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CHARCOT JOINTS

With a Foreword by PHILIP D. WILSON, M.D., Surgeon-in-Chief, Emeritus, Hospital for Special Surgery, New York, New York. A compendium of clinical knowledge on the Charcot joints . . . the only single source of such material in the modern literature. Controversial opinions are kept at a minimum. Emphasis is placed on factual clinical observations.

Throughout the entire book the interest of the reader is stimulated and provoked by "the maverick behavior" of the disease entity known as Charcot joint.

A general consideration of the subject is separated from examples of the disease as it involves various joints of the body. Pertinent differential diagnosis is included as a single subject and emphasized repeatedly by comparative examples throughout the text.

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CHARCOT JOINTS

Jean-Martin Charcot
(1825-1893)
(Courtesy of Dr. Harold Wainerdi)

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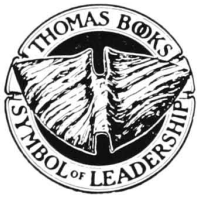

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Dedicated

to

Alice Anne

my wife, for her patience and understanding

and to

my parents

who made this monograph possible

Foreword

When Dr. Sidney Eichenholtz, the author of this book, first mentioned to me some years ago his interest in Charcot joints I could not have been less sympathetic. To me there was something frustrating or defeating about the very name, and this doubtless was born from my feeling that no new knowledge of the condition had been added since Charcot's original description, and, therefore, that nothing could be gained from any fresh study. But Dr. Eichenholtz could not easily be dissuaded from his interest in this condition. First of all he is an orthopedic surgeon and therefore concerned with all manifestations of joint pathology, and secondly for many years he had been connected with one of the Veterans Administration Hospitals where until a few years ago he had served as a full time Chief of the Orthopedic Service. In this role he had been called many times for consultation on patients who were disabled by Charcot joints and they had aroused his sympathy and curiosity. He is a man with the courage of his convictions and likes walking in fields where the paths are not charted. With admirable persistence he collected his observations and began making this material available for the information of his colleagues through the medium of scientific exhibits at orthopedic meetings. His material accumulated and he was able to draw some interesting deductions and conclusions. The expressions of interest that he received from the many orthopedic surgeons who visited his exhibits made him feel that it was desirable to publish the material and make it available for the information of his colleagues. I have reviewed his manuscript and found it full of interest and can recommend it to others.

At the same time he has made an extensive study of the existing literature on the subject and has become aware of its deficiencies. It was on these grounds that he has reached the conclusions that there is a need for new work on this subject and that, by including contributions on special types of neuro-arthropathy, he can bring the subject up to date and contribute something new and useful to our knowledge.

The paucity of scientific literature on Charcot joints is indeed strange. Almost nothing has been added to the description of the condition or the natural history of this disease since Charcot's time. Efforts have been made to explain the etiology of the disease on neuro-pathological grounds

and considerable reference to the condition has been made by those trying to prove the trophic function of nerves. The absence of study is probably accounted for by the lack of any method of treatment of the condition and the acceptance by physicians of the fact that it was a recognized complication of tabes dorsalis and that it was best prevented by early diagnosis of syphilis and intensive treatment to cure the disease before involvement of the central nervous system had occurred. To be sure the occurrence of neuropathic joints in syringomyelia was recognized and documented, and more recently the occurrence of similar pathological conditions affecting the distal joints of the lower extremities in diabetes mellitus has been described. Similar pathological changes in the joints of the hands and feet have been recognized in leprosy or Hansen's disease. But there have been no publications describing Charcot joints or their evolution in tabes dorsalis.

It is only the orthopedic surgeons who have kept the literature alive during the last twenty or more years with their reports of efforts to treat these joints and stabilize them by means of the operation of arthrodesis or fusion. Their interest was centered on the development of successful operative procedures and technical improvements, review of their results, and their percentages of failures or successes, but not upon the evolution of the disease itself or any attempt to catalogue the differences between the joints conditions that were treated. Their reports have contributed to our knowledge of therapeutic methods, but not to our knowledge of the condition.

With the improvement of anti-luetic treatment and the diminished frequency of central nervous system involvement the occurrence of Charcot joints has become less and less frequent. At the same time we cannot afford to let our guard down as the public authorities tell us there has been an upsurge in the occurrence of syphilis in recent years. However, the long range forecast for syphilis, tabes dorsalis, and Charcot joints is reassuring. Only recently Surgeon General of the Public Health Service, Luther Terry, has predicted that syphilis will be among a number of infectious diseases which will be eradicated from the population of the United States in the next twenty-five years. Even then we will continue to encounter Charcot joints in syringomyelia, diabetes mellitus and probably also in other uncategorized conditions. We can, therefore, welcome any further additions to our knowledge of Charcot joints.

PHILIP D. WILSON, M.D.

