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Hennoch Schönlein Purpura: Diagnosis and Management

Aim

The aim of the guideline is to provide guidance to safely diagnose and initiate management of HSP, and to provide a follow up plan for patients.

Definition of Terms

Purpura: a palpable bruise usually 3-10mm in diameter often due to a vasculitic process.

Introduction

Henoch-Schönlein Purpura (HSP) is the commonest small vessel vasculitis of childhood with an estimated incidence of 14–20.4/100 000.
The vasculitis (IgA-dominant immune deposits) affects small vessels and typically involves skin, gut and glomeruli and is associated with arthralgias or arthritis.
HSP typically presents with the triad of:

- purpuric rash on the extensor surfaces of limbs (mainly lower) and buttocks,
- joint pain/swelling in areas involved by rash and
- abdominal pain.

Abdominal pain or arthralgia sometimes may precede the rash.

The cause is unknown but there may be a recent history of an upper respiratory tract infection.

- This guideline is intended for Emergency department and medical staff.
- Commonest age group is 2-8yrs old but guideline suitable for use up to 16yrs old.
- Patient groups specifically excluded from guideline are: patients with platelet function disorders/ low platelet counts, patients with signs of evolving sepsis.

Assessment

Patient History

Often there is a history of a preceding viral URTI. Presentation is commonest in Spring/ Autumn. Group A Strep may be a trigger.

Physical

- Purpura: - usually symmetrical affecting lower limbs or buttocks first but may affect other parts of body (less commonly).
- Joint Pain: Swelling and arthralgia of large joints are often the patient's main complaint. In most situations this pain resolves spontaneously within 24-48 hours.
- Abdominal pain: Uncomplicated abdominal pain often resolves spontaneously within 72 hours. However serious abdominal complications may occur including intussusception (usually ileo-ileal), bloody stools, haematemesis, spontaneous bowel perforation, and pancreatitis.
- Renal disease: Renal involvement affects 60-70% of patients. Haematuria is present in up to 90%, but only 5% are persistent or recurrent. Less common renal manifestations include proteinuria, nephrotic syndrome, isolated hypertension, renal insufficiency and

renal failure (<1%). Renal involvement may only present during the convalescent period. Ureteric mucosal lesions are rarely seen but can cause renal colic, degree of Ureteric obstruction

- Subcutaneous oedema (scrotum, scalp, hands, feet, sacrum): This can be very painful.
- Rare complications - pulmonary and CNS involvement

Note: Clinical presentation is not reliable at predicting outcome. Outcome is excellent for the majority of patients.

Management

Investigations

Perform BP, urinalysis, check FBC and renal and liver function (including albumin).

Acute

- Discuss with nephrologist if:
  - Hypertension
  - Abnormal renal function
  - Nephrotic syndrome (oedema, low albumin, proteinuria)
  - Acute nephritis (Haematuria, proteinuria, oedema, hypertension, oliguria)
- surgical referral if abdominal/ testicular pain is dominant at presentation
- analgesia for joint pain (check degree of renal involvement before deciding on NSAID use)
- consider admission for bed rest if there is significant joint swelling/pain
- Steroids at a dose of 1mg per kg may be considered for patients who have predominant joint or abdominal pain at presentation- if you are considering commencing steroids, consider admission.
- Steroids are no longer routinely recommended for HSP (RCT Dudley et al 2013).
- Hypertension in the absence of renal involvement suggests CNS vasculitis and needs urgent assessment

Ongoing management

- Weekly BP and urine dipstick (early am urine) check for 1 month at GP
- BP and urine check then each 4 to 6 weeks for 6 months (GP also appropriate for this follow up)
- Some sources recommend yearly BP and urine check for life where there has been significant renal involvement at outset
- Nephrology referral if:
  - Hypertension
  - Persistent frank haematuria
  - Persistent/worsening proteinuria for >1 month

Follow up

- The required frequency of follow up review has not been defined.
- Follow up monthly for 6 months is reasonable as renal disease can present late, but is unlikely to develop after 6 months
- Follow up checks may be performed by the family doctor in the absence of problems at presentation such as significant renal involvement (i.e more than mild transient haematuria).

- Should symptoms evolve or should the patient develop renal involvement the GP should have a clear referral plan for the patient. This may be to the local paediatric ED to facilitate referral to renal services

Special Considerations

Recurrence

Rarely a recurrent form of HSP may occur. Patients typically present with recurrent purpuric rash a few months to greater than a year after the initial episode. The lower limbs are usually affected. Such patients require BP and urine check and careful consideration of differentials including meningococcal disease. There are no clinical or lab markers indicative of the likelihood of recurrent disease. Annual follow up review is suggested.

Renal disease

Long term prognosis is determined predominantly by the extent of renal involvement. Severe early renal involvement is more likely to be associated with a poor outcome.

Companion Documents

Link to References

Link to Parent Information Leaflet

Link to Literature Search Strategy

Link to GP Letter template