Punctate Palmoplantar Keratoderma (PPK) often appears in adolescence with epidermal thickening of the palms and soles characterized as hyperkeratotic growths. PPK occurs in 1 in 100,000 people and is acquired or inherited in an autosomal dominant inheritance pattern. A mutation in the AAGAB gene leads to keratinocyte proliferation, resulting in thickened skin. PPK is divided into subtypes: diffuse, focal, and punctate. There are three forms, each of which varies slightly by clinical features: PPK type 1 (Buschke-Fischer-Brauer), PPK type 2 (Spiny Keratoderma), PPK type 3 (Acrokeratoelastoidosis). PPK type 1 is characterized as small, hyperkeratotic, yellow or brown papules on palms and soles of pressure-bearing sites. The number of papules can increase in number over time, if papules fall off it can result in small pits. PPK type 2 appears as small spines projecting from the palms and soles and PPK type 3 presents with tiny flat or umbilicated keratotic shiny papules along the margins of palms, soles, and digits. All three subtypes may be associated with underlying malignancies, including renal, lung, gastrointestinal and cutaneous carcinomas.

Some PPKs only affect the skin of the palms and soles while others have associated features including changes in hair, teeth, nails, hearing loss, and cardiomyopathy.

Positive family history is a strong clinical indicator for diagnosis; however, skin biopsy can help to confirm the diagnosis. Differential diagnosis includes porokeratosis palmaris et plantaris, warts, arsenical keratosis, calluses and corns, palmer pits, Cole disease, and Darier disease. It is important to obtain an extensive history to assess for inherited disease. This includes inquiring about age of onset, palmoplantar pain, blistering, sweating, infection, hearing loss, abnormal hair, nails, or teeth, mucosal abnormalities, and family history of cancer. Given the potentially complex systemic presentation, genetic counseling may be essential for the patient and their family.

Management of PPK includes educating patients on the benign nature of the condition that can persist throughout life, but also making them aware that further workup may be necessary to rule out underlying malignancy. Symptomatic treatment options are available to improve thickened skin, which includes moisturizing creams, keratolytic agents (salicylic acid, lactic acid, urea), topical retinoids, topical vitamin D ointment (calcipotriene), topical 5-fluorouracil cream, oral retinoids including acitretin. Surgical intervention with cryosurgery, mechanical debridement, and excision may also be helpful to remove painful horns. If it is determined that the disease is acquired in nature and not genetic, it is important to treat underlying disease and remove possible triggers.