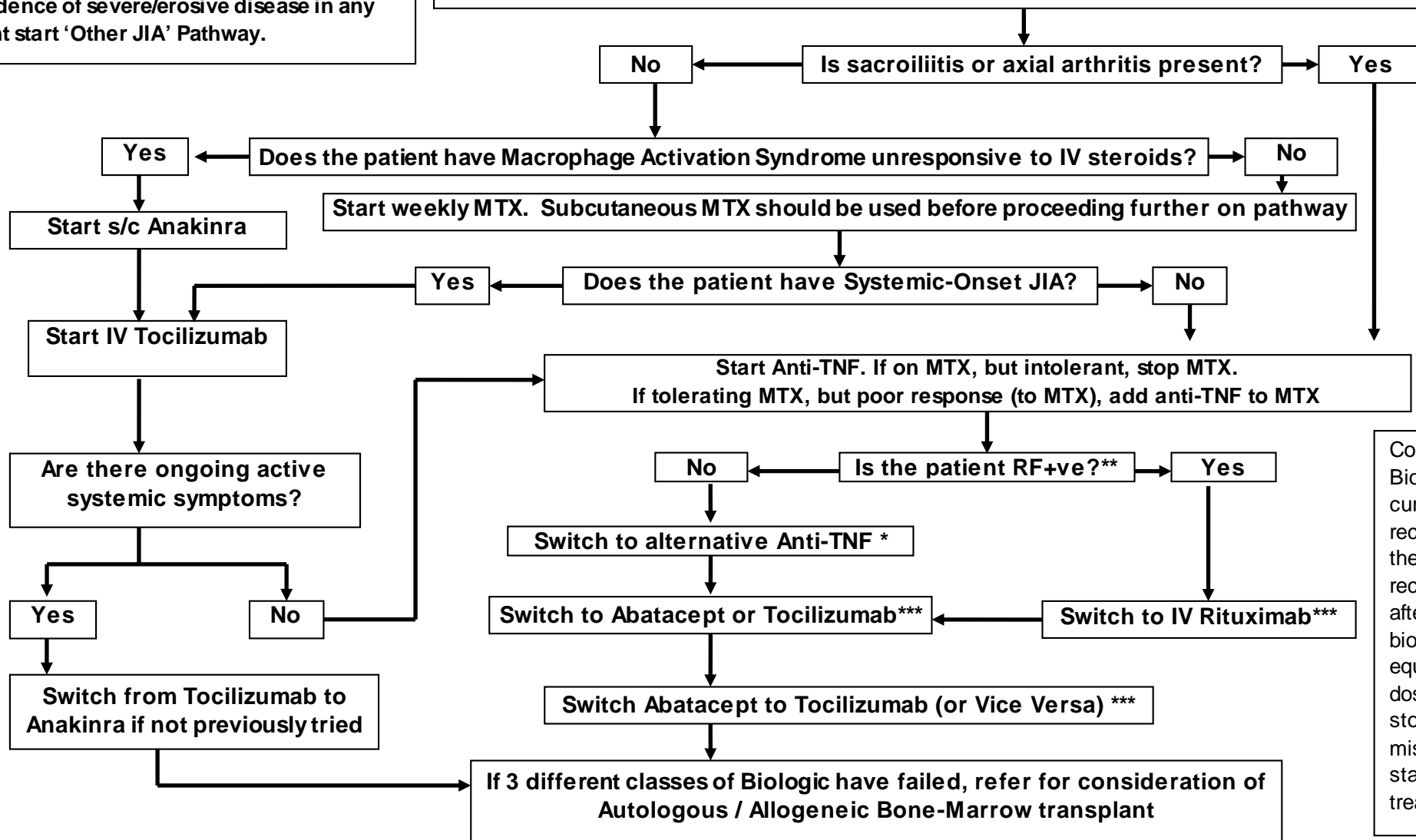


## Appendix A: Suggested Treatment Flow-chart for JIA

**Oligo-Articular JIA:** All Patients should receive intra-articular steroids to all affected joints.

Re-assess 3-6 monthly and re-inject as needed. If extension of arthritis to >4 joints, more than 2 injections within 12 months, or evidence of severe/erosive disease in any joint start 'Other JIA' Pathway.

