

Supplementary Document 5: Cross-sectional survey

Participant study code _____

Country of residence: UK

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

Xeroderma Pigmentosum National Service

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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, (*****) 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.

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"/>
Masters	<input type="checkbox"/>

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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

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

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

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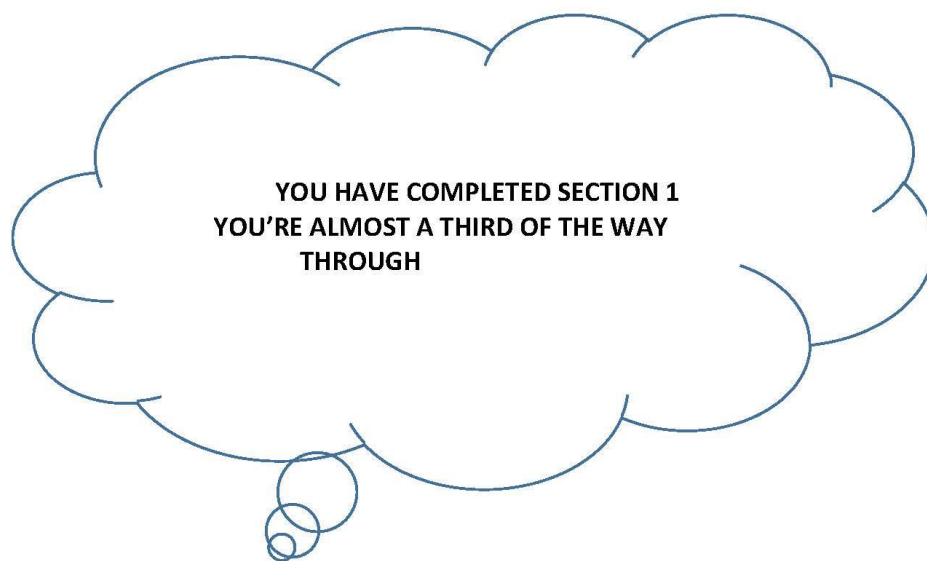
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Please circle the number that best corresponds to your views:

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>

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>



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ABOUT YOUR PROTECTION FROM ULTRAVIOLET (UV) IN DAYLIGHT

27. What is the date today? (day/month/year)/...../.....
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28. Which is your nearest town? <u>If you do not wish to give this information please move to the next question.</u>	
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These questions focus on the LAST 7 DAYS

29. Over the last 7 days, what is your best guess for <u>how many days</u> it has been sunny for at least half the day?Days (out of 7)
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30. Thinking about a <u>typical day</u> in the last 7 days, what is your best guess on average for the <u>number of hours</u> you have spent outside in daylight (with or without UV protection)?hours per day
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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
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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
--	---

33. Over the last 7 days, when at home to what extent have you	Not at all
--	------------

<p>stayed in rooms with UV protection (Please circle the answer)</p>	<p>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>
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<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>
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<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>
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<p>42. "Every time I got ready to go outside, wearing lip sunblock was something I did automatically without thinking"</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 lip sunblock</p>
<p>43. "Every time I got ready to go outside, wearing a scarf or "face buff" was something I did automatically without thinking"</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 scarf or "face buff"</p>
<p>44. "Every time I got ready to go outside, wearing a hoodie (worn up) was something I did automatically without thinking"</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 hoodie (worn-up)</p>
<p>45. "Every time I got ready to go outside, wearing long sleeves* was something I did automatically without thinking"</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 long sleeves</p>
<p>46. "Every time I got ready to go outside, wearing gloves was something I did automatically without thinking"</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 gloves</p>
<p>47. "Every time I got ready to go outside, wearing long trousers or thick tights was something I did automatically without thinking"</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 long trousers or thick tights</p>

**sleeves would include coats or jackets*

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

<p>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).</p> <p><i>Completely very well a fair amount a little Not at all</i></p>	<p>I did not do anything to protect myself from UV in the last 7 days</p>
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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

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

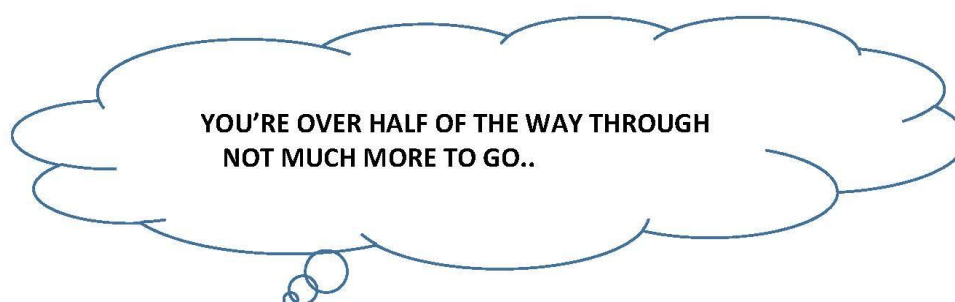
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When you go outside (Please circle the answer)	Cloudy days	Sunny days
58. How often do you wear a hoodie (worn -up)?	Never rarely sometimes often always	Never rarely sometimes often always
Thinking about your ARMS OR HANDS...		
59. How often do you 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

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	



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

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

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



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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

**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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122. ANXIETY / DEPRESSION


I am not anxious or depressed

I am slightly anxious or depressed

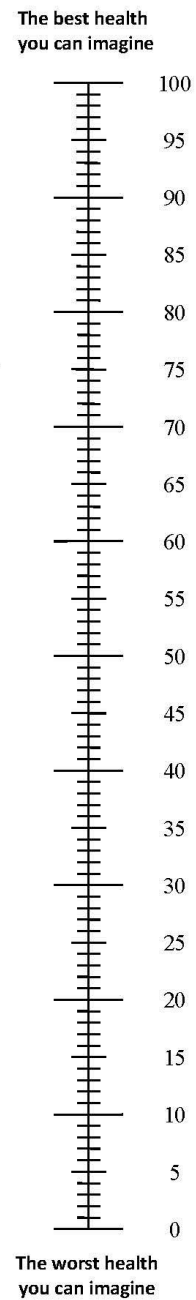
I am moderately anxious or depressed

I am severely anxious or depressed

I am extremely anxious or depressed

- 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 =



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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:

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