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REVIEW ARTICLE

Clinical practice guidance for juvenile dermatomyositis (JDM) 2018-Update

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ABSTRACT

Juvenile dermatomyositis is the most common type of juvenile idiopathic inflammatory myopathy mainly affecting the skin and proximal muscles. We have published the Japanese version of 'Clinical practice guidance for juvenile dermatomyositis (JDM) 2018 'consisting of a review of articles in the field and evidence-informed consensus-based experts' opinion on the treatment strategy in collaboration with The Pediatric Rheumatology Association of Japan and The Japan College of Rheumatology under the financial support by 'Research on rare and intractable diseases, Health and Labor Sciences Research Grants'. This article is a digest version of the Japanese guidance.

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KEYWORDS
Diagnosis; pathophysiology; management; interstitial lung disease; myositis-specific autoantibodies

1. Introduction

Juvenile idiopathic inflammatory myopathy is an umbrella entity comprising juvenile dermatomyositis (JDM), juvenile polymyositis (JPM), immune-mediated necrotizing myopathy (IMNM), inclusion body myositis (IBM), and connective tissue disease-associated myositis, which develops before the age of 18 [1]. JDM is the most common form of juvenile idiopathic inflammatory myopathies (JIIMs) currently affecting 1.7 per 100,000 children in Japan [2]. Recently, Single Hub and Access point for pediatric Rheumatology in Europe (SHARE) has published consensus-based recommendation for the management of JDM [3]. However, there are several differences in the frequency of complications and drug use between Europe or United States and Japan such as the frequency of anti-melanoma differentiation-association gene 5 (MDA5) antibody-positive rapidly progressive (RP)-interstitial lung disease (ILD) and the available route of methotrexate (MTX) administration, respectively [4–6]. These facts prompted us to develop clinical practice guidance for JDM suitable for patients in Japan. The Scientific Research Group for Pediatric Rheumatic Diseases (SRGPRD) was organized to standardize the diagnosis, evaluation of severity, and management of pediatric rheumatic diseases under the support by the Japanese Ministry of Health, Labor and Welfare. The JDM Research Group of SRGPRD consisting of 13 pediatric rheumatologists, a dermatologist and a pathologist has published the Japanese version of the guidance in collaboration with The Pediatric Rheumatology Association of Japan (PRAJ) and The Japan College of Rheumatology (JCR). Because evidence on the management of JDM is limited, this guidance mainly consists of narrative review of articles in the field and evidence-informed consensus-based experts' opinion on the treatment strategy. Articles were searched for the consensus-based part in MEDLINE and PubMed in 2017. Articles after the publication of the original Japanese version were hand-searched. Each section was reviewed and approved by all the JDM research group member. Final version of the manuscript was confirmed after public comments and approved by the steering committee of both PRAJ and JCR. The present English version summarizes the Japanese version with an update consisting of several recently published articles.

2. Definition

JDM is a symmetrical inflammatory myositis predominantly affecting proximal muscles with characteristic skin lesions [7]. However, some cases lack muscle weakness despite skin lesions characteristic of JDM and this is currently called juvenile clinically amyopathic dermatomyositis (JCADM) [8]. JCADM comprises juvenile amyopathic dermatomyositis

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ORIGINAL ARTICLE

Disease activity, treatment and long-term prognosis of adult juvenile idiopathic arthritis patients compared with rheumatoid arthritis patients

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ABSTRACT

Objective: To evaluate the difference between adult juvenile idiopathic arthritis (JIA, starting at <16 years) and rheumatoid arthritis (RA).

Methods: Data on 128 adult JIA patients were from the National Database of Rheumatic Diseases in Japan (NinJa), 2014, divided into 4 groups by period of disease onset (Group 1: 2000–2013, $n = 32$; Group 2: 1981–1999, $n = 32$; Group 3: 1966–1980, $n = 31$; Group 4: ~1965, $n = 33$). Disease activity, treatment and long-term prognosis of adult JIA patients were compared with RA patients matched for sex- and disease duration in each era.

