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# **A review of photodiagnostic investigations over 26 years: experience of the National Scottish Photobiology Service (1989-2015)**

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## **Abstract**

**Background and Aims:** The Scottish Photobiology Service is the national referral pathway for patients with cutaneous photosensitivity diseases in Scotland. We reviewed the pattern of diagnosis of photosensitivity diseases and investigations performed between 1989 and 2015.

**Methods and Results:** Data were collected from the Photodiagnostic Database, annual reports and paper records. The total number of patients assessed each year was stable over the period studied (median 242 [range 231-266]), with most being new patients (median 69 [range 62-73] %).

Monochromator phototesting was the most utilised investigation, although the use of provocation testing and photopatch testing has increased. The commonest diagnosis was polymorphic light eruption and there was a trend to increasing diagnosis of photoaggravated atopic eczema.

**Conclusions:** The pattern of diagnosis of photosensitivity diseases remains fairly stable in Scotland and we wish to emphasise the importance of this Scottish specialist service for patients with photosensitivity diseases and referrers.

**Keywords:** Photosensitivity diseases, photodermatosis, photobiology, National Service

## Introduction

Abnormal photosensitivity typically presents either as a heightened exaggerated sunburn-like reaction to sunlight or daylight exposure, for example in patients taking photosensitising drugs, or as an abnormal rash, which occurs after sunlight exposure, such as with the commonly occurring “prickly heat” (polymorphic light eruption; PLE). Thus, the photosensitivity diseases are a heterogeneous group of disorders, which include common conditions, such as PLE and the uncommon and rare photosensitivity diseases: chronic actinic dermatitis (CAD), idiopathic solar urticaria (SU), actinic prurigo (AP), hydroa vacciniforme (HV), DNA repair diseases, particularly xeroderma pigmentosum, and some of the cutaneous porphyrias. They may range also from causing a minor disruption to life through to being a cause of major morbidity or rarely mortality, as for example in xeroderma pigmentosum. Although patients are often aware of the association between their condition and sun exposure, this is not always the case, particularly if the condition is caused by UVA and/or visible light, meaning that the patient will have problems all year round. Investigation of patients with suspected photosensitivity, in order to define conclusively whether they are or are not abnormally photosensitive and, if so, which wavelengths they are sensitive to and the degree and type of photosensitivity, is thus essential for accurate diagnosis, optimised management and holistic patient care.

Photodiagnostic testing is thus carried out in patients suspected to have abnormal cutaneous photosensitivity. It is undertaken worldwide in selected specialist centres, but in only a few sites in the UK, with there being only one in Scotland, the Scottish Photobiology Service (SPS). This Service is offered as a National Service for patients with photosensitivity diseases in Scotland and is funded through NHS National Services Scotland (NSS) and the National Services Division (NSD), which provides commissioning for Scotland’s health and facilitates access for patients to investigations and treatments of a highly specialised nature, often for rare diseases. The Photobiology Unit (PBU) at Ninewells Hospital in Dundee hosts the SPS and thus accepts referrals from across Scotland.

Phototesting involves use of an irradiation monochromator to determine if there is sensitivity to narrow wavebands of ultraviolet (UVA or UVB) radiation and/or visible light, with light irradiated on small areas of back skin.(1)

Other investigations may include:

\*Provocation testing to larger areas of skin using broadband UVA, UVB and indoor lighting sources

\*Minimal erythema dose (MED) testing to light sources that can be used therapeutically

\*Patch and photopatch testing

\*Serology to screen for lupus

\*Plasma spectrofluorimetry and porphyria biochemical analysis of blood, erythrocytes, urine and faeces

\*Vitamin D levels

\*HLA typing

\*Skin biopsy

Prevalence of photodermatoses can be estimated based on data from such photodiagnostic units, if it is assumed that the majority of cases pass through such a unit (likely to be a correct assumption for rare and severe diseases, but not for common conditions with a great range of severity such as PLE). For example, a study based on diagnoses made in the SPS, on the prevalence of photodermatoses in Scotland was published in 2009.(2) Other units have published data on photodermatoses as a percentage of total photodermatoses diagnosed, for example, in New York in 1995,(3) in Singapore in 2005, (4) or in Athens, Greece in 2003.(5) None of these studies attempted to estimate prevalence or incidence of photodermatoses; all found PLE to be the most common diagnosis made. Prevalence estimates have also been made from questionnaire-based surveys of populations, such as a study of the resident populations of areas within the Yunnan province in China,(6) and a survey of Swedish pharmaceutical company employees screened for PLE.(7)

