingers or toes or fused bones in the forearm, hammer toes and small stature.

Not all patients have these outward characteristics. There can also be differences within one family. And there are also descriptions of family members who have these outward characteristics but no HNA.

Gene research

Over the past few years, more scientific insight has been gained into the genetic changes that can lead to HNA. Thus, it is now known that HNA can be caused by a mutation in several places in the DNA. The condition is, in medical terms, '*genetically heterogeneous*.' It has, however, recently been indisputably established that one gene can lead to HNA attacks.

This *gene*, or piece of hereditary material, is called SEPT9 and is located on the long arm of the 17th chromosome (17q25). In this *gene*, doubling or very small changes in the DNA material are found in families with HNA. It is, however, not yet clear precisely how this makes one more sensitive for neuritis. We do know that the typical features and other outward characteristics in some families appear to be caused by one very specific mutation in the SEPT9 *gene*, the so-called 'R88W' mutation.

Mutations in the SEPT9 gene are estimated to be present in approximately half of the HNA patients (or families). Alongside this, there are, therefore, also families who do not have these mutations, thus there must be more genes on other chromosomes that cause HNA when they undergo a mutation.

Because a mutation in the SEPT9 *gene* only occurs in half (and in the Netherlands, probably even fewer) of HNA patients, it is not yet worthwhile to test Dutch patients for this. After all, if no mutation is found, you still do not know for sure whether or not you have HNA. For the time being, the diagnosis is, therefore, primarily dependent upon whether or not there are other family members with NA. There are no other characteristics that can clearly differentiate between the hereditary and non-hereditary forms for the individual patient. Thus, in both forms, repeated attacks can occur and, in both forms, nerves other than those in the arm can also be affected.

Treatment

It is clear that the treatment of NA is not easy. Over the past years, however, more information has become available on which treatment can be worthwhile. That treatment varies per each phase of the condition. We differentiate between three phases:

1. Acute phase

Painkillers

The pain in the acute phase reacts best to a combination of two strong painkillers: a so-called *long-acting NSAID* and a *long-acting opiate*. These drugs must be prescribed by a physician. If your pain score is equal to or greater than 7 on a scale of 10, it is generally pointless to try only paracetamol or drugs such as ibuprofen or naproxen. Most NA patients say that these ordinary painkillers do nothing at all in the acute phase of an attack.

Prednisone and other drugs that influence the immune system

In the first days of an attack, a short therapy course using prednisone has a favourable effect with some NA patients. However, it does not work with everyone; prednisone provides around five out of ten people with fast pain reduction and, in one or two out of ten, with a somewhat faster recovery of strength than you would normally expect. It is, therefore, no miracle drug. Prednisone is a drug that must always be prescribed by a doctor. When prescribing it, one must always pay attention to the *contraindications*; those are the reasons why someone must not use the drug (such as poorly regulated diabetes, a stomach ulcer or depression). There are, as yet, still no data on a possible favourable effect of other drugs that influence the immune system in the case of NA. Therefore, the use of these drugs cannot be recommended.

Advice for your doctor in the acute phase

To combat pain during an NA attack or a relapse, a combination of long-acting morphine (for example, MS Contin 2 dd (twice daily) 10-20 mg) with a long-acting NSAID (for example, Diclofenac 2 dd 100 mg Retard) is advised. If there are no contraindications, a short course of prednisone per os can be considered. We use

60 mg per day for this (with children: 1 mg/kg) for 7 days, in the following week to be reduced by 10 mg/day to nothing. If necessary, laxatives (with morphine use) and stomach protection (with prednisone and NSAID).

Physical therapy

Physical therapy can be useful in the acute phase or to ensure that the affected arm does not stiffen in the joints because you can no longer move it. In medical terms, this is called *contracture prevention*. It is important that the mobility in the joints of the shoulder, elbow, wrist and fingers are maintained and that the muscles do not shorten. Nothing can be done to combat the muscles belonging to the affected nerves becoming thinner. Other forms of physiotherapy, such as manipulation, traction and resistance or weight training are usually not yet an option in this stage and often only cause more pain.

