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REHABILITATION OF PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

REHABILITACIJA PACIJENATA S MIOZITISOM U SKLOPU UPALNE REUMATSKE BOLESTI

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The primary idiopathic inflammatory myopathies (IIMs) are classified as dermatomyositis (DM) and myositis of the antisynthetase syndrome (ASyS), two conditions with extra-muscular involvement, and immune-mediated necrotizing myopathy (IMNM) and inclusion body myositis (IBM), the later two being muscle-specific autoimmune diseases. Myositis can overlap with systemic lupus erythematosus (SLE), systemic sclerosis (SSc), and less often with rheumatoid arthritis (RA) and Sjögren's disease (SSjo), and is a part of mixed connective tissue disease (MCTD) associated with a high titer ANA and anti-U1 RNP.

The estimated prevalence of IIM is 14 do 30 per 100 000 inhabitants. The peak incidence is at the age of 50 to 60 years, but the onset is possible at any age, including in children (juvenile IIM). The female to male ratio is about 2:1 except for IBD with male predominance and onset over the age of 40.

The disease onset is in most cases subacute over weeks or months. Muscle weakness typically is proximal, including neck flexors, bilateral and symmetrical except for IBM. Patients with IBM present with proximal (quadriceps) and distal (fingers flexors) and asymmetrical muscle weakness with amyotrophy. Up to 20% of patients with DM can present with amyopathic disease. Extra-muscular involvement includes skin changes in DM, intestinal lung disease (ILD), arthralgia and arthritis, calcinosis and Raynauds syndrome. Typical skin changes in DM (Gottron's papules and sign, and typical distribution of skin erythema) precede the muscular weakness for 3 to 6 months. DM is associated with cancer.

The diagnosis is made based on elevated muscle enzymes, presence of myositis specific antibodies (MSA), myogenic changes on electromyography, magnetic resonance (MRI) findings of muscle edema and thypical myopathological findings on muscle biopsy. MSA are found only in IIM patients, and in

up to 60% of cases. Usually, only one MSA is present in IIM, making them a useful tool for the diagnosis of a specific IIM. Moreover, some are indicative of exstra-muscular features or malignancy, and some MSA titers correlate with disease activity.

Glucocorticoids are usually combined with methotrexate or azathioprine as the first-line treatment. Intravenous immunoglobulin (IVIg) should be considered for refractory DM and IMNM, and in patients severely affected with life-threatening complication. Second-line therapy includes rituximab, mycophenolate mofetil and calcineurin inhibitors. Topical glucocorticoids, calcineurin inhibitors, and hydroxychloroquine should be considered for DM skin rash. There is no validated treatment for calcinosis.

In 2016 ACR/EULAR proposed core set measures (CSMs) in IIM and clinical response criteria in adult and juvenile IIM. CSMs include: physician global activity score (MD Global); parent's global evaluation of the child's overall well-being score (Parent Global); muscle strength assessment (MMT or CMAS); serum muscle enzyme or physical summary score of the child health questionnaire-parent form 50 (Enzyme or CHQ-PhS); the most abnormal serum muscle enzyme value among CPK, aldolase, ALT, AST and LDH; extra-muscular disease activity (ExtraMusc/DAS); and physical function (C-HAQ). At least 20% improvement in three of six CSMs, with no more than one or two worsening (muscle strength is not allowed to worsen), is required. In adult IIM a total improvement score of 0-100 can be calculated using absolute percent change in CSMs. Thresholds for minimal (≥20 points), moderate (≥40), and major improvement (≥60) are given.

Muscle weeknes, muscle atrophy, dysphagia, dysphonia, pain, fatigue, reduced aerobic capacity, calcinosis and joint contractures, frequent falls, osteoporosis and fractures, and avascular osteonecrosis cause damage and functional impairment, and patients develop sustained disability and reduced quality of life. Muscle inflammation (edema detectable on MRI) and damage (atrophy and fatty replacement) and consequently muscle prognosis is worse in IMNM than in DM and ASyS especially in early-onset of IMNM. Patients with IBM present with slowly progressive loss of muscle strength and atrophy, and they need a wheelchair for ambulation in the late stage of the disease with limited use of their hands and marked dysphagia. Life span in IBD is mildly reduced with aspiration pneumonia and respiratory complications as the most common cause of death.

Exercise is recommended for all IIM. In IBM, where glucocorticoids and immunosuppressants or immunomodulators drugs are not effective, exercise represents the core therapy. Exercise has an anti-inflammatory effect instead of exacerbating muscle inflammation. Thus, exercise does not alter

inflammatory markers or cause exacerbation of IIM. Exercise is safe in all stages of IIM and in all age groups of patients. Exercise improves muscle strength, muscle endurance, aerobic capacity, reduce glucocorticoid induced muscle atrophy, is important in reduction of falls and fractures in IIM, may exhibit a positive influence on BMD and osteoporosis, and exercise improves cognitive and psychological health of patients with IIM. Muscle strengthening exercises, aerobic exercise, aquatic exercise, and home exercises are part of the exercise program in IIM.

Keywords: Myositis; Idiopathic Inflammatory Myopathy; Exercise; Muscle Strength; Physical Functional Performance

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