In 2009, the Scottish study undertaken in the population reported a prevalence of 16.5 cases per 100000 population for chronic actinic dermatitis (CAD), 4.0 per 100000 for idiopathic solar urticaria (SU), and 3.3 per 100000 for actinic prurigo (AP). This study took into account the whole Scottish

population when estimating the prevalence of rarer diseases likely to lead to assessment in the PBU, finding a prevalence of 0.5 per 100000 for patients with hydro vacciniforme and 0.2 per 100000 cases of xeroderma pigmentosum.(2)

The **aim** of this study was to look at the diagnoses made in the Unit over time and whether there were any trends in diagnosis of the photodermatoses. We also assessed the incidence of diagnosis of photodermatoses in the Tayside population from data collected in the unit over 26 years. A secondary **aim** of the study was to assess the activities of the **SPS**, including the frequency and trends of investigations undertaken.

## **Materials and Methods**

Patients are referred from across Scotland to the SPS. The photodiagnostic database spans 1973 until the present day, with information on new patients investigated in the SPS, their investigations, results and final diagnoses. **The database originally consisted of patient records and investigation findings being kept on a card filing system (1972-1984) but this evolved into an in-house electronic database, which was adapted and transferred to an Access database in 2007.** This database was used to obtain retrospective data for the numbers of patients referred to the SPS, both new and return patients, **using predominantly digital data retrieval, with additional manual retrieval of data as required.** The location from which patients were referred to the unit, and how this has changed over time, were also assessed. Data on the number of new cases of photodermatoses assessed annually over 26 years were collected, and trends examined. The pattern of investigations undertaken over time was also assessed.

Each new patient is given a unique identifying number in the database. This ensures that return patients' records are not duplicated. Their record is amended, but not overwritten, so that their year of diagnosis remains the original year of assessment. Thus, our figures for diagnosis are only for new patients investigated in the SPS. Each patient is given a primary diagnosis and some may also have a secondary diagnosis. We have used the primary diagnosis for our figures.

Searches were performed for each photodermatosis individually, from 1989 until August 2015. The numbers for primary diagnosis of each photodermatosis per year were recorded. We also used the

search function in the database to split this into patients who were resident within the NHS Tayside catchment area and those from out with the NHS Tayside Health Board area. Census data (2001) were then used to calculate incidence per 100000 for Tayside patients, for each photodermatosis.

Database queries were performed to look into the numbers of each investigation performed from 1989 until 2015, in new patients investigated. The number of each investigation per year was also recorded.

'Others' in the database represented approximately 30% of diagnoses. The cards on which diagnoses were recorded originally were reviewed to obtain information on the actual diagnosis in these cases.

Access to NSS figures provided the total numbers of new/return patients and total investigations performed each year (April – March). Total investigations included minimal erythral dose (MED) testing for both TL01 and UVA which are not always part of the investigation of photodermatoses, but were included when they were performed in order to give a complete picture of the workload of the SPS. It should be stressed that of patients with cutaneous porphyrias, only the minority are phototested and, as the data assessed in this study only relate to patients who were phototested, the figures do not reflect the prevalence of the porphyrias.

## Results

The total number of patients seen in the SPS has remained stable over the period studied, ranging from 231-273 (median 244) per year, with the majority of these being new patients (62%-73%, median 69%). This reflects the agreed (with NSS/NSD who commission the service) service level of activity of 245 NHS patients in Scotland per year. The majority of referrals are from within Scotland, with less than 12% from the rest of the UK. Over time, the percentage of patients referred from outwith Scotland has reduced, reflecting the development of other units elsewhere in the UK.