2. The first few months

When the acute, extremely severe pain begins to subside, the nerves in the affected arm(s) often remain hypersensitive to stretching and movement for awhile. You will notice this because you get shooting pains or tingling in the arm if you move it. This pain is the consequence of the hypersensitivity of the affected nerves and does not mean that the nerves are breaking down even further. The pain is often irritating but tolerable for most patients, certainly once they know why it still hurts.

Co-analgesics

If, however, these shooting pains are extremely unpleasant, you can try to treat them with so-called *co-analgesics*. These are drugs that were originally used to combat either epilepsy or depression. They muffle, as it were, the nerve stimuli in the brain, but they also do this in the damaged nerves in your arm or leg. That is why they work with nerve pain in NA. These drugs are, however, no ordinary pain-killers. That means that you cannot and must not take them at the moment that the pain suddenly occurs, but that they must be used continually in order to build up a certain amount of the medication in the blood. In the first weeks, they often have side effects in the form of vertigo or disturbed concentration. This generally passes. They only start working after a number of weeks. You must use them for at least four to six weeks and gradually increase the dosage. If, after six weeks

in the correct dosage, they do not help, it is better to cut back and then to stop. These drugs can also only be obtained with a prescription and in consultation with your physician.

Physical therapy

When the acute pain has subsided somewhat, it is often a good idea to ask for advice and support from a physical therapist. Furthermore, it is important that it is clear to both you and the physical therapist that the pain and loss of muscle strength with NA is not the same as 'ordinary' shoulder pain. In the case of ordinary shoulder pain, the therapy often consists of strengthening the muscles surrounding the shoulder by exercises with elastic bands or weights. This standard treatment, however, does not generally work with NA, or even causes more symptoms. This is because the muscles that are already weakened by the nerve damage are often already being forced to function at their maximum level. Normal movements are, for these weakened muscles, already top-class sports. If they are then forced to train even more, they quickly become overtaxed and that causes a fatigued or numb feeling and subsequently even more muscle pain.

The general rule is that you should not be bothered by a specific training or activity for longer than the activity itself has lasted. Thus, thirty minutes of exercise or household work should not cause a numb feeling or pain for three hours. If this does happen, then the activity or exercise was too much and/or too long and must be adjusted. Talk to your therapist if this is the case.

The physical therapist can explain to you what exactly is wrong with the shoulder movements and help you to keep those movements as normal and smooth as possible. Because the body has the tendency to quickly compensate weakness in the shoulder by moving differently, a completely deviating way of using the shoulder is often – unconsciously – developed. In the long term, this can lead to persistent pain symptoms in the shoulder, neck and the back of the head. Your physical therapist can help you to again normalize such an awkward way of moving and compensating. Furthermore, it is important to know that compensation, in itself, is not a bad thing, but that it can certainly lead to an increase in the symptoms. Your therapy should concentrate chiefly on providing insight into moments of overexertion and how you can reduce these moments. Moreover, patients often tighten all sorts of compensating muscles too much when that is actually not at

all necessary. Learning how to carry out a movement with the correct timing of muscle tightening and relaxing is important (muscle coordination training). Often, too much thought is given to the muscle weakness and the strengthening of the muscles and, in the therapy, one 'forgets' that tightening must be alternated with relaxation.

3. Late phase (residual symptoms)

Factors that maintain the symptoms

Approximately two thirds of the NA patients who were examined at the UMC St. Radboud had, after a few weeks to months, severe pain in the area surrounding the shoulder blade, the neck and the back of the head. This was caused by overexertion of both the weakened and the compensating muscles. Moreover, pain and reduced mobility in the shoulder joint are common. This is caused by the different way of moving the shoulder joint because a number of the muscles that control this joint are reduced in strength. The joint thus becomes – both in rest and during movement – unbalanced. Because of this, the movement between the socket joint of the shoulder (glenoid) and the ball of the shoulder is no longer fluid and the tendons and muscles surrounding the shoulder can become pinched and damaged.

