Accordant
A CVS Caremark Company

Clinical Practice Guidelines

Dermatomyositis Polymyositis

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Approved by:
Andrew Krueger, MD
Mark B. Bromberg, MD, PhD
Donald Sanders, MD

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Dermatomyositis/Polymyositis

Table of Contents

INTRODUCTION

The AccordantCare™ program works with health plans to assess, monitor, and support those with certain complex, chronic conditions. The focus of the program is to improve health outcomes and prevent or limit disease-related complications. AccordantCare offers unique services at no additional charge to the patient, putting them in a strong position to adhere to their treatment plan.

There are several ways AccordantCare augments physicians' efforts. Through regular telephone contact, AccordantCare nurses:

- Keep patients informed about the disease process
- Coach patients in self-motivation and self-care skills
- Encourage patients to alert their physician when new symptoms arise
- Direct patients to resources that help pay for medication, transportation, home modifications, etc.
- Ensure preventive and screening measures are accomplished
- Provide emotional support to patients and caregivers
- Screen for depression
- Find local support groups

We invite physicians to make use of the services offered by AccordantCare and to suggest ways we can further patients' treatment goals. To offer feedback, get more information, ask questions, or voice concerns, call toll-free 1-800-948-2497 to speak with a program representative from 8 a.m. to 9 p.m., Monday through Thursday, and from 8 a.m. to 5 p.m. on Friday, Eastern Time. Messages left after hours will be returned the next business day.

Intent of Guidelines

The purpose of this Clinical Practice Guideline is to describe current patterns of practice where there is no fully established national guideline for diagnosis and management. It is not meant to dictate care of patients. Decisions about care are made by the physician and the patient based on the individual needs of that patient.

A patient's health plan may or may not pay for the all medicines, tests, equipment, or services mentioned in this document. Benefits should be checked with the individual's health plan to assure payment.

DISEASE OVERVIEW

Myositis (chronic inflammation of muscle tissue) is a term used to describe a group of acquired and potentially treatable muscle diseases. This group of diseases is also known as the myositis syndromes and as the idiopathic (because their exact cause is not known) inflammatory myopathies.

Because of the chronic inflammation of muscle fibers, the dominant clinical characteristics of the myositis diseases are muscle weakness and fatigue.[1] Myositis symmetrically attacks a person's skeletal muscles,[2] most commonly those closest to the trunk of the body.[1]

In addition, the inflammation can also affect the blood vessels that run through muscles.[1] Myositis frequently is a systemic disease.[3] The systemic complications of myositis can involve an individual's skin, bones, and joints.[3] In addition, myositis can affect the pulmonary, gastrointestinal, and cardiac systems.[3]

Incidence/Prevalence/Demographics of disease[4]

Dermatomyositis

The incidence of dermatomyositis is estimated to be 1 case per 100,000 of population. The prevalence of dermatomyositis is estimated at 1 to 10 cases per million in adults and 1 to 3.2 cases per million in children. Dermatomyositis is more common in females than males by a 2 to 1 ratio. The onset of disease has two peaks, between the ages of 5 and 14 years in children and an average adult age at diagnosis of 40.

Polymyositis

The exact incidence of polymyositis is unknown. Polymyositis is more common in females than males. It occurs mostly in adults over age 20, and it is very rare in children.

Types of the disease

The idiopathic inflammatory myopathies have been classified variously into clinical groupings or subtypes.[5] The following is one meaningful grouping:

- Polymyositis—develops gradually and rarely affects people younger than 18 years of age[4] (see below for details)
- Dermatomyositis—inflammation affects the patient's skin as well as muscles (see below for details)[4]
- Connective tissue myositis—also called overlap syndromes, in which myositis occurs in patients who have connective tissue disease[6]
- Cancer-associated myositis—individual has cancer within two years of myositis diagnosis[3]
- Juvenile myositis—diagnosis occurs before the person is 18 years of age[3]
- Inclusion body myositis (IBM)—disease progresses very slowly, usually beginning in people age 50 and older.[4] Further, it affects distal forearm muscles (grip) and proximal leg muscles in an asymmetric distribution. **Accordant does *not* enroll members with IBM.** However, patients with IBM occasionally get into the program. Some patients are initially diagnosed with polymyositis but actually have IBM, as it is more common than other types of inflammatory myositis.

DIAGNOSIS OF DISEASE

The diagnosis of myositis depends on a combination of clinical presentation, lab studies, and pathological findings.

