VITILIGO UPDATE

By Elaine Moore

Vitiligo is an autoimmune skin disorder, scientifically known as a leukoderma, that causes a loss of the skin pigment melanin. Vitiligo is characterized by blanched or white dots, macules or patches of skin. The degree of pigmentation loss (depigmentation) in vitiligo varies in different individuals and may be mild (slight hypopigmentation) or severe (complete depigmentation). Although the skin is most likely to be affected, the hair, retina of the eyes and the mucous membranes of the mouth, nose, genital and rectal tissue may also be affected. Like most autoimmune disorders, symptoms wax and wane and the severity of the condition varies, changing over time. The cause of depigmentation in melanoma is uncertain.

Older theories focused on the presence of melanocyte-destroying autoantibodies contributing to vitiligo. Newer studies support the involvement of a cell-mediated autoimmune response related to an increase in suppressor T cells (CD8 cells) and a decrease in helper T cells (CD4 cells) in association with the presence of a type-1 cytokine. It has also been proposed that increased release of norepinephrine melanocytotoxin by the autonomic nerve endings near melanocytes causes their destruction.

Who Develops Vitiligo?

Approximately 2 percent of the world’s population (40-50 million people) have vitiligo. Vitiligo is more likely to occur in people with another autoimmune condition, particularly autoimmune thyroid disease, adrenal insufficiency, alopecia areata, which causes baldness, and pernicious anemia. For instance, about 7 percent of people with Graves’ disease develop vitiligo compared to a 1 percent rate among the general population. Studies show that 95% of those affected by vitiligo develop symptoms before age 40, with most new patients between 10 and 30 years. Vitiligo occurs in all races, and it affects both sexes equally.

Symptoms

Early symptoms of vitiligo include white or blanched patches of skin usually affecting skin that’s exposed to the sun. The white areas may blend into the skin and not be distinct, and the affected area may itch. Occasionally, the affected areas may have hyperpigmented borders. Because vitiligo is often a progressive disorder, these patches tend to spread to other areas of the body. The rate at which the vitiligo spreads varies in different patients. Overall, most patients develop several areas of depigmentation on different areas of skin. This is known as a generalized vitiligo pattern. However, some patients develop a focal pattern of vitiligo, in which one or two areas of unpigmented skin persist for many years in the absence of other skin changes. Other patients may notice patches of vitiligo on only one side of the body, which is called a segmental pattern.
Segmental vitiligo usually occurs early in life and spreads rapidly. Non-segmental vitiligo has a slower course. Often, early patches of vitiligo persist for life although there is no spreading.

Some patients with vitiligo may also develop premature graying or whitening of hair, including scalp hair, eyelashes, eyebrows, axillary hair, pubic hair, and beard. Vitiligo often occurs around body orifices such as the lips, genitals, gingival, areolas, and nipples. Loss of pigment in the oral mucosa may also become prominent and tend to be more noticeable in people with darker skin. Some patients report an exacerbation or worsening of symptoms in times of both physical and emotional stress, including the stress of sunburn.

**Diagnosis**

Vitiligo is suspected in patients with characteristic skin changes who report recent rashes or skin trauma in the affected areas or premature hair graying and in patients with a history of autoimmune disease. Patients without a prior history of autoimmune disease are usually tested for blood cell counts, thyroid hormone levels, antinuclear antibodies, and adrenal function.

**Treatment**

Treatment for vitiligo focuses on restoring the function and appearance of the skin. Most medical treatments work slowly and are administered for 3-18 months before there is noticeable improvement. Corticosteroid treatments are usually the first-line treatment approach, followed by topical and/or oral psoralen photochemotherapy (Psoralen and ultraviolet A therapy, or PUVA used to increase skin pigmentation) and depigmentation treatments. Topical photochemotherapy may be sufficient for children or adults with small areas of depigmentation, but in adults a combination of oral and topical treatment is usually used.

Side effects of photochemotherapy include severe sunburn and blistering. Oral PUVA therapy is used for patients whose vitiligo affects more than 20 percent of their body. Oral therapy is not recommended for children younger than 10 years. Oral PUVA therapy may be used with natural sunlight although it is generally more effective when used in combination with exposure to artificial PUVA light.

Depigmentation therapy involves fading the skin to blend in with the depigmented areas. Depigmentation therapy is usually reserved for patients with more than 50 percent skin involvement. The drugs monobenzylether or hydroquinone are commonly used, applying the preparations to pigmented skin twice daily. Side effects include redness, swelling, itching and dryness. Treated areas also experience permanent sensitivity to sunlight.

Recent studies show that the anticonvulsant medication phenytoin (Dilantin), used as a high dose topical preparation in combination with topical corticosteroids, improves the
appearance of skin in patients with vitiligo. Phenytoin works by inhibiting the release of norepinephrine, and by stimulating collagen production and inhibiting its breakdown.

Patients who do not show a good response to medical therapies may be treated with skin grafts, micropigmentation, which is a form of tattooing, and autologous melanocyte transplants using cells removed from unaffected areas of the patient’s skin.

**Resources:**

NIAMS branch of the National Institutes of Health publication, online.