Treatment of the above symptoms is not easy, certainly not if they have been going on for months or are getting worse. Often, NA patients get caught up in a negative spiral in which, for example, the pain increases so that the patient is forced to rest until the pain subsides, and then the patient again takes on all sorts of activities that, once again, lead to pain. The first step in the approach to these on-going symptoms is to determine what factors play a role in the patient's life in maintaining that vicious circle.

Four factors can be differentiated that keep the pain and restrictions going in patients with NA.

Physical factors

First of all, there is the physical factor: strength and sensation in the individual muscles and their recovery ('*hardware*'). What is important here is that the nerve recovery leads to a muscle that can be controlled again, but that this, at the same time, is at the expense of the possibility of the muscle to persevere and keep up a certain movement or contraction. You will notice that you can carry out a certain movement a couple of times, but that the arm then quickly becomes heavy, tired and less strong and that moving it becomes increasingly difficult.

Way of moving

The following factor is the way of moving that the entire body uses to carry out tasks. Shoulder movements demand an extremely complex control (the '*software*') from the brain. This happens, for the most part, unconsciously.

If there is damage to the hardware, the body immediately shifts to another way of steering (a *compensation strategy*) so that the position or movement can still be carried out as well as possible. For example, you might find that in order to reach above shoulder height your body is pulling the shoulder very close and you lean over with your trunk to get the hand up where it needs to be. This compensation strategy, however, costs more energy than the normal way of moving. The movement will, therefore, work at first but often cannot be maintained for long and ultimately leads to overburdening.

The load/capacity ratio

The third factor by which the symptoms are maintained is the ratio between the load and the capacity in daily life and work (or school/training). Because nerve recovery takes so long (several months to a few years), NA patients often want, nonetheless, to get back to their tasks again at home and at work as well as possible. This also depends, of course, on other circumstances, such as caring for a partner or children, or an employer's demands. It is better for the muscles affected by NA to change position often during work and to alternate the weight of the load; also, take frequent short breaks or do something else for a short while ("*mini-breaks*"). In the daily situation of many people, such flexibility in their schedule is often hardly possible – or only with a great deal of adjustment (also from others). A common problem is then that patients, because of these external circumstanc-

es, 'simply keep going' and thus, over the course of months, completely exhaust their bodies – and thus themselves. At first, the pain is often only in the hours after the exertion but it then returns more quickly and continues for longer periods of time, until a situation is reached in which pain and exhaustion become a steady state.

Finally, the manner in which someone deals with his symptoms and limitations also determines whether the symptoms decrease or remain. This manner of dealing with the condition – or, 'coping style', is determined by the personality of the person himself. It has already been determined from a study of NA patients with continual symptoms that, as a group, there was no observation of a deviant personality ("psychopathology") or chronic pain syndrome. In practice, we do see that many NA patients have the tendency, in spite of their symptoms, to still carry on with their normal activities ("if I do my very best, I can get over it") and that it is this attitude that throws them into a vicious circle or downwards spiral of continuous overburdening, which then causes the symptoms that force them to slow down, then struggle back up again and then, once again, get back to work fanatically only to, once again, overburden themselves, and so forth.

Therapy plan

Once someone with NA develops chronic symptoms, they can generally no longer be treated with simple measures or medication. It is, then, often necessary to map out – together with your consulting physicians – all of the symptoms and their contributing factors. Thus, an assessment can be made of what the recovery possibilities are and what must be done to make that happen: physiotherapy, occupational therapy, a rehabilitation trajectory, psychological coaching, consulting with the company doctor or a combination of all of the above. Such an action plan must then lead to concrete advice and steps that the patient can take. This is often done within a rehabilitation trajectory but it can also be examined together with the family doctor or one's own neurologist, depending upon his experience with this condition. In the Netherlands, such rehabilitation for NA is offered by the multidisciplinary team ('the Plexus out-patient clinic') at the Rehabilitation Department of the UMC St. Radboud. In order to register for this, it is necessary to have a referral from your family doctor, neurologist or other specialist.

