

Review Article

Physiotherapy in Juvenile Dermatomyositis

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Abstract

Juvenile dermatomyositis (JDM) is a relatively rare, multi-systemic autoimmune disease, which combines inflammatory myopathy, characteristic cutaneous findings and symptoms of other organ systems. In JDM the basic characteristic is the skin and muscle vasculitis, although the first sign of JDM is usually a skin rash in a V-shaped pattern that may be red and patchy, with typically red or purplish color on the eyelids (“heliotrope” rash), cheeks, or both. The medications used for the relief of symptoms of dermatomyositis are corticosteroids, cyclosporine, intravenous immunoglobulin, cyclophosphamide, methotrexate, and biological agents. Evidence shows that children with low-activity dermatomyositis should be able to safely perform aerobic exercise and resistance-based exercise regimes to reduce disease activity and improve circulation in the resting phase, but also during exercise. Until now, both pharmaceutical therapy and physiotherapy have been proven the best way to treat the disease so that patients achieve complete remission without disease-related complications or any use of steroids.

Keywords: Exercise, Children, Juvenile dermatomyositis, treatment, Physiotherapy

Introduction

There are some rare diseases such as idiopathic inflammatory myopathies (IIM) that primarily affect skeletal muscles. In grown-up people, IIMs are separated in polymyositis (PM), dermatomyositis (DM) and inclusion body myositis (IBM)¹. Dermatomyositis is an inflammatory disease of the skin and muscles that progresses rapidly. Juvenile dermatomyositis (JDM) is a regular, non-infectious, inflammatory malady, beginning before the age of 16 and principally impact on the skin and the muscles².

Etiology

No infectious agent been clearly implicated to date with JDM, until now the cause of the illness remains unbeknown, while we have not fully understood the role of genetic factors^{3,4}.

Epidemiology

JDM is a rare autoimmune disease that affects all nations, with a prevalence of 2.5-4.1 children per million in the United States⁵. The average age of onset is approximately 7 years and commonly leads a chronic course^{6,8}. JDM accounts for more than 80% of the idiopathic inflammatory myopathy childhood cases, thus making it the most common idiopathic inflammatory myopathy among children with a 5:1 ratio of female to male^{5,9}.

Pathogenesis

The pathogenesis of JDM involves several infectious agents (coxsackie virus, Toxoplasma spp, echovirus, parvovirus) and environmental triggers (sun exposure, medications, and vaccines). A suggested possibility for the immune response is a molecular imitation⁷. In JDM patients, peripheral blood cells have been shown to lyse autologous target cells presenting a binding peptide derived from skeletal myosin. It has also been found that the amino acid sequence of Protein M5 of hemolytic A β group streptococci is homologous to the human skeletal heavy chain. So we can understand that the muscular fiber disaster can participate to these cytotoxic T lymphocytes and that the myosin heavy chain epitope may be a tee autoantigen in JDM.

JDM basic characteristic is the skin and muscle vasculitis. There are several reports of various human

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Symptom	Number of patients					
	Diagnosis	1 yr	2 yr	3 yr	4 yr	5 yr
Rash	30	20	3	3	0	1
Gottron's papules	22	12	3	2	1	0
Weakness	28	10	4	1	0	0
Myalgia	30	13	3	1	0	0
Systemic features	31	18	9	5	1	0
Palatal weakness	12	3	1	0	0	0
Ulceration	8	0	0	0	0	0
Lipoatrophy	0	1	0	1	0	0
Calcinosis	1	2	2	2	2	2

Table 1. Number of patients with symptoms over a 5 yr period from a group of 36 patients followed prospectively from diagnosis. (Adopted from McCann LJ et al. 2006)⁶.

leukocyte antigens (HLA), with a polymorphism of the tumor necrosis factor (TNF)- α gene (TNF- α -308) and an increased expression from genes of type-1 interferon- in most of the patients. What shows us the activity of the JDM disease at the beginning and after three years is interferon in peripheral blood of patients JDM as well as overexpression of the adhesion molecules ICAM-1 and VCAM-1 that has been described on endothelial cells, inflammatory cells and keratinocytes².

