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Original research

Identifying the psychosocial predictors of ultraviolet exposure to the face in patients with xeroderma pigmentosum: a study of the behavioural factors affecting clinical outcomes in this genetic disease

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ABSTRACT

Background For patients with xeroderma pigmentosum (XP), the main means of preventing skin and eye cancers is extreme protection against ultraviolet radiation (UVR), particularly for the face. We have recently developed a methodology for objectively measuring photoprotection behaviour ('UVR dose to facial skin') and have found that the degree of photoprotection varies greatly between patients with XP. We have previously identified factors affecting photoprotection behaviour in XP using a subjective measure of photoprotection. Here, we have used this objective methodology to identify the factors which determine photoprotection behaviour in XP.

Methods We studied 29 psychological, social, demographic and clinical variables in 36 patients with XP. We have previously objectively measured UVR protection (by measuring the dose of UVR reaching the skin of the face over a 3-week period) in these patients. Here, we use linear mixed-effects model analysis to identify the factors which lead to the differences in degree of photoprotection observed in these patients.

Results Psychosocial factors accounted for as much of the interindividual variation in photoprotection behaviour (29%) as demographic and clinical factors (24%).

Psychosocial factors significantly associated with worse UVR protection included: automaticity of the behaviours, and a group of beliefs and perceptions about XP and photoprotection known to associate with poor treatment adherence in other diseases.

Conclusions We have identified factors contributing to poor photoprotection in XP. Identifying these potentially reversible psychosocial features has enabled us to design an intervention to improve photoprotection in patients with XP, aiming to prevent skin and eye cancers in these patients.

INTRODUCTION

Xeroderma pigmentosum (XP) is an autosomal recessive inherited disorder in which patients develop multiple skin cancers from childhood, eye disease (corneal and conjunctival scarring and malignancy) and progressive neurological degeneration.¹ The incidence in Western Europe is low (2.3 per million live births).² XP is more common in Japan³ and

Key messages

What is already known on this topic

- ⇒ Ultraviolet radiation photoprotection is critical to prognosis in xeroderma pigmentosum (XP).
- ⇒ We have recently found that many patients with XP photoprotect poorly.

What this study adds

- ⇒ Out of 29 factors examined, 9 are strongly associated with poor photoprotection in patients with XP.
- ⇒ Seven of these nine factors are psychological or social.

How this study might affect research, practice and/or policy

- ⇒ Since these seven psychosocial factors are potentially reversible, we are designing a psychological intervention targeting these factors, in order to improve photoprotection in patients with XP.

North Africa.⁴ Mean life expectancy in the USA is 32 years.⁵ The prognosis in tropical countries is much worse: in South Africa 80% of patients develop multiple squamous cell carcinomas, frequently metastatic, by the age of 6.⁶ In 80% of patients, the disease is caused by defects in nucleotide excision repair (NER), required for repair of ultraviolet radiation (UVR)-induced mutagenic photo-products in nuclear DNA in cells.⁷ The other 20% of patients are 'XP variants', who have normal NER but defective translesion synthesis past UVR-induced DNA damage.⁷ The genetic defects can involve any of eight disease-causing genes. Each gene corresponds to one XP complementation group (XP-A to G, and XP-V). There is clinical heterogeneity between and within complementation groups.¹

In XP, disease phenotype correlates to some extent with the mutations,¹ but, because the molecular defect specifically impairs the cellular response to DNA damage caused by UVR, UVR exposure of the skin and eyes plays the critical role in determining clinical outcomes.

Since there is no therapy for the underlying molecular defect, the main means of preventing eye and skin cancers to minimise UVR exposure. This is achieved through absolute and lifelong photoprotection. Since 5% of daylight, even when cloudy, is UVR, patients require rigorous avoidance of daylight, protection of the face using visors made of UVR-protective transparent film and of the rest of the body with clothing, hats, gloves and high factor sunscreens.⁸ The advice for photoprotection is considerably more rigorous in XP than is recommended for other photosensitive conditions or for other groups at increased risk of skin cancer. Since 80% of skin cancers in XP are on the face, head and neck,⁹ and skin cancers on the face have the highest surgical morbidity, photoprotection of the face is particularly important in XP.

If there are any patients with XP who photoprotect poorly, it is likely that this will significantly worsen their prognosis. Over the past 20 years, it has become clear that patients' 'non-adherence' to medical treatments contributes to worse clinical outcomes in many diseases.¹⁰ It is estimated that 30%–50% of all prescribed treatments are not taken as directed¹¹ and that rates might be higher in dermatological conditions^{12–13} and for preventive health behaviours.¹⁴ There are many known determinants of non-adherence (capability ('is the patient able to adhere to the treatment?'), opportunity ('is the treatment available and affordable for the patient?') and motivational ('does the patient wish to adhere to the treatment?')).¹⁵ Out of all these factors, a small group of psychological and social factors, including perceived necessity and concerns about treatment, the extent to which adherence becomes habitual and patients' mood and level of confidence to enact the behaviour, are consistent predictors of non-adherence in a range of chronic diseases.^{16–18} Since these factors can be modified by behaviour change interventions,¹⁹ identifying factors associated with poor photoprotection has therapeutic implications in XP.

Prior to our current programme of research, there had been no studies of how well patients with XP photoprotect, or of the factors determining their photoprotection behaviour.

In our previous work, we have used a simple self-report questionnaire to subjectively assess UVR protection for the face in 156 patients with XP in Europe and the USA.²⁰ The results suggested that UVR protection for the face was suboptimal in one-third of these patients. That questionnaire study of XP²⁰ also identified several psychosocial factors which appeared to be associated with poor adherence to photoprotection. However, the significance of the results from that study is unclear because the measure of adherence to photoprotection was so subjective.

In the study presented in this paper, we identify the psychosocial factors associated with poor photoprotection, using the 'gold standard' of UVR protection in XP, an objective and quantitative measure of the dose of UVR to which the patient's face is exposed.

Since there has previously not been a way to objectively measure the dose of UVR reaching the face, we recently developed a method to do this.^{21–22} It measures the total dose of UVR reaching the skin of the face per day, taking the methods of face photoprotection used by the patient into account. For each 15 min period a patient spends outside (window glass efficiently protects against UVR when inside), we measure the environmental UVR (using a wrist-worn UVR dosimeter), and combine that with the proportion of environmental UVR which will penetrate the method of face photoprotection being used during that period (using the 'face protection factor' associated with the protection method recorded by the patient in an activity diary for that period).^{21–22} We have used this method to objectively

measure photoprotection behaviour in 36 patients with XP in the UK over a 21-day period.²² We identified a wide range in photoprotection behaviour: the patient with XP with the highest mean daily UVR exposure dose to the face had 120-fold higher exposure than the patient with the lowest. The worst protecting patients had UVR exposure similar to the mean in a group of healthy individuals.²²

In summary, our recent study has shown that poor adherence to photoprotection, assessed using this 'gold standard' objective measure, is a common problem in XP.²² In this current study, we have gone further in this same group of 36 patients, to examine their psychosocial, and other, characteristics in detail, in order to identify the factors which may explain why photoprotection is so unexpectedly poor in so many of them.

METHODS

In this prospective observational study, patients completed a structured questionnaire to measure psychological and social variables. Clinical and demographic variables were collected from patients' medical records. The objective measure of photoprotection was the mean daily dose of UVR to which the skin of the face was exposed over the 3-week period of the study.

Recruitment

Patients were recruited from the UK National XP Clinic. The inclusion criterion was a diagnosis of XP made by identifying reduced unscheduled DNA repair or typical XP-variant changes in cultured skin fibroblasts^{23–24} in a patient with typical clinical features,¹ confirmed by genetic testing (mutations analysed in genomic DNA by massively parallel Illumina sequencing, using a platform containing the coding exons and splice sites (–30/+20 bp) of the 8 XP genes¹).

Eligible patients were contacted by a research nurse. For patients under 16 years, and for adults lacking capacity to consent (due to XP-related cognitive impairment), their carer was contacted. Patients, or carers, were only recruited if they understood sufficient English to complete questionnaires and activity diaries.

Procedure

UVR measurement and activity diary

Each patient wore a UVR electronic dosimeter (SunSaver 3^{25–26}) on the wrist, and completed a simple one page per day UVR protection record in which the patient selected which method or methods of face photoprotection, from a list of 7 possibilities, they used for each 15 min period spent outdoors (or whether they had not protected at all).²² They did this throughout a 3-week period in 2016, during the months when environmental UVR levels are highest in the UK (6 May–6 August). All patients were provided with their usual SPF50 high UVA protection sunscreen to use for the duration of the study, and were trained to apply it by an XP clinic nurse. For each 15 min period, the dose of UVR to the face was calculated by multiplying the UVR exposure recorded by the wrist-worn dosimeter by the 'face protection factor' associated with the face photoprotection behaviour recorded in the diary for that interval.²² The UVR measurements from the dosimeter were expressed as 'standard erythemal doses' (SEDs), a measure chosen because it is weighted towards the wavelengths of UVR most damaging to DNA, that is, the SED is the clinically relevant measure of UVR exposure in XP.^{22–27–28} Details of the UVR face exposure measurement methodology are described elsewhere.^{21–22}

Demographic and clinical factors

Clinical and physical variables collected from patients' medical records from the XP clinic included: XP complementation group, genotype, cultured skin fibroblast unscheduled DNA repair activity, presence/absence of XP-related cognitive impairment, severity of eye and neurological disease, degree of photosensitivity ('XP Sunburn score'²⁹), number and type of skin and eye cancers, age at diagnosis, years since diagnosis, age when photoprotection started. Demographic data included age, gender, ethnicity and family history of XP.

