

A PATIENT'S JOURNEY

Pemphigus vulgaris

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Twelve years ago, **Siri Lowe** developed pemphigus vulgaris, a potentially fatal autoimmune blistering disorder. As her story shows, it can have a devastating impact on patients' lives, as can the side effects of the necessary treatments

When my mouth and tongue suddenly became badly blistered in 1995, I never suspected there could be anything seriously wrong with me—this was surely just some crazy infection in my mouth. But it was pemphigus vulgaris, a rare, life threatening, autoimmune disease affecting skin and mucous membranes, and, of course, I'd never heard of it in my life.

I deteriorated quickly and in only five weeks moved from being strong and healthy to being unable to eat solid food, drinking through a straw, and being in agonising pain. I remember walking the London streets in a total daze thinking, "This can't be true, it's like the script of a bad television play." I had no idea how living with a chronic disease would change my life.

There's no cure for pemphigus. No smart drug to take it all away. It is nearly always controllable, but that control comes at a heavy cost. At that time, my only options were high dose corticosteroids or heavy immunosuppressant treatment. Not only had I a life threatening disease but my future suddenly became ruled by serious prescription drugs.

The early days

During the first three years, the drugs themselves brought their own health problems, some serious and some just hard to live with. I was hugely grateful that my disease wasn't as bad as some people's—at least my skin hadn't literally blistered off my body—but it gradually began to affect nearly every area of mucous membrane: mouth, nose, throat, eyes, vagina, cervix.

Coping with the condition was extremely difficult. The slightest change in drug regimen could disturb the precarious balance, and instead of controlling the disease I'd be tumbling down into another crisis. Those years were a constant nightmare until my consultant dermatologist found a drug combination that I could tolerate and which worked (with only minor flares).

I secretly hoped my "journey" would be over once I got this far. Consequently, I found it very hard to

accept that my health was badly damaged and I couldn't return to my old self. High dose corticosteroids and immunosuppressants had been necessary, and undoubtedly saved my life. But there were side effects, and I was now disabled with problems with walking and pain. Prescription analgesics and pain management techniques could do no more than modify the pain levels. I had changed.

Six months after becoming ill, it became clear I couldn't return to work. I had to retire. Suddenly I was no longer part of a dynamic publishing company. I was also far too ill to continue with the voluntary work I'd trained to do, and the hobbies I'd enjoyed before were now either physically impossible or too exhausting. By the time my condition was more stable, I'd lost many of the things that defined who I was. I'd lost my financial viability and my identifiable role in the world. I was also a claimant, constantly worrying about the next set of forms to be filled in. All I had was the illness. I was an illness and nothing but an illness.

The people around me

But that's the downside. I also had wonderful companions. My surviving family were elderly and frail and needed to be protected from some of the unpleasant facts, but I had the constant presence of many loving and supportive friends. With them, I was able to explore new ways of relating to people. I'd usually been the "helper." Now I had to learn to accept help, and it wasn't an easy lesson. I started to realise that I could still have a sense of identity but that it would have to be a different one.

USEFUL RESOURCES

- International Pemphigus & Pemphigoid Foundation (www.pemphigus.org) in the US has an excellent medical advisory board; provides information and support; offers an email discussion group and online live chat
- Pemphigus Vulgaris Network (www.pemphigus.org.uk) is the support group started for the UK, which provides information and support and focuses on things specific to the UK (we work closely with the International Pemphigus & Pemphigoid Foundation)
- The Electronic Medicines Compendium (www.emc.medicines.org.uk) gives full and up to date drug information, essential for pemphigus patients, and is produced by the Association of the British Pharmaceutical Industry (ABPI)
- The Skin Care Campaign (www.skincarecampaign.org) for the wider picture of issues affecting pemphigus patients

This is one of a series of occasional articles by patients about their experience of traumatic medical events that offer lessons to doctors. The *BMJ* welcomes contributions to the series. Please contact Peter Lapsley (plapsley@bmj.com) for guidance.

A doctor's perspective

Pemphigus is a serious autoimmune blistering disorder caused by circulating autoantibodies to the epithelial adhesion proteins desmoglein 1 and 3. These antibodies result in a failure of epidermal cells to adhere correctly to each other. This causes flaccid blisters and subsequently painful erosions in the skin and mucous membranes.

