ORIGINAL ARTICLE

Characteristics and Outcomes of Juvenile Dermatomyositis (JDM) in Thai Children: Experience from a Tertiary Referral Center

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Background: Juvenile dermatomyositis (JDM) is the most common autoimmune myositis in children. However, there is a paucity of data on the clinical characteristics of JDM patients in Thailand.

Objective: To describe the clinical manifestations, investigations, treatments, complications, and outcomes of JDM in Thailand.

Materials and Methods: A bidirectional descriptive study based on past medical records review. All children younger than15 years diagnosed with JDM by Bohan and Peter criteria at King Chulalongkorn Memorial Hospital between January 2010 and December 2022 were enrolled.

Results: Thirteen cases were identified with 69% female. The median age at diagnosis and median time to diagnosis were 3.4 (IQR 2 to 6.5) years and 3.5 (IQR 1.2 to 8) months, respectively. The common clinical presentations at disease onset were weakness at 100%, Gottron papules at 69%, and heliotrope rash at 46%. Lactate dehydrogenase was elevated in all cases at diagnosis. Evidence of muscle inflammation was found in 78% of biopsies, 80% of EMGs, and 100% of MRIs. The most common myositis specific Ab was anti-NXP2 at 64%. Most patients had polyphasic disease courses. More than half of the patients, or 62%, developed calcinosis. Six of thirteen cases received recommended treatment with prednisolone and methotrexate at diagnosis. One patient with calcinosis had a good response with infliximab.

Conclusion: The present review underscores that JDM patients with younger age at diagnosis will need to be monitored closely and need aggressive long-term treatment. Prompt treatment should be managed according to the severity of symptoms to improve clinical outcomes and prevent serious complications.

Keywords: JDM; Calcinosis cutis; Myositis autoantibody; Children; Developing countries

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Juvenile dermatomyositis (JDM) is a heterogeneous autoimmune-mediated disease characterized by proximal muscle weakness and rashes, typically heliotrope rash and Gottron papules. JDM is the most common inflammatory myopathy in children and can lead to significant morbidity, particularly in cases of delayed diagnosis and treatment[1]. An estimated incidence of 1.9 to 4 per million per year and prevalence of 2.5 per 100,000 was reported worldwide in a previous study[2]. Demographic data and clinical presentation may differ by race and geographic region. The etiology and pathogenesis of this disease remains unknown, but it has been found associated with multiple genetic risk factors and environmental exposure to infectious agents or medications[3].

Patients sometimes present with other manifestations such as fatigue, fever, arthritis, abdominal pain, gastrointestinal tract bleeding, and interstitial lung disease. The disease course is variable and with advanced therapeutic approaches now available, mortality has declined[4]. However, calcinosis remains a frequent long-term complication[5].

The clinical manifestations of JDM have been extensively described in different countries[6-8]. However, there is a paucity of data from developing countries, especially Southeast Asian countries.

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During the past ten years, there was only one published study of JDM in Thai children. Advanced investigations like magnetic resonance imaging (MRI) or myositis autoantibodies were scarcely mentioned in that paper[9]. The present study primary objective was to identify clinical features, progression, and treatment outcomes of JDM in Thai children from a large, single tertiary referral center.

Materials and Methods

The author performed a retrospective medical chart review of children younger than 15 years diagnosed with JDM by Bohan and Peter criteria over a 12-year period between January 2010 and December 2022 at King Chulalongkorn Memorial Hospital, Bangkok, Thailand. The diagnostic criteria established by Bohan and Peter included a classic skin lesion as heliotrope rash or Gottron papules, plus at least three of the following conditions, symmetrical proximal muscle weakness, one or more muscle enzymes elevated, electromyographic abnormalities, and consistent muscle biopsy findings[10]. Patients were excluded from the study if they had other connective tissue diseases or overlapping syndrome.

Ethical approval was obtained from the Institutional Review Board, Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand (COA No.324/2020, IRB No.017/63).