Personal experiences

A 'luxury pain': Experiences of a woman with NA

I am now 54-years old. Three years ago was the first time that I had the feeling that I had a 'hot spot' in my left shoulder that was initially not so hard to take but gradually started to give increasingly more problems. The pain became so bad that I went to see my family doctor who, because of my years of chronic neck pain, prescribed a pain killer for the umpteenth time and left it at that. I left it at that, as well, because I also thought that the pain was probably coming from my neck. Because of the painkillers, the feeling was suppressed temporarily but it certainly did not disappear.

I continued to feel the pain for the entire following year. I had my normal physiotherapy for my neck and everything went on as usual, but sometimes I got a cramp in my left arm and my fingers, thumb and the palm of my left hand started to tingle. Meanwhile, I simply muddled along.

A year later, after a severe pain attack, I went back to the family doctor who then agreed to have x-rays taken at the hospital and referred me to the neurologist.

I then received the results of the x-rays which were then discussed with me by an alternate neurologist. The x-rays indicated wear and tear of the vertebrae in the neck, but not to such a degree that it could cause that degree of pain. The alternate neurologist acted like this was a 'luxury pain' in these luxurious times.

According to him, people used to get this but they did not complain so much!

One year later, the pain was so intensely white-hot and ice cold that I was hospitalized and even had symptoms of paralysis in my left arm.

In the hospital, I met Doctor Van Alfen from the UMC St Radboud in Nijmegen.

She recognized the condition and prescribed medication; a course of treatment with prednisone and gabapentin, in order to help me get rid of that excruciating pain. I was ultimately released from the hospital with gabapentin, which I am still taking.

It is now May 2001, and, since the attack last September, I had another attack in February of this year. This one was less severe but I felt it coming on and could quickly start a course of treatment again with the medication so that I was saved from experiencing even more misery.

How do I manage this?

I have been given help at home, something I initially had problems with because I'm a 'do-it-yourself' person. If I am in a lot of pain, I can, however, now let things roll off me much more easily. The pain fluctuates, as well. Sometimes, I get up and then I feel that I am going to have an 'off-day.' In that case, I won't be able to do much and will have to rest a lot. But I also have days when I can certainly get things done: hang up the laundry; lift heavy things, such as a full pan, a coffee or teapot or a vase of flowers. Those types of actions have, however, become extremely difficult.

How I care for myself has also changed. Putting on pantyhose, underwear and shoes, washing and combing my hair have all become difficult. I see now that I put on jackets and blouses differently; in short, there are a great many limitations in my life now. Sleeping is also one of them. The pillow is thrown out of the bed and put back at least ten times per night and I can no longer sleep very well on my left side. Sitting is only comfortable if I have sufficient support for my left shoulder; I absolutely cannot tolerate any pressure on my arm or head or a strange position, at all.

How do those around me experience this?

It dominates part of our lives. The care that my husband and children provide for me has become different. They clean up more after themselves and they are continually thinking about how they can make it easier for me. They help where they can, when they have time.

Within the family, they always ask about my health. This is logical and normal but it is also sometimes 'annoying'. There are so many other things, better things, in life.

Future

Sometimes, I worry a great deal about the future. If this continues like this – getting one attack after another – I will have to keep taking medication. If the symptoms of paralysis occur more often and get worse, what then?

As long as it is possible, I want to keep doing everything that I can, everything I am allowed to do and whatever is possible. I try to maintain a positive attitude as much as possible, for myself and for others.

I am not strong, but I am certainly very tenacious!

Hereditary neuralgic amyotrophy (HNA)

Experience of a patient, born in 1964

How HNA started and what precisely caused it is not completely clear to me. What I can say is that, in the four experiences that I have had thus far, the muscles that are affected continue to feel tired for a number of weeks (around two to four), resembling the fatigue after playing intensive sports. This fatigue decreases gradually to a nagging pain that is constantly present.

After two to four weeks, I notice that the pain is abnormal (too constant and too severe) and then I start to suspect that it could be HNA. After around four to six weeks, the pain is so present and severe that normal functioning is barely (or not at all) possible anymore (at work and in private life). I become irritable and drained; I lose interest in everything, until the pain, from one moment to the next, is gone; at least this is how it seems. I think that this is a period of a couple of days in which the pain subsides and the muscle paralysis sets in.

