**Section 5.13**

**Rheumatology**

**Box 5.13.1 Minimum standards**
- Penicillin.
- Aspirin.
- Prednisolone.
- Haloperidol, diazepam and lorazepam.
- Anti-endocarditis measures.
- IV gamma globulin if at all possible.
- Non-steroidal anti-inflammatory drugs (NSAIDs).
- Sulphasalazine.
- Ocular steroids and mydriatics.
- Intra-articular steroids.
- Physiotherapy and family support.

**Introduction**

Making a diagnosis of a rheumatic disease in a child relies primarily on clinical skill and experience, as there are few diagnostic laboratory tests. Although these diseases are rare in children, symptoms that raise the possibility of rheumatic disease are common. Rheumatic symptoms may be relatively specific, such as joint swelling, or relatively non-specific, such as fever, lethargy, pallor, anorexia, failure to thrive, muscle weakness, musculoskeletal pain, rash, headache and abdominal pain. The interpretation of these clinical features requires a meticulous approach to characterising the nature of each feature and considering the overall pattern of all the clinical features in the individual patient. The aims of this section are to assist in the recognition of common patterns of clinical features, and to provide guidance for appropriate treatment and monitoring of rheumatic disease in children.

pGALS (paediatric Gait, Arms, Legs and Spine) is a simple quick approach to joint examination and helps to discern abnormal from normal joints; this is especially useful in the context of non-specific features such as limp or fever. pGALS includes incorporates a series of simple manoeuvres to assess all joints quickly (takes approximately 2–3 minutes). It has been validated in school-aged children (although can be performed in younger children) and has been shown to be effective when performed by non-specialists in detecting significant joint abnormalities in acute paediatric practice (including in Africa). The interpretation of pGALS requires knowledge of normal musculoskeletal development and the clinical context to facilitate a differential diagnosis (www.arthritisresearchuk.org/health-professionals-and-students/video-resources/pgals.aspx).

**Rheumatic fever**

Rheumatic fever is an abnormal immune response to group A streptococcal infection in genetically susceptible individuals. It is most common between the ages of 6 and 16 years. Symptoms of acute rheumatic fever follow streptococcal pharyngitis after a latent period of approximately 3 weeks. The disease usually presents with joint pain, but may have an insidious onset, especially if carditis is the predominant feature. There is no definitive test, and diagnosis depends on recognition of clinical signs known as the Jones criteria.

**Further reading**


Jones criteria for diagnosis of rheumatic fever

<table>
<thead>
<tr>
<th>Major criteria</th>
<th>Minor criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carditis</td>
<td>Previous history of rheumatic fever</td>
</tr>
<tr>
<td>Migratory large-joint polyarthritis</td>
<td>Fever</td>
</tr>
<tr>
<td>Erythema marginatum</td>
<td>Arthralgia</td>
</tr>
<tr>
<td>Subcutaneous nodules</td>
<td>First-degree heart block</td>
</tr>
<tr>
<td>Chorea (onset 2–6 months after pharyngitis)</td>
<td>Elevated acute-phase reactants (ESR and CRP)</td>
</tr>
</tbody>
</table>

ESR, erythrocyte sedimentation rate; CRP, C-reactive protein.

Diagnosis: Jones criteria

The diagnosis in an individual is made by the Jones criteria (revised by the WHO in 2003) on the basis of the presence of either two major criteria (which include polyarthritis, erythema marginatum rash, subcutaneous nodules, carditis and chorea) or one major criterion and two minor criteria (minor criteria include persistent fever, arthralgia, raised ESR or CRP, persistent leukocytosis, abnormal ECG except if carditis is the major feature, and previous episode of rheumatic fever). Each combination must also include evidence of streptococcal infection, usually a rising titre of antistreptolysin O.

