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Recommendations

Treating juvenile dermatomyositis to target: Paediatric Rheumatology European Society/Childhood Arthritis and Rheumatology Research Alliance-endorsed recommendations from an international task force

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ABSTRACT

Objectives: Despite the recent prognostic improvement, a sizeable proportion of patients with juvenile dermatomyositis (JDM) respond suboptimally to contemporary therapies. This study aimed to develop recommendations for treating JDM to target.
Methods: A Steering Committee formulated a set of provisional recommendations based on evidence derived from a systematic literature review and a retrospective chart review of patients. These were discussed, amended, and voted on by an international Task Force, including 28 paediatric rheumatologists, 2 specialists in neuromuscular diseases, 1 dermatologist, 1 physical therapist, 1 research nurse, 2 patients with JDM, and 1 parent of a patient with JDM. Items that achieved at least an 80% majority vote were accepted as final recommendations.
Results: Although the literature review did not reveal trials that compared a treat-to-target strategy with a nonsteered approach, it provided indirect evidence about specific end points that could serve as targets that facilitated development of recommendations. The group reached consensus on 7 overarching principles and 12 recommendations. It was agreed that both patients/parents and treaters should share decisions in setting treatment targets and therapeutic strategies, with inactive disease as the preferred target and minimal disease activity an alternative

one. Inactive disease is targeted to be achieved within 12 months after treatment start. Interim targets include minimal and moderate clinical improvement within 6 weeks and 3 months, respectively, and normalisation of muscle strength within 6 months. High-dose glucocorticoids remain fundamental in the initial management, but progressive tapering and discontinuation within 12 months through optimisation of concomitant immunomodulatory therapy was advised. A research agenda was formulated.

Conclusions: The Task Force developed recommendations for treating JDM to target, being aware that the evidence is not strong and needs to be expanded by future research. Implementation of the recommendations in clinical practice will help to reach optimal outcomes for JDM.

INTRODUCTION

Juvenile dermatomyositis (JDM) is the most common idiopathic inflammatory myopathy in childhood. It is a multisystem vasculopathic inflammatory disease of presumed autoimmune aetiology that involves primarily the skin and the skeletal muscles [1,2]. Despite the markedly reduced mortality rates and improvement of functional outcomes for JDM over the last 30 years, there are still many patients who respond suboptimally to available therapies and experience chronic disease activity. These patients are at risk of developing cumulative disease-related or treatment-related damage and physical disability, which may have a profound impact on their quality of life [3–7].

In recent years, 2 randomised controlled trials of traditional and novel medications have been conducted in JDM [8,9]. In addition, there have been several consensus initiatives aimed to provide expert advice on the optimal management of JDM and to harmonise the therapeutic approaches internationally [10–16]. This progress has been paralleled by the development and validation of novel standardised assessment tools (Table 1) [17–21]. However, the treatment remains largely empiric and widely variable across paediatric rheumatology centres. In addition, the optimal therapeutic goals and the ideal timing of their achievement are not well established.

Studies in adults with rheumatoid arthritis have shown that patient outcomes are improved if low levels of disease activity are targeted by frequent adjustments of therapy according to quantitative indices, regardless of the therapeutic agent chosen (so-called treat-to-target [T2T] strategy) [22–25]. In recent years, the T2T approach has been introduced in the management of juvenile idiopathic arthritis and childhood-onset systemic lupus erythematosus [26–30]. Since a similar effort has not yet been made for JDM, an international Task Force (TF) was convened to reach a consensus on a set of recommendations aimed at defining a T2T strategy for JDM.

METHODS

The project was initiated by authors AR and BMF, who invited paediatric rheumatologists from Europe and North America (CH, LMC, AMR, and LGR), selected on the basis of publication records, expertise in treating JDM and previous participation in similar activities, to form a Steering Committee (SC). The first online meeting of the SC (May 11, 2023) was aimed to discuss the unmet needs in the treatment of JDM and the potential for adopting a T2T strategy. It was unanimously agreed that defining therapeutic goals and an appropriate targeted treatment approach would be beneficial, but there was concern that underlying evidence could be lacking. It was, therefore, decided that – in accordance with the European Alliance of Associations for Rheumatology (EULAR) standardised operating procedures for recommendation efforts [31,32] – a comprehensive systematic literature review (SLR) to address the current evidence in JDM was a necessary initial step of the process. After the formulation of a series of search questions (Supplementary Table S1), the SLR was performed by authors SR, JMM, TB, and AIR-G, with the help of a research librarian.

The SLR results (which will be described in a separate manuscript) were presented to the SC at a subsequent online meeting (March 27, 2024). The literature search revealed that no randomised controlled trials that addressed a target-oriented, steered therapy in comparison with conventional management had been published in JDM. Indirect evidence on optimal therapeutic approaches was, however, available to inform the next stages of the process. On this basis, the SC formulated a provisional set of recommendations.

