GUY'S & ST THOMAS' CHARITY
A CATALYST FOR INNOVATION IN HEALTH

Guy's and St Thomas' NHS
NHS Foundation Trust

UK National XP Service

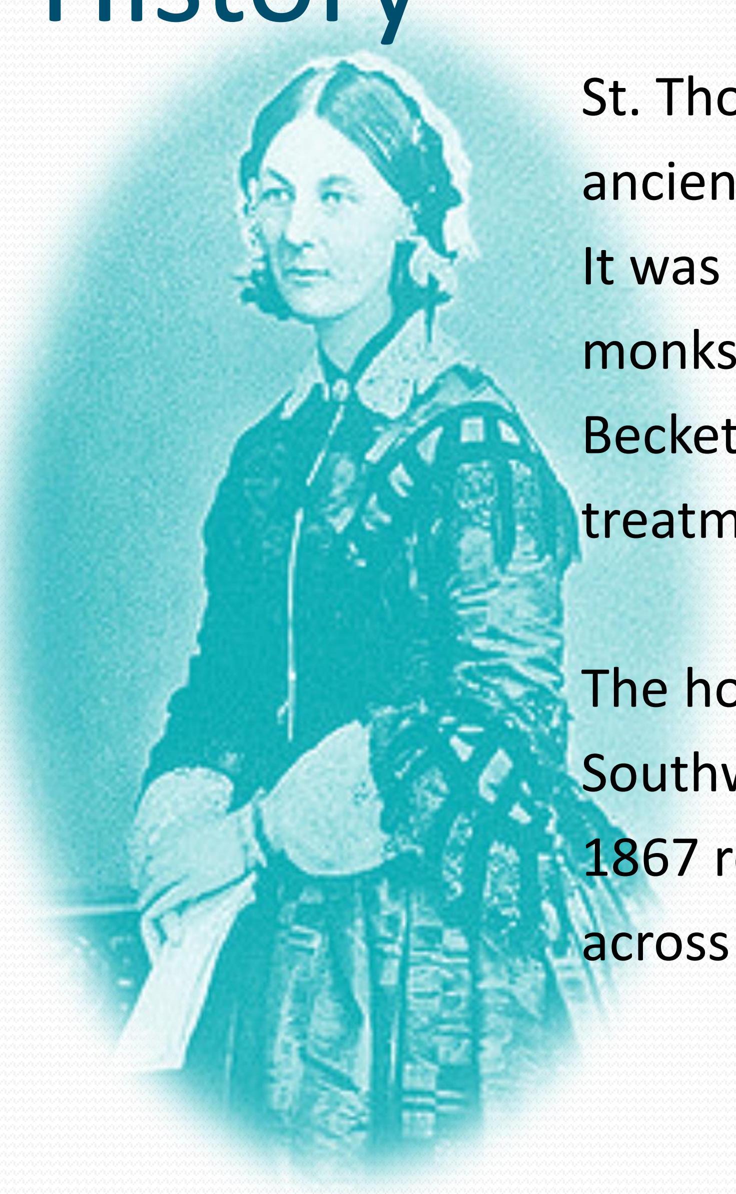
Guy's & St Thomas' Hospital

St Johns Institute of Dermatology

NHS England

Tanya Henshaw XP CNS for Adults 2016

History



St. Thomas's Hospital was described as ancient in 1215.
It was a mixed order of Augustinian monks and nuns, dedicated to Thomas Becket which provided shelter and treatment for the poor and sick.

