

ATLAS OF SARCOIDOSIS



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Pathogenesis, Diagnosis, and Clinical Features

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“What the mind does not know, the eyes cannot see”
—*Johann Wolfgang von Goethe*

PREFACE

This *Atlas of Sarcoidosis* is designed to complement and provide a visual supplement to already existing excellent texts on pulmonary medicine and sarcoidosis. Medical students, postgraduate candidates, general practitioners, internists, pulmonologists, dermatologists, and practitioners of many other disciplines of medicine who have to treat sarcoidosis patients will find the book useful and rewarding. In addition, paramedical personal such as nurses, physician's assistants, and respiratory therapists will enjoy browsing the book.

Whatever the speciality of the reader, he or she must learn to obtain a careful history and perform a thorough physical examination. The next step is to link these observations with pertinent radiographic and laboratory information. If this amalgamation is not achieved the treatment of the patient will remain incomplete and ineffective. The atlas aspires to facilitate this process.

Sarcoidosis is a complex multisystem disease. The lungs are the most commonly involved organs by sarcoidosis, but no structure of the body is immune to its ravages. Each organ involvement is dealt in a brief and easy to comprehend manner. Various radiographic and laboratory abnormalities are then linked to the clinical features in order to encourage a smooth and easy integration at the bedside. Finally it is worth remembering that the Atlas is not a repository of all that is known about sarcoidosis. Its goal is to provide the reader a tantalizing visual interpretation of a fascinating and mysterious illness.

Violeta Mihailovic-Vucinic
Om P. Sharma
November 2004

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The special photographic work of Mr Josip Chanady speaks for itself.

The authors are particularly grateful to the whole medical staff of the V Th Clinical Department- Institute of Pulmonary Diseases, Clinical Center Belgrade, and Keck School of Medicine, LAC-USC Medical Center Los Angeles, CA, USA who helped me with this work.

The authors also appreciate all sarcoidosis patients for their assistance. They are permanently teaching us about many faces of this mysterious disease.

*Violeta Mihailovic-Vucinic
Om P. Sharma*

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CONTENTS

	Preface	vii
	Acknowledgments	ix
Chapter 1	The Pioneers of Sarcoidosis	1
Chapter 2	Pathogenesis	5
Chapter 3	Diagnosis of Sarcoidosis	9
Chapter 4	Clinical Features	17
Chapter 5	Laboratory Investigations and Immunologic Testing	21
Chapter 6	Pulmonary Sarcoidosis	25
Chapter 7	Pleural Involvement in Sarcoidosis	43
Chapter 8	Sarcoidosis of the Skin	47
Chapter 9	Ocular Sarcoidosis	61
Chapter 10	The Parotid and Other Salivary Glands and Lachrymal Glands	63
Chapter 11	The Spleen in Sarcoidosis	65
Chapter 12	The Liver in Sarcoidosis	69
Chapter 13	Renal Sarcoidosis	73
Chapter 14	Sarcoidosis of the Heart	77
Chapter 15	Neurosarcoidosis	83
Chapter 16	Bone Sarcoidosis	93

Chapter 17	Endocrine Involvement	103
Chapter 18	Gastrointestinal Tract in Sarcoidosis	105
Chapter 19	Sarcoidosis of the Upper Respiratory Tract	107
Chapter 20	Sarcoidosis and Malignancy	109
	Index	113

CHAPTER 1

The Pioneers of Sarcoidosis

More than a century ago, Sir Jonathan Hutchinson (1828–1913), who was a surgeon dermatologist born in Selby, Yorkshire, England, identified the first case of sarcoidosis at King’s College Hospital, London, England (Figures 1.1 and 1.2). Because of his wide range

of interests covering most medical and surgical specialties, Hutchinson became one of the most renowned consultants of his time. Considered for decades an exotic illness, sarcoidosis has now blossomed into a commonly seen multisystem clinical syndrome.

FIGURE 1.1 Jonathan Hutchinson caricatured elegantly by Spy in *Vanity Fair*.¹



FIGURE 1.2 The first patient with sarcoidosis described by J. Hutchinson had multiple, raised, dusty-red patches on his feet, fingers, and arms.



