Erythrokeratodermia with ataxia (EKDA) is a hereditary disorder of the skin and nervous system (neurocutaneous syndrome) characterized by groups of hard, red plaques that develop during infancy and childhood. When these skin lesions heal, the disorder seems to become dormant for several years, after which the neurological symptoms and signs emerge in the form of a typically awkward gait (ataxia) when the affected individual is around 40 years of age or older. Erythrokeratodermia with ataxia starts during early infancy. This disorder is characterized by groups of red, hardened scaly skin plaques (ichthyosis) that remain throughout childhood, but disappear during young adulthood. These plaques tend to develop most often on the skin of the extremities. They usually disappear during the summer. A progressive neurologic syndrome develops during adulthood, consisting of impaired muscle coordination (ataxia), imperfect articulation of speech (dysarthria), involuntary rhythmic oscillation of the eyes (nystagmus), and decreased tendon reflexes. Erythrokeratodermia with ataxia is an extremely rare disorder that is thought to affect males and females in equal numbers. The diagnosis of erythrokeratodermia with ataxia may be suspected soon after birth by the appearance of characteristic skin lesions. Neurological abnormalities associated with this disorder may not occur until adulthood.