1. **Aim/Purpose of this Guideline**

1.1. This guideline aims to give clear guidance to medical and nursing staff who are caring for children and young people with *Henoch-Schönlein Purpura* (HSP).

2. **The Guidance**

2.1. **Definition:**
Palpable purpura often in the presence of either (1) diffuse abdominal pain, (2) arthritis or arthralgia (3) renal involvement (any haematuria and/or proteinuria) (4) any biopsy showing predominant IgA deposition.

2.2. **Epidemiology:**
Present in all age groups from few months to late adulthood, but more common in younger children. (>50% in under 5 years, >75% in under 10 years)

**Incidence:** Commonest vasculitis of childhood. 10-20 per 100,000 children. Caucasians are more often affected than other ethnic groups. Male predominance (2:1).

Approx. 2/3 have a possible infective trigger.

2.3. **Clinical Features:**
Classical rash is sign that usually leads to diagnosis, but in approx. 40% the rash is preceded by abdominal pain and/or arthritis by up to 2 weeks.

**Skin** (100%)
- Typical rash is palpable purpura, often symmetrically distributed over extensor surfaces of lower limbs and buttocks.
- Purpura range in size from petechiae to large ecchymoses. May be preceded by urticarial or erythematous macular papular lesions.
- Rarely haemorrhagic bullae and vesicles, subcutaneous oedema of scalp, ears, periorbital area, dorsum of hands / feet and genitalia.

**Gastrointestinal involvement** (50-75%)
- Most commonly with colicky abdominal pain.
- May have vomiting, diarrhoea or GI haemorrhage. Massive bleeding is rare [2%]
- Intussusception (4-5%).
- Protein losing enteropathy, haemorrhagic pancreatitis are other rare complications.

**Joints** (80%)
- Arthralgia and peri-articular oedema.
- Knees, ankles most commonly affected, also elbows, wrists and fingers.
- For 15-25% arthralgia / arthritis is the presenting symptom. Does not result in permanent damage.

**Renal** (20-60%)
- Haematuria, proteinuria, nephritic or nephrotic syndrome, renal impairment or hypertension.
- Develops within 4 weeks in 75-80% and within 3 months in 97-100%
- Severe renal disease in 5-7% (acute nephritis, nephrotic syndrome, renal impairment)
- Hypertension may develop without evidence of renal disease.

Other:
- **Urological**: Orchitis in up to 24% of boys. Swollen, tender scrotum, swelling of penis is not uncommon.
- **Neurological** (rare): encephalopathy, seizures, intracranial haemorrhage, infarction.
- **Pulmonary** (rare, but more common in adults): Diffuse alveolar haemorrhage.
- **Carditis**.
- **Parotitis**.

2.4. **Differential Diagnoses**;
- Sepsis (systemically unwell)
- Systemic vasculitides (microscopic polyarteritis, Wegeners granulomatosis, SLE etc)

2.5. **Investigations**:
There are no specific tests that have been shown to be helpful in making the diagnosis of HSP, which is based on clinical features. Investigations can be useful in excluding other diagnoses and determining the extent of renal disease.

2.6. **Initial investigations**:
- Weight, height, BP.
- FBC, clotting, U&E, LFT, bone profile, CRP & ESR. Urinalysis.

2.7. **Additional investigations**
To be considered if renal disease (haematuria or proteinuria) present.
- Complement (C3 & C4)
- pANCA and cANCA
- Autoimmune profile
- ASOT titres and antiDNAse B titres
- Renal USS
To be considered if Intersusception suspected
- Abdominal ultrasound

Other:
- **Urological**: Orchitis in up to 24% of boys. Swollen, tender scrotum, swelling of penis is not uncommon.
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**Differential Diagnoses**;
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2.8. Clinical Pathway

HSP: suggested clinical pathway for detection and referral of patients with HSP nephritis

W1-W4: weekly GP* review (BP, EMU dipstick)
- Hypertension/
  macroscopic haematuria/
  proteinuria

W5-W12: fortnightly GP* review (BP, EMU dipstick)
- Hypertension/
  macroscopic haematuria/
  proteinuria

6–12 month GP* review (BP, EMU dipstick)
- No renal involvement
  Microscopic haematuria
  Discharge
- Hypertension/
  macroscopic haematuria/
  proteinuria
  Annual GP review (BP, EMU dipstick)

General paediatrician:
- Baseline:
  • W1, Ht, BP, urine dipstick
  • EMU UP:UC
  • U&E, creat, albumin
  • FBC, clotting
  • ASOT antiDNAse B
- Consider
  • C3, C4, AIP, ANCA, lgs
  • Thereafter (pending results), weekly:
    • EMU UP:UC
    • BP and weight monitoring
    • Clinical assessment

