What Researchers Say About the Treatment and the Management of Juvenile Dermatomyositis (JDM)

The diagnosis of Juvenile Dermatomyositis raises a lot of questions. Patients and families wonder, how do I know what is the best treatment plan?

A treatment plan is based on many factors, including the severity and expression of the JDM. Each case is different, and the symptoms can change over time. Some patients suffer more skin rashes, and others suffer from more muscle weakness. Physicians create treatment plans to address individual needs, but all plans are designed to provide the greatest benefit to the patient and minimize side effects.

Recently, some of the best physicians in the world that diagnose and treat JDM patients pooled their collective experience and published a series of consensus plans to standardize and optimize treatment for a variety of JDM patients. These plans can help you discuss treatment options with your doctor.

The plans are divided into categories, which are based on patient diagnosis.

1. **Patients with new onset JDM, and patients diagnosed with JDM classified as moderate.**

Paper Title: Protocols for the Initial Treatment of Moderately Severe Juvenile Dermatomyositis: Results of a Children's Arthritis and Rheumatology Research Alliance Consensus Conference

Link to Paper: https://onlinelibrary.wiley.com/doi/epdf/10.1002/acr.20071

For patients that met the criteria of moderate JDM, the panel of physicians agreed on three, possible treatment plans (Figure 1).

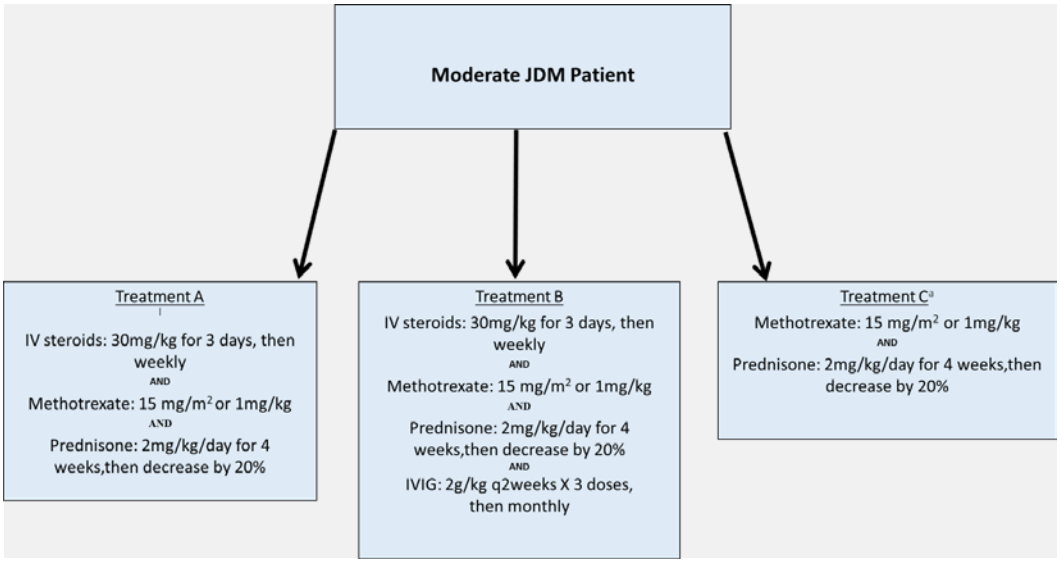

Figure 1. Consensus treatment plans for newly diagnosed patients classified with moderate JDM.

The panel of physicians also created a consensus treatment plan for 2-months after initial treatment.

Paper Title: Consensus Treatments for Moderate Juvenile Dermatomyositis: Beyond the First Two Months. Results of the Second Childhood Arthritis and Rheumatology Research Alliance Consensus Conference

Link to Paper: https://onlinelibrary.wiley.com/doi/epdf/10.1002/acr.20695

This a follow up paper to the original, intended to describe consensus treatment plans for the same group of patients with moderate JDM beyond the first 2 months of treatment. Given the critical role of steroids in the treatment of JDM, one of the key components of this work was consensus on a tapering regimen for steroids, balancing the risk of inadequately treating the JDM and the risk of side effects. This tapering was modified depending on whether patients were improved, unchanged or worse. Some additional consensus was reached on further follow up and investigations and is reported in this paper.

2. **Patients with JDM that have improved symptoms of muscle weakness but have a continued skin rash.**

Following initial treatment, some JDM patients get much better. For example, some patients muscle weakness disappears. However, some symptoms persist, in particular, a skin component of the disease. This subset of patients often requires changes to their treatment to improve their rash (Figure 2). For patients that met the criteria, the panel of physicians agreed on three, possible treatment plans (Figure 2).

Paper Title: Childhood Arthritis and Rheumatology Research Alliance Consensus Clinical Treatment Plans for Juvenile Dermatomyositis with Persistent Skin Rash.

