Chronic mucocutaneous candidiasis (CMC) defines a heterogeneous group of orphan and inherited syndromes characterised by chronic and recurrent infections of the skin and mucosa with the yeast Candida. Increasing evidence suggests that this inefficient defence against Candida species is reflected by a DC/T cell defect which results in an impaired Th17 and Th1 immune response and, consecutively, a failed immune instruction of tissue cells. Little is known about the incidence and prognosis of CMC. Clinically, the main complications are debilitating hands (Candida granuloma) and oesophageal stricture with potential mal-digestion/-absorption. Furthermore, the chronic infections are likely a risk factor for the development of squamous cell carcinoma. Since resistance to anti-mycotic drugs evolves rapidly, efficient and flexible therapeutic management is essential for CMC patients.