KIDS GET SCLERODERMA TOO!!!!!
www.jsdn.org

For Kids' Sake

A Support Network Helping Children With Scleroderma & Their Families Summer 2004

Why Would A Child Have Raynaud's?

Written by, Thomas J.A. Lehman, M.D.
Pediatric Rheumatologist
Hospital for Special Surgery
New York, NY

Everyone reading this newsletter already knows that adults may get Raynaud's, but how many of you realize that children may get it too? Fortunately Raynaud's in childhood seems to be rare. (a complete review of the literature failed to produce a single estimate of its prevalence). In my large pediatric rheumatology practice I see fifteen to twenty new cases every year. Pediatric rheumatologists see many children with Raynaud's, but there are probably many more who never see a doctor because their hands get cold or are dismissed by their doctors without further investigation because, "it happens sometimes."

Many people think Raynaud's only happens in the cold, but I've treated children who had Raynaud's in 80 degree weather. It can happen with embarrassment, air conditioning, or any other event that alters blood flow. It's not just fingers. Some children have toes that blanch. Some children will see the tip of their nose turn white, or even their ear lobes.

Raynaud's in children occurs because the blood vessels in the fingers (or elsewhere) are clamping down in spasm. It's the same thing that happens in adults. But children are supposed to have young and healthy blood vessels. They aren't using jack hammers or working with chemicals that may cause Raynaud's. Does that mean the children all have diseases like lupus or scleroderma? No fortunately not!! Many thin teenage girls have Raynaud's, just like their mothers. How can you be sure?

There are a number of important steps to take if you have a child with Raynaud's. First make sure that's what is happening. Remember Raynaud's is not simply cold hands. True Raynaud's requires a three phase color change. The fingers have to blanch white, then turn red as they warm up, then get a bluish/purplish dusky color as the poor circulation returns. If the fingers don't blanch white, it's not Raynaud's.

If a child definitely has Raynaud's they should be evaluated by a rheumatologist, preferably a pediatric rheumatologist with experience in the area. Studies have been done on a number of these children and it appears to be possible to separate those we should from those we shouldn't worry about. All children with Raynaud's should be tested for antinuclear antibodies (ANA). Many with nothing wrong will test positive, but if there are no findings on exam it is unlikely that a child who tests negative for ANA has any underlying condition. However, a negative test once, is not a lifetime guarantee. If things change, get tested again. In addition, primary Raynaud's is infrequent in boys. A boy with Raynaud's always deserves extra attention.

continued on page 3

Inside This Issue: *Treatment of Juvenile Scleroderma, page 4
***A Personal Look At Juvenile Scleroderma, page 6 *Summer Camp, page 7**

For Kids' Sake
Published by:
Juvenile Scleroderma Network, Inc
1204 West 13th Street
San Pedro, CA 90731
Helpline: (800) 297-4756 ext. 74
Tel: (310) 519-9511
Email: OutreachJSDN@jsdn.org
Website: www.jsdn.org

Parent Collaborative
Officers 2003-04
Kathy Gaither, President
Carmela Candella, Vice-President
Sherry Gaither, Treasurer
Board of Directors
Jerry Gaither, Chairman
Carmela Candella
Kathy Gaither
Sherry Gaither
Roxanne Nichols

Medical Advisory Board
Balu Athreya, M.D., P.R.
Wilmington, DE
Lawerence Eichenfield, M.D., P.D.
San Diego, CA
Donald Gaither, R.N., B.S.N
San Antonio, TX
Julie James, CNC
Long Beach CA
Bernice Krafchik, M.D., P.D.
Toronto, Canada
Ronald Laxer, M.D., P.R.
Toronto, Canada
Thomas J.A. Lehman, M.D., P.R.
New York, NY
Vanessa Malcarne, Ph.D.
San Diego, CA
Richard Mier, M.D., P.R.
Lexington, KY
Terry Moore, M.D., P.R.
St. Louis, MO
Egla Rabinovich, M.D., P.R.
Durham, NC
James R. Seibold, M.D.
New Brunswick, NJ
Kim Striegel, R.N., F.N.P.
New York, NY

Newsletter
Kathy Gaither, Editor

Disclaimer: We must remind you that the JSDN in no way endorses any drugs or treatment that we report to you. It is our wish only to keep you informed. We ask you to check any treatment with your child's physician.