Discuss with nephrologist if:
- Hypertension
- Abnormal renal function
- Macroscopic haematuria—5 days
- Nephrotic syndrome:
  • UP: UC>250 mg/mmol
  • Plasma albumin<25 g/l
  • Oedema
- Acute nephritic syndrome:
  • Haematuria/proteinuria/oedema,
    hypertension/oliguria
  Or if persisting abnormalities with:
  • Persistent proteinuria:
    0 UP: UC>250 mg/mmol for 4 weeks
    0 UP: UC>100 mg/mmol for 3 months
    0 UP: UC>50 mg/mmol for 6 months

Indications for consideration of renal biopsy:
1. Acute nephritic syndrome/ARF
2. Nephrotic syndrome/nephrotic range
   proteinuria (UP:UC>250 mg/mmol)
   for 4–6 weeks

EMU = (Early Morning Urine)  UP:UC = (Urine protein: creatinine ratio)
2.9. Treatment:
The natural history of HSP is predominantly resolution of all symptoms except for the renal disease, which may be associated with long-term complications.

2.10. General:
1. Supportive (hydration, electrolytes, nutrition)
2. Simple analgesia. (Use NSAIDs with caution)
3. Severe abdominal pain (secondary to intussusception), arthritis, pulmonary haemorrhage - consider prednisolone 1mg/kg (after appropriate investigation) and referral to appropriate specialist.
4. For treatment of severe skin disease on the advice of a dermatologist: Dapsone 2mg/kg once daily. Maximum 100mg..

2.11. Renal:
There is no evidence that corticosteroids or cyclophosphamide is beneficial in those with renal involvement. Treatment may be beneficial however in crescentic nephritis.

2.12. Outcome:
- 33% have recurrence of symptoms.
- Long-term renal impairment in ~20% of those with nephritic or nephrotic syndrome.
- HSP nephritis accounts for 2-3% of children with chronic kidney disease.
- Renal involvement at presentation relates to likelihood of developing long term renal disease.
- Presentation of renal disease can occur several months after presentation.
- 97% of those with renal involvement have presented within 6 months.

2.13. Long-term follow up (Local Arrangement):
- Arrange for acute paediatric outpatient appointment at 6 months after diagnosis.
- If BP and urinalysis are normal at 6-12 months, they can be safely discharged.
- All with a history of HSP nephritis should have lifelong follow-up – annual BP and urinalysis.
3. Monitoring compliance and effectiveness

<table>
<thead>
<tr>
<th>Element to be monitored</th>
<th>Compliance with section 2.8 of the guideline. Compliance with initial and additional Investigations</th>
</tr>
</thead>
</table>
| Lead                    | Audit lead  
Dr. C. Williams |
| Tool                    | Notes audit review.  
Outpatient follow up. |
| Frequency               | At point of patient contact as rare occasion |
| Reporting arrangements  | Paediatric consultant  
Child Health Department audit and guidelines meeting |
| Acting on recommendations and Lead(s) | Paediatric consultant  
Department audit and guidelines meeting  
Required actions will be identified and completed within 3-6 months. |
| Change in practice and lessons to be shared | Required changes to practice will be identified and actioned within 3-6 months. A lead member of the team will be identified to take each change forward where appropriate. Lessons will be shared with all the relevant stakeholders |

4. Equality and Diversity

4.1. This document complies with the Royal Cornwall Hospitals NHS Trust service Equality and Diversity statement.

4.2. Equality Impact Assessment

The Initial Equality Impact Assessment Screening Form is at Appendix 2.
Appendix 1. Governance Information

<table>
<thead>
<tr>
<th>Document Title</th>
<th>Clinical Guideline for the management and investigation of Henoch-Schonlein Purpura (HSP) in children.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Date Issued/Approved:</td>
<td>31 January 2013</td>
</tr>
<tr>
<td>Date Valid From:</td>
<td>31 January 2013</td>
</tr>
<tr>
<td>Date Valid To:</td>
<td>1 January 2017</td>
</tr>
<tr>
<td>Directorate / Department responsible (author/owner):</td>
<td>Dr. Chris Williams Paediatric Consultant</td>
</tr>
<tr>
<td>Contact details:</td>
<td>01872252463</td>
</tr>
<tr>
<td>Brief summary of contents</td>
<td>Clear guidance for the management and investigation of Henoch-Schonlein Purpura (HSP) in children.</td>
</tr>
<tr>
<td>Suggested Keywords:</td>
<td>HSP, Children, Paediatrics</td>
</tr>
<tr>
<td>Target Audience</td>
<td>RCHT</td>
</tr>
<tr>
<td>Executive Director responsible for Policy:</td>
<td>Medical Director</td>
</tr>
<tr>
<td>Date revised:</td>
<td>December 2013</td>
</tr>
<tr>
<td>This document replaces (exact title of previous version):</td>
<td>Henoch-Schonlein Purpura</td>
</tr>
<tr>
<td>Approval route (names of committees)/consultation:</td>
<td>Paediatric consultants, Paediatric audit and guidelines meeting</td>
</tr>
<tr>
<td>Divisional Manager confirming approval processes</td>
<td>Sheena Wallace</td>
</tr>
<tr>
<td>Name and Post Title of additional signatories</td>
<td>Not Required</td>
</tr>
<tr>
<td>Signature of Executive Director giving approval</td>
<td>{Original Copy Signed}</td>
</tr>
<tr>
<td>Publication Location (refer to Policy on Policies – Approvals and Ratification):</td>
<td>Internet &amp; Intranet</td>
</tr>
<tr>
<td>Document Library Folder/Sub Folder</td>
<td>Paediatrics</td>
</tr>
<tr>
<td>Links to key external standards</td>
<td>none</td>
</tr>
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</table>
Clinical Guideline for the management and investigation of Henoch-Schonlein Purpura (HSP) in children.