New York, New York

Preface

Paris, France, was indeed fortunate to have been the city wherein Jean-Martin Charcot was born on November 29, 1825. His association with the Salpêtriere created the most famous neurological and psychopathological clinic of the nineteenth century, and one which even by modern standards could compete successfully with any similar clinic in the world today. The very foundations of modern psychopathology may be attributed to his scientific approach in the fields of hypnotism and hysteria. His propensity for scientific study produced reports of amazing accuracy on such widely diverse medical subjects as gout, arthritis, diseases of the nervous system, lead intoxication, and diseases of the lungs, liver, kidneys, and joints. The names of those who studied with this brilliant teacher are familiar to any neophyte in the medical sciences. These include Bauchard, a co-investigator with Curie of radium emanations, Hanot of Hanot's cirrhosis, Marie of Charcot-Marie-Tooth disease, Marie-Robinson and Marie-Strumpell's disease, Bechterew of Bechterew-Strumpell's disease, Sachs of Tay-Sach's disease, Freud, and Babinski. When he died in 1893, a giant among other medical geniuses such as Dupuytren, Laennec, Pasteur, Claude Bernard, and Duchenne, he well deserved the title of "Charcot-Caesar of Salpêtriere." Later medical historians call him "the most brilliant physician of his day in France." One cannot embark upon a study of Charcot joints without being overwhelmed by the intellectual achievements of the man for whom they were named. Much confusion persists concerning the nature of the joint changes described by him although comparatively little has been added to his description of the disease.

Interest in the subject was stimulated by the controversial statements made in the literature relative to all aspects of the disease. One investigator believed that the condition of Charcot joints was similar to osteo-arthritis "only more so." Similarly, much confusion arises from a consideration of pathological changes frequently found in the hip joints of paraplegic patients. These joints may become quite disrupted, dislocated, and degenerated, and have been classified by some as Charcot joints. It seemed illogical that such widely different pathological processes could produce identical joint changes. Accordingly, a study was made at the Veterans

Administration Hospital in the Bronx and the Hospital for Special Surgery in New York City of all patients treated for Charcot joint disease. It rapidly became apparent as the confusion was magnified that some attempt at clarification would be worthwhile. This manuscript does not pretend to eliminate all the confusion but merely to point up some of the obvious misconceptions. Some of the material in this manuscript is available in various publications, much is original and never previously published. It is hoped that the reader will find in this manuscript information of interest to all fields of medicine concerned with the disease called Charcot joints.

Acknowledgments

No publication in the modern medical literature is the exclusive product of any single individual. All manuscripts reflect the exchange of ideas which is so peculiarly inherent in the field of medicine. Wherever two or more physicians can assemble, be it on the battlefield or on safari, some medical knowledge will be exchanged and subsequently reflected in the thinking of each participant. The author readily acknowledges all such information and is grateful to the many associates who added specific examples for illustration of the subject matter. Once embarked on a clinical study, the investigator soon becomes deluged by the case studies referred to him eagerly and unselfishly from various sources to swell the fund of general knowledge.

Inspiration and guidance was provided by Dr. Philip D. Wilson, Sr., Surgeon-in-Chief (now Emeritus) at the Hospital for Special Surgery and Senior Orthopaedic Consultant at the Veterans Administration Hospital in the Bronx, one of the giants of modern orthopaedic surgery. Eighteen years of association with this truly great teacher have been of inestimable value in molding the author's philosophy of orthopaedics.

Most of the illustrations were provided by the Medical Illustrations Department of the Bronx Veterans Administration Hospital. Mr. David Lubin and the other members of that department have my eternal gratitude for their patience and courteous cooperation.

My secretary, Mrs. Elaine Fedorowich, typed and arranged the manuscript in addition to all her regular duties with never a murmur of complaint. Her remarkable fortitude is deeply appreciated. Publishers are expected to provide guidance for authors and mine were not exceptions. In addition, they freely offered encouragement and suggestions without which this monograph would not be possible.

S. N. E.

Contents

CHARCOT JOINTS

CHAPTER I

General Considerations

Definition

The term "Charcot joint" was originally designated for destructive joint changes found in the presence of and related to the neuropathic involvement of patients with tabes dorsalis. By common usage this term is also accepted for similar changes found in other disease entities with neurological abnormalities and included under the designation of neuropathic arthropathy. Other terms used to characterize the same condition are neuropathic joint and neuro-arthropathy.

For our purposes the term "Charcot joint" shall be synonymous with all the names listed above to designate the progressive destructive changes involving joints of patients with or without neurologic disorders provided those changes follow the pathologic sequence to be described.

Etiology

It has been stated that 90 per cent of neuropathic joints are caused by tabes dorsalis but this has not proven to be true in the author's experience with a series of sixty-eight patients. Slightly over half of these patients had a confirmed diagnosis of tabes dorsalis. More than a fifth of the patients had diabetes mellitus with neuropathic changes characteristic in this disease. Similar joint changes are found in peripheral neuritis secondary to nutritional factors such as alcoholism and avitaminosis. The occurrence of neuropathic joint changes in syringomyelia, pernicious anemia, and leprosy are well documented. Additional causes for neuro-arthropathy are the rare cases of congenital indifference to pain and bilateral cervical ribs. At times there is no known specific cause for the joint changes. Recently neuropathic changes have been ascribed to repeated intra-articular injections of cortisone derivatives. Similar changes have occurred in two patients included in this series, one with extensive spina bifida, and the other following surgical cordotomy for intractable hip pain.

Some authors have incriminated such diseases as acute myelitis, anterior poliomyelitis, progressive muscular dystrophy, and even vertebral tuberculosis as causes of Charcot joints. It is highly doubtful that any of these disease entities can justifiably be proven responsible for the neuropathic joint. Similarly, confusion arises from a consideration of pathological changes frequently seen in the hip joints of paraplegic patients, which are fallaciously characterized as Charcot joints. In the series of sixty-eight patients studied by the author the etiology distribution was as follows:

Syphilis	34
Diabetes Mellitus	12
Nutritional Deficiency (including alcoholism)	4
Anemia	3
Syringomyelia	3
Leprosy	1
Cauda Equina Tumor	1
Spina Bifida	1
Surgical Cordotomy	1
Unknown	8

Of particular interest is the last group of eight patients in whom no definite systemic disease could be found to account for the neuropathic joint changes, despite every imaginable pertinent consultation or diagnostic aid. Nevertheless, the joint changes in this group of patients were characteristic of those which will be described for other neuro-arthropathies.