1. Muscle Enzyme Tests[7,8]
   - serum creatine kinase (CK or CPK)
   - LFTs (liver function tests)
   - AST (aspartate aminotransferase)
   - ALT (alanine aminotransferase)
   - lactate dehydrogenase
   - aldolase
2. Autoantibodies[8]
   - anti-Jo-1 antibodies
   - antinuclear antibodies (should cause further consideration of connective tissue disease such as systemic lupus erythematosus)
3. Electromyography[7]
4. Muscle Biopsy[7]

Dermatomyositis
A patient with dermatomyositis shows a pattern of symmetrical proximal limb weakness—although this pattern is not distinctive for dermatomyositis.[8] Typical skin findings include[4]:

- Gottron's papules
- Heliotrope rash on upper eyelids
- Erythematous rash on face
- Rash on back and shoulders (shawl sign), neck, chest, knees, and elbows
- Telangiectasia
- Mechanic's hands: palmar and lateral aspects of fingers rough with "dirty" horizontal lines

In addition, for a diagnosis of dermatomyositis, the muscle biopsy should indicate

- the presence of small atrophic muscle fibers at the edges of the muscle bundles that are darker or bluish in hematoxylin- and eosin-stained sections located (perifascicular atrophy),[8] is diagnostic of dermatomyositis even in the absence of inflammation.[2]
- the absence of multiple muscle fibers surrounded by inflammatory cells.[8]
- capillary abnormalities showing deposits from the membrane attack complex.[9]

Polymyositis[8]

Patients with polymyositis show progressive symmetrical muscle weakness without skin rash. The muscle biopsy should include:

- prominent inflammatory cells surrounding many of the muscle fibers.
- degeneration of otherwise normal appearing muscle fibers with invasion by macrophages.
- absence of perifascicular atrophy.
- infiltrates affecting the connective tissue surrounding the individual muscle fibers (the endomysium) are composed of CD8+ T cells and macrophages.[9]
- absence of rimmed vacuoles (the presence of rimmed vacuoles indicates a diagnosis of inclusion body myositis).

APPROACH TO MANAGEMENT OF PRIMARY CONDITION

The goals of therapy for myositis are to decrease inflammation in targeted tissues and allow for the rebuilding of degenerated muscle fibers leading to reduced symptoms (improving muscle strength and rebuilding endurance), so the patient can carry out activities of daily living.[2,3] Another important goal is to minimize the effects of any extramuscular complications, such as skin disorders and lung, heart, or gastrointestinal issues.[2]

Types of Treatment

Much more research is needed to learn about the effectiveness of various immunosuppressant and immunomodulatory agents in treating dermatomyositis and polymyositis.[10] Accumulating patients for large-scale trials is challenging because myositis is rare. Consequently, the current treatment models for myositis use medicines that have worked well in treating other systemic inflammatory disorders.[11]

A muscle biopsy is required to confirm inflammation before starting treatment.[4]

First-Line Therapy

Corticosteroids are the first-line, principle treatment for dermatomyositis and polymyositis.[4,10,12] However, myositis is sometimes refractory to steroid treatment.[10]

Second-Line Therapy

Second-line agents for treating myositis are methotrexate (Rheumatrex®), azathioprine (Imuran®), or IVIG (human). The decision to begin therapy with a second-line agent can be based on the following criteria[2]:

- If a steroid-sparing effect is needed because the patient develops complications even though responsive to corticosteroid therapy
- If lowering the high steroid dose causes a relapse

- If a course of two to three months of high-dose prednisone is ineffective
- If the patient suffers from rapidly progressive weakness and respiratory failure

Third-Line Therapy

The main third-line agents used to treat myositis include cyclophosphamide (Cytoxan®), cyclosporine (Sandimmune®), tacrolimus (Prograf®), and mycophenolate mofetil (CellCept®).[13]

Physical Therapy Treatment

Adjunctive treatment for myositis includes physical therapy to maintain range of motion and increase muscle strength.[11] Exercise that is guided by the physician as well as physical and occupational therapists may help to regain some muscle mass.[14] However, the benefit only lasts as long as exercise is continued.

Treatment-Resistant Myositis

When a patient does not respond to therapy, consider the following possibilities[14]:

1. incorrect diagnosis—a muscle re-biopsy may be required to confirm original diagnosis
2. insufficient drug dosing or presence of steroid myopathy
3. non-adherence to treatment plan
4. muscle weakness caused by muscle damage or muscle atrophy, not persistent disease activity
5. malignancy (especially in DM)
6. true treatment resistance

PREVENTION AND MANAGEMENT OF COMPLICATIONS

Accordant helps patients prevent and manage complications by teaching early warning signs, encouraging adherence to treatment plans, offering supportive care, and recommending physician contact where needed. The list of goals and cooperative interventions listed below does not represent a comprehensive list of complications but reflects some of the more common clinical situations specific to PM/DM. General health topics (e.g., age-appropriate cancer screening) are beyond the scope of this document.

