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Henoch-Schoenlein Purpura

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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.