Monochromator phototesting was the most frequently used investigation and is considered the gold standard photodiagnostic test used in the SPS. Use of monochromator phototesting remained stable

over the 26 years (median 162 (range 127-185) patients who underwent monochromator phototesting per year) (Figure 1a). Provocation testing was increasingly undertaken since 1992 (for example from only 77 patients who underwent provocation testing in 1991 through to a median of 111 (range 83-131) per year for the period 1992-2015) (Figure 1a), and photopatch testing similarly increased in use since 1995 (in only 29 patients in 1994; through to a median of 78 (54-107) per year for the period 1995-2015) (Figure 1a). However, solar simulator testing was performed less frequently (maximum use was in 1990 when 158 patients underwent solar simulator phototesting; with only a median of 5 (range 0-24) patients per year undergoing this investigation in the period 1993-2015) (Figure 1a). Skin biopsy was part of the investigation plan for a small number of patients (median of 13 (range 0-57) patients per year). Biochemical and immunological investigations were also performed where relevant, most commonly lupus screening since 1994, and Vitamin D testing since 2009 (Figure 1b).

We only collected information on the primary diagnosis. However, some patients will have had more than one diagnosis, for example an immunological photodermatosis combined with photocontact allergy.

We looked in detail at 2011 as we selected this as a typical, representative and relatively recent year within the time period studied (Figure 2a; Table 1). The commonest diagnosis was PLE (31%). This was followed by: photoaggravated atopic eczema (AE, 7%); CAD (6%); SU (2%) and AP (1%) (Figure 2a). 20% of patients had not received a formal diagnosis, usually because of atypical or evolving clinical and investigation findings (these patients would have returned for further investigation on other occasions) or had photosensitivity as a diagnosis excluded. 'Other' diagnoses included photoaggravated seborrhoeic eczema (3), rosacea (5), psoriasis (3) or non-specified dermatitis (13) or contact allergy and lupus erythematosus, and constituted 30% of the patients seen in 2011 (Table 1).

Thus, PLE was the commonest diagnosis made. However, the numbers diagnosed in the SPS with this condition have fallen over time (Figure 3a), although the numbers per 100,000 population diagnosed in Tayside remained stable at 4-11 per 100000 per year (Figure 4a).

Comparing the diagnoses of CAD and AE, showed a trend for an increasingly frequent diagnosis of AE (Figure 3b). In Tayside, the number of new cases referred and diagnosed with CAD per 100,000



per year was 0-2, with no clear trends (Figure 4b). There was no trend in the numbers of patients diagnosed with solar urticaria in the SPS as a whole.

Regarding the other rare photodermatoses, there was no trend in the numbers of patients diagnosed with xeroderma pigmentosum. There were fewer patients diagnosed with actinic prurigo and hydroa vacciniforme over time (Table 2).

Finally, the numbers of patients with drug-induced photosensitivity being referred and diagnosed in the unit reduced over time. This was true for both total numbers of patients to the SPS and the local incidence of diagnosis in Tayside (Table 2, Figure 4c).

## **Discussion**

The SPS, hosted by the Photobiology Unit at Ninewells Hospital, Dundee is the National Service for investigation of patients with photosensitivity diseases and is available for all patients throughout Scotland.

Although we have data for numbers of diagnoses per year, we cannot extrapolate these data to cover the whole of Scotland, or even the area of Tayside in the case of many conditions. An example is PLE, as most patients with this condition are not referred for phototesting, the diagnosis being made clinically instead. It is likely that the numbers of diagnoses of PLE in the SPS has fallen due to increased education and training of referrers and referral only for those with severe or atypical disease, and to exclude other photodermatoses. Thus, our data show a reduction in referral rates to the SPS for this condition. The stability of the numbers of cases diagnosed in the local population may be explained by the fact that patients may have a higher threshold for travelling long distances if they have milder seasonal PLE as we do not consider that we are over-diagnosing PLE in the local population.

We were encouraged to see increased numbers of patients diagnosed with photoaggravated atopic eczema as this reflects increased referrals of this patient group in whom it may be clinically impossible to distinguish between CAD and photoaggravated AE. It is important to phototest in these

cases as rare photodermatoses, in particular CAD, must be excluded in this group, as the management approach would be very different. For example, in photoaggravated AE, monochromator phototesting will typically be normal and UVB phototherapy can be very effective, whereas in CAD, monochromator phototesting will be abnormal, typically showing broadband and often disproportionately UVB-weighted abnormal photosensitivity and UVB phototherapy would be contraindicated as a therapeutic approach.