Psychological and social factors: assessed from a self-report questionnaire

The self-report questionnaire assessed psychological and social factors which might be determinants of photoprotection (see online supplemental materials). The self-report questionnaire assessed psychological and social factors which might be determinants of photoprotection. Self-caring adults completed the questionnaire themselves. For non-self-caring adults, and for children (below 18 years), the questionnaire was completed by the main carer, since UVR exposure of non-self-caring adults and children is largely decided by the main carer. The questionnaire was the same in both cases except for use of personal pronouns, with the carer providing answers about the patient's XP and the carer's beliefs and perceptions.

Decisions about which psychosocial factors to include were based on the psychosocial factors already known to affect treatment adherence in other diseases,¹⁴ the factors already known to affect photoprotection behaviour in healthy individuals³⁰ and on psychological theories relevant to treatment adherence^{31 32}:

- ▶ Patients' perceptions of XP ('consequences', 'timeline', 'personal control of XP', 'photoprotection control of XP', 'treatment control', 'identity', 'negative emotional representation' and 'perceived understanding') were examined using single items from the Adapted Brief Illness Perception Questionnaire,³³ scored on an 11-point scale (0–10), higher scores representing a stronger and more negative perception.
- ▶ Beliefs relating to the patient's perception of the need for photoprotection were examined with an adapted version of the Beliefs about Medicines Questionnaire.³⁴ Participants indicated their strength of agreement with each statement on a 5-point scale from 1 ('strongly disagree') to 5 ('strongly agree'), and a mean score for the two subscales ('necessity' and 'concern') is calculated, higher scores indicating stronger beliefs.
- ▶ 'Intention', 'self-efficacy' and 'automaticity' for each photoprotection behaviour when outside (eg, wearing a face visor, using sunscreen, wearing a hat) were assessed by recording the strength of agreement with statements on a 7-point scale. The 10-item 'intention' and 'self-efficacy' scales were adapted from a manual based on the Theory of Planned Behaviour,³⁵ and the 10-item automaticity scale was adapted from the Self-Report Habit Index.³⁶ The mean score across items in each scale was calculated, higher scores indicating stronger agreement. A single item about the habit of avoiding going outside during the day was assessed with the same 7-point scale.
- ▶ The level of social support received (1 ('no support') to 5 ('comprehensive support')) and satisfaction with social support (1 ('very dissatisfied') to 5 ('very satisfied')) were adapted from the Social Support Questionnaire.³⁷

- ▶ Emotional well-being was measured using the short-form Warwick-Edinburgh Mental Well-Being Scale (SWEMWBS).³⁸ It consists of seven items measuring how often in the past 7 days participants experienced various positive aspects of mental well-being, each item scored on a 5-point scale (1 ('none of the time') to 5 ('all of the time')). Total scores range from 7 to 35, higher scores indicating greater mental well-being.

We have previously established the reliability of these psychological scales in patients with XP: Cronbach's α internal reliability values within a sample of 156 patients with XP were good: 0.73 for necessity, 0.80 for concern, 0.73 for intention, for 0.75 self-efficacy and 0.71 for automaticity.²⁰

Analysis

Descriptive information for continuous variables was reported as mean and SD, unless data were heavily skewed in which case the median and IQR was considered more appropriate. Categorical variables were reported as the count of non-missing observations and the percentage.

Factors associated with mean daily UVR exposure to the face were assessed using linear mixed-effects models. Daily total UVR exposure to the face was the outcome variable (ie, up to 21 repeated daily values per person). The model incorporated a random intercept to account for the repeated daily observations within individuals. A first-order autoregressive error structure was modelled to account for cross-day effects within individuals. Separate models were estimated for each predictor to assess the individual association of each predictor with UVR dose to the face. A multivariate model included all predictors identified in those models to estimate the overall variance in UVR dose to the face explained by clinical, demographic and psychosocial factors. Models controlled for weekend effects, whether the respondent rated the day as sunny, and the type of report (patient or carer). All analyses were conducted in Stata V.16 and the 5% level used to determine statistical significance.

Exploratory analysis of qualitative groupings

We also carried out an exploratory analysis of the relationship between the data from this study and qualitative findings from a previous study. Twenty of the 21 adults in this study had previously taken part in a qualitative study in which semi-structured interviews with patients were analysed using a framework approach.³⁹ That study identified three psychologically distinct groups within the XP population: a 'dominated' group, who were very worried about the risks of XP and described photoprotection dominating their lives to a distressing extent; a 'resistant' group, for whom photoprotection was not important, and whose priorities were to avoid stigma, be accepted and engage in normal social activities and an 'integrated' group, for whom photoprotection was a routine and accepted part of their life.

Here, we have analysed the relationship between the results from this study (mean daily UVR dose to the face, and psychosocial variables from the questionnaire) and the group ('dominated', 'resistant' or 'integrated') to which each patient was found to belong in the previous qualitative study. Patient numbers in the three subgroups were too low to enable rigorous statistical analysis.

RESULTS

Of the 93 patients with XP known to the XP clinic, 78 were eligible, 47 of whom consented to take part. Six withdrew before dosimeter fitting, one provided <14 days of data, in two

Table 1 Demographic and clinical characteristics of the patients

	Self-caring adult Sample (n=21)	Patients cared for by a caregiver (children; non-self-caring cognitively impaired adults) Sample (n=15)	Total (n=36)
Demographic variables			
Male, n (%)	14 (67%)	9 (60%)	23 (64%)
Female, n (%)	7 (33%)	6 (40%)	13 (36%)
Age, mean (SD)	40.0 (16.0)	14.1 (9.9)	29.2 (18.8)
Ethnicity			
White British	9 (43%)	8 (53%)	17 (47%)
Asian British	8 (38%)	7 (47%)	15 (42%)
'Other mixed': mixed Caribbean	2 (9%)	0 (0%)	2 (5%)
'Other': Turkish	1 (5%)	0 (0%)	1 (3%)
'Other': Arab	1 (5%)	0 (0%)	1 (3%)
Clinical variables			
Complementation group			
A	5 (24%)	3 (20%)	8 (22%)
C	6 (29%)	5 (33%)	11 (31%)
D	1 (5%)	4 (27%)	5 (14%)
E	2 (10%)	0 (0%)	2 (6%)
F	1 (5%)	0 (0%)	1 (3%)
G	0 (0%)	2 (13%)	2 (6%)
V	6 (28.6%)	0 (0%)	6 (17%)
Unknown	0 (0%)	1 (7%)	1 (3%)
Age at diagnosis (years), mean (SD)	20.4 (15.9)	4.2 (3.4)	13.7 (14.7)
Age at which started photoprotection (years), mean (SD)	18.9 (14.0)	3.7 (3.2)	12.6 (13.2)
Abnormal sunburn reaction (XP sunburn severity score ≥ 1), n (%) ^{30*}	6 (29%)	10 (67%)	16 (44%)
Previous skin, eye or oral malignancy, n (%)	12 (57%)	2 (13%)	14 (39%)
Cognitive impairment, n (%)	2 (10%)	5 (33%)	7 (19%)

*Patients with complementation groups A, B, D, F and G have abnormal, increased sunburn reactions. Groups C, E and V are associated with normal visible reactions to sun exposure. XP, xeroderma pigmentosum.

dosimeters malfunctioned and two did not provide a full analysable dataset. In total, 36 of the 47 patients providing consent provided sufficient data to be included in the analysis. This occurred despite the demands on patients of the study. Because of the rarity of the disease, the sample size was based on the maximum number of patients that could be recruited, rather than on a calculation of statistical power.

