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Behçet’s disease

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1. WHAT IS BEHCET

1.1 What is it?

Behçet’s syndrome, or Behçet’s disease (BD), is a systemic vasculitis (inflammation of blood vessels throughout the body) of unknown cause. The mucosa (tissue that produces mucous, which is found in the lining of the digestive, genital and urinary organs) and skin are affected and the main symptoms are recurrent oral and genital ulcers and eye, joint, skin, blood vessel and nervous system involvement. BD was named after a Turkish doctor, Prof. Dr. Hulusi Behçet, who described it in 1937.

1.2 How common is it?

BD is more common in some parts of the world. The geographical distribution of BD coincides with the historical ‘silk route’. It is mainly observed in countries of the Far East (such as Japan, Korea, China), Middle East (Iran) and Mediterranean basin (Turkey, Tunisia, Morocco). The prevalence rate (number of patients in the population) in the adult population is 100-300 cases/100,000 people in Turkey, 1/10,000 in Japan, and 0.3/100,000 in Northern Europe. According to a study conducted in 2007, the prevalence of BD in Iran is 68/100,000 inhabitants (2nd highest in the world after Turkey). Few cases are reported from the United States and Australia.
BD in children is rare, even in high risk populations. The diagnostic criteria are fulfilled before the age of 18 in approximately 3-8% of all BD patients. Overall, the age of disease onset is 20-35 years. It is equally distributed between females and males, but the disease is usually more severe in males.

1.3 What are the causes of the disease?

The causes of the disease are unknown. Recent research carried out in a large number of patients suggests that genetic susceptibility may have some role in the development of BD. There is no known specific trigger. Research into the cause and treatment is being carried out in several centres.

1.4 Is it inherited?

There is no consistent pattern of inheritance of BD, although some genetic susceptibility is suspected, especially in early onset cases. The syndrome is associated with a genetic predisposition (HLA-B5), especially in patients originating from the Mediterranean basin and the Far East. There have been reports of families suffering from this disease.

1.5 Why does my child have this disease? Can it be prevented?

BD cannot be prevented and its cause is unknown. There is nothing that you should have done less or more to prevent your child from getting BD. It is not your fault.

1.6 Is it infectious?

No, it is not.

1.7 What are the main symptoms?

Oral ulcers: These lesions are almost always present. Oral ulcers are the initial sign in about two-thirds of patients. The majority of children develop multiple, minor ulcers, indistinguishable from recurrent ulcers, which are common in childhood. Large ulcers are rarer and may be very difficult to treat.

Genital ulcers: In boys, the ulcers are located mainly on the scrotum and, less frequently, on the penis. In adult male patients, these almost always leave a scar. In girls, external genitalia are mainly affected. These ulcers resemble the oral ulcers. Children have fewer genital

ulcers before puberty. Boys may have recurrent orchitis (testicular inflammation).

Skin involvement: There are different skin lesions. Acne-like lesions are present only after puberty. Erythema nodosum are red, painful, nodular lesions, usually located on the lower legs. These lesions are more frequent among children before puberty.

Pathergy reaction: Pathergy is the reactivity of the skin of BD patients to a needle prick. This reaction is used as a diagnostic test in BD. After a skin puncture with a sterile needle on the forearm, a papule (raised round circular rash) or pustule (round raised pus-containing rash) forms within 24 to 48 hours.

Eye involvement: This is one of the most serious manifestations of the disease. While the overall prevalence is approximately 50%, it increases to 70% in boys. Girls are less frequently affected. The disease involves both eyes in most patients. Eyes are involved usually within the first three years after the disease onset. The course of the eye disease is chronic, with occasional flares. Some structural damage occurs as a result of each flare, causing gradual vision loss. Treatment is focused on controlling the inflammation, preventing the flares and avoiding or minimizing vision loss.

Joint involvement: Joints are involved in about 30-50% of children with BD. Usually ankles, knees, wrists and elbows are affected, and typically fewer than four joints are involved. The inflammation can cause joint swelling, pain, stiffness and restriction of movement. Fortunately, these effects usually last only a few weeks and resolves on their own. It is very rare for this inflammation to cause joint damage.

Neurological involvement: Rarely, children with BD can develop neurological problems. Seizures, increased intracranial pressure (pressure inside the skull) with associated headaches and cerebral symptoms (balance or gait) are characteristic. The most severe forms are seen in males. Some patients may develop psychiatric problems.

Vascular involvement: Vascular involvement is seen in about 12-30% of juvenile BD patients and can signal a poor outcome. Both veins and arteries may be involved. Any vessel size in the body can be involved; hence the classification of the disease as "variable vessel size vasculitis". The vessels of the calves are commonly affected, becoming swollen and painful.

Gastrointestinal involvement: This is especially common in patients from the Far East. Examination of the bowel reveals ulcers.

1.8 Is the disease the same in every child?

No, it is not. Some children may have mild disease with infrequent episodes of oral ulcers and some skin lesions, while others may develop eye or nervous system involvement. There are also some differences between girls and boys. Boys usually experience a more severe disease course, with more eye and vascular involvement, than girls. Besides the different geographic distribution of the disease, its clinical manifestations may also differ throughout the world.

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

BD is rare in children compared to adults, but there are more familial cases among children with BD than in adults. The disease manifestations after puberty are more similar to the disease in adults. More generally, in spite of some variations, BD in children does resemble the adult disease.