UK National XP Service
Guy’s & St Thomas’ Hospital
St Johns Institute of Dermatology

Tanya Henshaw XP CNS for Adults 2016
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
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: 97 patients
- 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 throughout 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, workplace
- 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
Work with XP Support Group

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


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


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.
Contact us

Guy’s and St Thomas’ NHS Foundation Trust

St. John’s Institute of Dermatology

www.gstt.nhs.uk/xp

Healthunlocked.com/xp-uk
Thank you

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