Baseline characteristics

Demographic and clinical characteristics (table 1): the age range was wide (mean age 29.2, SD=18.8). Most patients (21 of the 36) were self-caring adults, with a wide range of educational attainment. Of the 15 non-self-caring patients, 11 were children, and 4 were cognitively impaired adults. There were more male patients than female patients. Seventeen of the patients were white British and 19 were non-white British (15 British Asian (13 Pakistani, 1 Sri Lankan, 1 Indian), 2 mixed Caribbean ancestry, 1 Arab and 1 Turkish), with a similar ethnic breakdown in the self-caring adult and cared-for groups. All XP complementation groups were included apart from B, which is very rare. Cognitive impairment and eye problems due to XP were common. XP sunburn severity scores show that 44% of patients described abnormally severe sunburn reactions, as expected given the

known links between sunburn responses and complementation group (only groups C, E and V have normal sunburning responses³⁴). Around two-thirds of patients had started photoprotection by the age of 13 (mean 12.6, SD=13.2). Nearly 40% had already suffered a mucocutaneous malignancy, which had occurred at a wide range of ages, reflecting the clinical heterogeneity of this disease.¹

Psychological and social characteristics (table 2): overall, participants perceived XP to be a chronic condition that can be controlled by photoprotection and medical procedures. They believed photoprotection to be necessary, but had moderate concerns about protecting. Respondents reported moderate intention to photoprotect (mean score=4.7, SD=1), rated photoprotection as fairly automatic (ie, happened without thinking: mean score=4.4, SD=1.6) and were confident that they were able to carry out each photoprotection activity (mean score=6.4, SD=1.3). They were less confident that they could avoid going outdoors (mean score=4.2, SD=2). Overall, the sample had a similar level of emotional well-being (mean score=23.0, SD=5.5) to the general population in England (mean score=23.6, SD=3.9)⁴⁰ and felt that they were well supported (mean score=4.1, SD=1.1). In comparison to the self-caring adults, the cared-for sample (n=15) thought XP had more serious consequences (cared-for sample mean score=9.2, SD=1.4; self-caring adults' mean score=6.0, SD=3.3), felt that they had less control (cared-for sample mean score=4.1, SD=4.1; self-caring adults' mean score=6.1, SD=2.6) and thought that they had a poorer understanding of XP (cared-for sample mean score=6.3, SD=3.7; self-caring adults' mean score=8.5, SD=1.4).

Associations between demographic, clinical and psychosocial factors and mean daily UVR dose to the face

Out of the 29 demographic, clinical and psychosocial factors examined, 9 had statistically significant associations with photoprotection behaviour: 2 of them were clinical factors and 7 were psychosocial (figure 1, table 3).

The clinical factors associated with worse photoprotection (ie, higher face UVR exposure) were as follows:

- ▶ Being older when diagnosed.
- ▶ Experiencing a normal sunburn response (ie, an abnormal severe sunburn response was associated with better photoprotection).

The psychosocial factors associated with worse photoprotection were as follows:

- ▶ Holding a strong belief that photoprotection can be beneficial for health ('photoprotection control').
- ▶ Holding a strong belief that clinical treatment can control XP ('treatment control').
- ▶ Greater satisfaction with social support received for photoprotection,

The psychosocial factors found to be protective, that is, associated with a lower UVR dose to the face (figure 1 (the numbers represented in figure 1 are in table 3)) were:

- ▶ perceiving XP to be a life-long condition ('timeline');
- ▶ acceptance that photoprotection is necessary ('necessity');
- ▶ carrying out photoprotection automatically ('automaticity');
- ▶ avoiding going outdoors automatically ('automaticity').

A strong belief that XP has serious consequences ('consequences') was associated with better photoprotection but did not quite reach statistical significance.

Worse photoprotection was associated with better psychological well-being, although not reaching statistical significance.

Table 2 Psychological and social characteristics of the patients

Psychological and social variables, mean score (SD)	Self-caring adult Sample (n=21)	Patients cared for by a caregiver (children; non-self-caring cognitively impaired adults) Sample (n=15)	Total (n=36)
<i>Illness perceptions (0–10)</i>			
'Consequences' of XP on life (0 no effect; 10 severe effect)	6.0 (3.3)	9.2 (1.4)	7.3 (3.1)
Timeline or duration of XP (0 a very short time; 10 forever)	9.4 (2.2)	9.7 (1.3)	9.5 (1.9)
Personal control of XP (0 absolutely no control; 10 extreme amount of control)	6.1 (2.6)	4.1 (4.1)	5.3 (3.4)
Photoprotection control of XP (0 not at all helpful; 10 extremely helpful)	8.0 (2.5)	9.8 (0.6)	8.8 (2.1)
Treatment control of XP (0 not at all helpful; 10 extremely helpful)	8.8 (1.8)	9.2 (2.1)	8.9 (1.9)
'Identity', that is, symptom experience (0 no symptoms at all; 10 many severe symptoms)	5.7 (2.9)	7.0 (3.4)	6.2 (3.1)
Illness concern (0 not at all concerned; 10 extremely concerned)	6.6 (2.9)	7.5 (2.8)	7.0 (2.8)
Understanding of XP (0 do not understand at all; 10 understand very clearly)	8.5 (1.4)	6.3 (3.7)	7.6 (2.8)
Emotional representation/impact of XP (0 not at all affected emotionally; 10 extremely affected emotionally)	5.0 (3.5)	7.0 (3.5)	5.9 (3.6)
<i>Beliefs about photoprotection (1 strongly disagree; 5 strongly agree)</i>			
Necessity (importance of UVR protection to the person's health)	3.9 (0.8)	4.6 (0.8)	4.2 (0.8)
Concern (concerns about negative impacts of UVR protection to the person's health)	2.9 (1.1)	3.7 (0.6)	3.2 (1.0)
<i>Intention to photoprotect (1 strongly disagree; 7 strongly agree)</i>			
Intention to avoid going out in the next 7 days	4.4 (2.4)	4.5 (2.4)	4.5 (2.4)
Intention to photoprotect outdoors (a variety of methods of photoprotection) in the next 7 days	4.5 (0.9)	5.2 (0.9)	4.7 (1.0)
<i>Confidence about ability to photoprotect (1 strongly disagree; 7 strongly agree)</i>			
Self-efficacy to photoprotect (Over the next 7 days I am confident I could ... (a variety of methods of photoprotection))	6.2 (1.4)	6.6 (1.1)	6.4 (1.3)
Self-efficacy to avoid going outside	4.2 (2.0)	4.1 (2.5)	4.2 (2.2)
<i>Degree to which photoprotection is enacted automatically (1 strongly disagree; 7 strongly agree)</i>			
Automaticity of photoprotection (a variety of methods of photoprotection)	3.8 (1.5)	5.3 (1.2)	4.4 (1.6)
Automaticity to avoid going outside	3.7 (2.9)	4.9 (2.9)	4.2 (2.9)
<i>Social support (1 no support/very dissatisfied; 5 comprehensive support/very satisfied)</i>			
Level of social support	3.9 (0.9)	3.8 (1.3)	3.8 (1.0)
Satisfaction with support	4.1 (1.0)	4.0 (1.3)	4.1 (1.1)
Psychological well-being (SWEMWBS) (7–35)	22.9 (5.8)	23.0 (4.6)	23.0 (5.3)

SWEMWBS, short-form Warwick-Edinburgh Mental Well-Being Scale; UVR, ultraviolet radiation; XP, xeroderma pigmentosum.

None of the 20 other demographic, clinical or psychosocial factors analysed reached the 5% level of statistical significance for association with the UVR dose to the face. These included: sex, age, ethnicity, previous mucocutaneous malignancy and psychological measures of confidence and intention to photoprotect.

Exploratory analysis indicated that the protective effect of an abnormal sunburn reaction depended on the severity of the reaction, which differs greatly between complementation groups²⁹: the most severe photosensitivity (XP sunburn severity score of 3) was associated with mean UVR dose to the face of 0.07 SED/day, <50% of the mean dose (0.15 SED/day) in those with less severe or no photosensitivity (XP sunburn severity scores 0–2). Patients with the less severe sunburn responses (XP sunburn scores 1–2) had a similar mean UVR dose to the face as patients without abnormal sunburn responses (sunburn score 0): 0.15 vs 0.14 SED/day.

Multivariate analysis indicated that 53% of the variance in mean daily UVR dose to the face (SED) could be explained by

the demographic, clinical and psychosocial factors measured in this study. Psychosocial factors accounted for a greater share of this variance (29%) than demographic and clinical variables (24%).

Analysis of qualitative groups

In the 20 of the 21 adults in this study who had previously taken part in our qualitative interview study,³⁹ we carried out exploratory analysis of the relationship between their results from the qualitative study and the data from this study (table 4).

