

AUTOIMMUNE SKIN DISEASES

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INTRODUCTION

The skin is the largest organ in the human body, functioning as protection covering the underlying tissues and organs from impact, chemicals, and infections. It also maintains the body's normal temperature by regulating heat exchange with the environment, stores nutrients, detects touch, pressure, pain and temperature stimuli and relays the information to the nervous system, and excretes salts, water and organic wastes. With the skin being the largest organ in the human body comes disorders and diseases connected to it, some minor and other major. One of the major ones is autoimmune skin diseases which are diseases that affect the body by causing the body's immune system to attack healthy cells. It occurs when the body's immune system attacks its own healthy tissues, with autoimmune skin diseases autoantibodies attack skin cells or collagen tissues. Autoimmune skin diseases are no different as there are many diseases within the category. First discovered over a hundred years ago by Paul Ehrlich he proposed this idea as the existence of mechanisms to prevent the immune reaction against their own elements. These mechanisms prevent or stop the production of autoantibodies to avoid self-damage, he named this concept the *horror autotoxins*. The diseases include Pemphigus, Bullous pemphigoid (bp) diseases, Dermatitis herpetiformis (DH), Epidermolysis bullosa acquisita (EBA), Vitiligo, and Psoriasis. Autoimmune skin diseases tend to not have a specific or exact cause and are not fully understood by medical researchers. Although the cause is not clear, there are risk factors for many of the autoimmune diseases including genetic factors, infections, hormones, and drugs. Genetic factors are commonly polygenic, so when multiple genes combine it can increase risk, infections can trigger an autoimmune process by mimicking a self-antigen, certain drugs can precipitate various autoimmune skin diseases.

Autoimmune skin diseases affect anyone from women to men or children to adults. The way different autoimmune diseases affect a child can be differentiated from how it affects an adult. Pemphigus is an autoimmune skin disease that consists of various parts such as Pemphigus Vulgaris (PV), Pemphigus Foliaceus, Pemphigus erythematosus (PE), and more. This disease affects mostly middle aged to late adulthood and affects men and women equally. When it comes to how a patient gets Pemphigus can vary from drug reactions such as antibiotics to genetics or other illnesses that can cause the beginning of an autoimmune skin disease. The disease dermatitis herpetiformis is more prevalent in Caucasians, affecting them in any age, but more in their fourth decade of life or middle aged and is more frequent in men than women. The reason or how they get the disease could be caused by a wheat allergy, where wheat proteins can trigger their diseases symptoms. The Bullous pemphigoid disease mainly affects the elderly, but a small number of cases have been reported in children, the disease affects both men and women. People can get BP disease with trauma, burns, radiation, ultraviolet radiation (UV), and a variety of drugs. The epidermolysis bullosa acquisita disease has been reported to have a higher prevalence in Koreans and

can present a wide range of ages from childhood to middle age, however most cases start at 40 and 50 years of age, they can get this disease due to family genetics as it can be passed down. The vitiligo disease is distributed between men and women equally, the disease starts during the second decade of life, but one can get it through genetics or environmental factors. The psoriasis disease can affect anyone, men and women or adults and children, they can get it with stress, infections, medication, weather such as cold and dry weather, skin injuries and exposure to the sun causing sunburn. (Ávalos-Díaz).

There are many clinical signs that show the existence of autoimmune skin diseases, while others cannot be found on the surface of the skin but on the inside of the skin barrier. The disease pemphigus, in greek pemphix means blister, which is a group of blistering diseases with an organ-specific autoimmune disease that affects the skin and mucous membranes. The disease is characterized by the blisters, erosions and lesions that can be found when the disease is occurring. The disease bullous pemphigoid is a blistering autoimmune disease as well, with the blisters being large and tight causing erosions and scabs. The disease dermatitis herpetiformis can be shown by rashes and blisters and in some cases there are lesions which usually appear on the surface of the skin. The epidermolysis bullosa acquisita disease can develop autoantibodies that cause skin fragility, erosions, and blisters. Vitiligo is the destruction of melanocytes, causing depigmentation, but can be presented clinically as a primary disease or a component of multiple other autoimmune processes such as lupus, adult onset autoimmune diabetes, and Addison's disease. Psoriasis is clinically presented as the surface of the skin covered with whitish scales and lesions distributed mainly on the scalp, elbow, knees, and gluteal creases. The psoriasis scales and lesions have a pattern of improving during the summer and worsening during the winter. With all the clinical signs in mind not everyone who gets an autoimmune disease may show the same signs as another person with the same autoimmune disease. (Ávalos-Díaz).