The data collected included demographic information, age of presentation, time to diagnosis (duration between onset of symptoms and diagnosis), follow-up duration, clinical and laboratory findings, treatment, and treatment outcomes. Periungual telangiectasia was documented by a dermoscopy or direct examination. The investigation profile included muscle enzymes, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), myositis autoantibodies, electromyography (EMG), muscle biopsy findings, MRI, and other tests as applicable. Patients who achieved normal muscle power, normal enzyme levels, and no new rashes or systemic features were graded as complete remission (CR), while at least one grade improvement in muscle power was graded as partial remission (PR). No improvement in muscle power was graded as non-response (NR)[11]. The disease course was classified as monocyclic when there was a full recovery and no relapses while being off medications within two years of diagnosis. The disease course was classified as polycyclic if there was disease recurrence after achieving a definite remission by PRINTO criteria for inactive disease. Chronic continuous (CC) was defined if disease activity was persistent for more than two years after diagnosis. The PRINTO criteria for inactive disease includes creatine kinase (CK) of less than 150 U/L, childhood myositis assessment scale (CMAS) greater than 48, manual muscle testing score (MMT) greater than 78, and physician global visual analog scale (VAS) of less than 0.2[12]. Patients satisfying any three of these criteria are classified as having clinically inactive disease.

All statistical analyses were performed using IBM SPSS Statistics for Windows, version 21.0 (IBM Corp., Armonk, NY, USA). Statistical significance was set at p-value less than 0.05, with a confidence interval (CI) of 95%. Descriptive statistics were reported as number and percentage (%), mean and standard deviation (SD), or median and interquartile range (IQR), as appropriate.

Results

Patient characteristics and diagnostic features

Thirteen patients with JDM were identified over the 10-year period. All patients were compatible with the criteria for classification as definitive JDM proposed by Bohan and Peter[10]. The overall gender ratio was female to male at 2.3 to 1. The median age at diagnosis was 3.4 (range of 2 to 6.5) years and median time to diagnosis was 3.5 (range of 1.2 to 8) months. Most of the patients had regular follow-ups with a mean follow-up period of 5±2.3 years.

Proximal muscle weakness was noted in all subjects and other frequent clinical features included Gottron papule, heliotrope rash, muscle pain, periungual telangiectasia, malar rash, vasculitis, fever, and skin ulcer (Table 1). Eight patients (62%) developed calcinosis cutis, which is a poor prognostic feature and sign of refractory to treatment. The authors also found this skin feature at disease onset in two of the eight patients. Neurological manifestations were seen in one male patient (case 10), who developed status epilepticus requiring intensive care unit admission. His MRI brain showed findings compatible with cerebral vasculitis. Renal involvement, which is rare presentation in JDM, was observed in the same patient (case 10) who had transient glomerulonephritis during a flare of his disease (Table 2). In this case, repeated antinuclear antibody (ANA), and complement levels did not support the diagnosis of systemic lupus erythematosus (SLE) or overlap syndrome.

Laboratory investigations

At least one muscle enzyme was elevated in all

Table 1. Demographic features and clinical manifestations of JDM patients

	n (%)
Patient	13
Sex	
Female	9 (69)
Age at diagnosis (years); median (IQR)	3.4 (2 to 6.5)
Time to diagnosis (months); median (IQR)	3.5 (1.2 to 8)
Follow-up duration (years); mean±SD	5 ± 2.3
The most common presentation at disease onset	
Weakness	13 (100)
Gottron papule	9 (69)
Heliotrope rash	6 (46)
Muscle pain	6 (46)
Other clinical presentation	
Periungual telangiectasia	10 (77)
Malar rash	7 (54)
Vasculitis (CNS, GI, or palmar vasculopathy)	4 (31)
Photosensitive rash (V-sign or shawl sign)	4 (31)
Fever	3 (23)
Skin ulcer	3 (23)
Restrictive lung disease	2 (15)
Renal involvement: glomerulonephritis	1 (8)
Calcinosis (n=8)	
At onset	2 (25)
At disease course	6 (75)

CNS=central nervous system; GI=gastrointestinal; IQR=interquartile range; SD=standard deviation

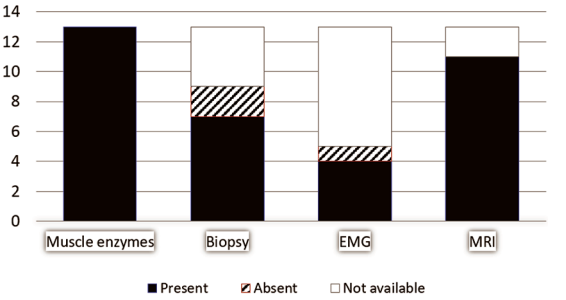

Figure 1. Diagnosis modalities of JDM patients in the study.

