

Scleroderma Foundation Chat 7.14.04

Guest: Dr. Daniel Furst

Chathost 1 Hello everyone and welcome to the Scleroderma Foundation chat room! Tonight we are lucky to have Dr. Daniel Furst of UCLA as our guest! Dr. Furst is a member of our Medical Advisory Board as well as a volunteer at the local chapter in Southern California. He is committed to finding better treatments for the complications of Scleroderma through clinical trials and he is a pioneer in autologous stem cell transplants for scleroderma. Welcome Dr. Furst!

DEFURST Good evening everyone, you will have to abide by my poor typing so please be kind.

Brock Dr. Furst, I am interested in your bovine collagen trials and how are they proceeding; Do you have any preliminary results?

DEFURST We are testing bovine Type 1 collagen for treating SSc. We have been joined by 168 SSc patients in a 15 month trial. We do not yet have any results but at least know it is not toxic.

Brock Thanks.

Jimw Is there any validity in the claims I have been reading about antibiotics for treatment for SD?

DEFURST Minocycline has been claimed to be effective in SSc but a recent open study of 53 patients has not shown any positive results. Other antibiotics have even less data... So I would not use antibiotics to treat SSc. They may be useful for complications like infections, of course.

Sclero What are stem cell transplants?

DEFURST Stem cell transplants are the use of one's own cells, in a special and purified form to help treat the disease. Sometimes a relative's cells are used. This is totally different from the embryonic cells that are so controversial at the present time.

Elizabeth I have had scleroderma for four years with kidney failure and lung involvement. Last fall I had bilateral dissected arteries. Could this have been due to scleroderma?

DEFURST The blood vessels are very much involved in SSc. Some think that large vessels may be involved, not just the small vessels such as those in the hands

or kidneys. However, dissected arteries are not part of SSc and it sounds like you had the bad luck to have two separate and serious problems.

Deedee Is there anything to be gained from a stem cell transplant? Are there any risks?

DEFURST For the 10-15% of SSc patients with the most severe form of SSc, these transplants may be life-saving. In our 25+ patients skin has improved by about 70%, while ability to function has improved by nearly 90%. However, this is an open study and we are about to embark on a controlled study. The potential for very serious side effects exist and about 10% of patients die from the procedure... This is only for the most severe disease and in a controlled trial.

Sschubby My wife has had SSc for nine years. She has been recently diagnosed with PAH and interstitial fibrosis of the lungs. She is taking Tracleer for the PAH. She also had kidney failure and is on dialysis. Is there anything else that can help with the lung involvement?

DEFURST Sometimes we add Sildenafil for the lungs and Prostacyclin derivatives like Flolan can also help. At the same time, other therapy for the whole of her SSc may be useful. Finally, some patients are undergoing lung and/or lung/heart transplants.

BC I underwent stem cell transplant at FHCRC in Seattle two years ago. I am doing great! What should I be doing for long term care?

DEFURST Hello BC, glad to hear from you. You should be getting your childhood vaccinations and should enjoy life. You need regular follow-up to be sure the disease doesn't come back... and if it does (it has in about 10%), that treatment is re-started.

Cmwhitten Does the stem cell transplant actually stop the progress of disease or does it reverse the symptoms?

DEFURST It seems to halt the internal organ involvement for at least three years (the average f/u to date), and improves function. We hope it halts the disease and it may reverse at least parts of it.

Michelle How common is facial changes in those with CREST? I know everyone is different, but is this something that the majority of us will experience at some point? I was diagnosed in 2001 and so far have the puffy fingers along with Raynauds.

DEFURST Facial changes are common with CREST (now called limited SSc). They may be minimal, but the skin is frequently involved on the face.

Jimw My wife was diagnosed about two months ago. She has an appointment with Dr. Clements at UCLA in August. What is important for us to do at this early stage?

DEFURST She needs to try to clarify with him what type of SSc he thinks she has. She needs to find out what degree of involvement she can expect. She needs to ask about what kind of treatments are available. If she wants to talk to some patients or read up on the disease, she should go through the Scleroderma Foundation as you are obviously doing.

Lashea Is there any medication for hypopigmentation? I have it on my hands and neck and it seems to be spreading.

DEFURST The best we can do at present are cover-ups and make-up. This is a tuff problem and we don't have good answers. Remember, however as trite as it sounds, that you are more than the way your skin looks... don't forget that.

Lashea Thank you.

OCCalifornia It was an honor to meet you and your wife at National Conference in LA a few weeks ago. Congratulations on "Doctor of the Year".
I was diagnosed with probable SSc in Indiana in February of 1997, even though my ANA was negative. The Rheumatologist suggested I move back to warm California to control Raynauds. When I returned to California, MD's denied SSc diagnosis due to negative ANA, and skin had softened. Two months ago on my second opinion a UCLA Rheumatologist diagnosed SSc (ANA still Negative).