It is also apparent that the etiology distribution in any series of Charcot joints will vary considerably in other parts of the world depending upon the relative frequency of such diseases as leprosy, diabetes, and other neurological disorders.

Age Incidence

Less than 20 years	2 Patients
20-30 years	3 Patients
31-40 years	11 Patients
41-50 years	13 Patients
51-60 years	23 Patients
61-70 years	15 Patients
over 70	1 Patient
Total	68 Patients

The patients with tabes dorsalis were generally older than those with diabetes mellitus. However, when consideration was given to the area of involvement it was especially interesting to note that, whereas tabes dorsalis could produce neuropathic joint changes in any location, diabetes mellitus limited its effect solely to the ankle and foot. Only three instances of diabetic neuropathic involvement of other areas have been reported by other investigators. Zucker (1952) reported a well documented case of neuropathic spine in a diabetic patient in whom the diagnosis was supported by autopsy findings. Shore (1947) and Spear (1947) each reported a case of diabetic neuro-arthropathy of the left knee in a letter to the editor of Lancet.

Areas of involvement were as follows:

Knee	32
Foot and toes	31
Ankle	13
Hip	7
Spine	5
Shoulder	3
Elbow	2
Wrist	1
	94

In this series the preponderance of lower extremity involvement (eighty-three of the ninety-four joints) is striking but not necessarily significant. Obviously, in areas of endemic leprosy a greater percentage of finger joint involvement could alter the ratio.

Symptoms

The duration of symptoms prior to the initial examination varied so greatly as to lose all significance. Most patients with syphilis had had the disease for many years so that the neuropathic arthropathy was a late development. But the patients with diabetes mellitus followed no set pattern at all. Four of the twelve patients had not known of the

presence of their diabetes prior to the examination for the joint involvement, although all had positive signs of neuropathy. It is generally believed that neuropathy in diabetics always follows a prolonged period of poor control of the disease. Yet one of our patients was a "mild" diabetic easily and well controlled. Ellenberg, in a study of a large group of patients with diabetic neuropathy, observed a number of patients whose diabetes was well controlled and in whom there was no correlation between neuropathy and duration or severity of the diabetes. In fact some of his patients developed the neuropathy *after* institution of good control and this observation was also noted by other investigators (Rundles, Sprague). Ellenberg suggests the possibility that neuropathy in diabetes may occur independent of the presence, degree, or duration of hyperglycemia and glycosuria. He regards the neuropathy as an integral part of the diabetic syndrome, a concomitant feature rather than a complication. A striking example of similar nature is the list of diabetic complications that have been so thoroughly documented by Ellenberg as occurring in the "prediabetic" phase of patients prior to any manifest disorder of carbohydrate metabolism. These include renal disease, complications of pregnancy, vascular changes, and diseases of the eye and skin. He emphasized that "the detectable loss of carbohydrate tolerance by standard means represents a late stage of the diabetes syndrome and *actually may never occur in some instances*" (italics by the author). This may account for some or all of our eight patients in whom no systemic disease could be diagnosed.

Among the chief complaints when first seen were the following:

Painless swelling	28
Pain and swelling	24
Instability	17
Ulceration	14
Numbness	11
Weakness	6

There seems to be a general misconception that all Charcot joints are painless. As can be seen from the above list of presenting chief complaints almost as many patients sought medical aid for painful swelling as did those for painless swelling. Obviously, the presence or absence of pain in a swollen joint can play little importance in establishing a diagnosis. However, the intensity of pain is usually less than one would expect in the presence of similar gross joint disruption from other causes.

Pathology of Charcot Joints

The histopathological pattern is one of degeneration of all elements about the joint. Ligaments and capsule are infiltrated with areas of fibroblastic proliferation, edema, and small round cell infiltration. Elastic fibres are scarce or non-existent. The synovium is similarly involved and becomes quite thickened. Areas of hemorrhage in various stages of resolution with residual scarification and pigmentation are seen. Characteristically, and pathognomonic of this disease, one finds bits of dead bone and dead or living cartilage within the layers of the synovium. (Fig. 1). The articular cartilage in a Charcot joint has undergone various stages of degeneration.

Much of it has been replaced by fibrous tissue elements but even the displaced fragments have living cartilage cells. The subchondral bone is usually grossly necrotic, fragmented, and avascular and, where separation has occurred, dead bone is in evidence (Figs. 2, 3, and 4 .

The gross pathological changes may be readily apparent in a well established neuropathic joint or quite elusive in an early case. However, some of the following pathological changes should be apparent either by roentgenological study or clinical examination, particularly of the di-arthrodial joints. While the vertebrae and feet may not demonstrate these changes as readily, the process is identical.

a) There should be signs of chronic synovitis with induration and thickening of the synovium.

b) Joint effusion should be readily apparent in any neuropathic di-arthrodial joint.

c) Ligamentous relaxation and capsular distention of varying degrees should be present leading to subluxation or dislocation.

d) Gross evidence of bits of cartilage and bone debris embedded within the synovium is pathognomonic of a Charcot joint.

e) Later there may be subchondral sclerosis, cartilage erosion and eburnation of bone ends.

f) Loose bodies within the joint result from marginal fractures.

g) Finally, there may be gross instability leading to complete disorganization.