**Other JIA:** At diagnosis all patients should receive either intra-articular steroids to all affected joints, or systemic (preferably IV) corticosteroids. Apart from the two specific circumstances of Macrophage Activation Syndrome or Sacroiliitis (see main document page 5) all patients should start treatment with Methotrexate (MTX) as the first line DMARD and be re-assessed every 3/12. 'Flares' of arthritis should be treated with steroids as above, but if >2 joints are affected, or the patient is intolerant of the treatment, they should proceed to the next step of the pathway. Consider access to clinical trials or research studies at all treatment decision points. All patients on biologics should be counselled to consent to be enrolled into Registries.



Co-administration of Biologics is not currently recommended. It is therefore recommended that after stopping any biologic therapy the equivalent of 2 doses of the drug stopped should be missed before starting the new treatment

## **Notes on the Pathway:**

- \*- Etanercept is the usual first choice for JIA. It is licensed and approved by NICE. However there is increasing anecdotal evidence that it should not be used in children with Chronic Anterior Uveitis as it has been associated with severe, and even sight-threatening worsening of uveitis. In those circumstances Adalimumab would be the usual first-choice as it has the best evidence for efficacy in uveitis as well as JIA and is currently the focus of a multi-centre trial. Infliximab also has some evidence of effectiveness against uveitis, but has a higher risk of infusion-related reactions and is more expensive. It would normally be considered as an alternative to either Etanercept or Adalimumab only when adherence to Etanercept or Adalimumab is suspected to be poor or when treatment with Adalimumab has not proven efficacious in a child with Uveitis and Arthritis.
- \*\* - NICE guidance in adults with Rheumatoid Arthritis, of which RhF +ve Poly-Articular JIA is usually considered to be analogous, suggests Rituximab should be used after failure of any anti-TNF treatment. Usual practice in other forms of JIA would be a trial of an alternative Anti-TNF before proceeding further along the pathway.
- \*\*\* - Abatacept and Tocilizumab both have good evidence to support their use in Poly-articular forms of JIA which have failed anti-TNF therapy but there is no head to head comparative trial to assess effectiveness. Tocilizumab appears to have a faster onset of action than Abatacept and therefore may be preferred in cases where arthritis is widespread and severe. However Tocilizumab has been associated with liver function abnormalities, and appears in clinical practice to have a greater risk of infections being under-recognised by medical staff due to its effect on suppressing both the symptoms of fever and measurement of acute phase reactants. The choice of which to use should therefore be at the discretion of the paediatric rheumatologist. A patient should fail 3 different classes of biologic before being referred for autologous stem cell rescue or bone-marrow transplantation.

## **Usual doses used in Paediatric Rheumatology**

1. Methotrexate – oral or s/c:  $10\text{-}15\text{mg}/\text{m}^2$  given once a week.
  - a. Maximum dose usually 25mg
2. Etanercept – s/c only:  $0.4\text{mg}/\text{kg}$  given twice a week, or  $0.8\text{mg}/\text{kg}$  given once a week.
  - a. Maximum dose usually 50mg/week
3. Adalimumab – s/c only:  $24\text{mg}/\text{m}^2$  given once every 2 weeks.
  - a. Maximum dose 40mg
4. Infliximab – IV only:  $6\text{mg}/\text{kg}$  given 4-8 weekly depending on response.
  - a. Maximum dose  $10\text{mg}/\text{kg}$  given 3-weekly

5. Abatacept – IV Only: 10mg/kg given every 4weeks
  - a. Maximum dose 1000mg
6. Tocilizumab – IV only (s/c trial in children under way): 8-12mg/kg IV every 2-4 weeks
  - a. Maximum dose – under 30kg body weight – 12mg/kg 2-weekly, over 30kg 8mg/kg 4-weekly
7. Anakinra – s/c only: 1-2mg/kg once a day
  - a. Maximum dose 100mg od
8. Rituximab – IV only: 750mg/m<sup>2</sup> given twice, 2-weeks apart
  - a. Maximum dose - 1000mg/ dose