

Clinical Guidance

Management of Henoch Schonlein Purpura (HSP)

Summary: This document guides assessment and management of children with Henoch Schonlein Purpura including follow up arrangements

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Author(s)	Joy Tan, Mark Butler
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HENOCH-SCHONLEIN PURPURA (HSP)

Incidence

- HSP is the most common vasculitis of childhood, with an incidence of 20.4 per 100,000 in the UK¹
- It is more common in Asian and Caucasian children. ¹
- There is a similar incidence between boys and girls ⁸
- The mean age of presentation is 6 years old and generally affects age 2-10 years of age ⁸

Aetiology

- Its cause is unknown ⁸
- Usually presents following an infection (GAS, HSV, adenovirus, CMV, Mycoplasma, HPV, B19 or vaccination (MMR, PCV, Influenza) ¹
- Group A Streptococcus (GAS) has been found in around one third of patients with HSP ⁸

Presentation

- HSP is a clinical diagnosis of rash, and one or more of:
 - arthritis/arthralgia, ⁸
 - abdominal pain, ⁸
 - haematuria, proteinuria +/- High BP⁸

Less Common but serious- could present with:

- Intussusception^{1, 8}
- Haematemesis and Melaena^{1, 8}
- Scrotal/ testicular swelling^{1, 8}
- Pancreatitis/ gall bladder involvement^{1, 8}
- Vasculitis involvement of brain and lung^{1, 8}

Pathophysiology

- Small vessel systemic vasculitis. IgA mediated²

Investigation

- Height
- Weight
- Blood pressure
- Urinalysis - dipstick & MSU
- Urine Profile
- Urine protein: creatinine ratio (UPCR)
- FBC
- Serum Albumin
- **Also should consider the following post discussion with senior**
 - CRP, ESR
 - Blood culture
 - Coagulation Profile
 - ASOT
 - Throat Swab
 - ANCA, ANA, dsDNA, C3, C4 (if there is significant renal involvement)
 - Immunoglobulin profile
 - Abdominal Ultrasound if suspecting intussusception

Differential Diagnosis:

- Sepsis (meningococcal septicaemia)
- ITP
- Others vasculitis (SLE, Polyarteritis Nodosa, ANCA associated vasculitis, hypersensitivity vasculitis)

Management :

- Symptomatic treatment of pain with analgesia.
 - Paracetamol/Ibuprofen for pain relief. (NSAIDs should be given with caution if evidence of renal involvement)
- Rest for arthritis/arthralgia
- Strict input and output (*if admitted as inpatient*)
- Steroids
 - No evidence for their use routinely to prevent HSP Nephritis.
 - ***This may be considered for children with severe abdominal or arthritis + significant systemic disease including rash under the guidance of senior review/ consultant.*** ^{9,10}
 - Surgical review essential before using steroids for children with abdominal pain. ^{4,5,6.}
- Continue f/u of BP and urine for at least 6 months as 97% of renal involvement presents by that time

BP measurements and BP Centile Charts

Original tables and additional advice can be found at [2016 European Society of Hypertension guidelines for the management of high blood pressure in children and adolescents](#) ¹¹