Pathogenesis

Much has been written concerning the pathogenesis of neuropathic joints, and many theories postulated as to the evolution of these joints. Charcot believed the joint destruction was the indirect result of central nervous system "trophic" disorder. Others believed the disease to be a result of local involvement of the joint by the systemic disease. Still others are of the opinion that trauma, in a previously denervated joint, played the major role in its subsequent disorganization. It is not the purpose of this presentation to attempt to solve or clarify the differences in these three theories.

It would be preferable to propose for your consideration a course of events leading to the formation of a neuropathic joint. It has been shown that in patients who demonstrate the sequence of joint changes described previously, the underlying disease has produced a loss or marked diminution of proprioception. This in turn permits an increased and unphysiological range of motion which results in stretching and tearing of the adjacent articular soft tissues. Continuation of this process leads to grinding of the articular cartilaginous surfaces. Many of these patients demonstrate diminution of pain sensation in the presence of intact muscles permitting unrestricted use of joint motion with subsequent increased synovial reaction and cartilaginous degeneration. Continued use following early mechanical disruption hastens the degenerative changes. The resulting effusion causes further distention with relaxation of ligaments and capsule and finally produces

the late findings described previously.

Recent reports have given support to the concept of trauma in a denervated joint as the primary factor in the pathogenesis of Charcot joints. Chandler and co-workers described a case of rapid destruction of an osteoarthritic hip joint, previously followed for nineteen years after eighteen months of intra-articular injections of 50 mg. of Hydrocortisone acetate at monthly intervals. They postulated that the suppression of pain by prolonged treatment with hydrocortisone encourages a damaging degree of movement and weight bearing, leading to the development of rapid joint destruction.

The theory of "proprioceptive defect" as a basic underlying cause of Charcot joints is tenable but still doesn't fit all the cases. Rose and Petrie described cases of neuropathic joints in patients with congenital indifference to pain but no other neurological abnormality. Feindel described a patient with neuropathic arthropathy and congenital absence of pain in whom he was able to demonstrate the normal number of free nerve terminals—these terminals being responsible for pain sensation. He postulated that the indifference to pain and the arthropathy must therefore be ascribed to some central defect rather than a denervation of the joint. One must therefore surmise that the final word has not been written on the pathogenesis of Charcot joints. No one theory fits all the cases; each theory fits some cases.

Evolution of a Charcot Joint

To the interested observer, privileged to follow the changes in a neuropathic joint by means of serial roentgenograms, a logical and usually predictable sequence of changes can be detected. No other pathological entity demonstrates the same course of events which are believed to be pathognomonic of Charcot joints. For purposes of clarification these changes have been divided into three stages during which various gross pathological findings described previously can be elicited.

Stage of Development

Roentgenograms of the early formative stage of a Charcot joint will show some evidence of debris formation usually beginning at the articular margins (Fig. 4). Synovial biopsy at this point will demonstrate microscopic evidence of the debris embedded within the synovium and pathognomonic of the disease (Fig. 5). This will be followed by fragmentation of the subchondral bone and attached articular cartilage (Fig. 6). As this process is repeated further disruption and capsular distention results in subluxation or dislocation.

Stage of Coalescence

This is characterized by absorption of much or all of the fine debris. Most of the larger fragments fuse together and then adhere to and coalesce with the adjacent bones (Fig. 7). This process together with the loss of vascularity resulting from the previous disorganization and fragmentation produces the characteristic sclerosis of the bone ends of a Charcot joint.

