A PATIENT'S JOURNEY

Intensive care

Emilie Perrier

London

Correspondence to: E Perrier
azanome@hotmail.com

BMJ 2007;334:796-7

doi: 10.1136/bmj.39157.723090.BE

I am a 27 year old French woman and have lived in London since 2002. I came here to learn English, to meet people from other countries and to work. I had never been ill in my life, never been allergic to anything, and had no worries about my health.

In February 2003, on holiday in France, I had severe diarrhoea. After a few days, I was admitted to hospital with peritonitis. A few weeks later I was diagnosed with Crohn's disease. It changed my life completely. I spent three months in hospital before returning to England, where my husband was waiting for me.

As I learnt about my new life and new diet, I also started to become an old lady in my body. I developed a lot of joint pain from arthritis. At the end of October, back in France, my gastroenterologist prescribed sulfasalazine for my pain. The treatment went well for two weeks and I returned to London, but then I began to feel really sick. I had a high fever, pain everywhere in my body and I vomited everything I ate. I had shivers, could not sleep, and collapsed repeatedly.

On 20 November, we called an ambulance, but they said it was probably flu and that I should see my general practitioner, who prescribed paracetamol. Within two days, I was very ill; my skin had become very red and I was collapsing more frequently.

At the St Charles Hospital in London, the doctor gave me penicillin, believing I had meningitis. I was sent to St Mary's Hospital, where I saw several doctors, who thought my illness could be a bladder problem related to Crohn's disease. I had several CT scans and was told I was a "very interesting case." Nobody wondered

about the fact that I had a rash. I began to feel better; the rash disappeared and I wanted to go home.

I was discharged on 28 November, with a check-up appointment for the following Monday. I started my treatment again. On the Monday, my skin was very itchy and red. I told the doctor, who gave me anti-itch pills and I had some blood tests done.

That night the fever returned. I felt terribly tired and had no appetite. I was supposed to see my general practitioner on the Tuesday afternoon but was far too tired and sick. I called my husband and told him I needed to go to hospital.

When I arrived I began to feel really unwell and very hot; I collapsed in front of the lift. A doctor who saw me put me straight in a wheelchair and sent me to the accident and emergency department, where I was admitted. During the night I collapsed again and vomited a lot. I had a fever, and the on-call doctor became very worried.

The next morning the consultant arrived but really did not know what to do; the medical team still believed my illness had something to do with my Crohn's disease and I was scheduled for surgery the next morning.

My fingers started to swell, and the nurse had to cut one of my rings. Even at this point, we did not ask ourselves why I was swelling up. That night, the nurse needed to take some blood and tried to find a vein. Desperately she tried and tried again but with no success. I began to lose consciousness.

At this point I became really spaced out. I knew I had been moved to intensive care and that there were people talking to me, but I didn't care; my mind was somewhere else. The nurse told me I had to be brave as they were going to put some lines between my legs. I didn't care—I was so sick, they could do anything they wanted. I didn't feel the pain, I was in another world. I could hear and see people, but my mind was not there. On Thursday, I went for surgery.

After surgery, my skin blew up and burned. I was slipping into a coma, unaware of everything around me. I was put onto a life support machine and transferred to the intensive care unit in St Thomas' Hospital.

During my coma I dreamed a lot. Strangely, I was always in a hospital bed during my dreams. I remember my father speaking to me, saying my mother and my family were there. My husband was my guide, speaking to me by telepathy. It was never scary, but I know I prayed a lot, asking the Lord to help me. It's funny because I dreamed of doctors and nurses who were good or bad during my coma; when I woke up, there they were, for real.

I have no clear memory of my first awakening. It was

HOW THINGS ARE NOW: THE BAD AND THE GOOD

Three years on, I am still much affected by my experience. I am still very tired, my body is still very weak, my legs are extremely painful, and my skin is sensitive. I don't sunbathe or do sports, and I need help for most activities.

I am allergic to a lot of medications; the list is not definitive. Every time I try a new pill, it's a challenge. I have been allergic to every new treatment except those for Crohn's disease. When I have an allergic reaction, the rash comes six or seven hours after taking the medication, and I need to be treated with steroids at once.

It is not easy to explain my condition to people who are not familiar with my case; so many of them do not see the gravity of it. I have had some problems in emergency departments, where doctors didn't think it necessary to call a dermatologist.

