Norfolk and Norwich University Hospitals NHS Foundation Trust

James Paget University Hospitals NHS Foundation Trust

Joint Trust Guideline for the Management of: Henoch – Schönlein Purpura (HSP) in Children

A clinical guideline recommended for use

For Use in:	Jenny Lind Children's Hospital, Emergency Department
By:	Junior and Senior medical staff, Nursing staff
For:	Children aged 0 – 16 years
Division responsible for document:	Women and Children Division
Key words:	Child, Henoch-Schönlein, Purpura
Name of document author:	Chris Upton
Job title of document author:	Consultant Paediatrician (NNUH)
Name of document author's Line Manager:	Dr Mary-Anne Morris (NNUH)
Job title of author's Line Manager:	Chief of Services, Paediatrics (NNUH)
Supporter:	Dr Abbie Reeve Paediatric Consultant. (JPUH).
Assessed and approved by the:	Clinical Guidelines Assessment Panel (CGAP) If approved by committee or Governance Lead Chair's Action; tick here ☑
Date of approval:	20/10/2020
Ratified by or reported as approved to (if applicable)**:**	Clinical Safety and Effectiveness Sub-board
To be reviewed before: This document remains current after this date but will be under review	20/10/2023
To be reviewed by:	Dr Chris Upton (or successor)
Reference and / or Trust Docs ID No:	1219
Version No:	JCG0082 v2.1
Compliance links: *(is there any NICE related to guidance)*	None
If Yes - does the strategy/policy deviate from the recommendations of NICE? **If so why?**	N/A

Trust Guideline for the Management of:
Henoch – Schönlein Purpura (HSP) in Children

Version and Document Control:

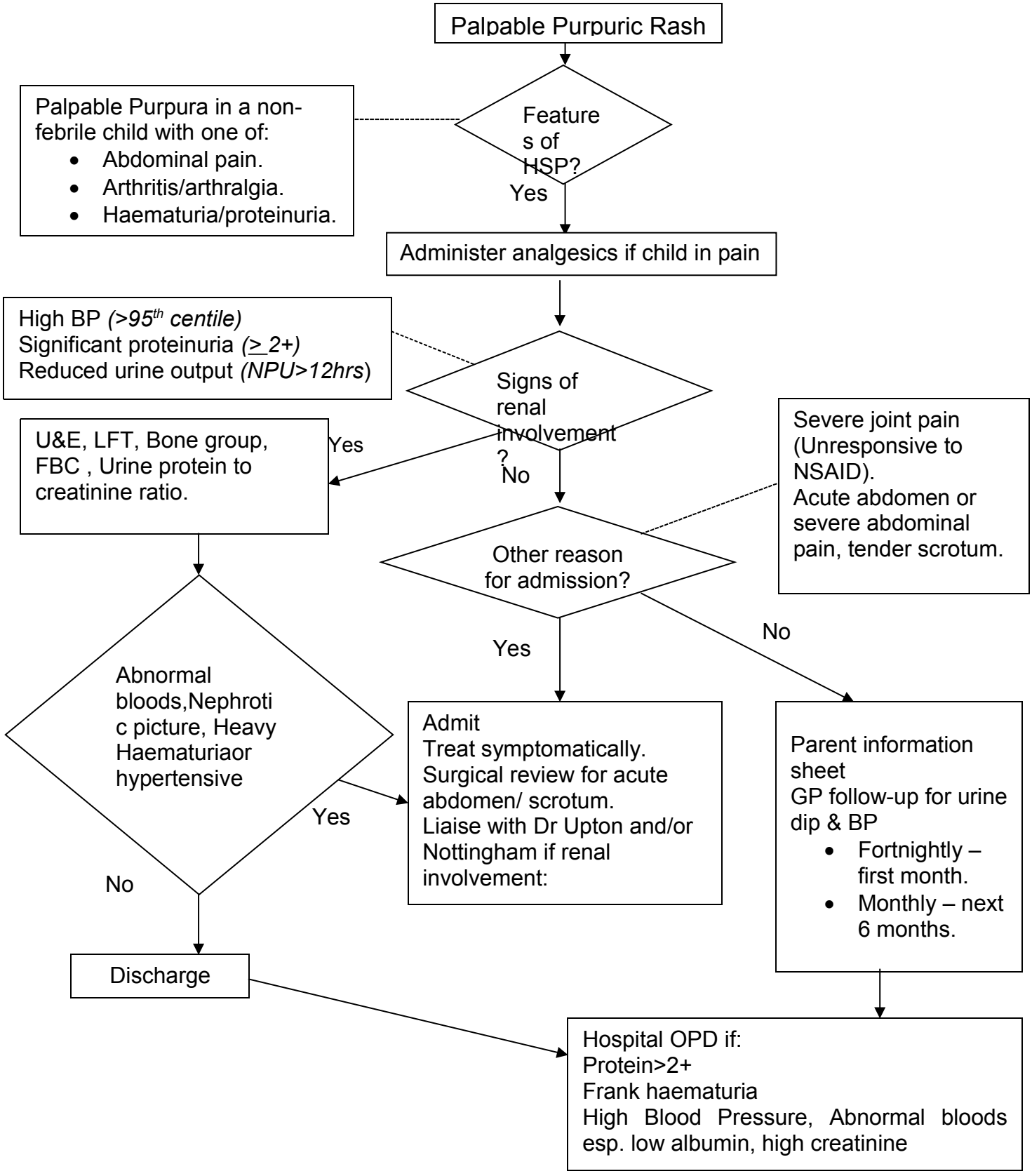
Version Number	Date of Update	Change Description	Author
2.1	20/10/2020	Document reviewed, no clinical changes	Chris Upton