Drug-induced photosensitivity is being referred to the unit less, and this is likely to be a reflection of increased education and training of referrers and awareness of the entity of drug photosensitivity by clinicians, thus appropriately stopping incriminating drugs and reassessing without the need for formal investigation. We continue to see our role in education and training as being important in this regard.

Although we have data for 26 years' worth of referrals to the unit, it was not possible to obtain figures on incidence of the rarer photodermatoses, for example, xeroderma pigmentosum, which has an incidence quoted in the literature of 2.3 per million live births in Western Europe.(8) However, it is likely that for these rarer severe diseases that most cases would be referred to the SPS and the reduction in the number of cases of hydroa vacciniforme and actinic prurigo diagnosed over recent years, may well reflect a true reduction in incidence, although we are aware of the need for ongoing training to raise awareness of these rare diseases and to reduce delays in diagnosis.(9)

Finally, the SPS also offers the National Porphyrria Service. However, many patients with suspected porphyria are not phototested, and instead have biochemical investigations and diagnosis. Importantly, the data presented only relate to patients who were phototested and not to the majority of porphyria patients who are investigated through laboratory biochemical investigations alone.

In summary, we have shown that there is a trend for increasing diagnosis of photoaggravated atopic eczema, reflecting an important increase in the numbers of these patients with difficult complex presentations being referred to the SPS for investigation and management. We have shown reduction in the numbers of patients referred with drug-induced photosensitivity, which likely reflects improved education and awareness of this condition among clinicians. Finally, for rare diagnoses such as hydroa vacciniforme and actinic prurigo, we have shown a reduced number of patients diagnosed with these conditions over time.

Chronic and severe photosensitivity of any cause has major adverse impact on quality of life for patients.(10) The numbers of patients seen with the rare photosensitivity diseases of CAD, SU, HV, AP, XP and other DNA repair disorders and the rare cutaneous porphyrias are low and as such it is even more important to highlight awareness of these diseases. An ongoing challenge and priority for the SPS is to increase understanding and awareness of the cutaneous photosensitivity diseases and that they can present in a myriad of heterogeneous and often atypical ways. Awareness of the availability of the SPS and uptake by referrers for centralised, coordinated multidisciplinary investigations and management of patients with severe, complex and often rare disabling photosensitivity disorders is of crucial importance for optimised care. We are exploring ways to disseminate knowledge and uptake of the SPS through Scotland-wide multi-disciplinary team meetings and at international level through involvement in the European Rare Disease Network for Skin, in which we will play key roles in developing the inclusion of the photosensitivity diseases.

### **Acknowledgements**

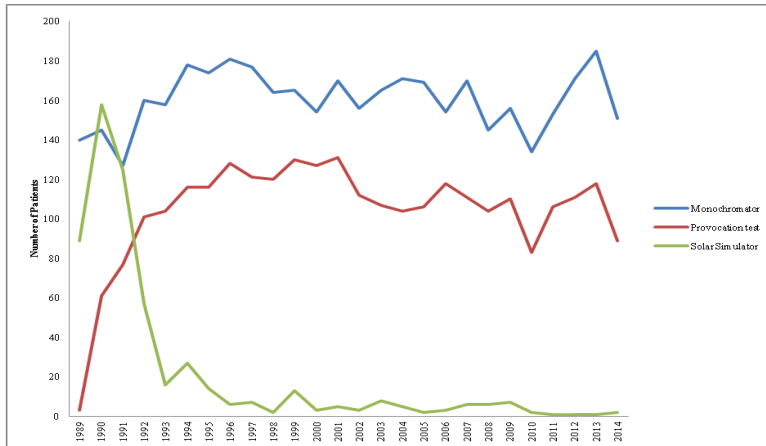
We would like to thank Lynn Fullerton, senior clinical technologist in the Photobiology Unit, for her help with the use of the photodiagnostic database and all technical queries.