doi.org/10.1097/HJH.0000000000001039

Boys

Age (years)	BP percentile	SBP (mmHg) percentile of height							DBP (mmHg) percentile of height						
		5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
1	90th	97	97	98	100	101	102	103	52	53	53	54	55	55	56
	95th	100	101	102	104	105	106	107	56	57	57	58	59	59	60
	99th	108	108	109	111	112	113	114	64	64	65	65	66	67	67
2	90th	98	99	100	101	103	104	105	57	58	58	59	60	61	61
	95th	102	103	104	105	107	108	109	61	62	62	63	64	65	65
	99th	109	110	111	112	114	115	116	69	69	70	70	71	72	72
3	90th	100	100	102	103	104	106	106	61	62	62	63	64	64	65
	95th	104	104	105	107	108	109	110	65	66	66	67	68	68	69
	99th	111	111	113	114	115	116	117	73	73	74	74	75	76	76
4	90th	101	102	103	104	106	107	108	64	64	65	66	67	67	68
	95th	105	106	107	108	110	111	112	68	68	69	70	71	71	72
	99th	112	113	114	115	117	118	119	76	76	76	77	78	79	79
5	90th	103	103	105	106	107	109	109	66	67	67	68	69	69	70
	95th	107	107	108	110	111	112	113	70	71	71	72	73	73	74
	99th	114	114	116	117	118	120	120	78	78	79	79	80	81	81
6	90th	104	105	106	108	109	110	111	68	68	69	70	70	71	72
	95th	108	109	110	111	113	114	115	72	72	73	74	74	75	76
	99th	115	116	117	119	120	121	122	80	80	80	81	82	83	83
7	90th	106	107	108	109	111	112	113	69	70	70	71	72	72	73
	95th	110	111	112	113	115	116	116	73	74	74	75	76	76	77
	99th	117	118	119	120	122	123	124	81	81	82	82	83	84	84
8	90th	108	109	110	111	113	114	114	71	71	71	72	73	74	74
	95th	112	112	114	115	116	118	118	75	75	75	76	77	78	78
	99th	119	120	121	122	123	125	125	82	82	83	83	84	85	86
9	90th	110	110	112	113	114	116	116	72	72	72	73	74	75	75
	95th	114	114	115	117	118	119	120	76	76	76	77	78	79	79
	99th	121	121	123	124	125	127	127	83	83	84	84	85	86	87
10	90th	112	112	114	115	116	118	118	73	73	73	74	75	76	76
	95th	116	116	117	119	120	121	122	77	77	77	78	79	80	80
	99th	123	123	125	126	127	129	129	84	84	85	86	86	87	88
11	90th	114	114	116	117	118	119	120	74	74	74	75	76	77	77
	95th	118	118	119	121	122	123	124	78	78	78	79	80	81	81
	99th	125	125	126	128	129	130	131	85	85	86	87	87	88	89
12	90th	116	116	117	119	120	121	122	75	75	75	76	77	78	78
	95th	119	120	121	123	124	125	126	79	79	79	80	81	82	82
	99th	127	127	128	130	131	132	133	86	86	87	88	88	89	90
13	90th	117	118	119	121	122	123	124	76	76	76	77	78	79	79
	95th	121	122	123	124	126	127	128	80	80	80	81	82	83	83
	99th	128	129	130	132	133	134	135	87	87	88	89	89	90	91
14	90th	119	120	121	122	124	125	125	77	77	77	78	79	80	80
	95th	123	123	125	126	127	129	129	81	81	81	82	83	84	84
	99th	130	131	132	133	135	136	136	88	88	89	90	90	91	92
15	90th	120	121	122	123	125	126	127	78	78	78	79	80	81	81
	95th	124	125	126	127	129	130	131	82	82	82	83	84	85	85
	99th	131	132	133	134	136	137	138	89	89	90	91	91	92	93
16	90th	121	122	123	124	126	127	128	78	78	79	80	81	81	82
	95th	125	126	127	128	130	131	132	82	82	83	84	85	85	86
	99th	132	133	134	135	137	138	139	90	90	90	91	92	93	93
17	90th	122	122	123	125	126	127	128	78	79	79	80	81	81	82
	95th	125	126	127	129	130	131	132	82	83	83	84	85	85	86
	99th	133	133	134	136	137	138	139	90	90	91	91	92	93	93

Modified from Task Force on High Blood Pressure in Children and Adolescents [7]. Boxed area corresponds to reference values of boys 16 years or older in which the reference values for adults are recommended. BP, blood pressure.

