East and North Hertfordshire NHS Trust

Department Guideline

For

Management of Henoch-Schonlein purpura in Children and Young People under 16 years

A Policy recommended for use

In: All areas of Children's Services

By: All Medical and Nursing staff

For: Children with Henoch-Schonlein Purpura

Key Words: Children ,vasculitis , rash , Joint and renal involvement , GIT symptoms , purpuric rash on extensor surface , follow up , petechiae

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Trust Ratification:

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Version	Date	Comment
1	2011	Guideline Developed
2	January 2016	Overdue review
3	September 2019	Reviewed

Equality Impact Assessment
This document has been reviewed in line with the Trust's Equality Impact Assessment guidance and no detriment was identified. This policy applies to all regardless of protected characteristic - age, sex, disability, gender-re-assignment, race, religion/belief, sexual orientation, marriage/civil partnership and pregnancy and maternity.

Dissemination and Access
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Associated Documentation
None

Review
This document will be reviewed within three years of issue, or sooner in light of new evidence.

Key messages

- Henoch-Schonlein Purpura (HSP) is a disease of exclusion
- Characteristic purpuric rash appears in 100% of cases
- Joint involvement is in 60-80% of patients
- Treatment of HSP is mainly supportive but children, especially with renal involvement need proper follow up for at least 6 months.

1. INTRODUCTION

Henoch-Schonlein purpura (HSP) is a disease involving inflammation of small blood vessels. This document outlines the presentation, diagnosis, management and monitoring of these children.

2. SCOPE

For all children under 16 years diagnosed with HSP.

3. PURPOSE

This document will ensure that children with HSP have the necessary monitoring and follow-up arranged in a timely manner. Also ensures that parents receive standard evidence based advice.

4. DEFINITIONS

- A generalized allergic vasculitis (IgA mediated vasculitis) of small vessels of the skin, gastrointestinal (GI) tract, kidneys, joints, and, rarely, the lungs and central nervous system (CNS)
- Described by 2 German physicians:- Johan Schönlein in 1837 described purpura with arthritis; 30 years later, Edouard Henoch pointed out GI manifestations.

5. DUTIES

Medical and nursing teams are expected to consider these guidelines to provide the most appropriate and standardised care to these children.

6. METHOD

6.1 Etiology and Pathophysiology:

- Etiology is unclear. More than 75% of patients report previous upper respiratory or gastrointestinal infections
- Many bacteria, viruses, drugs and vaccinations have been associated
- Thought to be IgA-mediated autoimmune phenomenon with an unknown antigenic stimulant. Antigen-antibody complexes deposit throughout the body and activate pathways leading to necrotizing vasculitis

Sex: In children, M: F ratio is 2:1. In adults, M: F ratio is closer to 1:1.
Age: 75% of cases are aged 2-11 years. The median age is 5 years

6.2 Clinical Features:

Prodrome of headache, anorexia, fever is followed by a rash, abdominal pain, and/or arthritis.

Skin:

- **Rash appears in 100% of cases and is the presenting feature in 50%.**
- Lasts an average of 3 weeks.
- Consists of erythematous macules, urticarial papules, pruritic papules, and plaques.
- Cardinal feature is a palpable purpuric rash over the buttocks and extensor surfaces of the lower limbs. May involve arms, face and ears. May appear maculopapular before becoming purpuric. Skin lesions tend to appear in crops. Varying stages of eruption are usually present simultaneously.

GI:

- 50-70% of patients will have GI symptoms, including colicky abdominal pain, nausea, and vomiting.
- May be Occult blood positive stool or overt bleeding.
- Abdominal examination generally is unremarkable. Occasionally, the abdomen may be very tender. An acute abdomen is rare.
- Gastrointestinal complications include intussusception (usually ileo-ileal), bowel infarction, bowel perforation, hydrops of the gallbladder, pancreatitis, massive GI bleed & protein losing enteropathy.

Joints:

- Involved in **60-80%** and is the presenting sign in about 25%. Knees, ankles, and (less commonly) wrists most commonly involved, with pain and swelling.

Renal:

- Involved in 50% of patients - **develops within 4 weeks in 75-80% and within 3 months in 97-100%.** A few cases develop even years later.
- Involvement ranges from mild haematuria/proteinuria to nephrotic syndrome, oliguria and renal failure (5%).

- Approximately 5% of patients develop end-stage renal disease (ESRD). Patients with only haematuria do not develop ESRD. 15% of patients with haematuria and proteinuria develop ESRD. Approximately 50% of patients with nephritic/nephrotic syndrome develop ESRD.

