

Henoch-Schonlein Purpura (HSP)

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****Please print off the renal follow-up guide and parent hand-held record and give to parents on discharge****

Background

HSP is the most common small vessel vasculitis of childhood. It is IgA mediated and has multi-system involvement.

HSP is more common in young children with over 50% under 5 years and over 75% under 10 years.

The aetiology is unclear but in many cases there appears to be an infectious trigger, in particular there may be a history of a recent upper respiratory tract infection. However, other pathogens have been implicated as triggers along with some cases being described following vaccinations & some drugs.

HSP is self-limiting in most cases. Although most symptoms resolve renal disease may be associated with long term complications. A third of patients may relapse.

Classification

The diagnosis of HSP is based on the European League Against Rheumatism (EULAR) and Paediatric Rheumatology European Society (PReS) consensus criteria for the classification of childhood vasculitis.

HSP should be diagnosed in the face of **palpable purpura** & the **presence of at least one of the following four features**:

- Diffuse abdominal pain
- Arthritis or arthralgia
- Renal involvement
- Any biopsy showing predominant IgA deposition

Assessment

Clinical Features

HSP most commonly affects the skin, joints, gastrointestinal tract & kidneys. Clinical features are often atypical at the extremes of age. Children under the age of 2 years are less likely to develop nephritis or abdominal complications.

Skin

The characteristic rash is diagnostic – typically palpable purpura symmetrically distributed over the extensor dependent surfaces of the lower limbs & buttocks.

It may affect the arms and face but usually spares the trunk.

Purpura may be preceded by urticarial or erythematous, maculopapular lesions.

Subcutaneous oedema occurs

Gastrointestinal involvement (occurs in 50 – 70% of cases)

Abdominal pain – if uncomplicated resolves spontaneously in 72 hours

Serious abdominal complications include:

- Intestinal bleeding
- Intussusception
- Protein losing enteropathy
- Pancreatitis

Joints

Up to 80% have joint involvement at some point during the illness. Arthritis/arthralgia can be the presenting symptom.

Arthritis typically affects the large joints of the lower limbs.

Renal

Renal disease manifests as: haematuria, proteinuria, nephritic syndrome/nephritis, renal impairment & hypertension.

May only present during convalescent period

Hypertension can develop without evidence of renal involvement.

Urological

Orchitis

Testicular torsion

Rare Complications

Pulmonary and Neurological involvement

Differential Diagnosis

If the purpura has an atypical distribution or if a child appears systemically unwell consider:

- Meningococemia
- Thrombocytopenia
- Other vasculitides – including microscopic polyarteritis and Wegner's granulomatosis

Management

Investigations

The main aims of the investigations are to exclude any other potential differential diagnoses and to assess the extent of the renal involvement.

Investigations for all
➤ FBC
➤ Clotting
➤ U&Es / LFTs / Bone Profile
➤ ESR + CRP
➤ Blood Pressure
➤ Height (needed to work out normal BP range)
➤ Weight
➤ Urinalysis – if proteinuria ($\geq 1+$) is identified the urine should be sent for urine protein:creatinine ratio

If systemically unwell, evidence for a site of infection should be sought

- Blood cultures
- Urine microscopy + culture
- Viral serology
- Throat swab

In the presence of renal involvement (haematuria ± proteinuria or hypertension)

- ASOT + antiDNase B titres

If the diagnosis is not clear or there is severe renal impairment (hypertension, acute renal impairment or low albumin) from the outset a full autoimmune screen should be done including ANA, dsDNA, ANCA, Immunoglobulins & C3, C4.

When to Admit

Few cases require admission but it may be necessary if:

- Significant joint pain
- Severe Abdominal pain
- GI haemorrhage
- Neurological symptoms
- Hypertension
- Evidence of acute glomerulonephritis
 - Nephrotic syndrome
 - Abnormal renal function

System specific management

Joint pain: Simple analgesia including non-steroidal anti-inflammatory drugs but caution with NSAIDs if renal impairment or hypertension.

Gastrointestinal: Non-specific abdominal pain – simple analgesia
 Severe symptoms – early steroids may prevent complications.
 Prednisolone 1mg/kg daily (max. 50mg) for 2 weeks & then wean over 2 weeks.
 Protein losing enteropathy or GI bleed – consider Methylprednisolone

If any concerns about abdominal pain or possible testicular involvement, refer to the surgeons.