Present: supportive of myositis, Absent: not supportive of myositis, Not available: not done or results not accessible

EMG=electromyography; MRI=magnetic resonance imaging

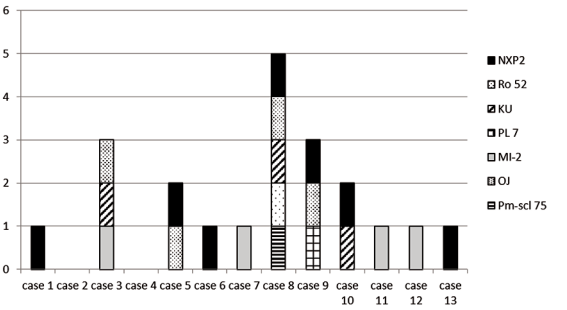

Figure 2. Myositis autoantibodies of JDM patients in the study (n=11; case 2 and 4 not tested).

subjects at diagnosis, which served as a marker of muscle injury. Lactate dehydrogenase (LDH) was the most frequently raised enzyme at 13 (100%), followed by aspartate aminotransferase (AST) in 10/11 (91%), CK in 9/13 (69%), and alanine aminotransferase (ALT) in 6/11 (55%). Serum aldolase testing was not performed due to non-availability at the authors' institute. Tests for AST and ALT were not performed in two patients. Nine of eleven patients (82%) had raised ESR greater than 30 mm/hour, while there were no patients displayed abnormal CRP at presentation.

Nine patients underwent muscle biopsy, with seven (78%) showing characteristic features of JDM, including perivascular inflammatory cell infiltration, perifascicular atrophy, and degenerative muscle fibers. EMG was performed on five patients with four (80%) demonstrating abnormal patterns consistent with inflammatory myopathy. Eleven of thirteen patients (85%) completed MRI study with all having abnormal findings, including edema, signal abnormalities in the affected muscles, and muscle atrophy with fatty replacement (Figure 1).

Myositis autoantibody testing was completed in 11 of 13 JDM patients in the present study. Results revealed a positive finding of at least one myositis-specific autoantibody (MSA) in all patients. The most common myositis autoantibodies found in the present study cohort were anti-NXP2 in 64%, anti-Ro52 in 36%, anti-Mi2 in 36%, anti-Ku in 27%, anti-PL7 in 9%, anti-OJ in 9%, and anti-PM-Scl75 in 9% (Figure 2).

Treatment and outcome

The medications administered at diagnosis were corticosteroid plus methotrexate (MTX) in 6/13 (46.2%), corticosteroid monotherapy in 3/13 (23.1%), corticosteroid plus hydroxychloroquine in 3/13 (23.1%), and corticosteroid plus cyclophosphamide 1/13 (7.7%). All patients were treated with corticosteroids at different dosages. Other immunosuppressive medications were added as a second line therapy during the disease course, such as azathioprine in nine (69%), mycophenolate mofetil in seven (54%), and intravenous immunoglobulin (IVIG) in four (31%). MTX was used in 13 patients (100%), either at diagnosis or during disease course.