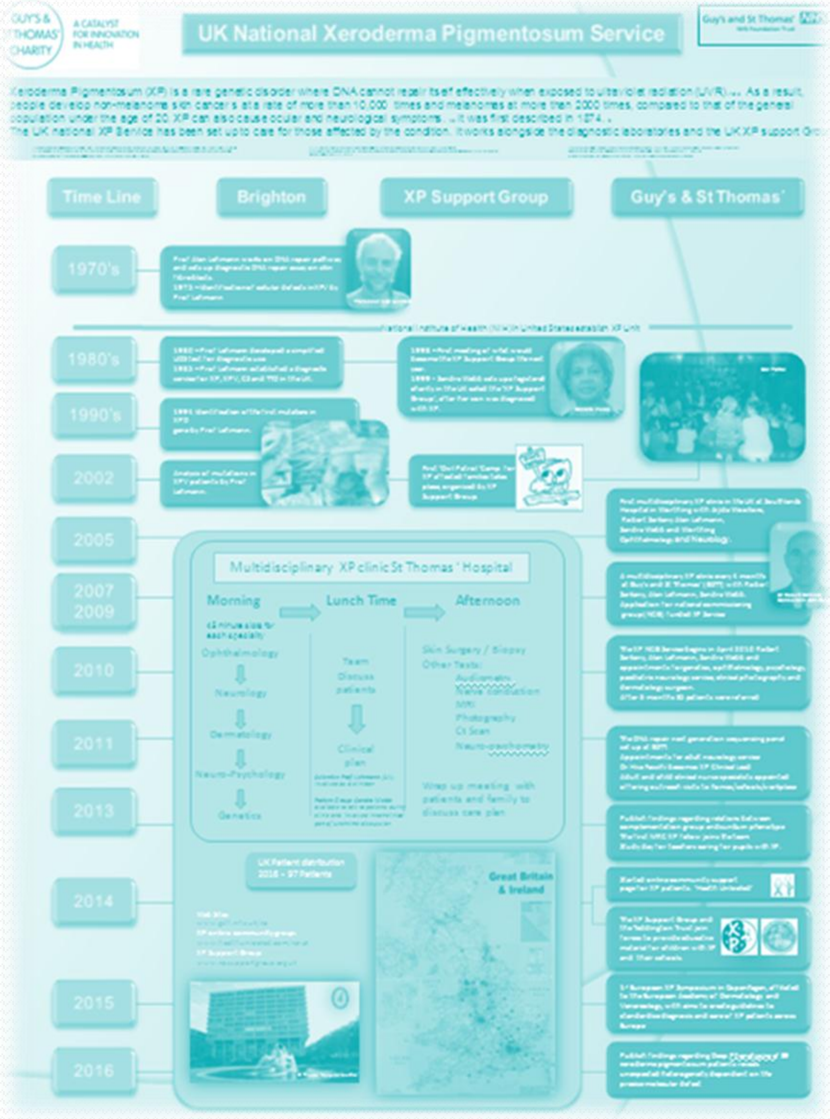
The hospital was originally located in Southwark, just south of London Bridge.
1867 rebuilt at the present Lambeth site across from the houses of Parliament.

Before XP National Service

- Patients seen by different dermatologists around UK
- Very few had easy access to a wider medical team
- No recognised standards of care
- Misconceptions about diagnostic indicators
- Misdiagnosis , late diagnosis or not diagnosed at all

- XP Support Group - advice and support

Evolution of the clinic



UK National Xeroderma Pigmentosum Service

Xeroderma Pigmentosum (XP) is a rare genetic disorder where DNA cannot repair itself effectively when exposed to ultraviolet radiation (UVR)... As a result, people develop non-melanoma skin cancer's at a rate of more than 10,000 times and melanomas at more than 2000 times, compared to that of the general population under the age of 20. XP can also cause ocular and neurological symptoms. ..It was first described in 1874..

The UK national XP Service has been set up to care for those affected by the condition. It works alongside the diagnostic laboratories and the UK XP support Gr

Multidisciplinary XP clinic St Thomas' Hospital

Ophthalmology → Neurology → Dermatology → Neuro-Psychology → Genetics

Team Discuss patients → Clinical plan

Skin Surgery / Biopsy
Other Tests:
Audiometry
Nerve conduction
MRI
Photography
Ct Scan
Neuro-psychometry

Wrap up meeting with patients and family to discuss care plan

Founders:

- Professor Alan Lehmann
- Sandra Webb
- Dr Robert Sarkany

After 5 years of planning and various formats, the XP NCG Service begins in April 2010.

