

Erythrokeratolysis hiemalis is an extremely rare form of ichthyosis that was first described in South Africa but has subsequently been identified in other countries. In such cases, a link to South Africa has been determined. The disorder is characterized by periodic attacks of red (erythematous) plaques that are distributed equally on both sides of the body. A layer of skin can be peeled from these plaques. Symptoms usually improve with age. The disorder tends to become worse with the cold weather. Erythrokeratolysis hiemalis is characterized by periodic attacks of red skin plaques. A thick layer of skin can be peeled off these plaques from the center outward. As new skin forms, the plaques are pushed outward. Symptoms may start during infancy or adolescence, and the disorder usually improves with age. Only the palms of the hands and the soles of the feet are involved in most cases. However, the disorder may spread to the skin of the back or elsewhere on the body surface. Appearance of new plaques may be precipitated by fever or surgery. Erythrokeratolysis hiemalis primarily affects descendants of farmers from the Oudtshoorn district in South Africa. This disorder starts during infancy or adolescence and usually improves after age 30. It is thought to affect males and females equally. One estimate is that 37 babies are born with this disorder annually. In most instances, the appearance of the skin determines the diagnosis, but a family history and physical examination are often required to rule out other possible causes of scaly, dry skin. Some physicians may examine skin tissue under a light microscope or even under an electron microscope.