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Book reviews

La Sclerosi Sistemica Progressiva. By Mario Giordano. (Pp. 150; illustrated + tables. Lire 8000.) Edizioni Luigi Pozzi: Rome. 1977.

The official birthday of systemic sclerosis was 1753 when Carlo Curzio, a Neapolitan physician working at a Hospital for Incurables, was the first accurately to describe a patient with scleroderma, although Zacutus Lusitanus had published an incomplete description of the disease several centuries before. The name 'scleroderma' was suggested by Gintrac in 1847 and has persisted although 'progressive systemic sclerosis' (PSS) introduced by Grotz in 1945 better reflects our acceptance today of the multisystem involvement. Nevertheless, the adjective 'progressive' seems inappropriate because in the majority of patients the disease evolves slowly or remains static for long periods and in only a few cases does it proceed rapidly to a fatal outcome. These are some of the points made at a symposium on scleroderma held in Venice in 1976, the proceedings of which have been edited by Dr Giordano.

Among the systemic diseases of connective tissue PSS is roughly as frequent as systemic lupus erythematosus, while in the population as a whole the annual incidence of onsets of PSS varies between 2.7 and 12 per million. In the Campania region around Naples, PSS appears to be the most prevalent of the connective tissue diseases and within this region the prevalence is greatest in the district of Irpinia. The sex incidence also varies throughout the world, varying from 2.9 to 9.4 women for every man affected.

Under the electron microscope dermal collagen is normal. However, polarised light microscopy shows widening of the interspaces between fibres and blurring of the outline of the fibres, producing an appearance of homogenisation. Physically, the compliance of sclerodermatous skin is reduced whilst chemically measurements of hydroxyproline in sclerodermatous skin by many workers has produced inconsistent results but it seems that there is an increase in 3-hydroxylysine associated with an increased amount of hydroxylase and of procollagen hydroxylase, especially in rapidly progressive disease.

There is, nevertheless, evidence that young collagen is being synthesised as shown by the electron microscopic (200-400 Å) appearances and the solubility in saline, whilst studies with labelled proline show increase in incorporation by the fibroblasts. There is also an increase in urinary hydroxyproline and in serum collagen-like proteins. In the skin there is an increased level of reducible crosslinks particularly in the initial phases of the disease.

Increased synthesis of young collagen is found in the subcutaneous and adipose tissues (100-400 Å). Tissue cultures of fibroblasts from affected skin produce large quantities of soluble collagen and glycoprotein. Serotonin stimulates this fibroblast activity, but the question of whether PSS fibroblasts activity is quantitatively or qualitatively different is not yet settled. Certainly PSS fibroblast morphology, whether in light phase contrast or electron microscopy, is normal and their growth in culture is also normal. There is surprisingly little evidence of any causal or mediating immune mechanism in this disease. Various serological abnormalities have been described but none of them are constant or specific. For example, in one series of 40 patients, there was a slight increase of IgG. Complement was reduced in a few, but positive titres of rheumatoid factor were found in 50%. Antibodies against thyroid, gastric parietal cells and cell nuclei have occurred in between 36 and 87% in various series.

A mild lymphocytic infiltration of the skin is also described.

Raynaud's phenomenon, telangiectasia, and various angiopathies are common features. They are discussed in detail but the important question of whether the disease is the cause of the result of the vascular abnormalities remain unanswered.

The largest part of this book is devoted to highly detailed descriptive reviews of organs and tissues affected by PSS in which the pathological, radiological, and immunological features are related as far as possible to the clinical presentation. Reproductions of x-rays, ECGs, and colour photographs abound but it is only in the final few pages that perspective is regained, prognosis being dealt with in 2 pages and treatment in 5.

One author quotes benefit in Raynaud's phenomenon, dysphagia, and joint problems in a personal series of 50 patients treated with griseofulvin. D-penicillamine is advised only at the onset of the disease. Immunosuppressive therapy does not help and corticosteroids are still controversial. Combined therapy with aldosterone, spironolactone, and potassium and magnesium aspartates was reported to benefit the Raynaud's phenomenon, the skin lesions, the joint and muscle manifestations, the abnormalities of oesophageal motility, and the cardio-pulmonary complications. Progesterone therapy is also put forward as increasing collagen breakdown with consequent improvement in cutaneous and visceral manifestations. Other treatments discussed include salazopyrin, piacledine (an extract of avocado pears and soya beans), ethelene diamine tetra-acetic acid, potassium amino benzoate, and colchicine.

Mixed connective tissue disease and eosinophilic fasciitis and the conception of the Crest syndrome are all discussed in the differential diagnosis. A new nomenclature for PSS based on the distribution of affected fibrous tissue is proposed.

A very useful feature of this book is a bibliography of no less than 506 references, most of them published in the last 20 years, indicating the relatively recent explosion of interest in this, the most mysterious of the systemic diseases of connective tissue.

J. GRABER

The Rheumatic Diseases of Childhood. *Proceedings of the 1st ARA Conference, Vol. 20, March, No. 2, Supplement.* Edited by Jane G. Schaller and V. Hanson. (Pp. 491; illustrated + tables. US \$ 15.00, soft covers.) Arthritis Foundation: Atlanta. 1977.

This book represents the proceedings of a meeting held in March 1976, to which many prominent physicians contributed. The contributors are largely American and the views expressed and subjects chosen reflect this selection. There are chapters, based on extensive personal series, covering most of the connective tissue diseases. It is for this accumulation of material that this book is primarily of value. Within its 491 pages will be

found much of the present knowledge on the topics covered, including in certain cases the conflicting views.