- Evidence of streptococcal infection (usually a pharyngitis secondary to group A beta-haemolytic streptococcus) with positive throat swab culture or, preferably, a positive serology for recent streptococcal infection. This is usually accompanied by a prolonged fever and followed by other clinical features after a 2- to 3-week period.
- Arthritis of the large joints. This is a reactive arthritis (rather than a septic arthritis), often affecting many joints, and it is migratory in nature. It usually responds dramatically to aspirin, up to 120 mg/kg/day in four to six divided doses by mouth after food, but do not exceed 75–80 mg/kg/day if facilities for assay of salicylate levels are not available. Alternatively, use non-steroidal anti-inflammatory drugs (NSAIDs; see below). The presence of joint pain without swelling (i.e. arthralgia alone) may still indicate rheumatic fever in the presence of the other clinical features compatible with a diagnosis of acute rheumatic fever.
- Rash and subcutaneous nodules: erythema marginatum is an uncommon feature. It has a ‘snake-like’ appearance, usually over the trunk, and occurs early in the disease, is usually transient, and disappears within a few hours. Subcutaneous nodules are not uncommon, occurring over bony prominences such as the elbows and knees.
- Carditis: this may range from a tachycardia with a prolonged PR interval seen on the ECG through to myocarditis with a systolic apical mitral murmur, pericarditis or cardiac failure. Cardiac inflammation may involve the endocardium (valvulitis mostly affecting the mitral and aortic valve), the myocardium (impaired cardiac function) or the pericardium in severe cases (pericarditis). Examination may reveal a pericardial friction rub, an apical pansystolic murmur from mitral regurgitation, or an early diastolic decrescendo murmur from aortic regurgitation. As the valves heal they may scar and fibrose. Mitral regurgitation, mitral stenosis and aortic regurgitation are the commonest long-term consequences of acute rheumatic fever.
- Chorea is an involuntary movement disorder, often of the face, tongue and upper limbs. It may appear as dysarthria or clumsiness, and is associated with emotional lability. It is a late manifestation of acute rheumatic fever, and is more common in girls.

The disease may be prevented by detecting group A streptococcus in cases of pharyngitis (throat swab or rapid antigen test) and treating with penicillin (see below).

Treatment

Management of acute rheumatic fever

- Eradicate streptococcal infection (give oral penicillin V 10–12.5 mg/kg/dose (maximum dose 500 mg) three times a day for 10 days).
- Commence aspirin 90–120 mg/kg a day in four divided doses after food. Monitor serum salicylate levels (the optimal level is 15–25 mg/dL). Reduce the dose to two-thirds of the original dose when there is a clinical response.
- When the CRP and ESR decrease to normal levels, taper the aspirin dose over 2 weeks.
- Give prednisolone 2 mg/kg/day (maximum 60 mg/day) in place of aspirin if there is moderate to severe carditis or pericarditis.
- If prednisolone is given, continue treatment for 3 weeks, and then taper the dose over a further 2–3 weeks. As the prednisolone dose starts to taper, commence aspirin 50 mg/kg/day in four divided doses and stop aspirin 1 week after prednisolone is stopped.
- Treat heart failure as described in Section 5.4.B.
- Urgent valve replacement is sometimes required.

The requirement for bed rest during the acute attack is controversial; it is also very difficult to enforce on young children.

For arthralgia, give aspirin as described above or an NSAID (e.g. ibuprofen 30–60 mg/kg daily up to a maximum of 2.4 G in three to four divided doses after food). Naproxen at 20 mg/kg/day in two divided doses appears to be a better alternative.

Treatment of streptococcal infection with IM benzathine penicillin (1.2 million units as a single injection, often given as 0.6 million units in each thigh) or a 10-day course of oral penicillin at high dose (12.5 mg/kg four times a day). Once there has been one episode of rheumatic fever a recurrence is likely. The recurrence risk is minimised by giving long-term penicillin prophylaxis, preferably for life. This is usually given as intramuscular injections of 1.2 million units of benzathine penicillin every 3 weeks (this drug must not be given IV). If oral penicillin is required, the highest dose generally recommended is 250 mg twice daily for all ages, as doses of oral penicillin in children below the age of 5 years need not be given because rheumatic fever does not occur in this age group. For patients who are allergic to penicillin, erythromycin in the same doses can be used.