©Copyright 2004 JSDN

A Message from the Board Of Directors

Albert Einstein once said, "In the middle of difficulty lies opportunity." *Most of us are reading the For Kids' Sake newsletter because we have a child or loved one with juvenile scleroderma. This difficulty lead us to the "source of help and hope" for families of children who have juvenile scleroderma. Lets take a look at some of the opportunities we have available through the JSDN.*

Internet Friends: *Parents of children with JSD are invited to connect with other parents and children worldwide for information, advice & emotional support with our JSD online community.*
Family Outreach Program: *Our program will put in touch with other families in your geographic area. You will receive the support when you need it by a Family Outreach Volunteer Parent. By the way, Family Outreach Volunteer Parents we need you!*
Medical Questions: The Doctor is In: Q&A: *Dr. Lehman has donated his time to answer your medical question(s) through our email form. Please feel free to send your questions anytime.*

The JSDN website has what you need. If you can't find it, let us know and we'll find it for you. We have a great Resource List and excellent articles under the Archive of Medical Articles. Take a look at our website to learn more about our programs and services for the whole family at www.jsdn.org.

Please keep in mind, the JSDN programs and services would not be possible for families if it weren't for our generous donors and membership to the JSDN!

There are many ways you can help the juvenile scleroderma cause…Join the JSDN! Make a donation! Volunteer to raise awareness!

Just so you know, we value your privacy! Your privacy is the utmost concern to us. To ensure your privacy, it is our policy ***NOT*** *to* ***RENT****,* ***SELL****, or* ***EXCHANGE*** *our mailing list to any person or company.*

We greatly appreciate everything you can do for the JSDN and the juvenile scleroderma cause!

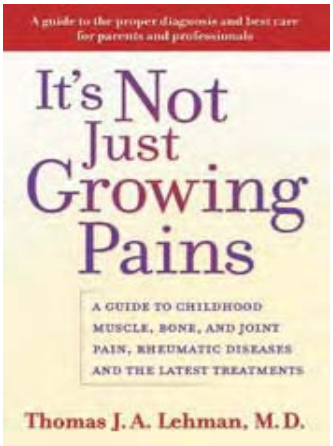
It takes a nation to bring an organization together…

Warm Hugs & Support,
Kathy ♥ Jerry ♥ Carmela ♥ Sherry ♥ Roxanne

How does your tax-deductible donation help?

P*rovides membership packets with valuable information*
P*rovides educational newsletters twice a year*
P*rovides literature on juvenile scleroderma*
P*rovides a 24 hour 800-number help line*
P*rovides 24 hour online support*
P*rovides a Website that helps the JSDN give worldwide educational and emotional support*
P*rovides Pediatric Rheumatology referrals*
C*onnects families to a support network for children with juvenile scleroderma*

New!



"A must-have for any family with a child who has a rheumatic disease! I personally found this book to be very informative. Dr. Lehman, as always, gives families a clear understanding of their choice of treatments and what they can expect from their child's rheumatic disease." – Kathy Gaither, Juvenile Scleroderma Network

GET YOUR COPY
TODAY
AND LEARN ABOUT

~ diagnostic tests
~ discusses the most effective medications
~ discusses physical therapy
~ alternative therapy
~ surgical options that are available

This book is available online at:
www.goldscout.com
www.jsdn.org

Also available at fine bookstores

Members Only Contest

Send in or email your name, address & phone number to the JSDN, and you may be eligible to win It's Not Growing Pains.
All entries must be received by August 1, 2004.

continued from page 1

If a child is ANA positive further evaluation should be done to make sure they have normal blood work including a normal blood count, a normal 'chemistry panel,' a normal sedimentation rate, and a normal urine. Many doctors will do all these tests when they draw the initial ANA. Once the physician knows the ANA is positive they should test for anti-centromere antibodies, anti-scl 70, and the subtypes of ANA. I also test for antiphospholipid antibodies, RF, and anti-thyroid antibodies. Any further testing will depend on the clinical picture. Some children with Raynaud's will have a significant underlying connective tissue disease (secondary Raynaud's). All of these children should be under the care of an experienced pediatric rheumatologist. So should all children with significant abnormal blood tests. Most parents' children will have normal tests. Positive tests for ANA in low titer are very common. If the ANA is positive, but the physical exam is normal and there are no other laboratory findings, it's still primary Raynaud's.