Analysis of UVR dose to the face in relation to these psychologically distinct subgroups³⁹ (n=20), indicated, as expected, that the 'dominated' group had the smallest mean UVR dose to the face (0.04 SED/day), the 'integrated' group had a threefold higher dose (0.12 SED/day) and the 'resistant' group had a fivefold higher dose (0.20 SED/day). The 'dominated' group scored highest on the two psychosocial factors identified by this study

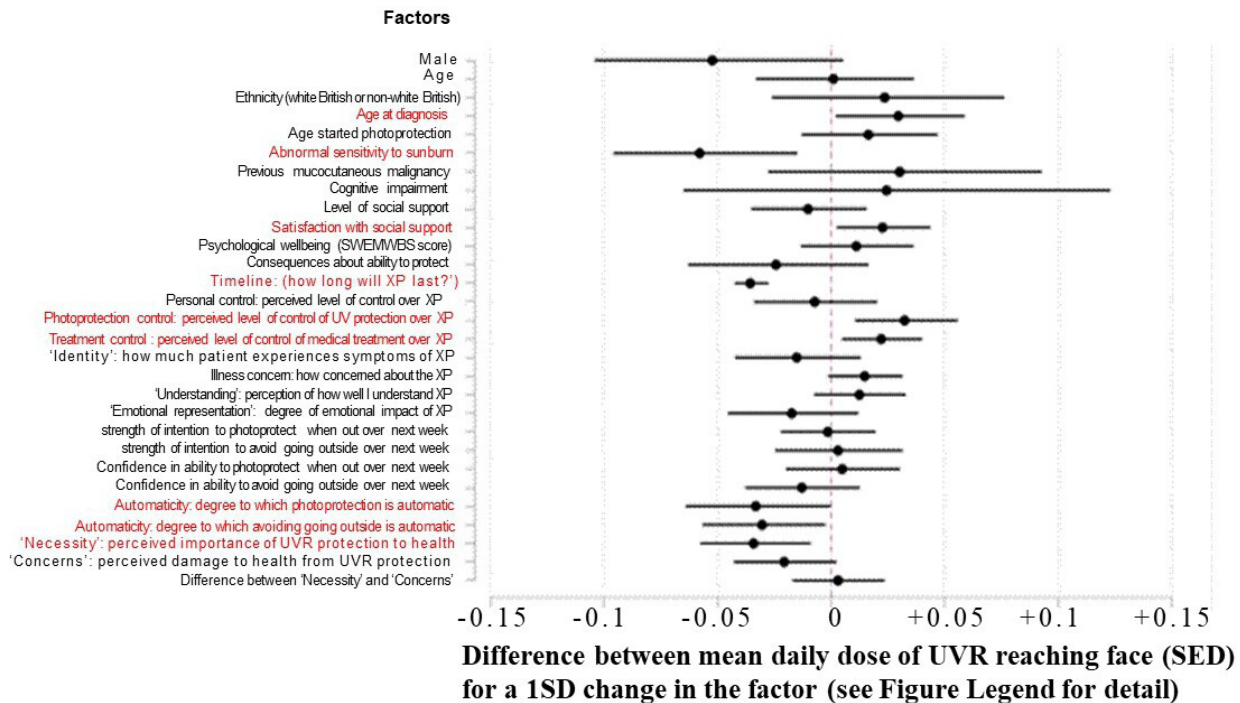


Figure 1 The associations between demographic, clinical and psychosocial factors and the calculated mean daily dose reaching the face (standard erythemal dose (SED)) for the patients with xeroderma pigmentosum (XP). The bars represent 95% CIs for the difference between the mean daily dose of ultraviolet radiation (UVR) reaching the face (SED) for a 1 SD change in the predictor variable for continuous variables, and change relative to reference group (eg, 'males' vs 'females') for categorical variables. The factors on the figure which can be seen to have a significant association with mean daily dose of UVR reaching the face are highlighted in red for clarity. SWEMWBS, short-form Warwick-Edinburgh Mental Well-Being Scale.

as most protective: acceptance of the necessity of photoprotection and automaticity of photoprotection, whereas the 'resistant' group scored lowest on these factors.

DISCUSSION

We recently used a quantitative and objective measure of photoprotection ('the UVR dose to the face') to show that adherence to photoprotection is unexpectedly poor in many patients with XP. In this study, we have identified the factors which determine how well patients with XP photoprotect. UVR protection, particularly of the face, is crucial to prognosis.

In this study, multivariate analysis showed that the psychosocial factors examined are at least as important as clinical and demographic factors in determining objectively measured face photoprotection behaviour. The key psychosocial factors we have identified here, including perception of disease chronicity, treatment necessity beliefs and automaticity, are similar to but not identical with those identified in our previous questionnaire study in 156 patients with XP across Europe and the USA, which had used a subjective and qualitative self-report measure of face photoprotection.²⁰ These are also some of the key determinants of non-adherence identified more broadly in other chronic diseases.^{16 41}

In this study, we found that a strong belief that XP can be controlled by photoprotection was associated with a higher dose of UVR to the face. We suspect that this apparently paradoxical finding may be because individuals holding this belief strongly may overestimate the effectiveness of photoprotection measures taken while outside and therefore go outside more. Sunscreen and most other methods of face photoprotection let through 20% or more of UVR, so any factor that increases confidence to spend time outside, even with good protection, will

increase overall UVR exposure of the face. Similarly, having a strong belief that clinical treatment is effective (with clinicians monitoring and removing skin cancers before they metastasise) was associated with an increase in UVR dose to the face. These unintended consequences of patients developing overconfidence in medical interventions has been shown to cause similar problems in cardiac patients: patients with the highest confidence in the effectiveness of medical intervention are less likely to adopt a healthier lifestyle after cardiac surgery.⁴² This highlights the tension between patients' and health professionals' responsibilities for health outcomes.

Exploratory analysis of the results from this study in the 20 adult patients who had also previously taken part in our qualitative interview study found that group differences in clusters of psychosocial factors (as demonstrated by the three patterns of adaptation to photoprotection identified in that qualitative study) were associated with differences in UVR dose-to-face larger than those ascribed to any individual psychosocial factors studied. This is consistent with health psychology theories,^{31 32} which suggest that psychosocial factors are not independent of each other, and tend to cluster together.

The complexity of the relationship between photoprotection in XP, well-being and social support is indicated by the finding that greater satisfaction with social support is associated with higher UVR exposure of the face. The qualitative interview findings in these patients³⁹ had suggested that greater satisfaction with social support might reflect a more active social life, and that this might increase the time spent outdoors, and that finding has been confirmed by our results.

The lack of an association between lower UVR exposure and improved psychological well-being is unexpected. In many chronic diseases, better adherence to treatment is associated

Table 3 The associations between demographic, clinical and psychosocial factors and the calculated mean daily dose reaching the face (SED) for the patients with XP

Variable	Difference in mean daily UV dose to face for 1 SD change in predictor variable (SED)	95% CI		P value
		Lower	Upper	
Timeline (how long will XP last?)	-0.036	-0.042	-0.028	0
Photoprotection control: perceived level of control of UV protection over XP	0.032	0.011	0.056	0.004
Abnormal sensitivity to sunburn	-0.058	-0.096	-0.015	0.008
'Necessity': perceived importance of UVR protection to health	-0.034	-0.058	-0.009	0.008
Treatment control: perceived level of control of medical treatment over XP	0.022	0.005	0.04	0.012
Satisfaction with social support	0.023	0.002	0.044	0.028
Automaticity: degree to which avoiding going outside is automatic	-0.03	-0.057	-0.002	0.033
Age at diagnosis	0.03	0.002	0.059	0.036
Automaticity: degree to which photoprotection is automatic	-0.033	-0.064	0	0.048
Illness concern: how concerned about the XP	0.015	-0.001	0.031	0.074
Male	-0.052	-0.104	0.005	0.075
'Concerns': perceived damage to health from UVR protection	-0.021	-0.043	0.002	0.079
Understanding: perception of how well I understand XP	0.012	-0.007	0.033	0.222
Consequences of XP on life	-0.024	-0.063	0.016	0.241
Emotional representation: degree of emotional impact of XP	-0.017	-0.046	0.012	0.246
Age started photoprotection	0.016	-0.013	0.047	0.275
'Identity': how much patient experiences symptoms of XP	-0.015	-0.042	0.013	0.289
Previous mucocutaneous malignancy	0.03	-0.028	0.093	0.311
Confidence in ability to avoid going outside over next week	-0.013	-0.038	0.013	0.32
Ethnicity (white British or non-white British)	0.024	-0.026	0.076	0.355
Psychological well-being (SWEMWBS score)	0.011	-0.013	0.036	0.376
Level of social support	-0.01	-0.035	0.016	0.438

Continued

Table 3 Continued

Variable	Difference in mean daily UV dose to face for 1 SD change in predictor variable (SED)	95% CI		P value
		Lower	Upper	
Cognitive impairment	0.024	-0.065	0.123	0.602
Personal control: perceived level of control over XP	-0.007	-0.034	0.02	0.605
Confidence in ability to photoprotect when out over next week	0.005	-0.02	0.03	0.704
Difference between 'necessity' and 'concerns'	0.003	-0.017	0.023	0.769
Strength of intention to avoid going outside over next week	0.003	-0.025	0.031	0.833
Strength of intention to photoprotect when out over next week	-0.002	-0.022	0.02	0.887
Age	0.001	-0.033	0.036	0.956

This is the numerical data represented in figure 1. For clarity, the factors are presented in order of increasing p value, and those which can be seen to have an association with mean daily dose of UVR reaching the face, significant at the 5% level, are highlighted in red.

*Difference between the mean daily dose of UVR reaching the face (SED) for a 1 SD change in the predictor variable for continuous variables and change relative to reference group (eg, 'males' vs 'females') for categorical variables.