Girls

Age (years)	BP percentile	SBP (mmHg) percentile of height							DBP (mmHg) percentile of height						
		5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
1	90th	94	95	97	99	100	102	103	49	50	51	52	53	53	54
	95th	98	99	101	103	104	106	106	54	54	55	56	57	58	58
	99th	105	106	108	110	112	113	114	61	62	63	64	65	66	66
2	90th	97	99	100	102	104	105	106	54	55	56	57	58	58	59
	95th	101	102	104	106	108	109	110	59	59	60	61	62	63	63
	99th	109	110	111	113	115	117	117	66	67	68	69	70	71	71
3	90th	100	101	103	105	107	108	109	59	59	60	61	62	63	63
	95th	104	105	107	109	110	112	113	63	63	64	65	66	67	67
	99th	111	112	114	116	118	119	120	71	71	72	73	74	75	75
4	90th	102	103	105	107	109	110	111	62	63	64	65	66	66	67
	95th	106	107	109	111	112	114	115	66	67	68	69	70	71	71
	99th	113	114	116	118	120	121	122	74	75	76	77	78	78	79
5	90th	104	105	106	108	110	111	112	65	66	67	68	69	69	70
	95th	108	109	110	112	114	115	116	69	70	71	72	73	74	74
	99th	115	116	118	120	121	123	123	77	78	79	80	81	81	82
6	90th	105	106	108	110	111	113	113	68	68	69	70	71	72	72
	95th	109	110	112	114	115	117	117	72	72	73	74	75	76	76
	99th	116	117	119	121	123	124	125	80	80	81	82	83	84	84
7	90th	106	107	109	111	113	114	115	70	70	71	72	73	74	74
	95th	110	111	113	115	117	118	119	74	74	75	76	77	78	78
	99th	117	118	120	122	124	125	126	82	82	83	84	85	86	86
8	90th	107	109	110	112	114	115	116	71	72	72	73	74	75	76
	95th	111	112	114	116	118	119	120	75	76	77	78	79	79	80
	99th	119	120	122	123	125	127	127	83	84	85	86	87	87	88
9	90th	109	110	112	114	115	117	118	72	73	74	75	76	76	77
	95th	113	114	116	118	119	121	121	76	77	78	79	80	81	81
	99th	120	121	123	125	127	128	129	84	85	86	87	88	88	89
10	90th	111	112	114	115	117	119	119	73	73	74	75	76	77	78
	95th	115	116	117	119	121	122	123	77	78	79	80	81	81	82
	99th	122	123	125	127	128	130	130	85	86	86	88	88	89	90
11	90th	113	114	115	117	119	120	121	74	74	75	76	77	78	78
	95th	117	118	119	121	123	124	125	78	78	79	80	81	82	82
	99th	124	125	127	129	130	132	132	86	86	87	88	89	90	90
12	90th	115	116	118	120	121	123	123	74	75	75	76	77	78	79
	95th	119	120	122	123	125	127	127	78	79	80	81	82	82	83
	99th	126	127	129	131	133	134	135	86	87	88	89	90	90	91
13	90th	117	118	120	122	124	125	126	75	75	76	77	78	79	79
	95th	121	122	124	126	128	129	130	79	79	80	81	82	83	83
	99th	128	130	131	133	135	136	137	87	87	88	89	90	91	91
14	90th	120	121	123	125	126	128	128	75	76	77	78	79	79	80
	95th	124	125	127	128	130	132	132	80	80	81	82	83	84	84
	99th	131	132	134	136	138	139	140	87	88	89	90	91	92	92
15	90th	122	124	125	127	129	130	131	76	77	78	79	80	80	81
	95th	126	127	129	131	133	134	135	81	81	82	83	84	85	85
	99th	134	135	136	138	140	142	142	88	89	90	91	92	93	93
16	90th	125	126	128	130	131	133	134	78	78	79	80	81	82	82
	95th	129	130	132	134	135	137	137	82	83	83	84	85	86	87
	99th	136	137	139	141	143	144	145	90	90	91	92	93	94	94
17	90th	127	128	130	132	134	135	136	80	80	81	82	83	84	84
	95th	131	132	134	136	138	139	140	84	85	86	87	87	88	89
	99th	139	140	141	143	145	146	147	92	93	93	94	95	96	97

BP, blood pressure. Modified from Task Force on High Blood Pressure in Children and Adolescents [7]. Boxed area corresponds to reference values of boys 16 years or older in which the reference values for adults are recommended.

Prognosis

- 95% self-limiting illness within 4-6 weeks
- Relapse in 1/3 usually within 4 months of initial episode.
- Renal disease in 1/3
- Long term renal disease in 3 %

Discharge and follow-up plan

- Arrange follow up so as not to miss persistent renal disease/nephritis.
- Nephritis is not always present on initial presentation.
- 90% of children with nephritis develop this by two months and 97% by six months.

