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Scleroderma

New Zealand support group

newsletter

Winter Issue 2013

Winter has started but no complaints because we have had such a great summer. Who needs to go to Brisbane when we have such good weather here? Long may it last!

On the 5th May we had our first seminar.

This was held in the Learning Centre Room at Lower Hutt Hospital. It was well attended and we had people as far away as Dipton, (which is in southland for those North Islanders who have never heard of it before) to Auckland, Palmerston North, Kapiti and most of the Wellington group were there too.

Also attending was an Arthritis NZ educator, a nurse and a podiatrist. There was a good line up of speakers as you will read further on covering relevant topics.

It was good to see so many attending and also to see so many people in one room with Scleroderma.



Scleroderma seminar

Scientific health and experts' jargon could have flown thick and fast at the seminar, held most successfully on May 4, but through it all, the featured speakers managed to minimise it and talk to us in plain English.

The first speaker, Andrew Harrison, talked candidly about how to get the most out of available health services. He described an overstretched

health system in which your initiative will play a big part in your treatment.

Richard Steele, gave the laboratory perspective on scleroderma and the tests and laboratory detective work that confirm it.

He described this monitoring as "critical to well being for those with scleroderma.

Dianne Purdie thoroughly examined Raynaud's and the physiological reasons why it affects us.

Gordon Purdie wrapped up a full day, testing a theory that exposure to solvents seems to result in incidents of scleroderma.

Reports on the seminar continue over the page.

Seminar reports

Andrew Harrison, Head of Department of the Wellington Regional Rheumatology Unit and Associate Professor in Medicine at University of Otago, Wellington



Scleroderma, Andrew Harrison says, is poorly understood and it lacks a high profile. Drug companies prefer to research medicines to treat common diseases because they make a lot more money. The Beehive can overlook scleroderma too.

"There's low political incentive for funding because there just isn't enough of you to vote governments in and out," he says.

So there's no cure for scleroderma and no race to find one.

"On the bright side, if a drug were to be funded, it wouldn't break the bank," he says but academic interest rather than the promise of large financial rewards is still motivating research into the disease.

Scleroderma's MVPs

With a general lack of knowledge amongst primary health care providers, Andrew says you, the person with the disease, are top of the Most Valuable People list.

1. **You** know a lot more about it than the health professionals.
2. **Your general practitioner** will recognise that a rheumatologist or immunologist might be useful.
3. **Rheumatologists and immunologists** can coordinate your care and lead you on to other specialists (whose expertise may lie with just one organ) as the need arises.
4. **Specialists** include those in podiatry, cardio, renal,

physiotherapists.

5. **Facilitators** make things happen: nurse specialists, practice nurses.

Self help

Andrew's advice: don't put too much faith in health system.

"You will know more about your condition than most doctors," he says, many of whom may hear of it as 5th year medical students but may not have heard of it again till they met you.

"So the more you know, the better your care will get," is this specialist's advice.

"You are your lead health care advisor"

Rheumatology advice is stretched to capacity. Andrew says the recommended ratio of rheumatologists to the population is 1 to 100,000. In 2004 there was one per 238,000 people; in 2012, 1 for 162,000. So things are improving but we are still not doing well.

Having advocated self help, he outlined how to apply it.

How to get urgent help

In extreme urgency, call your local hospital's emergency department first. They can ring your specialist. You can call the hospital's rheumatology department or even go directly to your GP and get him or her to contact specialists.

Andrew Harrison says he is in favour of patients planning should they run out of medication or experience severe scleroderma symptoms. Plan with your

specialist and GP. For example, you need a plan if your blood tests suddenly show extreme readings.

"It really is a case of the squeaky wheel getting the oil and you've got to be prepared to squeak occasionally," he told the seminar.

Andrew also advised us to keep our own copies of records, particularly if doctors from different DHBs are involved. He advises you to take your records with you to ensure a complete history of your treatment is available.