Alongside the muscle paralysis, I am greatly hindered by a numbness of the skin. It is always at the place where the paralysis was. The recovery after a paralysis in the arms takes around a year (90%). The final 10% of recovery takes a number of years and is dependent upon the degree to which the paralysed muscles are used.

How does HNA manifest itself?

In my case, I have had three attacks in one arm and one attack in the area of the vocal cords.

Year:	Where:	Recovery:
1977	Right arm, complete, including numb skin on forearm and hand	one year
1985	Left arm, complete, including numb skin on forearm and hand	one year
2000	Left arm partial, including numb forearm on inside	three months
2000	Both vocal cords (inability to talk for six months) and part of oesophagus	six months.

What now?

One can live with HNA. I do notice every time that the recovery is less complete. I have had two attacks in my left arm; it is also my worst arm. Some arm movements remain difficult. Physical therapy can help one to recover as much as possible after an attack. Perseverance and (sensible) exercise have been, in my

case, extremely important. During the attacks, I have also been very helped by pain control with TENS. I now have a TENS of my own.

HNA in my case

I am a 50-year old man and had my first attack at the age of eight. As far as I can remember, I had no pain, but there was considerable loss of strength in the arms. Starting at the age fifteen, I went to work as a construction fitter. In the period that I worked, I was regularly home for a few months with attacks of pain and loss of strength. At the age of twenty-seven, I ended up in the WAO (Disablement Insurance), 80-100% disabled.

Between the ages of thirty and fifty, I have had a series of attacks in the arms. My last attack was only recently.

Progression of an attack

First of all, there is pain. This scores 8-10 on the pain rating scale. It keeps me awake at night. I am also bothered by muscles shutting down. The entire attack lasts for around six to ten weeks. I do not use any medication, except sometimes two paracetamol of 500 mg. Why not? Because there is actually no medication for HNA. As far as recovery is concerned, with me there is always residual damage manifested as loss of strength.

In conclusion

It is not so difficult for me to live with this. I was able to accept that I have HNA fairly quickly. I believe that it does not help to rebel against it, in spite of all of my limitations.

More information

For additional information, you can go to the website of the UMC St. Radboud, via <http://www.umcn.nl/Zorg/Ziektebeelden/Pages/neuralgischeamyotrofie.aspx> and to the Vereniging Spierziekten Nederland [Society of Muscular Diseases] (VSN) via www.vsn.nl.



Muscular diseases Infoline (Spierziekten infolijn)

At the Muscular diseases infoline of the VSN (Telephone number 0900 548 04 80, € 0,15 per minute or e-mail infolijn@vsnl.nl) you can obtain information on a variety of matters concerning muscular diseases and their consequences. This also includes contact with fellow-sufferers via the VSN.

Information for the family doctor and physiotherapist

This condition is now generally called *neuralgic amyotrophy*, but is also known under the somewhat older name of 'Parsonage-Turner syndrome' and 'plexus neuritis'. There is an idiopathic and a (rare) autosomal dominant genetic form of the condition. The main characteristic of this condition is an acute beginning, starting with extremely severe pain in the neck, shoulder and/or arm. As a pain score, (VAS), patients often give 8 - 10 out of 10. These are often people who have never before had shoulder or neck complaints. The pain is much more severe than 'normal' shoulder or neck complaints and hardly reacts at all to painkillers (including opiates). In the beginning, the pain is present constantly and can barely be influenced by a change of position or anything like that. Characteristically, the arm is held protectively against the body and patients often lie awake for nights on end because of the pain.



Figure 2. Winged scapula on the right



Figure 3. Distal atrophy and paresis on the right

Generally, after a few hours to days – but sometimes not for a few weeks – a patchy paralysis of the shoulder blade arch and arm muscles develops. There is also often, strikingly quickly, a considerable atrophy of the affected muscles. The so-called shoulder blade alata is very characteristic (see figure 2), but this is certainly not present in every patient. As well as the classical 'proximal' form in which lifting the arm and exorotation of the arm and the shoulder are affected, there are also more distal forms in which stretching in the wrists and fingers or the small muscles of the hand is often severely curtailed (figure 3).