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## Figures & Tables

**Figure 1a. Phototesting, 1989-2014.**



**Figure 1b. Other Investigations, 1989-2014. \*Vit D only recorded 2009 onwards**

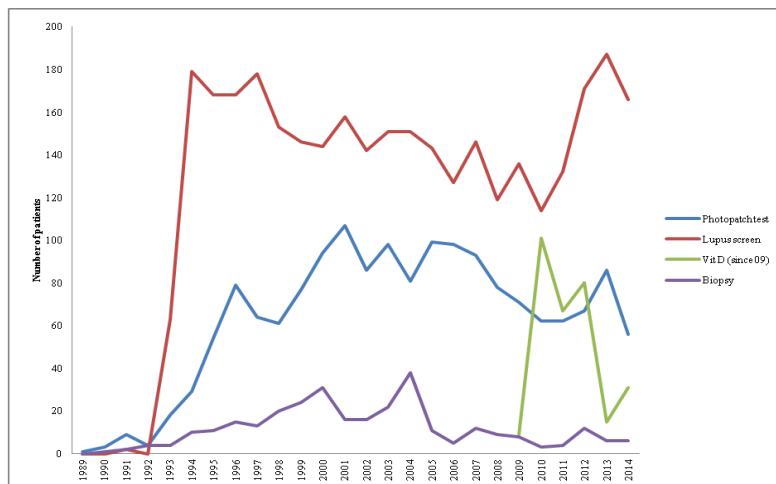


Figure 2a. All diagnoses, 2011

("Others" are detailed in Table 1)

