

PAEDIATRIC ACUTE CARE GUIDELINE

Henoch-Schonlein Purpura		
Scope (Staff):	All Emergency Department Clinicians	

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Henoch-Schonlein Purpura

Emergency Department

Background

Scope (Area):

- **Henoch-Schönlein Purpura** (HSP) is the most common cause of non-thrombocytopenic purpura in children.
- The cause is unknown, but it is an IgA-mediated vasculitis of small vessels, usually following an upper respiratory tract infection (URTI).
- It is more common in children than in adults, with a peak incidence in the 2-8 year age group, and a seasonal peak in winter.
- The onset of the illness may be acute, with simultaneous involvement of several organ groups, or insidious, with sequential occurrence of features over weeks to months.
- It is usually a self-limiting disease.
- Many children will need only a urine test and can be sent home with GP follow up.

Assessment

Clinical Features

- The child is generally well-looking
- There is often a history of a recent URTI
- Small-vessel vasculitis may involve several organ groups, in particular the skin, joints, gastrointestinal tract and kidneys
- Rarely, there may be CNS or pulmonary involvement

Rash	 Initially blanching pink maculopapules which may be discrete or confluent Progresses to petechiae or purpura, which are often raised ("palpable purpura") The distribution of the purpura typically involves gravity-dependent body areas, particularly the buttocks, legs and extensor surfaces of the arms.
Oedema	 Vasculitis of dermal vessels also results in angio-oedema Non-pitting, often painful, oedema of dependent areas (hands and feet) as well as the eyelids, lips or scrotum.
Arthritis	 Occurs in two-thirds of patients Self-limiting serous joint effusions which resolve over several days Typically involving the gravity-dependent joints (elbows, wrists, knees, ankles) – swollen, painful joints; may cause difficulty in weight-bearing
GIT	 Abdominal pain is common, and is generally intermittent and colicky in nature Diarrhoea (with occult blood) is common Occasionally, frank haematemesis or melaena may occur More serious, but infrequent, complications include spontaneous bowel perforation or intussusception
Renal	 90% of patients have microscopic haematuria, but this is persistent or recurrent in only 5% of cases Serious complications may include acute glomerulonephritis, nephrotic syndrome, acute renal failure (<1%), or isolated hypertension Renal involvement may be a late manifestation, up to 6 months after initial presentation.

Differential Diagnosis

- Meningococcal septicaemia
- Thrombocytopenia
- Other vasculitides
- Patients initially presenting with only with abdominal pain or arthralgia may pose a diagnostic challenge
- Patients initially presenting with haematuria only should have other causes of haematuria excluded

Investigations

• There is no diagnostic investigation for HSP. Investigations are used to detect complications or other causes of purpura

Urine

- Microscopic haematuria is common (90%)
- If macroscopic haematuria is present, urine should be checked for RBC casts (nephritis) and protein (nephrotic syndrome)

Consider

- FBC to exclude thromocytopaenia as cause of purpura if diagnosis is uncertain
- Blood culture, white cell count, CRP, meningococcal PCR if meningococcal septicaemia is suspected
- Urea, creatinine and electrolytes if renal impairment is suspected
- Complement levels and ASO-titres if nephritis is present

Management

Most patients with HSP will be managed as an outpatient with symptomatic treatment (analgesia) and follow up.

- Document BP
- Surgical consult for abdominal complications or testicular pain (testicular pain from vasculitis may be difficult to differentiate from testicular torsion)
- Abdominal X-Ray or ultrasound may be needed to exclude abdominal complications
- There is some evidence to suggest that prednisolone 1 mg/kg/day for 2 weeks benefits in abdominal and joint pain (and oedema)
 - There is very limited evidence in preventing renal complications
 - $\circ\,$ The decision whether to start prednisolone should be made by the admitting Paediatric Consultant

Admission criteria

- Abdominal complications (intussusception, perforation, haematemesis or bloody stools)
- Renal complications (nephritis, nephrotic syndrome, hypertension, renal failure)
- Symptomatic relief (severe joint pain or abdominal pain, painful oedema)

Referrals and follow-up

- Patients should receive follow up with their GP or Paediatrician for at least 6 months to ensure symptom relief and disease resolution
- Check BP and urinalysis weekly for one month, then monthly for 6 months after presentation as renal disease may present late
 - If still present at 6 months refer to the Renal team
- Provide <u>Henoch-Schonlein Purpura Health Facts</u>

Nursing

Routine nursing care.

Tags

abdominal, angio oedema, arthritis, blanching, haematuria, henoch schonlein purpura, hsp, maculopapules, oedema, petechiae, purpura, rash

References 1. Dudley J, Smith G, Llewelyn-Edwards A, Bayliss K, Pike K and Tizard J. Randomised, double-blind, placebo-controlled trial to determine whether steroids reduce the incidence and severity of nephropathy in Henoch-Schonlein Purpura (HSP). Archives of Disease in Childhood 2013, July. Accessed online: http://adc.bmj.com/content/early/2013/07/10/archdischild-2013-303642.abstract 2. Shin JI and Lee JS. Steroids in Henoch-Schonlein purpura and abdominal pain. Archives of Disease in Childhood. 2006 Aug; 91(8):714. Accessed online: http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2083041/ 3. Weiss PF, Klink AJ, Localio R, Hall M, Hexam K, Burnham JM, Keren R and Feudtner C. Corticosteroids may improve clinical outcomes during hospitalisation for Henoch-Schonlein purpura. Paediatrics. 2010 Oct, 126 (4): 627-81. Accessed online: http://www.ncbi.nlm.nih.gov/pubmed/20855386

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