After 6 months 35 patients were referred

Present day XP

- XP in the UK: 97patients
- UK population: 60 million
- Prevalence: 1.48/million

2010: National Health Service
‘ Rare Disease funding ’ to establish a National XP Service

Diagnostic Laboratories

DNA Repair assays

Next generation sequencing of all DNA repair genes

Patient Support Group

Multidisciplinary Clinic

- Photodermatology
- Dermatological Surgery
- Ophthalmology
- Neurology
- Neuropsychology
- Clinical Genetics
- Dermatopathology
- Visiting clinicians

Nurse-run outreach network

visit patients in their homes, schools and work place

The XP Multidisciplinary Clinic

2-3 clinics per month

Morning:

45 mins with specialist

- **Eyes**
- **Skin**
- **Neurology**
- **Neuropsychology**
- **Genetics**

→

Lunchtime:

Team Discuss Patients
Clinical plan agreed

Scientist: Prof. Alan Lehmann: *fully involved as a clinician*

→

Afternoon:

- Skin surgery
- Documentary Photography
- Blood tests
- *Audiometry*
- *MRI*
- *Nerve conduction*
- *Neuropsychometry*
- *CT Scanning*

Wrap up:

Summarising discussion with family

Patient Support Group: Sandra Webb: *available to talk to patients through out the day. Involved in non-clinical part of lunchtime discussion*

Aim of XP Service

- All Pts diagnosed with XP are offered referral to the service
- Continuity of care
- Standardised care
- Regular Skin monitoring patient/clinic
- Minimise any surgery, confocal microscopy/Mohs surgery if required.
- Support
- Advise local teams
- Attending conferences spreading the word
- Research

Getting the message across

- Eight complementation groups
- XP presents in different ways
- Not all patients have sun burning
- Distinct freckling patterns with areas of sparing
- Hyper and Hypo pigmentation
- Ocular surface damage
- Freckling from the age of two
- Multiple skin cancers of all types
- Early skin cancer
- Prolonged and severe sun burn
- Neurological degeneration of unknown causality

Role of the XP CNS

- Key worker for XP patients
- Organise all-day multi-disciplinary XP clinics
- Ensure equitable access to service
- Outreach visits to homes, schools, work place
- Promote awareness of XP
- Develop patient pathways
- Patient information leaflets
- Research
- Service evaluation and development
- Audit
- Skin biopsy and basic surgery
- Camouflage advice
- Social services

Nursing issues

Paediatric

- Getting photo protection in schools
- liaising with schools
- Growing families and genetics counselling
- Transition years
- Age appropriate education

