013 Pigmentosa xeroderma group D: A clinical and genetic study of 19 Japanese cases
H Tanaka1, K Tera1, S Yamaguchi1, K Takahashi1 and S Moriwaki1 1 Department of Dermatology, Osaka Medical College, Takatsuki, Japan and 2 Department of Dermatology, University of the Ryukyus, Okinawa, Japan
Xeroderma pigmentosa (XP) is a rare autosomal recessive genetic disorder of DNA repair in which the ability to repair damage caused by ultraviolet light with an incidence of 1/22,000 in Japan. From the clinical symptoms, XP is classified into 3 types, such as XP cutaneous disease, XP neurologic disease, and XP/Cockayne syndrome (CS) complex. Genetically, XP is classified into seven genes, A (XPA), B (XPB), C (XPC), D (XPD), D2 (XPD), E (XPE), and F (XPF). Though XPA and XPC are the majority (55%) and XPV follows it (25%) in Japanese XP cases. Although the frequency of XPD is only 8% in Japan, there are the most diverse clinical symptoms. In this study, we classified and classified Western XP patients that reported to our department as an XP (7 cases), XP associated with internal malignancies (3 cases), the age at onset of skin cancer was 44.4 years for cutaneous disease, 12 years for neurological disease, and 2 years for XPCS complex. By the genetic analysis, we found that XP cases were associated with cutaneous disease, and NRRs were associated with severe XPCS complex, and R36W was associated with mild XPCS complex. In conclusion, we have conducted the clinical, epidemiological, and genetic studies on 19 Japanese XP cases. The type of XP cutaneous disease is dominant in Japanese XP cases. And clinical type (phenotype) and genotype association is implied in Japanese XP cases from the confirmed gene mutation.

015 The impact of pemphigus on health-related quality of life: First results with the EQ-5D questionnaire
B Tamasi1, V Brodsky2, K Hadju1,2, S Karpal1, I Galuta1, A Szegedi1,2, Z Beltagy1, G Kocsis1, A Kincses1,3,4, V Fencsik1 and F Renczi1 1 Department of Dermatology, Venerology and Allergology, Semmelweis University, Budapest, Hungary, 2 Department of Health Economics, Corvinus University of Budapest, Budapest, Hungary, 3 Department of Dermatology, University of Debrecen, Debrecen, Hungary, 4 Department of Dermatological Allergology, University of Debrecen, Debrecen, Hungary, 5 Department of Dermatology and Allergology, Albert Szent-Györgyi Medical Centre, University of Szeged, Szeged, Hungary, and 6 Department of Dermatology, Venerology and Oncoendematology, University of Pécs, Pécs, Hungary
The objective of our study was to evaluate health-related quality of life (HRQoL) in pemphigus patients using the EQ-5D and to analyse its convergent validity with other outcome measures. Between 2014 and 2017, a cross-sectional study was conducted at the Department of Dermatology, University of Pécs. The HRQoL was completed on the five-level version of EQ-5D (EQ-5D-5L) and Dermatology Life Quality Index (DLQI). Average pain at the time of the survey was evaluated by an 11-point visual analogue scale (VAS). Disease activity was measured by the Autimmune Bullous Skin Disorder Intensity Score (ABSIS). A total of 91 consecutive patients with pemphigus (pemphigus vulgaris: n=69, pemphigus foliaceus: n=29, other: n=2) participated in the study. Median age was 57 years (range 19-93), and 69% were women. Overall, 47%, 44%, 43, 40% and 17% of the patients reported problems regarding pain, mobility, anxiety/depression, usual activities and self-care, respectively. Median EQ-5D-5L index scores of patients with limited (n=47), moderate (n=22), severe (n=17) and extensive (n=4) pemphigus were as follows: 0.94, 0.86, 0.76 and 0.70 (p<0.005). The EQ-5D-5L showed a strong correlation with the average pain VAS (r=-0.60, p<0.001), and a moderate correlation with DLQI (r=-0.540, p=0.001) and ABSIS scores (r=-0.411, p=0.001). This is the first study in the literature to use the EQ-5D questionnaire in pemphigus. The EQ-5D is a valuable instrument for the assessment of HRQoL in pemphigus patients that can be also used to generate health utility values for economic evaluations of pemphigus treatments.