Visiting a specialist

Andrew Harrison has treated scleroderma and related diseases probably more than most. He had suggestions for those for whom these visits are brief and overwhelming.

Decide what you want out of the appointment. Don't waste time describing symptoms: concentrate on your main concern. If you prefer, write up a list of what you hope to cover to avoid leaving the room kicking yourself because you forgot to ask some questions.

Stay informed on scleroderma as it relates to your specific problems and don't waste time wondering why you have scleroderma - it happened so focus on the things that matter, such as managing Raynaud's, reflux, skin ulcers, respiratory involvement and maintain a healthy lifestyle.

Conclusion

You are the lead health care adviser for yourself and, according to Andrew, you need to understand both your condition and the health system and be proactive.

Don't wait for the big things to happen.

Seminar reports

Dr Richard Steele, Clinical Immunologist and Immunopathologist at Aotea Pathology and Capital & Coast Health.

Systemic sclerosis: A Laboratory Perspective

Scleroderma occurs mostly in women aged 30 - 50 years, with perhaps 50 to 300 cases per million, according to Dr Richard Steele. He says there will be about one new case per million people each year. When men contract it, they usually do so at a younger age compared with women.



Dr Richard Steele address the seminar

No one, it seems, knows why you have scleroderma but Dr Steele says an infection with a similar structure to the human protein may trigger an immune response that directs itself against the human protein while fighting the intruder. It's called molecular mimicry.

It's possible that chronic infection or your environment may stimulate the disease. Researchers, have investigated such environmental agents as are found in silica, gold and coal mining environments. They look long and hard at organic solvents found in paint products, acetone and degreasers.

One theory is that a mother and baby may exchange cells during pregnancy and one of them may treat the other's cells as foreign and attack them.

The roll of the laboratory

Dr Steele ran through those short acronyms that appear on all blood tests that scleroderma sufferers learn to recognise.

ANA - anti-nuclear antibodies

Systemic scleroderma causes the body to produce 'autoantibodies' or, antibodies. They are immune system proteins that attack the body's own tissues. He says nearly all those with scleroderma have a positive ANA. When your body directs auto-antibodies against particular proteins, the doctors infer that you are suffering from systemic sclerosis or its variants.

After ANA, Dr Steele says the next step is an ENA test. ENA stands for Extractable Nuclear Antigen Antibodies. There is a mixture of 3 routine and around 15 non-routine tests. Dr Steele says some of the tests are complicated and need to be done overseas.

ESR - Erythrocyte Sedimentation Rate

ESR is a reliable test that measures how fast in millimetres an hour red blood cells sink down a measuring tube.

CRP - C reactive protein

Your liver produces CRP in response to inflammation caused by autoimmune and other conditions. Its levels rise and fall quickly, much more so than ANA's. ESR and CRP levels are higher in those with systemic sclerosis and a raised ESR is to be monitored closely.

The routine monitoring, the blood tests and doctor's visits, is, says Dr Steele, "Critical for maintaining well being." Monitoring ensures correct treatment.

Studies suggest two types of systemic sclerosis: inflammatory and fibrotic. Neither has a cure as yet. Regardless of the type, Dr Steele says his profession needs to balance the severity of the condition against possible

side effects of medication.

"The prognosis remains guarded when the condition is extensive and involves internal organs," Dr Steele says. In these advanced cases he says there is a 5-year survival rate of 90% of patients.

Although New Zealand's health system lacks resources, Dr Steele says international advances in research and treatment do filter through to here and they're adopted quickly.

"about one new case
per million people
each year"

Seminar reports

Adrienne Burleigh releases the results of her survey into your experiences with scleroderma and health services

There could be 1000 people with scleroderma in New Zealand with many unaware that they have one or other form of systemic sclerosis. That's not a large number so it's not surprising that many medical personnel have not ever come across the condition and do not have a great deal of information about it.