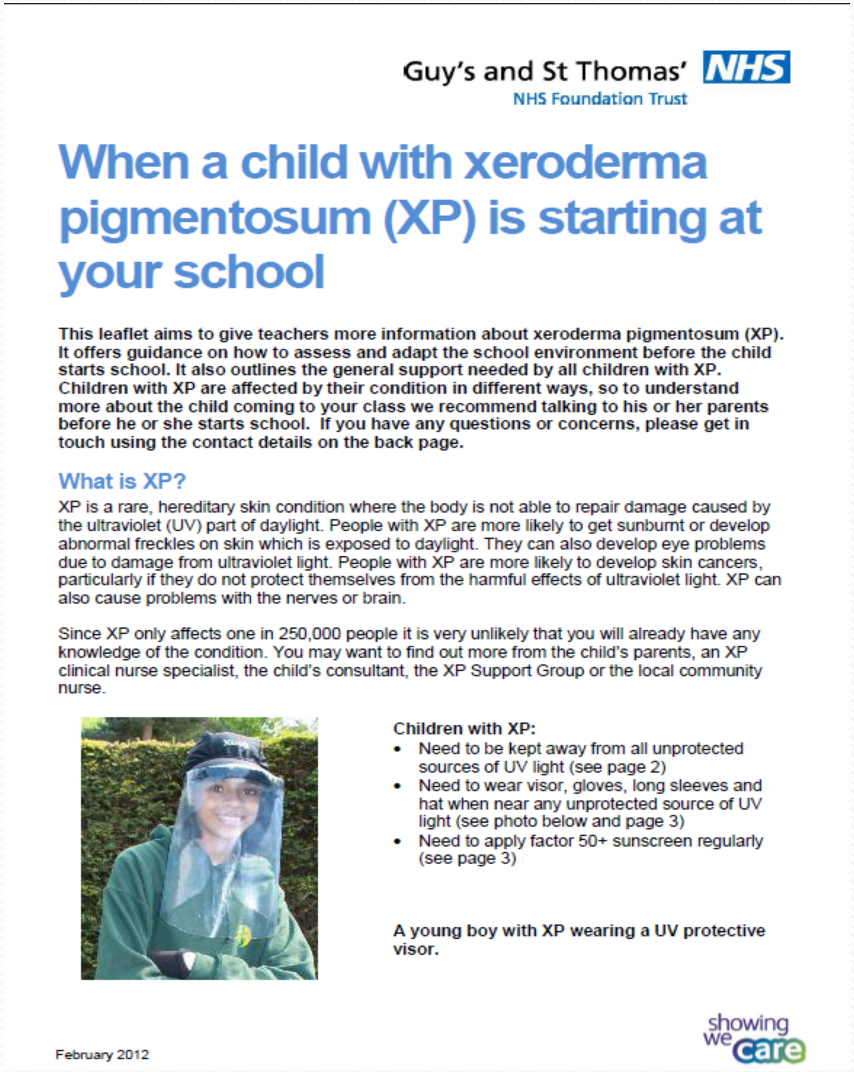
Adult

- Social services
- Education, encouraging adults to protect
- Helping in the work place
- Neurological degeneration support
- Multiple surgeries

Teachers study days

- Written information about XP and
- Schooling

- Started yearly XP study day for teachers



Guy's and St Thomas' NHS
NHS Foundation Trust

When a child with xeroderma pigmentosum (XP) is starting at your school

This leaflet aims to give teachers more information about xeroderma pigmentosum (XP). It offers guidance on how to assess and adapt the school environment before the child starts school. It also outlines the general support needed by all children with XP. Children with XP are affected by their condition in different ways, so to understand more about the child coming to your class we recommend talking to his or her parents before he or she starts school. If you have any questions or concerns, please get in touch using the contact details on the back page.

What is XP?

XP is a rare, hereditary skin condition where the body is not able to repair damage caused by the ultraviolet (UV) part of daylight. People with XP are more likely to get sunburnt or develop abnormal freckles on skin which is exposed to daylight. They can also develop eye problems due to damage from ultraviolet light. People with XP are more likely to develop skin cancers, particularly if they do not protect themselves from the harmful effects of ultraviolet light. XP can also cause problems with the nerves or brain.

Since XP only affects one in 250,000 people it is very unlikely that you will already have any knowledge of the condition. You may want to find out more from the child's parents, an XP clinical nurse specialist, the child's consultant, the XP Support Group or the local community nurse.

Children with XP:

- Need to be kept away from all unprotected sources of UV light (see page 2)
- Need to wear visor, gloves, long sleeves and hat when near any unprotected source of UV light (see photo below and page 3)
- Need to apply factor 50+ sunscreen regularly (see page 3)

A young boy with XP wearing a UV protective visor.

February 2012

showing we care

Work with XP Support Group

- Day at Owl Patrol each year.
- Contribute to the news letter
- Collaborate to produce leaflets

Kira-Nerys...Nathaniel...Mark...

The positives

"Having XP or a related condition means that you know that image isn't everything."

"It makes you a more caring and accepting person."

If you have a sibling with XP or a related condition it can make you more thoughtful and accepting of others."

If you have XP or a similar condition you probably spend more time doing things as a family and that can bring you closer."

Alex...Awatif...Alex...Ben...Colli

The information in this leaflet was generated by teenagers on the Owl Patrol Camp organised by the XP Support Group in 2014 and compiled by Dr Isabel Garrood (Neuro psychologist) and Hermanus Louw (XP Clinical Nurse Specialist) at the XP Service at Guys and St Thomas' Hospital, London

Want to find out more? Got any questions?

Contact the XP Support Group on

01494 456192 or 01494 459888

Email: info@xpsupportgroup.org.uk

or contact

0207 188 6339

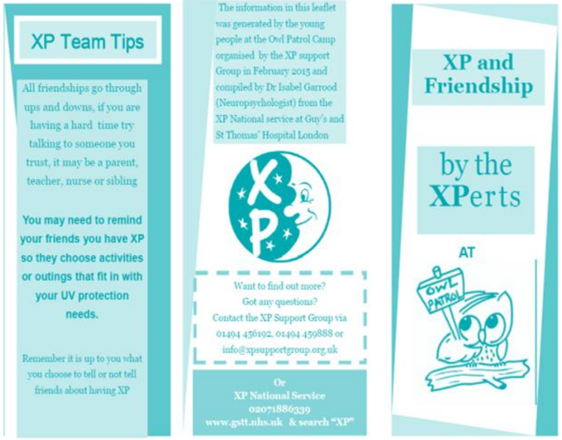
Email: sally.turner@gstt.nhs.uk

2014

Living with XP and other related conditions

by the **xp**erts

2014



XP Team Tips

All friendships go through ups and downs, if you are having a hard time try talking to someone you trust, it may be a parent, teacher, nurse or sibling