In the meantime, a TF including 22 additional paediatric rheumatologists practicing in various areas of the world (Africa, Asia, Australia, Europe, South America, and North America), 2 specialists in neuromuscular diseases, 1 dermatologist, 2 allied health professionals (1 physical therapist and 1 research nurse), 2 patients with JDM and 1 parent of a patient with JDM was gathered. These invitations were based on individuals' contributions to the field, experience with JDM clinical care and research, and deliberations among members of the SC.

The provisional overarching principles and recommendations were, then, shared with the TF members through an online consensus survey, which remained open from July 8 to August 28, 2024. Respondents were asked to provide their agreement or disagreement about each recommendation, as well as the reasons for disagreement and suggestions for changes. The survey was completed by 92.5% of TF members, and the percentage of agreement was >80% for 17 of the 19 proposed statements (Supplementary Tables S2 and S3). The results of the survey were examined by the SC at an online meeting (September 4, 2024). Based on the comments provided by the respondents, some recommendations were amended or reworded.

To provide a further background to the process from the real world of clinical practice, a retrospective chart review of patients with JDM followed up at 2 tertiary care paediatric rheumatology centres was performed by 4 authors (SR, JMM, TB, and AIR-G). The objective of this exercise (whose findings will be reported in a separate manuscript) was to gain insights into optimal targets [17–21,33,34] and timing of their achievement during the first 2 years after disease diagnosis. The key data obtained from the retrospective chart review are summarised in Supplementary Table S4.

The preliminary recommendations, as well as the results of SLR and retrospective chart review, were presented and discussed at a final consensus conference, which was convened in Genoa, Italy, on October 7-8, 2024 and was attended in person by all but 1 member of the TF. A modified nominal group technique [35] was used to ensure equal participation among TF members. Participants were randomly divided into 3 breakout

Table 1

Instruments and criteria used for assessment of disease activity and treatment response in JDM

	Items included														
	PhGA	PaGA	CMAS	MMT8	hMC	Skin VAS	Skin DAS	Total DAS	EM VAS	CK	Muscle enzymes[a]	CHAQ	CHQ-PhS	Requirements for classification	Ref
Criteria for clinically inactive disease															
PRINTO criteria	X		X	X						X				At least 3 of the following 4 criteria: CK ≤ 150 U/L, CMAS ≥ 48, MMT8 ≥ 78 and PhGA ≤ 0.2 cm of 10 cm	[19]
Modified PRINTO criteria	X		X	X						X				PhGA ≤ 0.2 cm of 10 cm and at least 2 of the following 3 criteria: CK ≤ 150 U/L, CMAS ≥ 48, MMT8 ≥ 78	[20]
JDMAI1 criteria	X	X			X	X								Total score ≤ 2.4	[18]
JDMAI2 criteria	X	X			X		X							Total score ≤ 5.2	[18]
Criteria for minimal (or low) disease activity															
JDMAI1 criteria	X	X			X	X								Total score ≤ 6.6	[18]
JDMAI2 criteria	X	X			X		X							Total score ≤ 8.5	[18]
2016 ACR/EULAR JDM response criteria															
Based on IMACS core set	X	X		X					X		X	X		Minimal ≥ 30	[21]
Based on PRINTO core set	X	X	X					X				X	X	Moderate ≥ 45 Major ≥ 70	

ACR, American College of Rheumatology; CHAQ, Childhood Health Assessment Questionnaire; CHQ-PhS, Physical Summary Score of the Child Health Questionnaire; CK, creatine kinase; CMAS, Childhood Myositis Assessment Scale; DAS, Disease Activity Score; EM, extramuscular; EULAR, European Alliance of Associations for Rheumatology; hMC, hybrid MMT8/CMAS; IMACS, International Myositis Assessment and Clinical Studies Group; JDM, juvenile dermatomyositis; JDMAI, Juvenile DermatoMyositis Activity Index; MMT8, Manual Muscle Testing 8; PaGA, Parent's global assessment of child's overall wellbeing; PhGA, Physician's global assessment of overall disease activity; PRINTO, Paediatric Rheumatology International Trials Organisation; VAS, visual analogue scale.

[a] Muscle enzymes: CK, lactate dehydrogenase, aspartate aminotransferase, alanine aminotransferase, aldolase.

tables, moderated by 3 authors (AR, BMF, and AC), who did not vote. Extensive discussions took place during these sessions, and the suggested wording was reformulated as deemed appropriate, with majority votes where controversy emerged. The results obtained by the breakout groups were reported to the whole TF, which then discussed the proposals, amended them, and arrived at final wordings that were subjected to an electronic poll (Poll Everywhere) [36]. Items that achieved at least an 80% majority vote were accepted as final recommendations.

After the consensus meeting, all participants were asked to adjudicate via e-mail their level of agreement with each overarching principle and recommendation on a 0 to 10 scale (0 = no agreement at all; 10 = full agreement). The evaluation of the level of evidence (LoE) and strength of recommendation was based on the Oxford Evidence Based Medicine categorisation [37].

