Henoch Schönlein purpura

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Henoch–Schönlein purpura

E J Tizard,1 M J J Hamilton-Ayres2

Henoch–Schönlein purpura (HSP) is the common-est vasculitis of childhood. The first description of this disorder was probably that of a young boy with “bloody points” over the shins of his legs, abdominal pain, blood in the stools and urine and painful subcutaneous oedema, described by William Heberden in 1801. In 1837 Johann Schönlein described the association of purpura and joint pain as “Peliaosis rheumatica”. Eduard Henoch, Schönlein’s former student, noted gastrointestinal involvement in association with purpura and arthritis in 1868 and subsequently he recorded renal involvement too.1

DIAGNOSIS

Until recently the 1990 American College of Rheumatology criteria for HSP were the most commonly used criteria for diagnosis.2 This classification was modified but has now been superseded by the European League Against Rheumatism (EULAR) and Paediatric Rheumatology European Society (PReS) consensus criteria for the classification of childhood vasculitis. The group reached consensus that the classification of HSP should be the finding of palpable purpura in the presence of one of either diffuse abdominal pain, a biopsy showing predominant IgA deposition, arthritis or arthralgia and/or renal involvement (any haematuria and/or proteinuria) (table 1).3

EPIDEMICOLGY

Henoch–Schönlein purpura is found in all age groups, from the age of a few months to late adulthood but it is more common in young children, with over 50% under 5 years and over 75% under 10 years. The clinical features of HSP are often atypical at extremes of age and the illness is more severe in adults while children under 2 years old are less likely than older children to have nephritis or abdominal complications. The reported incidence of HSP varies from 10–20.4 per 100 000 children.4–7 It is highest in younger children with 22.1/100 000 in the under 14 years and 70.3/100 000 in the 4–7 year age group being reported in one series from the West Midlands.4 In this study there was a significantly lower annual incidence in black children compared with white or Asian children. A male predominance is found in most series in a ratio of up to 2:1 but female predominance has also been reported. HSP is more common in the winter, autumn and spring than the summer, which supports the view that an infectious trigger may have a role in its pathogenesis.8–9

In a large Italian cohort two thirds of patients had a possible infective trigger before the onset of HSP, 63/150 had a respiratory tract infection and 37/150 had other infections or fever.9 Many organisms have been implicated in precipitating HSP but Group A β hemolytic streptococcus has been the commonest organism cultured in up to 36% of those tested in one series.6 Other organisms that have been identified include hepatitis A and B, CMV, HIV, adenovirus, Mycoplasma, herpes simplex, helicobacter pylori, toxocara canis, human parvovirus B19, varicella, and scarlet fever but many of the reports are anecdotal. HSP has also followed vaccinations including measles, mumps, rubella (MMR), pneumococcal, influenza, meningococcal and hepatitis B.6–9 Various drugs have been implicated as triggers although none has been proved to be associated in controlled studies.

CLINICAL FEATURES

It is the classical rash which often precipitates presentation of patients with HSP. Other system involvement may be present from the onset, or alternatively an evolving picture may develop over the course of several days to several weeks. In one study abdominal pain or arthritis preceded the rash by 1–14 days in 43/100 patients.8 As many cases follow an upper respiratory tract infection, the onset of the disorder may be accompanied by systemic symptoms such as fever and malaise.

Skin

The typical rash is of palpable purpura often symmetrically distributed over the extensor, dependent surfaces of the lower limbs and buttocks. It may involve the arms, face and ears but usually spares the trunk. The purpura range from petechiae to large ecchymoses and may be preceded by urticarial or erythematous, maculopapular lesions. Severe bullous lesions are rare in children, occurring in about 2% of patients (figs 1–3).

Gastrointestinal involvement

The reported incidence of gastrointestinal involvement is generally between 50–75% of cases with the most common presentation being colicky abdominal pain.8–10 Other symptoms include

Table 1 Classification criteria for Henoch–Schönlein purpura

<table>
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<tr>
<th>Palpable purpura (mandatory) in the presence of at least one of the following four features:</th>
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<tr>
<td>Diffuse abdominal pain</td>
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<tr>
<td>Arthritis (acute) or arthralgia</td>
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<tr>
<td>Renal involvement (any haematuria and/or proteinuria)</td>
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<tr>
<td>Any biopsy showing predominant IgA deposition</td>
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vomiting and gastrointestinal haemorrhage manifesting as overt bleeding or positive stools for occult blood. Massive gastrointestinal haemorrhage is rare, being reported in about 2% of patients. The symptoms result from oedema and haemorrhage of the bowel wall due to vasculitis. Intussusception is also a rare complication but is important to exclude as delay in management can result in ischaemic bowel. A protein losing enteropathy, pancreatitis and hydrops of the gall bladder are also described. It must be remembered that oedema secondary to hypoalbuminaemia is likely to be due to either nephrotic syndrome or a protein losing enteropathy or a combination of both.

