Raynaud's Phenomenon in Scleroderma Treated with Hyperbaric Oxygen

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(for G B Dowling FRCP)
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Miss P C, aged 46. Clerk
History: Generalized itching for ten years and signs of scleroderma with pigmentation on the trunk for over four years. She had had typical Raynaud's phenomenon of hands and feet for four years, but there had never been any sclerosis or œdema of the hands, although about half a dozen macular telangiectases can be seen.

Investigations: Barium swallow normal; standard tests for systemic sclerosis and connective tissue disorders all negative.

Effects of hyperbaric oxygen on bone-marrow function (Dr P Crome): The rate of clearance of ^{59}Fe citrate from the plasma was measured immediately before and after the first treatment with hyperbaric oxygen. The readings shown in Fig 1 suggest that the rate of hæmopoiesis may be temporarily slowed but more work is being done to test the significance of these findings.

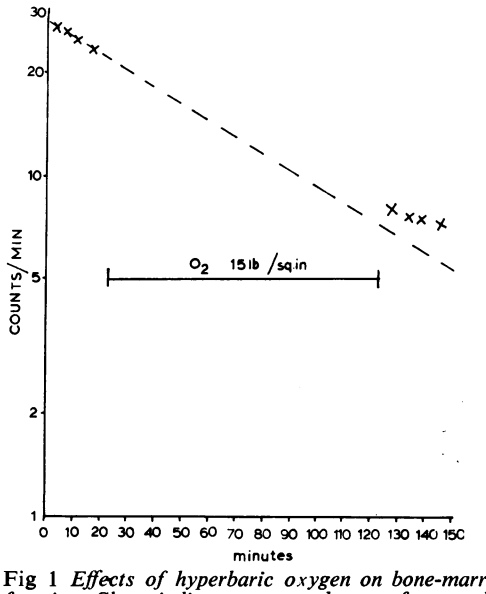

Fig 1 *Effects of hyperbaric oxygen on bone-marrow function. Slope indicates expected rate of removal of* ^{59}*Fe* (x x)

Comment
The patient is presented for the following reasons:
(1) To emphasize that generalized itching seems to be a common precursor of scleroderma and dermatomyositis. Three of our last 7 patients with scleroderma and 2 of the last 3 with dermatomyositis have had puritus for one to five years before these diagnoses were made.
(2) To invite comment about the diagnosis as to whether this is generalized morphœa with Raynaud's phenomenon or systemic sclerosis with Raynaud's phenomenon but without apparent alteration of the skin of the hands or other stigmata of systemic disease.
(3) To mention the benefits of hyperbaric oxygen in scleroderma patients with ischæmic symptoms and signs.

Hyperbaric Oxygen
Six women between the ages of 46 and 71 (Table 1) were treated for ten to fourteen days with oxygen at 2 atmospheres absolute (which gives a Po_2 of approximately 1,400 mmHg) at two sessions a day each of two hours. The symptoms of Raynaud's phenomenon were improved, and in all cases the improvement lasted for longer

Table 1
Diffuse systemic sclerosis: 6 women treated with hyperbaric oxygen

Case No.	*Age*	*History (years)* SS	R	*State of fingers*	*Sympathectomy (years ago)*	*Result*
1	46	5	5	Œdema, acro., nec.	1	Improvement for 3 months then relapse and O_2 unhelpful
2	56	30	30	Acro. + +, nec. + +, loss of terminal phalanges	30	Improvement for 2 months – painless mobility – Raynaud improvement
3	70	30	40	Acro. + nec. +	2	Greatly improved painless mobility, ulcers healed. Two acute admissions: (*a*) Fingers; pain + + (septic gangrene, bone necrosis). (*b*) Right leg; pain and ischæmic ulcer
4	51	2	2	Acro. +, œdema +	–	Improved mobility and comfort, Raynaud better
5	62	4	4	Acro. + +	–	Improved mobility
6	71	?	3	Œdema, nec. +	–	Finger ulcers healed

SS = diffuse systemic sclerosis. R = Raynaud's phenomenon.
Acro. = acrosclerosis. Nec. = necrosis and ulceration of skin

than a month. The 5 with stiffness of the skin all volunteered that the skin was more mobile. The 4 patients with painful ulcers, ischæmic and infected, were relieved of pain within forty-eight hours and the ulcers healed in one to three weeks which was far more rapidly than was to be expected, as for example in Case 3 who had complete loss of the terminal phalanges.

