KERATOSIS FOLLICULARIS SPINULOSA DECALVANS

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Keratosis follicularis spinulosa decalvans (KFSD) is an uncommon inherited skin condition. Most cases are found in men. Females who have the disease tend to have milder symptoms. KFSD is characterized by dry, rough, thickened skin, and scarring hair loss occurs as a result. This condition begins in infancy, initially appearing on the face and neck, and then progresses to the torso, arms and legs. Scarring hair loss of the eyebrows and scalp become evident in childhood and progress until teenage years.

KFSD is a form of ichthyoses, which are a group of inherited disorders of the skin. The skin tends to become dry, rough and thick, giving a scaly appearance. The thickening of the skin damages the hair follicles causing scarring hair loss. Complications of this disease include vision problems, nail disorders, and cavities.

KFSD is inherited as an X-linked dominant trait. Chromosomes carry genetic information in the form of genes. Males have one X and one Y chromosome, and females have two X chromosomes. X-linked disorders are caused by an abnormal gene on the X chromosome. Due to its X-linked dominant mode of inheritance, the most severe manifestations are found in males.

KFSD is difficult to treat and there are no specific treatments. Patients may be prescribed retinoid creams that help smooth the skin, as well as emollients that help hydrate the skin. Occasionally steroid creams are used for symptomatic relief. However, treatment is generally considered unsatisfactory.