RESULTS

The individual statements that received a positive vote comprised 7 overarching principles and 12 recommendations. These items are shown in Tables 2 and 3, respectively, together with the percentage of positive votes obtained at the consensus conference, LoE, strength of recommendation, and level of agreement, and are discussed in detail further.

The evidence base

The SLR led to retrieve 2714 studies from the scrutinised databases, with additional 47 abstracts identified through citation searching and 23 flagged by SC members. Of these, 300 underwent a full-text review process and 220 were included in the final review (Supplementary Fig S1). The SLR revealed that no randomised controlled trials had compared a targeted therapeutic approach with conventional therapy in JDM. There was, therefore, no direct evidence that a T2T strategy was superior to nontargeted management. However, indirect support regarding the time that should elapse before escalating treatment in a patient with persistently active disease was provided by 2 randomised clinical trials, noncontrolled therapeutic studies, observational investigations, and therapeutic recommendations formulated by expert panels [8–15,38–43]. Nevertheless, the LoE for the development of recommendations was anticipated to be low and mainly based on expert consensus.

Overarching principles

A. The treatment targets and the therapeutic strategy should be based on shared decisions between patients, caregivers and the multidisciplinary healthcare team.

It was acknowledged that involvement of the parents/caregivers and, where appropriate, the patient in therapeutic decision-making is crucial and may enhance adherence to treatment and potentially improve the outcome. Although it was highlighted that the management of patients with JDM should be ideally conducted by professionals with specific paediatric expertise, it was argued that not all children will have access to paediatric rheumatology care and that adolescents and young adults should also be considered. For these reasons, it was decided to remove the adjective paediatric, which was placed in the initial formulation before the multidisciplinary health care team. This item was endorsed unanimously.

B. Treatment approaches should be based on all of an individual patients' disease characteristics, taking into account all available findings (including, but not limited to, laboratory findings, autoantibody profile, capillaroscopy findings, imaging, and histopathology features).

It is well established that considerable heterogeneity exists among patients with JDM in terms of clinical phenotype, disease severity, course, and outcomes. This variability implies that the therapeutic choices, optimal targets, and treatment strategy should be tailored to the prevalent forms of organ involvement and the overall level of disease activity. In recent years, it has been shown that this heterogeneity also involves the presence of a wide array of myositis-specific autoantibodies, which can be identified in ~60% of children with JDM and have been found to define homogeneous subgroups of patients with similar clinical features, response to therapy, and prognosis [44–48]. Several studies have demonstrated the diagnostic and prognostic value of nailfold capillaroscopy in JDM [49–57]. In recent years, magnetic resonance imaging (MRI) has played an important role, both in the diagnosis of JDM and in helping monitor myositis activity [58–60]. Muscle biopsy remains an important investigation in the diagnostic workup of a subset of patients with JDM, especially in the absence of skin rash or in an otherwise atypical presentation [61,62]. Histopathology findings may predict disease prognosis [63]. Because emerging evidence indicates that the abovementioned tools may influence the therapeutic approach, participants believed it important to include them all in the statement. However, concern was raised that some diagnostic methods may not be available in all centres, particularly in lower-resource countries. It was also argued that novel JDM biomarkers, such as galectin-9, CXCL10, and Siglec-1 expression on monocytes, may deserve consideration in the future [64,65]. For this reason, the sentence 'but not limited to' was incorporated. The term imaging was preferred over muscle MRI in the consideration of the growing adoption of muscle ultrasonography [66–68]. After merging these suggestions, this item achieved 97% of participants' votes.

C. The goals of treating patients with JDM are to control signs and symptoms, to prevent organ damage and disease complications, including calcinosis and lipodystrophy, and to prevent flares. These objectives should be coupled with avoidance of drug toxicity and treatment complications and with optimisation of physical function, mental wellbeing, growth and development, quality of life, education and social participation.

This item was unanimously endorsed by TF members, after several amendments. It was agreed to give priority to the control of inflammatory signs and symptoms, followed by prevention of organ damage and disease complications, especially calcinosis and lipodystrophy. All participants concurred that caution should apply particularly to systemic glucocorticoids, whose side effects may have a devastating impact in the paediatric age. Optimisation of mental wellbeing, as well as growth and development, was incorporated in the therapeutic goals to highlight the paediatric specificity of the recommendations.

D. The management of JDM requires an understanding of its diverse manifestations and should be tailored, using patient-centred, individualised treatment strategies and a multidisciplinary approach.

The original term targeted was replaced by tailored, which was deemed more appropriate to express the need to adapt the therapeutic approach to the particular disease phenotype. It was also emphasised that the management of patients with JDM requires the involvement of a multidisciplinary team of specialists. This item was unanimously endorsed.