Stage of Reconstruction

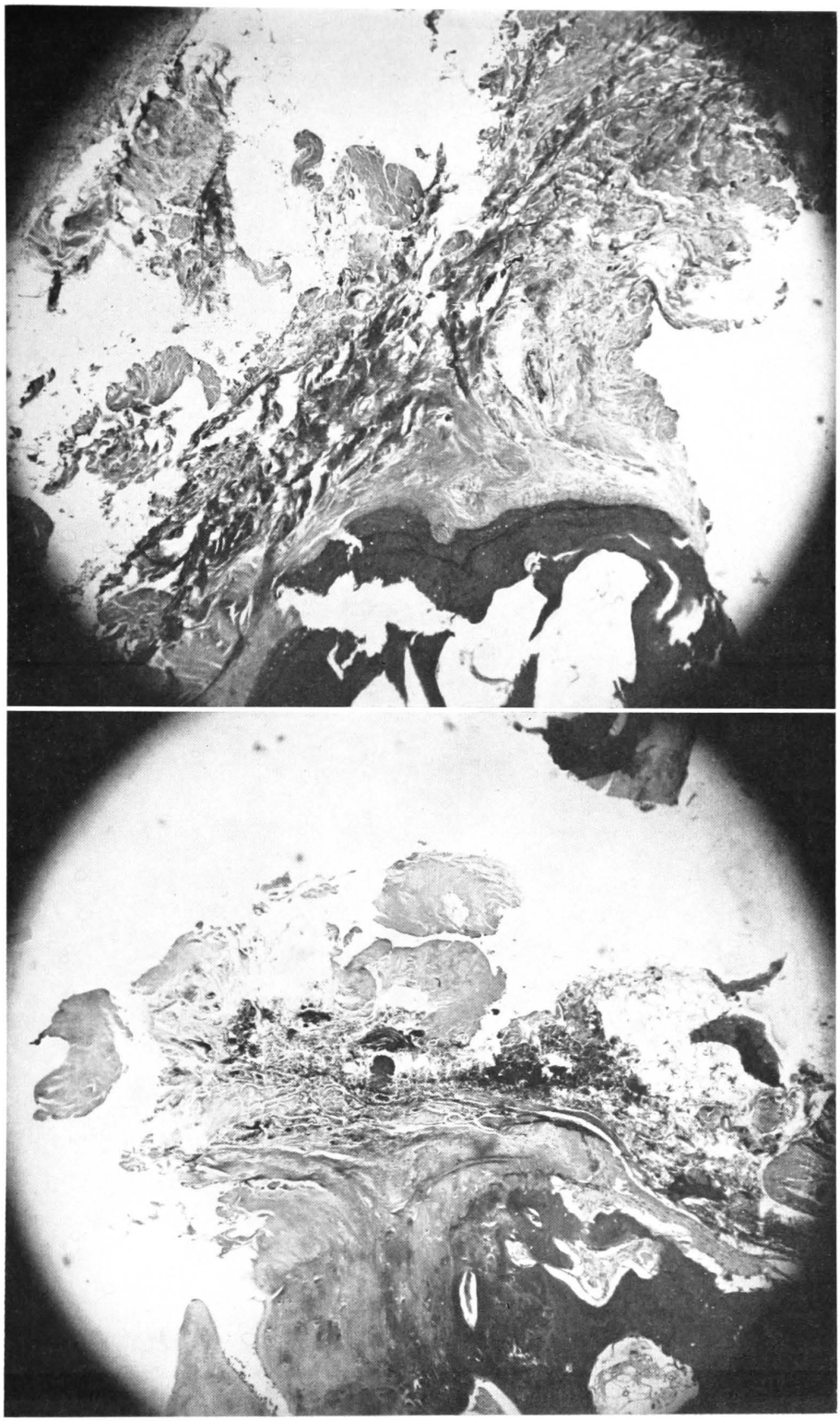
The bone ends and major fragments

become rounded; re-vascularization produces a diminution in the degree of sclerosis. As more viable bone is reconstituted some attempt at reformation of joint architecture becomes apparent (Fig. 8).

The process outlined above occurs in all Charcot joints although it may be difficult to visualize completely in roentgenograms of the spine because of overlapping gas and soft tissue shadows. The time interval between and during these stages may vary from a matter of weeks to many years, with no apparent correlation between the severity of the disease and the time required for completion of all three stages. Furthermore, as was demonstrated in several instances, the process may literally grind to a halt in the first or second stage and remain unchanged for many years. In other instances the process may repeat itself and once again go through all three stages. Multiple areas of joint involvement in a single patient occurred only in the presence of syphilitic central nervous system disorder. The only exception to this in our series was in the diabetic patients who frequently demonstrated multiple joint involvement in one or both feet, but not the widespread involvement of the tabetics (Fig. 9 a, b, c, d, e, f). In some patients who demonstrated multiple areas of involvement, each of the joints was in a different stage of development (Fig. 10). No characteristic pattern has as yet been manifest to aid in an accurate prognosis as to the eventual outcome of any given neuropathic joint. This is most unfortunate since an accurate prognosis is so essential to the final result of any contemplated surgery on these joints. It seems obvious that the performance of an arthrodesing procedure to stabilize a Charcot joint during the active phase of the Stage of Development would be doomed to failure at its inception. Similarly, the optimal time for surgery should be at the completion of the Stage of Reconstruction unless the surgical trauma stimulates an entirely new series of all three stages.

FIGURE 1. Charcot Joint. Note the seams of bony fragments overlying degenerated cartilage and subchondral bone. The latter is fragmented and few living bone cells can be seen.

FIGURE 2. Charcot Joint. Note the debris of dead bone and bits of cartilage. Below this one sees living but degenerated cartilage covering avascular necrotic bone.