016 Psoriatic arthritis and hearing loss
Y Semenov1, E Hsiao2, A Huang2, X Hui1, SK Kwatra3, BA Cohen4 and MJ Anadkat5 1 Dermatology, Washington University School of Medicine in St. Louis, Saint Louis, MO, USA, 2 Department of Dermatology, Johns Hopkins University School of Medicine, Baltimore, MD, USA and 3 Department of Epidemiology, Johns Hopkins University Bloomberg School of Public Health, Baltimore, MD
Psoriasis is associated with other conditions such as musculoskeletal and cardiovascular diseases and these comorbidities are frequent and significantly associated with the burden of psoriatic arthritis. Psoriatic arthritis was present in 1.1% of the study population. Individuals with psoriatic arthritis were more likely to report having hearing difficulties (OR 1.74, p = 0.005), seeing a mental health provider (OR 1.94, p = 0.019), have an average of 2.37 more days of poor mental health over last month (p = 0.002), and were more likely to be depressed (OR 2.6, p = 0.001), than normal controls. SEM analysis revealed that hearing impairment mediated 7.0%, 8.6%, and 6.4% of the effect of psoriatic arthritis on days of poor mental health. Psoriatic arthritis was present in 1.1% of the study population. Individuals with psoriatic arthritis were more likely to report hearing difficulties (OR 1.74, p = 0.005), seeing a mental health provider (OR 1.94, p = 0.019), have an average of 2.37 more days of poor mental health over last month (p = 0.002), and were more likely to be depressed (OR 2.6, p = 0.001), than normal controls. SEM analysis revealed that hearing impairment mediated 7.0%, 8.6%, and 6.4% of the effect of psoriatic arthritis on days of poor mental health. Psoriatic arthritis was present in 1.1% of the study population. Individuals with psoriatic arthritis were more likely to report hearing difficulties (OR 1.74, p = 0.005), seeing a mental health provider (OR 1.94, p = 0.019), have an average of 2.37 more days of poor mental health over last month (p = 0.002), and were more likely to be depressed (OR 2.6, p = 0.001), than normal controls. SEM analysis revealed that hearing impairment mediated 7.0%, 8.6%, and 6.4% of the effect of psoriatic arthritis on days of poor mental health. Psoriatic arthritis was present in 1.1% of the study population. Individuals with psoriatic arthritis were more likely to report hearing difficulties (OR 1.74, p = 0.005), seeing a mental health provider (OR 1.94, p = 0.019), have an average of 2.37 more days of poor mental health over last month (p = 0.002), and were more likely to be depressed (OR 2.6, p = 0.001), than normal controls. SEM analysis revealed that hearing impairment mediated 7.0%, 8.6%, and 6.4% of the effect of psoriatic arthritis on days of poor mental health. Psoriatic arthritis was present in 1.1% of the study population. Individuals with psoriatic arthritis were more likely to report hearing difficulties (OR 1.74, p = 0.005), seeing a mental health provider (OR 1.94, p = 0.019), have an average of 2.37 more days of poor mental health over last month (p = 0.002), and were more likely to be depressed (OR 2.6, p = 0.001), than normal controls. SEM analysis revealed that hearing impairment mediated 7.0%, 8.6%, and 6.4% of the effect of psoriatic arthritis on days of poor mental health.

018 Kim-1 biomarker of kidney injury in patients with psoriasis
I Niculae1, CD Ere2, M Tampa3, M Cirstea4 and SR Georgescu5 1 Victor Babes Clinical Hospital of Tropical and Infectious Diseases, Bucharest, Romania, 2 Nephrology Hospital “Dr. Carol Davila”, Bucharest, Romania and 3 Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania
The mechanisms mediating renal insufficiency in patients with psoriasis are controversial. The authors aim to establish if kidney injury molecule-1 (Kim-1) might represent a specific serologic marker useful in identifying those patients with psoriasis with a higher risk to develop renal dysfunctions. We performed a prospective study which included 45 patients with untreated psoriasis vulgaris and 45 controls. All selected subjects were adults, normolipidemic, normoponderal, with a balanced diet. The serum level of Kim-1 (ELISA method) was analysed in correlation with the severity of the cutaneous disease and the classical markers of renal dysfunctions. The cutaneous disease was assessed using the PASI score. Kim-1 was defined as mild chronic plaque-type psoriasis (15 cases), PASI>7-12 as moderate chronic plaque-type psoriasis (15 cases) and PASI>12 as severe chronic plaque-type psoriasis (15 cases). The renal function was evaluated through serum evaluation of urea and creatinine and urinary assessment of estimated glomerular filtration rate (eGFR), the albumin/creatinine ratio and the uric acid/creatinine ratio. The serum levels of Kim-1 (mg/ml) were significantly increased in psoriasis (938±102) versus control (356±43, p=0.001). No association between Kim-1 and the severity of the cutaneous disorder was observed. A negative association was however obtained between the value of Kim-1 and eGFR (r=-0.540, p=0.013). These conclusions plead for the concept that psoriasis is a risk factor for developing renal dysfunctions.