Joints
Arthritis or arthralgia may be the presenting symptom in 15–25% of cases but overall up to 82% of patients have a degree of joint involvement during the illness. The arthritis usually affects the large joints of the lower limbs including knees, ankles, feet and hips although upper limbs may be affected too. In a retrospective review of 100 patients, 72% had feet and ankle involvement, 50% had knee involvement, 26% had hands and wrist involvement and 10% had elbow involvement. Symptoms include pain, swelling and decreased range of movement and although joint involvement can be debilitating it does not result in permanent damage.

Renal
Renal involvement in HSP is reported to occur in 12–92% of cases but in unselected series it is more commonly between 20–60%. Renal disease is manifest as haematuria, proteinuria, nephrotic syndrome/nephritis, renal impairment and hypertension. This develops within four weeks in 75–80% and within three months in 97–100% of cases. However, a few cases will develop even years after the initial presentation. In unselected series the incidence of severe renal disease including acute nephritis, nephrotic syndrome or renal impairment is 5–7%. Hypertension may be found in the presence of renal involvement. However, occasionally patients with HSP may develop hypertension without evidence of renal involvement. If it does not resolve as the HSP resolves further investigations are warranted.

Neurological
Neurological symptoms are rare although non-specific headache followed by subtle encephalopathy with minimal changes in mental status, such as labile mood, apathy and hyperactivity may be more common than previously thought. In a prospective study 31% of 26 patients had headache and 46% had abnormal EEGs. Seizures occurred in 2 out of 100 patients in one series. Subdural haematoma, subarachnoid haemorrhage, cerebellar haemorrhage, intraparenchymal bleeding and infarction have also been documented in case reports.

Pulmonary
Pulmonary involvement in HSP is rare. Although it occurs more often in adults, isolated case reports have been published describing pulmonary complications in children. The most common and severe manifestation is diffuse alveolar haemorrhage.
(DAH) but it may present as interstitial pneumonia or interstitial fibrosis.

Urological
Orchitis is a relatively common finding in boys with HSP being reported in up to 24%. In a series of 93 boys with HSP, 27 had involvement of the male genital system. Twenty-two of these had a swollen and tender scrotum, 10 bilateral and 12 unilateral and five had swelling of the penis. Three of the 22 patients with scrotal swelling were investigated radiologically and eight had surgical exploration. None of these patients had torsion but surgical exploration remains indicated if the diagnosis of the syndrome is not beyond doubt and torsion cannot be excluded on clinical grounds. Genital swelling may also be due to more generalised oedema secondary to hypoalbuminaemia from renal or gastrointestinal involvement.

INVESTIGATIONS
There are no specific laboratory tests that have been shown to be helpful in making the diagnosis which is usually based on clinical features. The investigations are aimed at excluding other diagnostic possibilities and assessing the extent of organ involvement such as the kidneys.

Initial investigations should include a full blood count, clotting, urea, electrolytes and creatinine, liver and bone profile and urinalysis to identify haematuria and proteinuria. If proteinuria is identified on dipstick then an early morning protein:creatinine ratio should be requested. If the diagnosis is in doubt then a full autoimmunity profile (AIP) including ANA, dsDNA, ANCA, immunoglobulins, C3 and C4 should be performed.

These investigations may show anaemia, leucocytosis, a raised erythrocyte sedimentation rate and in a few, abnormal renal or liver function tests. Thrombocytosis has been associated with more severe disease. Standard coagulation studies are usually normal; however Factor XIII activity is usually reduced and associated with more severe disease, but is currently not recommended in routine assessment.

IgA is elevated in half the children; however there is no correlation with disease severity. C3 and C4 have been reported to be low in a few patients but in general the basic immunological investigations are normal.

