Amplified Musculoskeletal Pain or Reflex Neurovascular Dystrophy (RND)
A guide for parents of children with RND
By David D. Sherry, MD

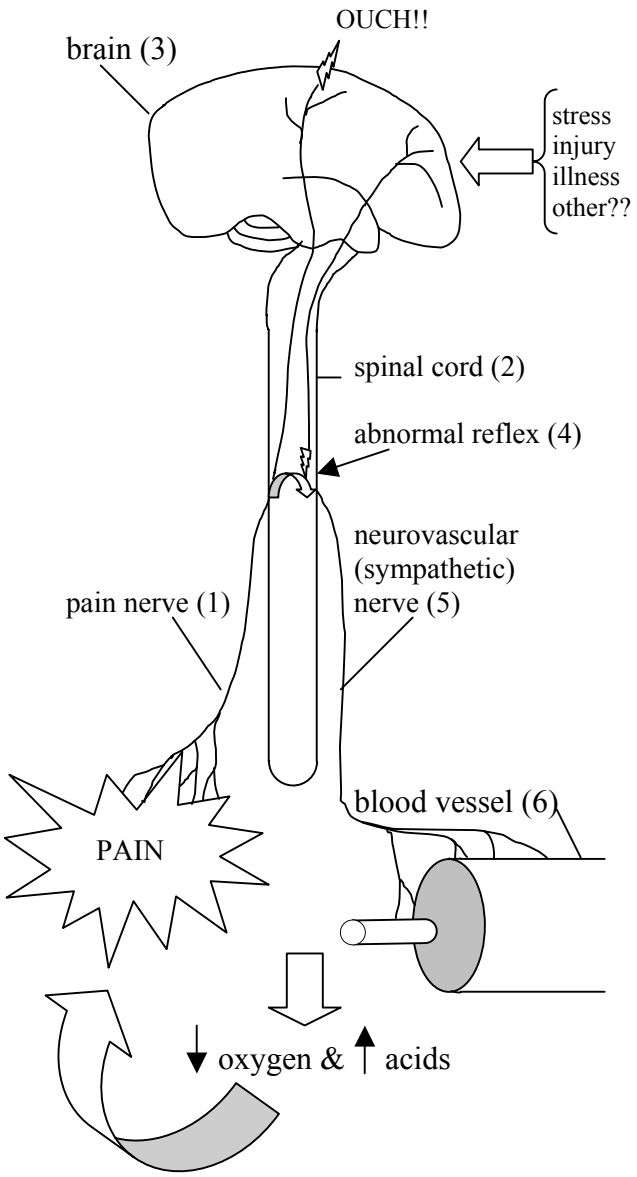
What is Amplified Musculoskeletal Pain?
Amplified Musculoskeletal Pain or Reflex Neurovascular Dystrophy (RND) is a very painful medical condition. It usually affects a limb (a foot or leg more commonly than a hand or arm), but can cause pain anywhere on the body. Some children have pain all over and a few have intermittent attacks of pain. It has multiple manifestations and each form can be named separately, but herein I will use the term RND to encompass the spectrum of amplified musculoskeletal pain syndromes. The pain these children experience, however, is much more intense than one would normally expect because the pain signal is amplified.

How is the pain signal amplified?
First, look at the figure to see how we normally feel pain. Usually pain is in response to tissue damage, such as stepping on a tack. The damage sends a signal through the pain nerve (1), to the spinal cord (2), that then sends the signal up to the brain (3). The brain then recognizes the signal as being painful.

In RND there is an abnormal short circuit in the spinal cord (4). Therefore, the pain signal not only travels up to the brain, but also goes to the neurovascular nerves (5) that control blood flow through the blood vessels (6). These nerves, however, cause the blood vessels to constrict thus decreasing blood flow. The decreased blood flow deprives the skin, muscles, and bones of oxygen and also leads to a build-up of acid waste products such as lactic acid. This lack of oxygen and acid build-up causes pain. This new pain signal also goes across the abnormal reflex and causes a further decrease in blood flow, thus leading to more pain. Thus, the pain is greatly amplified. RND is a very painful condition.

Frequently the part of the body with RND will become cold, blue or purple, and even swollen due to the decreased blood flow. Some children will have a test called a bone scan that may show this decreased blood flow (although this test may also be normal – there is no specific test for RND). Other findings frequently seen are osteoporosis due to a decrease in the calcium from the bones and muscle wasting (atrophy). Rarely, the skin will become thick and waxy, a so-called dystrophic change that gives RND part of its name.