Results: In Groups 1 and 2, adult JIA patients had significantly lower clinical disease activity indices (CDAI) (Group 1: adult JIA 1.5 [0.4–6.9]-vs-RA 5.3 [2.5–10.3], $p = .001$, Group 2: 2.6 [0.6–9.0]-vs-6.9 [3.5–11.0], $p = .001$, shown as median [quartile range], p-value, respectively), and had higher CDAI remission rates than RA patients (Group 1: 54.8%-vs-28.2%, $p = .002$, Group 2: 51.7%-vs-17.0%, $p < .001$). More adult JIA than RA patients in Group 1 used biologics (62.5%-vs-24.7%, $p < .001$). However, there were no adult JIA-vs-RA differences in joint destruction and physical function in any group.

Conclusions: Adult rheumatologists must recognize that adult JIA patients are different from RA patients even when disease duration is the same.

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KEYWORDS
Transition; adult Juvenile idiopathic arthritis (JIA); rheumatoid arthritis (RA)

Introduction

Juvenile idiopathic arthritis (JIA) is the most common form of arthritis in children. JIA is of unknown aetiology, beginning before the 16th birthday and persisting for at least 6 weeks [1]. Recently, treatment of JIA has been improved by employing biological therapies, as in rheumatoid arthritis (RA). Advances in pediatric medicine have resulted in increased numbers of adult patients who had childhood-onset chronic disease [2]. However, long-term follow-up data in other countries showed that JIA is still ongoing in 34–50% of JIA patients after they reach adulthood [3,4]. Therefore, a seamless transition in medical care from adolescence to adulthood (i.e. transitional care) is important [5,6]. However, adult rheumatologists who take over the care of adult JIA patients commonly have little knowledge of the pathogenesis, treatment and characteristics of JIA [7]. Moreover, it is unclear whether adult JIA patients should be treated similarly to RA patients [8]. To provide appropriate medical care, we should establish evidence-based management strategies for adult JIA.

In the pre-biologics era, only one study comparing the prognosis of JIA and RA has been published [9]. That study, which dealt with disease subtypes and the presence of antibody, indicated that oligoarticular JIA had the best outcome according to radiographic changes, whereas seropositive RA had the worst [9]. However, little evidence is available with regard to the difference in prognosis between adult JIA and RA in the biologics era. Although the long-term prognosis of JIA still requires elucidation, to the best of our knowledge, there are no large databases on adult JIA patients in Japan which could be explored for this purpose. For this reason, here we extracted data on adult JIA patients (defined as onset at <16 years of age) who were registered in the RA database as 'oligoarticular JIA (oligoarthritis) or polyarticular JIA (polyarthritis)'. We then compared their current status and prognosis with RA patients (defined as starting at ≧16 years of age) who had the same disease duration.

The National Database of Rheumatic Diseases in Japan (NinJa) was established in 2002 to reveal trends and problems associated with RA [10]. Nationwide, attending physicians in multiple centres register patients diagnosed with RA in this database, which includes disease activity, drug use, physical function, joint outcome and other data which are collected annually. Adult JIA patients (oligoarthritis or

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Pediatric Rheumatology

RESEARCH ARTICLE Open Access

Tocilizumab modifies clinical and laboratory features of macrophage activation syndrome complicating systemic juvenile idiopathic arthritis

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Abstract

Background: This study aimed to determine the influence of tocilizumab (TCZ) in modifying the clinical and laboratory features of macrophage activation syndrome (MAS) complicating systemic juvenile idiopathic arthritis (s-JIA). Furthermore, we assessed the performance of the 2016 MAS classification criteria for patients with s-JIA-associated MAS while treated with TCZ.

Methods: A panel of 15 pediatric rheumatologists conducted a combination of expert consensus and analysis of real patient data. Clinical and laboratory features of s-JIA-associated MAS in 12 TCZ-treated patients and 18 untreated patients were evaluated. Possible MAS was defined as having characteristic laboratory features but lack of clinical features of MAS, or atypical MAS, or early treatment that prevented full-blown MAS.

Results: Clinically, the TCZ-treated patients with s-JIA-associated MAS were less likely febrile and had significantly lower ferritin, triglyceride, and CRP levels than the untreated patients with s-JIA-associated MAS. Other laboratory features of MAS including lower platelet counts and lower fibrinogen were more pronounced in TCZ-treated patients. The TCZ-treated patients with s-JIA-associated MAS were less likely to be classified as MAS based on the MAS classification criteria (25% vs 83.3%, $p < 0.01$). This is ascribed to the absence of fever or insufficient ferritin elevation, compared with the untreated patients.