Table 2
Overarching principles for T2T in JDM

Overarching principles	Positive votes at consensus conference (%)	Level of evidence	Strength of recommendation	Level of agreement (mean ± SD)
A. The treatment targets and the therapeutic strategy should be based on shared decisions between patients, caregivers and the multidisciplinary healthcare team.	100	5	D	10 ± 0.2
B. Treatment approaches should be based on all individual patients' disease characteristics, taking into account all available findings (including, but not limited to, laboratory findings, autoantibody profile, capillaroscopy findings, imaging, and histopathology features).	97	5	D	9.9 ± 0.5
C. The goals of treating patients with JDM are to control signs and symptoms, to prevent organ damage and disease complications, including calcinosis and lipodystrophy, and to prevent flares. These objectives should be coupled with avoidance of drug toxicity and treatment complications and optimisation of physical function, mental wellbeing, growth and development, quality of life, education, and social participation.	100	5	D	10 ± 0.2
D. The management of JDM requires an understanding of its diverse manifestations and should be tailored, using patient-centred, individualised treatment strategies and a multidisciplinary approach.	100	5	D	9.9 ± 0.4
E. Use of glucocorticoids remains key to achieve early disease control. The dosage should be progressively reduced to the lowest level needed to maintain disease control and, when possible, discontinued through the optimisation of concomitant immunomodulatory therapy.	94	2b	B	9.9 ± 0.3
F. Treat-to-target requires regular assessment of the patient and adjustment of therapy until the desired target is reached.	91	5	D	9.8 ± 0.4
G. Patient monitoring should include assessment of treatment acceptability, adherence, and adverse events.	88	5	D	9.7 ± 0.9

JDM, juvenile dermatomyositis; T2T, treat-to-target.
Level of evidence ranges from 1a (systematic review of randomised controlled trials) to 5 (expert opinion); strength of recommendation ranges from A (directly based on category I evidence) to D (directly based on category IV evidence or extrapolated recommendation from category II or III evidence); agreement: percentage of experts agreeing with the recommendation statement; level of agreement ranges from 0 (fully disagree) to 10 (fully agree).

E. Use of glucocorticoids remains key to achieve early disease control. The dosage should be progressively reduced to the lowest level needed to maintain disease control and, when possible, discontinued through the optimisation of concomitant immunomodulatory therapy.

High-dose glucocorticoids remain the mainstay of the initial management of JDM, especially in patients with acute or life-threatening manifestations. However, in consideration of the serious side effects related to the prolonged administration of glucocorticoids, the recommendation to decrease progressively their dosage to the minimum level that ensures maintenance of adequate disease control was widely supported. This goal may be facilitated by the concomitant administration of immunomodulatory drugs that have shown a glucocorticoid-sparing potential in JDM. This item obtained 94% of votes.

F. Treat-to-target requires regular assessment of the patient and adjustment of therapy until the desired target is reached.

Although the SLR had produced only indirect evidence for the utility of the T2T strategy in JDM, the large majority of TF members were in favour of including regular measurement of disease activity and adjustment of therapy with persistently active disease, in the overarching principles. Nonetheless, the value of broadening the concept of patient assessment to encompass, beside disease activity, the whole burden/impact of the disease on patient's daily life was underscored. This item achieved 91% of participants' votes.

G. Patient monitoring should include assessment of treatment acceptability, adherence, and adverse events.

During breakout sessions, a debate arose about whether this statement should be kept separated or merged with principle F. It was finally decided to leave it as a standalone principle. However, because treatment adherence can be affected by tolerability and side effects, the terms acceptability and adverse events were added to the original formulation. Furthermore, the previous word therapy was replaced with the broader term treatment to extend the advice to non-medication therapies, such as physical therapy. After these refinements, approval was granted by 88% of the participants.

Recommendations

1. The primary target for treatment of patients with JDM is the state of inactive disease, which means the absence of signs and symptoms of inflammatory disease activity in muscles and skin as well as in other organs/systems.

Thus far, no clinical trial in JDM has compared progression of cumulative organ damage or improvement in quality of life when inactive disease rather than another state is targeted. However, there is indirect evidence that disease-related damage or complications are more effectively prevented when inactive disease is achieved [3,4,55,56,69–71]. Considering that the recent therapeutic advances enable reaching clinical remission in the majority of patients with JDM [72,73], inactive disease was set as the primary therapeutic target. The definition of inactive disease was intended to be quite strict, that is, as complete absence of all signs and symptoms of inflammatory disease activity not only in the skin and muscles but also in other affected organs. Although 91% of voters agreed with this statement, concern was raised that setting the state of inactive disease as principal therapeutic target may imply that only physician-centred goals should be pursued. Many participants thought that restoring normal function may be equally