Adrienne's survey, conducted in October 2012, was responded to by people who have been told they have some form of systemic sclerosis. Space permits only a sample: the full survey is available on the support group's website at:

www.scleroderma.org.nz/scleroderma-patient-survey-october-2012/ or Adrienne can email to you: adrienne.burleigh@xtra.co.nz

Diagnosis

Scleroderma	# respondents	Notes
Limited	10	Two respondents have both diffuse and myositis (overlap)
Diffuse	9	
Other (please state)		
CREST	1	
Raynard's	1	
Sjogren's	1	
Total	22	

Number of times admitted to hospital for scleroderma related condition/s

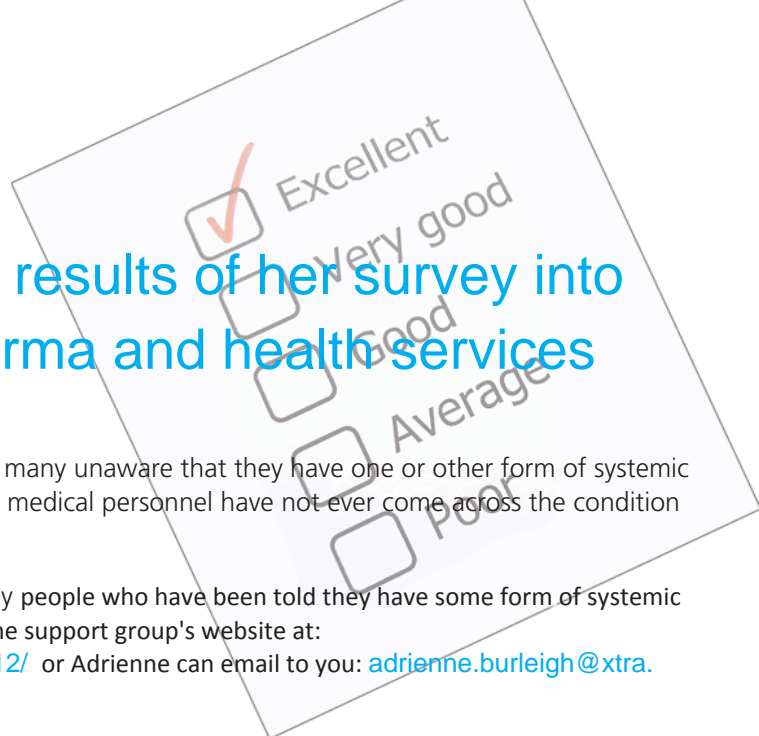
Admissions	0	1	4	10	12	More than 12
#	4	2	1	1	2	1

Rate your level of satisfaction with information received from each of these specialists on a scale of 1-10

Specialist	# responses	Range	Average level of satisfaction with information received
Cardiology	12	0-10	6.6
Dermatology	4	0-8	4.3
Gastroenterology	7	1-9	4.0
Immunology	1	-	8
Renal (Urology)	4	1-8	2.75
Respiratory	11	1-9	5.9
Rheumatology	18	1-10	6.2
Dental	3	2-7	5.0
Muscle biopsy	1	-	7
Neuro surgeon	1	-	8

No information received: 1

Not rated: 2



Survey summary cntd.

How would you rate your own personal knowledge of scleroderma and related conditions on a scale of 1-10 where 10 is the highest?

Scale	1	2	3	4	5	6	7	8	9	10
#Respondents	1	-	-	1	3	-	3	6	2	3

Respondents: 19

Range: 1-10

Average personal knowledge rating: 7.2

On a scale of 1-10, what level of encouragement do you receive to manage your condition from your care providers?