DEFURST The diagnosis of SSc is not made on the basis of an ANA. It is a combination of symptoms, signs and other lab data. I can't say whether you have the disease but I can say no single lab test defines SSc.

Carole Have you heard of any success in treating diffuse SSc with either Methotrexate or Cellcept?

DEFURST There are two well-controlled trials that show that Methotrexate works for some parts of SSc- early skin, joint,??lung. There are only anecdotal studies of Cellcept to date, but the rationale for it is there and I am sure a trial of it will start.

Pacjoyseygirl I had sudden hearing loss four years ago. I currently wear hearing aids. Can Scleroderma cause hearing loss? Some doctors have said yes and others have said no.

- DEFURST** Hearing is not a part of the many problems that SSc can cause. There are associated “overlap” syndromes where “autoimmune hearing loss” can occur. You might ask about that as a possibility, rather than SSc.
- Kitty** Why the weight loss and excessive fatigue and where can I find information on stem cell transplants?
- DEFURST** Weight loss and fatigue are very common. Particularly early in SSc- depression, heartburn, bacterial overgrowth and other GI problems, Kidney, lung, heart problems, or anemia can all cause these symptoms. It is best to find out what your disease is like, then attack one area at a time... getting a sense of control can help anyone a lot.
- Carrie** What is the difference in Diffuse and CREST and also how does D-pen help the disease?
- DEFURST** Diffuse disease refers to involvement of a lot of skin (above the elbows and knees) while limited is the opposite (Diffuse is often more dangerous disease early on, while limited can cause problems later), The use of D-pen has been shown not to be effective.
- Chathost1** *Carrie, we have some publications at the National Office that may help you further understand the differences between limited and diffuse scleroderma. You can call the National Office (800) 722.4673.*
- Deedee** I was interested in the risks with stem cell transplants?
- DEFURST** As I noted, risks include a 10% chance of dying from the procedure... Other problems are increased possibility of infections. This is only for the 10-15% of patients with the worst disease and should be done within four years of starting the disease as longer duration decreases the chance of response, we believe.
- Jimw** Is there anything regarding diet and nutrition you should change or watch with having SSc?
- DEFURST** One area that needs to be watched is spicy or dry food for the patient with esophageal problems. Some people with lower GI problems develop lactose intolerance. If there is malabsorption, fat soluble vitamins need to be given in larger doses.
- Reelapple** What is the healing process of a transplant?
- DEFURST** After transplant, many of our patients feel much better within two months. They need to stay around the transplant center for three months to be sure their system doesn't get infected. By one year they need re-vaccination and

during that year they need prophylaxis against infections. They continue to feel better and better for over one year.

Michelle Does weight loss in scleroderma occur rapidly? May be a silly question... but how would my doctor determine if my weight loss is from SSc?

DEFURST This is actually a difficult question. It often occurs very rapidly early on, secondary to anxiety and depression and then recovers. If it is due to malnutrition or bacterial overgrowth, it may be gradual and may or may not be associated with diarrhea. The pancreas can also be involved and this can be tested for.

Lynn I had a Hepatitis B Vaccine and I did not develop any antibodies. I have SSc. Does this mean my immune system can not develop the antibodies? Should I have the vaccine repeated?

DEFURST Most of the time vaccination works, although not necessarily as well. I would strongly consider re-doing the vaccination, but check with your doctor to be sure there is not some other reason it didn't occur or that there is not a reason that it could be dangerous to do it again.

Tekakwitha Can you have nerve damage due to scleroderma?

DEFURST Nerve damage is relatively common and is associated with numbness, tingling or a "bad" feeling along the arms or legs whenever someone touches you. It does not affect the brain.

OCCalifornia Do you know of any relationship between SSc and fibroids in the uterus during pregnancy?

DEFURST There is no relationship to fibroids that we know of. Even though they are fibrous, we do not believe they are connected.

Marg What is the rationale behind the thinking that Cellcept would be helpful as a medication- I have had a kidney transplant and am already on Cellcept?

DEFURST Cellcept affects the cells we think helps cause SSc.

Debbied How do you know when your blood vessels are hardening? What does this mean?

DEFURST Hardening blood vessels usually is associated with less blood flow- so it can cause pain and decreased function wherever it happens. Legs, lungs, heart, kidneys, hardened arteries per se are not part of SSc though less blood flow

from the collagen and overgrowth may be. This is a large topic and I can't cover it well in just a few minutes. Sorry. Thank you all for your attention.

Chathost 1 Thank you Dr. Furst for your perseverance. We are so glad that you could join us tonight. If anyone has any further questions please feel free to contact us at the National Office at (800) 722.4673 and we will be glad to help in anyway possible. Don't forget to join us for our open chat the last Wednesday of the month. Thank you all!