SED, standard erythemal dose; SWEMWBS, short-form Warwick-Edinburgh Mental Well-Being Scale; UVR, ultraviolet radiation; XP, xeroderma pigmentosum.

with improved psychological well-being, including patients with malignant melanoma in whom reduced sunbathing is associated with improved well-being.⁴³ We suspect that this reflects the high demands of photoprotection in XP: the qualitative study found that excellent photoprotection was associated with considerable emotional distress among the 'dominated' group.³⁹ This implies that there is a trade-off between dermatological and psychological health. The high social and psychological costs of rigorous photoprotection point to the importance of finding ways to cope with issues of stigma and self-identity as part of behavioural interventions to increase adherence among patients with XP, since adherence to photoprotection marks patients with XP out as visibly different, often leading to adverse social and psychological impacts.⁴⁴

Studies of rare diseases are frequently limited by a sample size that restricts design and analysis, and present challenges for recruitment.⁴⁵ Despite recruiting nearly half the known cases of XP in the UK, caution is needed when interpreting the associations between factors and UVR exposure outcomes, as our sample was small for capturing statistical significances and limited the types of statistical tests that could be conducted. For qualitative subgroup analysis, the sample was too small for statistical testing. Caution is also needed when interpreting the average scores for the psychological and social factors as some variables were not normally distributed. In addition, while we have accounted for possible measurement confounders in the mixed-effect models, the CIs were large across factors. If tested in a larger sample, the strength and direction of the associations could potentially be influenced by the interaction between demographic and clinical factors and/or other confounders.

The group of patients in this study are representative of the overall XP population in the UK demographically and clinically,

Table 4 UV dose to face, and mean scores for some key psychosocial factors divided according to grouping from the qualitative interview study

	'Dominated' group	'Integrated' group	'Resistant' group
Number of patients	4	10	6
Mean daily UV dose to face (SED/day)	0.04	0.12	0.2
<i>Psychosocial factor</i>	<i>Mean score for psychosocial factor</i>	<i>Mean score for psychosocial factor</i>	<i>Mean score for psychosocial factor</i>
'Timeline or duration of XP' (0 a very short time; 10 forever)	9.5	10	8.8
'Necessity' of photoprotection (score 1–7)	4.5	4.1	3.4
'Automaticity' of photoprotection (score 1–7)	4.7	3.4	2.8
'Consequences' of XP on life (0–10)	8.3	5.5	5.9
Level of social support (1–5)	4.0	4.1	3.1

SED, standard erythemal dose; UV, ultraviolet; XP, xeroderma pigmentosum.

including the proportion of adults to children.¹ The previously published questionnaire study of adherence to photoprotection in XP was an international study of 156 patients with XP in the UK, France, the USA and Germany.²⁰ In that study, 10 factors were identified which significantly affected photoprotection behaviour. Four of those factors (age at diagnosis, 'necessity' and both types of automaticity) make up four of the nine factors identified in this UK-based study with its stronger methodology. This suggests that the results of this UK-based study may have application to groups of patients outside the UK.

Although disease phenotype in XP correlates to some extent with the nature of the mutations,¹ the eventual phenotype and clinical outcomes result from the interaction of the underlying genetic defect with acquired and environmental factors. Because the molecular defect in XP specifically impairs the cellular response to DNA damage caused by UVR, the environmental factor affecting disease expression is unusually well-defined. Exposure of the skin and eyes to environmental UVR, and protection from it, is entirely dependent on behaviour. An understanding of photoprotection behaviour and its underlying psychosocial causes provides an understanding of the acquired factor modulating phenotype in XP, to complement the understanding of the genetic factors. This study has defined the psychosocial basis of photoprotection behaviour in XP.

We have used the findings from this study to design a therapeutic behaviour change intervention ('XPAND') to target psychosocial determinants of UVR dose to the face in patients with XP.⁴⁶

The unexpectedly large variation in photoprotection behaviour between patients with XP raises the possibility that there may be a similarly dramatic range in behaviour affecting acquired disease-modulating factors between patients with other monogenic disorders, particularly in diseases where the avoidance of a disease-aggravating environmental factor or exposure to a protective environmental factor (such as a therapy) may be a potential source of psychological or social stress. If this is the case, identifying the psychological and social determinants of this variation in behaviour in order to design behaviour change interventions may be a useful approach to improving clinical outcomes in other monogenic disorders.

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Contributors RS played a joint lead role in conceptualisation, planning and funding acquisition, and led in the writing of the original draft and subsequent editing of the paper, and played an equal lead role in interpreting data; SN played a

lead role in data analysis and a supporting role in review of the paper; MC played a supporting role in data analysis and in review of the paper; MM led on qualitative aspects of this work and played a supporting role in review of the paper and a support role in interpretation of data; LF played a lead role in subject recruitment and an equal lead role in acquiring data from patients and played a supporting role in review of the paper; KS played a supporting role in project design, review of the paper and interpretation of data; VA-S played a supporting role in project design, review of the paper and interpretation of data; HCW played a supporting role in project design and data interpretation, particularly photobiological aspects, and in review of the paper; JWe played a joint lead role in conceptualisation, planning and funding acquisition, and a support role in writing of the original draft and review of the paper, and in interpretation of data; JWa played a joint lead role in conceptualisation and planning, a support role in subject recruitment, equal lead role in acquiring data, a support role in interpretation of data and a lead role in reviewing the paper. Guarantor: RS.

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Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

Understanding Ultraviolet Protection (UV) in Xeroderma Pigmentosum (XP) Questionnaire

Xeroderma Pigmentosum National Service

2nd Floor, South Wing

St Thomas' Hospital

Westminster Bridge Road

London SE1 7EH



We are trying to understand more about how having to protect against daylight affects the lives of people who have XP. From the answers we hope to be able to find ways to help patients and their families.

Please answer as many questions as you can. Where you are not sure of an answer, it is fine if you put your best guess.

Remember all the information you give will be treated with the strictest confidence and will be analysed anonymously. The people who look after you in the XP clinic will not see the answers you give.

If you are 12-15 years old and take responsibility for your own UV protection, please look through the questionnaire FIRST to see whether you feel able to complete it on your own. If you do not think you can, or if you are unsure, it is best for your parent or carer to complete one for you instead. They will complete a different version of questionnaire. Please contact our Research Nurse, Lesley Foster (Lesley.Foster@gstt.nhs.uk) if you do not have the version for parents or carers.

Thank you for taking the time to answer this questionnaire. Because XP is rare, every questionnaire that is completed and sent back to us makes a big difference for this research.

Filling out the questionnaire takes about 25 minutes. There are questions on both sides of each page.

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

The questionnaire is split into different topics:

- **XP and your thoughts about it**
- **What you do to protect yourself from Ultraviolet (UV) in daylight**

Some questions ask about what you usually do and others ask about the last few days or weeks. Some questions look the same as others but are actually different, so please can you try to answer as accurately as you can.

- **Your views about UV protection**
- **General things about you and your quality of life**

Before we ask about your XP, we'd like to find out a few details about you.

1. How old are you now? _____ Years

2. Are you: (please tick) Male or Female

3. I am completing the questionnaire because: (please tick one option)

I have been diagnosed with XP and I am 16 years or over	<input type="checkbox"/>
I have been diagnosed with XP and I am 12 - 15 years old and have chosen to complete the questionnaire myself	<input type="checkbox"/>

4. Do you have any of these qualifications? (Please tick all that apply).

If you have no formal qualifications please tick the box below

NVQ or equivalent	<input type="checkbox"/>
GCE / GCSE / O-level or equivalent	<input type="checkbox"/>
A-level or equivalent	<input type="checkbox"/>
Pre-degree foundation course/ diploma / HND or equivalent	<input type="checkbox"/>
Undergraduate degree	<input type="checkbox"/>

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

Masters	<input type="checkbox"/>
Doctorate/ PhD	<input type="checkbox"/>
Professional qualification	<input type="checkbox"/>
Other	<input type="checkbox"/>
Please write in _____	<input type="checkbox"/>
No Formal qualifications	<input type="checkbox"/>

ABOUT YOUR XP

Please complete or circle the answer

5. How old were you when you were diagnosed as having XP? Years
6. Have you ever had a skin cancer?	Yes / No
7. Have you had to see an Eye Specialist doctor because of any problems with the eyes?	Yes / No
8. Has the XP caused problems with your hearing, walking, or speaking?	Yes / No
9. Do you sunburn more easily than other people in your family who do not have XP?	Yes / No
10. Have you ever had sunburn so badly you needed to see a doctor about it?	Yes / No
11. Have you ever had sunburn in the winter in the country where you live?	Yes / No
12. Have you ever had sunburn which took more than 3 days to go away?	Yes / No
13. Do you take special measures to avoid some light bulbs?	Yes / No

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

14. How would you describe your skin?	
	Fair or light coloured
	Asian
	Light brown
	Afrocaribbean
	Dark brown
15. What colour are your eyes?	
	Blue
	Green
	Brown
	Other
16. Do you know what complementation group of XP you have?	Yes / No
17. If Yes please circle the group	
	A
	B
	C
	D
	E
	F
	G
V	

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

We are interested in your own personal views of how you see all aspects of your XP.