We were surprised at the prolonged effect of the treatment. We cannot explain our findings but wonder whether the increased oxygen supply to the arterial wall resulted in less spasm and whether the capillary oxygen beyond the arterial blocks was sufficiently raised to improve materially the oxygenation of the tissues. The method is well established and safe. The only complication which we encountered was a transient myopia lasting three weeks in one patient. Notwithstanding the success of the treatment we advocate that this time-consuming method which needs special skill and equipment should be kept for acute episodes in the disease.

Dr G B Dowling: I find this case interesting as a possible example of the 'systemic sclerosis' type of scleroderma in which the skin change has appeared primarily on the trunk. Otherwise you must assume that Raynaud's phenomenon appearing first well in middle age and scleroderma at about the same time are disconnected events.

Dr R E Church: What is the temperature inside the hyperbaric oxygen chamber and has Dr Copeman tried the effect of enclosing patients with scleroderma in the chamber without giving hyperbaric oxygen?

In Sheffield we have a room in which the temperature can be controlled and we have treated a number of patients with scleroderma by keeping them at a constant temperature of about 85°F(29°C). The effects are most striking in the relief of pain in digital ulcers as the pain disappears in about 24 hours and there is an improvement in their skin condition which lasts for several months.

Dr E J Moynahan: We have used hyperbaric oxygen for these cases and also for stasis ulcers at Guy's Hospital, but have abandoned it, as we found in the case of the latter that exposure of the ulcer to oxygen in a polythene bag was more effective.

I am a little surprised that your patients are allowed to read in the chamber and I would expect a catastrophe like the recent one at Cape Kennedy, especially as the pressure of oxygen inside the chamber is far higher than that in the Apollo spacecraft.

Dr P W M Copeman: Regarding the temperature inside the Vickers Hyperbaric Oxygen Bed, this is maintained at the same level as the surrounding air, and hyperthermia is not part of the treatment. As regards putting patients inside without the oxygen, we thought this would be unjustified until there were a reasonable number of cases who had improved with the oxygen. On the point of reading in the chamber, this is perfectly safe, and the accident at Cape Kennedy was not caused by it.

Systemic Sclerosis of the Small Intestine

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(for Professor C D Calnan FRCP)
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Mrs J C, aged 56. Housewife

History: In 1961 she was investigated for right-sided abdominal pain and backache. A uterine fibroid was found and her symptoms attributed to this. Soon afterwards she started to get Raynaud's phenomena in all four limbs and noticed that her face, arms and hands were swollen. In February 1962 she had an abdominal hysterectomy. Later in the same year she had joint pains and when seen in October slight but definite sclerosis was present in the skin of her face, forearms and hands. Systemic sclerosis was diagnosed. A group of about twelve shiny, flat-topped, mauve papules, each 0·5 cm in diameter, then evolved on her forearms. These showed the histological changes of scleroderma with a marked cellular infiltration by eosinophils and lymphocytes. In May 1963 her ESR reached 70 mm in 1 hour (Westergren) and her plasma globulin 4·1 g/100 ml. In December 1963 abdominal pain after meals returned and after a few weeks she started to vomit small quantities of bile-stained fluid. Examination of her abdomen showed guarding and rebound tenderness maximal in the right iliac fossa. There was a wide-necked incisional hernia resulting from her hysterectomy. Straight X-ray of the abdomen showed a distended large bowel and a diagnosis of incomplete obstruction was made. She was treated expectantly with intravenous fluids and suction. Barium enema two weeks later showed no abnormality.

In February 1964 water brash was troublesome and in April diarrhœa started three or four times a day, usually after meals. By June she was again getting abdominal cramps in her right side. Barium swallow showed normal œsophageal peristalsis and a hiatus hernia. In October 1964 she had dysphagia for solids. Her incisional hernia was repaired in April 1965. In May diarrohœa returned and she was again passing three or four watery stools a day with some mucus. She vomited occasionally. This lasted for two months. In April 1966 she had a further attack of diarrhœa. Her ESR had now reached 128 mm in 1 hour (Westergren). X-rays of the hands showed no bone absorption or soft tissue calcification. Manometry showed œsophageal aperistalsis. In December 1965 the diarrhœa restarted with mucus and a little fresh blood.

In March 1967, at routine outpatients attendance, she complained of severe abdominal pain; signs of peritonitis were found and she was referred for a surgical opinion.

On examination: Apyrexial. Pulse rate 72/min.