Conclusion: TCZ could modify the clinical and laboratory features of s-JIA-associated MAS. When evaluating the s-JIA patients while treated with TCZ, it is not applicable to use MAS classification criteria. Care must be taken to not underdiagnose MAS based on the MAS classification criteria.

Keywords: Macrophage activation syndrome, Systemic juvenile idiopathic arthritis, Tocilizumab, Classification criteria

Background

Macrophage activation syndrome (MAS) is a severe, potentially life-threatening complication of rheumatic diseases, which is clinically characterized by fever, hepatosplenomegaly, lymphadenopathy, profound depression of all three blood cell lines, deranged liver function, intravascular coagulation, and central nervous system dysfunction. The hallmark of MAS is an uncontrolled and dysfunctional immune response with excessive activation and expansion of T lymphocytes and macrophages exhibiting hemophagocytic activity, which leads to overproduction of numerous proinflammatory mediators, thereby eliciting a cytokine storm. MAS is complicated with many rheumatic diseases. However, MAS is most commonly seen in systemic juvenile idiopathic arthritis (s-JIA) and occurs in approximately 10% patients with s-JIA [1]. Furthermore, subclinical or occult MAS may occur in as many as 30–40% patients with active s-JIA [2].

s-JIA is a severe systemic inflammatory disorder of unknown etiology characterized by arthritis and systemic features such as spiking fever, skin rash, generalized

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Pediatric Rheumatology

RESEARCH ARTICLE Open Access

Clinical features and characteristics of uveitis associated with juvenile idiopathic arthritis in Japan: first report of the pediatric rheumatology association of Japan (PRAJ)

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Abstract

Background: Although there are many reports on Juvenile Idiopathic arthritis-associated uveitis (JIA-U) from various countries, especially from Europe and North America, there are few reports from Asia. Our aim was to investigate the epidemiology, characteristics and predictors of JIA-U in Japan.

Methods: Data were retrospectively collected on 726 patients with JIA from medical records as of April 2016 at 15 medical centers specialized in pediatric rheumatic diseases. Of these, patients with uveitis were further investigated for the specific characteristics of this manifestation.

Results: The prevalence of uveitis was 6.1% in the 726 JIA patients examined. Incidence of uveitis was significantly higher in patients with an earlier arthritis onset (2.6-vs.-5.8 years, $P < 0.0001$), oligoarthritis (16.1%-vs.-1.6%, $P < 0.001$), or anti-nuclear antibodies. On the contrary, it was significantly less common in patients with rheumatoid factor or anti-cyclic citrullinated peptide antibodies. A history of using methotrexate (MTX), infliximab or adalimumab was also associated with uveitis occurrence. The median age at uveitis diagnosis was 5 years, and the median time from arthritis onset to uveitis diagnosis was 2 years. The occurrence of anterior and bilateral uveitis was 79.3 and 53.7%, respectively. There were no symptoms at uveitis diagnosis in 58.5% of cases. Complications arising between the time of uveitis diagnosis and the last observation increased from 31.7 to 56.1%; in particular, cataract was increased 3-fold. While no patients lost their vision, 61.9% did not recover normal vision (≥ 1.0), and in many cases active uveitis persisted, especially in males. In addition to steroid eye drops (97.6%) and MTX (15.4%), biological agents were used for treating the uveitis in 41.5% of patients.

Conclusions: The epidemiology, characteristics and predictors of JIA-U in Japan are described here for the first time. Although the prevalence of JIA-U in Japan is lower than in predominantly Caucasian cohorts, as reported from North America and Europe, the epidemiology, characteristics and predictors were found to be similar.

