APPENDIX VIII:

PAEDIATRIC HOSPITAL LEVEL STANDARD TREATMENT GUIDELINES FOR RHEUMATOID ARTHRITIS, 2006 EDITION

12.2 JUVENILE RHEUMATOID ARTHRITIS (JRA)/JUVENILE IDIOPATHIC ARTHRITIS (JIA)

DESCRIPTION
Juvenile rheumatoid arthritis is a chronic non-suppurative inflammatory condition of synovium.

Different clinical subgroups are recognised according to the pattern of onset:

- **Systemic onset**
  - extra articular features are most striking
  - characteristic spiking fever and erythematous macular rash
  - serositis, i.e. pericarditis and pleuritis
  - hepatosplenomegaly and lymphadenopathy
  - 50% of patients will have destructive polyarthritis with poor response to treatment

- **Pauciarticular**
  - typical patient is a pre-school girl
  - involves the large joints, i.e. wrists, knees, ankles or elbows
  - often asymmetrical distribution
  - ≤ 4 joints are involved
  - prognosis is good, depending on management
  - 15–30% develop chronic iridocyclitis, which is asymptomatic, but eventually may lead to severe visual impairment/blindness
  - There is an increased risk of iridocyclitis/uveitis in patients with positive antinuclear antibodies.
  - All children with pauciarticular disease must be examined at each visit and may need slit lamp examinations 3–4 times yearly for at least the first 5 years of disease.
  - a subgroup will develop polyarthritis, i.e. > 4 joints affected, which is then classified as extended oligo-articular JIA

- **Polyarthritis (Rheumatoid factor negative)**
  - affects ≥ 5 joints in first 6 months of disease
  - typical patient is a pre-school girl
  - symmetric arthritis of multiple joints typically including small joints of the hands
  - temporomandibular joints and cervical spine may become involved later on
  - onset may be insidious with gradual development of joint stiffness, swelling and loss of motion, or fulminant, with sudden appearance of symptomatic arthritis

- **Polyarthritis (Rheumatoid factor positive)**
  - affects ≥ 5 joints in first 6 months
  - involves large and small joints
  - follows pattern of adult RA with nodules and bony erosions
• aggressive form of disease with chronic course persisting into adulthood

**Enthesitis related arthritis** (HLA B27 positive or family history thereof)
  - mostly pre-teen and teenage boys
  - onset of arthritis in boy > 8 years
  - asymmetrical arthritis of lower limb joints and enthesitis
  - enthesitis, presenting with heel pain, plantar fasciitis, Achilles tendonitis, pain at bases of 1st and 5th metatarsals and tibial tuberosity
  - sacroiliac joint tenderness and inflammatory spinal pain
  - anterior uveitis associated with pain, redness and photophobia
  - family history of arthritis, bad backs or ankylosing spondylitis
  - associated with inflammatory bowel disease

**Clinical**
  - exclude other forms of arthritis
  - age of onset < 16 years
  - arthritis in one or more joints
  - duration > 6 weeks
  - any of the patterns of onset

**Differential diagnosis**
  - JRA is a clinical diagnosis and depends on the persistence of arthritis or typical systemic manifestations for ≥ three consecutive months and excluding other diseases:
    - pyogenic and tuberculous joint infection and osteomyelitis
    - arthritis associated with other acute infectious illnesses
    - acute leukaemia and other malignancies
    - acute rheumatic fever
    - auto immune disorders, SLE or mixed connective tissue disease
    - Reiter syndrome, i.e. arthritis, urethritis and conjunctivitis
    - ulcerative colitis or arthritis associated with enteritis

**Investigations**
  - there is no diagnostic test
  - full blood count: differential and platelet count
  - bone marrow aspiration must be done before starting disease modifying drugs, including steroid therapy and methotrexate
  - C-reactive protein and erythrocyte sedimentation rate
  - serum urea, creatinine and electrolytes
  - muscle enzymes, albumin, calcium, phosphate and alkaline phophatase
  - auto-antibodies, complement C3 and C4, rheumatoid factor, IgG and IgA levels
  - plain X–ray
  - specialist may advise arthroscopy, synovial biopsies, ultrasound and CT scan in appropriate circumstances

**NON-DRUG TREATMENT**
  - occupational and physiotherapy are essential to provide:
    - exercises to increase range of movements of joints and to maintain muscle strength
    - daily exercise programmes, hot water baths, swimming pool exercises
splints, e.g. nocturnal splints, for pain relief and prevention of contractures
- shoe inserts/raises
- advice on aids for activities of daily living
- orthodontic treatment if joints of jaw are involved

**DRUG TREATMENT**

**NSAID, e.g.:**
- ibuprofen, oral, 10 mg/kg/dose 3–4 times daily

Efficacy is determined within weeks to months unless there is aggressive progression or severe adverse effects, i.e. gastric irritation, peptic ulcer, hepatic toxicity, renal impairment or platelet dysfunction.

If arthritis is not adequately controlled

**ADD**

- methotrexate, oral, 0.3 mg/kg/week as a single dose on an empty stomach. Specialist initiated.
  Increase at monthly intervals up to 1 mg/kg/week until there is satisfactory response.
  Maximum dose: 25 mg/week.
  Adverse effects include: nausea, mood changes, raised liver enzymes, bone marrow toxicity and protein/haematuria.
  Monitor FBC, LFT, U&E and urine test strips monthly.

**PLUS**

- folic acid, oral, 5 mg daily for the duration of the treatment

If arthritis still not controlled and to control acute flare

**ADD**

- prednisone, oral, 2 mg/kg as a single daily dose for 1–2 weeks. Specialist initiated.
  Continue with 0.3–0.5 mg/kg/day as single dose.
  Try to wean off over next 3 months.

**Disease modifying drugs**

All patients requiring disease-modifying drugs must be referred to specialist rheumatologists.

**REFERRAL**

- all for confirmation of diagnosis
- patients with pauciarticular disease for slit lamp examination, if not locally available
- patients with iridocyclitis
- all with complicated JRA or uncontrolled disease
- adverse reaction to NSAID
- for orthopaedic or orthodontic treatment