Many diseases have similar symptoms, causes or even clinical signs and most of the autoimmune skin diseases have similar causes and clinical signs. There are many diseases that could be mistaken for autoimmune diseases. Psoriasis can be mistaken for many skin diseases including eczema, hives, acne, rosacea, keratosis pilaris, skin cancer and more. These diseases have similar appearances when the condition progresses. (Davis).

Diagnostic tests vary between different autoimmune skin diseases because each disease has its own symptoms that result in different testing to diagnose whether the disease is present or not. Pemphigus, the diagnosis of autoimmune blistering diseases is based on the evaluation of clinical outcomes, histopathology, and direct or indirect immunofluorescence. Bullous pemphigoid diagnosis is similar to

pemphigus's diagnosis where it is based on the clinical outcomes and immunofluorescence. Epidermolysis bullosa acquisita can be diagnosed by clinical outcomes, family history, and skin biopsy. Vitiligo, psoriasis and dermatitis herpetiformis can all be diagnosed by clinical evaluations and outcomes, as well as skin biopsies. (Ávalos-Díaz).

Treatments can vary between each autoimmune skin disease, but the results can also vary where one treatment works and the other doesn't depending on the level of the disease a person is on, but is focused not only on curing but maintaining the symptoms to become more curable. To treat pemphigus it involves the administration of corticosteroids and immunosuppressive agents and if the case of pemphigus is resistant then using IVIG and Rituximab is effective. Bullous pemphigoid is difficult to treat due to the progressive condition of the disease. Dermatitis herpetiformis treatment consists of gluten-free diets and dapsone. For epidermolysis bullosa acquisita the treatment depends on the clinical features to avoid causing skin trauma, so the treatment leans towards therapy. Vitiligo is treated using various therapeutic approaches including the administration of oral photosensitizing molecules such as psoralen. Psoriasis uses traditional treatments to maintain the effects of the disease. (Ávalos-Díaz).

Therapy is an additional form of treatment that can be as or more effective than the original medications or treatments to cure an autoimmune skin disease. For pemphigus additional therapy includes anti-idiotypic antibodies or apoptosis inhibitors that can be used to control blisters. Bullous pemphigoid disease therapy includes systemic steroids, dapsone, immunosuppressants. Dermatitis herpetiformis therapy includes a gluten-free diet combined with dapsone or steroids at low doses. Epidermolysis bullosa acquisita disease is cured using original therapy that consist of colchicine, dapsone, steroids, and immunosuppressive therapy. Vitiligo uses different therapeutic methods such as traditional therapy using corticosteroids, calcineurin inhibitors. Psoriasis is currently being treated using therapeutic approaches that include the use of chimeric or human anti-TNF monoclonal antibodies and recombinant anti-cytokine receptors. (Ávalos-Díaz).

Autoimmune skin diseases cannot be cured to the point where the patient does not have it, however treatments whether that's medication or therapeutic treatments can help manage clinical features such as blisters, lesions, erosions and so on. With most autoimmune skin diseases being a progressive condition it is not possible to clear without it returning again or worsening. Many people who have autoimmune skin diseases tend to live normal lives while maintaining their condition to avoid flaring of the symptoms.

There have been experiments done in the past year that are promising against autoimmune skin diseases. One of those is a treatment in the form of a drug called litifilimab, which is a long term treatment for autoimmune skin diseases, such as cutaneous lupus erythematosus (CLE). This disease affects the skin to the point where it severely impacts a patient's quality of life,, this drug cannot cure the disease, but it can lessen the progression of the condition. (“New Drug Shows Promise against Autoimmune Skin Disease - Penn Medicine”).

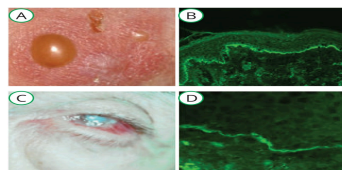
Conclusion

The autoimmune skin diseases that have been mentioned in this article each need different treatments and therapy to maintain the condition of the disease. These diseases consist of mostly blistering autoimmune skin diseases, so a patient should focus on maintaining the blisters to avoid flare ups that cause further problems. With the patient or their family I would discuss their condition, treatment, therapy options, and not to expect the disease to clear up, but instead to maintain the symptoms of the condition by staying consistent with treatments. Although these diseases are continuously being researched and experimented on there are many treatments and therapeutic methods that help a patient live a normal life.

Pictures (Ávalos-Díaz).



Pemphigus



Bullous Pemphigoid (bp) Disease



Vitiligo



Psoriasis

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