After thorough questioning, it can turn out that there are often sensory symptoms in the form of tingling or a slight hypesthesia, but these symptoms are generally not the most prominent.

The pain generally progresses in three phases: First of all, there is the continuous, severe neuropathic pain that lasts for several days to weeks, then the pain disappears in rest, but increases with movement or stretching of the damaged plexus; finally the patients are then extremely hampered by a persistent muscle pain through overburdening of the paretic – but also the compensating, healthy -- muscle groups.

The paresis recovers slowly over a period of months to a few years. This slow recovery and the development of muscle symptoms through 'surmenage' or overwork, often lead to long-term (partial) disability and necessitates good supervision and guidance of these patients in rehabilitation and employment.

Treatment of neuralgic amyotrophy (NA) in the acute phase

The pathophysiology of an episode or attack is probably a patchy infection inside the plexus brachialis based on an auto-immune reaction. In order to try to limit or prevent damage to the nerves as much as possible, we advise, as a painkiller in the acute phase, a combination of long-acting morphine (for example, MS Contin 2 dd 10-20 mg) with a long-acting NSAID (for example, Diclofenac 2 dd 100 mg Retard). If there are no contraindications, a short course of oral prednisone can be considered. We use 60 mg per day for this (i.e. and for children: 1 mg/kg) for 7 days, in the following week to be reduced by 10 mg / day to nothing. Prednisone is no miracle drug; on average, 5 out of 10 NA patients have a favourable reaction, with a fast decrease of pain, and, in 1 or 2 out of 10 people, the recovery is

somewhat faster than would have been expected. At this time, however, there is no other alternative so we still advise it if there are no contraindications. Co-analgesics, such as carbamazepine or gabapentin are, in theory, the most suitable for the neuropathic pain but, in practice, the effect often takes too long (4-6 weeks) to truly be useful in the acute phase of an attack of an NA.

Treatment

It goes without saying that, first of all, a treatable cause of the plexopathy should be sought. If this cannot be found, attention is then turned to pain control, prevention of complications and creating the most optimum conditions for spontaneous recovery.

Pain medication

With acute pain, a combination of a long-acting NSAID and an opiate often works best. If it is expected that the pain will continue for longer than a few weeks, it is a good idea then to add a co-analgesic to subdue the neuropathic component of the pain. Be wise, but not too frugal with painkillers: this neuropathic pain can be extremely severe and debilitating! Progression of the pain, in spite of pain medication, compels us to search for an underlying progressive etiology or the development of complicating factors, such as joint problems or tendo-myogenic overexertion.

Type of neuropathic pain	Medication
Acute	Diclofenac retard 2 dd 100 mg + MS Contin 2 dd 10 mg (also consider leave : stomach protection and laxative)
Chronic, especially shooting/stabbing	the above and: carbamazepine 2 dd 100-200 mg, or gabapentin 3 dd 600-1200 mg
Chronic, chiefly burning	see acute pain, and: amitriptyline 2-3 dd 10-25 mg

Figure 4. Example of pain medication for plexopathy

TENS

If medicinal painkillers alone do not help sufficiently, a TENS (Transcutaneous Electrical Neuro-Stimulation) appliance can sometimes provide even more relief. This device, a sort of 'walkman' with a number of electrodes that are affixed to the painful areas with stickers, drowns out, as it were – via electrical stimuli – the pain signals that reach the spinal cord. In the Netherlands the patient must apply for the device himself – with an authorization – with the insurance provider and then, with a new authorization, have the physical therapist instruct him in its use.

As the nerves recover, the patient can practice more actively. In time, the number of repetitions and, later if necessary, the resistance can be expanded. Patients need not be afraid that exercise will further damage the nerves; but be careful not to strain them. An exception to this is the group of people with hereditary pressure neuropathy, or HNPP; they are born with nerves that are hypersensitive to stretching, pressure and pulling. Specialist advice is essential in these cases.