If the child is systemically unwell evidence for a site of infection should be sought with blood cultures, swabs, urine for microscopy and culture and a chest x-ray if clinically indicated. Evidence of recent streptococcal infection may be indicated by raised ASOT and antiDNase B titre. If renal disease is present it is also appropriate to measure these as sometimes a post-streptococcal glomerulonephritis may complicate the picture. Viral investigations (serology and throat swabs) may reveal a precipitating cause of HSP and should be considered in the unwell child but are unlikely to alter management.

SPECIFIC ORGAN INVOLVEMENT MAY WARRANT MORE DETAILED INVESTIGATIONS
Renal
If there is evidence of renal involvement, including haematuria, proteinuria, renal impairment or hypertension the pathway suggested in figure 4 is recommended.

Abdominal ultrasonography may show thickening of both small and large intestinal walls. As the most common form of intussusception is within the small bowel, its frequent inaccessibility to demonstration by contrast enema means that ultrasonography is the investigation of choice.

PATHOLOGY
In the skin there is a leucocytoclastic vasculitis with perineural infiltration of polymorphs and mononuclear cells. There may be necrosis of small blood vessels and platelet thrombi. IgA is found in most purpuric lesions and can also be found in non-affected areas. In practice skin biopsies are rarely required to make the diagnosis but may be useful in severe, atypical or recurrent disease when the diagnosis is in doubt.

The renal lesion of Henoch–Schoenlein nephritis is indistinguishable from that of IgA nephropathy characteristically showing a focal and segmental proliferative glomerulonephritis. The proliferation of extracapillary cells may result in crescent formation. Findings are graded according to the classification of the International Study for Kidney Diseases in Children (ISKDC). Immunofluorescence usually reveals mesangial IgA with IgG, C3 and fibrin. It is the non-renal clinical features that differentiate IgA nephropathy from HSPN.

Indications for renal biopsy include acute nephritic syndrome/acute renal impairment or nephrotic syndrome/nephrotic range proteinuria (urine protein:urine creatinine ratio >250 mg/mmol) for 4–6 weeks (fig 4).

IMMUNOPATHOLOGY
Although the aetiology of HSP is not clear there is evidence to support immunopathological mechanisms. Widespread abnormalities in IgA have been described including altered levels of IgA, increased levels of IgA class Ab such as IgA RF, IgA ANCA, IgA immune complexes and IgA deposition in renal and skin biopsies. It has been suggested that IgA ANCA could have a role in the diagnosis of HSP. IgA occurs in two isotypes—IgA1 and IgA2. Sixty per cent of IgA in secretions is IgA2 and generally polymeric, while serum IgA is predominantly IgA1 and 90% is monomeric. However in HSP there is predominantly deposition of polymeric IgA1 in skin, gastrointestinal and glomerular capillaries.

In patients with HSP-associated nephritis there have been reports of significantly elevated levels of abnormally glycosylated IgA1 compared with those without nephritis and controls. This abnormality has also been shown in adults with...
IgA nephropathy supporting the view of a common immunogenetic pathway in IgA-mediated glomerular disease.

Clinically the likelihood of a common pathogenesis is supported for example by the cases of patients with IgA nephropathy developing HSP and of simultaneous development of IgA nephropathy and HSP within sibships.25

GENETIC POLYMORPHISMS

A number of studies have tried to identify genetic polymorphisms that may be associated with the development of HSP or associated with disease severity or development of HSP nephritis.

Interleukin 1β expression has been found in skin biopsies of patients with HSP and a polymorphism of this gene has been associated with severity of nephropathy and renal sequelae.26 Polymorphisms of cytokine genes may mediate an abnormal inflammatory response leading to severe sequelae.

Investigation of the association of renin-angiotensin system gene polymorphisms with susceptibility to HSP and HSP nephritis (HSN) has produced conflicting results,27 28 with the polymorphisms of the ACE gene and angiotensinogen (Agt) gene possibly influencing the risk of developing HSP and Agt being shown to be associated with HSN. However further studies are needed to investigate this finding.

Differential Diagnosis

The diagnosis of HSP is usually obvious due to the characteristic rash. However, in the event of an atypical picture other conditions should be considered. These include other forms of vasculitis, particularly small vessel vasculitis such as microscopic polyarteritis, Wegener’s granulomatosis and isolated cutaneous leukocytoclastic vasculitis. Systemic lupus erythematosus (SLE) may also be associated with a similar vasculitic rash. Some of the vasculitides and SLE will be distinguished by the presence of ANCA or ANA/dsDNA respectively.

Children with HSP are usually systemically well whereas the child with septicaemia is generally clinically obvious. However, if there is any doubt systemic antibiotics should be given pending results of cultures. Other clotting disorders or thrombocytopenia should be identified through appropriate haematological investigations.