Table 3
Recommendations for T2T in JDM

Recommendations	Positive votes at consensus conference (%)	Level of evidence	Strength of recommendation	Level of agreement (mean ± SD)
1. The primary target for treatment of patients with JDM is the state of inactive disease, which means the absence of signs and symptoms of inflammatory disease activity in muscles and skin as well as in other organs/systems.	91	2b	C	9.7 ± 0.7
2. Minimal (or low) disease activity may be an acceptable primary target when inactive disease cannot be achieved, as in more refractory cases.	87	2c	D	9.6 ± 0.8
3. Setting the target, selecting the measurement tools and decisions about treatment should all be based on individual patient characteristics and agreed upon with the caregivers/patient.	94	5	D	9.8 ± 0.5
4. Disease activity must be assessed and documented regularly using standardised and validated assessment tools.	94	2c	C	9.9 ± 0.4
5. Initial target I. Within 6 wk after starting treatment, at least a minimal clinical improvement[a] should be achieved. In patients with life-threatening conditions, a significant clinical improvement should be targeted in a shorter time frame.	97	5	D	9.5 ± 0.7
6. Interim target II. Within 3 mo after starting treatment, at least a moderate clinical improvement[b], as well as normalisation of muscle enzymes, should be achieved. Patients should be free of life-threatening conditions at this time.	88	5	D	9.2 ± 0.9
7. Interim target III. Normalisation of muscle strength should occur within 6 mo.	86	5	D	9.2 ± 0.9
8. The primary target, the state of inactive disease[c], should be achieved within 12 mo.	100	5	D	9.6 ± 0.6
9. Glucocorticoids should be aggressively tapered as long as interim targets are reached; glucocorticoids should be discontinued, ideally, within 12 mo or sooner[d].	100	2b	C	9.8 ± 0.5
10. Regular assessment is recommended to ensure that the patient is on the correct trajectory to reach the target. The frequency of assessments depends on the level of disease activity and severity and the presence of complications.	100	5	D	9.9 ± 0.3
11. Treatment should be adjusted until the desired target is achieved.	97	2b	C	9.9 ± 0.3
12. Once the primary target has been achieved, it should be sustained. Ongoing monitoring should occur to ensure maintenance of the target.	100	2b	C	10 ± 0.0

JDM, juvenile dermatomyositis; T2T, treat-to-target.
Level of evidence ranges from 1a (systematic review of randomised controlled trials) to 5 (expert opinion); strength of recommendation ranges from A (directly based on category I evidence) to D (directly based on category IV evidence or extrapolated recommendation from category II or III evidence); agreement: percentage of experts agreeing with the recommendation statement; level of agreement ranges from 0 (fully disagree) to 10 (fully agree).

[a] As defined by a validated response criteria or 25% improvement in a composite disease activity measure or in the physician global assessment.
[b] As defined by a validated response criteria or 50% improvement in a composite disease activity measure or in the physician global assessment.
[c] As defined in recommendation 1.
[d] If target is not achieved, favour additional/change of immunomodulatory treatment, over repeatedly increasing glucocorticoid dose.

important as reaching inactive disease in the management of patients with JDM. Furthermore, some experts would have preferred to name the primary target clinically inactive disease to highlight the potential role of biomarkers or imaging techniques in defining disease remission more reliably than clinical assessment. However, despite emerging evidence for biomarkers and MRI reflecting subclinical inflammation, at present, there is no established role for these methodologies in defining remission.

2. Minimal (or low) disease activity may be an acceptable primary target when inactive disease cannot be achieved, as in more refractory cases.

Although the TF did not intend to replace the target of inactive disease with that of low disease activity, it was recognised that stringent inactive disease, as defined in recommendation 1, may be difficult to achieve in some patients, especially those with long-standing or refractory disease. Low disease activity is differentiated from the state of inactive disease by the persistence of a few residual signs and symptoms. However, it is assumed that physical function and quality of life would not be substantially worse than in inactive disease and that progression of organ damage, while possibly not halted, would be minimal. This item achieved 87% of participants' votes.

3. Setting the target, selecting the measurement tools and decisions about treatment should all be based on individual patient characteristics and agreed upon with the caregivers/patient.

This recommendation emphasises the need to individualise the therapeutic target, the method used for its assessment, and the therapeutic decisions based on patients' characteristics, which include the severity and extent of muscle and skin disease, involvement of visceral organs, presence of disease-related complications, especially dystrophic calcinosis and lipodystrophy, and comorbidities (eg, osteoporosis, growth failure, hypertension, and infection). Therapeutic decision-making may be guided by published consensus-based recommendations and treatment plans for the management of JDM [10–15]. This item was approved by 94% of the participants.

4. Disease activity must be assessed and documented regularly using standardised and validated assessment tools.

There was unanimous consensus that the use of standardised and validated assessment tools is the best method to evaluate disease activity and response to therapy in JDM. During the consensus meeting, the relative measurement properties and suitability for the T2T strategy of the existing tools [17–21,74–76] were discussed. It was finally decided not to recommend the use of a specific instrument, but to leave the choice open for the clinician. Hence, the neutral term standardised and validated assessment tool was endorsed by 94% of voters.