Goal: Improve Self-management Skills
Cooperative interventions: Teach patients to:

- strengthen their personal motivation skills;
- develop prevention-focused, self-management skills;
- develop purposeful communication skills and maintain open, ongoing communications with their physician; and
- work with Accordant for education, information, and self-care needs.

Goal: Limit the impact of skin involvement in dermatomyositis
Cooperative interventions: Teach patients to[15]:

- understand the dermatomyositis disease process;
- practice proper protective dress while outside;
- understand the importance of obtaining sunscreens that block both ultraviolet A and ultraviolet B;
- use tepid sponge baths and compresses to relieve itching;
- avoid high potency topical corticosteroids where they are very easily absorbed, such as the face or on areas inflamed by the chafing of adjacent areas of skin;
- report draining ulcers in the skin to a doctor or nurse immediately;
- have regular eye exams if taking hydroxychloroquine therapy;
- understand the adverse side effects of long-term corticosteroids and report occurrence to a nurse or doctor;
- understand and report the side effects of immunosuppressive agents;
- understand the teratogenic effects of medicines and take appropriate contraceptive measures as needed; and
- take all medicines as prescribed.

Goal: Educate about the potential for cardiovascular involvement and promote early detection.
Cooperative interventions: Teach patients to:

- understand that cardiovascular involvement can be asymptomatic;
- have regular clinical exams (including blood pressure and cholesterol[16]) and electrocardiograms;
- reduce the risk of heart attack and stroke by making efforts to keep blood pressure below 120/80 mm Hg and total cholesterol below 200 mg/dL.[17]
- ask MD if heart medications such as nitrates, β-blockers, and calcium channel blockers are needed to lower poorly controlled blood pressure.[18]
- ask MD about low-dose aspirin
- report any cardiovascular-related abnormalities to a doctor or nurse as soon as possible; and
- take all medications as prescribed.

Goal: Detect and treat interstitial lung disease (ILD) early
Cooperative interventions: Teach patients to:

- recognize not only the signs and symptoms of ILD but also that it may also be asymptomatic;
- understand that ILD can be life-threatening;
- know the risk factors for ILD (e.g., anti-Jo antibodies[19]) and discuss testing with doctor;
- understand the adverse side effects of corticosteroid and immunosuppressants and the need to report them to a doctor or nurse; and

- take all medicines as prescribed.

Goal: Prevent or ensure prompt diagnosis of infections
Cooperative interventions: Teach patients to:

- understand that the myositis disease process and the immunosuppressant medicines used to treat the disease make patients more vulnerable to infections[20];
- know the signs and symptoms of infections and the need to report them to a doctor or nurse as soon as possible;
- talk to their child's rheumatologist about the risks/benefits of the varicella (chickenpox) vaccine[21];
- talk to their rheumatologist about the risks/benefits of the herpes zoster (shingles) vaccine if ≥60 years old[22,23];
- understand and practice good aseptic procedures; and
- take all medicines as prescribed.

Goal: Facilitate early detection of cancer-associated myositis (CAM)
Cooperative interventions: Teach patients to:

- understand the increased risk of malignancy, especially in those with DM who test negative for the antibodies normally associated with myositis[24,25];
- understand the general signs and symptoms of cancer (e.g., change in a mole, persistent cough);
- know the importance of regular clinical exams and lab studies that are appropriate for their age, gender, and ethnicity to evaluate for malignancies[24];
- understand that if cancer is found and successfully treated, myositis symptoms may improve[26]; and
- take all medicines as prescribed.

Goal: Avoid hospitalization, surgery, and disability associated with osteoporotic fractures
Cooperative interventions: Teach patients to:

- know the importance of BMD testing[27];
- recognize the importance of calcium and vitamin D supplements;
- understand the importance of smoking cessation;
- learn the importance of reducing alcohol consumption;
- discuss treatment options to prevent and/or repair bone loss with a doctor;
- participate in exercise that improves balance, agility, strength, and posture[28];
- learn fall prevention strategies (e.g., check and correct vision problems)[28,29]; and
- improve home safety.[30]

PATIENT EDUCATION

Accordant Health Communities Web site at:
https://www.accordant.com

The Myositis Association Web site at:
http://www.myositis.org/

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