Nephrology referral criteria

Criteria for referral to/discussion with Paediatric Nephrology
ANY OF THE FOLLOWING:

1. Nephrotic syndrome:
 - Urine protein:creatinine ratio >250mg/mmol
 - Plasma albumin <25g/L
 - Oedema
2. Hypertension.
3. Abnormal renal function.
4. Macroscopic haematuria ≥5 days.
5. Persistent proteinuria:
 - Urine protein:creatinine ratio >250mg/mmol for 4 weeks
 - Urine protein:creatinine ratio >100mg/mmol for 3 months
 - Urine protein:creatinine ratio >50mg/mmol for 6 months

Henoch-Schonlein Purpura (HSP) flow chart

How to approach a patient with suspected Henoch-schonlein purpura (HSP)

□ **Perform:**

- History and examination
- Height and Weight
- Vital signs including BP
- Urinalysis

□ **Look for:**

- **Cutaneous-** Palpable purpura predominantly of lower limbs and buttocks

Plus one of

- **Gastrointestinal** - Abdominal pain
- **Articular** - Arthritis and/or arthralgia
- **Renal** - Haematuria and/or Proteinuria (>2+) and high BP (>95th centile)

EXCLUDE other causes of purpuric rash (meningococcal septicaemia, Disseminated Intravascular Coagulation, Neoplasm, Non Accidental Injury etc.)

If patient meets criteria above, please also perform

- Urine Protein: Creatinine Ratio (UPCR)
- FBC, Basic Renal Profile and Albumin

Also should consider following discuss with senior

- CRP, ESR
- Coagulation Profile
- ASOT
- Throat Swab
- ANCA, ANA, dsDNA, C3, C4
- Immunoglobulin profile
- Abdominal Ultrasound

If any of the following present

- Hypertension
 - Oedema
 - UPCR > 250mg/mmol
 - Macroscopic Haematuria
 - Serum Albumin <25g/dl
 - eGFR < 90ml/min/1.73m²
- [Formula: $31 \times \text{height (cm)} / \text{serum creatinine (mmol/L)}$]**

Refer to Renal Team

- Bleep 1141/ mobile 07990 800587

If any of the following present

- Atypical features- Myalgia, Myositis, atypical organ involvement eg respiratory (upper or lower), scleritis
- Persistent, recurrent or atypical rash
- Persistent musculoskeletal pain
- Recurrent non-renal flares