Link to paper: https://ped-rheum.biomedcentral.com/track/pdf/10.1186/s12969-016-0134-0

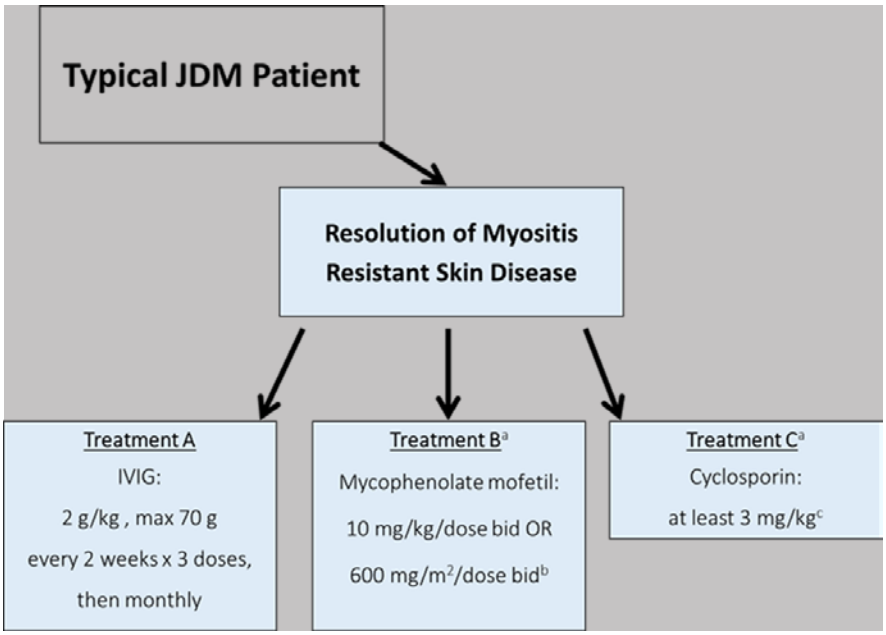

Figure 2. Consensus treatment plans for patients classified with JDM with persistent Skin Rash

3. **Patients with new onset JDM classified as amyopathic, skin predominant or juvenile dermatomyositis sine myositis.**

Some patients with JDM have a significant skin rash, but no significant weakness, or muscle inflammation. Some physicians think that this group of patients can be treated differently than those children with apparent muscle involvement (like weakness, or abnormal tests like biopsy or MRI). The panel of JDM experts considered this subtype of JDM patients and developed consensus treatment plans and published their findings (Figure 3).

Paper title: Childhood Arthritis and Rheumatology Research Alliance consensus clinical treatment plans for juvenile dermatomyositis with skin predominant disease

Link to Paper: https://ped-rheum.biomedcentral.com/track/pdf/10.1186/s12969-016-0134-0

The treatment plans recommended in this publication had a number of options, depending on diagnosis and severity of disease. Specific plans are to be chosen based on the judgment of the treating physician.

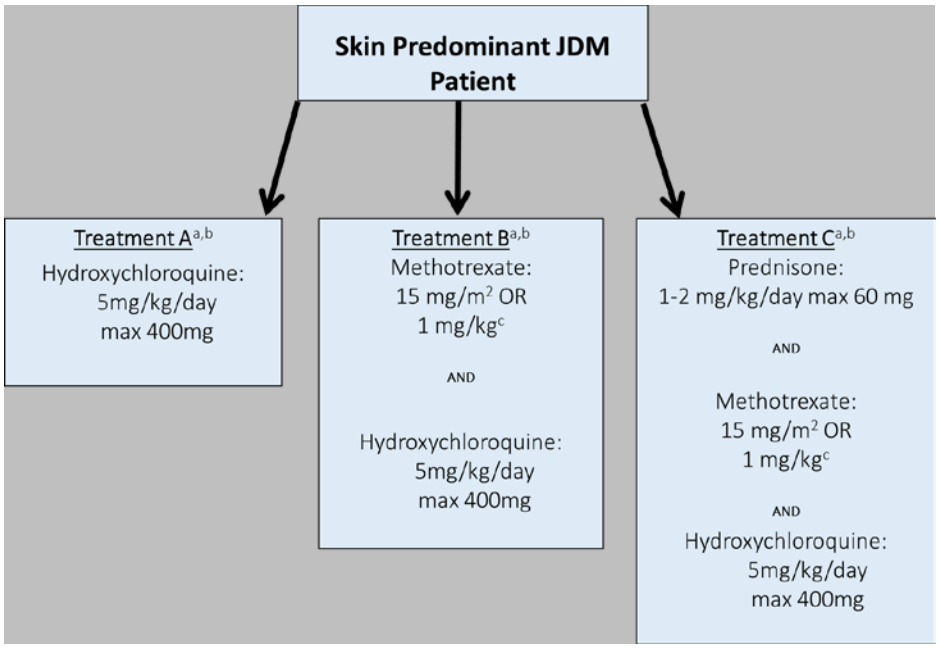

Figure 3. Treatment plans for patients with JDM that have resolved some components, but still have a skin component of the disease.

4. European experience in management of JDM.

The consensus treatment plans you just read were completed by Childhood Arthritis and Rheumatology Research Alliance (CARRA). CARRA (carragroup.org), is a North American group of clinicians and researchers focused on the care of pediatric rheumatologic diseases. But, JDM is global and groups outside of North America have made specific recommendations for the management of JDM. A European initiative called Single Hub and Access for pediatric Rheumatology in Europe (SHARE) have optimized and shared diagnostic and management plans for *European* children, and young adults with JDM. The results of the SHARE initiative resulted in a peer reviewed paper, which outlines specific, European approaches to care.

Link to paper: https://ard.bmj.com/content/annrheumdis/76/2/329.full.pdf

Since these recommendations were completed by and for European patient populations, it is not known if the standards can work in North America.

It is our hope that these plans will serve as a good starting point for conversations with your healthcare provider. The Cure JM Foundation is currently evaluating these consensus treatment plans and is actively funding research programs to expand treatment options and the number of drugs used to treat and manage JDM.