Keywords: Juvenile idiopathic arthritis, Uveitis, Epidemiology, Asian

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REVIEW ARTICLE

Clinical practice guidance for juvenile idiopathic arthritis (JIA) 2018

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ABSTRACT

Juvenile idiopathic arthritis (JIA) is the most common disease in pediatric rheumatism. There is no specific symptom or examination finding for JIA, and the diagnosis is made by exclusion and differentiation. Because non-pediatric rheumatologists are sometimes involved in medical care, 'proposal for JIA guidance on diagnosis and treatment for primary care pediatricians and non-pediatric rheumatologists' was first published in 2007. In these 10 years, a number of new findings on pathophysiology and treatment of JIA have been published; therefore, we propose this guidance of 2018th edition aiming at updating and standardization of JIA medical care in Japan. This edition included the management of uveitis, macrophage activation syndrome, infectious diseases before and during treatment. Moreover, details of biologics are also described. Although this guidance is tailored to adaptation of examinations and drugs, we do not purpose to limit the physicians' discretion in clinical practice. This guidance should be viewed as recommendations and be individualized according to the condition of the patient. We hope that medical care for JIA will advance and more patients will get benefit based on this guidance. Then, further revisions are needed due to changes in future conditions.

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KEYWORDS
Juvenile idiopathic arthritis; clinical practice guidance; algorism of treatment; macrophage activation syndrome; uveitis; biologics

1. General considerations and classification of pediatric patients with chronic arthritis

Juvenile idiopathic arthritis (JIA) is defined as chronic arthritis of unknown etiology beginning before the 16th birthday and persisting for at least 6 weeks when other known conditions are excluded.

The current classification of JIA was proposed by the Pediatric Standing Committee of the International League of Associations for Rheumatology (ILAR) [1], which published an initial revision in 1997 [2] and subsequently a second revision in 2001 [3]. This classification includes seven categories of JIA (Table 1) which mainly fall into two types according to the differences in clinical symptoms and pathophysiology, namely systemic arthritis (systemic JIA) and the other six JIA categories. The latter consist of oligoarthritis, rheumatoid factor-negative polyarthritis, rheumatoid factor-positive polyarthritis, psoriatic arthritis, enthesitis-related arthritis, and undifferentiated arthritis, and are often considered as 'articular-type JIA' in clinical practice in Japan. We therefore use this term in this guide. Within systemic arthritis, we can clearly differentiate a form where only arthritis remains after systemic inflammation subsides (fever, eruption, hepatosplenomegaly, serositis, etc.) from articular-type JIA. Here, we will use the term 'systemic arthritis with active arthritis (and without active systemic features)' in the present guide, according to the '2011 American College of Rheumatology Recommendations for the Treatment of Juvenile Idiopathic Arthritis' [4].

Recently, the term 'spondyloarthritis (SpA)' has been widely used in children. The main manifestations of this disease are axial arthritis (such as spondylitis and sacroiliitis), peripheral arthritis, and enthesitis of tendons and ligaments [5]. This is an umbrella disease which includes ankylosing spondylitis [6], psoriatic arthritis [7], arthritis of inflammatory bowel disease and reactive arthritis. Enthesitis-related arthritis, psoriatic arthritis, children with some undifferentiated arthritis in JIA categories are equivalent to SpA [5]. Because the categories which are excluded from the JIA classification (e.g. arthritis of inflammatory bowel disease) could be also diagnosed using SpA criteria,

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BRIEF REPORT

Validation of Classification Criteria of Macrophage Activation Syndrome in Japanese Patients With Systemic Juvenile Idiopathic Arthritis

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Objective. **To validate whether the 2016 American College of Rheumatology/European League Against Rheumatism classification criteria of macrophage activation syndrome (MAS) complicating systemic juvenile idiopathic arthritis (JIA) is practical in the real world.**

Methods. **A combination of expert consensus and analysis of real patient data was conducted by a panel of 15 pediatric rheumatologists. A total of 65 profiles comprised 18 patients with systemic JIA–associated MAS and 47 patients with active systemic JIA without evidence of MAS. From these profiles, 10 patient data points for full-blown MAS, 11 patient data points for MAS onset, and 47 patient data points for acute systemic JIA without MAS were evaluated.**

Results. **Evaluation of the classification criteria to discriminate full-blown MAS from acute systemic JIA without MAS showed a sensitivity of 1.000 and specificity of 1.000 at the time of full-blown MAS. Sensitivity was 0.636 and specificity was 1.000 at the time of MAS onset. The number of measurement items that fulfilled the criteria increased in full-blown MAS compared to that at MAS onset. At MAS onset, the positive rates of patients who met the criteria for platelet counts and triglycerides were low, whereas those for aspartate aminotransferase were relatively high. At full-blown MAS, the number of patients who met the criteria for each measurement item increased.**

