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HENOCH- SCHOENLEIN PURPURA

What is it?

Henoch-Schoenlein Purpura (HSP) is a condition in which small blood vessels get inflamed. This inflammation is called vasculitis and usually affects the small blood vessels in the skin, bowel and kidneys. The inflamed blood vessels may bleed into the skin causing a deeply red or purple rash called purpura. They can also bleed in the intestine or kidneys causing blood-stained stools or urine (hematuria).

HSP was named after Drs Henoch and Schoenlein who both described it independently more than one hundred years ago.

How common is it?

HSP, although not a frequent illness of childhood, is the most common systemic vasculitis in children aged between five and 15 years. It is more common in boys than in girls with a ratio of 2:1.

There is no ethnic or geographic distribution for this disease. Most cases in Europe and the Northern Hemisphere occur in winter, but some cases are seen during fall or spring.

What are the causes of the disease?

No one knows what causes HSP.

Infectious agents, such as viruses and bacteria, are thought to be potential triggering factors for the disease, because it often appears following an infection. However, HSP has also been seen following prescription of medicines, insect bites, exposure to cold, chemical toxins and the intake of specific foods that can cause allergies. For these reasons the term allergic purpura was previously used, as HSP was thought to be an allergic reaction to all these agents. In some countries, it is also called rheumatoid purpura due to symptoms related to joints and muscles (see the section on symptoms below).

The presence of specific products of the immune system, such as Immunoglobulin A (IgA), in lesions of HSP, suggest that an abnormal response of the immune system attacks small blood vessels in the skin, joints, the gastrointestinal tract, kidneys, central nervous system or testis and causes the disease.

Is it inherited? Is it contagious? Can it be prevented?

HSP is not an inherited disease. It is not contagious and cannot be prevented.

What are the main symptoms?

The main symptom is a characteristic skin rash present in all patients with HSP. The rash usually begins with small hives, red patches or red bumps, which change to a purple bruise over time. It is called palpable purpura, because the raised skin lesions can be felt. Purpura usually covers the lower extremities and buttocks, although some lesions may appear elsewhere in the body (upper limbs, trunk etc).

Painful joints (arthralgia) or painful and swollen joints with limitation of movement (arthritis), are found in the majority of patients (>65%). Arthralgia and arthritis are accompanied by soft tissue swelling and tenderness located near and around the joints. Soft tissue swelling in hands and feet, forehead and scrotum may occur early in the disease, particularly in very young children.

The symptoms effecting joints are temporary and disappear within a few days.

When the vessels of the bowel become inflamed, abdominal pain is present in more than 60% of cases. The pain appears intermittent around the umbilical cord and may be accompanied by mild or severe gastrointestinal bleeding (haemorrhage). Very rarely an abnormal folding of the bowel called intussusception can occur causing obstruction of the intestine and may need surgery.

When the vessels of kidneys become inflamed, they may bleed (in about 20-35% of patients) and mild or severe hematuria and proteinuria (protein in urine) may occur. Usually, kidney problems are not serious. In rare cases renal disease may last for months or years and may progress to renal failure (one to five %). In such cases, a consultation with a nephrologist (kidney specialist) and cooperation with the patient's physician are necessary.

The symptoms described above usually last for about four to six weeks. Occasionally, they may precede the appearance of skin rash by a few days. They may appear simultaneously, or gradually in a different order.

Other symptoms, such as seizures, brain or lung haemorrhage and swelling of the testis, which are due to inflammation of the vessels in other organs of the body are rarely seen.

Is the disease the same in every child?

The disease is more or less the same in every child, but the extent of cutaneous and organ involvement may vary among patients. HSP can present as one single episode, or several recurrent relapses.

Is the disease in children different from the disease in adults?

The disease in children is not different from the disease in adults, but it is rarer.

How is it diagnosed?

Diagnosis of HSP is mainly clinical and based on the classic purpuric eruption, usually confined to the lower limbs and buttocks. Other diseases that can cause a similar picture have to be excluded.

What laboratory and other tests are useful?

There are no specific tests that confirm the diagnosis of HSP. Erythrocyte sedimentation rate (ESR), or C-reactive protein (a measure of systemic inflammation) may be normal or elevated. Blood in stools may be positive due to intestinal haemorrhage. Urinalysis should be performed during the course of the disease to detect kidney involvement. Low-grade hematuria is common and resolves with time.

A kidney biopsy is required if kidney involvement is severe (renal insufficiency or heavy proteinuria).

Can it be treated?

Most HSP patients do fine and do not need any medication at all.

Treatment, when needed, is mainly supportive with control of pain, either with simple analgesics (pain killers), or with non-steroidal anti-inflammatory drugs such as ibuprofen and naproxen when joint complaints are more prominent.

Administration of steroids (prednisone) is indicated in patients with severe gastrointestinal symptoms or haemorrhage and in rare cases with severe symptoms from other organs. If renal disease is severe, a renal biopsy (the removal of a small part of the effected tissue) has to be performed and, if indicated, a combined treatment with steroids and immunosuppressive drugs is started.

What are the side effects of drug therapy?

In most cases of HSP, drug treatment is not necessary, or given only for a short time and there are no severe side effects to be expected. In the rare cases with severe renal disease, that require prednisone and immunosuppressive drugs for a long time, drug side effects may be a problem (see therapy section).

How long will the disease last for?

The entire course of the disease is about four to six weeks. Half the children have at least one recurrence within six weeks, which is usually briefer and milder than the first one. Rarely, relapses can last longer. The majority of patients recover completely.

What kinds of periodic check-ups are necessary?

Urine samples should be checked several times during the disease and after HSP goes away in order to detect kidney problems as, in some cases, renal involvement may occur several weeks after the disease onset.

What is the long-term outcome of the disease?

In most children the disease is self-limited and does not cause long term problems. The small percentage of patients presenting with persistent or severe renal disease, may have a progressive course with possible renal failure.

What about school and sports?

During acute illness physical activity is usually limited, but after recovery the child is able to go to school again and lead a normal life. Vaccinations should also be postponed and rescheduled by the child's paediatrician.