Clinical Description There are many reviews of the clinical features of XP [1,4–6].These individuals have the severe neurological and developmental problems charac– teristic of Cockayne Syndrome together with the solar– induced pigmentation changes of XP.In the absence of rigorous protec– tion from the sun, areas of hyper– and hypo–pigmenta– tion will result, followed by accelerated photo–ageing, warty lesions, in–situ melanocyte and keratinocyte malig– nancy, and eventually multiple basal cell carcinomas and invasive squamous cell carcinomas and melanomas.Sunny climates, outdoor living, fair skin, smoking, poor availability of diagnostic facilities, delayed diagnosis and poor protection from sunlight will exacerbate the cuta– neous features, resulting in multiple pigmentation changes, multiple skin cancers and early death.The neurological abnormalities are the result of progressive neuronal degeneration resulting in sensorineural deafness, ataxia, areflexia, microcephaly and intellectual deficiency as well as impaired eyesight.Continued sunlight exposure may result in Page 2 of 6 severe keratitis, leading to corneal opacification and vas– cularization, and in neoplasms (epithelioma, squamous cell carcinoma, and melanoma) [5,7].Ocular abnormalities are almost as common as the cutaneous abnormalities, but they are strikingly limited to the anterior, UV–exposed structures of the eye (lids, cornea, and conjunctiva).In XP patients there is also a greatly increased fre– quency of cancer of the oral cavity, particularly squa– mous cell carcinoma of the tip of the tongue, a presumed sun–exposed area [5].Conver– sely, less sunny climates, indoor living, pigmented skin, early diagnosis and good solar protection, can result in relatively mild skin features.In these individuals, this sunburn reac– tion can happen in the first weeks of life and is often blamed on neglect or labelled wrongly as cellulitis or impetigo.Lentigines increase in number and darken and are difficult to distinguish clinically from the many, flat, pigmented seborrhoeic warts, which also proliferate and become warty.The early age of onset and the frequency of skin cancers in an otherwise normal individual should trigger further assessment for XP. In addition to the very large increase in skin cancer there is an approximately 50–fold increase in internal neoplasms, especially of the central nervous system.The clinical features are dependent on exposure to sunlight, the complementation group, the precise nature of the mutation as well as unknown factors.In these cases, the first manifestation, often by two years of age, is an unusually .increased number of lentigines (freckle–like pigmentation) in sun– exposed areas