5. Initial target I. Within 6 weeks after starting treatment, at least a minimal clinical improvement (as defined by a validated response criteria or 25% improvement in a composite disease activity measure or in the physician global assessment) should be achieved. In patients with life-threatening conditions, a significant clinical improvement should be targeted in a shorter time frame.

This statement was subject to intense debate. Some attendees argued that the application of validated response criteria in clinical practice is time-consuming and proposed the alternative use of the simpler physician global assessment. There was much interest for composite disease activity measures, such as the Juvenile DermatoMyositis Activity Index (JDMAI) [17], which were thought to be feasible for regular assessments in routine clinics. A 6-week time frame was considered too long for the most severe patients (eg, those with intestinal vasculitis or rapidly progressive interstitial lung disease). Conversely, other participants objected that the proposed magnitude and time of improvement could be too ambitious for some patients. In the end, it was widely agreed that although setting a therapeutic goal that fit all patients is unrealistic; it is, nevertheless, fundamental to prompt an early therapeutic change if a satisfactory improvement is not observed within 6 weeks after treatment start. This item was approved by 97% of the participants.

6. Interim target II. Within 3 months after starting treatment, at least a moderate clinical improvement (as defined by a validated response criteria or 50% improvement in a composite disease activity measure or in the physician global assessment), as well as normalisation of muscle enzymes should be achieved. Patients should be free of life-threatening conditions at this time.

As for interim target I, there was strong discussion about this proposal for interim target II. Some TF members wanted to eliminate the requirement of muscle enzymes normalisation within 3 months as it may be difficult to attain and does not apply to amyopathic disease. Some participants were in favour of keeping patients with more aggressive disease separated from those with mild-to-moderate involvement. Others argued that setting diverse targets for different degrees of disease severity could make the recommendations too complex and cumbersome. To reconcile these disparities, a sentence was added to underscore the need to ensure that life-threatening disease manifestations should no longer be present at 3 months after treatment start. After this change, the item was endorsed by 88% of the participants.

7. Interim target III. Normalisation of muscle strength should occur within 6 months.

Some participants contended that a 6-month time frame is excessively brief for patients who present with extreme muscle weakness, for whom a 9-month interval could be more acceptable. Consensus was finally reached to maintain the proposed timeline for all disease phenotypes. It was, however, advised that in case complete recovery of muscle strength is not obtained by the suggested time point, the treating physician should evaluate whether not only the patient needs an intensification of drug regimen but also a focused physiotherapy intervention could help to improve energy and stamina or whether persistent muscle weakness is due to glucocorticoid-induced myopathy. In patients with long-standing disease, the possible contribution of muscle damage or atrophy should be accounted for. Approval of this item was provided by 86% of the participants, with contrary votes being mostly explained by the disagreement about the 6-month time frame.

8. The primary target, the state of inactive disease (as defined in recommendation 1), should be achieved within 12 months.

This statement was endorsed unanimously. Indeed, all participants agreed that the target of inactive disease should be set at 12 months. It was emphasised that normalisation of muscle strength is not necessarily equivalent to complete abrogation of muscle inflammation.

9. Glucocorticoids should be aggressively tapered as long as interim targets are reached; glucocorticoids should be discontinued, ideally, within 12 months or sooner (if target is not achieved, favour additional/change of immunomodulatory treatment, over repeatedly increasing glucocorticoid dose).

Although all participants agreed on the merit of addressing glucocorticoid tapering and to consider it as a treatment target,

this statement was discussed in depth and modified several times, due to divergence about schemes of glucocorticoid tapering. Bearing in mind that the T2T strategy is not intended to provide indications for use of specific treatments, it was finally agreed to make a general statement that underlined the key importance of aggressive tapering and discontinuation of glucocorticoids. As a cautionary note, the need for verifying attainment of interim targets before decreasing glucocorticoid dosage was included. However, the concept that aggressive glucocorticoid tapering should be pursued was strengthened by indicating that the ideal timing for glucocorticoid discontinuation is 12 months after their start. The final version of the statement was endorsed unanimously.

10. Regular assessment is recommended to ensure that the patient is on the correct trajectory to reach the target. The frequency of assessments depends on the level of disease activity and severity and the presence of complications.

Owing to the clinical heterogeneity of JDM, the intervals between clinical evaluations vary in relation to the disease phenotype, level of disease activity, and presence of complications or comorbidities. In patients with profound muscle weakness and extensive skin rash or life-threatening disease manifestations, such as interstitial lung disease or intestinal vasculitis, frequent evaluations (even daily) of the disease status and course are required to ensure the necessary therapeutic adjustments in a timely manner. Patients with less severe disease require less frequent evaluations, which may occur weekly to every 3 months, depending on the degree of disease activity. Patients in sustained remission can be assessed at looser intervals, approximately every 4 to 6 months, to ensure maintenance of the target reached and, in those who are still receiving therapy, to watch the occurrence of adverse events and avoid overtreatment. This recommendation was approved by 100% of the participants.