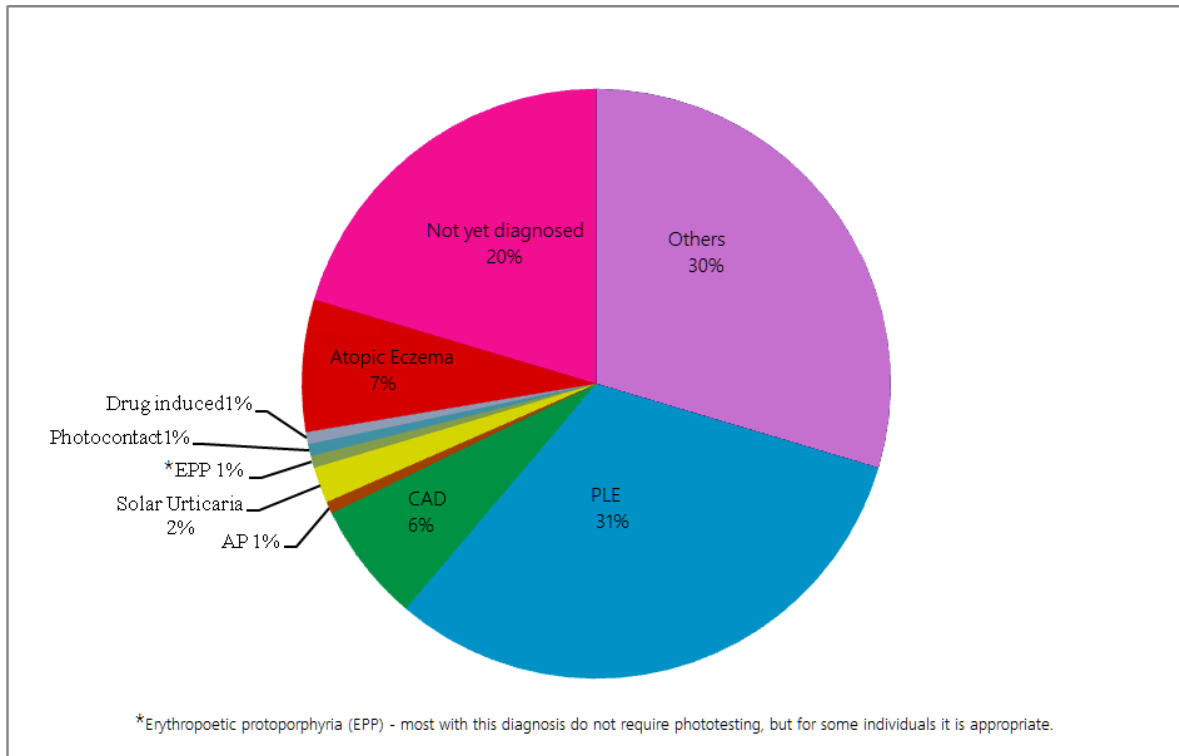
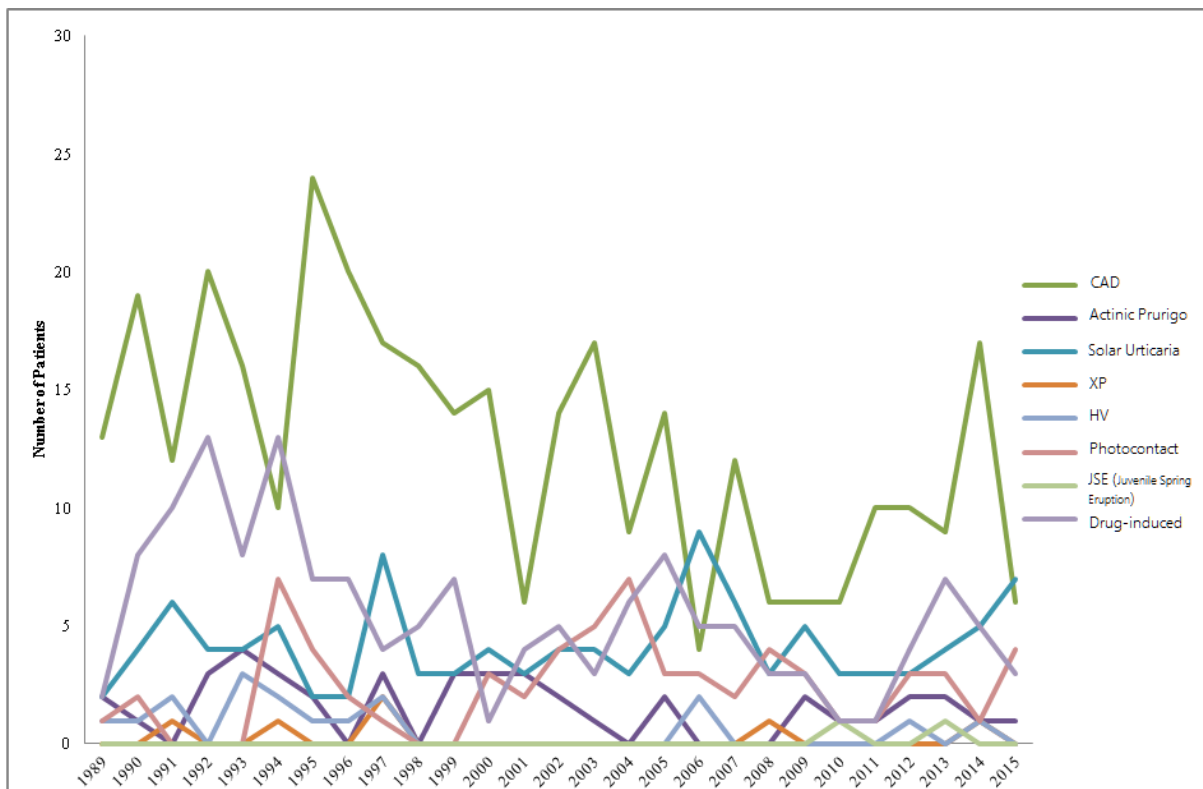
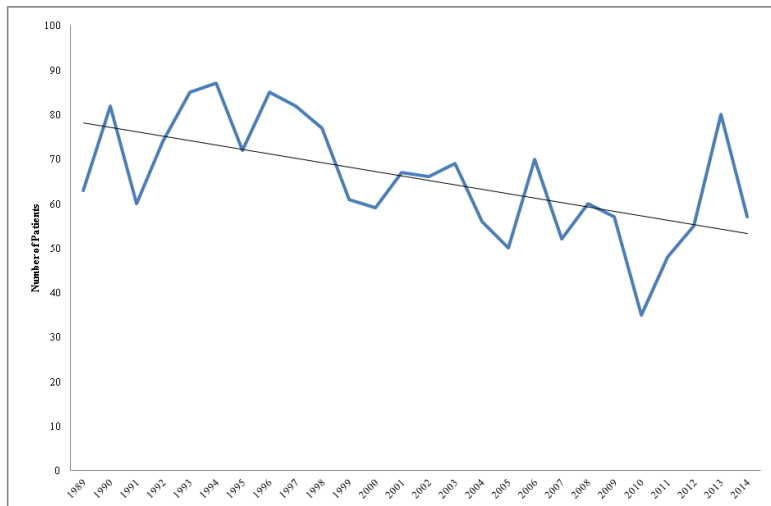