Please circle the number that best corresponds to your views:

18. How much does your XP affect your life?	0	1	2	3	4	5	6	7	8	9	10
<i>no affect at all</i>											<i>severely affects my life</i>
19. How long do you think your XP will continue?	0	1	2	3	4	5	6	7	8	9	10
<i>a very short time</i>											<i>forever</i>
20. How much control do you feel you have over the way XP affects your health?	0	1	2	3	4	5	6	7	8	9	10
<i>Absolutely no control</i>											<i>extreme amount of control</i>
21. How much do you think daylight/UV protection can help your skin or eye health?	0	1	2	3	4	5	6	7	8	9	10
<i>not at all</i>											<i>extremely helpful</i>
22. How much do you think XP treatment in the clinic (e.g. surgery, creams) can help your skin or eye health?	0	1	2	3	4	5	6	7	8	9	10
<i>not at all</i>											<i>extremely helpful</i>
23. How much do you experience symptoms related to your XP?	0	1	2	3	4	5	6	7	8	9	10
<i>no symptoms at all</i>											<i>many severe symptoms</i>
24. How concerned are you about your XP?	0	1	2	3	4	5	6	7	8	9	10
<i>not at all concerned</i>											<i>extremely concerned</i>

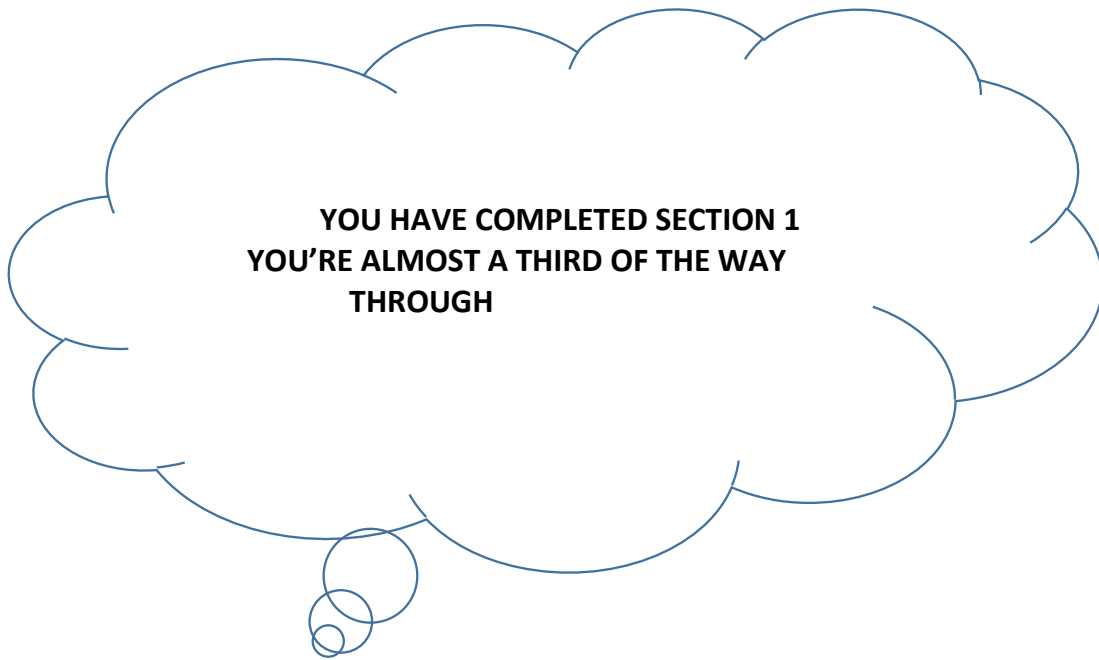
Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

Please circle the number that best corresponds to your views:

25. How well do you feel you understand your XP?											
0	1	2	3	4	5	6	7	8	9	10	
<i>don't understand at all</i>										<i>understand very clearly</i>	
26. How much does your XP affect you emotionally? (does it make you angry, scared, upset or depressed?)											
0	1	2	3	4	5	6	7	8	9	10	
<i>not at all affected emotionally</i>										<i>extremely affected emotionally</i>	



Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

ABOUT YOUR PROTECTION FROM ULTRAVIOLET (UV) IN DAYLIGHT

27. What is the date today?
(day/month/year)

...../...../.....

28. Which is your nearest town?

If you do not wish to give this information please move to the next question.

These questions focus on the LAST 7 DAYS

29. Over the last 7 days, what is your best guess for **how many days** it has been sunny for at least half the day?

.....Days (out of 7)

30. Thinking about a **typical day** in the last 7 days, what is your best guess on average for the **number of hours** you have spent outside in daylight (with or without UV protection)?

.....hours per day

31. We know it can be difficult to protect against UV all the time. Over the last 7 days when you went outside how often did you protect yourself against UV?
(Please circle the answer)

Never
rarely
sometimes
often
always

32. Do you have UV protective window film on home windows?
(Please circle the answer)

I have protective film on:
ALL windows
SOME windows
NO windows
I don't know

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

<p>33. Over the last 7 days, when at home to what extent have you stayed in rooms with UV protection (Please circle the answer)</p>	<p>Not at all rarely sometimes often always</p>
<p>34. Is there UV protective window film on windows at school or work? (Please circle the answer)</p>	<p>There is film on: ALL windows SOME windows NO windows I don't know I don't work or go to school outside the home</p>
<p>35. Over the last 7 days, when at work or school to what extent have you stayed in rooms with UV protection (Please circle the answer)</p>	<p>Not at all rarely sometimes often always</p>
<p>36. Do you have UV protective window film on your car's windows? (Please circle the answer)</p>	<p>There is film on: ALL windows SOME windows NO windows I don't know I don't have a car</p>

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

We know it can be difficult to protect against UV in daylight all the time.

These items ask how much you agree or disagree with the statements about the different things you could do to protect yourself from UV.

They are very similar statements about each way you could have protected yourself over the PAST 7 DAYS. Please read each question carefully and circle the number that best corresponds to your views. If you did not protect from UV in that way please circle the box "I did not..."

OVER THE LAST 7 DAYS.....

<p>37. <i>"Avoiding going outside during the day was something I did automatically without thinking"</i></p> <p style="text-align: center;">1 2 3 4 5 6 7</p> <p>Strongly Disagree Strongly Agree</p>	<p>I did not avoid going outside</p>
<p>38. <i>"Every time I got ready to go outside, wearing a face visor was something I did automatically without thinking"</i></p> <p style="text-align: center;">1 2 3 4 5 6 7</p> <p>Strongly Disagree Strongly Agree</p>	<p>I did not wear a face visor</p>
<p>39. <i>"Every time I got ready to go outside, wearing a hat was something I did automatically without thinking"</i></p> <p style="text-align: center;">1 2 3 4 5 6 7</p> <p>Strongly Disagree Strongly Agree</p>	<p>I did not wear a hat</p>
<p>40. <i>"Every time I got ready to go outside wearing glasses was something I did automatically without thinking"</i></p> <p style="text-align: center;">1 2 3 4 5 6 7</p> <p>Strongly Disagree Strongly Agree</p>	<p>I did not wear glasses</p>
<p>41. <i>"Every time I got ready to go outside, wearing sunscreen was something I did automatically without thinking"</i></p> <p style="text-align: center;">1 2 3 4 5 6 7</p> <p>Strongly Disagree Strongly Agree</p>	<p>I did not wear sunscreen</p>

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Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

42. "Every time I got ready to go outside, wearing lip sunblock was something I did automatically without thinking"	I did not wear lip sunblock
<p style="text-align: center;">1 2 3 4 5 6 7</p> <p>Strongly Disagree Strongly Agree</p>	
43. "Every time I got ready to go outside, wearing a scarf or "face buff" was something I did automatically without thinking"	I did not wear a scarf or "face buff"
<p style="text-align: center;">1 2 3 4 5 6 7</p> <p>Strongly Disagree Strongly Agree</p>	
44. "Every time I got ready to go outside, wearing a hoodie (worn up) was something I did automatically without thinking"	I did not wear a hoodie (worn-up)
<p style="text-align: center;">1 2 3 4 5 6 7</p> <p>Strongly Disagree Strongly Agree</p>	
45. "Every time I got ready to go outside, wearing long sleeves* was something I did automatically without thinking"	I did not wear long sleeves
<p style="text-align: center;">1 2 3 4 5 6 7</p> <p>Strongly Disagree Strongly Agree</p>	
46. "Every time I got ready to go outside, wearing gloves was something I did automatically without thinking"	I did not wear gloves
<p style="text-align: center;">1 2 3 4 5 6 7</p> <p>Strongly Disagree Strongly Agree</p>	
47. "Every time I got ready to go outside, wearing long trousers or thick tights was something I did automatically without thinking"	I did not wear long trousers or thick tights
<p style="text-align: center;">1 2 3 4 5 6 7</p> <p>Strongly Disagree Strongly Agree</p>	

*sleeves would include coats or jackets

OVER THE LAST 7 DAYS WHEN OUTSIDE.....