Other Complications:

- Cardiac: Coronary artery vasculitis resulting in myocardial infarction (MI)
- CNS: Headache, CNS bleeding, subtle encephalopathy , seizures
- Other: Pulmonary haemorrhage, Orchitis which may lead to surgical exploration of the scrotum

6.3 Diagnostic features:

Mainly clinical

Palpable Rash with one of these:

- Diffuse abdominal pain
- Acute arthritis/arthralgia
- Renal involvement
- Biopsy showing predominant IgA deposition

Differential Diagnosis:

HSP should remain in the differential diagnosis of any patient presenting with abdominal pain with or without a skin rash. This is one of the reasons why urinalysis in children presenting with non-specific abdominal pain is so important.

It is important to consider other causes of non-blanching rash:

- Disseminated intravascular coagulation
- Meningococcal sepsis
- Other sepsis

Other vasculitides

- Cutaneous small vessel vasculitis
- Granulomatosis with polyangitis (formerly Wegener's)
- Systemic lupus erythematous (SLE)
- Microscopic polyarteritis
- Polyarteritis nodosa

Causes of thrombocytopaenia

- Reduced production: leukaemic, aplastic anaemia
- Increased destruction: Idiopathic thrombocytopaenic purpura, haemolytic uraemic syndrome

Hypersplenism Examination: Examine fully for joint swelling, abdominal tenderness and check BP

6.4 Investigations:

- Urine dip for blood and protein. Send urine for microscopy and casts
- Only if diagnosis not certain or to assess renal involvement. E.g. U/E's, Creatinine, FBC, clotting, LFTs.
- Additional tests rarely needed: ASO titre, Monospot, Antinuclear antibody, Rheumatoid factor, complement C3/C4 levels, blood cultures

6.5 Management:

- Treatment is largely **supportive** with monitoring of blood pressure, pulse, respiration and temperature.
- Paracetamol or Ibuprofen **(avoid if significant proteinuria or hypertension)** may reduce joint and abdominal discomfort. Refer to BNFc for doses and frequency.
- Steroids – There is no strong evidence supporting the routine use of prednisolone to reduce the long-term renal outcome in children with HSP ,
- In children with severe abdominal or joint pain not responding to simple analgesia, a course of prednisolone 1mg/kg daily for 1-2 weeks may be considered after discussion with treating consultant
- Surgical review must be considered prior to the use of steroids in HSP abdominal pain.
- Provided there is no frank haematuria, and no severe abdominal tenderness or severe joint pain, the child may be discharged with paracetamol or Ibuprofen
- If discharging, note arrangements for follow up below
- If frank haematuria is present initially, admit for observation, urine output measurement, and BP checks.
- If severe abdominal pain is present or if symptoms suggest intussusception or GI bleeding occurs, discuss with consultant, admit for observation and consider surgical assessment

6.6 Prognosis:

Fifty percent of patients have at least 1 recurrence. Younger patients (<3 y) usually have a better prognosis. Prognosis is best for patients with minimal or no renal involvement at the outset of the illness.

Detection and referral of Nephritis:

- Nephritic or Nephrotic presentation, raised creatinine, hypertension or oliguria: require close attention to fluid balance, electrolytes, and use of anti-hypertensives (if indicated) **Require urgent referral to Nephrologist**.
- If admitted, check **early morning** urine for blood and protein daily
- **Parents to bring Early Morning Urine (EMU) sample to all clinics and GP.**

