Chapter 10
Musculoskeletal system

CONTENTS

1 Arthritis

Juvenile idiopathic arthritis

Management

Rheumatic diseases require symptomatic treatment to relieve pain, swelling, and stiffness, together with treatment to control and suppress disease activity. Treatment of juvenile idiopathic arthritis may involve Non-steroidal anti-inflammatory drugs p. 703 (NSAIDs), a disease modifying anti-rheumatic drug (DMARD) such as methotrexate p. 588 or a cytokine modulator, and intra-articular, intravenous, or oral corticosteroids.

Rheumatic disease, suppressing drugs

Overview

Certain drugs, such as methotrexate p. 588, cytokine modulators, and sulfasalazine p. 36, are used to suppress the disease process in *juvenile idiopathic arthritis* (juvenile chronic arthritis); these drugs are known as disease-modifying antirheumatic drugs (DMARDs). In children, DMARDs should be used under specialist supervision.

Some children with juvenile idiopathic arthritis do not require DMARDs. Methotrexate is effective in juvenile idiopathic arthritis; sulfasalazine is an alternative but should be avoided in *systemic-onset juvenile idiopathic arthritis*. Gold and penicillamine p. 666 are no longer used. Cytokine modulators have a role in *polyarticular juvenile idiopathic arthritis*.

Unlike NSAIDs, DMARDs can affect the progression of disease but they may require 3–6 months of treatment for a full therapeutic response. Response to a DMARD may allow the dose of the NSAID to be reduced.

DMARDs can improve not only the symptoms of inflammatory joint disease but also extra-articular manifestations. They reduce the erythrocyte sedimentation rate and C-reactive protein.

Antimalarials

The antimalarial hydroxychloroquine sulfate p. 690 is rarely used to treat juvenile idiopathic arthritis.
Hydroxychloroquine sulfate can also be useful for systemic or discoid lupus erythematosus, particularly involving the skin and joints, and in sarcoidosis.

Retinopathy rarely occurs provided that the recommended doses are not exceeded.

Mepacrine hydrochloride is used on rare occasions to treat discoid lupus erythematosus [unlicensed].

Drugs affecting the immune response

Methotrexate, given as a once weekly dose, is the DMARD of choice in the treatment of juvenile idiopathic arthritis and also has a role in juvenile dermatomyositis, vasculitis, uveitis, systemic lupus erythematosus, localised scleroderma, and sarcoidosis; for these indications it is given by the subcutaneous, oral, or rarely, the intramuscular route. Absorption from intramuscular or subcutaneous routes may be more predictable than from the oral route; if the oral route is ineffective subcutaneous administration is generally preferred. Folic acid may reduce mucosal or gastro-intestinal side-effects of methotrexate. The dosage regimen for folic acid p. 620 has not been established—in children over 2 years a weekly dose [unlicensed indication], may be given on a different day from the methotrexate.

Azathioprine p. 558 may be used in children for vasculitis which has failed to respond to other treatments, for the management of severe cases of *systemic lupus erythematosus* and other connective tissue disorders, in conjunction with corticosteroids for patients with severe or progressive renal disease, and in cases of *polymyositis* which are resistant to corticosteroids. Azathioprine has a corticosteroid-sparing effect in patients whose corticosteroid requirements are excessive.

Ciclosporin p. 559 is rarely used in juvenile idiopathic arthritis, connective tissue diseases, vasculitis, and uveitis; it may be considered if the condition has failed to respond to other treatments.

Cytokine modulators

Cytokine modulators should be used under specialist supervision.

Adalimumab p. 693, etanercept p. 696, and infliximab p. 38 inhibit the activity of tumour necrosis factor alpha (TNF-α). Adalimumab can be used for the management of active polyarticular juvenile idiopathic arthritis and enthesitis-related arthritis. Etanercept is licensed for the treatment of the following subtypes of juvenile idiopathic arthritis: polyarticular juvenile idiopathic arthritis in children who have had an inadequate response to methotrexate or who cannot tolerate it, oligoarthritis in children who have had an inadequate response to methotrexate or who cannot tolerate it, psoriatic arthritis in children over 12 years who have had an inadequate response to methotrexate or cannot tolerate it, and enthesitis-related arthritis in children over 12 years who have had an inadequate response to conventional therapy or cannot tolerate it. Infliximab has been used in