48. Thinking about all the things you did to protect yourself over the past 7 days (e.g., wearing sunscreen, wearing a hat), how well do you think they protected you from UV? (Please circle).	I did not do anything to protect myself from UV in the last 7 days
<p>Completely very well a fair amount a little Not at all</p>	

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Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

We know that people adapt their UV protection to fit in with everyday life.

These questions are about what you USUALLY do throughout the year to protect yourself from UV in daylight when it is CLOUDY AND then what you do on days when it is SUNNY.

Please circle how often you do the different things on the days when it is CLOUDY and then on the days when it is SUNNY, throughout the year.

49. To what extent do you try to avoid going outside during the day?	Cloudy days	Sunny days
	Never	Never
	rarely	rarely
	sometimes	sometimes
	often	often
	always	always

WHEN YOU GO OUTSIDE... (Please circle the answer)

	Cloudy days	Sunny days
Thinking about your FACE....		
50. How often do you wear a face visor?	Never rarely sometimes often always	Never rarely sometimes often always
51. How often do you wear a hat?	Never rarely sometimes often always	Never rarely sometimes often always
52. How often do you wear glasses?	Never rarely sometimes often always	Never rarely sometimes often always

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

When you go outside (Please circle the answer)	Cloudy days	Sunny days
53. How often do you use sunscreen on your face?	Never rarely sometimes often always	Never rarely sometimes often always
54. If you use sunscreen on your face, what sun protection factor does it have? (or circle I do not....)	SPF number	SPF number
	I do not wear sunscreen on my face	
55. How often do you reapply the sunscreen on your face during the day? (or circle I do not....)x/dayx/day
	I do not wear sunscreen on my face	
56. How often do you put on lip sunblock?	Never rarely sometimes often always	Never rarely sometimes often always
57. How often do you wear a scarf or "face buff"?	Never rarely sometimes often always	Never rarely sometimes often always

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

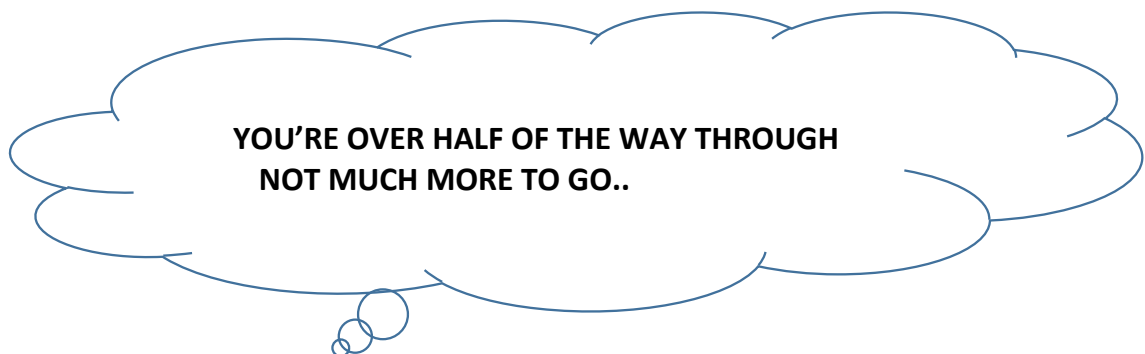
When you go outside (Please circle the answer)	Cloudy days	Sunny days
58. How often do you wear a hoodie (worn -up)?	Never rarely sometimes often always	Never rarely sometimes often always
Thinking about your ARMS OR HANDS...		
59. How often do you wear long sleeves?	Never rarely sometimes often always	Never rarely sometimes often always
60. How often do you use sunscreen on your arms or hands?	Never rarely sometimes often always	Never rarely sometimes often always
61. How often do you wear gloves?	Never rarely sometimes often always	Never rarely sometimes often always
Thinking about your LEGS...		
62. How often do you wear long trousers or thick tights?	Never rarely sometimes often always	Never rarely sometimes often always

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

When you go outside (Please circle the answer)	Cloudy days	Sunny days
63. How often do you use sunscreen on your legs?	Never rarely sometimes often always	Never Rarely sometimes often always
Thinking about using sunscreen on your BODY (NOT FACE), ARMS, HANDS OR LEGS....		
64. If you use sunscreen on any of these areas (body, arms, hands or legs) what sun protection factor does it have? (or circle I do not....)	SPF number	SPF number
	I do not wear sunscreen on my body, arms, hands or legs	
65. How often do you reapply the sunscreen to any of these areas (body, arms, hands or legs) during the day? (or circle I do not....)x/dayx/day
	I do not wear sunscreen on my body, arms, hands or legs	



Participant study code _____ Country of residence: UK
 Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

**We would like to ask your personal views about protecting against UV in daylight.
 This means whatever you do to protect yourself, including:**

- avoiding going outside during the day
- putting on sunscreen and lip sunblock
- wearing glasses
- wearing protective clothes (e.g., hat, scarf or face-buff, gloves, long sleeves, long trousers/thick tights)
- or wearing a face visor

These are statements other people have made about their UV protection.

Please indicate the extent to which you agree or disagree with what they have said by circling the answer that best corresponds to your views.

There are no right or wrong answers. We are interested in YOUR PERSONAL VIEWS.

66. <i>"My health depends on protecting against UV"</i>	Strongly Agree	Agree	Uncertain	Disagree	Strongly Disagree
67. <i>"Having to protect against UV worries me"</i>	Strongly Agree	Agree	Uncertain	Disagree	Strongly Disagree
68. <i>"My life would be impossible without protecting against UV"</i>	Strongly Agree	Agree	Uncertain	Disagree	Strongly Disagree
69. <i>"If I did not protect against UV I would be very ill"</i>	Strongly Agree	Agree	Uncertain	Disagree	Strongly Disagree
70. <i>"I sometimes worry about other people's reactions to the things I have to do to protect against UV (e.g., wearing a visor, using sunscreen, wearing a hat)"</i>	Strongly Agree	Agree	Uncertain	Disagree	Strongly Disagree
71. <i>"I sometimes worry about long-term effects of having to protect against UV"</i>	Strongly Agree	Agree	Uncertain	Disagree	Strongly Disagree

Participant study code _____ Country of residence: UK
 Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

72. <i>"I don't really understand why I need to protect against UV"</i>	Strongly Agree	Agree	Uncertain	Disagree	Strongly Disagree
73. <i>"My health in the future will depend on protecting against UV"</i>	Strongly Agree	Agree	Uncertain	Disagree	Strongly Disagree
74. <i>"Having to protect against UV disrupts my life"</i>	Strongly Agree	Agree	Uncertain	Disagree	Strongly Disagree
75. <i>"Protecting against UV stops my health getting worse"</i>	Strongly Agree	Agree	Uncertain	Disagree	Strongly Disagree
76. <i>"I sometimes worry about the impact on family and friends of having to protect against UV"</i>	Strongly Agree	Agree	Uncertain	Disagree	Strongly Disagree

We are now asking about the NEXT 7 DAYS.

These questions are about the different things you might do to protect yourself from UV. We know that people want to do some things more than others, and they find some things easier to do than others. We want to find out what YOU THINK.

These statements are very similar to each other about the ways you might protect yourself over the next 7 days. Please read each question carefully and think about each way in turn.

77. Over the next 7 days I <u>intend</u> to protect myself by avoiding going outside during the daytime	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree

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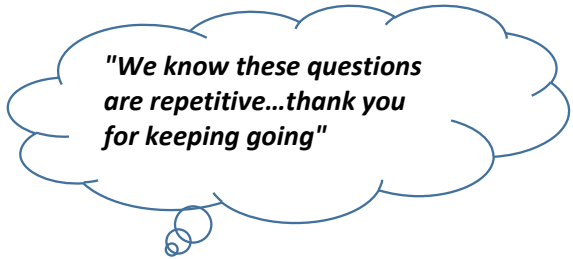
Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

When I am outside in the NEXT 7 DAYS....**(If you are not intending to go outside in the next 7 days please skip to question 88.)**

78. I <u>intend</u> to protect myself by wearing a face visor	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree
79. I <u>intend</u> to protect myself by wearing a hat	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree
80. I <u>intend</u> to protect myself by wearing glasses	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree
81. I <u>intend</u> to protect myself by putting on sunscreen	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree

When I am outside in the NEXT 7 DAYS....