You may need to remind your friends you have XP so they choose activities or outings that fit in with your UV protection needs.

Remember it is up to you what you choose to tell or not tell friends about having XP

The information in this leaflet was generated by the young people at the Owl Patrol Camp organised by the XP support Group in February 2015 and compiled by Dr Isabel Garrood (Neuropsychologist) from the XP National service at Guy's and St Thomas' Hospital London

Want to find out more?
Got any questions?
Contact the XP Support Group via
01494 456192, 01494 459888 or
info@xpsupportgroup.org.uk

Or
XP National Service
02071886339
www.gstt.nhs.uk & search "XP"

XP and Friendship

by the **XP**erts

AT

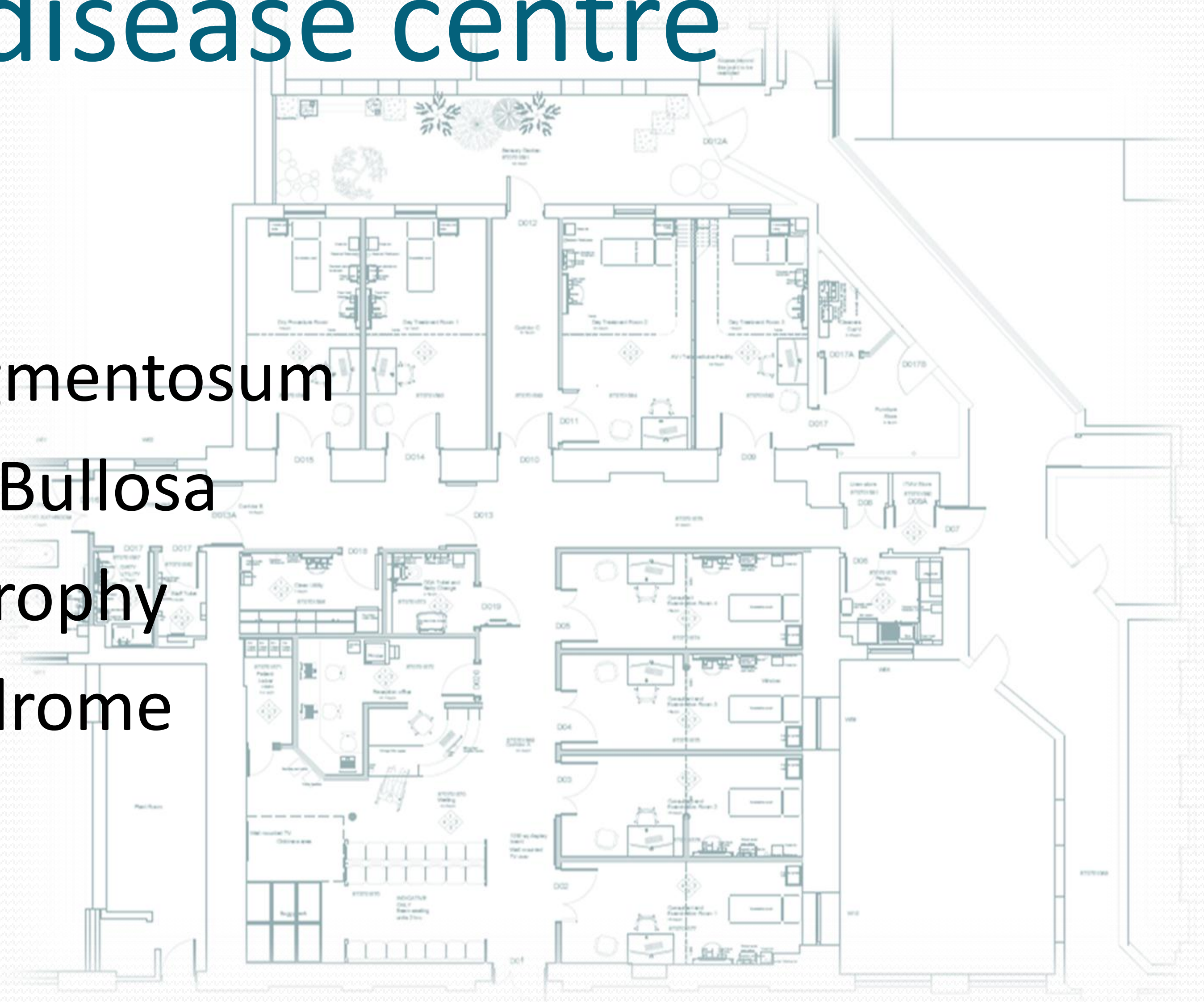
First European XP Society meeting Copenhagen 2015

