bpacnz PRIMARY CARE UPDATE SERIES

SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)
– when the immune system misidentifies its target

Key practice points:

- SLE is a chronic antibody-driven autoimmune condition that can affect organs and tissues throughout the body; there is no single discernible cause
- Diagnosing SLE can be difficult due to its wide-ranging effects and non-specific features that overlap with other conditions; identifying this condition relies heavily on excluding other more likely causes:
  - Consider SLE as a possibility in patients with persistent cutaneous features and arthritis lasting >6–8 weeks that cannot be sufficiently explained by alternative diagnoses
  - A range of different cutaneous presentations can occur in patients with SLE (often triggered by sun exposure), however, the most common and distinctive type is the "butterfly rash" which is a red or purple malar rash with well-defined margins, typically on the nose/cheeks with nasolabial sparing
  - Clinical suspicion of SLE should be increased further if the patient fits within the expected demographic, i.e. an Asian female of reproductive years, or if any additional systemic features are present, including renal, neuropsychiatric, serosal and haematological symptoms, fatigue and fever
- If after evaluation of the patient's history and clinical features SLE is still the most likely diagnosis, ANA testing should be requested (this test is positive in >95% of patients with SLE), in addition to urinalysis, serum creatinine, electrolytes, FBC, ESR and CRP tests (if not already done)
  - The absence of ANA does not exclude SLE; further testing is required (see below)
  - Additional SLE-specific antibody testing, e.g. for anti-dsDNA or anti-Sm antibodies, and complement level analysis is often performed by laboratories at the same time as ANA testing; the presence of these antibodies, or low complement levels, should add to clinical suspicion
- Following ANA testing, any patient for whom there is a high clinical suspicion of SLE should be referred to a rheumatologist and initiated on a NSAID while awaiting their appointment
  - Once a diagnosis is confirmed, the rheumatologist will guide the initial pharmacological management depending on the patient's specific pattern of symptoms and signs; this often involves hydroxychloroquine and/or NSAIDs, and corticosteroids (topical or oral) may be considered in some cases.
  - For patients with severe presentations or refractory SLE, additional immunosuppressive medicines or biologics may also be used
- Subsequent monitoring can then predominantly occur in primary care; if patients experience flares, these can usually be managed by increasing the dose of existing medicines (or adding new ones such as immunosuppressive medicines, as advised by rheumatology)
- Females with SLE have higher risk pregnancies and should ideally have a rheumatology review prior to planning a pregnancy (or as soon as possible if they unexpectedly become pregnant)
- In the long-term, the patient's prognosis usually depends on the manner with which they present, e.g. patients who present with mild and localised cutaneous SLE without organ involvement have a better prognosis than those who initially have cerebral or kidney involvement

Abbreviations: anti-dsDNA, anti-double stranded DNA; anti-Sm, anti-Smith; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; FBC, full blood count; NSAID, non-steroidal anti-inflammatory drug.