Henoch-Schonlein Purpura Guidelines

Henoch-Schonlein purpura (HSP) is the commonest vasculitis of childhood which is self-limiting in majority of cases.

Epidemiology:
Incidence varies from 10 -20 per 100000 children. Male to Female ratio 2:1
More common in young children with over 50% (< 5 years) and over 75% (<10 years). More common in winter, autumn and spring rather than summer

Diagnosis:
The agreed criteria for HSP diagnosis (1) is as follows

Palpable purpura (mandatory) in the presence of at least one of the following

- Diffuse abdominal pain
- Arthritis (acute) or arthralgia
- Renal involvement (Hematuria and/or proteinuria)
- Biopsy showing predominant IgA deposition

Clinical Features:
Classical skin rash often precipitates presentation of patients with HSP
Cutaneous

- Palpable purpura – symmetrically distributed over extensor, dependent surfaces of lower limbs & buttocks. Can also involve arms, face & ears but usually spares trunk.
- Purpura can range from petechiae to large ecchymoses and maybe preceded by urticarial or erythematous, maculopapular lesions.
- Bullous lesions are rare but seen.

Gastro intestinal

- Colicky abdominal pain
- Vomiting & GI bleeding
- Intussusception
- Rarely – protein losing enteropathy, pancreatitis & hydrops of gall bladder

Joints

- Pain, swelling (peri-articular oedema) & decreased range of movement. Synovial effusions are typically absent.
- Usually affects large joints & lower limbs (Knees, ankles, feet & hips).Less commonly wrists, fingers and elbows.
- Although can be debilitating, does not result in permanent damage.

Renal –

- Haematuria, proteinuria, nephritic syndrome/nephritis & renal impairment
- Hypertension

Other features
Genital- orchitis, genital oedema, testicular pain, cord heamatoma
Neurological –Nonspecific headache & mood changes. Rarely – encephalopathy, seizures and stroke.

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Differential Diagnosis:
Sepsis – especially meningococcal disease
ITP
In atypical presentations connective tissue disorders (SLE, microscopic polyarteritis, and Wegener's granulomatosis) should be considered

Investigations:
No specific laboratory tests for diagnosis, but these investigations are aimed at excluding other diagnosis & assessing the extent of organ involvement.
All patients need weight, height, BP and urine dipstix

Initial
FBC, Coag, Urea & electrolytes, Albumin
Urine dipstick – for proteinuria and haematuria. If proteinuria more than 0.3 gm/L (>1 + on Siemens Multistix 10 SG) , then early morning urine protein:creatinine ratio.

If renal disease present, also appropriate to measure ASOT and anti-DNAse B as a post streptococcocal glomerulonephritis may complicate picture.
If diagnosis in doubt -
Autoimmune screen (ANA, dsDNA, ANCA), Immunoglobulins, C3 & C4 levels.

If suspecting Intussusception – Abdominal U/S is the investigation of choice

Admission
Consider admission if the patient is unwell, has significant joint pain, severe abdominal pain, G I haemorrhage, hypertension, evidence of acute glomerulonephritis, nephrotic syndrome, abnormal renal function or neurological symptoms.

Treatment:
Usually drug therapy - aimed for symptomatic relief

GIT For relief of significant abdominal pain- Prednisolone 1mg/kg/day for 2 weeks followed by 0.5mg/k/day for 1 week and then 0.5 mg/kg alt days for 1 week (2). Prednisolone as per local guidelines / liaise with tertiary gastroenterologists

In severe GIT disease (protein losing enteropathy or severe GIT hemorrhage) – IV Methylprednisolone followed by oral prednisolone as above.

Joints Treat with paracetamol. Avoid NSAIDs if at risk of dehydration, any renal impairment or hypertensive.

HSP nephritis
Most patients with renal involvement make a good recovery with long term morbidity being <5%. HSP nephritis accounts for < 3% of end stage renal failure (2).
Treatment of HSP nephritis is debatable. In severe renal involvement cases discuss with UHW Paediatric nephrologists because of potential long term consequences.

Severe abdominal, GIT bleeding or joint problems etc may require further review by Paediatric team. Patient with recurrent episodes of HSP should be monitored as for first episode of HSP.