What causes the abnormal reflex?
There are three major reasons for the abnormal reflex: injury, illness, and psychological stress. There may be other reasons such as age, genetics, or hormones (80% of children with RND are girls). More research needs to be done.



What kind of injury leads to RND?
Injury leads to RND in approximately 10 - 20% of children with RND. The majority of children with post-traumatic RND have a specific injury such as a broken bone, piercing injury, or surgery. The symptoms develop immediately or within a few weeks after the injury. Frequently as the injury is beginning to heal the pain starts and then continues for weeks to months even though the injury fully heals.

What kind of illness leads to RND?
Illness is an infrequent cause of RND. Most commonly I have seen it in conjunction with inflammatory illnesses of the musculoskeletal system such as arthritis, tendinitis, or enthesitis. Other illnesses may be the initial cause of pain, most are infections such as mononucleosis, influenza, gastroenteritis, or streptococcus.

What kind of psychological stress leads to RND?
In fully 80% of children with RND, psychological factors seem to be playing a role. There is a whole host of possible stresses that may play a role in causing (or perpetuating) RND including the psychological consequences of having the pain (and perhaps not having a timely diagnosis). All change is stressful, even a change for the good. Feelings like fear, worry and shyness are also stressful as well as the stresses of the events occurring in the world today. The most common stresses we see in these children are those that arise from family and school issues. Many families are undergoing changes such as an older sibling leaving the home, new jobs, or marital issues. School is also becoming more demanding as well as peer pressure and social demands. Developmentally most children with RND are gaining more independence and responsibilities. Some children deal with stress by keeping it inside and this stress can lead to bodily symptoms, including RND.

Is the RND all in my child's head?
No. The pain is very real and very intense. Even in those children in whom psychological stress plays a major part in causing the RND, the decreased blood flow to the area of RND causes the extreme pain.

Are there any laboratory tests to prove it is RND?
No. Blood tests are normal unless there are other conditions present (for example, an infection can alter the blood counts and sedimentation rate). In some children a bone scan can show abnormalities that are very suggestive of RND but the bone scan may also be normal.

What can be done to help my child?
It is most important that someone with RND begin to use his or her body in a normal way. The abnormal reflex is broken by intense exercise therapy. Some children are able to do this on their own by exercising at home. Although it hurts to do the exercises, it does not cause damage, and some children are able to work through the pain.

Most children will need to have a formal exercise program. It just hurts too much for him or her to do it on his or her own. Most require daily out-patient therapy. A few children require hospitalization. The exercise program lasts 5-6 hours a day and involves physical and occupational therapy as well as pool therapy.

In addition to the exercise therapy we do an evaluation to see if stress plays a role in your child's RND. This involves filling out questionnaires and talking to the psychologist. Some children may also need to have formal school testing. It is through an evaluation that we can either give you more specific advice about formally addressing stress or determine that stress does not play a role in your child's RND.

A very few children will need a behavior modification program. It we think your child needs this we will speak to you about this in depth.

How long does the program take?
The average is 2 to 3 weeks but an occasional child will require many weeks. It is impossible to predict until we see the rate of progression once in the exercise program. Function comes back first; the pain usually takes longer to decrease. Most children are fully functioning by two weeks and pain-free by four weeks.

What will my child be doing in the program?
Your child will have an exercise program designed specifically for him or her. It will be intense and will focus on the body areas that are painful or do not function properly. If there are body areas that are painful to touch, these areas are desensitized with rubbing and massage. In the beginning this exercise program will take up to six hours a day. Physical and occupational therapists that are experts in RND direct this therapy. A program of exercises to do at home is also part of the exercise treatment. There will also be scheduled appointments with the psychologist for the child and parents and there may be schoolwork evaluations scheduled.

Are parents allowed in the exercise sessions?
No. We have found that it is best that the parents not be present during the exercise therapy. Some children respond differently in a parent's presence. While the child is being treated the parents can go about their usual activities. Younger patients may need some supervision by a parent during lunch and other breaks.

Do the exercises have to be done here?
For most children seen in Philadelphia, yes. The type of therapy is different from what most physical and occupational therapists are taught. Most local therapists are not able to deliver the amount of therapy required. However, there are situations where we can work with a local therapist and, as stated above, not all children need the full five-hour a day program. We are very supportive of having other therapists develop an expertise in treating children with RND. More treatment centers need to be established so children can be adequately treated close to home in a timely fashion. We can supply your physician and therapist with written descriptions of our exercise program. A video or DVD for physical and occupational therapists explaining the exercise program in detail is available on loan (or purchase).