Care provider

GP

Scale	1	2	3	4	5	6	7	8	9	10
#Respondents	1	-	-	2	2	2	2	2	-	8

Respondents: 19

Range: 1-10

Average encouragement rating: 7.4

Specialist

Scale	1	2	3	4	5	6	7	8	9	10
#Respondents	-	1	-	2	4	3	2	6	-	3

Respondents: 19

Range: 2-10

Average encouragement rating: 7.4

Family

Scale	1	2	3	4	5	6	7	8	9	10
#Respondents	3	2	-	1	2	2	-	4	2	3

Respondents: 19

Range: 2-10

Average encouragement rating: 5.9

Comments

- Family ignore my condition – don't want to know. GP is most helpful and cheers me up.
- My GP is encouraging and supportive but even he sees me as a complex patient with many different conditions
- Don't need any encouragement
- I have learned to cope each day and am better than I was in the first 15 years
- I insisted that my specialist start me on 100 gms of minocycline which I take Monday, Wednesday and Friday. I have a noticeable change in skin hardness, lung function is stable, and hand use has improved.
- Counselling has helped. I have brave motto too
- Other: work 1 – they are terrible!! They just say look after yourself.
- After a number of years of gradual improvement my specialist classes me as "stable".
- It is a continuing learning pattern for all my family have concerns about my health and I try to ensure they are well informed. They now tend to be less concerned as they see me managing well in a partnership with my health providers

Remember that this is a survey sampler and you can find the full survey on our website

Seminar reports

Dianne Purdie explains the importance of battling cold when Raynaud's attacks

When it comes to comfort, the human body temperature thrives at 37 degrees Celsius. At that temperature, the main organs are protected from damage and operate at best efficiency. As the air warms and cools, the body accommodates itself to maintain 37 degrees.

As the body's environment cools, Dianne says a whole lot of things start happening. Receptors in the blood and on the skin tell the brain whether it's warming or cooling outside. Sufferers of Raynaud's, says Dianne

(and that's 95% of people who have scleroderma), have extreme reactions to cold.

When receptors defend against cold, Dianne says, their signals arrive at an almond-sized part of the brain called the hypothalamus, which is situated just above the brain stem. The hypothalamus controls body temperature, hunger, important aspects of parenting and attachment behaviours, thirst, fatigue, sleep, and circadian cycles (24 hour clock).

The hypothalamus, triggers some automatic and some voluntary responses. It calls on other parts of the brain, the cortex and limbic system to reduce heat loss. They are partly why you might rub your hands to generate friction when you're cold, pace and draw nearer a heater.

The body's metabolic rate increases and warms up until the crisis is over the and hypothalamus calms down and reduces its heat-producing signals.