Acute haemorrhagic oedema of infancy (AHEI) is sometimes considered a variant of HSP.29 There are large purpuric lesions in a target-like pattern with oedema mainly affecting the face, auricles and extremities. Although the histological findings are similar only a third have IgA deposition and it is generally regarded as a distinct entity which rarely involves other organs.
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. Therefore drug therapy is aimed at more rapid symptomatic relief, except in the case of significant renal disease or severe organ involvement such as cerebral disease when treatment may have a significant impact on long-term outcome.

Gastrointestinal disease

Steroids are often used for the relief of abdominal pain although until recently there have been no randomised controlled studies on which to base this treatment. In a retrospective analysis children treated with 2 mg/kg prednisolone were more likely to have resolution of their abdominal pain than those on no treatment in the first 24 h (44% vs 14%) but by 72 h pain had resolved in 75% of both groups. Other retrospective studies and case series have also suggested a benefit of steroids in the treatment of abdominal pain. In one prospective, randomised, placebo controlled study of oral prednisolone in 40 children there was no significant difference in the development of gastrointestinal complications, although two in the placebo group developed intussusception compared with none in the steroid group. There was also a tendency for abdominal symptoms to be more prolonged in the prednislolone group. In the most recently published prospective study, prednisolone 1 mg/kg daily (maximum dose 50 mg) for two weeks with subsequent weaning over two weeks was effective in reducing the intensity of abdominal pain and the duration by a mean of 1.2 days. In severe gastrointestinal disease associated with protein-losing enteropathy a combination of intravenous methylprednisolone (10 mg/kg (max dose 500 mg) daily for 5 days, followed by 2 mg/kg oral prednisolone (max dose 80 mg) daily and weaning) and an elemental diet has been successful (personal experience).

Severe gastrointestinal haemorrhage has also been treated with high dose methylprednisolone (1 g × 3 days) followed by 40 mg prednisolone daily for a week in a 15-year-old boy. It is important to exclude gastrointestinal complications which may be exacerbated by steroids, before treatment and, in view of the association of gastrointestinal haemorrhage with non-steroidal anti-inflammatory drugs, it is recommended that these should not be used in combination with steroids in HSP.

Abdominal pain usually settles within a few days with or without treatment. Occasionally chronic abdominal pain may persist and methotrexate has been reported to be a useful steroid sparing agent in this situation. Mycophenolate has also been used for unresponsive and recurrent abdominal pain. However the potential gastrointestinal side effects of Mycophenolate should be considered.

Joint involvement

Joint pain is often managed with non-steroidal anti-inflammatory drugs but in severe cases steroids have been used. In one prospective study prednisolone appeared to reduce the severity of pain compared with the placebo group. Non-steroidal drugs should, however, be avoided in the presence of renal disease.

Skin involvement

Skin disease rarely requires specific treatment but bullous lesions have anecdotally been reported to respond to steroids. Dapsone, an anti-leprotic drug, has also been shown to be of benefit as a steroid sparing agent, particularly for skin, but has no effect on renal disease. A combination of aspirin and colchicine has also been used for chronic skin and joint disease.

HSP nephritis

Many have looked for prognostic factors which might help to select those patients who could benefit from treatment. The natural history of HSP is that most patients make a good recovery with the incidence of severe long-term morbidity/mortality being less than 5%. However, long-term renal impairment occurs in 19.5% of those with a history of nephritic or nephrotic syndrome. In the past HSP nephritis (HSPN) has been reported to account for up to 15% of children with end-stage renal failure (ESRF) (now termed chronic kidney disease (CKD) stage 5). More recent data show that HSPN accounts for 1.8–3% of children with CKD stage 5, possibly suggesting that more aggressive treatment may have had a beneficial impact on the outcome.

The presence of nephrotic/nephritic syndrome and renal impairment has been shown to be associated with a poor prognosis. Tarshish reported the outcome of 79 patients with heavy proteinuria and ISKDC grade III histopathological changes, some of whom were treated with cyclophosphamide as part of a prospective trial. Only 5/28 with nephrotic level proteinuria and severe histopathology recovered completely and no patient with >50 crescents on renal biopsy recovered completely. In a review of 114 patients with a diagnosis of HSPN 69 patients with normal and 25 with minor urinary abnormalities were designated to be the favourable outcome group, and 15 with active renal disease and five with renal failure the unfavourable group. Nephrotic syndrome, decreased Factor XIII activity, hypertension and renal failure at onset were more frequent in the unfavourable group. Other risk factors for development of renal disease include older age, abdominal pain and persisting purpura. However, in a follow-up study of 219 adults and children Coppo found, on multivariate analysis, that persisting proteinuria as opposed to disease markers at onset is the best predictor of outcome.