Clinical presentation

In JDM we can identify over the medial malleolus, the knee and the metacarpophalangeal and interphalangeal joints to develop some typical dermal far-reaching amendments consisting of heliotrope dermatitis and Gottron's papule¹⁰. Muscle biopsy, electromyography (EMG) demonstrating denervation and myopathy and the increase of serum level of one of the "muscle" enzymes are some of the criteria used for the diagnosis¹⁰. In most cases, the first sign of JDM is a skin rash in a V-shaped pattern that may be red and patchy, with typically red or purplish color on the eyelids ("heliotrope" rash), cheeks, or both⁷. The criteria they use Bohan and Peter say that a patient can be diagnosed with JDM if they present all three plus the rash, or a alternative case if an ailing meets two of the tests and the rash¹⁰. Thus, it is clear that persistent rash is a problem for a large number of children with JDM¹¹. A malar rash may involve the nasolabial fold but characteristics are different from Systemic lupus erythematosus (SLE), which makes the differential diagnosis from SLE or scleroderma an absolute necessity.

The clinical presentation of JDM may also involve the following symptoms: alopecia, skin and oral ulcers, gingivitis, Raynaud's phenomenon, limb edema, livedo reticularis, skin nodules, poikiloderma, subcutaneous calcinosis, arthralgia, arthritis involving major joints, asthenia, dysphonia,

osteopenia as a result of dysphagia or anorexia that often cause major weight loss². Symptoms from other organs' involvement (with incidence as shown in Table 1) that may contribute to morbidity and mortality is hepatomegaly, lymphadenopathy, fever, esophagitis, gastritis, bowel disease and colitis with a risk of perforation in a few cases, pancreatitis, anasarca, contractures, interstitial lung disease, cardiac involvement, renal disease, or seizures¹¹.

In JDM we can often find symmetrical weakness of the proximal muscles¹⁰. Muscle weakness often, but not always, occurs at the same time or after the rash. Patients usually experience weakness in walking or climbing stairs, inability to get up from a sitting position, inability to raise their hands above the shoulders and inability to lift objects. Early physical examination includes Gower's and Trendelenburg's test as muscle weakness affects all the muscle groups but mostly the trunk muscles, the anterior neck flexors and the limb-girdle musculature⁷.

Illness operation and impairment assessment

Laboratory tests

In JDM, C-reactive protein (CRP) and erythrocyte sedimentation rate test (ESR) levels may in some cases be elevated. Serum creatine kinase (CK), galacto-dehydrogenase, aldolase, serum glutamate oxalate transaminase (SGOT) and glutamate pyruvate serum transaminase (SGPT) in the laboratory tests appear to be elevated, but this is not an indication of disease activity. Measurements of insulin and glucose levels are needed in order to detect the risk of other metabolic disorders and of lipodystrophy. Anti-PM-Scl, anti-U-RNP, anti-SSa, and anti-p155 antibodies are found in subsets of patients with either typical JDM or overlapping connective tissue diseases¹².

Phase	I	II	III
Aim	<ul style="list-style-type: none"> Familiarizing the patient with the treadmill Improving aerobic exercise capacity Developing proper strength training techniques 	<ul style="list-style-type: none"> Improving aerobic exercise capacity Improving muscle strength 	<ul style="list-style-type: none"> Improving aerobic exercise as in phase 2 albeit with a higher intensity Improving muscle strength as in phase 2
Frequency [week- 1]	3	3	2
Time/session [minutes]	40-60	45-60	50-60
Type	Light interval training on treadmill and strength training	Moderate interval training on treadmill and strength training	Vigorous interval training on treadmill and strength training
Interval training on treadmill			
Intensity [HR as % of HRpeak]	65-70	70-80	80-90
Interval duration [minutes]	3	2-2.5	1-2
Number of intervals	4-7	6-10	10-12
Strength training			
Number of exercises	3	3	3
Number of sets/exercise	3	3	3
Number of repetitions or time/set	Week 1: 3 repetitions Week 2-12: as much repetitions in 20 or 30 seconds.	As much repetitions in 20 or 30 seconds	As much repetitions in 20 or 30 seconds

Table 2. Description of the exercise training program. (Adopted from Habers EA et al. 2012)¹⁸.

