

Henoch-Schönlein purpura (HSP) is a systemic vasculitis that causes the blood vessels in the skin to become inflamed, causing red spots. When the blood vessels in the skin get inflamed, they can bleed, causing a rash that is called purpura. This rash is typically seen on the lower legs or arms. The specific skin lesion is characterized by the tissue deposition of an immune system product, called IgA immunoglobulin, which is also found in kidneys of patients with a renal disease, called IgA nephropathy.

HSP occurs more often in children than in adults, and many cases follow an upper respiratory tract infection (infection in your sinuses and /or lungs). Half of affected children are under age five, although kidney involvement is more likely to be severe in older children. Compared to children, adults had more severe and frequent kidney involvement.

Symptoms occur over a period of days to several weeks: skin rash, joint aches and pains, usually in knees and ankles, occasional swelling, abdominal pain and renal disease manifesting mostly as hematuria (blood in your urine), proteinuria (abnormal excretion of proteins in urine), edema (swelling) or alteration in the volume of urine. The hematuria may be noticed as red or tea-colored or cola-colored urine or the amount may be so small that it can only be seen under a microscope. The brain or the lung may also be involved in HSP.

Gastrointestinal symptoms are present in the majority of patients including abdominal pain that is frequently associated with vomiting. The pain typically develops within eight days of the appearance of the rash. Bleeding of the gastrointestinal tract presenting with black or bright red color in stools is seen in these patients. Although rare, more serious complications may develop like intussusception, a situation in which one portion of the bowel slides into the next creating an obstruction in the bowel, leading to swelling, inflammation, and decreased blood flow to the intestines involved or inflammation of other organs leading to pancreatitis, cholecystitis, and enteronephrotic pathy.

Renal (kidney) involvement is common, occurring in 30-70 % of patients. Kidney disease is usually noted after the onset of systemic symptoms. More marked findings may also occur including nephrotic syndrome, a situation characterized by abnormal excretion of proteins and lipids in urine, swelling (edema), low level of albumin in blood and hyperlipidemia. High blood pressure (hypertension) and acute kidney failure may also be seen. Worsening of the kidney symptoms and biopsy-confirmed worsening of the kidney lesions may be observed in patients with repeated attacks of rash or hematuria (blood in the urine).

Even though the symptoms of HSP make it easier to diagnose in children, confirmation of the diagnosis of HSP requires evidence of tissue deposition in the skin or kidney of IgA immunoglobulin. Renal biopsy is another method to establish the diagnosis, but is reserved for patients in whom the diagnosis is uncertain or in whom there is evidence of more severe renal involvement.

The overall **outcome** is good in most patients. All of the manifestations of active HSP usually resolve spontaneously, although recurrent episodes of skin rash and hematuria may be seen. Among those with kidney involvement, only a minority have persistent disease. The kidney prognosis is excellent in most patients. However some patients will have persistent protein in their urine, high blood pressure, and renal insufficiency. It is estimated that HSP accounts for approximately 3% of cases of end-stage kidney disease in children. Poor renal prognosis is more common among those with the nephrotic syndrome, renal insufficiency, and more advanced findings on biopsy.

Recurrences are common, occurring in approximately one-third of patients. Since complete recovery occurs in 94% of children and 89 % of adults, respectively, most patients receive no specific therapy. There is suggestive evidence that corticosteroids enhance the rate of resolution of the arthritis and abdominal pain, although they do not appear to prevent recurrent disease.

However, specific **treatment** is recommended in patients with marked proteinuria (protein in the urine) and/or impaired kidney function during the acute episode. A kidney biopsy can be performed to reveal the severity of the lesions which appears to be the best indicator of prognosis. Advanced disease, usually defined as crescentic nephritis, is treated with a regimen consisting of pulse intravenous methylprednisolone followed by oral prednisone.

Other regimens that have been evaluated in children with kidney disease include corticosteroids and azathioprine and multidrug regimens such as corticosteroids, cyclophosphamide, and dipyridamole, or corticosteroids, cyclophosphamide, heparin/warfarin, and dipyridamole. However, since spontaneous recovery is often observed in these patients, it remains unknown whether these regimens are superior to no or less aggressive therapy.

Plasmapheresis has also been used in a number of patients with severe disease although its efficacy is uncertain. Intravenous immune globulin has been tried in a small number of patients with heavy proteinuria and a progressive decline in kidney function.

Kidney transplantation can be performed in those patients who progress to end-stage kidney disease, although recurrent disease can occur. This appears to be more likely in patients with aggressive initial disease who progressed to end-stage kidney disease in less than three years after the onset of HSP. Therefore it is recommended the transplantation to be delayed for 12-24 months after the disappearance of the rash. Some observations suggest that the risk of recurrent disease also may be higher in living-related donors.