Conclusion. **The classification criteria for MAS complicating systemic JIA had a very high diagnostic performance. However, the diagnostic sensitivity for MAS onset was relatively low. For the early diagnosis of MAS in systemic JIA, the dynamics of laboratory values during the course of MAS should be further investigated.**

Introduction

Macrophage activation syndrome (MAS) is a severe complication of systemic juvenile idiopathic arthritis (JIA), which is clinically characterized by fever, hepatosplenomegaly, lymphadenopathy, depression of all 3 blood cell lines, deranged liver function, intravascular coagulation, and central nervous system dysfunction (1). MAS is a potentially life-threatening disease, and thus, a timely and prompt diagnosis is essential to initiate life-saving treatment. However, it can be difficult to distinguish MAS from systemic JIA flares, sepsis, or other secondary hemophagocytic lymphohistiocytosis. Differentiation of MAS from these conditions is essential for the selection of an appropriate therapeutic intervention in a timely

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Review Article

Proposal for the development of biologics in pediatric rheumatology

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Abstract In order to assess the development, approval and early introduction into clinical practice of biologics in the pediatric field, we herein describe the current status of the development to approval of biologics as anti-rheumatic agents for children in Japan, discuss the present problems and provide a proposal for the future. It has become apparent that the duration of the review period required for the preparation of clinical trials and Pharmaceuticals and Medical Devices Agency approval is clearly reduced compared with the past. Thus, it was speculated that a rate-limiting step in the process from development to approval was the duration of clinical trials from start to end. Hence, we focused on the following key words with regard to promotion of the development of biologics and their early practical use: "registry", "centralization", and "global cooperation", all of which are related to the reduction of duration of a clinical trial. In conclusion, to reduce the duration of a clinical trial, it is essential to complete a world-scale registry system by developing the registry system established by the Pediatric Rheumatology Association of Japan. The next step is then to carefully plan to participate in the international network using the world-scale registry system, and develop global cooperative trials in which we can ensure a sufficient number of entries from Japan.

Key words biologic, centralization, international cooperation, pediatric rheumatology, registry.

Rheumatic diseases in childhood are regarded as incurable even now, and their pathogenesis has not been clarified. These are serious diseases that fulfill the following four conditions: (i) the pathogenesis has not been clarified; (ii) the therapeutic method has not been established; (iii) they are rare; and (iv) they need long-term medical treatment. Pediatric rheumatic diseases are systemic inflammatory diseases involving antoinflammation and autoimmunization, and their medication and treatment have dramatically advanced due to marked progress in diagnostic technology in inflammatory science and rheumatology. Therefore, favorable outcomes in the inflammatory state are expected without carrying over organ failure to adulthood if the principles of early diagnosis and early therapeutic intervention are maintained, and it is not too much to say that the advent of biologics facilitated this improvement. In the pediatric rheumatology field, four biologics (tocilizumab, etanercept, adalimumab and palivizumab) were approved in Japan by June 2017. In particular, new approval for pediatric indications for tocilizumab, etanercept and adalimumab – anti-rheumatic agents – has greatly changed medical treatment in the pediatric rheumatology field, by which many clinicians have realized that treatment has changed from care to cure.[1]

In this article, to assess the development, approval and early introduction into clinical practice of biologics in the pediatric field, we herein describe the current status of the development to approval of biologics as anti-rheumatic agents for children in Japan, discuss the present problems and provide a proposal for the future.

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Positioning of biologics as anti-rheumatic agents for children in Japan

In the early first decade of the 2000s, the major treatment for systemic-onset juvenile idiopathic arthritis (JIA) was steroid, and that for polyarticular JIA was methotrexate.[2] It was recognized, however, that 30–40% of pediatric patients did not respond to these treatments, nor did their symptoms easily resolve.[1]

Recently, indications for the use of biologics have been described according to guidelines for initial treatment[3,4] and guidelines for the use of biological preparations.[5–7] Namely, for systemic JIA, tocilizumab, an anti-interleukin (IL)-6

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ORIGINAL ARTICLE

Survey of the awareness of adult rheumatologists regarding transitional care for patients with juvenile idiopathic arthritis in Japan

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ABSTRACT

Objectives: To understand the current status of adult rheumatology care for patients who had previously had juvenile idiopathic arthritis (JIA) (excluding systemic JIA), and to identify issues interfering with the transition from pediatric to adult care in Japan.