So what should you do now? The most important thing for parents of children with primary Raynaud's is common sense. Keep their hands warm. Make them wear gloves. You want to make sure they pay attention to their fingers, but you don't want to make them, "different from everyone else." Let them go out and play in the snow with the other kids. Just make them dress more warmly and come in to be checked on periodically. Don't let them take a job at the supermarket where they have to work in the cold storage room or freezer cabinet. If a child with Raynaud's has a finger that turns white and numb that doesn't get better after an hour head for the doctor's office or the local emergency room.

Even children with primary Raynaud's can have real problems with cold finger tips. Children with significant problems can be treated with calcium channel blockers or other agents to relieve their symptoms. There aren't established pediatric doses for these medications, so you need to find a physician with the experience to use them in children. However, most children with primary Raynaud's don't require medication. If a child has severe primary Raynaud's or secondary Raynaud's make sure they are under the care of an experienced rheumatologist.♥

For more information about Raynaud's contact:
Raynaud's Association, Inc
94 Mercer Avenue
Hartsdale, NY 10530
(800) 280-8055
www.raynauds.org

Did you know

Dr. Lehman –
Best Doctors in New York
New York Magazine –
June 2000

To view Dr. Lehman's Pediatric Rheumatology home page go to:
www.goldscout.com

Treatment of Juvenile Scleroderma

Written by, Balu Athreya, M.D.
Pediatric Rheumatologist
DuPont Hospital for Children
Wilmington, DE

Our current approach to the medical management of juvenile scleroderma is based on anecdotal reports and personal experience and not on solid scientific evidence. This is because of several reasons. We do not have a good understanding of the basic mechanism of this group of diseases. There is no consensus on the classification of the various subtypes. It is difficult to measure the activity of the disease. Natural remission is very common particularly with the localized variety. In addition there are very few children with these diseases in any one pediatric rheumatology center and therefore it is difficult to perform controlled clinical trials. However several new developments in basic sciences give us hope for specific therapy. In addition, European Pediatric Rheumatology Society (PRES) has already conducted a world-wide study to collect data and efforts are being made to establish criteria for classification. Hopefully the next step will be multicenter clinical trials of potentially useful drugs.

Localized Scleroderma: *This variety is also called morphea by some rheumatologists. There are several subtypes within this group, morphea en plaque, linear morphea or scleroderma, deep morphea and en coup de sabre. Decision to treat this type should be based on the specific subtype and the fact that majority of patients enter remission in about 3 to 5 years. Morphea, which is superficial, is more of a cosmetic problem and it is preferable not to treat this condition with potentially toxic medications. Local ointments (such as Vitamin A and E) to soften the skin or local application of glucocorticoid creams are adequate. However, children with linear scleroderma, deep morphea and those whose lesions are active and spreading rapidly may benefit with the use of oral glucorticoids particularly during the early phase of the disease. This is used for only 2 to 3 months and tapered off. Other drugs used to treat localized scleroderma include d-Penicillamine, methotrexate, colchicine and hydroxychloroquine. One of these drugs is started early together with glucocorticoids and continued for 2 to 3 years at the least. At present, methotrexate seems to be the favorite although d-penicillamine has a longer track record.*

Several other agents have been tried with limited or no success. These include phototherapy with UV A1 with and without psoralen, which seems to be effective. However long term adverse effects are not known. Oral calcitrol therapy injection of interferon gamma into the skin lesion were found to be ineffective. Topical calcipotriene (a synthetic form of Vitamin D) seems too effective but needs further testing.

As mentioned earlier local treatment with emollients may be helpful to keep the skin soft. The lesions may involve deeper tissues and cause atrophy of muscle and contractures. When skin lesions cross- joints, contractures may develop. Therefore proper physical therapy program is necessary to stretch affected muscles and tissues, maintain position and maintain function.

Systemic Scleroderma (SS)*: There are two subtypes of this syndrome – diffuse cutaneous variety, which is the more common type in children and the less common limited cutaneous variety. Both these types are much less common than in adults. Once again no drug has been shown to be effective in these subtypes based on controlled clinical trials. However, better management of the internal organ involvement has contributed to better quality of life and better survival as shown initially with the use of angiotensin-converting Enzyme (ACE) inhibitors to treat kidney disease of scleroderma.*

Treatment of SS can be divided into two portions: 1.strategies aimed at modifying the basic disease process and 2. Strategies aimed at prevention and treatment of specific organ involvement.

