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Henoch-Schoenlein Purpura

Version of 2016

1. WHAT IS HENOCH-SCHOENLEIN PURPURA

1.1 What is it?

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

1.2 How common is it?

HSP, although not a frequent illness of childhood, is the most common systemic vasculitis in children aged between 5 and 15 years. It is more common in boys than in girls (2:1).
There is no ethnicity preference or geographical distribution of the disease. Most cases in Europe and the Northern Hemisphere occur in winter, but some cases are also seen during fall or spring. HSP affects approximately 20 in 100,000 children per year.

1.3 What are the causes of the disease?

No one knows what causes HSP. Infectious agents (such as viruses and bacteria) are thought to be a potential trigger for the disease because it often appears after an upper respiratory tract infection. However, HSP has also been seen following prescription of medicines, insect bites, exposure to cold, chemical toxins and intake of specific food allergens.

HSP may be a reaction to an infection (an overly aggressive response from your child’s immune system).
The finding of deposition of specific products of the immune system such as Immunoglobulin A (IgA) in lesions of HSP suggests that an abnormal response of the immune system attacks small blood vessels in the skin, joints, gastrointestinal tract, kidneys and seldom central nervous system or testes and causes the disease.

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

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

1.5 What are the main symptoms?

The leading symptom is a characteristic skin rash, which is present in all patients with HSP. The rash usually begins with small hives; red patches or red bumps, which in time change to a purple bruise. 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 also appear elsewhere in the body (upper limbs, trunk, etc.).
Painful joints (arthralgia) or painful and swollen joints with limitation of movement (arthritis) - usually knees and ankles and less commonly wrists, elbows and fingers - are found in the majority of patients (>65%). Arthralgia and/or 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 joint symptoms are temporary and disappear within a few days to weeks.
When the vessels become inflamed, abdominal pain is present in more than 60% of cases. It is typically intermittent, felt around the belly button (umbilicus), and may be accompanied by mild or severe gastrointestinal bleeding (haemorrhage). Very rarely, an abnormal folding of the bowel called intussusception may occur, causing an obstruction of the intestine that may need surgery.
When the kidneys’ vessels become inflamed, they may bleed (in about 20-35% of patients) and a mild to severe haematuria (blood in the

urine) and proteinuria (protein in the urine) may occur. Kidney problems are usually not serious. In rare cases, renal disease may last for months or years and may progress to kidney failure (1-5%). In such cases, consultation with a kidney specialist (nephrologist) and cooperation with the patient's physician are needed.
The symptoms described above may occasionally 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 testes due to inflammation of the vessels in these organs, are rarely seen.

1.6 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 significantly from patient to patient.

1.7 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 occurs rarely in adults.

2. DIAGNOSIS AND TREATMENT

2.1 How is it diagnosed?

Diagnosis of HSP is primarily clinical and based on the classic purpuric eruption, usually confined to the lower limbs and buttocks, and generally associated with at least one of the following manifestations: abdominal pain, joint involvement (arthritis or arthralgia) and renal involvement (most often haematuria). Other diseases that can cause a similar clinical picture must be excluded. A skin biopsy is rarely needed for the diagnosis in order to show the presence of immunoglobulin A in histological examinations.

2.2 What laboratory and other tests are useful?

There are no specific tests that contribute to the diagnosis of HSP. Erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP, a measure of systemic inflammation) may be normal or elevated. Occult blood in the stools may be an indication of a small intestinal haemorrhage. Urinalysis should be performed during the course of the disease to detect kidney involvement. Low-grade haematuria is common and resolves with time. A kidney biopsy may be required if kidney involvement is severe (renal insufficiency or significant proteinuria). Imaging tests such as ultrasound may be recommended to rule out other causes of abdominal pain and to check for possible complications, such as a bowel obstruction.

2.3 Can it be treated?

Most HSP patients do fine and do not require any medication at all. Eventually, children can bed rest while symptoms are present. Treatment, when needed, is mainly supportive, with control of pain either with simple analgesics (painkillers) such as acetaminophen, or with non-steroidal anti-inflammatory drugs, such as ibuprofen and naproxen, when joint complaints are more prominent. Administration of corticosteroids (orally or sometimes intravenously) is indicated in patients with severe gastrointestinal symptoms or haemorrhage and in rare cases of severe symptoms involving other organs (i.e. testes). If renal disease is severe, a renal biopsy must be performed and, if indicated, a combined treatment with corticosteroids and immunosuppressive drugs is initiated.

2.4 What are the side effects of drug-therapy?

In most cases of HSP, drug treatment is not necessary or is administered only for a short time; hence, no severe side effects are expected. In rare cases, when severe renal disease requires the use of prednisone and immunosuppressive drugs for a long time, drug side effects may be a problem.

2.5 How long will the disease last?

The entire course of the disease is about 4-6 weeks. Half of children with HSP have at least one recurrence within a 6-week period, which is

usually briefer and milder than the first episode. Relapses rarely last longer. A recurrence is not indicative of the severity of the disease. The majority of patients recover completely.

3. EVERYDAY LIFE

3.1 How might the disease affect the child and the family's daily life and what kinds of periodic check-ups are necessary?

In most children, the disease is self-limited and does not cause long-lasting problems. The small percentage of patients who present persistent or severe renal disease may have a progressive course, with possible renal failure. In general, the child and family are able to lead a normal life.

Urine samples should be checked several times during the course of the disease and 6 months after the HSP is no longer present: this is to detect potential kidney problems as, in some cases, renal involvement may occur several weeks or even months after the disease onset.

3.2 What about school?

During acute illness, all physical activity is usually limited and bed rest may be needed; after recovery, children can go to school again and lead a normal life, participating in all the same activities as their healthy peers. School for children is equivalent to work for adults: a place where they learn how to become independent and productive young people.

3.3 What about sports?

All activities can be performed as tolerated. Therefore, the general recommendation is to allow patients to participate in sports activities and to trust that they will stop if a joint hurts, while advising sports teachers to prevent sport injuries, in particular for adolescents. Although mechanical stress is not beneficial for an inflamed joint, it is generally assumed that the small risk of damage is outweighed by the psychological impact of being prevented from playing sports with friends because of the disease.

3.4 What about diet?

There is no evidence that diet can influence the disease. In general, the child should observe a balanced, normal diet for his/her age. A healthy, well-balanced diet with sufficient proteins, calcium and vitamins is recommended for a growing child. Overeating must be avoided in patients taking corticosteroids, because these drugs may increase appetite.

3.5 Can climate influence the course of the disease?

There is no evidence that climate can affect the disease manifestations.

3.6 Can the child be vaccinated?

Vaccinations should be postponed and the time of missed vaccinations will be decided by the child’s paediatrician. Overall, vaccinations do not seem to increase the disease activity and do not cause severe adverse events in PRD patients. However, live attenuated vaccines are generally avoided because of the hypothetical risk of inducing infection in patients receiving high dose immunosuppressive drugs or biologics.

3.7 What about sexual life, pregnancy, birth control?

The disease presents no restrictions on normal sexual activity or pregnancy. However, patients taking medications should always be very careful about the possible effects of these drugs on a foetus. Patients are advised to consult their physician about birth control and pregnancy.