Other procedures

Some of the examinations performed to evaluate the patients' condition are: Electromyography, Quantitative nail fold capillaroscopy which is an approved noninvasive method to evaluate small vessel vasculitis and, to some extent, assess JDM activity, Magnetic resonance imaging (MRI) of the lower limb, Ultrasound and Power Doppler ultrasonography that is a technique that can give a lot in the evaluation of muscle hyperemia and vascularization. Muscle histopathology is what will help us with differential diagnosis, as well as the prognosis of JDM's disease². Esophageal manometry, lung function tests, EKG, and cardiac ultrasound contribute interesting information to evaluate a multisystemic disease. Osteodensitometry is useful to assess the effect of the disease on bone mineralization and body mass composition¹³.

Muscle strength and functional assessment

Muscle strength is prescribed as the maximum ability of a muscle or muscle group to exert power under a given set of conditions, and we use it to evaluate muscle disease. We can measure muscle strength with Manual muscle testing (MMT). It evaluates strength on an ordinal 6-point, 0-5, scale or a 12-point, 0-10, scale². Muscle strength differs in children, depending on age and gender, that's why we must be very careful when using the MMT test in children with dermatomyositis and when using resistance. In addition, the evaluation of muscle strength, muscle fatigue and the

evaluation of daily activities will give us a comprehensive picture of the damage the disease has caused. The tools for the capability in achieving daily movements are the childhood health assessment questionnaire (CHAQ) and for the muscle function is the Childhood Myositis Assessment Scale (CMAS)^{14,15}. The evaluation of disease activity regarding the muscular system and various other organ systems of the body, such as constitutional, articular, cardiac, pulmonary, gastrointestinal, cutaneous systems is accomplished with the JDM disease activity score (DAS)¹⁶.

Treatment

Medication

Drug therapy of JDM is based primarily on clinical experience and studies. The medication used for the relief of symptoms of dermatomyositis are corticosteroids, cyclosporine, intravenous immunoglobulin, cyclophosphamide, methotrexate, and biological agents¹⁷. In typical cases of dermatomyositis, at the onset of the disease, lofty -dose prednisone (2 mg/kg per day) in 4 months is administered in order to rapidly control the disease but it can cause serious side effects such as cataract, osteoporosis, symptoms and typical appearance as in the Cushing syndrome, increase in blood pressure or delayed growth. Into patients recently diagnosed with JDM, methotrexate is another option. In a comparative study of 31 patients, the team cured by prednisone (2 mg/kg

per day) and methotrexate (15 mg/m² per week) stopped much more rapidly prednisolone and with an average lower cumulative dose than those control groups given only prednisolone. Both treatment groups had good results in terms of recovery of strength and physical function but the group receiving the combination therapy had fewer side effects, small weight gain, best tallness growth, and reduced danger of evolving cataracts contrasted by controls. For this reason methotrexate is utilized from physicians as a steroid-sparing agent⁷. When corticosteroids and methotrexate can not stop the progression of the illness or the child's case is severe, some physicians use intravenous immunoglobulin. Cyclosporine or biological agents, such cyclophosphamide and the anti-CD20 antibody rituximab, can also be used in chronic cases⁷.

Physiotherapy

Children with chronic inflammatory diseases such as dermatomyositis have decreased physical activity due to the symptoms of the disease. The very disease causes drowsiness, which in turn further reduces the patients' physical activity, resulting in the risk of developing complications such as obesity, diabetes or cardiovascular disease. In chronic inflammatory disease the use of exercise training is increased as a non-pharmacological interference. The goals of physiotherapy in JDM in the inflammatory phase are prevention of joint contractions and strengthening of the respiratory system whereas in the phase of revitalization the goals are to strengthen muscle fibers suffering and to treat myopathy caused by corticosteroids. Physical activity is safe and a significant component in medication for those receiving medical treatment for JDM and probably effective in improving muscle damage. Kinesiotherapy is often added to the treatment program to help patients make progress with their long-term motor skills, total strength, aerobic exercise capacity, and muscle function¹⁸.