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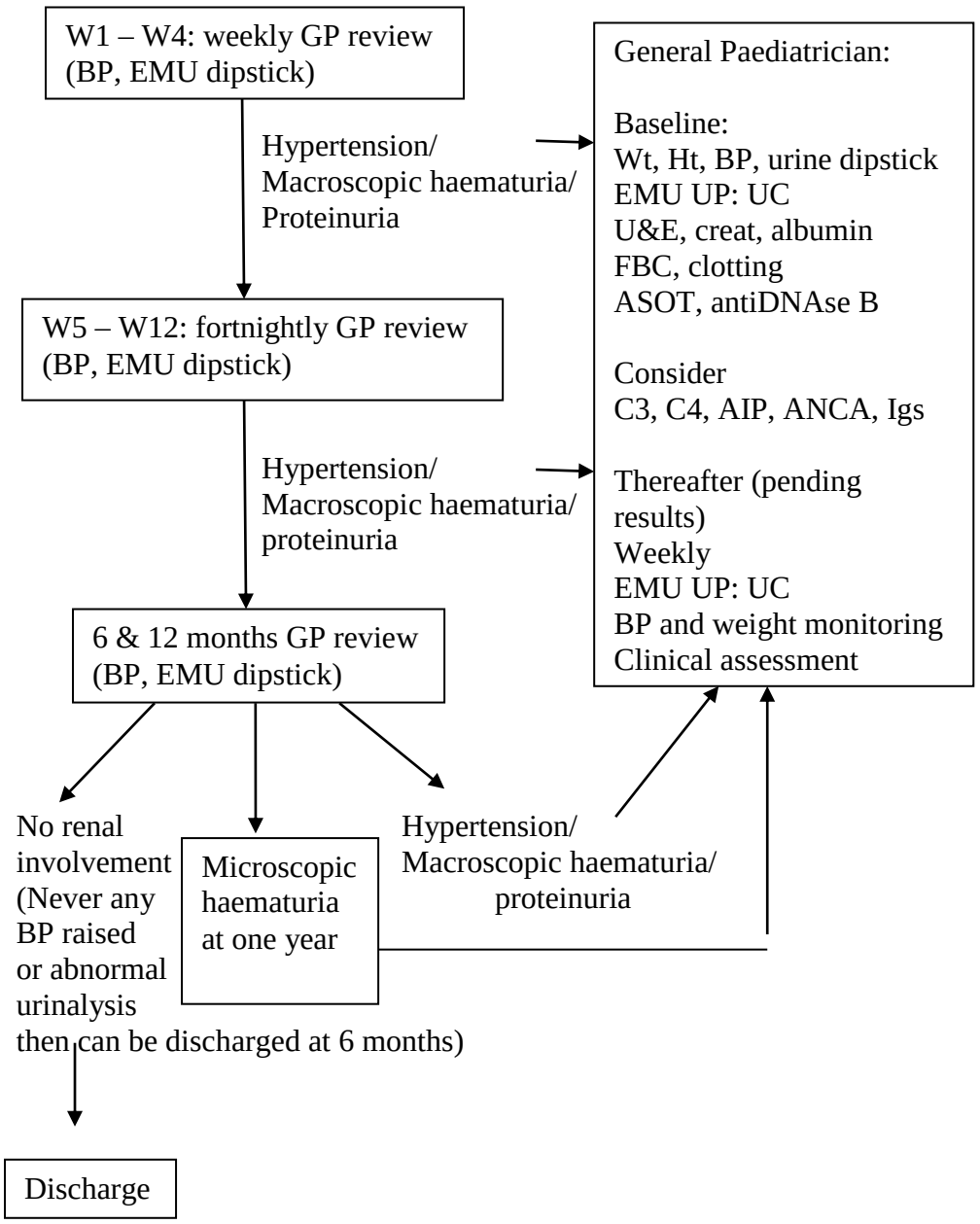
Suggested clinical pathway for detection and referral of patients with HSP nephritis (4)



Discuss with UHW nephrologists if:

Hypertension
Abnormal renal function
Macroscopic haematuria – 5 days
Nephrotic syndrome:
UP: UC > 250mg /mmol
Plasma albumin < 25g/l
Oedema
Acute nephritic syndrome:
Haematuria / proteinuria / oedema / hypertension / oliguria

Or

If persisting abnormalities with:

Persistent proteinuira:
UP: UC >250 mg / mmol for 4 w
UP: UC > 100 mg / mmol for 3 m
UP: UC > 50 mg / mmol for 6 m

Persistant microscopic haematuria at one year

Note

- Different dipsticks may have slightly different values for 1+, 2+.
- Proteinuria is > 0.3g/L (At RGH – Siemens Multistix 10 SG are used and proteinuria of 1+ is 0.3g/L). Levels of 2+ (1.0g/l)
- All children with HSP should be reviewed by a senior paediatrician (middle grade or above). Please discuss with the Paediatric Consultant regarding follow up in clinic. Simple cases with no urinary abnormalities, normal bloods and blood pressure can be followed up by GP and do not need hospital follow up unless there is any other indication.