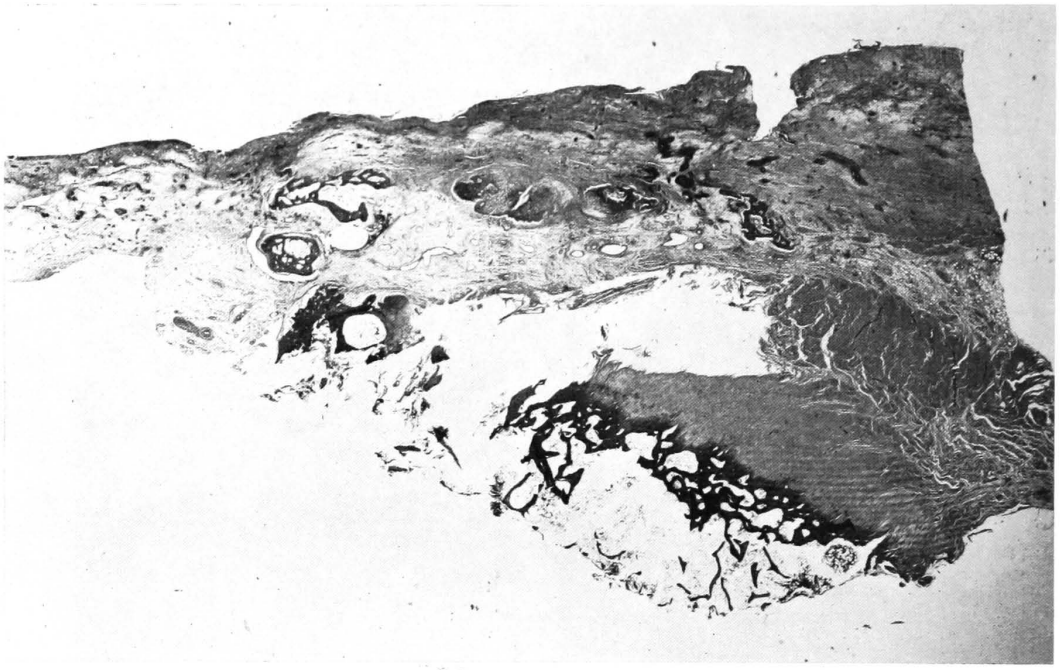
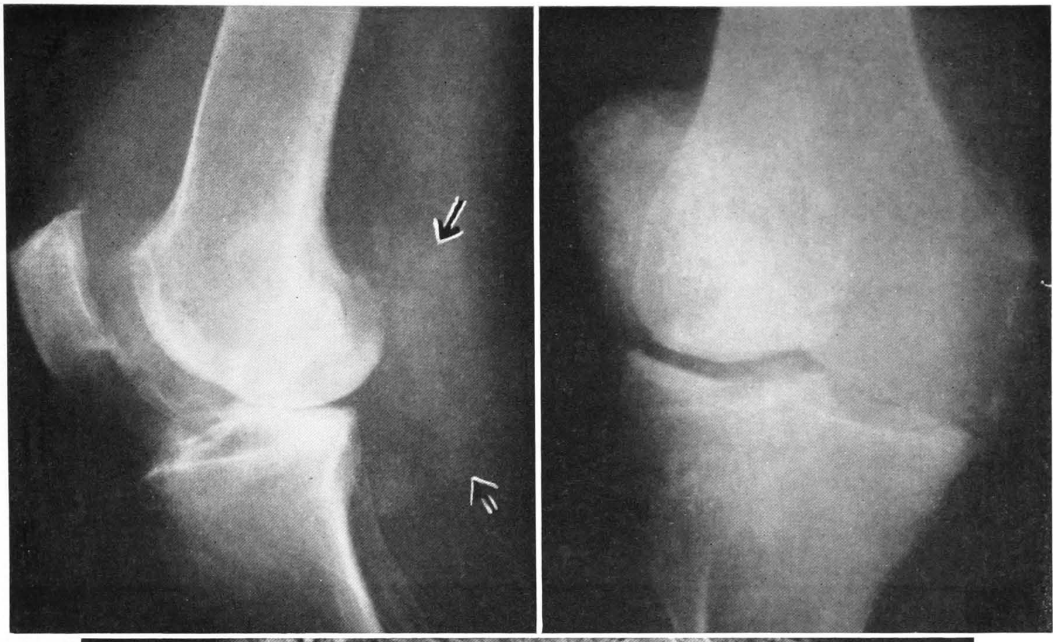
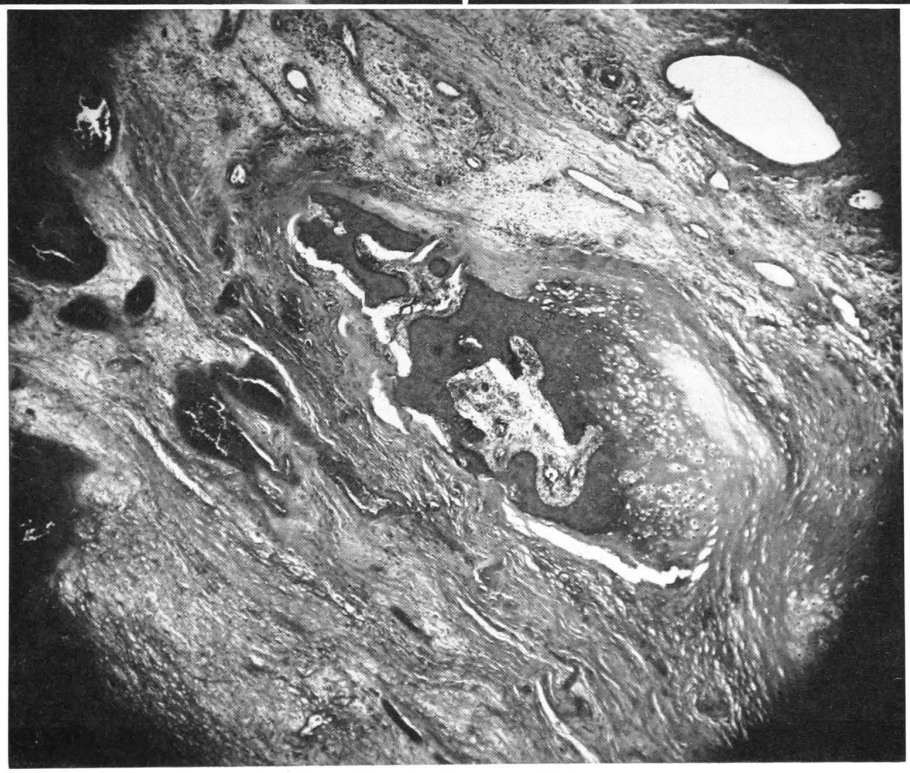


FIGURE 3. Charcot Joint. The well maintained but atrophic subchondral bone (*lower right*) with degenerated articular cartilage is apparent. But note the thickened synovium (*above*) with bits of bone and cartilage embedded within it.