Table 2. Clinical profile and outcomes of JDM patients in the study

Case	Age at diagnosis (year)/sex	Duration before diagnosis (months)	Rash/skin manifestation		Muscle weakness	Muscle enzyme elevation	Electromyo-graphic changes	Muscle biopsy	Positive myositis antibodies		Follow up (months)	Medication	Outcome	Complication
			Heliotrope	Gottron papule					Anti-NXP2	Others				
1	2/male	1	+	+	+	CK, AST, LDH	N/A	No significant change	+	-	77	MP, OP, MTX, AZA, MMF	PR	Obesity, NASH
2	6/female	24	+	+	+	AST, LDH	N/A	N/A	N/A	N/A	68	OP, HCQ, AZA	PR	Calcinosis cutis, maculopathy, NASH, DM
3	10/female	8	+	+	+	AST, ALT, LDH	Myopathy	Perivascular infiltration, perifascicular atrophy	–	Ro52, MI2, KU	78	MP, OP, MTX, HCQ, AZA, MMF, IVIG, PAM	PR	Calcinosis cutis, osteoporosis, Neoplasm sclerosing pneomocytoma
4	5/female	8	+	–	+	LDH	Myopathy	N/A	N/A	N/A	94	OP, AZA, MMF	PR	Calcinosis cutis
5	8/female	2	+	+	+	CK, AST, ALT, LDH	No myopathy	N/A	+	Ro52	24	MP, OP, MTX, HCQ, AZA, MMF	CR	Calcinosis cutis, obesity
6	2/female	14	+	+	+	CK, LDH	N/A	Perivascular infiltration, perifascicular atrophy	+	-	91	OP, HCQ, MTX, AZA, PAM	AD	Calcinosis cutis
7	3/male	1	–	+	+	CK, AST, ALT, LDH	N/A	Inflammatory cell infiltration, no perifascicular atrophy	N/A	MI2	51	MP, OP, MTX, MMF	CR	Obesity, NASH
8	1.5/female	6	+	+	+	AST, LDH	N/A	Perivascular infiltration, perifascicular atrophy	+	Ro52, KU, OJ, PM-Scl 75	40	MP, OP, MTX, CYC, IVIG, AZA, INF, PAM, MMF	AD	Calcinosis cutis, lipodystrophy, joint contracture, LTBI
9	2/female	1	+	+	+	CK, LDH	N/A	Perivascular infiltration, perifascicular atrophy	+	Ro52, PL7	41	MP, OP, CYC, MMF, MTX, IVIG, INF, PAM	PR	Calcinosis cutis, HT
10	6/male	2	+	+	+	CK, AST, ALT, LDH	N/A	N/A	+	KU	32	MP, OP, MTX, IVIG, AZA	CR	GN, DVT, HT, AVN, MDD, CNS vasculitis
11	2/male	3	–	+	+	CK, AST, ALT, LDH	N/A	Perivascular infiltration, perifascicular atrophy	–	MI2	12	MP, OP, MTX	PR	Calcinosis cutis
12	3/female	4	–	–	+	CK, AST, LDH	Myopathy	Perivascular infiltration, perifascicular atrophy	N/A	-	Lost to follow up	MP, OP, MTX	PR	HT
13	10/female	2	+	–	+	CK, AST, ALT, LDH	Myopathy	No significant change	+	-	3	MP, OP, HCQ, MTX, AZA	PR	Skin ulceration

AD=active disease; ALT=alanine aminotransferase; AST=aspartate aminotransferase; AVN=avascular necrosis; AZA=azathioprine; CK=creatinine kinase; CR=complete remission; CYC=cyclophosphamide; DM=diabetes mellitus; DVT=deep vein thrombosis; GN=glomerulonephritis; HCQ=hydroxychloroquine; HT=hypertension; INF=infliximab; IVIG=intravenous immunoglobulin; LDH=lactate dehydrogenase; LTBI=latent tuberculosis infection; MDD=major depressive disorder; MMF=mycophenolate mofetil; MP=methylprednisolone; MTX=methotrexate; NASH=nonalcoholic steatohepatitis; OP=oral prednisolone; PAM=pamidronate; PR=partial remission

Of note, two patients who had severe persistent disease course and extensive calcinosis received infliximab therapy (cases 8 and 9) (Table 2).

At the time of last follow-up, complete remission was achieved in three patients (23%), partial remission in eight patients (62%), and two patients (15%) still had active disease. Among the active cases, case 8 experienced a suboptimal response to corticosteroid and various immunosuppressive medications (Table 2) and was put on infliximab with partial response. The authors' next plan was to commence her on Rituximab, which was limited use in the present study due to financial constraint. There was no mortality in the present study. The disease course was classified as polycyclic in eight patients (62%) and chronic persistent in four patients (31%). There was only one patient with a monocyclic disease course. One patient (case 9) was non-responsive to routine treatment modalities and developed extensive calcinosis cutis. It was important to note that after infliximab administration regularly for six months, the patient showed a dramatic response in resolving the calcinosis.