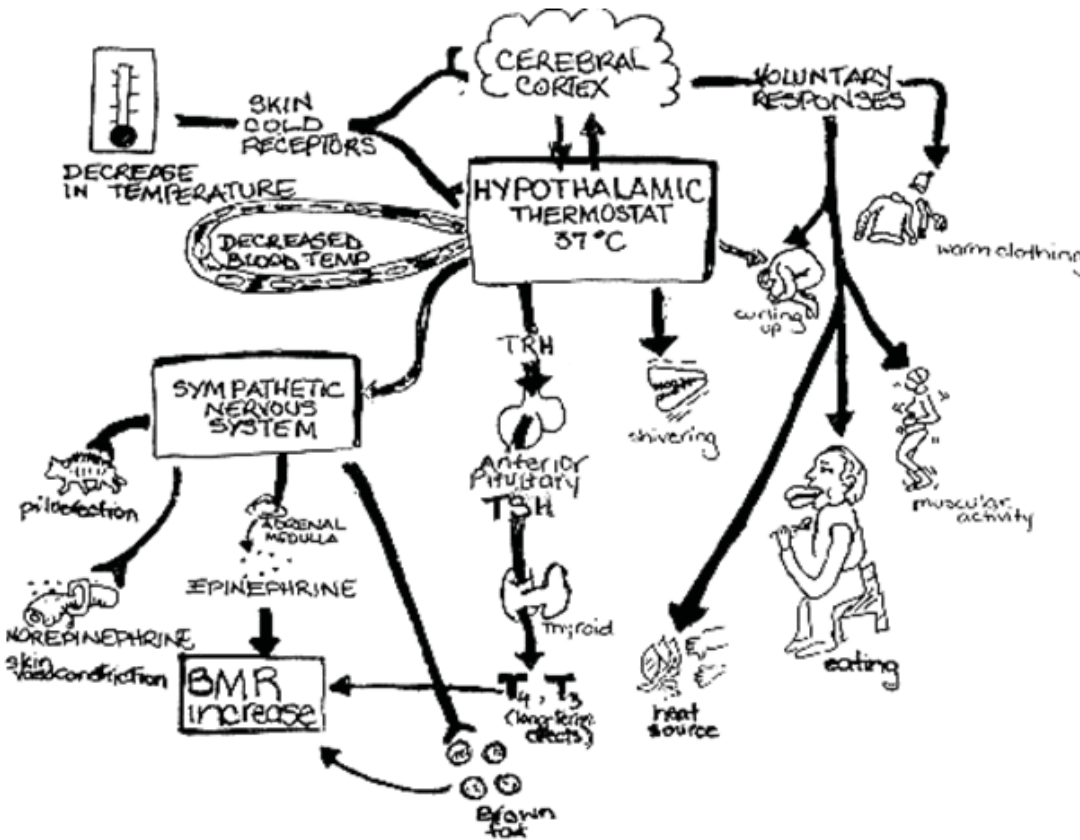
In scleroderma and with

Raynaud's in particular, Dianne says, blood vessels become damaged and narrow. The pattern of blood flow to extremities changes. First the cells that line the blood vessels become damaged and the basement membrane thickens. Then the middle thickens and finally the outside layer becomes excessively fibrous.

With narrowed blood vessels, Dianne says we are already a step ahead of people who do not have scleroderma and it doesn't take much to constrict our blood vessels when cold signals race off to the brain. Raynaud's phenomenon describes these episodic constrictions in the fingers and toes. In some people it may also affect the tip of the nose and earlobes. Exposure to cold triggers attacks or it may be even a smaller drop temperature, emotional stress, and vibration.

Raynaud's consists of two types: Primary which means no underlying disease and secondary which is a complication of scleroderma and other medical conditions. So it's possible to suffer from Raynaud's without scleroderma but if you have scleroderma, it's almost forgone that Raynaud's will appear too.

The secondary variety that goes hand-in-hand with scleroderma is clinically more severe than primary and tends not to develop in people under 30, However, it's more severe, attacks more often, is prolonged and painful.



continues next page

Raynauds *cntd.*

It is frequently associated with lesions caused by inadequate blood supply (ischaemic) and death of tissue in the fingers and toes.

Fighting Raynaud's

Dianne's advice is to get warm and stay that way. Use gloves before you feel cold. Warm your house to 22 degrees, even the loo and eat well and eat enough to keep your blood sugar levels constant.

Exercise daily to improve circulation and:

- Use thermal gloves, socks, and boots to prevent both the drying effects of weather and Raynaud's attacks - look for wool, sheepskin.
- Try technology warmers like silver gloves and silver socks
- Use hand and foot warmers - invaluable in cold weather
- Choose shoes and boots with thick soles
- Exercise a lot to improve blood supply to the skin

- Wear rubber gloves to protect the hands with thin cotton gloves inside them to prevent the skin sweating
- Use barrier creams and use skin cream after washing

If Raynaud's attacks:

- Place hands/feet in lukewarm water
- Rub hands/feet briskly
- Drink a large hot beverage
- Shake arms and legs
- Walk around indoors
- Put your hands in your armpits
- Curl your hands and blow on them
- Take a hot bath
- Get really warm all over

Seminar reports

Gordon Purdie on a possible link between solvents and scleroderma

Gordon began looking for a link between exposure to solvents and scleroderma after he noticed his wife, Dianne, who'd spent 25 years working in laboratories, had scleroderma and she had noticed that Raynaud's was common amongst her fellow workers.

After Dianne read though all the studies and their advice, Gordon says Dianne thought the evidence was strong enough to leave her career as a medical cytologist.