FIGURE 4. Stage I. Fine debris is distributed throughout the joint and tends to collect in the natural pouches (*arrows*) when the process is more rapid than the note of synovial engulfment and absorption.

FIGURE 5. Charcot Joint. In this higher magnification one can readily see the embedded bone and cartilage fragment surrounded by fibrous elements with small round cell infiltration and vascular channels entering the bone fragment. Note that the cartilage cells are alive and able to maintain themselves from the adjacent tissue fluids.





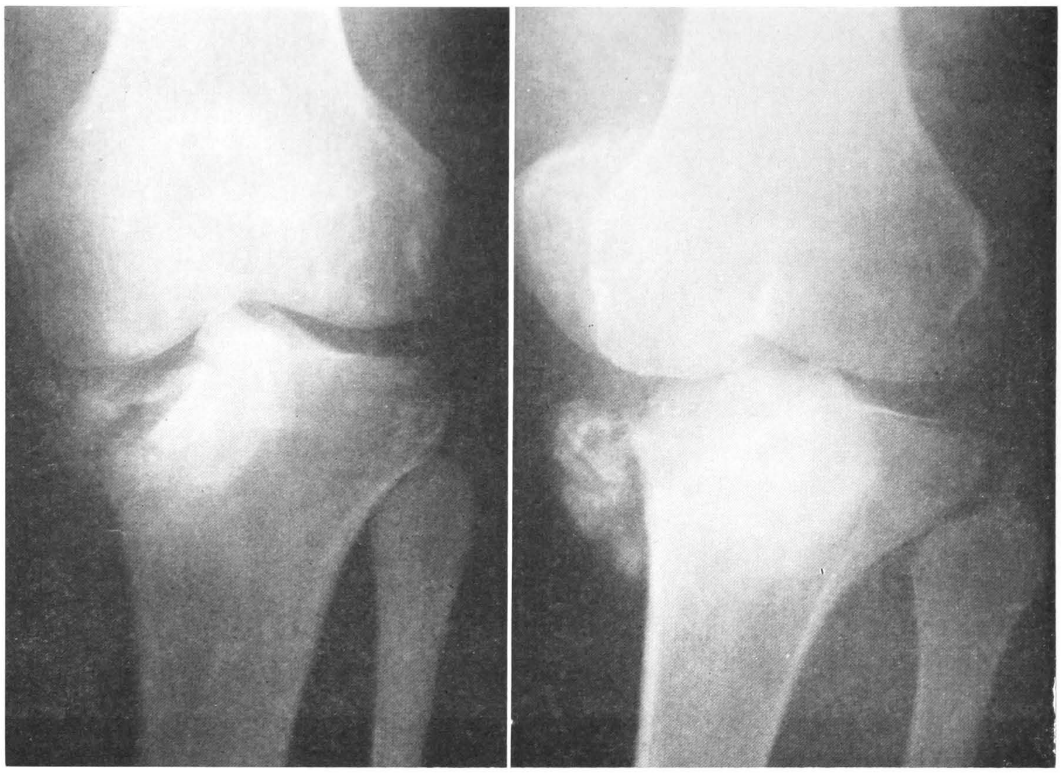


FIGURE 6. Stage I. Fragmentation of subchondral bone and adjacent articular cartilage rather than fine debris formation initiated the first stage of the disease process in this patient.

FIGURE 7. Stage II. Fragmentation has stopped and sclerosis of bone occurs concomitantly with the coalescence of fragments and adherence to adjacent bone.