For acute carditis, prednisolone given orally (2 mg/kg/day) for 2–3 weeks or by intravenous infusion is effective.

Chorea may respond to haloperidol, 12.5–25 micrograms/kg twice daily (maximum 10 mg a day). Extrapyramidal side effects may occur. Chorea usually becomes less of a problem within a few weeks.
Vasculitis in children

Vasculitis in childhood may be primary, including Henoch–Schönlein purpura, Kawasaki disease and the rare vasculitides, or secondary to multisystem connective tissue diseases, including juvenile dermatomyositis and systemic lupus erythematosus (SLE). In all of these diseases, skin manifestations are usually prominent, but the combination with other clinical features helps to ascertain the diagnosis.

Henoch–Schönlein purpura (HSP)

Presentation

- Purpuric rash: a palpable purpuric rash is most commonly seen over the buttocks and around the ankles and legs. The purpura occurs in crops and may range from small petechiae-like lesions to large ulcerating ecchymoses. Oedema and urticaria may precede the purpura, particularly at the ankles, scrotum and face.
- Gastrointestinal pain: abdominal pain is a prominent feature early in the disease, and is often accompanied by vomiting. Occasionally, frank gastrointestinal haemorrhage may occur.
- Arthritis: this typically affects the large joints of the lower limb, especially the ankles. Ankle swelling may be difficult to interpret in the presence of tissue oedema. The joint pain is usually transient. Arthritis in HSP is never erosive.
- Renal disease: haematuria and proteinuria are common manifestations of the disease, but are usually only detected on dipstick urine analysis. A small proportion of children (1–3%) may develop renal failure secondary to severe glomerulonephritis. Clinically significant renal disease is uncommon below 5 years of age.

Treatment

Henoch–Schönlein purpura is usually a self-limiting disease, requiring supportive care and symptomatic treatment with simple analgesia only. If the abdominal pain is severe, prednisolone (1–2 mg/kg/day) for 1 week may be helpful.

Kawasaki disease

Kawasaki disease is characterised by a combination of most of the following features in a young child (usually less than 5 years old) who is extremely irritable.

- Fever: an irregular spiking fever that persists for 1–3 weeks despite antibiotics is characteristic during onset.
- Skin involvement: rash is variable and polymorphic, ranging from diffuse erythema of the trunk and face to minimal macular lesions on the limbs. The rash in Kawasaki disease is never vesicular. Tissue oedema of the dorsal surfaces of the hands, feet and perineum is characteristic. These changes are followed within days to weeks by desquamation, usually of the finger and toe tips (periungual desquamation), but occasionally more widespread.
- Mucositis and conjunctivitis: inflammation of the mucous membranes of the mouth and eyes results in a characteristic appearance of red eyes (conjunctival ‘injection’ rather than conjunctivitis) and red swollen cracked lips.
- Lymphadenopathy: this usually affects the cervical lymph nodes, often unilaterally.
- Cardiac disease: myocarditis with heart failure or pericarditis is a rare but serious complication of Kawasaki disease. Coronary artery aneurysms may be present from early in the disease process. Clinical manifestations are relatively non-specific, but the two-dimensional echocardiography appearances are diagnostic of the condition. However, echocardiography may be completely normal in Kawasaki disease.

Kawasaki disease is a rheumatological emergency. Delays in recognition and treatment of this condition can result in the development of coronary artery abnormalities with disastrous long-term consequences, including fatalities.