He found many studies showing an increased risk of scleroderma amongst solvent workers. Because scleroderma is rare, Gordon says, that the link needs to be studied with "case-control" studies.

He found at least 11 such studies into solvent exposure from 1989 to 2004. All found that people with scleroderma were more likely to have worked with solvents than those who

"Doctors noticed that some of the patients they were seeing had worked with solvents and thought their solvent exposure had caused their scleroderma"



did not have scleroderma. Averaged, they showed the risk of contracting scleroderma by working with solvents was between 2 and 3 times greater than for those who did not work with solvents. Several of the studies showed that the more people work with solvents the more likely they are to get scleroderma.

Gordon says the evidence is strong that exposure through work to solvents makes people susceptible to scleroderma. Even removing results from 3 questionable studies still produces a relative risk of 2.1 greater likelihood.

Gordon gave the example of a South Australian woman, Yvonne Hazeal, who was sprayed accidentally with a weedkiller, Tordon 50D, which contains a solvent. Within days she developed scleroderma-like symptoms and was confirmed

Exposure to solvents *cntd.*

with it in a few years.

Gordon's conclusion: it looks like solvents cause scleroderma.

Minimising solvent exposure has little cost and is probably risk free. Although there is no evidence, Gordon says, that it is possible that by avoiding solvent exposure your health might be better than if you don't.

They are difficult to avoid as they're used in petrol, beauty products, nail polish and remover, hair dye and even alcohol is classed as an organic solvent. Wellington Regional Council found the following airborne solvents when it tested air quality at Seaview: Toluene, xylene, benzene, ethylbenzene, pentane, trichloroethene, 2-butanone, 1,3,5-trimethylbenzene, acetone, hexane, n-propylbenzene, trichlorofluoromethane.



In summary, Gordon told the seminar:

- There is good evidence that solvents cause scleroderma
- No evidence on whether or not avoiding solvents will help if you have scleroderma
- Very difficult to avoid solvent exposure

Just relax...

Staying calm through stress, illness or excitement can be learnt and at the seminar, Manik Batish gave a small taste of just how relaxing stress management can be. He quoted the World Health Organisation as saying stress can cause up to 50% of illnesses. Manik says you deal with stress most effectively with a calm mind. He took



attendees through some basic stress management techniques and soon had them smiling and stress-free.

Manik is a volunteer with Art of Living, which works in 30 countries teaching

meditation and stress management.

He says the key to managing stress is that it should be effortless: achieve the rhythm of relaxation and you will relieve stress. The method of teaching that Manik follows is explained fully at the New Zealand branch's website:

www.artofliving.org/nz-en

Noticeboard



World Scleroderma Day
29th June Spread the
word and tell one person

Adrienne has something special for 29 June. Come to the usual meeting rooms anytime between 9.30 and 4pm Lunch at Dowse Art gallery, but bring morning and afternoon tea.

Anyone want to know how to make earrings (for pierced ears) Barbara will show you how at the Scleroderma Day rooms 29 June. Bring some beads and she will bring the rest.

We need to do some fund raising to cover costs of running Scleroderma NZ,

If you have any practical ideas please let us know we would love to hear them.

Next meeting date is August 10th.
Usual place
Te Awakairangi building
cnr Laings and Myrtle Street
Lower Hutt

Hamilton Seminar will be held on the
29th of June

Contact Linda Bell for details:
07 8535434
linda.bell@hotmail.co.nz



They're all ears: Manik reduces their stress levels



Graham and Heather Milligan flew all the way from Southland to attend



Kapiti and Palmerston North flew the flag with Maree Drogemüller, Chris Carlyon and Catherine Thompson



Dianne Purdie, seminar organiser, compere and presenter at work.



A seminar, it seems, marches or sits on its stomach and there was plenty to choose from at lunchtime

CONTACTS

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Links: www.arthritis.org.nz | www.scleroderma.org.nz | www.sjogrensnewzealand.co.nz