Based on current knowledge, disease modifying therapy includes the use of drugs to suppress the immune system, to dilate blood vessels and increase blood flow and to prevent scarring (fibrosis). Several immunosuppressive drugs have been used including azathioprine, cytoxan, methotrexate and cyclosporin. There are no controlled trials to show any of the work. The only exception is the beneficial effect of intravenous cytoxan for the lung disease of scleroderma. Photapharesis and interferon gamma have not been found to be effective.

continue on next page

continued from page 4
Controlled trials of methotrexate in adults show conflicting results. However this is one of the commonly used drugs in systemic scleroderma. Finally autologous stem cell transplantation has been tried to treat severe scleroderma with limited success.

There are several exciting new developments in the treatment of Raynaud's Phenomenon and pulmonary hypertension – both caused by obstruction of blood flow. Nifedepine and amlodipine are the drugs of choice to treat Raynaud's Pphenomenon. . Since occlusion of small vessels is the cause of ulcers of the fingertip and loss of fingertips, this should be treated aggressively with avoidance of cold exposure, use of nifidepine or amlodipine and small doses of aspirin. Patients with severe Raynaud's Phenomenon, intravenous or subcutaneous treatment with prostacyclin and its analogues is recommended. These treatments should be given under strict medical supervision. Some of the more recent developments include oral agents such as Bosentan and Viagra and nitrous oxide to treat pulmonary hypertension.

Among the antifibrotic drugs, d penicillaiine is the most studied. Although a large study in adults showed that it was not effective, some authors find fault with that study and still use this drug. In our own experience, this drug is as effective as any other drug proposed. The main concern is with its toxicity.

Newer understanding of the mechanism fibrosis has shown that a protein produced by the fibroblast called Transforming Growth Factor (TGF) is responsible for abnormal collagen production. Inhibition of this biological agent by antibodies has been shown to correct the abnormality in experimental models. Exciting new possibilities are being explored to control fibrosis using antibodies and chemicals that interfere with factors similar to TGF.

In treating individual organs affected in SS, the first step is early identification. For kidney involvement, angiotensin-converting enzyme inhibitors such as captopril and some of the newer agents that act on the receptors will help prevent progression of renal disease.

Patients with reflux in the esophagus should be treated with proper posture after meals, antacids and proton-pump inhibitors. Severe constipation may be treated with stool softeners and enema as needed.

Lung disease and increased pressure in the blood vessels of the lung are major problems for these patients. Some of the newer developments have made for better prognosis and longer survival. This includes the use of cytoxan and newer agents to treat pulmonary hypertension. Patients with interstitial lung disease the treatment of choice is the use of intravenous cytoxan once a month for 6 months.

Arthritis and joint pain may be treated with non-steroidal anti-inflammatory agents such as ibuprofen.

General measures should include avoidance of cold exposure by keeping the temperature at home at a warmer level an the use of mittens and double socks when outdoors in winter. The child should be encouraged to lead as normal a life as possible. General skin care to keep it moist and supple should include emollient creams. Physical therapy measure such stretching exercises may be needed to maintain range of movement of joints and function. Splints may be needed to treat contractures. Fingertip ulceration should be prevented with good hygiene, aggressive treatment of Raynaud's Phenomenon and small dose of aspirin if they get infected, treatment with antibiotics is indicated. ♥

Just a reminder...

Summer time is here and we all love to go out and play in the sun. Before doing so read your child's prescription for any precautions. Some medications can make you more sensitive to the sun. Avoid prolonged sun exposure, tanning booths or sunlamps. The sun is most intense, usually between the hours of 10am to 4pm, use sunscreen with an SPF of 15 or greater, recommended an SPF of 30. If playing in water put more sunscreen on every hour or two. Sunlight also damages the eyes. Wear sunglasses that provide 100% UV ray protection. Remember that you can become sunburned even on cloudy days.