11. Treatment should be adjusted until the desired target is achieved.

Indirect evidence indicates that early clinical improvement or the achievement of inactive disease is associated with better long-term outcome [3,4]. It is, thus, likely that pursuing the best possible target through treatment adjustments improves prognosis. Several TF members emphasised the value of non-pharmacologic interventions, particularly physiotherapy and rehabilitation, occupational therapy, optimisation of bone health and muscle fitness, management of fatigue, and psychological support. This recommendation was supported by 97% of the participants.

12. Once the primary target has been achieved, it should be sustained. Ongoing monitoring should occur to ensure maintenance of the target.

There are rationale and evidence, in JDM, that sustained/persistent remission leads to an optimal quality of life, enhances physical function, and stops progression of organ damage [3,4,55,69–71,77,78]. Conversely, an increase in disease activity during follow-up may lead to reduced quality of life and progression of cumulative damage [79,80]. Maintenance of the treatment target does not necessarily imply maintenance of treatment. Unfortunately, there is currently a lack of evidence-based data or guidelines to aid in the withdrawal of medications after disease remission in JDM. Furthermore, no reliable clinical, biomarker, or imaging indicators are currently available to identify patients at higher risk of experiencing disease flare after treatment discontinuation. This item was unanimously approved.

Adjudication of the level of agreement after the consensus meeting

The level of agreement on overarching principles and recommendations adjudicated by the TF members after the Genoa consensus meeting was very high, as all items achieved an average score greater than 9, and only 2 items had an average score lower than 9.5 (Tables 2 and 3).

DISCUSSION

We present in this study the overarching principles and specific recommendations for the T2T strategy in JDM, which were developed by a TF including a broad range of international experts and were endorsed by the Paediatric Rheumatology European Society (PReS) and the Childhood Arthritis and Rheumatology Research Alliance (CARRA). These were based on published evidence available to date, as well as on a retrospective review of 2 patient series, and were derived from an online consensus survey and extensive discussions conducted using a modified nominal group technique approach. Considering the wide international representation within the TF and the excellent level of agreement reached for all statements, the result of the effort will likely gain widespread acceptance.

The recommendations are primarily intended to provide guidance on general treatment approaches, with the aim of allowing for an optimal outcome of JDM in the individual patient. A remarkable difference with the previous treatment recommendations or treatment plans is the absence of indications or advice regarding specific agents or classes of medications – apart from the suggestion about tapering and discontinuation of glucocorticoid therapy and avoidance of long-term glucocorticoid administration. Hence, these recommendations should be applicable in all regions and countries, irrespective of medication availability. Although the recommendations are primarily aimed at paediatric rheumatology practitioners and other health professionals involved in the care of patients with JDM, they may also be of interest to official bodies, such as health authorities or payers, who may wish to explore clinical practice in their environment, as well as to clinical trialists and regulatory agencies, owing to the growing interest of pharmaceutical industries in trials evaluating a targeted therapeutic approach in comparison with conventional therapy in JDM.

The process was initiated by a SC, which followed the EULAR standardised operating procedures for the development of recommendations [31,32]. A large TF of 36 additional members was convened, which comprised 28 paediatric rheumatologists, 2 specialists in neuromuscular diseases, 1 dermatologist, 1 physical therapist, 1 research nurse, 2 patients with JDM, and 1 parent of a JDM patient. Ensuring input from a multidisciplinary group of paediatric specialists and allied health professionals was an important strength of the project. Participation of patients and parents was also critical to incorporate the view of these stakeholders and ensure that their needs were addressed, particularly when considering T2T potential side effects and quality of life as well as prevention of damage and disability.

Ideally, treatment recommendations should be based on available evidence. However, clinical trials focused on the comparison between continued treatment modification to reach a prespecified treatment target and a nontargeted approach do not exist for JDM. Although the SLR and the retrospective chart review provided indirect evidence about specific end points that could serve as targets, the proposed recommendations can only

be regarded as consensus-based expert opinion and, therefore, require additional research efforts.

Several TF members argued that the recommendations did not account for potential financial constraints or access to costly therapies. However, studies in adult patients with rheumatoid arthritis have shown that a good outcome can be obtained in a large proportion of patients with easily accessible and affordable therapies, as long as a strategic treatment approach is implemented [22,23,81]. A similar issue was raised regarding the availability of certain investigations, such as MRI, myositis-specific autoantibodies, or capillaroscopy, which may be valuable to define the best therapeutic approach. This problem was accounted for by recommending consideration for their results, but only when obtainable.