Aerobic exercise capacity

Exercise in children is considered to be more enjoyable and more effective when it is not continuous but with breaks. The exercise program for patients with JDM should be such that during the course of the program the breakdown frequency increases while the duration of each exercise decreases. The training program usually takes place in the treadmill as walking is a basic function that can affect the daily routine of patients with JDM¹⁸.

Strength training

To achieve an improvement in the strength of muscle groups most affected by dermatomyositis and large muscle groups we should design a program regime that includes the following exercises: push-ups, ventral crunch, and squat. To avert muscle malfunction, the exercises are against gravity but without additional weights, and a ball is often used as a toy to make the exercises more enjoyable and pleasantly

to children. Exercises for power recovery and exercise in the treadmill must be alternated during each session¹⁸.

Frequency

The frequency with which patients perform the exercises in the program as shown in Table 2 depends on the phase in which they were. Thus, the frequency of sessions in phase 1 and 2 is 3 times per week, in phase 3 is 2 times per week and duration of the session which consists of preheating, exercise on the treadmill, exercises to increase strength and rest periods are 40-60 minutes¹⁸.

Also, in two open trials with patients with JDM evaluated two exercise programs, the first containing 5 minutes walking on a treadmill followed twenty minutes of resistance training at 8-12 VRM, in three sets compounded with aerobic treadmill exercise at 70% of peak oxygen consumption (VO₂peak) for 30 min, straining, the exercise was initiated with two sets of 15-20 VRM and the second programme containing a schedule with aerobic home gymnastics which assessed with stable cycling at 65% of VO₂max every other day for 12 weeks¹⁹. Children with low-activity dermatomyositis have been shown to be able to safely apply aerobic gymnastics and impedance to restrict illness potency and better circulation during the resting phase, as well as, in the process of gymnastics¹⁹.

Family impact

Studies have shown that caring for children with chronic illnesses such as JDM affects parents physically and emotionally, which may eventually lead to increased levels of anxiety and generally lower quality of life. Especially mothers of children with JDM have statistically significantly higher levels of anxiety and depression compared to a standard reference group (p <0.001), and this fact may in turn affect the child in terms of monitoring and physiotherapies¹⁷.

Prognosis and outcome

The mortality of children with dermatomyositis today is 2% due to significant intervention from medical care and physiotherapy. Once medical treatment has started and muscle pain restrained, physical therapy assists recuperating muscle strength, initially through isometric exercises in critically affected patients, then through more challenging exercises².

Factors associated with a bad vital or functional prognosis are: belated start of rapid treatment, critically skin disease and vasculitis with ulcerative lesions, visceral involvement, in particular digestive tract, and lung involvement. Calcinosis, which is typically found in juvenile dermatomyositis is difficult to treat, however clinical experience has shown that daily massage can soften and prevent those injuries. Osteoporosis is present before corticosteroid treatment in one third of the patients and vertebral collapse is not uncommon. Osteodensity assessment, ideally together with body composition assessment (which may reveal excessive

fat mass), is important to follow these patients. JDM patients therefore need regular physical activity, with the help of physiotherapists, sufficient calcium and vitamin D intake, whereas the place of bisphosphonates and other treatment affecting bone remodeling needs appropriate assessment².

Conclusion

Juvenile dermatomyositis is a chronic poly-systemic disease that concerns patients at a very early age, so its treatment should be separated from adult dermatomyositis. Early diagnosis and start of the treatment is of utmost importance. Until now, both pharmaceutical therapy and physiotherapy are the best ways to treat the disease so that patients achieve complete remission without disease-related complications or steroids.

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