Methods: Questionnaire-based survey among 30 adult rheumatologists.

Results: Eighty-seven percent of adult rheumatologists responded that they had provided medical care to adults who had had JIA; 44% of them had felt hesitation or anxiety when providing such care. The reasons for this included lack of independence of the patients, lack of knowledge and experience among adult rheumatologists, and lack of preparation for accepting such patients. Many adult rheumatologists believed that the timing of transition from pediatric to adult rheumatology care must be considered based on therapeutic regimens or clinical conditions/disease states, not solely chronological age. A majority of adult rheumatologists showed great interest in transitional care for JIA patients and desired to communicate better with pediatric rheumatologists.

Conclusion: Transitional care for JIA patients is not sufficiently developed in Japan. Education and advocate campaign of transitional care is required for adult rheumatologists as well as patients and their parents.

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KEYWORDS
Juvenile idiopathic arthritis; transitional care; rheumatologist; questionnaire

Introduction

Progress in pediatric medical care has made it possible for many children suffering from intractable diseases to survive into adulthood [1,2]. At the same time, the number of children is increasing who have grown up to adulthood with a primary disease and its complications [3]. The transition from pediatric to adult health care systems has recently received significant attention worldwide [3–5]; however, Japan lags behind the United States and Europe.

Medical care for juvenile idiopathic arthritis (JIA) has also made significant progress with the increased use of MTX and biological anti-inflammatory agents. This has helped to delay or inhibit joint destruction in childhood and to improve their activity of daily life. Nonetheless, treatments must be continued for the majority of JIA patients even after transition to adult rheumatology care [6]. Although transitional care program for JIA have been developed and operated in other countries, it has rarely been applied in Japan [7]. In a recent survey among Japanese non-pediatric rheumatologists, 'inadequacy of adult rheumatology care' and 'lack of independence from parents' were extracted as the main factors to prevent smooth transitions of patients with pediatric rheumatic diseases [8]. That survey revealed the current status and issues regarding adult rheumatology care for pediatric rheumatic diseases in general, but not regarding specialized for JIA.

Therefore, we conducted this survey to understand the current status of adult rheumatology care for patients who had previously had JIA (excluding systemic JIA), and to identify issues interfering with the transition from pediatric to adult care in Japan.

Subjects and methods

A questionnaire-based survey was conducted anonymously among 30 adult rheumatologists working in 23 hospitals across Japan. All these hospitals had contributed to a nationwide Japanese database of rheumatic diseases (*NinJa*: National Database of Rheumatic diseases in Japan) [9]. The questionnaire asked adult rheumatologists for the

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Early use of alendronate as a protective factor against the development of glucocorticoid-induced bone loss in childhood-onset rheumatic diseases: a cross-sectional study

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Abstract

Background: Bisphosphonates are recommended for use as first-line therapy for the prevention and treatment of glucocorticoid-induced osteoporosis in adults. However, the appropriate usage of bisphosphonates for the prevention or treatment of glucocorticoid-induced osteoporosis in children remains unclear.

Methods: We performed a cross-sectional study to clarify the factors associated with the development of glucocorticoid-induced bone loss and osteoporosis in patients with childhood-onset rheumatic disease and to investigate the impact of the early use of alendronate. We recruited 39 patients with childhood-onset rheumatic disease who were evaluated to detect bone loss or osteoporosis at 3 months to 1.5 years after the initiation of treatment. The primary outcome of the study was the presence of bone loss or osteoporosis at the initial evaluation of the bone mineral density after at least 3 months of glucocorticoid therapy.