Treatment

- Hospitalisation and monitoring of cardiac status.
- Aspirin, 50–75 mg/kg/day in 4 divided doses after food until the acute inflammatory phase of the disease settles, then 1–10 mg/kg/day (usually 3–5 mg/kg/day) (antiplatelet doses).
- Intravenous gamma globulin 2 grams/kg immediately on diagnosis, if available. Every effort must be made to procure intravenous gamma globulin for the treatment of these children, as this is the only effective therapy for Kawasaki disease. This treatment reduces the likelihood of coronary artery aneurysms if given as early as possible during the illness (several inexpensive brands of intravenous gamma globulin are now available in resource-limited countries).
- Corticosteroids (e.g. prednisolone 1–2 mg/kg/day) may have a role in controlling the acute inflammation of Kawasaki disease, but are generally not recommended.
- Follow-up clinical examination and echocardiography (if available) is recommended at 6–8 weeks, as coronary artery aneurysms may appear after the initial presentation.

Juvenile idiopathic arthritis

Juvenile idiopathic arthritis is one of the more common physically disabling chronic diseases of children. The most prominent clinical features include joint swelling, restriction of joint movement, joint pain and tenderness at the joint margins, muscle wasting and any of the features mentioned below. The most common mistake is to diagnose arthritis in the absence of objective evidence of persistent joint swelling.

Diagnosis of juvenile idiopathic arthritis

All of the following four criteria are required:

1. The presence of arthritis, defined by swelling of a peripheral joint. Loss of joint range of movement and pain on movement are sufficient for the definition of arthritis involving the hip or spine (in the absence of other causes for the pain).
2. Persistence of arthritis for more than 6 weeks.
3. Onset of arthritis before the child’s 16th birthday.
4. The absence of any known cause for the arthritis.

Classification and differential diagnosis

There are a variety of different forms of juvenile idiopathic arthritis that are important to consider when advising on the prognosis and most appropriate treatment of the illness.

- Arthritis affecting only a few joints: oligo-arthritis
Arthritis affecting many joints: polyarthitis is likely to persist into adulthood in 40% of cases.

Arthritis affecting few or many joints with prominent extra-articular features:
- Systemic arthritis: with fever, rash, and enlargement of the liver, spleen and lymph nodes. Pericarditis and macrophage activation syndrome are life-threatening complications. Macrophage activation syndrome presents with persistent fever, encephalopathy, liver failure and clotting abnormalities and low platelet counts. The persistence of arthritis with this illness carries the worst prognosis: over 50% of these children have arthritis as adults.
- Psoriatic arthritis: often associated with a psoriatic rash, nail pitting and a family history of psoriasis. This has a similar outcome to polyarthitis.
- Enthesitis-related arthritis: the clinical manifestations of enthesitis include pain, tenderness and occasionally swelling localised to the exact site of tendon insertion. Other features include back pain, red painful eyes and urethritis. There is a 60% risk of development of ankylosing spondylitis in adulthood.

Monitoring for complications and disease progress in juvenile idiopathic arthritis

There are several important complications of juvenile idiopathic arthritis, including joint failure, chronic anterior uveitis and local growth disorders, as well as the general complications of chronic inflammatory disease in children, such as anaemia, fatigue, delayed puberty and growth failure. Three of these complications, namely joint failure, chronic anterior uveitis and growth disorders, will be discussed in more detail.

**Joint failure**
- Inability to walk without pain and stiffness.
- Inability to write or perform activities of self-care without pain and stiffness.
- The integrity of joint cartilage and bone density is affected from the onset of the disease.
- If the inflammation remains poorly controlled, destruction of cartilage, joint space narrowing and erosion of bone will result in permanent loss of joint function.