Related Documents:

- Rosenblum N et al. Steroid effects on the course of abdominal pain in children with HSP. Pediatrics 1987;79:1018-21
- Huber A et al. A randomised, placebo controlled trial of prednisone in early HSP. BMC medicine 2004;2:7

Training Need Identified? No

Version Control Table

<table>
<thead>
<tr>
<th>Date</th>
<th>Version No</th>
<th>Summary of Changes</th>
<th>Changes Made by (Name and Job Title)</th>
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</thead>
<tbody>
<tr>
<td>October 2009</td>
<td>V1.0</td>
<td>Initial Issue</td>
<td>Dr. E. Morris - paediatrics</td>
</tr>
<tr>
<td>January 2014</td>
<td>V2.0</td>
<td>Review and re format</td>
<td>Dr. C. Williams – paediatric consultant Tabitha Fergus-Deputy ward manager</td>
</tr>
</tbody>
</table>

All or part of this document can be released under the Freedom of Information Act 2000

This document is to be retained for 10 years from the date of expiry.

This document is only valid on the day of printing

Controlled Document

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This page contains information regarding the management and investigation of Henoch-Schonlein Purpura (HSP) in children.
Appendix 2. Initial Equality Impact Assessment Form

<table>
<thead>
<tr>
<th>Name of the strategy / policy / proposal / service function to be assessed (hereafter referred to as policy) (Provide brief description): Clinical Guideline for the management and investigation of Henoch-Schonlein Purpura (HSP) in children.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Directorate and service area: child health Is this a new or existing Policy? existing</td>
</tr>
<tr>
<td>Name of individual completing assessment: T. Fergus Telephone: 01872252800</td>
</tr>
<tr>
<td>4. *How will you measure the outcome? audit</td>
</tr>
<tr>
<td>5. Who is intended to benefit from the policy? Children and families</td>
</tr>
<tr>
<td>6a) Is consultation required with the workforce, equality groups, local interest groups etc. around this policy? no</td>
</tr>
<tr>
<td>b) If yes, have these *groups been consulted?</td>
</tr>
<tr>
<td>C). Please list any groups who have been consulted about this procedure.</td>
</tr>
<tr>
<td>7. The Impact Please complete the following table. Are there concerns that the policy could have differential impact on:</td>
</tr>
<tr>
<td>Equality Strands:</td>
</tr>
<tr>
<td>Age</td>
</tr>
</tbody>
</table>

Clinical Guideline for the management and investigation of Henoch-Schonlein Purpura (HSP) in children.
| **Sex** (male, female, transgender / gender reassignment) | x |
| **Race / Ethnic communities / groups** | x |
| **Disability** - learning disability, physical disability, sensory impairment and mental health problems | x |
| **Religion / other beliefs** | x |
| **Marriage and civil partnership** | x |
| **Pregnancy and maternity** | x |
| **Sexual Orientation, Bisexual, Gay, heterosexual, Lesbian** | x |

You will need to continue to a full Equality Impact Assessment if the following have been highlighted:
- You have ticked “Yes” in any column above and
- No consultation or evidence of there being consultation - this excludes any policies which have been identified as not requiring consultation. or
- Major service redesign or development

8. Please indicate if a full equality analysis is recommended.  Yes  No x

9. If you are not recommending a Full Impact assessment please explain why. No relevant fields.

| Signature of policy developer / lead manager / director T.Fergus | Date of completion and submission January 2014 |
| Name and signatures of members carrying out the Screening Assessment | 1. |

_A summary of the results will be published on the Trust’s web site._

Signed _____T.Fergus__________

Date ____January 2014___________