This is a Controlled Document

Printed copies of this document may not be up to date. Please check the hospital intranet for the latest version and destroy all previous versions.

Joint Trust Guideline for the Management of: Henoch – Schönlein Purpura (HSP) in Children

Quick Reference



Joint Trust Guideline for the Management of: Henoch – Schönlein Purpura (HSP) in Children

Objective of Guideline

To promote evidence based management of children with HSP.

Rationale for the recommendations

HSP is the commonest vasculitis of childhood with an incidence of 20 / 100,000, thus making it a common diagnosis in the Jenny Lind Children's Hospital. This guideline aims to provide consistency in practice, to decrease Children's Assessment Unit (CAU) waiting times, to avoid unnecessary investigations, to identify the small group of children with HSP at risk of complications, to standardise follow-up arrangements, and decrease unnecessary inpatient admissions.

Glossary

Anti DNase-B	Additional serology for streptococcal infection – will be done if ASOT clicked on ICE
ASOT	Anti-Streptolysin Titre
BP	Blood Pressure
CAU	Children's Assessment Unit
LFT	Liver Function Tests
NSAID	Non-Steroidal Anti-Inflammatory
OPD	Out Patient Department
U&E	Urea and Electrolytes

Broad recommendations

Background

HSP is an Ig A mediated vasculitis and is the commonest vasculitis of childhood.
It is characterised by **inflammation** of small vessels leading to non-thrombocytopaenic purpura, arthritis/ arthralgia, GI haemorrhage and glomerulonephritis.

Definition of HSP

Palpable purpura in presence of one of the following:

- Diffuse abdominal pain.
- Acute arthritis /arthralgia.
- Renal involvement defined as haematuria or proteinuria.
- A biopsy of any involved tissue shows IgA deposition.

Assessment

The commonest age group is 2 - 8 years. The cause is unknown but there may be a recent history of an upper respiratory tract infection. Abdominal pain or arthralgias sometimes precede the rash.

Clinical Features

Skin: Palpable Purpura: a symmetrical rash on the extensor surfaces of limbs (mainly lower), or dependent and pressure bearing areas (ankles / buttocks). It can also occur in arms face & ears, but usually spares the trunk. The rash typically fades over weeks.

Subcutaneous oedema (scrotum, hands, feet, and sacrum): seen especially in children <3yrs, can be painful, also resolves spontaneously in 24 - 48hrs.

Joints: Joint Pain: Knees and ankles commonly affected with pain, stiffness and sometimes swelling. In most situations it resolves spontaneously within 24-48 hours. It can fluctuate rapidly.

Gastrointestinal Tract: Abdominal pain: Uncomplicated abdominal pain is transient and often resolves spontaneously within 72 hours. Complications include gastro-intestinal haemorrhage (haematemesis, bloody stools), intussusception, perforation, scrotal oedema, testicular torsion and pancreatitis.