**Differential diagnosis of juvenile idiopathic arthritis**
- Transient arthritides: irritable hip, reactive arthritis.
- Septic arthritis and osteomyelitis, including immunodeficiency.
- Acute lymphoblastic leukaemia, neuroblastoma, lymphoma and local neoplasia.
- Bleeding diatheses: haemophilia.
- Haemoglobinopathies: thalassaemia, sickle-cell crisis.
- Epiphyseal disorders: dysplasia, avascular necrosis, osteonecrosis, slipped upper femoral epiphysis.
- Metabolic and endocrine disorders.
- Traumatic joint disease, including non-accidental injury.
- Hypermobility and inherited connective tissue diseases.
- Systemic connective tissue diseases, including systemic lupus erythematosus, dermatomyositis and vasculitis.
- Idiopathic musculoskeletal pain syndromes.

<table>
<thead>
<tr>
<th>Joint affected</th>
<th>Type of contracture</th>
<th>Consequence of contracture</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tibio-talar gait</td>
<td>Plantar flexion</td>
<td>Circumduction or high-deforty stepping</td>
</tr>
<tr>
<td>Knee</td>
<td>Flexion</td>
<td>Quadriceps wasting, limping gait</td>
</tr>
<tr>
<td>Hip</td>
<td>Flexion</td>
<td>Limited 'swing-phase' gait</td>
</tr>
<tr>
<td>Wrist</td>
<td>Flexion</td>
<td>Poor writing</td>
</tr>
<tr>
<td>Neck</td>
<td>Flexion</td>
<td>Poor neck rotation</td>
</tr>
</tbody>
</table>

- Arthritis of inflammatory bowel disease.

**Note**: diseases shown in bold type in the above list are emergencies, and require prompt expert management.

**Initial minimal set of investigations for differential diagnosis**
- Full blood count, including white blood cell differential and platelet counts.
- Plain radiographs of affected joints.
- Synovial fluid aspiration, microscopy and culture.
- Blood culture.

**Eye disease**
- Chronic anterior uveitis is typically insidious and asymptomatic: all children with juvenile idiopathic arthritis (but especially those with oligo-arthritis) should undergo slit-lamp eye examination to detect cells in the anterior chamber and protein ‘flare’. Delay in the diagnosis can lead to blindness.
- Inflammation is treated with ocular topical cortico-steroids (hydrocortisone 1% eye drops or ointment 0.5%) three times daily and mydriatics (3 minutes after hydrocortisone) (atropine 0.5% eye drops or 1% ointment).
- Severe chronic anterior uveitis may require systemic treatment with corticosteroids or methotrexate.

**Growth disorders**
- Generalised growth failure may be due to inadequate energy intake (chronic inflammatory disease increases energy demands) or the adverse effects of medication. It is usually treated with dietary energy supplements.
- Local growth disturbance: bony overcrowth of the knee with an increase in leg length, sometimes with a valgus knee deformity. Arthritis of the small joints of the hands is likely to cause premature fusion of the epiphyses and reduced growth of the affected fingers.

**Treatment of juvenile idiopathic arthritis**
- The first priority is to exclude the differential diagnoses, especially the emergencies of septic arthritis, acute lymphoblastic leukaemia or other malignancies, and non-accidental injury. Septic arthritis will require large doses of intravenous antibiotics (see Section 5.17).
- The effective treatment of juvenile idiopathic arthritis usually requires a team of trained healthcare professionals, including therapists and medical staff.
- Education of the patient and family is important, especially concerning the risks and benefits of all treatment and the natural history of the disease.
- Physiotherapy, hydrotherapy and occupational therapy
work together to maintain joint function and muscle bulk, correct joint deformities and rehabilitate affected joints.

- Drug treatment should begin as soon as the diagnosis is made, with the following:
  - **Non-steroidal anti-inflammatory drugs (NSAIDs):**
    - Give ibuprofen up to 60 mg/kg/day up to a maximum of 2.4 g in three or four divided doses after food. Naproxen at 20 mg/kg/day in two divided doses is possibly a better alternative. Avoid using more than one NSAID at a time.
  - **Intra-articular corticosteroids:** Strict aseptic conditions, no-touch technique, appropriate sedation, and local or general anaesthetic must be given. Triamcinolone hexacetonide is the most effective steroid, at a dose of 1 mg/kg large joint (e.g. knee, hip or shoulder) or 0.5 mg/kg small joint (e.g. ankle, wrist or elbow). This technique requires an experienced operator.