82. I <u>intend</u> to protect myself by putting on lip sunblock	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree
83. I <u>intend</u> to protect myself by wearing a scarf or "face buff"	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree
84. I <u>intend</u> to protect myself by wearing a hoodie (worn-up)	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree
85. I <u>intend</u> to protect myself by wearing long sleeves	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree
86. I <u>intend</u> to protect myself by wearing gloves	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree
87. I <u>intend</u> to protect myself by wearing long trousers or thick tights	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree



"We know these questions are repetitive...thank you for keeping going"

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

Over the NEXT 7 DAYS.....88. I am confident I could **avoid going outside** during the daytime

	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree

When I am outside in the NEXT 7 DAYS....**(If you are not intending to go outside in the next 7 days please skip to question 99.)**89. I am confident I could **wear a face visor**

	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree

90. I am confident I could **wear a hat**

	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree

91. I am confident I could **wear glasses**

	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree

When I am outside in the NEXT 7 DAYS....92. I am confident I could put on **sunscreen**

	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree

93. I am confident I could put on **lip sunblock**

	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree

94. I am confident I could **wear a scarf or "face buff"**

	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree

95. I am confident I could **wear a hoodie (worn-up)**

	1	2	3	4	5	6	7
Strongly Disagree							Strongly Agree

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

96. I am <u>confident</u> I could wear long sleeves							
	1	2	3	4	5	6	7
	Strongly Disagree					Strongly Agree	
97. I am <u>confident</u> I could wear gloves							
	1	2	3	4	5	6	7
	Strongly Disagree					Strongly Agree	
98. I am <u>confident</u> I could easily wear long trousers or thick tights							
	1	2	3	4	5	6	7
	Strongly Disagree					Strongly Agree	

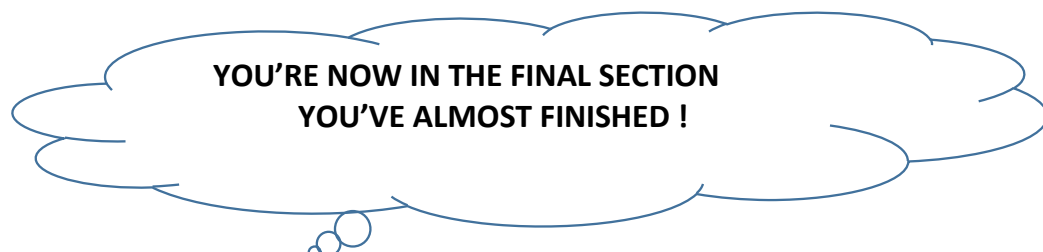
Overall in the NEXT 7 DAYS.....

99. I am confident I can do the things I want to do to protect myself from UV							
	1	2	3	4	5	6	7
	Strongly Disagree					Strongly Agree	

About your XP and the people around you.....

100. How much support or help do you have from the people around you with your UV protection?					
<i>No support</i>	<i>hardly any support</i>	<i>Some support</i>	<i>lots of support</i>	<i>comprehensive support</i>	

101. How satisfied are you with the support or help that you have to help you with your UV protection?					
<i>Very dissatisfied</i>	<i>fairly dissatisfied</i>	<i>uncertain</i>	<i>fairly satisfied</i>	<i>very satisfied</i>	



Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

ABOUT YOU AND YOUR QUALITY OF LIFE

These questions are about you, your thoughts about the present and future, your emotions and your quality of life.

Please circle the number that best corresponds to your views:

102. <i>"Thinking about the future is pleasant to me"</i>	1 2 3 4 5	Very true of me Very untrue of me
103. <i>"When I want to achieve something, I set goals and consider specific means of reaching those goals"</i>	1 2 3 4 5	Very true of me Very untrue of me
104. <i>"Meeting tomorrow's deadlines and doing other necessary work comes before tonight's play"</i>	1 2 3 4 5	Very true of me Very untrue of me
105. <i>"It seems to me that my future plans are pretty well laid out"</i>	1 2 3 4 5	Very true of me Very untrue of me
106. <i>"I think that it seems useless to plan too far ahead because things hardly ever come out the way you planned anyway"</i>	1 2 3 4 5	Very true of me Very untrue of me
107. <i>"If I do not get done on time, I do not worry about it"</i>	1 2 3 4 5	Very true of me Very untrue of me
108. <i>"I try to live one day at a time"</i>	1 2 3 4 5	Very true of me Very untrue of me
109. <i>"I feel that it is more important to enjoy what you are doing than to get the work done on time"</i>	1 2 3 4 5	Very true of me Very untrue of me
110. <i>"I do not do things that are good for me if they do not feel good now"</i>	1 2 3 4 5	Very true of me Very untrue of me

Participant study code _____ Country of residence: UK
 Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

Below are some statements about feelings and thoughts.

Please circle the answer that best describes your experience of each over the LAST 2 WEEKS

111. <i>"I've been feeling optimistic about the future"</i>	None of the time	Rarely	Some of the time	Often	All of the time
112. <i>"I've been feeling useful"</i>	None of the time	Rarely	Some of the time	Often	All of the time
113. <i>"I've been feeling relaxed"</i>	None of the time	Rarely	Some of the time	Often	All of the time
114. <i>"I've been dealing with problems well"</i>	None of the time	Rarely	Some of the time	Often	All of the time
115. <i>"I've been thinking clearly"</i>	None of the time	Rarely	Some of the time	Often	All of the time
116. <i>"I've been feeling close to other people"</i>	None of the time	Rarely	Some of the time	Often	All of the time
117. <i>"I've been able to make up my own mind about things"</i>	None of the time	Rarely	Some of the time	Often	All of the time

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

These questions are about how your quality of life is affected by the XP.**Under each heading, please tick the ONE box that best describes your health****TODAY.****118. MOBILITY**I have no problems in walking about I have slight problems in walking about I have moderate problems in walking about I have severe problems in walking about I am unable to walk about **119. SELF-CARE**I have no problems washing or dressing myself I have slight problems washing or dressing myself I have moderate problems washing or dressing myself I have severe problems washing or dressing myself I am unable to wash or dress myself **120. USUAL ACTIVITIES (e.g. work, study, housework, family or leisure activities)**I have no problems doing my usual activities I have slight problems doing my usual activities I have moderate problems doing my usual activities I have severe problems doing my usual activities I am unable to do my usual activities **121. PAIN / DISCOMFORT**I have no pain or discomfort I have slight pain or discomfort I have moderate pain or discomfort I have severe pain or discomfort I have extreme pain or discomfort

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Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

122. ANXIETY / DEPRESSION

I am not anxious or depressed

I am slightly anxious or depressed

I am moderately anxious or depressed


I am severely anxious or depressed

I am extremely anxious or depressed

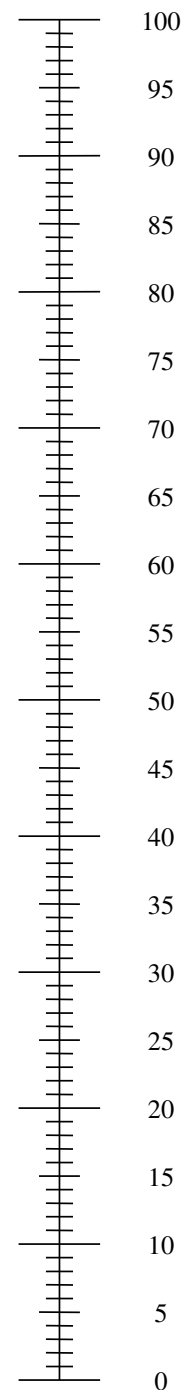
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Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

- We would like to know how good or bad your health is TODAY.
- This scale is numbered from 0 to 100.
- 100 means the best health you can imagine.
0 means the worst health you can imagine.
- Mark an X on the scale to indicate how your health is TODAY. 
- Now, please write the number you marked on the scale in the box below.

123. YOUR HEALTH TODAY =

The best health
you can imagineThe worst health
you can imagine

Participant study code _____

Country of residence: UK

Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

ONE MORE QUESTION ABOUT YOU...

This is an extra question, some people may not want to fill it out. If you don't want to do this question, just leave it blank.

124. Which of the following **best** describes your **ethnicity**? Please tick **one** of the following options.

White British	<input type="checkbox"/>	Black or Black British Caribbean	<input type="checkbox"/>
Any other White background please write in _____	<input type="checkbox"/>	African	<input type="checkbox"/>
Mixed		Any other Black background, please write in _____	<input type="checkbox"/>
White and Black Caribbean	<input type="checkbox"/>	Chinese or other ethnic group	
White and Black African	<input type="checkbox"/>	Chinese	<input type="checkbox"/>
White and Asian	<input type="checkbox"/>	Any other	<input type="checkbox"/>
Any other Mixed background please write in _____	<input type="checkbox"/>	Please write in _____	
Asian or Asian British			
Indian	<input type="checkbox"/>		
Pakistani	<input type="checkbox"/>		
Bangladeshi	<input type="checkbox"/>		
Any other Asian background please write in _____	<input type="checkbox"/>		

If you have any additional thoughts or comments please can you write them here:

Participant study code _____ Country of residence: UK
Questionnaire version 11 28/01/2016 Adult 16 yrs (12-15yrs opt)

**THANK YOU VERY MUCH FOR COMPLETING OUR
QUESTIONNAIRE.**

**If you have any questions about the questionnaire please
contact our Research Nurse Lesley Foster.**

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