PAEDIATRIC HOSPITAL LEVEL ESSENTIAL MEDICINES LIST
CHAPTER 12: RHEUMATOLOGY AND VASCULITIDES
NEMLC 20 OCTOBER 2022
Following comment – for final ratification

MEDICINE AMENDMENTS

SECTION	MEDICINE	ADDED/DELETED/NOT ADDED
12.2 Juvenile Idiopathic Arthritis	Methylprednisolone	Maximum dose added
	Prednisone	Retained for use in discussion with rheumatologist
12.5 Takayasu Arteritis	Prednisone	Maximum dose added

12.1 IMMUNOGLOBULIN A VASCULITIS (PREVIOUSLY HENOCH SCHÖNLEIN PURPURA)

An external stakeholder proposed that the referral criteria be updated for clarity.

The text was updated as follows:

HSP with complications, i.e. in patients with:
» Persistent proteinuria, persistent ~~macroscopic~~ microscopic haematuria, hypertension or ~~progressive nephritic syndrome~~ worsening renal function for age (renal biopsy indicated).
» Persistent abdominal pain.

12.2 JUVENILE IDIOPATHIC ARTHRITIS (JIA)

Methylprednisolone IV*: maximum dose added*

An external commenter proposed the addition of a maximum dose for the methylprednisolone. The text was amended as follows:

- Methylprednisolone, IV, 30 mg/kg/day (maximum 1 gram) for 3 days.

This maximum dose is in line with the British National Formulary for Children 2020/2021 (BNFc) recommendation for this indication. [1]

Prednisone: *retained for use in discussion with rheumatologist*
An external comment was received recommending that the management ofsevere of juvenile idiopathic arthritis with prednisone be removed, as early referral is needed for these children for consideration of DMARD therapy. It was however noted that these patients are often referred late, or there is a waiting period for an appointment. It was thus recommended that the prednisone recommendation and dose be retained, and recommended for use in discussion with a rheumatologist.

[1] BNF for Children 2020/2021. September 2020-2021. BMJ Group and Pharmaceutical Press.

The text was updated as follows:

If severe disease:

- Prednisone, oral, 1–2 mg/kg as a single daily dose for 2 weeks and wean over 2 weeks (in discussion with rheumatologist).
    - Refer all children early for consideration of a DMARD therapy.

12.5 TAKAYASU ARTERITIS

Prednisone: *maximum dose added*

An external commenter proposed the addition of a maximum dose for the prednisone. The text was amended as follows:

- Prednisone, oral, 2 mg/kg/day (maximum 60mg) for maximum of 4 weeks.

This maximum dose is in line with BNFc 2020/2021 recommendation for this indication. [1]

PAEDIATRIC HOSPITAL LEVEL ESSENTIAL MEDICINES LIST
CHAPTER 12: RHEUMATOLOGY AND VASCULITIDES
NEMLC 23 JUNE 2022

MEDICINE AMENDMENTS

SECTION	MEDICINE	ADDED/DELETED/NOT ADDED
12.1 Immunoglobulin A vasculitis	Paracetamol	Added
12.2 Juvenile Idiopathic Arthritis	Methotrexate	Dose titration removed
Management of a flare	Ibuprofen	Added
	Prednisone	Added
12.4 Systemic Lupus Erythematosus	Vitamin D and calcium supplementation	Moved from General and Supportive Measures to Medicine Treatment
	Chloroquine	Dosing amended
	Azathioprine	Dose amended

12.1 IMMUNOGLOBULIN A VASCULITIS (PREVIOUSLY HENOCH SCHÖNLEIN PURPURA)

Disorder name change
The disorder name was amended to Immunoglobulin A vasculitis (previously Henoch Schönlein Purpura.

Diagnostic criteria
This was updated in line with the 2010 EULAR/PReS/PRINTO criteria.[2]

Medicine Treatment
Paracetamol, oral: added

Previously only oral ibuprofen was included for arthritis, oedema, fever and malaise. This section was updated with the addition of oral paracetamol which is in line with other areas of the STGs for symptom management.

12.2 JUVENILE IDIOPATHIC ARTHRITIS (JIA)

Methotrexate: dose titration removed

The guidance on methotrexate titrating monthly was removed as dose increases should be only if there is poor response.

The text was amended as follows:

[2] Paediatric Rheumatology International Trials Organisation. EULAR/PRINTO/PRES criteria for Henoch- Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhool Takayasu arteritis: Ankara 2008. Part II: Final classification. Ann Rheum Dis. 2010, 69: 798-806.

- Methotrexate, oral, 10-15 mg/m^2/week as a single dose on an empty stomach. Specialist initiated.
  - ~~Increase dose at monthly intervals up to 1 mg/kg/week until there is satisfactory response, continue maintenance at the same dose.~~

Management of a flare of disease

Ibuprofen: Added

Prednisone: added

12.3 KAWASAKI DISEASE/MUCOCUTANEOUS LYMPH NODE SYNDROME

The following note was added to ensure this is considered.

Important: MIS-C, a complication of SARS-CoV-2, can mimic Kawasaki Disease

12.4 SYSTEMIC LYPUS ERYTHEMATOSUS

2012 SLICC criteria added:

The text was added as follows:

SLICC CLASSIFICATION FOR SLE
Requirements: 4>/ criteria (at least 1 clinical and 1 laboratory criteria)
OR biopsy-proven lupus nephritis with positive ANA or Anti-DNA

CLINICAL CRITERIA	IMMUNONOLOGIC CRITERIA
1. Acute Cutaneous Lupus 2. Chronic Cutaneous Lupus 3. Oral or nasal ulcers 4. Non-scarring alopecia 5. Arthritis 6. Serositis 7. Renal 8. Neurologic 9. Hemolytic anemia 10. Leukopenia 11. Thrombocytopenia (<100,000/mm^3	1. ANA 2. Anti-DNA 3. Anti-Sm 4. Antiphospholipid Ab 5. Low complement (C3, C4, CH50) 6. Direct Coombs' test (do not count in the presence of hemolytic anemia)

Vit D and calcium supplementation: Moved from General and Supportive Measures to Medicine Treatment.

Chloroquine: dosing amended

The chloroquine dosing was amended to a daily dose Monday to Friday, and a maximum dose of 200mg; to align with the available 200mg formulation

The text was amended as follows:

Chloroquine (as base), oral, 5 mg/kg/dose ~~once~~ daily, Monday to Friday.
- Maximum dose: ~~150~~ 200 mg.
- 6-monthly eye examination necessary.

Azathioprine: 1-2.5mg/kg/day, not 2-3 mg/kg/dose

Azathioprine dose aligned with the South African Medicine Formulary[3] and the British National Formulary.[4]

The text was amended as follows:

• Azathioprine, oral, ~~2-3~~ 1 – 2.5 mg/kg/dose as single daily dose.

12.5 TAKAYASU ARTERITIS

Diagnostic criteria
This was updated in line with the 2010 EULAR/PReS/PRINTO criteria.[5]

[3] Division of Pharmacology, Faculty of Health Sciences, University of Cape Town and Health and Medical Publishing group. South African Medicines Formulary, 12th Edition. 2016.
[4] BNF for Children, 2020-2021. BMJ Group and Pharmaceutical Press
[5] Paediatric Rheumatology International Trials Organisation. EULAR/PRINTO/PRES criteria for Henoch- Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhool Takayasu arteritis: Ankara 2008. Part II: Final classification. Ann Rheum Dis. 2010, 69: 798-806.