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***Blood Pressure Centiles* (5)**

Systolic blood pressure for boys by age and height centile

Age, y	BP Percentile	SBP, mm Hg						
		Percentile of Height						
		5th	10th	25th	50th	75th	90th	95th
1	50th	80	81	83	85	87	88	89
	90th	94	95	97	99	100	102	103
	95th	98	99	101	103	104	106	106
	99th	105	106	108	110	112	113	114
2	50th	84	85	87	88	90	92	92
	90th	97	99	100	102	104	105	106
	95th	101	102	104	106	108	109	110
	99th	109	110	111	113	115	117	117
3	50th	86	87	89	91	93	94	95
	90th	100	101	103	105	107	108	109
	95th	104	105	107	109	110	112	113
	99th	111	112	114	116	118	119	120
4	50th	88	89	91	93	95	96	97
	90th	102	103	105	107	109	110	111
	95th	106	107	109	111	112	114	115
	99th	113	114	116	118	120	121	122
5	50th	90	91	93	95	96	98	98
	90th	104	105	106	108	110	111	112
	95th	108	109	110	112	114	115	116
	99th	115	116	118	120	121	123	123
6	50th	91	92	94	96	98	99	100
	90th	105	106	108	110	111	113	113
	95th	109	110	112	114	115	117	117
	99th	116	117	119	121	123	124	125
7	50th	92	94	95	97	99	100	101
	90th	106	107	109	111	113	114	115
	95th	110	111	113	115	117	118	119
	99th	117	118	120	122	124	125	126
8	50th	94	95	97	99	100	102	102
	90th	107	109	110	112	114	115	116
	95th	111	112	114	116	118	119	120
	99th	119	120	122	123	125	127	127
9	50th	95	96	98	100	102	103	104
	90th	109	110	112	114	115	117	118
	95th	113	114	116	118	119	121	121
	99th	120	121	123	125	127	128	129
10	50th	97	98	100	102	103	105	106
	90th	111	112	114	115	117	119	119
	95th	115	116	117	119	121	122	123
	99th	122	123	125	127	128	130	130
11	50th	99	100	102	104	105	107	107
	90th	113	114	115	117	119	120	121
	95th	117	118	119	121	123	124	125
	99th	124	125	127	129	130	132	132
12	50th	101	102	104	106	108	109	110
	90th	115	116	118	120	121	123	123
	95th	119	120	122	123	125	127	127
	99th	126	127	129	131	133	134	135
13	50th	104	105	106	108	110	111	112
	90th	117	118	120	122	124	125	126
	95th	121	122	124	126	128	129	130
	99th	128	130	131	133	135	136	137
14	50th	106	107	109	111	113	114	115
	90th	120	121	123	125	126	128	128
	95th	124	125	127	128	130	132	132
	99th	131	132	134	136	138	139	140
15	50th	109	110	112	113	115	117	117
	90th	122	124	125	127	129	130	131
	95th	126	127	129	131	133	134	135
	99th	134	135	136	138	140	142	142
16	50th	111	112	114	116	118	119	120
	90th	125	126	128	130	131	133	134
	95th	129	130	132	134	135	137	137
	99th	136	137	139	141	143	144	145
17	50th	114	115	116	118	120	121	122
	90th	127	128	130	132	134	135	136
	95th	131	132	134	136	138	139	140
	99th	139	140	141	143	145	146	147

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2	50th	85	85	87	88	89	91	91
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4	50th	88	88	90	91	92	94	94
	90th	101	102	103	104	106	107	108
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5	50th	89	90	91	93	94	95	96
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	99th	117	118	119	120	122	123	124
8	50th	95	95	96	98	99	100	101
	90th	108	109	110	111	113	114	114
	95th	112	112	114	115	116	118	118
	99th	119	120	121	122	123	125	125
9	50th	96	97	98	100	101	102	103
	90th	110	110	112	113	114	116	116
	95th	114	114	115	117	118	119	120
	99th	121	121	123	124	125	127	127
10	50th	98	99	100	102	103	104	105
	90th	112	112	114	115	116	118	118
	95th	116	116	117	119	120	121	122
	99th	123	123	125	126	127	129	129
11	50th	100	101	102	103	105	106	107
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	90th	119	120	121	122	124	125	125
	95th	123	123	125	126	127	129	129
	99th	130	131	132	133	135	136	136
15	50th	107	108	109	110	111	113	113
	90th	120	121	122	123	125	126	127
	95th	124	125	126	127	129	130	131
	99th	131	132	133	134	136	137	138
16	50th	108	108	110	111	112	114	114
	90th	121	122	123	124	126	127	128
	95th	125	126	127	128	130	131	132
	99th	132	133	134	135	137	138	139
17	50th	108	109	110	111	113	114	115
	90th	122	122	123	125	126	127	128
	95th	125	126	127	129	130	131	132
	99th	133	133	134	136	137	138	139