Specific pain symptoms in the case of a plexopathy of the shoulder

In the case of a paresis of the muscles that fixate the shoulder blade to the trunk (specifically, the serratus anterior, trapezium and rhomboid), patients often complain about an increase in pain in the arm and shoulder if the limb hangs down for a long time, such as when walking or standing. During physical examination, it is generally also visible that the shoulder blade has slipped down on the trunk in a resting state, often laterally and downwards. Because of the weight of the arm, traction develops on the muscles fastened to the shoulder blade. Although patients should, even in such a case, try to keep the arm as pliable and mobile as possible, it could be advisable – certainly when standing for long periods of time or walking – to support the arm in the jacket pocket or in a sling or something similar in order to temporarily relieve pressure on the shoulder. This often provides sufficient relief.

Moreover, some patients are also bothered by a kind of chronic muscular pain in the paretic postural muscles. Most shoulder and pelvic girdle muscles are unconsciously actively occupied all day long in maintaining the posture of the body and, therefore, get 'no rest'. The ability to move our arms well, for example, especially to the front beyond our normal power, is largely dependent upon a stable shoulder blade which the arm can push against. If the muscles responsible for this are weakened, they must then tighten themselves to the maximum in order to provide any stability at all. And, they must do this all day, every day. The chronic muscular pain resulting from this, in our experience, hardly reacts to painkillers at all. Often, the best solution is to try to change position and movements frequently, not to do any one activity for too long and, in the case of an unstable shoulder blade, to rest by lying flat on one's back.

Often, topical heat is pleasant; sometimes a shoulder blade-brace offers some support.

Alongside the above advice, cognition with respect to the symptom can play a role in perpetuating it. Because patients cannot see and feel their shoulder movements well, sometimes ideas develop about what is going on that do not help in the recovery and in dealing with the symptoms. For example: 'whether or not I use it, I am still in pain', or 'I will avoid all use of the shoulder because every time I take something out of the cabinet, I am in so much pain'. In the chronic phase, the connection between what one does and the symptoms is often hard to explain to

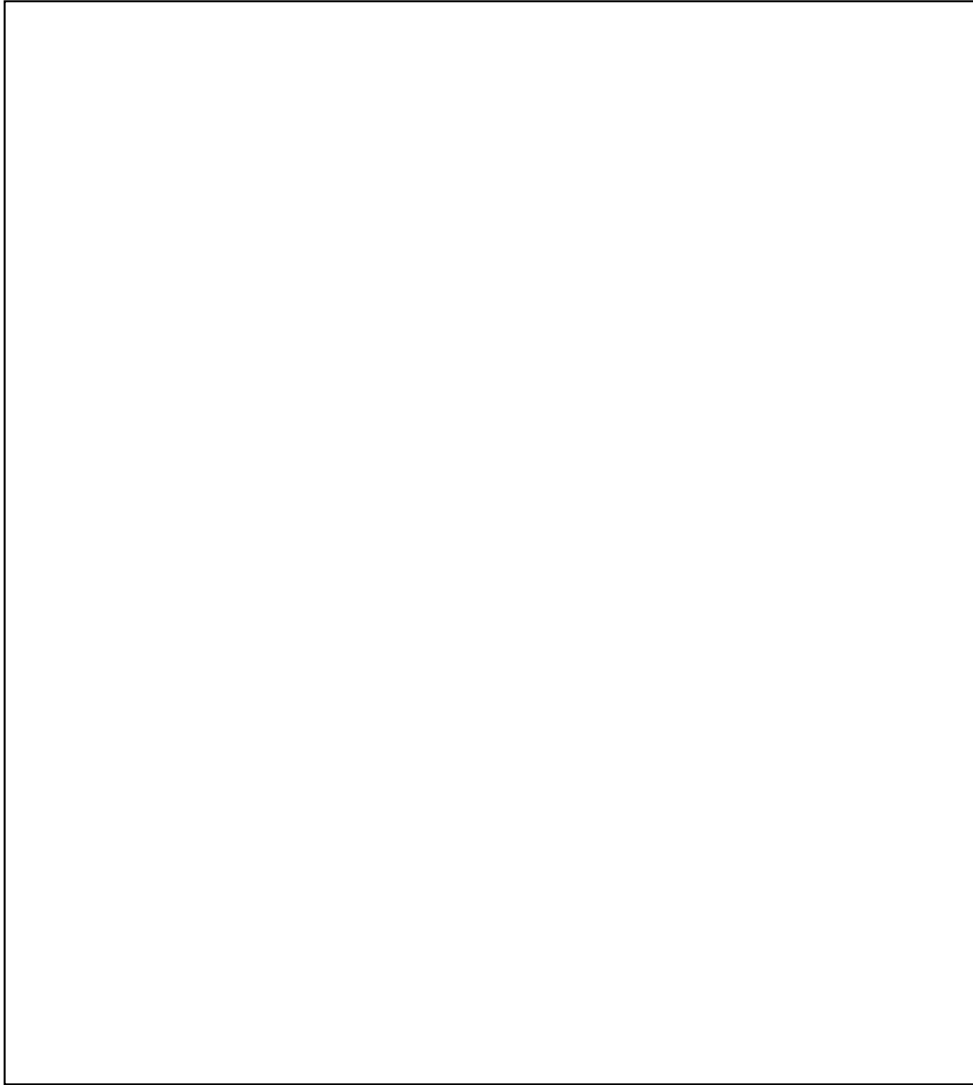
the patient. It is only after sufficient explanation and after having the patient feel and experience what overtaxing does and what using the shoulder in the correct manner does with the symptoms, does it become clear to the patients and the therapists how one can work gradually on building up the capacity and seeking the balance between load and capacity.