In a Finnish study of 19 patients with nephrotic range proteinuria the initial biopsy was not...
predictive of outcome. Although severe changes on early biopsy may lead to institution of successful treatment, a biopsy that is too early may be falsely reassuring and re-biopsy should be considered if the clinical condition does not improve.49

The treatment of HSPN remains controversial. In view of the evidence that severe proteinuria is a risk for long-term renal damage there have been attempts to prevent the development of renal disease with the early treatment with steroids. In a large study of 164 children with HSP but without evidence of nephropathy at the onset, patients were allocated alternately to receive prednisolone 1 mg/kg for two weeks or no treatment.50 None of the treated group and 11.9% of the untreated group developed evidence of nephropathy within six weeks of the onset of HSP. Two others developed nephropathy at two and six years after the initial illness. The difference in the two groups was highly significant suggesting a beneficial effect of steroids. Another study in support of the use of steroids was that of Kaku.47 In this retrospective study of 194 patients without nephritis at onset, 79 were treated with steroids for symptoms of abdominal pain or joint pain. A multivariate analysis identified the use of steroids as having a role in reducing the development of renal disease. In contrast Saulsbury found no benefit in the use of steroids in the prevention of nephritis. However, in this report steroids were used to treat patients with abdominal pain and as these are at higher risk of developing renal disease it may have obscured the benefit of steroids.51

The first randomised placebo controlled study of prednisolone in early HSP, which included 40 children, did not show a benefit in the use of steroids on the risk of renal involvement at one year.52 Ronkainen’s larger randomised, placebo controlled study of 171 patients showed no significant benefit of steroids in the development of renal symptoms overall.53 However, there was a slightly more rapid resolution of renal symptoms in the steroid treated group and there appeared to be a specific benefit in a small subgroup of children over 6 years who had or developed renal symptoms in the first month. A recent systematic review of the use of corticosteroids in HSP concluded that based on the prospective studies from Ronkainen,53 Huber54 and Mollica50 there was a benefit of steroids on reducing the persistence of renal disease.52 However the study by Mollica only included those without renal disease at presentation and the follow up in the Ronkainen study was only 6 months. In addition, in the latter study the definition of renal disease included those with microscopic haematuria alone and therefore renal disease of less significance in terms of long term outcome. The authors of this review suggested that larger prospective studies should be performed.

The largest prospective study to date which has been undertaken in SW England and Wales randomised 555 children to receive either prednisolone or placebo for two weeks. The preliminary results of this study have not shown any benefit of early steroids in the prevention of renal disease at one year.53 However, subgroup analysis is still awaited (Dudley et al, unpublished data). Data from this study was not available at the time of the systematic review.

For those with established renal disease there is again a lack of clear evidence of the benefits of treatment. This is in part due to the fact that relatively few patients have severe renal disease at onset and therefore randomised controlled trials are difficult to perform. However there are a number of uncontrolled case series showing some benefit of immunosuppressive therapy.

In a study of nine Japanese children with severe proteinuric HSPN associated with ISKDC Grade III or IV histological changes, early treatment with oral prednisolone and cyclophosphamide resulted in a favourable outcome in all patients.44 However, this is an uncontrolled small study with a relatively short follow-up period. Foster also demonstrated a benefit of steroids and azathioprine in patients, all of whom had significant proteinuria or nephrotic syndrome and three of whom had renal impairment at onset.55 There has also been anecdotal evidence in support of the use of cyclosporin.56

The evidence for the efficacy of treatment in the patients with the most severe disease is limited but the significant long-term consequences in some patients make this an important area to consider. There is again some evidence for the benefits of intensive immunosuppression. Niaudet treated 38 children with severe HSPN (nephrotic syndrome and/or 50% crescentic nephritis) with methylprednisolone; seven also had cyclophosphamide with or without plasma exchange. Four of these 38 compared with 11/29 historical untreated controls developed end stage renal failure.45 A good outcome was found in 11 of 12 patients with rapidly progressive glomerulonephritis, nine of whom had 60–90% crescents on biopsy, following aggressive treatment with methylprednisolone, cyclophosphamide, oral prednisolone and dipyridamole.57 Plasma exchange also appears to have a possible benefit when used in conjunction with steroids and cyclophosphamide if treatment is started early in the disease,58 as has a combination of methylprednisolone and urokinase with or without cyclophosphamide.59 In contrast a randomised study of 56 patients with severe nephritis showed no benefit of cyclophosphamide on renal outcome.60