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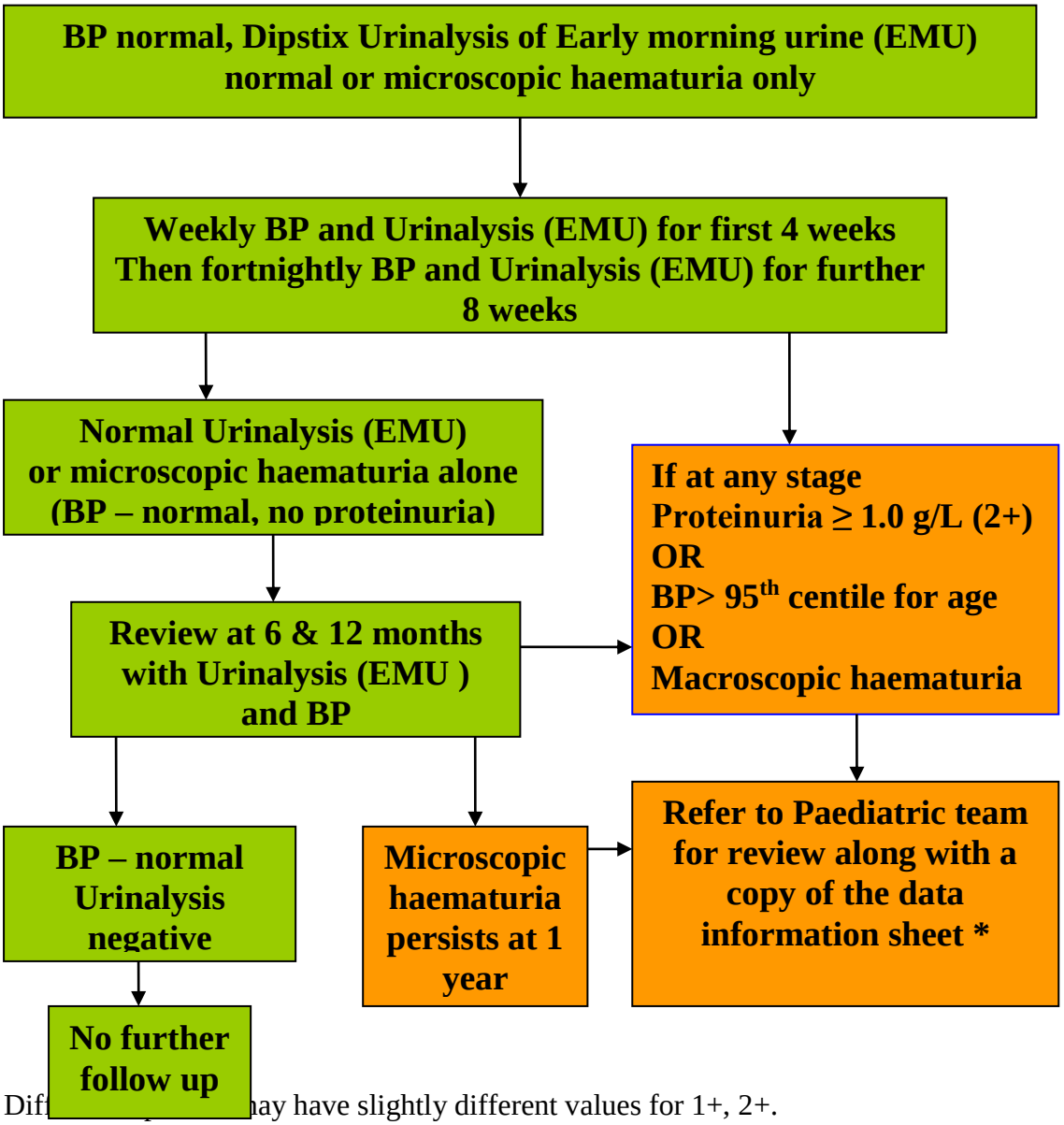
Appendix 1: Follow up algorithm for GP

Printout, complete and send it to GP along with the discharge letter and also give a copy to parent or carer to be submitted to GP surgery. *Please remember to file a copy in the notes as well.*

Patient Name: DOB: Hospital number: Name of G.P	Weight: Kg Height : Metres Systolic BP (95th Centile for age and height – **Please document legibly**) mm of Hg



Diff ay have slightly different values for 1+, 2+.
Proteinuria is ≥ 0.3g/L (At RGH – Siemens Multistix 10 SG are used and proteinuria of 1+ is 0.3g/L)

Appendix 2

Printout and give *a copy to GP and a copy* to *parent or carer*, regarding visits to GP.

Your child has been diagnosed with Henoch Schonlein Purpura. You need to attend your surgery for Blood Pressure and Urine testing of early morning urine. A copy of the data information sheet is enclosed. Please have your copy of data information filled up as well, during each visit to your GP.

* Data information sheet (A copy each to GP and parent or carer)

Week	Date	Systolic B P (in mm Hg)	Urine dipstix - Protein (g/L)	Urine dipstix - Blood
1				
2				
3				
4				
6				
8				
10				
12				
6 months				
12 months				

References

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