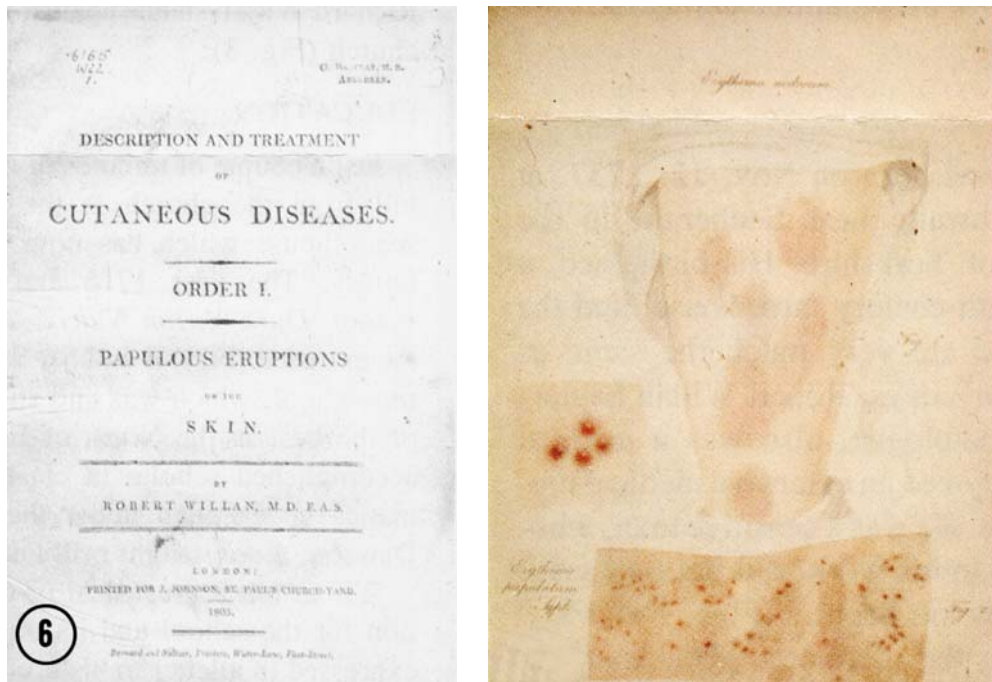


FIGURE 1.3 Robert Willan's *Cutaneous Diseases* was the first authoritative dermatology text.

The term erythema nodosum was first introduced by Robert Willan (1757–1812), generally considered to be the father of modern dermatology (Figure 1.3). He described the lesion as elevated, painful, red protuberances on the legs, occurring mostly in female servants. His description published in 1798 (Figure 1.4) remains a classic of dermatology to this day.² Much of the current terminology of skin disease can be attributed to Willan, who was also the first to use the term wheal for skin lesions that occur in nettle rash.

Ernst Henri Besnier (1831–1909) was the first to report lupus pernio. He described the skin lesions on his patient's face as lupus pernio de la face-synovites fougueusses (scrofulo-tuberculoses) symetriques des extrémités supérieures. The term biopsy—, in its more archaic “biopsie,” is said to have been introduced by him.

Caesar Peter Moller Boeck (1845–1917) was born in Norway and described skin lesions of a patient with lymphadenopathy as lymphoma cutis multiplex/multiple benign sarcoid of the skin. Jorgen Schaumann (1879–1953) was the first to report systemic sarcoidosis, calling it lymphogranulomatosis benigna (Figure 1.5). Sven Löfgren (1910–1978) a Swedish physician, was the first to link erythema nodosum with sarcoidosis (Figure 1.6). The association is now called Löfgren's syndrome.

FIGURE 1.4 Robert Willan (1757–1812).





FIGURE 1.5 Jorgen Schaumann (1879–1953).



FIGURE 1.6 Sven Löfgren (1910–1978).

Louis Eliot Siltzbach (1906–1980) established the specific diagnostic value of the Kveim test in 1954, which is now, appropriately called the Kveim Siltzbach test (Figure 1.7).

Carol Johnson Johns (1923–2000) was the first woman to organize an International Conference on Sarcoidosis (Figure 1.8). The conference, held at the Johns Hopkins Medical School in Baltimore in 1984, was attended by 305 delegates from 29 countries.³

In Japan, Keitzo Nobechi (1890–1978) was the first to report the uneven geographic distribution of Japanese cases of sarcoidosis (Figure 1.9). In 1960 he suggested a higher prevalence of the disease in the north of Japan than in the south. He graduated from the University of Tokyo School of Medicine in 1919 and, within seven years, he was awarded a Doctorate of Public Health from Harvard University. He worked on the epidemiology of infectious diseases and studied cholera in cooperation with the World Health Association.

FIGURE 1.7 Louis Eliot Siltzbach (1906–1980). This charcoal drawing was done by his wife Hansi Bohm.



FIGURE 1.8 Carol Johnson Johns (1923–2000).





FIGURE 1.9 Keitzo Nobechi (1890–1978).



FIGURE 1.10 Dr D. Geraint James, Founder President WASOG.

Today, information is available through WASOG, the World Association of Sarcoidosis and Other Granulomatous Diseases. The Founder President is D. Geraint James (Figure 1.10). At present, the President of WASOG is Om P. Sharma.

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