Before the introduction of systemic glucocorticosteroids in the early 1950s, pemphigus was a universally fatal disorder with extensive involvement of the skin and mucous membrane leading to prostration, sepsis, and failure of multiple organs. The use of steroid sparing immunosuppressants such as azathioprine, cyclophosphamide, methotrexate, and ciclosporin has been the turning point in the management of the disorder. Recently, rituximab (an anti-CD20 antibody which targets B cell differentiation) administered together with intravenous immunoglobulin has been shown to be another advance. Pemphigus is therefore now eminently treatable.

Ms Lowe's account does show, however, just what an unpleasant disorder pemphigus vulgaris is, and what a tremendous toll it has taken on her personal life. She describes how completely devastated she was by the diagnosis and how the treatment, particularly systemic corticosteroids, interfered with her wellbeing. She also had ready access to information particularly via the internet, and this must have been quite frightening to read.

As the physician central to her case for the past 10 years, I have found my role to be largely guiding Ms Lowe through her illness and adapting the use of steroids, azathioprine, and ciclosporin to fit her personal needs. It was possible to induce a complete remission fairly early on, although she did have a relapse five years ago, and she still worries that she might relapse again. During this time, she has also developed breast cancer, and was clearly concerned what effects surgery and intubation during anaesthesia might have on the disease. The dermatologist working in the hospital was able to liaise with her other specialists, explain her condition, and orchestrate her care, so that she managed psychologically to survive the trauma of her second illness.

The lesson that was reinforced to me about caring for her and for most patients with serious skin disorders is how essential adequately resourced specialist dermatology services are.

Ms Lowe has fought her illness with great fortitude. A measure of this has been the determination with which, with the support of the British Association of Dermatologists, she set up the first patient support group for pemphigus, which has been of immense help to other people with the condition.

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WHAT WORKS WELL AND WHAT DOESN'T

What works well

- The therapeutic alliance between doctor and patient when it happens. I don't know how I could have got through the past 12 years without the close alliance I have with my doctors, particularly my consultant dermatologist. I'm convinced it increases my healing resources and helps get me through the worst times. There doesn't seem to be a channel to thank any of them properly—which is a pity.
- Continuity of care. In the changeable world of pemphigus I need the stability of the same doctors treating me over a period of time and knowing my case.
- When doctors cut through bureaucratic red tape to share essential information, such as an ear, nose, and throat specialist directly contacting an anaesthetist regarding special procedures.

What works less well

- There's no psychological space to say how scared I am—permanently. I think I'd risk almost anything else happening rather than go back to that terrible place of pemphigus crisis—skin dissolving, unbearable pain, and 80 mg/day of steroids.
- When people don't take pemphigus seriously. For example, it doesn't matter how brilliant surgery is if healing is messed up in a hospital ward where no one listens if I say, "I'm immunosuppressed, you need to be really careful about infections," "I've got a serious skin condition, if you use that dressing I'll have a bad reaction." This is the downside of health care—when the patient isn't believed and skin disease doesn't count as important. Not only is it physically dangerous, but it pushes me to a point of emotional despair which hasn't got easier over time.
- Confusion of language. When doctors talk about "remission" they mean disease control with drugs, but the rest of us think remission means a cure and no further need for drugs.
- The pressure to always be positive. No one wants a "heartsink" patient, that was made clear to me very early on. So I end up saying I'm doing well when what I mean is that the pemphigus is doing well, but the unwanted effects of my drugs mean I feel awful most of the time.

One of the ways this happened was as a direct result of my condition. In the early days one of the things I had found most difficult was getting any information about pemphigus and what it was like to live with. How many people survived? Did everyone feel as awful as I did taking the drugs? Then a friend discovered the International Pemphigus Foundation in the United States, and I was able to get those myriad questions answered. It was so much easier to cope once there was someone else with pemphigus to correspond with. I still remember that sense of relief. So, together with friends, I started the UK patient support group, which is still running today.

Of course, my constant companions are the doctors who look after me, primarily my consultant dermatologist, who has stayed consistently committed to my care. Because pemphigus has affected my body in so many different ways, I also see seven other consultants in four different hospitals. Miraculously, I've mostly been able to stay with the same specialists. This continuity makes such a difference. It's grim enough spending so much time waiting in hospital outpatient clinics, but it's really awful having to explain, yet again, to yet another doctor, the problems that are important to me. Hospital notes should do this, of course, but somehow they never do.

Other serious medical conditions have added to the difficulties. I've had breast cancer in both breasts—five operations, radiotherapy, etc. Suddenly I can find myself seeing doctors or nurses who don't believe a skin disease is serious. At times it seems impossible for people to see that my needs as a cancer patient can't be dealt with in isolation from my needs as a pemphigus patient. My calm dissolves in the insanity of trying to communicate with a health professional who doesn't believe patients can know what they're talking about: I just have to remember all the fantastic doctors and nurses who can see the wider picture. Luckily this problem doesn't occur often.

