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IN BRIEF

Classifications in Brief: Eichenholtz Classification of Charcot Arthropathy

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History

Charcot arthropathy is a progressive, noninfectious destructive disease of joints, most commonly affecting the foot and ankle. When first described by Jean Martin Charcot in 1868, it mainly was associated with tabes dorsalis resulting from tertiary syphilis [4]. In 1936, Jordan first noted its relationship to diabetes [11], which now is recognized as the most common cause of Charcot arthropathy in developed countries [18]. Alcoholism, syringomyelia, and leprosy also have been associated with this process, albeit less frequently [7].

The exact pathophysiology of Charcot arthropathy remains unclear with several theories proposed [18]. In the neurotraumatic hypothesis, loss of neuroprotection is thought to cause repetitive microtrauma. Conversely, the neurotrophic hypothesis proposes that sympathetic neuropathy leads to denervation of arterioles, causing increased blood flow, hyperemia, bone rarefaction, and increased susceptibility to osteoclast-mediated resorption and fragmentation [1, 9, 18, 25]. While the development of Charcot arthropathy is likely attributable to a combination of these processes, other theories also may play a role. One such implicates inflammatory cytokines, such as tumor necrosis factor-α and interleukin-1, as stimulating osteoclast formation, resulting in bone resorption and weakening [1].

Purpose

Charcot arthropathy can be challenging to diagnose, as its earliest manifestations (eg, swelling, inflammation and warmth) are similar to those seen with simple sprains, deep venous thrombosis, osteomyelitis, cellulitis, and rheumatoid arthritis [16]. In addition to joint deformity and instability, foot ulceration, infection, and, in the most severe cases, amputation, patients with Charcot arthropathy have a median survival of approximately 12 years less than that of the general population [23]. Although this almost certainly reflects a more-severe diabetic condition rather than a direct effect of the neuropathic arthropathy, the pain, disability, and risk of infection associated with Charcot arthropathy point to the importance of accurate classification and staging of the varied presentations of this condition. The Eichenholtz classification is a temporal-based approach that has been used for this purpose; it can assist clinicians in diagnosis, staging, and selecting appropriate treatment [7].

Description

In 1966, orthopaedic surgeon Sidney N. Eichenholtz (1909–2000) published a monograph entitled "Charcot Joints" in which clinical, radiographic, and pathologic data of 68 consecutive patients were used to define three stages

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of Charcot arthropathy based on the natural history of the condition [7]. The three stages he described were (I) development; (II) coalescence; and (III) reconstruction and reconstitution (Table 1). Progression through these stages can range from several weeks or months to many years. Eichenholtz's cohort included patients with diabetes mellitus, syphilis, alcoholism, syringomyelia, and leprosy who had arthropathy develop in the foot and ankle, knee, and hip [7]. The diverse patient population studied by Eichenholtz elucidates the universal nature of Charcot arthropathy and the applicability of his classification to all joints affected.

The first stage of Charcot arthropathy described by Eichenholtz, which he called development, presents with radiographic evidence of osteopenia, periarticular debris and fragmentation, and joint subluxation or dislocation. Stage I also commonly is referred to as the "fragmentation" or "dissolution" stage. Physical examination of the involved joint reveals swelling, erythema, and/or ligamentous laxity [7]. Stage I arthropathy of the proximal humerus, which most often is caused by syringomyelia, may present as a "vanishing bone", with a predominantly resorptive pattern, marked bone loss, and little debris [17].

In the second stage, absorption of periarticular debris, early sclerosis, and bony consolidation of some of the larger fragments seen in Stage I are evident on radiographs. Additionally, new bone begins to form in this stage. Clinically, decreased warmth and swelling of the involved joint are observed [7].

The stage of reconstruction and reconstitution is the third stage described by Eichenholtz. This represents the progression of the joint to a more stable structure. Although deformity still may be evident on radiographs, new bone formation continues and becomes a more prominent feature at this time. Decreased sclerosis, rounding and smoothing of bone fragments, joint space narrowing with arthrosis, and fibrous and/or osseous ankylosis also will be present. On physical examination, swelling and erythema has dissipated, and the joint will seem stable in the setting of a fixed deformity [7].

Clinical signs (such as swelling, warmth, and erythema) regularly precede the radiographic findings seen with Eichenholtz Stage I arthropathy [5]. As such, in 1990 Shibata et al. [21] added a fourth stage, Charcot foot Stage 0, to the conventional Eichenholtz classification (Table 1). The addition of this prodromal stage has important therapeutic implications, because the immobilization and off-loading of feet with Stage 0 symptoms may prevent progression of skeletal destruction and deformity [13, 16].