Affiliated with the European Academy of Dermatology and Verenology (EADV)

Aims:

- Standardise diagnostic indices
- Agree on Standards of care
- Share knowledge
- Share research
- Working with the support groups

New Rare disease centre



- Xeroderma Pigmentosum
- Epidermolysis Bullosa
- Trichothiodystrophy
- Cockayne Syndrome

Articles /Publications

Fassihi H. **Spotlight on ‘xeroderma pigmentosum**’. *Photochem Photobiol Sci 2013, 12(1): 78-8*

Turner S, Mullard K, Fassihi H, Sarkany R. **Nursing patients with xeroderma pigmentosum in the UK**. *Dermatological Nursing 2013, 12(3): 20-6*

M.Sethi, A.R Lehmann, H.Fawcwtt, M.Stefanini, N Jaspers, K.Mullard, S.Turner, A.Robson, D.McGibbon, R.Sarkany, H.Fassihi (2013) **Patients with xeroderma pigmentosum complementation groups C, E and V do not have abnormal sunburn reactions**. *British Journal of Dermatology*

Sethi M, Lehmann AR, Fassihi H. **Xeroderma pigmentosum: a multidisciplinary approach**. *EMJ Dermatol 2013, **1: 54-63*** 10.

Turner S. **The Australian Travel Award: Just go for It!** *Dermatological Nursing. September 2015*

Fassihi H, Sethi M, Fawcett F, et al. **Xeroderma pigmentosum: Deep phenotyping of 89 xeroderma pigmentosum patients reveals unexpected heterogeneity dependent on the precise molecular defect**. *Proceedings of the National Academy of Sciences 2016, 113(9): 1236-45*

Morley S. (2016) Ocular Solar Protection in XP : **The role of untinted lenses in blocking Ultraviolet radiation**. BJD (2016) 175, pp 625-627

Henshaw T, Turner S. **XP and nursing management of children and adults**. *Dermatological Nursing 2016 15(2suppl): 25-26.*

Conclusion

- Raising awareness will lead to earlier diagnoses

- Meticulous UV protection, leading to better outcomes and quality of life.

- Having a national service for this rare condition means that patients and families can feel supported rather than isolated.

- Large cohort of patients with XP at one centre enables us to advance our knowledge of the condition

- Research that could lead to significant improvements in medical and nursing care.

- The XP CNS is central to the multi-disciplinary service, working with the patients' local health services to maintain continuity of care.

Sally Turner, Katie Mullard, Hiva Fassihi, Robert Sarkany Dermatological Nursing, 2013, Vol 12, No 3 PP21-26 Nursing patients with xeroderma pigmentosum in the UK

Contact us

Guy's and St Thomas' NHS
NHS Foundation Trust

St. John's Institute of Dermatology

Xeroderma pigmentosum (XP) service

Xeroderma pigmentosum (XP) is a rare, hereditary skin disorder affecting 1 in 250,000 people.