In this severe group there is some, albeit uncontrolled, evidence to suggest that aggressive therapy may be useful. In view of the potential long-term consequences, treatment is often instituted in these patients and therefore timely discussion/referral to a paediatric nephrologist is essential (fig 4). Intensive treatment including plasma exchange may also have a role in non-renal involvement such as neurological disease.60

The antihypertensive and renoprotective effects of angiotensin-converting enzyme inhibitors (ACEI) are well established in adults with hypertension and/or chronic renal failure. However, the experience in children is less well documented. One
study has shown a benefit of ACEI in the treatment of hypertension and reducing proteinuria in 397 children with mild to severe chronic renal failure. In addition, a study of 66 children and adults with IgA nephropathy but not HSPN demonstrated a benefit of ACEI on the progression of IgA nephropathy. A small case series showed a stabilisation of renal function in children with HSPN treated with fish oil with or without ACEI therapy. Therefore by extrapolation, there may be a benefit in treating patients with evidence of chronic disease following HSP, particularly with significant proteinuria/hypertension, with an ACEI. This should be discussed with a paediatric nephrologist.

There are no convincing data to date to support the use of fish oil in HSPN.

PROGNOSIS
Henoch–Schönlein purpura is generally a self-limiting disease but about 38% have recurrence of symptoms. Saulsbury reported 1–6 recurrences per child occurring from two weeks to 18 months after initial resolution of symptoms. Two per cent had recurrences after four months and patients with renal disease were more likely to have recurrences.

The long-term outlook for children with HSP is predominantly related to the renal disease. In two unselected series chronic renal disease developed in up to 1.4% of children at about eight years from diagnosis. Mortality is also very unusual occurring in <1% in one series. Although no feature is absolutely predictive of renal outcome, many studies confirm that features including nephrotic syndrome, renal impairment, hypertension and decreased Factor XIII at presentation are associated with a poorer prognosis.

In a follow-up study of 78 children who had had HSPN the severity of the clinical presentation and the renal biopsy were related to final outcome but nothing reliably predicted individual outcome. Forty per cent of those with nephritis or nephrotic syndrome at presentation followed for a mean of 23.4 years had hypertension or impaired renal function at follow-up whereas 82% of those with minimal involvement initially were normal at follow-up. However some patients who had apparently recovered initially deteriorated in the longer term follow-up. More optimistically about 50% of patients with severe disease at presentation had a favourable prognosis.

In this study 16/44 pregnancies were complicated with proteinuria and/or hypertension. This finding was confirmed in another long-term follow-up study of 52 children with HSP in which 16/23 (70%) of subsequent pregnancies were similarly complicated and five women had poor renal outcome.

In view of the potential late deterioration of renal disease it is recommended that all patients with a history of HSPN should have lifelong follow-up. In contrast, those with normal urinalysis and blood pressure at six months are unlikely to develop significant nephritis. A systematic review of 12 studies including 1153 children concluded that of the children developing renal involvement 97% did so within six months of presentation. No long-term renal impairment occurred after normal urinalysis. Therefore those with no evidence of renal disease can be safely discharged after 6–12 months. In cases where there is recurrence of non-renal symptoms (for example, rash with or without abdominal pain or arthritis) surveillance should continue until six months from the start of the last relapse.

CONCLUSION
Henoch–Schönlein purpura is the most common vasculitis of childhood which is self-limiting in the majority of cases. In the absence of either renal disease or ongoing extra-renal symptoms, children may be discharged at six months from diagnosis. In contrast, in those with a history of nephritis the risk of long-term renal damage warrants long-term surveillance with particular vigilance in women during pregnancy. Steroids appear to have a role in managing the symptoms of abdominal pain but are probably not indicated to prevent nephropathy. In view of the small number of patients with severe renal disease multicentre/international studies are required to establish the role of immunosuppressive therapy. On the current available evidence patients with significant renal or extra-renal disease should be assessed and considered for treatment with steroids with or without immunosuppressive therapy.

Competing interests: None.

REFERENCES