Anatomic-based classification systems for Charcot arthropathy also have been developed [2, 10, 12, 19, 22]. Of these systems, the most widely used is Brodsky's, which is specific to the foot and based on the most common regions affected (Table 2) [2]. Type 1 (midfoot) arthropathy involves the tarsometatarsal and naviculocuneiform joints; Type 2 (hindfoot) involves the subtalar and/or Chopart joint; Type 3A (ankle) involves the tibiotalar joint; and Type 3B (calcaneus) involves a fracture of the

Table 1. The modified Eichenholtz [7] classification

Stage	Radiographic findings	Clinical findings	Treatment
0 (prodromal)	Normal radiographs	Swelling, erythema, warmth	Patient education, serial radiographs to monitor progression, protected weightbearing
I (development)	Osteopenia, fragmentation, joint subluxation or dislocation	Swelling, erythema, warmth, ligamentous laxity	Protected weightbearing with total contact casting or prefabricated pneumatic brace. Cast or brace should be used until radiographic resolution of fragmentation and presence of normal skin temperature (usually needed for 2–4 months).
II (coalescence)	Absorption of debris, sclerosis, fusion of larger fragments	Decreased warmth, decreased swelling, decreased erythema	Total contact casting, prefabricated pneumatic brace, Charcot restraint orthotic walker, or clamshell ankle-foot orthosis
III (reconstruction)	Consolidation of deformity, joint arthrosis, fibrous ankyloses, rounding and smoothing of bone fragments	Absence of warmth, absence of swelling, absence of erythema, stable joint ± fixed deformity	Plantigrade foot: custom inlay shoes with rigid shank and rocker bottom sole. Nonplantigrade foot or ulceration: débridement, exostectomy, deformity correction, or fusion with internal fixation.

Stages I-III described by Eichenholtz, Stage 0 added by Shibata et al. [21], because clinical signs of Charcot arthropathy were found to precede radiographic changes.

Table 2. Brodsky [2] anatomic classification of Charcot arthropathy (Types 1–3B) with Trepman et al. [22] modification (Types 4 and 5)

Type	Location	Involved joints
1	Midfoot	Tarsometatarsal, naviculocuneiform
2	Hindfoot	Subtalar, talonavicular, calcaneocuboid
3A	Ankle	Tibiotalar
3B	Calcaneus	Tuberosity fracture
4	Multiple regions	Sequential, concurrent
5	Forefoot	Metatarsophalangeal

calcaneal tuberosity [2]. This system subsequently was modified by Trepman et al. [22] to include Types 4 (multiple areas) and 5 (forefoot), which correlate with sequential or concurrent involvement of multiple areas and metatarsophalangeal involvement, respectively (Table 2).

Validation

Reliability, which refers to the Eichenholtz classification's ability to consistently grade the radiographic deformity and stages across reviewers (interobserver reliability) and by the same reviewer across time (intraobserver reliability), has not been assessed to our knowledge. The validity of the classification, which refers to the concurrence between the radiographic and physical examination findings described by Eichenholtz at each stage, also remains unknown.

The literature pertaining to Charcot arthropathy has acknowledged this lack of validation, citing the subjectivity of the classification's stages and the difficulties encountered when attempting to distinguish the end of one stage and the beginning of the next as impediments to proper validation [6]. There are other challenges that complicate validation, including the limited prevalence of Charcot arthropathy, which is reported to be 0.08% to 7.5% in patients with diabetes, the high frequency of delayed and missed diagnoses, and the multitude of joints affected [14, 15]. Additionally, validation requires assessment of each patient's imaging and physical examination findings by multiple observers at one time, and with time, which can be difficult to facilitate.

Limitations

There are limitations of the Eichenholtz classification other than its subjectivity. As a temporal staging system, anatomic location is not taken into consideration. This was the impetus behind the development of other classification schemes, such as Brodsky's, because the specific location and extent of osseous destruction also can influence treatment decisions [2, 10, 12, 19, 22].

The stages for the Eichenholtz classification do not account for patients' symptoms and comorbidities, as it relates only to radiographic abnormalities with focused physical examination findings. Patient-specific symptoms and comorbidities cannot be ignored and must influence clinical decision-making. Many patients will present with additional musculoskeletal, ocular, renal, and/or vascular sequelae of diabetes, as longstanding, poorly controlled disease is intimately associated with Charcot arthropathy. This must be considered when developing a treatment plan, as certain conditions may prohibit surgical intervention.

Although plain radiographs remain effective in identifying the characteristic deformities of Charcot arthropathy, radiographs may be negative with early disease [20]. Advanced imaging modalities, such as MRI and bone scans, are of use in such instances, leading to earlier diagnosis, staging, and intervention [3]. MRI is considered the most sensitive modality in the detection of early changes of Charcot neuropathy, and will delineate soft tissue edema, arch collapse, joint effusions, and subchondral bone marrow edema of involved joints [24].

Despite its limitations and lack of validation, the Eichenholtz classification continues to be the most commonly used staging system for Charcot arthropathy. This is attributed to its ability to guide treatment selection and the timing of various interventions. For each of the stages, an algorithmic approach is available to clinicians, illustrating the classification's clinical utility (Table 1) [8, 24].

Conclusions and Uses

The Eichenholtz classification describes the pathophysiologic progression of Charcot arthropathy in a temporal staging system based on the natural evolution of this process [7]. Despite its subjectivity, the classification is well accepted and widely used by orthopaedic surgeons in addition to physicians of many other specialties [8]. It will be important for future studies to ascertain the reliability and validity of this staging system, because it is so widely used. In the future, there may be a role for advanced imaging in the classification of Charcot arthropathy in addition to incorporation of anatomic classification schemes.

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