Where am I now?

I've learnt to live in new ways: to celebrate the good days and try to use them. Equally, I have to accept the bad times—even if that means days or weeks when all I can be is the “sick” person. My condition can change rapidly, so I have to stay ready to adapt within days.

Emotionally it's hard to shift paradigms at the drop of a hat. There's no point being permanently optimistic, for then it feels catastrophically painful to slip down again. Conversely, there's no point being permanently pessimistic, for then there'd be no joy in life, no

possibility of experiencing each moment for itself.

I would love to have been more upbeat describing this journey, but the truth is more complex. Of course, I hope that medicine will provide a real cure, though there's no scientific indication of that on the horizon. So I can only hope that pemphigus, the drugs, pain, and disability will leave me enough space to live an altered, but still worthwhile, life.

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LESSON OF THE WEEK**Alcohol hand rubs: hygiene and hazard**

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Young or confused people and those dependent on alcohol are at risk from ingestion of alcohol hand rubs

Hospital acquired infections are common and increase morbidity, mortality, and length of stay.^{1 2} The benefit of hand washing has been highlighted,³ and studies have shown that alcohol hand rub is significantly better than soap in reducing hand contamination.⁴⁻⁷ In 2004, NHS trusts were advised to introduce “near patient” alcohol based hand rubs (for example, attached beside beds and at entrances to wards) by April 2005.⁸

With the widespread introduction of these hand rubs, risk assessments were made for the possibility of fire and ingestion. Although alcohol hand rub is flammable, no incidents involving fire have occurred. The risk of ingestion was thought possible in children and confused patients, but little advice was given on reducing this risk.⁸ Frequent dermal application of alcohol based hand rub has been shown not to raise blood ethanol levels.⁹ However, no further risk assessment has examined unintentional or intentional ingestion of alcohol rubs.

We report a case of considerable toxicity associated with the deliberate ingestion of alcohol based hand rub,

together with data from our poisons unit showing an increase in inquiries related to alcohol rubs.

Case report

A patient known to have a history of alcohol dependency presented with withdrawal symptoms, having not consumed alcohol for 24 hours. On examination the patient was alert, orientated (with a Glasgow coma score of 15/15), with tachycardia (95 beats/min), tremulous, and sweaty, but with no focal neurological abnormalities. Intravenous thiamine and chlordiazepoxide were started under the hospital alcohol withdrawal protocol. During this admission, the patient was found collapsed (coma score 3/15), having vomited in the bathroom, holding an empty 500 ml bottle of alcohol hand rub, and lying next to another such bottle (preparation 4 in the table).

The patient was intubated and ventilated and transferred to the intensive care unit, where intravenous antibiotics were started for aspiration pneumonia. Computed tomography of the head was normal, and blood ethanol concentration at the time of collapse was 7g/l (152.2 mmol/l). This is nine times over the legal UK driving limit (0.8 g/l) and is a potentially fatal concentration.¹⁰ The patient was normoglycaemic and had normal renal function; liver function tests were in keeping with chronic alcohol excess, and she had a mild metabolic acidosis. On questioning after extubation, the patient denied any intent to self harm.

We searched the Guy's and St Thomas' Poisons Unit's database to compare the numbers of inquiries related to both children and adults exposed to alcohol hand rub (both ingestion and eye exposure) during the 16 month periods before and after the widespread introduction of alcohol hand rubs (December 2003 to March 2005 and April 2005 to July 2006 respectively). The search found an

Alcohol hand rub preparations: volumes available, ingredients and their concentrations (%)

Preparation	Volumes available (ml)	Ethanol concentration (%)	Other ingredients (percentage concentration)
1	75, 100, 500, 1000	<5	Isopropanol (<25)
2	150, 500	30-60	Isopropanol (10-30), benzylalcohol (<10), hydrogen peroxide solution (<10), perfume (<10), polyoxyethylene caprylic/capric glycerides (<10), denatonium benzoate*
3	60, 125, 350, 1000	62	Isopropanol (<5)
4	60, 150, 500	65-75	Glycerol (<10), acrylic acid homopolymer (<1), triethanolamine (<1), denatonium benzoate*
5	60, 125, 350, 1000	80	Isopropanol (<5)

*Bittering agent.