Renal Tract : Microscopic haematuria is present in 90% of cases, and in up to 40% may be recurrent. Less common and more significant renal manifestations include proteinuria, nephrotic syndrome, hypertension, and ultimately renal failure (5-7% historically). Renal involvement may only become apparent months later.

Urogentital Tract: Manifest as scrotal oedema / orchitis or testicular torsion. Surgical review is required if there is any testicular involvement.

Investigations

In all children:

- Blood Pressure.
- Urine dipstick.

In children with renal involvement other than isolated microscopic haematuria:

- U & E, LFT, Bone group (NB LFTs include albumin), FBC.
- Consider ASOT, Anti DNAse B, complements, blood cultures, clotting screen if the diagnosis is unclear.

Management

- HSP is usually self-limiting and resolves over a period of 6 weeks and management is generally supportive.
- All patients presenting with a purpuric rash must be seen by a registrar, even if the child appears well.
- Document the child's blood pressure and urine dipstick.
- Simple analgesics for abdominal pain (Ibuprofen at 5-10mg/Kg/dose tds +/- Paracetamol at 10 -15mg/Kg/dose qds).
- If there are features of abdominal *complications* or painful scrotal swelling seek a surgical opinion (testicular haemorrhage is difficult to differentiate from testicular torsion).
- Simple analgesia and bed rest for joint pain (as above).
- Give the family an HSP fact sheet (see below).

Indications for admission

- Abdominal complications.
- Renal complications (other than microscopic hematuria and/or ≥2+ proteinuria):
    - Nephritic syndrome (frank haematuria (+/-casts), persistently elevated BP even after adequate analgesia, raised urea/creatinine, oliguria).
    - Nephrotic syndrome (oedema, proteinuria, hypoalbuminaemia).

  Discuss with on call consultant, who may consider further discussions with Dr Upton (if available) or Nottingham renal team (**********).
- Symptomatic treatment of severe joint pain, severe abdominal pain.

Early referral to Paediatric Nephrology

Children with:

- Acute nephritic syndrome.
- Nephrotic syndrome.
- Impaired renal function.
- Hypertension.

The aim is for early detection of those with severe renal involvement, for treatment with immunosuppression, prior to the development of scarring

Consider Prednisolone (2mg/Kg/day) for

Significant HSP with abdominal pain, severe GI haemorrhage, and testicular pain. There is no evidence that steroids prevent the development of long-term renal complications [1]. This is a *consultant* decision.

Discharge & Follow-up

If discharged from the CAU then it is imperative that appropriate follow-up is arranged to ensure adequate symptom control and resolution of the disease. The aim of follow-up is to detect any persistent renal inflammation that could progress onto permanent renal impairment.

Up to 82% who present with haematuria (with or without mild proteinuria) are normal on long-term follow-up.

Request the following primary care action

Copy & paste onto Electronic Discharge letter (EDL) the following box, and insert the 90th centile BP for age from the BP charts widely available on the unit. In very young children in which one might anticipate difficulties in measuring blood pressure in primary care, it may be appropriate to refer to Dr Upton at discharge to get BP checks done but in this instance please ask GP to monitor urine dips as below.GP follow-up – for uncomplicated HSP

Please monitor BP and urine dipstick according the regime below

- Fortnightly for the first month

- Monthly until 6 months or until 2 consecutive urine samples negative after 6 months

(The 90th centile for systolic BP for this age child is……and diastolic is……..

If unable to measure BP due to age, please refer back to admitting consultant)
Please refer to admitting consultant if:

- Proteinuria ≥2+
- Frank haematuria
- BP above 90th centile for age on two consecutive occasions

Discharge if 2 consecutive urine dipsticks normal with normal BP at 6 months

Secondary care follow-up

Not necessary for uncomplicated cases. Cases that have required in patient admission may need an OPD review at discretion of admitting consultant.

For children under secondary care follow up, refer to Paediatric Nephrology

- Any child with persistent proteinuria (≥2+ or more).
- Frank haematuria.
- Hypertension.
- Abnormal renal function or low albumin.

Outcomes in children with HSP and renal involvement

- Overall 1% progress to end-stage renal failure.
- HSP accounts for 5-15% of children entering end-stage renal failure.
- Age > 4, persistent purpura (> 1/12), and severe abdominal pain are significant risk factors for renal involvement .