Renal: **The long term outcome is related to the presence of renal disease**

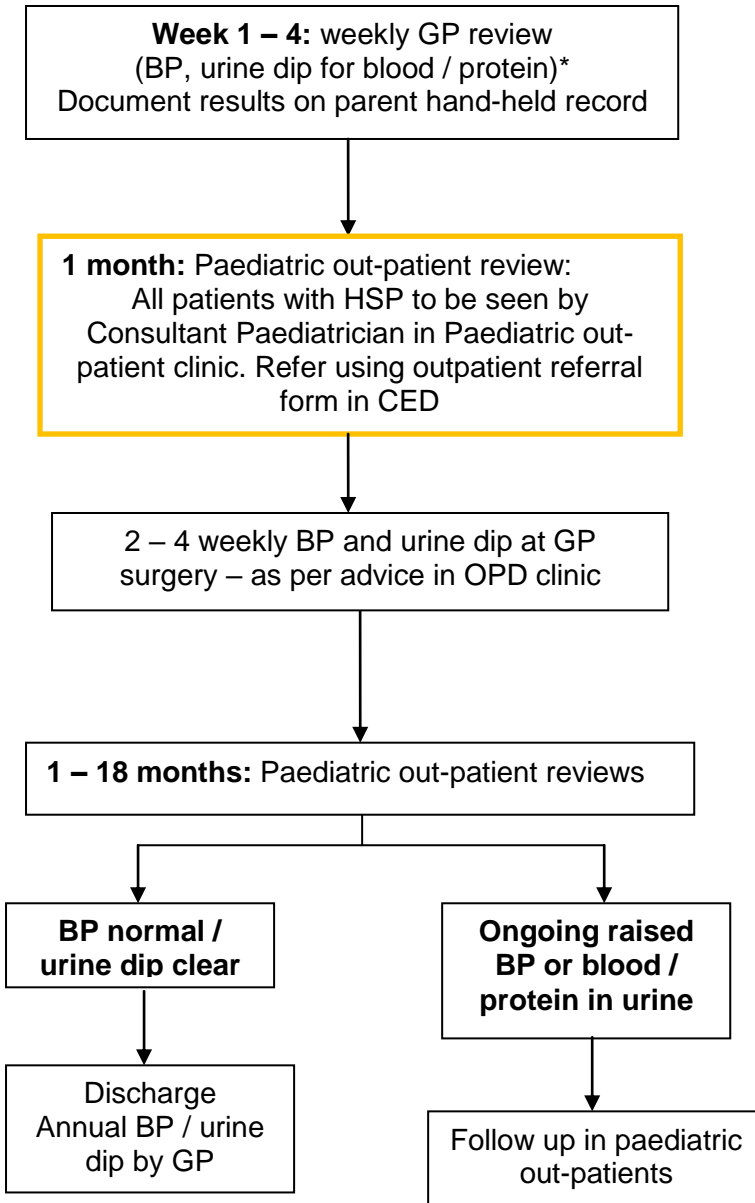
NB. Can develop long term renal impairment & hypertension and the renal features may only become apparent during the convalescent phase.

See flow chart for monitoring

Ensure the GP is aware of the child's target blood pressure

See next page for renal follow-up and parent hand-held renal results monitoring – print both and give to parent

GP and Outpatient follow up:



If BP high

- Re-check after 15 minutes (ensure cuff is best fit, child calm etc.)
- may need to arrange for repeat measurement in 2 – 3 days
- If concerned – contact Children's Emergency Department at RACH for advice

If blood in urine – document on parent-held record

If protein in urine – send for protein:creatinine ratio

If hypertensive or macroscopic haematuria / proteinuria ($\geq 2+$) or any atypical features on initial presentation in CED – arrange a review on Day Case Unit within a few days of presentation

Day case unit review to include:

Baseline:

- Weight, height, BP, urine dipstick
- Urine protein:creatinine
- U&E, creatinine, albumin
- FBC, clotting
- ASOT, antiDNase B (if not done initially)

Consider:

- C3, C4, AIP, ANCA, Igs (refer to main guideline)

Pending results or if necessary weekly review:

- Urine protein:creatinine
- BP + weight monitoring
- Clinical assessment

If normal – GP follow up to resume

Discuss with consultant of the week if:

- Hypertension
- Macroscopic haematuria – 5 days
- Nephrotic syndrome
- Acute nephritic syndrome

Or if persisting proteinuria with urine protein:creatinine:

- >250 mg/mmol for 4 weeks
- 100 – 250 mg/mmol for 3 months
- 50 – 90 mg/mmol for 6 months

Discussion with the Paediatric Nephrologists may be appropriate

***If found to be hypertensive or have macroscopic haematuria / proteinurea ($\geq 2+$) at any time in the community, please contact the Children's Emergency Department at RACH for advice.**

