intervals would be an efficient use of resources whilst being safe for patients. Furthermore, we provide evidence that less frequent blood monitoring may be feasible in patients on combination DMARDs, which is particularly relevant in an era where combination treatment is the standard of care in inflammatory arthritis.

Funding: No specific funding was received from any funding bodies in the public, commercial or not-for-profit sectors to carry out the work described in this manuscript.

Disclosure statement: The authors have declared no conflicts of interest.

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Accepted 25 March 2019

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Rheumatology 2019;58:2062–2064

doi:10.1093/rheumatology/kez226

Advance Access publication 14 June 2019

An international survey of developing classification criteria for juvenile dermatomyositis-scleroderma overlap

Rheumatology key message

- Definition of JDM-scleroderma overlap phenotype is important to tailor patients' management.

SIR, JDM is a heterogeneous autoimmune-mediated disease, which may overlap with other connective tissue disorders, including SSc, SLE and MCTD [1, 2]. However, there is no internationally agreed definition of 'JDM-scleroderma overlap' syndromes in children, and a paucity of published data on the clinical characteristics, laboratory features and outcome in this specific group. In adults with dermatomyositis, scleroderma overlap commonly presents with sclerodactyly, facial skin changes, and interstitial lung disease and gastrointestinal involvement, and may progress to include many SSc features [3]. Some authors suggest that myositis with features of scleroderma should be considered a separate entity known as 'scleromyositis' [4], but in JDM, 'overlap' features may develop after an initial classical JDM presentation [5]. The goal of this survey was to define the clinical characteristics of JDM-scleroderma overlap and identify whether experienced clinicians believe overlap features may affect outcome and management to support the development of clusters based on clinical features and biomarkers to help tailor patients' management.

An initial Delphi survey compiled by two clinicians (P.K. and S.C.-L.) and approved by the UK Juvenile Dermatomyositis Cohort & Biomarker Study (JDCBS) Steering committee was circulated to 152 members of the Juvenile Dermatomyositis Research Group and the Paediatric Rheumatology European Society JDM working group. The survey listed clinical criteria expected to be helpful and clinically relevant in making the diagnosis of JDM-scleroderma overlap. Each individual was asked to identify the most characteristic clinical manifestations, to rank them in terms of frequency, and to give an expert opinion with regards to outcome and management. The survey attempted to capture the clinical and serologic features that play a role in the diagnosis of overlap and whether autoantibodies influence the outcome and management. Survey respondents were also asked if they had treated JDM-scleroderma overlap patients. The survey was circulated to members by e-mail.

The survey had a response rate of 26.9% (41 individuals) from both the Juvenile Dermatomyositis Research Group and the Paediatric Rheumatology European Society JDM working party. Some 89% of participants who responded reported that they had seen patients with JDM-scleroderma overlap. Results of this survey are shown in Fig. 1. The most common features reported to occur in JDM patients with scleroderma overlap were sclerodactyly, sclerodermatous skin change proximal to MCP joints, RP and ulceration at the tip of fingers. A total of 95.1% of responders thought the scleroderma overlap presentation influenced the outcome of JDM, while 86.4% agreed that these features influence the choice of treatment.

Myositis-associated autoantibodies occurring in SSc overlap (anti-PM-Scl, anti-U1-RNP, anti-Ku) were thought to be the most commonly associated autoantibodies with JDM-scleroderma overlap (78%), followed by SSc-specific autoantibodies (anti-Scl70, anti-RNA polymerase III) (44%), anti-synthetase autoantibodies (anti-JO1, anti-PL7,

Fig. 1 Clinical features reported to occur in JDM-scleroderma overlap patients

The top four characteristics ranked in terms of frequency are sclerodactyly, sclerodermatous skin change proximal to MCP joints, RP and ulceration at the tip of fingers, respectively.

anti-PL12) (16.41%) and other antibodies, such as anti-Ro (8.21%). Interestingly, all responders (100%) thought that negative myositis autoantibodies did not exclude the diagnosis JDM-scleroderma overlap, but almost 11% of participants felt that myositis antibodies were needed to diagnose this subgroup of JDM. Although 83% felt that autoantibody profile influences the outcome, only 49% thought that autoantibody profile would influence the choice of medication.

To the best of our knowledge, this is the first attempt to develop criteria for JDM-scleroderma overlap syndrome in the paediatric population. In this survey, the majority of clinicians believed that defining overlap phenotypes (clinical and serological) influences their treatment, suggesting that further research is warranted to more fully understand this group. A limitation of our study is that less than half of the surveyed physicians responded to our initial survey, so our findings may not represent a true consensus. This subtype of JDM is rare: the number of children seen by each contributor varied from 1 to 20 cases. However, our survey targeted the international community and almost 90% of participants had experience in managing this specific group.

Over the last decade, autoantibodies such as myositis-specific autoantibodies and myositis-associated autoantibodies have become important biomarkers in the diagnosis of idiopathic inflammatory myopathies, the identification of overlap features and the monitoring of patients [2, 6]. In our previous report [7], we defined JDM-scleroderma overlap by the presence of two or more of RP, sclerodactyly or sclerodermatous skin changes. Myositis-associated autoantibodies, in particular anti-PM-Scl and anti-U1-RNP, were frequently detected in this group. Notably, these three features were ranked by clinicians as the top three in this survey.