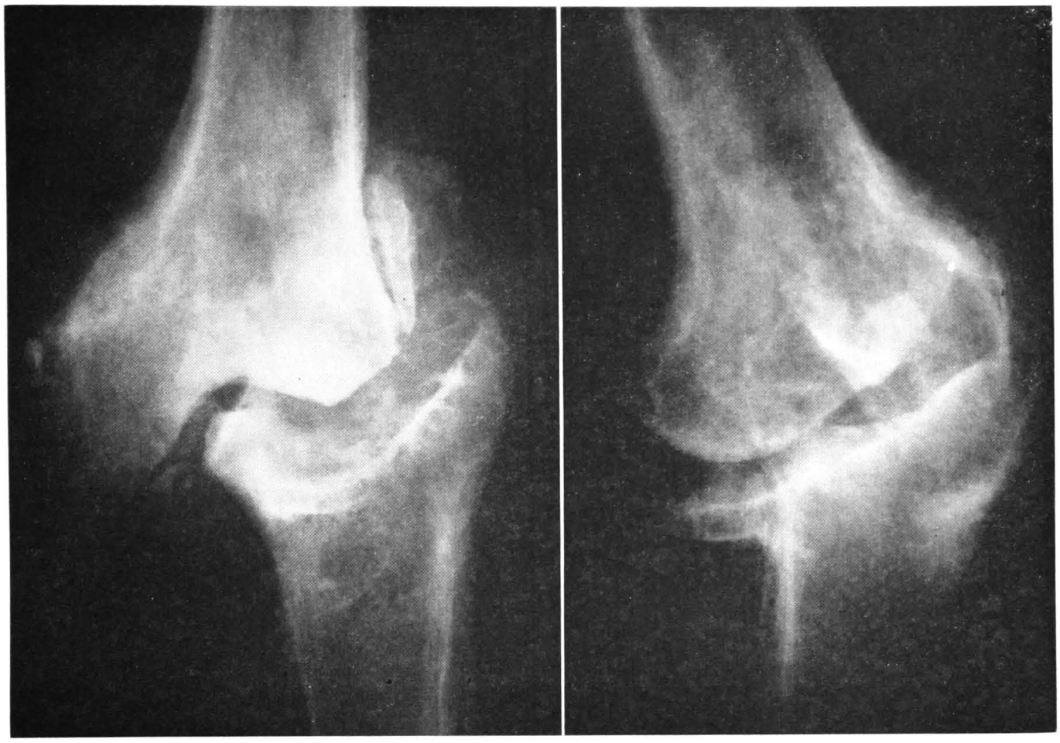


FIGURE 8. Stage III. Example of end stage in the formation of a Charcot joint with re-vascularization of sclerotic bone, re-formation of smooth rounded bone ends, and at-attempted joint reconstruction.

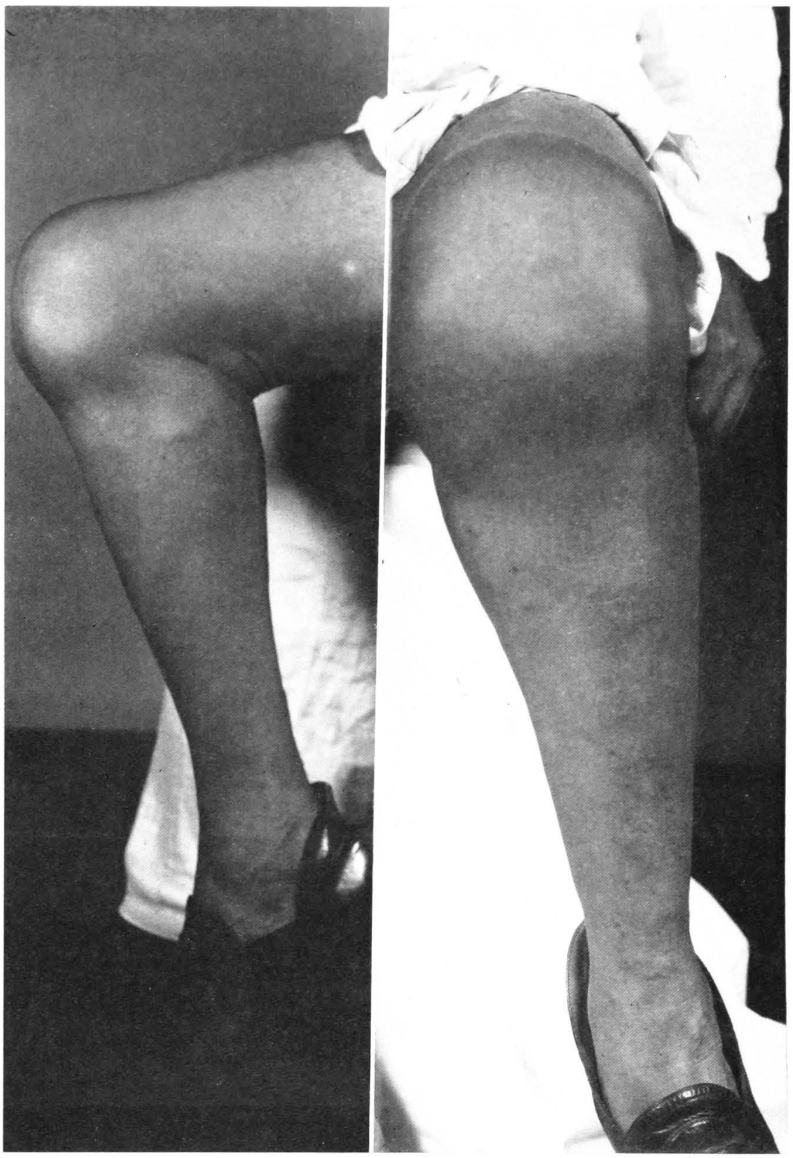


FIGURE 9 a. F. B. A sixty-eight-year-old male treated for teritary lues twenty-five years ago entered the Bronx Veterans Hospital because of painless swelling of the left knee. He had tabes dorsalis with multiple neuroarthropathies. The left knee was markedly swollen, but painless, and demonstrated abnormal mobility.

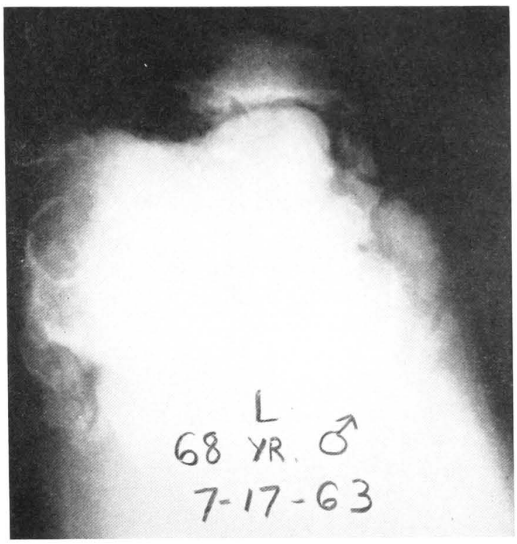


FIGURE 9 b. F. B. Patella view of the knee demonstrates a third stage of Charcot joint following extensive fragmentation, coalescence, and reformation.