Have a safe and fun summer and please, don't forget the sunscreen!♥

~JSDN

Hilary Abigail Morrissey
14 years old, 8th Grade
Macon, Missouri

A Personal Look At Juvenile Scleroderma - Hilary's Story

My name is Hilary Morrissey. I live in North Missouri and I am a eighth grader at Catholic School. Next year I will be a freshman in high school in public school. I have scleroderma, (had a skin punch biopsy in 1996 when I was 6 years old) and have problems with swallowing which really scares me when I can not swallow well on my bad days. I do not like it when I am unable to swallow. It scares me that I may choke to death. I also have problems with acid reflux.

Some mornings I am very, very stiff like an old lady and it is hard to get out of bed in the mornings. But my mom always makes me get ready for school even if I feel like I do not want to go. She may help me get ready which I am very grateful for. Mom is always there for me when I need help. I am glad she does make me get to school on the days I do not feel well because I really love school and I do not like to miss going to school. I also may get stiff like an old lady in the afternoon or evenings too. I seem like I am always tired even though I get a lot of sleep and rest. I do not like to feel tired, but I can't help it. I try to work hard to keep up with my classmates. Sometimes it is very hard to keep up with my friends. I wish I had their energy.
I have a bad bone problem too. My hands and my feet are very, very small for my age. I will be 14 years old in May 2004. I wear a size 7 1/2 infant/toddler shoe. My genetics doctor says my hands and feet have fused, so they will not be growing any bigger. The x-rays tell the story that my hands and feet will not be growing any larger. I have learned over the years that it is better to have tiny hands and feet than to have no hands or feet at all.

So I guess I really should not be complaining, but I do sometimes. I have a lot of pain in my joints and all over and in my shoulders too, which the bones are popping out like I am coming apart but my skin is keeping me together. My skin also hurts. I may get swelling in my joints, in my knees, in my hands, feet and ankles. I have numbness and tingling in my limbs, in my hands, arms, feet and legs. On June 22, 2004 I am going to have surgery on my elbows to release a ligament. I had a nerve conduction test that came back abnormal in March 2004 and I am having problems with numbness and tingling in my tiny hands. I hope the surgery helps me.

I am just going to continue to take one day at a time and live my life the best I can. After all things could always be worse, right? So I will be happy with MY life.
~Hilary

I wish people could jump into my skin, jump into my body to feel what I feel like. Then people would better understand what I am talking about. I have a pain management doctor who tried to help me. I also have some scoliosis. My back has been hurting me a lot too. Sometimes this can be really painful for me.
I have an insulin resistance problem too. I have acanthosis nigricans which, is dirty looking black skin on top of the tight, dry, shiny scleroderma skin. I have this condition on different places like my neck and/or my armpits which is black looking. I have had people make comments that I really should be cleaning my neck or armpits better with a washcloth and soap....By the way, I am a VERY clean person! My dermatologist told me not to let the comments bother me, but they do just the same. Sometimes people question me about my skin or why my hands and feet are so tiny or why I look different. I am also going through an advanced aging process. I also had all of my toenails removed because they were getting so thick and deformed from the connective tissue problem. The deformed toenails caused me a lot of pain too. My foot doctor said my toenails looked like toenails from a 70 year old. I am very glad I had my toenails removed. It took me four surgeries to have them all removed and this was the best thing I have ever done. I am happy with not having toenails.

Actually, I am getting to the point to tell everyone when they always question me the truth that my mom had silicone gel breast implants for a long time to correct a chest deformity when she was young. Her silicone breast implants harmed me because they were failed breast implants that had problems!

continued on next page

continued from page 6

She feels really bad too, because my mom would never, ever hurt me on purpose. She loves me a lot and is always helping me to have a better quality of life. I do not know what I would do without my mom.

My family, my teachers, and my friends love me very much. They all help me in some way to have a better quality of life. I appreciate this too. Besides not always feeling good I have a wonderful life. My parents help me and love me and so do my two older sisters Hannah and Haley. I have a great family! I have a wonderful grandpa too. My teachers and friends at Catholic School help me too, which I am grateful for. I sometimes use a wheelchair chair for distances and they help push me around if I need to use it. *I do not like to always use it because people may stare at me when I go to the mall. But sometimes I feel I have to use it to get around so I am not so tired or in so much pain. So the wheelchair does come in handy. I have two dogs, which help me get through my bad days. Freddie the Freeloader is a 15 year old "Westie". He listens to me and is great comfort to me when I do not feel good. My Chihuahua, Pee Wee who is 4 pounds is almost 6 years old. He has tiny paws like I have tiny hands. Pee Wee, (P.W. for short) is always there for me and he is always a comfort to me too. So I am very lucky and very happy with my life!*