The following clinical presentations are listed in order of increasingly poor outcomes:

1. Microscopic haematuria only.
2. Proteinuria (without nephrotic syndrome) + micro/macroscopic haematuria.
3. Acute nephritis (haematuria + ↑BP + ↑creatinine).
4. Nephrotic syndrome.
5. Mixed nephritic/nephrotic syndrome.

Clinical Audit Standards derived from guideline

Audit of children presenting with HSP to CAU or A&E:

Appropriate admissions, investigations, analgesia, follow up arrangements.

Summary of development and consultation process undertaken before registration and dissemination

The authors listed above wrote the original guideline in 2005, it was updated in 2007 and substantially revised in 2011. It has been presented to the paediatric directorate on each of the

above dates, who have agreed the final content. It was reviewed and updated in 2014 and again in 2017.

Distribution list/ dissemination method

CAU, Buxton ward, Intranet, Emergency Dept.

References/ source documents

Sano H, Izumida M, Shimizu H, Ogawa Y Risk factors of renal involvement and significant proteinuria in Henoch-Schonlein purpura.Eur J Pediatr. 2002 Apr;161(4):196-201

Dillon, MJ.Henoch-Schonlein purpura (treatment and outcome). Cleve Clin J Med. 2002;69 Suppl 2:SII121-3. Review.

Henoch – Schönlein Purpura E J Tizard, M JJ Hamilton Ayres – Arch Dis child Educ Pract Ed 2008;93:1-8. doi10 1136/adc.2004.066035

Narachi H.Arch Dis Child 2005;90:916-920. doi: 10.1136/adc.2005.074641 Watson L, Richardson ARW, Holt RCL, Jones CA, Beresford MW (2012) Henoch-Schonlein Purpura – A 5-Year Review and Proposed Pathway PLoS ONE 7(1): e29512 doi:10.1371/journal.pone.0029512

McCarthy HJ, Tizard EJ (2010) Clinical Practice: Diagnosis and management of Henoch-schonlein Purpura Eur J Pediatr 169:643-650

JENNY LIND CHILDREN'S HOSPITAL

Norfolk and Norwich University Hospitals NHS
NHS Foundation Trust

Patient Information Leaflet - Henoch-Schönlein purpura (HSP)

What is Henoch-Schönlein purpura (HSP)?
Your child has been diagnosed with a condition known as Henoch-Schönlein purpura. This is a condition which causes blood vessels to become inflamed (irritated and swollen). It usually affects the ***small*** blood vessels in the ***skin*** causing a ***rash*** that is called purpura. However, it may also affect blood vessels in the intestines, kidneys and joints.

What causes HSP ?
The cause of HSP is unknown. It occurs most often after an upper respiratory infection, like a cold. It appears most often in children from two to ten years of age, but it can occur in anyone. HSP is not contagious. Doctors don't know how to prevent HSP yet.

What can HSP cause?

- **Skin rash -** This occurs as reddish-purple spots, usually on the buttocks and lower legs. Many children with HSP also have swelling over the top of the feet and backs of hands, and around the scrotum in boys.
- **Pain in the joints** - mainly in the knees and ankles
- **Stomach pains**
- **Blood in the stool (poo) or urine**, This is caused by the blood vessels in the bowel and the kidneys becoming inflamed. Serious kidney problems do not happen very often, but they can occur.
- In rare cases, an abnormal folding of the bowel called **intussusception** can occur. This causes a blockage in the intestines that may need surgery.

Treatment
There is no specific treatment for HSP. Fortunately, HSP usually gets better without any treatment. Painkillers (e.g. Paracetamol) or anti-inflammatory medicines (e.g. Ibuprofen) can help relieve the pain in your child's joints. Medicines, such as antibiotics may be given to treat the infection that may have triggered HSP. Your doctor may also recommend a drug called Prednisolone (a steroid). This drug is only given to children who are so poorly that they require admission to hospital.

What to do next ?
Usually, HSP gets better on its own and does not cause any lasting problems. Unfortunately, about half of people who have had HSP once will get it again. Following your return home you will be asked to take your child back to your GP so that they can check your child's urine samples and blood pressure for any signs of kidney problems.

If your child has increasing tummy pains or becomes more unwell at home you should contact your **GP immediately** or call the **Children's Assessment Unit** on **01603 286319** for advice.

Henoch-Schonlein Purpura (HSP) - NNUH Fact Sheet Author: Dr Kate Armon, Consultant