Without exception the authors have kept to the point and wasted words are few. Some of the clinical descriptions are rather formal and stereotyped and the clinical picture of the disease does not come across well. Perhaps this is due to the papers being delivered to a peer audience rather than for wider consumption, the presenters forgetting that the diseases they describe may be unfamiliar to some members of this wider audience.

In addition to discussion of the principle diseases, unusual diseases and presentations are detailed and there is discussion of the attempts to define disease subgroups. The discussion following each section, although readable, would probably have been enhanced if a rapporteur system had been used.

The clinical aspects have been enhanced by the inclusion of chapters on background subjects, of which one might pick out those by Omenn on genetics and histocompatibility genes, Phillips on pathogenic mechanisms of infectious agents, and amyloidosis by Schnitzer and Ansell.

Interesting chapters deal with specific problems such as growth retardation and eye and cardiac involvement.

There are faults. It is often not clear when authors are extrapolating from results found in adults and when they base statements solely on childhood cases. There are surprising omissions such as scant reference to tenosynovitis and the enthesopathy of ankylosing spondylitis, both of great practical importance in diagnosis and management of these cases. The psychological aspects of childhood rheumatic diseases and their management are not discussed, nor are the dental complications. Haemophilia, the hereditary disorders of connective tissue and soft tissue disorders, and the many postural and orthopaedic problems are not dealt with.

It is because of the emphasis on quantitative facts over a limited range of the spectrum of disorders seen in a children's arthritis clinic and the lack of practical advice that this book cannot replace a textbook (nor was it meant to). As complementary to such a textbook it would be excellent. Unfortunately there is no textbook covering childhood arthritis at present. Perhaps the organising committee might think of inviting some

of their authors to reorientate their presentations, add other chapters, and produce the prototype textbook of childhood arthritis. Until that time this book should be freely available in every hospital dealing with these diseases. The price is highly competitive by today's standards.

P. J. L. HOLT

Peripheral Manipulation. 2nd edition. By G. D. Maitland. (Pp. 363; illustrated + tables. £13.50.) Butterworth: Sevenoaks. 1977.

This book, written by a manipulative physiotherapist, pairs with one by the same author on vertebral manipulation. I was able to review the latter favourably because experience had taught me that the common spinal mechanical syndromes can be easily recognised and do sometimes respond dramatically to empirical manipulative treatment, without needing to determine exactly the nature of the lesion. Similar lesions undoubtedly occur in peripheral joints as a late result of trauma, inflammation, or degenerative change, but I cannot accept the implication that passive manual procedures are the main key to treatment.

In the introduction Mr Maitland states that 'diagnosis will not be discussed as this is the province of the medical practitioner'. However, a large proportion of doctors referring patients for treatment are not skilled in orthopaedic or rheumatological diagnosis and physiotherapists must be constantly attuned to locomotor diagnosis. It cannot be sufficient to assess only pain and stiffness in the joint and set out to correct these by mobilising without constantly questioning the underlying cause. For example, pain and stiffness could arise in a hip with unremarkable x-rays from osteomyelitis, rheumatoid, osteoarthritis, aseptic necrosis, or slipped epiphysis.

The author is guiding his less experienced colleagues up a very narrow pathway. Although he is undoubtedly aware of the value of ancillary treatment—such as muscle re-education, heat, ice, and friction—these are not mentioned. The tenor of the book is such that it will tend to produce a cultist manipulative physiotherapist who could devote too much time to the elaborate mobilising procedures without utilising other important techniques.

D. A. H. YATES

International Co-ordination of Drug Trials. Rheumatological Research and the Fight against Rheumatic Disease in Switzerland. Edited by K. Fehr, E. C. Huskisson, and E. Wilhemi. (Pp. 290; illustrated + tables. Sfr. 32.00, paperback.) Monograph Series, No. 1. EULAR Bulletin: Basel. 1977.

The 30th Anniversary of the European League against Rheumatism was celebrated by a meeting in Zurich in April 1977. The papers and discussion are published in the first of a monograph series by EULAR Bulletin through the generosity of Merck, Sharp, and Dohme, Zurich. Almost two thirds of the monographs taken up by the first part of the meeting entitled 'The International Co-ordination of Drug Trials', a subject of great interest to the pharmaceutical industry and to many clinicians. Thirty-eight papers, almost exclusively from centres in Europe, were presented in 4 sessions grouped under the headings 'Methodology', Drug safety and effectiveness', 'What the health authorities require from clinical trials', and 'Design of protocols and other practical problems'.

The papers vary in quality, length, and clarity and, like many other conference proceedings, they fall short of the standard of formal papers for rheumatology journals; they can only hint at the valuable exchange of ideas that probably took place. Despite some editing there is repetition, particularly in measurement and trial design.

With international co-ordination it is hoped that reduplication of trials will be avoided but, on the other hand, clinicians have faith in the results when they know the centre where the trial was performed. Seldom is reference made to the researching experience or working conditions of the person carrying out a clinical trial. Standardisation of trials has hazards as well as advantages. There are thoughtful contributions at the beginning from Dr M. F. Grayson and at the end from Dr A. St. J. Dixon. Dr Grayson points out that although studies must be conducted in many places by many investigators in a wide variety of conditions the 'large and hectic multicentre studies now so popular in drug evaluation must be seen to be the juggling act and conjuring tricks that they are'. Dr Dixon explains the positive ethic in attempts to improve methods of treatment and in studying a new drug efficiently, but advises that, when considering a trial, the researcher should decide whether the company is interested in his opinion or whether he is taking part in a promotional exercise disguised as research.