The most common complication in the present cohort was calcinosis cutis. It is worth mentioning that four out of seven patients (57%) with anti-NXP2 autoantibodies positivity developed calcinosis.

Discussion

There are scarce data available in the literature on clinical characteristics and outcomes of JDM from Southeast Asian countries. The present study was designed to explore clinical features, investigations, and treatment outcomes of JDM in Thai children. In the present study cohort, the authors reported demographic data with female predominance, clinical presentations, and associated symptoms similar to existing literature from other regions of the world[6,8,13]. Contrary to the previous studies, the present study patients were younger at diagnosis, with a median age of 3.4 years, compared with other reports of around 6 to 7 years old. This might be a reason that JDM patients in the present study had severe disease progression and a tendency to develop calcinosis as mentioned in the study by Tansley et al. (2014), which showed that children diagnosed at young age had a high risk of calcinosis[14]. In addition, the authors found that the calcinosis group had a longer median time to diagnosis, at seven months (range of 2.3 to 12.5 months) compared to JDM patients without calcinosis at 1.5 months (range of 1 to 3.5 months). These findings suggest that early diagnosis and prompt treatment are crucial in decreasing the risk of developing calcinosis.

In the current study, the authors found a relatively high rate at 62% (eight out of 13 patients) of calcinosis, compared with previous reports[6-9]. Interestingly, two out of eight patients had early calcinosis at disease onset. It is noteworthy that one patient (case 8) diagnosed at one year of age, had chronic progressive disease and suboptimal response to treatment and developed calcinosis universalis. These findings are consistent with the research that found that younger age at diagnosis and longer disease duration with delay diagnosis increased the risk of calcinosis in JDM patients[5,14].

Elevation of LDH was identified in all JDM patients in the authors' cohort. In contrast to the LDH level, 31% of the patients had a normal CK at presentation despite having muscle weakness. The CK level alone is not enough to determine disease activity and does not always correlate with severity, particularly in chronic cases[1]. Therefore, it can be suggested that all muscle enzymes, including CK, LDH, AST, ALT, and aldolase, if available, be tested to obtain baseline evaluation and to monitor effectiveness of therapy. Additionally, the present study demonstrated that raised inflammatory markers such as ESR or CRP, are not good indicators of disease flare in JDM as similarly reported in the previous study[13].

The approach to diagnosis of JDM has changed in the last decade with less use of EMG and muscle biopsy as both are invasive procedures and difficult to perform in young patients[15]. However, according to the recent guidelines, muscle biopsy is still recommended in all cases where the presentation of JDM is atypical for confirmation of the diagnosis[16]. Recently, use of short tau inversion recovery (STIR) and fat-suppressed MRI is gaining popularity to assess inflammation in muscles[17]. It is interesting to note that all JDM patients in the present study had MRI showed abnormal findings consistent with myositis. This represents a promising tool to estimate disease burden, tailor treatment, and monitor treatment efficacy. However, in Thailand and other countries with limited resources, the use of MRI may be restricted and difficult to perform without sedation in younger patients.

In terms of myositis autoantibodies, all patients who completed this test had at least one MSA positivity in the present study. This result is divergent to the previous report from Thailand by Nitiyarom et al. (2021), which found only a minority of patients

(27.8%) had MSA positivity[9]. The most common MSA found in the present study cohort was anti-NXP2 autoantibodies at 64%. As mentioned in the literature review, this myositis antibody has been associated with a higher risk of developing calcinosis and severe disease phenotype[14]. This might explain poorer prognosis in the current study compared with the previous reports from Thailand[9,18]. While the authors did not find a statistically significant difference in the prevalence of calcinosis between anti-NXP2 positive and anti-NXP2 negative patients, the present study findings suggest that a larger study might provide sufficient power to detect a significant difference, with anti-NXP2 patients having a higher risk of calcinosis.