Refer to Rheumatology Team

- 07787842692 SpR

If normal, look for other indicators for admission

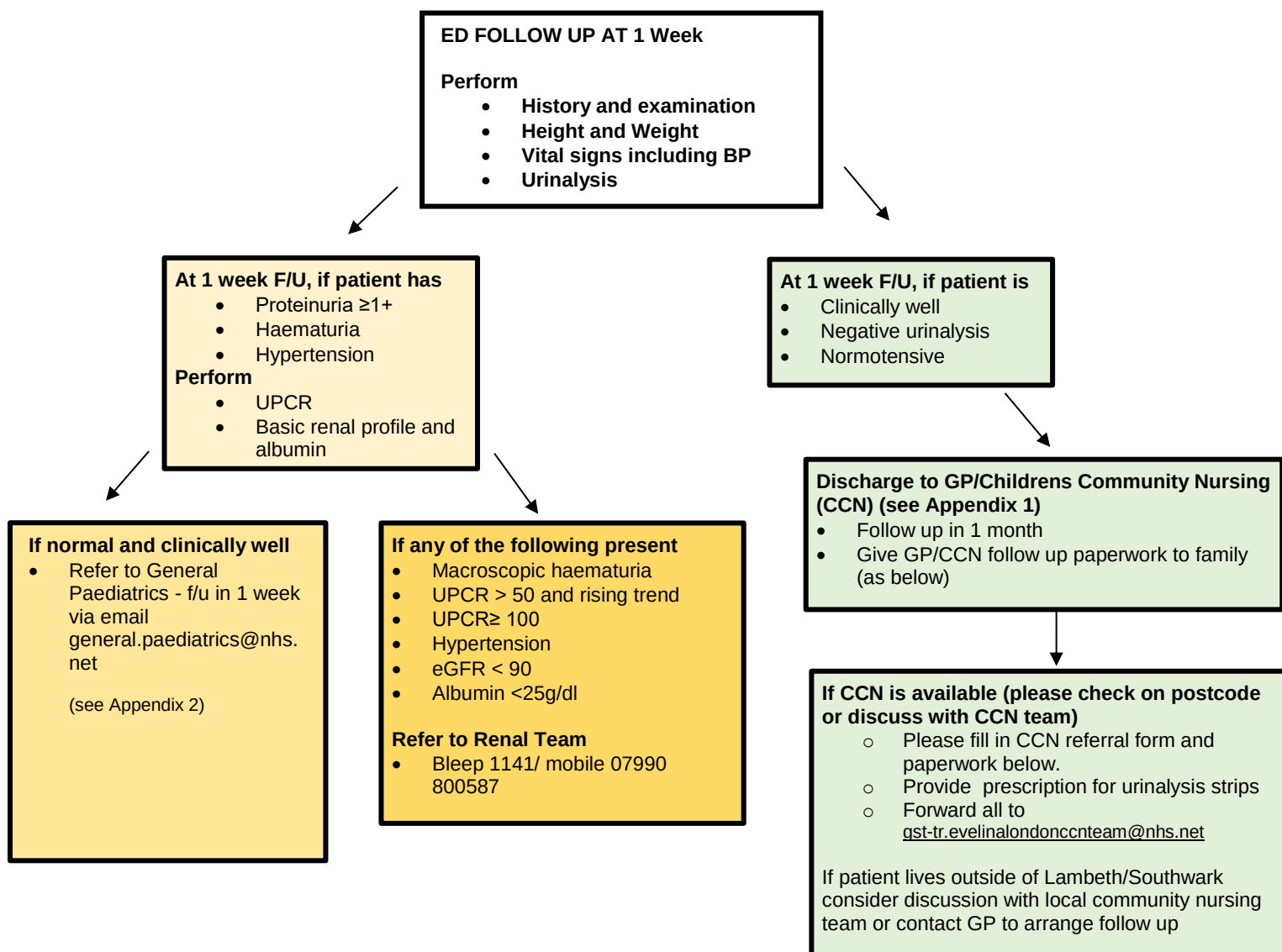
- Acute abdomen or severe abdominal pain
- Tender scrotum
- Malena

If Absent, ED FOLLOW UP AT 1 Week (Please see following page)

If present,

- Consider analgesia (No NSAIDS if renal involvement present)
- Consider surgical review
- Consider admission under general paediatrics (Bleep 0339)

ED Follow up at One Week



Appendix 1

GP or Children's Community Nursing Team Follow up

Patient Sticker 	Height: Weight: 	95th Centile for BP: Cuff size for BP:
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Dear Colleague,

Please will you continue to follow up this patient with HSP, they are normotensive and their urinalysis has been negative at presentation and at 1 week review.

However as you are aware all children with HSP require follow up to at least 6 months for HSP nephritis. We advise that children who do not show evidence of proteinuria at 1 week have their blood pressure checked and urine dipped 3 times in the next 5 months at 1month, 3 months and 6 months after initial presentation.

Therefore we would be very grateful if you could perform ongoing reviews of this patient as per the table below, and refer to the general paediatric team at Evelina if any signs of nephritis are present.

Thank you
The Evelina Paediatric Team

	Assessment	Outcome
1 month post diagnosis	Urine dip negative and BP normal	Follow up in 2 months
	Haematuria present	Refer to General Paediatric team [General.Paediatrics@nhs.uk]
	Proteinuria $\geq 1+$	
	BP > 95 centile on 3 readings	
3 Months post diagnosis	Urine dip negative and BP normal	Follow up in 3 months
	Haematuria present	Refer to General Paediatric team [General.Paediatrics@nhs.uk]
	Proteinuria $\geq 1+$	
	BP > 95 centile on 3 readings	
6 months post diagnosis	Urine dip negative and BP normal	NO further follow up required for HSP
	Haematuria present	Refer to General Paediatric team [General.Paediatrics@nhs.uk]
	Proteinuria $\geq 1+$	
	BP > 95 centile on 3 readings	