I want to tell you that on November 1, 2000 I donated 19 inches of my hair to Wigs for Kids. They make wigs for kids who have lost their hair to medical conditions or from cancer treatments. Last year on May 10th, 2003 I had cut off 16 inches of my hair, (second hair donation) for Wigs for Kids. Actually I did this as a Mother's Day present for my mom last year. Mother's Day 2003 was on Sunday, May 11th which was also my 13th birthday. My mom has multiple sclerosis, which is getting worse and she was having more health problems, so I cut off my hair so she would not have to french braid or fix my hair everyday. I thought she could use a vacation from always helping me with my hair, plus by donating my hair again I would also be helping a child feel better about what they looked like. So I felt like I was helping two people at one time. Since November 1st, 2000 I have donated 35 inches of my hair to Wigs for Kids. It is my hope that others may donate their own long hair to Wigs for Kids or to Locks for Love to help people of all ages feel better about themselves when they may not feel well or may feel different. I know what it feels like to look different. I am growing my hair out again and hope to make a third hair donation sometime in high school.

Well, this is my story and whatever happens to me is going to happen. I just have to be brave, keep positive and pray a lot! Besides mom says there is always someone worse off in the world than me with what I am dealing with....and she is right! I have a good life here in America and I am just going to continue to take one day at a time and live my life the best I can. After all things could always be worse, right? So I will be happy with MY life.♥

WIGS FOR KIDS

www.wigsforkids.org

Locks of Love

www.locksoflove.org

Hilary would love to hear from other kids at: **hil_bil90@hotmail.com**

Can't decide what to do for summer?

Why not send your child to camp this year? There are several camps for the summer and some are year round designed for children with a rheumatic disease and their families.

Some camps are FREE! and some have a fee, but for those who cannot afford camp usually there is a scholarship you can apply for.

The Arthritis Foundation has a wonderful summer camp. The children have theme days, arts and crafts, horseback riding, swimming, canoeing, archery, and so much more. There is always a pediatric rheumatologist and nurse on staff for when the children need them and they give the children their medication. Go to the Arthritis Foundation's website at: www.arthritis.org or call (800) 268-6942 for a camp location near you.

Hole in the Wall Camps, founded by actor, Paul Newman, also offer a wonderful summer camp for children with different disabilities, including rheumatic diseases. All children attend the camps FREE! of charge. Go to the Hole in the Wall Camps website at: www.holeinthewallcamps.org

Camp may be the only time during the year that lets them forget about doctor appointments and blood tests. They get a chance to meet other kids with similar experiences who they can talk openly about their fears. This is a their time for growth and independence, supported by dedicated people.

So grab your child, find a camp, and stock up on sunsreen! We'd love to hear about your child's experience at camp this summer.♥

Adults aren't the only one who get scleroderma!

Becoming a member to the JSDN connects people to a support network for children with JSD & their families. It is important to families to have a support network to exchange information and find emotional reassurance.

Yes! ***I want to join the JSDN. I understand this includes a one year tax-deductible membership to the JSDN.***

[ ] $20.00 U.S. **[ ] $30.00 Foreign & Canadian**
(U.S. Funds Only, Please)

Thank you for your support!

Name:_____**Email:**_____
Address:_____**C**
ity:_____**State:**_____**Zip:**_____

Please mail to:
JSDN – Membership
1204 West 13th Street
San Pedro, CA 90731

The JSDN is a separate non-profit organization from all other scleroderma organizations. The JSDN does not receive funds from other scleroderma organizations. The JSDN depends on your membership and donations for the JSDN programs and services. Thank you!

Published by: Juvenile Scleroderma Network, Inc
1204 West 13th Street
San Pedro, CA 90731
www.jsdn.org
A Support Network Helping Children With Scleroderma and Their Families

KIDS GET SCLERODERMA TOO!!!!!

For Kids Sake

Mission Statement:

It is hard enough to face the effects of Juvenile Scleroderma, but facing them alone can be devastating. The Juvenile Scleroderma Network, Inc invites families nationwide to become involved and help provide support and friendship to children who have Juvenile Scleroderma.