Results: Bone loss and a history of fracture were found in 56 and 18% of the participants, respectively. Weekly oral alendronate therapy (median, 25.4 mg/m^2) had been started by the time of the evaluation of osteoporosis in 46% of the participants and within 3 months after the start of glucocorticoid in 31% of the participants. There were no significant differences between the participants with bone loss (wBL group) and without bone loss (w/oBL group) in terms of gender, primary disease, or the age at the onset of primary disease. In terms of glucocorticoid use, there was no significant difference in the age at the start of glucocorticoid therapy, the length of glucocorticoid use, or the dose of glucocorticoids. The proportion of patients in the w/oBL group who received alendronate within 3 months after the start of glucocorticoid therapy was significantly greater than that in the wBL group. In the logistic regression analysis, only "alendronate therapy within 3 months after the start of glucocorticoid therapy" had a statistically significant effect on the development of bone loss (OR, 0.08; 95% CI, 0.02–0.43). The analysis did not reveal any factors associated with the development of osteoporosis.

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Survey of attitudes of non-pediatric rheumatologists among councilors of the Japan College of Rheumatology regarding transitional care

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ABSTRACT

Objectives: The transition from pediatric to adult healthcare systems has recently received worldwide attention. Surveys of the attitudes of Japanese non-pediatric rheumatologists regarding transitional care were conducted.

Methods: Non-pediatric rheumatologists among councilors of the Japan College of Rheumatology were enrolled in the surveys. Experiences of adult patients with childhood-onset rheumatic diseases, ideal medical care for these patients, and factors that made the transition to adult care difficult were examined via e-mail.

Results: Overall, 201 non-pediatric rheumatologists (21.2%) responded to the surveys. Ninety-one percent had previous experience with patients with childhood-onset rheumatic disorders. Transition to non-pediatric institutes was supported by about 90% of respondents. However, only 32% of non-pediatric rheumatologists had no hesitation about caring for adults with childhood-onset rheumatology disorders. Two main factors prevented smooth transitions to non-pediatric care: inadequacy of non-pediatric care (57%) and lack of independence from parents/family (53%). The majority of non-pediatric rheumatologists hesitated about medical care for patients with autoinflammatory syndromes, whereas they became familiar with articular juvenile idiopathic arthritis without hesitation (86.6%); 93% of respondents requested more opportunities to learn about pediatric rheumatology disorders.

Conclusions: Sharing additional knowledge about pediatric rheumatology within the non-pediatric rheumatology field is required.

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KEYWORDS
Attitude survey; autoinflammatory syndrome; childhood-onset rheumatic disease; pediatrics; rheumatology; transition

Introduction

Advances in medicine have dramatically improved the prognosis of childhood-onset chronic diseases, allowing patients to survive and reach adulthood [1,2]. In Japan, 1000 patients with childhood-onset chronic disease reach adulthood every year, and many of them survive without serious sequelae or disabilities [3]. The transition from pediatric to adult healthcare systems has recently received worldwide attention, and transitional care, as defined by the Society for Adolescent Medicine, is 'the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented healthcare systems' [4].

Pediatric patients with rheumatic disorders also frequently survive into adulthood [5–7]. Among pediatric rheumatic disorder, juvenile idiopathic arthritis (JIA) is the most common in Japan and develops in about one out of 100,000 children per year in Japan, with a prevalence of 9.74 out of 100,000 children less than 16 years of age according to a nationwide survey [8,9]. Some long-term studies of the outcomes of childhood-onset chronic rheumatic diseases have been conducted; however, most reports are retrospective, cross-sectional, or selective. Thus far, very few reports that followed patients beyond adolescence have been published, and studies of adult outcomes in other childhood-onset rheumatic diseases have shown high rates of active disease [10]. Moreover, most reports of transition were authored by pediatric health professionals.

Non-pediatric rheumatologists are expected to be primarily responsible for the transitional care of patients with childhood-onset rheumatic disorders. Prior to the establishment of supportive guidelines for the transition of patients to adult medical care in Japan, surveys of the attitudes of non-pediatric rheumatologists regarding transitional care were conducted.

Methods

Non-pediatric rheumatologists among councilors in the Japan College of Rheumatology were enrolled in the surveys. Experiences of adult patients with childhood-onset rheumatic diseases, ideal medical care for these patients, and factors that made the transition to adult care difficult were examined via e-mail. Regarding issues preventing a smooth

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