This process is the first step in developing meaningful criteria for the diagnosis of JDM-scleroderma overlap.

The next step would be to validate the criteria in a second cohort, and then to ask experts to submit patient cases that include the features we have identified as well as patients with other connective tissue disorders. The derivation of a proposed definition of scleroderma overlap would then require a consensus-based meeting of international experts to generate a proposal.

Acknowledgements

The authors would like to thank all members of JDCBS and the JDM Network of the Paediatric Rheumatology European Society for their dedication, contribution and support in this project. L.R.W. is supported by Arthritis Research UK and the NIHR Biomedical Research Centre at Great Ormond Street Hospital.

Funding: No specific funding was received from any funding bodies in the public, commercial or not-for-profit sectors to carry out the work described in this manuscript.

Disclosure statement: The authors have declared no conflicts of interest.

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Accepted 8 May 2019

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Rheumatology 2019;58:2064–2065

doi:10.1093/rheumatology/kez247

Advance Access publication 18 June 2019

Assessment of gastric emptying half time as a quantitative measure of gastrointestinal dysmotility in systemic sclerosis

Rheumatology key message

- Gastric emptying half time may be used as a quantitative measure to assess dysmotility in scleroderma.

SIR, Early identification and quantification of motility abnormalities in SSc patients can help in their better management. Gastric scintigraphy is a non-invasive method to assess the motility abnormalities. Delayed gastric emptying time has been demonstrated previously by multiple scintigraphy methods in SSc patients [1, 2]. The consensus recommendations by the American Neurogastroenterology and Motility Society and the Society of Nuclear Medicine, which are used to report gastric emptying, give a qualitative assessment [3]. We assessed the utility of gastric emptying half time (GE-T_{1/2}) as a quantitative measure to assess the

gastric emptying in a prospective, observational, single-centre study. Thirty-three patients with SSc were recruited from a rheumatology clinic. Patients with overlap syndrome, those who had prior abdominal surgeries and those with diabetes mellitus were excluded.

Patients were given a standard meal of 300 kJ calories labelled with technetium (Tc-99m sulphur colloid radio labelled 0.5–1.0 mCi). Anterior and posterior projections imaging was performed at 0, 1, 2 and 4 h. Delayed gastric emptying (gastric retention) was determined to be 90% at 1 h, 60% at 2 h, and 10% gastric retention at 4 h [3]. GE-T_{1/2} was calculated by linear interpolation based on abdominal images at 1, 2, 3 and 4 h after the meal. A minimum of two images were required to calculate the GE-T_{1/2}. A written informed consent was taken from the patients and the study was approved by local ethics committee of the Postgraduate Institute of Medical Education and Research. For normally distributed data, means were compared using *t* test and for skewed data, Mann-Whitney test. All statistical tests were two-sided and *P*-values ≤ 0.05 were considered significant.

The mean age was 36.4 (13.0) years, with a female : male ratio of 10 : 1. The baseline characteristics are given in Table 1. Six patients (18%) had a delayed gastric emptying (GE). Mean age, duration of illness and duration of gastrointestinal symptoms were not different among those with delayed GE. Mean GE-T_{1/2} was 112.4 (24.1) min in those with a delayed gastric emptying and was 75.2 (18.9) min in those with a normal emptying. The mean GE-T_{1/2} correlated with delayed gastric emptying by consensus recommendations (*P* ≤ 0.001). At a cut-off of 99.2 min, the area under the receiver operating characteristic curve for GE T_{1/2} was 0.904 and the sensitivity was 83% and specificity was 96% to detect a delayed gastric emptying as assessed by consensus recommendations.

GE-T_{1/2} has been suggested as a good measure for assessing gastric emptying as it quantifies the entire post-prandial period rather than the meal left at some specific time points [4, 5]. But this has not been attempted in SSc. GE-T_{1/2} gives a discrete value to follow up and compare. It has been shown to be accurate if calculated from the hourly gastric emptying at 1, 2 and 4 h or at all four time points (1, 2, 3 and 4 h). In our study, GE-T_{1/2} had a good correlation with consensus recommendations. The sensitivity and specificity of GE-T_{1/2} to detect a delayed emptying was good and may be considered for assessment of gastric emptying in this population. GE-T_{1/2} is a quantitative tool when compared with consensus recommendations. For a disease like scleroderma, regular assessment of the gastrointestinal system for treatment response or disease progression is needed. GE-T_{1/2} being quantitative may be more helpful than the consensus recommendations. In conclusion, GE-T_{1/2} is an under-utilized variable in gastric emptying scintigraphy and can be considered as a quantitative tool in assessing gastric emptying in SSc along with consensus recommendations.

Funding: No specific funding was received from any funding bodies in the public, commercial or not-for-profit sectors to carry out the work described in this manuscript.