Follow up Arrangements for all HSP cases

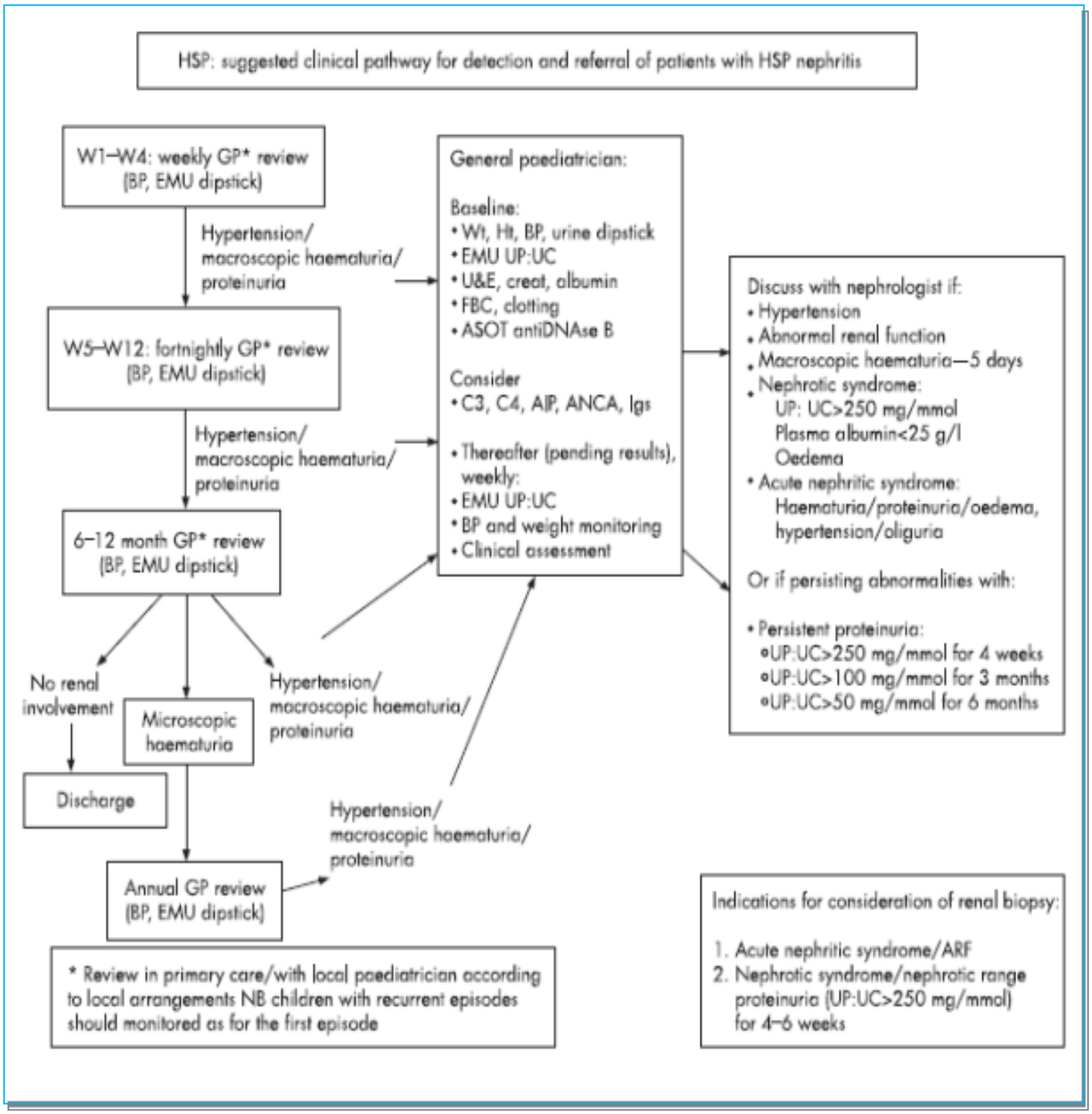
Print out 3 copies of the HSP Follow –up Guidance, one for the case notes, a copy for the parents, and one to be posted to the GP together with the discharge summary.

Give information leaflet to parents – print from KC add hyperlink

- Reg review clinic at 48-72 hours and at 2 weeks (for BP, EMU dipstick).
- GP to review weekly in weeks 1-4 (for BP, EMU dipstick), then fortnightly in weeks 5-12 (for BP, EMU dipstick).
- OPD Appointment at 3 months with Consultant. If, at that time, there has been no renal involvement, discharge to further GP follow up.
- GP should see child at 6 months (for BP, EMU dipstick) and discharge if no renal involvement.
- If at any time there is hypertension, macroscopic haematuria or proteinuria, GP should refer urgently to a paediatrician for investigations.

The Paediatrician arranges investigations detailed in algorithm and, depending on results, considers discussing with Nephrologist.

Follow algorithm below.



7. MONITORING COMPLIANCE

The specialist consultant will responsible for the monitoring of compliance with this guideline. Monitoring will be carried out via incident reporting and audit. Actions identified from monitoring will be discussed at the risk management meeting and any unresolved actions escalated to Children's Board and Risk Register.

8. REFERENCE:

- Tizard, Hamilton-Ayres. Henoch-Schonlein Purpura. Archives of disease in childhood Education and Practice; Feb 2008
- European League Against Rheumatism and Paediatric Rheumatology European Society; Annals of Rheumatic Diseases: 2006; 65: 936
- Nottingham Children's Hospital HSP Guidelines June 2016