Recently, consensus-based management recommendations of JDM have been published by SHARE (Single Hub and Access point for pediatric Rheumatology in Europe). Mainstay treatment is corticosteroid plus methotrexate. Second line medications for recalcitrant disease include hydroxychloroquine, cyclophosphamide, azathioprine, mycophenolate mofetil, cyclosporine, tacrolimus, IVIG, rituximab, infliximab, and adalimumab[16]. The present study found that a large proportion of patients (46.2%) were treated with corticosteroids plus MTX at diagnosis, compared to the earlier study from Thailand in 2001, where no patients had received MTX upfront[18]. At that time, standard treatment protocol for JDM was not available and management was mostly based on the experience of the physician. Another recent study from Thailand showed that 77.8% of JDM patients received additional immunosuppressive medication apart from steroids[9]. This represents an increase due to the new JDM guidelines. However, 23.1% of the present study patients had received only corticosteroid monotherapy at disease onset. This therapeutic regimen may not be enough to control active disease and may contribute to poorer outcomes in some patients.

The present study underscores the importance of early diagnosis and prompt initiation of appropriate treatment in JDM. As shown in case 10, who first presented with severe symptoms and serious organ involvement, after prompt and aggressive treatment, the patient achieved clinically inactive disease within one year of diagnosis and has experienced a good outcome.

Although there is no effective treatment for calcinosis yet, case reports showed an improvement in calcinosis using pamidronate[19], which was used frequently in the present study cohort (cases 3, 6, 8, and 9). Only one patient (case 9) showed a good response in resolving the calcinosis. It is important to note that this patient also received infliximab infusion as an adjunctive therapy to control disease activity. This finding supports evidence from the previous study by Campanilho-Marques et al. (2020) that reductions in muscle and skin disease, including calcinosis, were observed following treatment with infliximab and adalimumab[20].

The prognosis of JDM has remarkably improved with the advent of modern therapy. However, in developing countries, the disease still carries a risk for significant morbidity and mortality due to delay in diagnosis and resource constraints[7]. As shown in the present study, complete remission was achieved in only three patients (23%). This finding is consistent with the previous report by Sun et al. (2015) that only 33% of 39 JDM patients from a Taiwanese hospital were symptom free at last review[21]. Therefore, formulation of guidelines in resource poor settings is critical to enable appropriate timely interventions in JDM patients especially to manage patients with partial remission or chronic recurrence.

The present study findings should be interpreted in light of potential limitations. The study limitations included missing values for some tests and a relatively small number of cases identified. The reason could be the rarity of the disease in pediatric population. Under-reporting and under-documentation of cases due to the retrospective design are also to be considered. A further study with a prospective multicenter cohort is suggested to provide better quality data for better outcome assessments following treatment with the new international JDM guidelines.

Conclusion

In summary, the authors had described the demographic features, disease characteristics, diagnostic investigations, and treatment outcomes of Thai children with JDM from a large, single, tertiary referral center. With the advance of investigation tools, an MRI proved to be a sensitive tool that can assess muscle inflammation at the time of diagnosis and can also help to differentiate active and inactive disease during follow-up. The present study review underscores that JDM patients with a younger age at diagnosis will need to be monitored closely and are likely to need aggressive treatment. Prompt treatment should be managed according to the severity of symptoms to improve clinical outcomes and prevent serious complications.

What is already known on this topic?

The clinical manifestations of JDM have been extensively described in different countries. However, there is a paucity of data from developing countries, especially Southeast Asian countries.

What does this study add?

This study underscores that JDM patients with younger age at diagnosis will need to be monitored closely and are likely to need aggressive long-term treatment to prevent complications, particularly calcinosis cutis. Prompt and appropriate treatment should be managed according to the severity of symptoms to improve clinical outcomes.

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Ethical approval and consent to participate.

Written informed consent for publication was obtained from the guardian of patient. Ethical approval for the study was obtained from the Institutional Review Board (COA No.324/2020, IRB No.017/63), Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand.

Authors' contributions

CS contributed to concept and design, acquisition of data, analysis, interpretation of data, and drafting the manuscript. PK contributed to concept and design, analysis, interpretation of data, revising the manuscript, and corresponding author. All authors approved the final version of the manuscript.

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Conflicts of interest

The authors have declared no conflicts of interest.

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