For children with polyarthritis or systemic arthritis, in addition to the above, the following should be considered:

- **Methotrexate:** Begin with 500 micrograms/kg/week (up to 15 mg/week) starting dose, given orally 1 hour before food, and increased if necessary by 2.5 mg every month until 1.0 mg/kg/week (maximum 30 mg/week). Alternative dosage is 10–15 mg/m² once weekly starting dose and increased if necessary to a maximum dose of 25 mg/m² once weekly (see Section 9 for chart showing how to calculate m² from weight). The drug may be given by subcutaneous injection in severe cases. The patient should be monitored monthly for cytopenia (with full blood counts) and liver function abnormalities. Administration is sometimes accompanied by nausea, a side effect that can be improved with folic acid 1 mg once daily (not on the day of methotrexate treatment, but beginning the day after the methotrexate dose).

- **Intravenous methylprednisolone:** This may be needed for severe disease flares or for complications such as pericarditis. Give 30 mg/kg/dose (maximum dose of 1 gram) once a day for 3 days by slow intravenous infusion over a 2- to 3-hour period. Blood pressure monitoring for acute hypertension during the administration of this medication should take place every 30 minutes.

- **Sulphasalazine:** Begin with 12.5 mg/kg/day for the first week, increasing by 12.5 mg/kg/day each week until the maximum dose of 50 mg/kg/day in two divided doses is reached, or until adverse drug reactions occur. These may include a rash, nausea, abdominal pain and pancytopenia. Monitoring with 2- to 3-monthly full blood counts is a sensible precaution.

- More recently, a new group of drugs have been developed which appear to slow the progress of disease in some patients. They work by opposing tumour necrosis factor alpha, which contributes to cell damage, and are immunosuppressants. They include etanercept and infliximab. These drugs are currently very expensive.

**Paediatric systemic lupus erythematosus (SLE)**

**Pattern of clinical features in SLE**

There is a malar rash and erythema of the hard palate with hair loss in a child with multiple constitutional symptoms.

**Treatments**

- The first step is to rule out other conditions which can mimic SLE, such as infection, malignancy, post-streptococcal nephritis, other rheumatic diseases and drug-induced lupus-like syndromes.
- For mild musculoskeletal disease, NSAIDs (e.g. ibuprofen 20–40 mg/kg/day in three daily doses) are effective.
- For rapid control of acute moderate-to-severe disease, glucocorticoids (e.g. prednisone up to 2 mg/kg per day) are useful, tapering rapidly to the lowest tolerated dose.
- Hydroxychloroquine (5–7 mg/kg/day) is now a standard adjunctive therapy for limiting joint, skin and constitutional symptoms.
- Immunosuppressive agents (e.g. azathioprine, cyclophosphamide, mycophenolate mofetil) are useful additions in moderate to severe disease.
- Other general health measures that need to be considered include the following:
  - Bone health: weight-bearing exercises with calcium and vitamin D supplementation.
  - Cardiovascular health: education on modifiable risk factors for atherosclerosis, together with advice on reducing weight, smoking and cholesterol.
  - Health education (regarding vaccination, sun protection, dietary advice, exercise and reproductive health) and psychological support.
- Routine 2- to 3-month follow-up is necessary to monitor for complications. This should involve full blood count, renal and liver profiles, ESR, urinalysis, and urine protein:creatinine ratio, together with complement and antidsDNA antibody levels.

**Juvenile dermatomyositis (JDM)**

**Pattern of clinical features in JDM**

There is erythema over the face, shawl area, knuckles...
and knees, associated with proximal muscle weakness (which may be subtle).