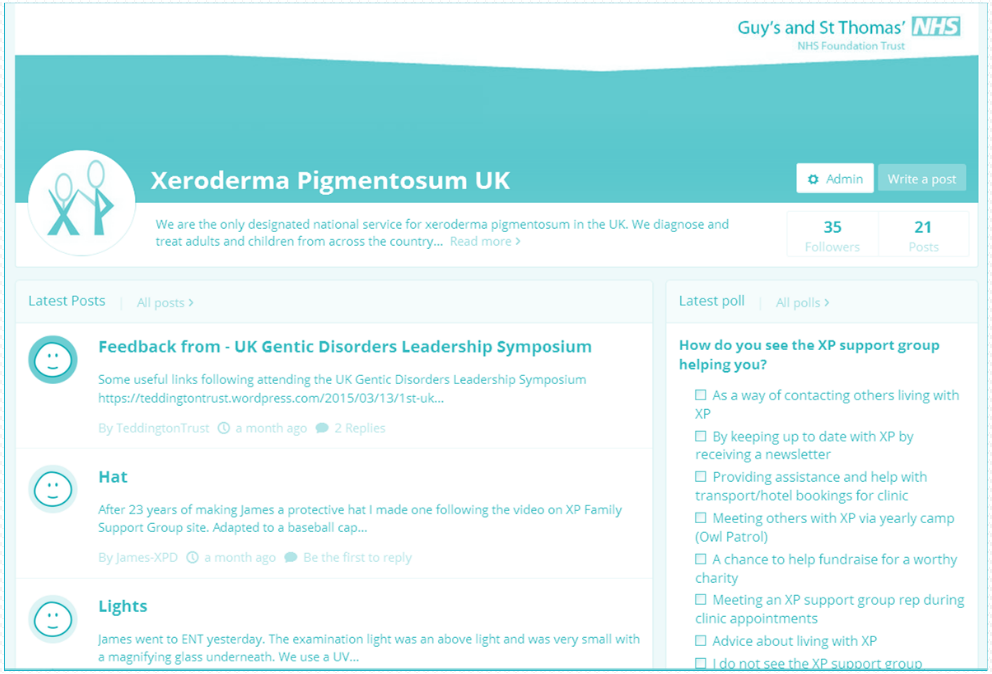
People with xeroderma pigmentosum are not able to repair the damage caused to their skin by the ultraviolet (UV) part of daylight. They can burn easily or develop abnormal freckles on skin that is exposed to UV. They can also develop eye, nerve or brain problems, and are more likely to develop skin cancers, especially if they do not protect their skin from the harmful effects of UV.

We are the only designated national service for xeroderma pigmentosum. We diagnose and treat adults and children from across the UK, working closely with our specialists in photodermatology, the dermatology surgery and laser unit, children's neurology, genetics, ophthalmology (eye), psychology and the neurogenetics department at University College Hospital.

Our patients see a wide range of health professionals at our clinic including dermatology specialists. Our specialist nurses visit patients at their home, school or workplace.

Join our online community

Follow the Xeroderma Pigmentosum UK community on HealthUnlocked, the social network for health.



Xeroderma Pigmentosum UK

We are the only designated national service for xeroderma pigmentosum in the UK. We diagnose and treat adults and children from across the country...

35 Followers
21 Posts

Feedback from - UK Gentic Disorders Leadership Symposium

Some useful links following attending the UK Gentic Disorders Leadership Symposium https://teddingtontrust.wordpress.com/2015/03/13/1st-uk...

By TeddingtonTrust a month ago 2 Replies

Hat

After 23 years of making James a protective hat I made one following the video on XP Family Support Group site. Adapted to a baseball cap...

By James-XPD a month ago Be the first to reply

Lights

James went to ENT yesterday. The examination light was an above light and was very small with a magnifying glass underneath. We use a UV...

How do you see the XP support group helping you?

- ☐ As a way of contacting others living with XP
- ☐ By keeping up to date with XP by receiving a newsletter
- ☐ Providing assistance and help with transport/hotel bookings for clinic
- ☐ Meeting others with XP via yearly camp (Owl Patrol)
- ☐ A chance to help fundraise for a worthy charity
- ☐ Meeting an XP support group rep during clinic appointments
- ☐ Advice about living with XP
- ☐ I do not see the XP support group

www.gstt.nhs.uk/xp

Healthunlocked.com/xp-uk

Thank you

- Guys and St Thomas Charitable Trust who enabled me to travel and attend the conference.

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ST THOMAS'
CHARITY

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IN HEALTH