Treatment advice

At the hardware level, the only worthwhile intervention is the optimization of the biomechanical situation. With the aid of the physiotherapist, one should try to recover a perishoulder blader movement pattern that is as normal and fluid as possible. Dysfunctional compensation strategies must be avoided or unlearned. Timely alternating periods of exertion and rest (the so-called "mini-breaks") are important in order to halt surmenage. The use of a foam rubber sling to support the arm while standing and walking is a useful part of this approach if the m. serratus is still weak (< MRC 4). It is certainly important then to have the patient exercise the passive mobility of the shoulder at least twice a day in order to avoid contracture. There are a few important pitfalls in the physical therapy for NA. Problems occur when the exercise programme give too much load, in particular if extra weights are used while the muscle strength is still < MRC 3. Moreover, it is, in practice, not possible with a perishoulder blader paresis, to strengthen the 'surrounding' muscle groups without (over) taxing paretic muscles. If this is done, the pain will often increase after the session.

When thinking about resuming work or hobbies, it must be explained to the patient that there is no reason to be afraid that the attacks will return or the nerve recovery will be hampered by physical exertion. At this time, it is simply not possible to predict whether or not the attacks will return, whatever one does or does not do. The level of the load follows, ideally, the amount of available muscle strength and stamina and must be adapted by the patient himself because, otherwise there is a great chance of over-taxation and, consequently, a reduction in physical ability. Gradual expansion of the activities, while dividing the amount of time that one spends in a certain position (such as sitting at a computer screen) and alternating these physically taxing activities with other less taxing activities is generally a good idea as long as there is still muscle weakness and physical limitations.

Write down your questions

A large, empty rectangular box with a thin black border, intended for writing down questions. It occupies the left half of the page.

Address

Visitors' address

UMC St Radboud
Neuromuscular Centrum Nijmegen
Radboud Oost
Reinier Postlaan 4, route 901
Nijmegen

Postal address

UMC St Radboud
Neuromuscular Centrum Nijmegen
935 Neurology
Postbox 9101
6500 HB Nijmegen

© UMC St Radboud
overname van (gedeelten uit)
deze tekst is uitsluitend
mogelijk na schriftelijke
toestemming van
Staf Concerncommunicatie