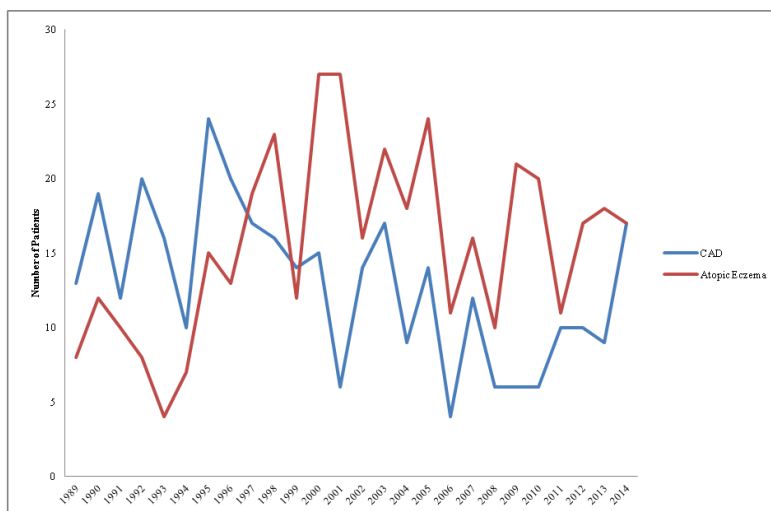
Figure 2b. All photodermatoses diagnosed, numbers per year, 1989 – 2015 (Excluding PLE)



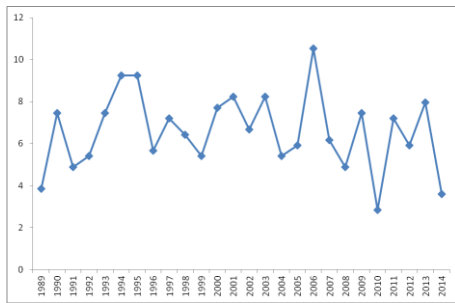
**Figure 3a. PLE Diagnoses, 1989 – 2014**



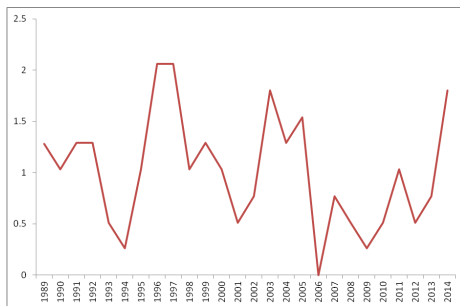
**Figure 3b. CAD and photoaggravated Atopic Eczema diagnoses in phototest patients, 1989 – 2014**



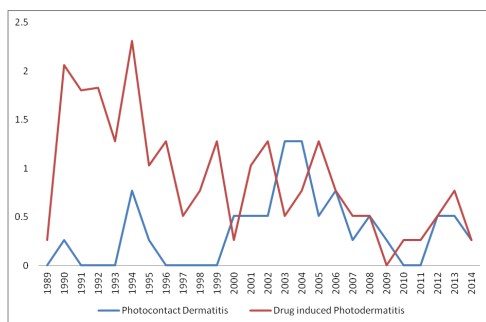
**Figure 4a. PLE**



**Figure 4b. CAD**



**Figure 4c. Photocontact Allergy and Drug Induced Photosensitivity**



**Figures 4a - 4c. Number of New Cases Diagnosed per 100,000 per year in Tayside patients referred for phototesting, 1989-2011**



**Table 1. Other Diagnoses, 2011**

<b>Diagnosis</b>	<b>Number of Patients</b>
Photoaggravated (including seborrhoeic dermatitis, other dermatitis, psoriasis, rosacea, cutaneous T-cell lymphoma)	24
Contact dermatitis / allergy	6
Lupus erythematosus	2
Dermatomyositis	1
Telangiectasia	1
Urticaria	1

**Table 2. Rarer Diagnoses, 1989 – 2015.**

<b>Diagnosis</b>	<b>Number of cases</b>		
	<b>1989-1997</b>	<b>1998-2006</b>	<b>2007-2015</b>
Solar urticaria	37	38	39
Actinic prurigo	18	14	10
Hydroa vacciniforme	13	2	2
Xeroderma pigmentosum	4	0	2
Photocontact	17	27	22
Drug-induced	72	44	32