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WHAT IS BENTA DISEASE?

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B cell expansion with NF-κB and T cell anergy (BENTA) represents a newly defined entity in immunology. It is a primary immunodeficiency (PID). This rare genetic disorder is transmitted in an autosomal dominant manner and classified as a predominantly antibody deficiency by the International Union of Immunological Societies (IUIS). The cause of the disease is a gain-of-function mutation in the Caspase recruitment domain-containing membrane-associated guanylate kinase protein-1 (CARMA1 (CARD11)) gene. Clinically, the disease is manifested at an early age with hepatosplenomegaly, lymphadenopathy, anemia, susceptibility to frequent respiratory tract infections, and a low response to certain vaccines. Lymphadenopathies can be part of the clinical spectrum of several PIDs and can pose a significant diagnostic dilemma. Patients with this disease carry a risk of developing chronic B cell leukemia. Thorough family history is an important element in the assumption of diagnosis of BENTA disease. Treatment options of BENTA disease are still being considered. They can include splenectomy, application of monoclonal antibodies such as rituximab to deplete B cell reserve, wearing special spleen guards when playing sports, and antibiotics for infections. Because it can present a burden for families, psychological support and counseling may be necessary. Each physician should be informed about the existence of this disease so they can eventually recognize it in their medical practice.

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