In 1991 Euwer and Sontheimer [1] proposed a revision of Bohan and Peter’s [2] original classification system of DM/PM, and included amyopathic DM as a sixth clinical type. Thereafter, the nomenclature of amyopathic DM is controversial. Some authors continue to use the historical term “DM sine myositis” [3, 4], others prefer the term “amyopathic DM” [5], a third group use the designations “premyopathic DM” [6] or “hypomyopathic DM” [5], but a few workers still prefer to use the terms “DM with normal muscle enzymes” [7, 8] or “DM without muscle weakness” [9, 10].

Amyopathic DM refers to a condition in which the typical cutaneous eruption of DM is present but muscle disease is lacking [1, 3, 5, 7]. This clinical variant of DM is the opposite of PM, in which muscle disease is present but cutaneous disease is lacking. The category of amyopathic DM is controversial, since according to the strict Bohan and Peter criteria patients with this presentation only fulfill one criterion and may have a suspected DM [11]. The clinical entity amyopathic DM includes patients with either cutaneous disease alone [7], or patients with cutaneous disease and minimal muscle involvement [12, 13]. However, the former definition lacks the criteria of Bohan and Peter for possible DM, and due to the low specificity of the skin lesions, it might be difficult to distinguish cases of amyopathic DM from those with other early connective tissue diseases such as lupus erythematosus [3, 14].

In 1975 when Bohan and Peter [1] published their five diagnostic criteria defining PM and DM, Krain [15] described the first six patients with “DM sine myositis,” four of whom were adults, who presented with typical DM skin lesions and lack of muscle disease; later on, all of them developed overt myositis at an interval of between 3 months and 10.5 years. In Krain’s series, one patient presented initially with cutaneous rash and weakness, which spontaneously remitted, and then presented again with only skin disease, but without muscle disease for 6 years. Another two patients who started with skin eruption and muscle weakness had normal muscle enzymes, demonstrated by EMG and a biopsy specimen. Muscle biopsy and EMG were performed, however, in only three patients with initial evolution of disease. Later on, Pearson [16] noted three cases of juvenile DM with “very mild muscular weakness and more prominent cutaneous features, but relatively stable course over a 6-month to 3-year period.” Bohan et al. [17] reported that, in their series, at the initiation of DM the muscle strength was normal in
48 of 173 patients (31%); there were three patients (~2%) who failed to develop significant muscle weakness but met other diagnostic criteria (e.g., abnormal enzyme, EMG, or muscle biopsy), and one woman with rash alone but without overt myositis who had cancer of the uterus. The term “amyopathic dermatomyositis” was introduced by Pearson in 1979 [12] for patients who had typical cutaneous findings of DM, but did not have any clinical or laboratory signs of muscle disease at least 2 years after the onset of the skin pathology. He reported five women with only cutaneous manifestations of DM, and six other patients (four women and two men) in whom the skin lesions were florid, whereas weakness and EMG changes were minimal [12]. Rockerbie et al. [9] described 28 of 50 patients (56%) who had DM with skin rash preceding muscle weakness, at intervals varying from 1 month to more than 4 years. Six patients (12%) only had cutaneous lesions more than 21 months before the onset of muscle weakness. Euwer and Sontheimer [1] described six patients who did not develop evidence of myositis for at least 2 years after the onset of cutaneous rash and defined cutaneous manifestation of amyopathic DM (“DM sine myositis”). However, EMG and muscle biopsy were not performed in any of these patients. Because five of these six cases were treated with moderate doses of prednisone and none of them developed overt myositis, Euwer and Sontheimer suggested that a “more aggressive approach to treating the skin disease may prevent the development of muscle disease in cases who initially have only cutaneous involvement.” Euwer and Sontheimer [13] divided patients with amyopathic DM into three groups: (i) Type 1 represents pure amyopathic DM patients who have only cutaneous manifestations, (ii) Type 2 are patients with skin lesions who have subjective myalgias and weakness, but without laboratory evidence of muscle disease (as seen in Krain’s series), and (iii) Type 3 are patients with eruptions, without features of muscle disease, but having evidence of abnormal laboratory tests at some time during their course. In 1993, Stonecipher et al. [7] reported 13 patients with typical DM rash and normal serum enzyme levels. The patients were separated into three groups: (i) four patients with only cutaneous changes without muscle involvement after 4–11 years follow-up (i.e., amyopathic DM), (ii) seven patients with cutaneous changes at baseline and subsequent development of myositis, and (iii) two patients with cutaneous changes and normal muscle enzymes in whom, however, the diagnostic evaluation showed subclinical evidence of muscle involvement demonstrated by EMG and/or muscle biopsy. This classification approach has been utilized by others [18]. Stonecipher et al. [7] have suggested the avoidance of therapy with systemic corticosteroids or other immunosuppressive drugs in patients with lack of overt muscle disease. In a retrospective study of 12 patients with typical DM rash but without reported weakness (i.e., with amyopathic DM) in France, Cosnes et al. [10] found eight cases with an increased enzyme levels, seven patients had myopathic finding on EMG and nine patients had abnormal muscle biopsy, of whom five had histopathologically confirmed myositis. After approximately 5 years follow-up, no patient developed overt muscle disease, and cutaneous manifestations improved in nine of all 12 patients [10]. Hydroxychloroquine sulfat was the first line therapeutic agent in all patients. None of the patients in this group received systemic corticosteroid treatment. Some clinically amyopathic DM patients have been observed to exhibit continuing skin disease activity for more than 20 years without ever developing clinically evident muscle weakness or other systemic features of DM [19].