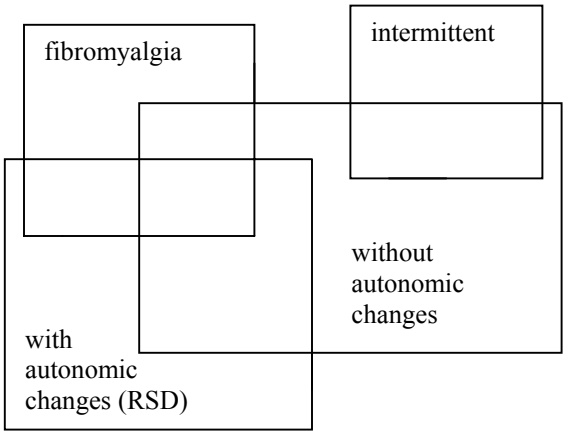
What do I tell others?
In simple terms you can explain that RND is a medical problem involving the nerves going to the blood vessels. These nerves are overactive and cause the pain. The treatment is a special series of exercises to retrain the nerves.

Are medicines used for pain?
No. Most patients take no medication because it seems to hinder the retraining of the nerves. Also, medication frequently causes side effects in these children and has minimal, if any, benefit. A very small number of children are depressed and will need anti-depressant medication. Some children may have sleeping difficulties and will need medication to help restore a normal sleep pattern; however, initially we try to avoid this since most will sleep better once they begin the exercise program. Before we use any medications, the risks and benefits will be fully discussed with you.

What other names does RND or amplified musculoskeletal pain go by?
There are various patterns or kinds of amplified musculoskeletal pain that are called a variety of names. I prefer the term RND since it refers to the abnormal reflex and neurovascular nerve. Most of the other names are related to where the pain is or to changes in the skin temperature and color (autonomic changes). These names include reflex sympathetic dystrophy, fibromyalgia, allodystrophy, complex regional pain syndrome types I and II, causalgia, Sudeck's atrophy, shoulder-hand syndrome, localized or diffuse idiopathic musculoskeletal pain, neuropathic pain, psychogenic pain or psychosomatic pain. The literature (and Internet) is quite confusing; most of it applies to adults with specific syndromes, especially

fibromyalgia and reflex sympathetic dystrophy. Children are much different than adults in the presentation of the illness, the response to treatment and the long-term prognosis and, on the whole, fare much better.

Various forms of RND can coexist in the same child or, if there is a reoccurrence, the second form may be different from the first form. A pictorial representation of the overlapping nature of the forms of RND is shown below:



How can I prevent RND from happening again?

Your child is going to have illnesses, injuries, and psychological stress in the future. However, if he or she starts to have excessive, increasing pain you should suspect RND. Also, if the area is tender to very light touch, suspect RND. Half of the children with a second attack of RND will say that the pain feels just the same as the first time; however, half will say that it feels different. Even if the pain feels different, it still may be RND. If you suspect RND, restart an exercise and desensitization program. If it is tender to light touch, desensitization with rubbing and massage should begin. The majority of second attacks occur within the first six months of the first episode. Therefore, when illness or injury occur, be sure that the pain is not out of proportion to the illness or injury.

If some form of psychological stress is present, having the stress addressed through formal counseling can be of great help.

Why did it take so long to diagnose the RND?

Early diagnosis may be difficult for several reasons:

1. Not all the RND symptoms may be present at first but may evolve over time.
2. Urgent problems such as fractures or infections need to be ruled out first to be 100% sure before starting an exercise program.
3. It may be hard to recognize since there is a wide spectrum of RND so unless the doctor has seen a variety of children with RND he or she may not diagnose it.

What is the long-term outcome?

There are few studies of long-term outcome. In one study where the children were treated with an intense exercise program, 88% of the children were pain free and fully functioning after an average of five years.

If psychological stress is a cause of RND, we have seen children subsequently develop a wide variety of other psychologically driven disorders such as anorexia nervosa, bulimia, other amplified pain syndromes (such as abdominal pain, headache, eye pain, sinus pain, tooth pain), pseudo-seizures, uncontrollable shaking or muscle spasms, suicide attempts and conversion reactions. In a conversion reaction, the body converts feeling and emotions into neurological symptoms such as paralysis, blindness, or numbness.

What is being done to find out more about RND?

Children's Hospital of Philadelphia is one of the few places doing research on children with RND. You and your child may be asked to take part in research. Before participating in any research project, it will be fully explained to you and is always entirely voluntary. Contributions toward research are always appreciated.