Patient's Henoch-Scholein Purpura (HSP) Follow-Up Record

Patient Sticker

You/your child has been diagnosed with HSP. It is important that we continue to monitor the kidney involvement of this disease. You will need to have your blood pressure measured and an early morning sample of urine (ie. the first urine passed that day) tested at regular intervals as suggested by the table below. This follow up will take place with your GP or by one of the hospital at home team members . Please keep this document safe and fully completed at each stage so that we can monitor you/your child closely. If you need more information, please refer to <https://www.nhs.uk/conditions/henoch-schonlein-purpura-hsp/>

Date of review	Place of Review	Height	Weight	Urine dipstick (Protein)	Urine dipstick (Blood)	Systolic blood pressure 95 th centile	Systolic BP (mmHg)

Appendix 2

General Paediatric Follow up (Patients with proteinuria):

Review at: • 2 weeks post diagnosis • 1 month post diagnosis • 2 months post diagnosis • 3 months post diagnosis • 4 months post diagnosis • 6 months post diagnosis • 12 months post diagnosis	<u>At each visit:</u> • Clinical Review and Examination • Urine Dip (provide patients with pots so they can bring an early morning sample with them) • BP (manual) <u>If urine dip $\geq 1+$ protein or $\geq 1+$ haematuria, please undertake the following basic tests:</u> • UPCR (on early morning sample) • Urine MCS • Basic renal profile • Serum Albumin
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Refer to Nephrology if:

- Macroscopic Haematuria ≥ 5 days or more than one discrete episode
- BP > 95th centile on three readings
- Urine PCR > 50mg/mmol and on a rising trend
- Urine PCR > 100mg/mmol
- Abnormal GFR
- Serum Albumin <25g/dl

And when decision made to refer undertake the following investigations:

- Renal USS
- FBC
- Clotting
- CRP
- ESR
- ASOT
- ANA
- ANCA
- dsDNA
- C3 & C4
- Immunoglobulin profile
- Group & Save

End of Pathway:

At 12 months if the patient has a negative urine dip and normal BP they can be discharged with advice for yearly GP review consisting of BP and urine dip for protein.

If there are ongoing concerns with microscopic haematuria or proteinuria please refer to the paediatric nephrology team.

Proteinuria follow up table for reference:

	Assessment	Outcome
2 weeks post diagnosis	Urine dip negative and BP normal	Follow up at 1 month
	Haematuria present or Proteinuria $\geq 1+$	Undertake basic tests as listed
	BP > 95 centile on 3 readings	Refer to Paediatric Nephrology team
1 months post diagnosis	Urine dip negative and BP normal	Follow up at 2 months
	Haematuria present or Proteinuria $\geq 1+$	Undertake basic tests as listed
	BP > 95 centile on 3 readings	Refer to Paediatric Nephrology team
2 months post diagnosis	Urine dip negative and BP normal	Follow up at 3 months
	Haematuria present or Proteinuria $\geq 1+$	Undertake basic tests as listed
	BP > 95 centile on 3 readings	Refer to Paediatric Nephrology team
3 months post diagnosis	Urine dip negative and BP normal	Follow up at 4 months
	Haematuria present or Proteinuria $\geq 1+$	Undertake basic tests as listed
	BP > 95 centile on 3 readings	Refer to Paediatric Nephrology team, ELCH
4 months post diagnosis	Urine dip negative and BP normal	Follow up at 6 months
	Haematuria present or Proteinuria $\geq 1+$	Undertake basic tests as listed
	BP > 95 centile on 3 readings	Refer to Paediatric Nephrology team, ELCH
6 months post diagnosis	Urine dip negative and BP normal	Follow up at 12 months
	Haematuria present or Proteinuria $\geq 1+$	Undertake basic tests as listed
	BP > 95 centile on 3 readings	Refer to Paediatric Nephrology team ELCH
12 months post diagnosis	Urine dip negative and BP normal	Discharge to yearly GP follow up
	Haematuria present or Proteinuria $\geq 1+$	Undertake basic tests as listed & refer to renal team
	BP > 95 centile on 3 readings	Refer to Paediatric Nephrology team ELCH

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