Morphea (Localized Scleroderma)

Morphea is an autoimmune disease that causes sclerosis, or scarlike, changes to the skin. Autoimmune diseases occur when the immune system, which normally protects us from bacteria, viruses, and fungi, mistakenly attacks a person's own body. Morphea lesions have 5 main presentations: (1) circumscribed (few circles on the trunk or limbs); (2) generalized (many circles on the trunk and limbs); (3) linear (lines of involvement on the limbs or head); (4) mixed (combination of circumscribed and linear or generalized and linear); and (5) pansclerotic (involvement of all of the skin). Morphea is usually limited to the skin, but it may extend deeper to involve muscle or bone. Morphea may also involve the inside of the mouth, the genitals, and the eyes. Morphea often first occurs in childhood or middle adulthood. Treatment decreases the risk of forming new lesions and the expansion of lesions. There is currently no cure for morphea.

Symptoms
Morphea is usually asymptomatic, with occasional itch and rarely pain. Morphea usually begins as a red or purple area of skin that then becomes thickened and white. The thick white areas usually thin out over time and turn brown. Once a lesion has formed, it is unlikely to completely go away. In children, linear morphea on the arms and legs can affect the underlying bone and interfere with bone growth. Linear morphea on the head (also called en coup de sabre) may extend inwards to the brain and cause seizures. If morphea crosses a joint, the thickening may limit joint movement. Morphea tends to have a waxing and waning course.

Diagnosis
Morphea is diagnosed based on findings of skin examination and skin biopsy. Patients with morphea do not have Raynaud phenomenon (fingers turning red, white, and blue with cold exposure), redness and swelling of both hands (red puffy hands), changes in the blood vessels along the nail edge (nailfold capillary changes), or tightening of the skin of the fingers (sclerodactyly). Patients who have these findings are likely to have systemic sclerosis, also called scleroderma. Systemic sclerosis and morphea are different diseases. Patients with morphea are not at increased risk of developing systemic sclerosis.

Treatment
Treatments help to control itch and decrease the growth and formation of new lesions. The treatment will depend on morphea type. Generally, creams and ointments are given to patients with circumscribed morphea or linear morphea that involves just the skin. Generalized morphea, or morphea that involves the muscle and bone, is usually treated with phototherapy (regular exposure to UV light in a special machine) or a combination of oral steroids and methotrexate (medications that suppress the immune system).