The TF agreed unanimously that a pivotal aspect in the care of JDM is the shared decision-making with the patient and the caregivers. There was also full consensus that control of sign and symptoms is the main objective in the treatment of JDM. However, other key goals should be pursued concomitantly, including prevention of organ damage, disease flares, and complications. A further item specific to paediatric patients is the optimisation of linear growth and pubertal development. The TF emphasised that care of adolescent patients be tailored in accordance with the broader process of transition from paediatric to adult rheumatology care. Parent/patient representatives put a particular emphasis on restoring physical and emotional functioning and wellbeing as well on avoidance of medication side effects.

The TF set the state of inactive disease as the primary target for treatment, with minimal (or low) disease activity as alternative goal. Although some experts highlighted the potential superiority of more stringent targets, such as remission by biomarkers or muscle MRI over clinical assessment, the majority considered that at present remission should be defined on clinical grounds through the use of one of the existing criteria, as the evidence for other methods is still limited.

Beside the overall therapeutic goals, interim targets as well as the timing for their achievement after treatment start were set. These included minimal and moderate clinical improvement within 6 weeks and 3 months, respectively, normalisation of muscle strength within 6 months, and inactive disease within 12 months. Although some consensus conference participants argued that the time frame of initial targets was too long for the most severe patients, whereas others warned that reaching a normal muscle strength within 6 months could prove difficult in patients with profound muscle impairment, all proposals were approved, with the commitment of verifying the feasibility of the targets by future investigations in clinical settings. In setting the timeline for treatment adjustments, the variability of time of onset of effect of some medications used to treat JDM should be taken into account.

It was recognised that the initial management of JDM relies on high-dose glucocorticoids, especially in patients with the most severe presentations. However, in consideration of the devastating side effects related to the prolonged administration of glucocorticoids in children, the indication for their gradual tapering till reaching the minimum dosage required to maintain the disease control and, when possible, discontinuation through the optimisation of concomitant immunomodulatory therapy was incorporated. The ideal time for their discontinuation was set at 12 months. The established treatment targets and the optimal time of their achievement are outlined in Supplementary Figure S2.

In conclusion, the recommendations to treat JDM to target have been developed by an international TF. Their implementation in clinical practice may improve patient care and outcomes for patients with JDM. The research agenda should prioritise testing of the recommendations in randomised controlled trials that compare a targeted therapeutic approach with conventional treatment and other objectives listed in the Box.

Box Research agenda

- Implementation of randomised trials aimed to show the superiority of a targeted treatment approach based on treat-to-target over a nontargeted approach
- Acceptance and applicability of treat-to-target strategies in standard clinical practice
- Acceptance and feasibility of treat-to-target strategies in low-resource countries
- Evaluation of the impact of treat-to-target in terms of cost, safety, and treatment burden compared with usual care
- Impact of parent/patient evaluation, particularly in the presence of marked fatigue, in the assessment of targets
- Impact of diagnostic delay in the achievement of targets
- Clarification of the role of myositis-specific autoantibodies in designing treatment strategies and defining targets
- Clarification of the role of capillaroscopy findings in designing treatment strategies and defining targets
- Role of emerging biomarkers (eg, interferon-related biomarkers) in the assessment of targets
- Role of imaging methods in definition and assessment of targets, especially remission of muscle disease
- Role of muscle histopathology in designing treatment strategies and defining targets
- Analysis of the optimal modalities of tapering and/or withdrawing treatments in patients reaching inactive disease
- Development and validation of widely accepted outcome measures for skin disease
- Evaluation of additive targets meaningful to patients and parents, such as restoring normal physical and emotional function and wellbeing

Competing interests

All authors declare they have no competing interests related to the present work.

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Contributors

AC, AR, AMR, BMF, CHH, FB, LGR, LJM, and SR developed the online consensus surveys, organised and facilitated the consensus meeting, and wrote the first version of the manuscript. AIR-G, JMM, SR, and TB performed the systematic literature review and the retrospective chart review. AMH, ANM, BB-M, CB, CF, CM, CPa, CPi, ES, GT, HS, LC, LRW, MA, MJ, MMK, NAH, NT, OK, PL, RC-M, RM, RN, SC, SET, SK, SM, SS, and TM participated as voting members of the consensus meetings, contributing to decisions regarding items to be included in the overarching principles and recommendations. All authors reviewed and approved the final draft of the manuscript.

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Patient and public involvement statement

Patients and parents from Europe and the USA actively participated in the design and conduct of this research. The Task Force included 1 parents and 2 young adults with juvenile dermatomyositis, now older than 18 years, providing patient-centred perspectives. They participated in the online consensus survey and attended the in-person consensus meetings with full, equal voting rights. They were involved in all discussions that led to the final formulations of overarching principles and recommendations. Patients and parents reviewed and approved the manuscript and are appropriately included as named authors.

Patient consent for publication

Not applicable.

Ethics approval

The study protocol was approved by the Ethics Committee of Regione Liguria on 18/09/2023, number 209/2023.

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Supplementary materials

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