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TURNING POINTS IN SCLERODERMA CARE

By Leslie Berger

Dr. Philip J. Clements, Professor of Medicine at the David Geffen School of Medicine at the University of California, Los Angeles, has been treating scleroderma patients for more than 30 years. With his colleague, Dr. Daniel E. Furst, he co-edited the seminal medical textbook "Systemic Sclerosis."

Q: What has changed, and what do you consider the major advancements, in scleroderma treatment?

A: One of the first major turning points was the introduction of a medicine called captopril. It was a new antihypertensive that had a new mode of action, and it happened to be particularly well suited to the 10 to 20 percent of patients with scleroderma who would get renal crisis, which is typified by kidney failure and accelerated hypertension. That was 1980. Before that, 95 percent of people who developed that crisis would die within a year. Once it became available, better than 70 percent lived at least a year and most longer than that. If you can make it to a year, you're likely to live five years or more. So I'm still taking care of people who had their renal crisis just after captopril became available.

Another significant thing was that before the mid-1980s, scleroderma was never scientifically studied. At U.C.L.A., we were the first to do it. Studies weren't done because there weren't good, standardized measures to tell how people were doing. Since then, in the U.S., Europe and Australia, there's been an emphasis on how to get good, reliable measures. We have measures of skin thickness, lung and kidney function test, quality-of-life measures that are commonly used and considered valid.

The third big advancement were the reliable measures. One of those is the modified Rodnan Thickness Score. Rodnan was a big name in the '60s and '70s in scleroderma, and he pioneered this method for measuring thickness all over the body. You pinch the skin — a trained set of fingers does it. You use a 0 to 3 scale, with 3 being very thick. You measure in 17 areas so you can get a score from 0 to 51. We know from longitudinal studies the score changes over time in each patient. And you can predict in each patient what the skin will do over time. About half the patients have small amounts of skin involvement, now called limited cutaneous scleroderma. It's limited to the face and extremities. The other half have diffuse scleroderma, or all over. That tends to have more trouble associated with it.

If I do nothing else in the first visit, it's to get a sense of what folks know and what they don't know and kind of coach them through. What's real to worry about, and what isn't. This is very reassuring for most people. Even if it's bad news, it's the correct news rather than a diffuse worry about everything.

The last major advancement is the biochemical revolution, or our growing understanding of the chemicals the different parts of our bodies make to communicate with one another. We're beginning to learn the biology of what goes on in scleroderma, we're getting the chemical messages we think trigger the fibrotic system and can direct other chemicals to neutralize them.

Q: What's involved with stem cell transplant?

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A: It's adult stem cells, not embryonic; you need to make that distinction. Not the ones the religious right don't want to have used. You take the patient's own stem cells, freeze them, they get them back. They're very young cells that haven't decided what they want to do when they grow up. They're very malleable. The theory behind this is that [scleroderma] isn't genetically transmitted. Something happened to the immune system. The trick is how to reprogram the immune system to behave itself.

Q: What are the risks of stem cell transplant?

A: Infection's the big one. We have to give a wide spectrum immunosuppressant because we don't have a specific molecule to target, and that diminishes the body's ability to fight off infection of all kinds. Pneumonia in the lungs, sepsis in the blood, staph and meningitis in the spinal cord are among the possibilities. In addition, with advanced scleroderma a lot of organ function is already compromised, so it can be hard to tease out whether it's the disease itself or the transplant that's causing the patient to fail rather than improve. The radiation and chemotherapy given during the transplants can also cause skin and organ damage.

Q: What is the most important thing a scleroderma patient can do to improve her life expectancy and quality?

A: The first few years are the critical times for seeing whether the patient will have heart, lung or kidney involvement, so those are the most important years to be monitored. Now there are good treatments for kidney disease and reasonable ones for lung disease. The heart, I'm not sure. So you should be seen by someone who knows what he's doing, and you do it early and get regular lung function tests and echocardiograms. Anyone in the first five years should have lung function tested every six months and an echocardiogram once a year. This is to pick up stuff early, before symptoms appear. So it's getting to see someone, getting monitored and managing complications as they start arising.

Dr. Philip Clements is scheduled to speak at the SFNE 2014 Patient Education Seminar.

RAYNAUD PHENOMENON

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In 1865, a French medical student, Maurice Raynaud, described this condition, in which an individual experiences repeated episodes of color changes in the fingers (and sometimes the toes) during cold exposure or emotional stress.

It is normal for blood vessels supplying the skin or extremities to narrow (constrict) in response to cold temperatures, and this is termed "vasoconstriction." In Raynaud phenomenon, the body's blood vessels constrict in an ex-

WHAT CAN I DO TO MANAGE RAYNAUD SYMPTOMS?

Patients should take an active part in managing their Raynaud symptoms including:

- Keep the core body (chest, abdomen) and head warm by dressing warmly and wearing a turtleneck, scarf and hat in cold weather
- Avoid rapidly changing temperatures (such as the frozen food section of the grocery store).
- Avoid cold breezes.
- Stop cigarette smoking and try to avoid second hand smoke.
- Minimize emotional stress, if at all possible.
- Avoid repeated occupational or recreational injury (trauma) to the fingertips. Some patients may experience vibration-induced Raynaud, and for these patients vibrating tool use should be avoided.
- Practice methods to stop a Raynaud attack after it begins (placing hands under warm water or under the armpits, rubbing hands together, or rotating arms in a windmill pattern).

THE BIG CHILL

New England is known for its winter winds and freezing weather, and they will soon be upon us. 'Tis the season for both Raynaud attacks and very dry skin from having the heat up high.

Dr. Domsic's advice on managing Raynaud attacks is excellent **IF** you remember to put the hats, gloves, or mittens on **BEFORE** you leave the house. Struggling in the cold to put them on or waiting until you get into your cold car is enough time to bring on an attack. A remote car starter is also very helpful - put one on your Christmas list!

Dry, itchy skin is another winter problem. Moisturizers and other lotions/creams work best if applied frequently and liberally, especially after washing or bathing. While skin is still damp, moisturize your hands, feet, arms, and legs which have fewer sebaceous glands to lubricate them naturally. Through the years our readers have recommended using such products as Dove or Basis soap and moisturizers such as Eucerin, CeraVe, Lubriderm, Bag Balm, Vaseline, and even Crisco vegetable shortening! Experiment until you find what works best for you, and then remember to use it regularly!