Juvenile dermatomyositis is the most common inflammatory myopathy of childhood, and the diagnosis is based on the following criteria:

- **Muscle weakness**: a symmetrical, usually progressive weakness affecting proximal muscles.
- **Skin rash**: erythematous rashes occurring over the face or extremities, heliotrope rash over the eyelids, and Gottron’s papules over extensor joint surfaces. More severe complications include skin ulceration and calcinosis at pressure points, causing functional disabilities. Capillary loop abnormalities seen proximal to the cuticles with an auroscope are a very characteristic sign if present.
- **Laboratory evidence of muscle disease**: this can include increased activity of muscle enzymes in the blood (creatine kinase, lactate dehydrogenase, transaminases), or results from more invasive tests, such as muscle biopsy or electromyography (if available).

**Treatment**

- High-dose corticosteroids are the standard treatment, namely early IV methylprednisolone 30mg/kg per day (maximum 1 gram daily) with or without low-dose daily oral corticosteroid (500 micrograms/kg per day).
- It can be useful to add methotrexate (15mg/m²/week orally or subcutaneously) as a steroid-sparing agent and intravenous immunoglobulin in resistant cases (where available).
- Skin disease may also be helped by routine photoprotective agents and topical corticosteroids or tacrolimus.
- Physiotherapy and aerobic exercise are helpful for improving function and strength.

**TABLE 5.13.A.3** Differential diagnosis of childhood idiopathic inflammatory myopathies

<table>
<thead>
<tr>
<th>Weakness alone</th>
<th>Weakness with or without rash</th>
<th>Rash alone</th>
</tr>
</thead>
<tbody>
<tr>
<td>Muscular dystrophies (e.g. Duchenne’s, limb-girdle)</td>
<td>Viruses (enterovirus, influenza, coxsackie, echovirus, polio)</td>
<td>Psoriasis</td>
</tr>
<tr>
<td>Metabolic myopathies (e.g. glycogen- or lipid-storage disorders)</td>
<td>Bacterial (Staphylococcus, Streptococcus, Lyme disease)</td>
<td>Eczema</td>
</tr>
<tr>
<td>Endocrine myopathies (hypothyroidism, hyperthyroidism, Cushing’s syndrome, diabetes mellitus)</td>
<td>Parasitic (toxoplasmosis, trichinosis)</td>
<td>Allergy</td>
</tr>
<tr>
<td>Drug-induced myopathies (e.g. glucocorticoids, hydroxychloroquine, growth hormone)</td>
<td>Other rheumatic conditions (SLE, mixed connective tissue disease, scleroderma, juvenile idiopathic arthritis, vasculitis)</td>
<td>Other inflammatory conditions (coeliac disease, inflammatory bowel disease)</td>
</tr>
<tr>
<td>Neurological (myasthenia gravis, spinal muscular atrophy)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**5.14 Cancer in children**

**BOX 5.14.1 Minimum standards**

- Local enthusiastic clinical lead.
- Supporting team of doctors and nurses.
- Basic diagnostic pathology and imaging (X-ray and ultrasound).
- Centre with provision for some chemotherapy and surgery.
- Access to antibiotics.
- Access to blood products.
- Access to palliative care drugs (see Section 1.16).

**Ideal extra requirements**

- Imaging with computed tomography (CT) scan.
- Access to radiotherapy.
- Indwelling long-term vascular access.

**Introduction**

- More than 85% of all newly diagnosed children with cancer and 95% of deaths in children with cancer occur in low- and middle-income countries.
- With an increasing global population, principally in resource-limited countries, the number of children will continue to increase both in terms of absolute numbers and proportionally in these countries.
- As malnutrition and infection decline, particularly in young children, the worldwide contribution to mortality from cancer will increase.
- Only a limited proportion of all children with cancer in resource-limited countries receive curative therapy, and most do not even receive any form of palliative care.
- A child diagnosed with cancer who lives in one of the poorest countries has an 80% probability of dying, compared with less than 30% in the most well-resourced countries (see Figure 5.14.1).