But this experience brings me lots of positive stuff as well. Fortunately I have lots of great doctors around me. My family and friends have been extremely supportive. They are all aware of my disease and my problems.

I have become closer to my sister and my mother. It has also given me the chance to appreciate the importance of life. I am more sensitive to nature, to the environment, to this amazing planet, and to other people, and I cry easily.

When I look back it is difficult to believe that all this really happened; at the same time, it is as if it happened only six months ago. I shall always be grateful to all the medical staff, and to my husband and family, for everything they have done for me.

A DOCTOR'S PERSPECTIVE

Emilie Perrier's story was striking. She was a healthy 24 year old who had been well until April that year, when she was diagnosed with Crohn's disease. This was well controlled with mesalazine until October, when she had started to experience joint pains, and she was switched to sulfasalazine. Four weeks later she developed a diffuse maculopapular rash and abdominal pain. She was admitted to hospital, and a computed tomogram showed bilateral pleural effusions and free fluid in the abdomen and pelvis. She was treated on a presumptive diagnosis of intra-abdominal infection with cefuroxime and metronidazole. Notably, during admission the sulfasalazine was stopped and she improved. On returning home, she restarted all drugs, and within 24 hours she collapsed. On readmission she was noted to have diffusely "flushed" skin and a low blood pressure of 89/49 mm Hg. She had right upper quadrant tenderness. Her sulfasalazine was again stopped. In view of her abdominal pain she had a laparotomy. This showed no abnormality. She rapidly deteriorated, developing marked erythroderma and facial oedema. She developed bullous lesions over pressure points. There was no oral or genital involvement. Nikolsky's sign was negative (Nikolsky's sign is positive if there is intradermal blistering, which can develop when apparently normal skin is rubbed (this can be seen in toxic epidermal necrolysis)).

On admission to intensive care she had a metabolic acidosis, with a bicarbonate concentration of 14. Her white cell count was raised, with a neutrophilia $9.3 \times 10^9/l$. Notably she had a raised eosinophil count at $1.93 \times 10^9/l$. Her liver function tests were deranged (alanine aminotransferase 357 U/l and alkaline phosphatase 178 U/l). Creatinine concentration was $198 \mu\text{mol/l}$ and albumin concentration was 19 g/l. Her chest radiograph showed bilateral pleural effusions with interstitial shadowing. Acute respiratory distress syndrome was diagnosed, and the patient was intubated.

The differential diagnosis included toxic epidermal necrolysis, but

the negative Nikolsky's sign and lack of oral and genital involvement suggested otherwise. The neutrophilia and relapsing remitting nature of the presentation also argued against this diagnosis. With the combination of rash, peripheral eosinophilia, and hepatic and pulmonary symptoms, we diagnosed DRESS (drug reaction eosinophilia and systemic symptoms) syndrome in response to sulfasalazine.

It is important to make the differential diagnosis between DRESS and toxic epidermal necrolysis, as steroids are successful in the former but can be deleterious in the latter, where they have been associated with infection and gastrointestinal bleeding.

We started pulsed methylprednisolone 500 mg daily for two days, followed by ciclosporin 50 mg twice daily. Fortunately for this patient, she made a full recovery, although she now has an increased sensitivity to various drugs, and needs longer term ciclosporin.

What can we learn from this case?

- At first presentation, start from the beginning; don't rely on previous notes. In retrospect, the relation between Emilie's deterioration and starting and restarting sulfasalazine is compelling
- All clinical signs are important even if you can't at first explain them. She had a rash from first presentation
- The differentiation between toxic epidermal necrolysis and DRESS syndrome was vital in this case. Clear history taking and examination of clinical and laboratory findings went against our initial diagnosis of toxic epidermal necrolysis (which we had made on the basis of her being very sick with a severe skin reaction). This allowed vital steroids to be given. In my view, this turned the case around.

Justine Hextall, consultant dermatologist, Worthing and Southlands Hospital, and honorary consultant, St John's Institute of Dermatology, St Thomas' Hospital, London
justine@doctors.org.uk

all very confusing, and I think the drugs were mixing reality and dreams. I had a chest infection and remember the physiotherapist coming and making me cough really hard. It's one of the most difficult things I have done, it was so exhausting.

After 24 hours, they put me back to sleep. I was still not aware of what had happened or of the gravity of the situation. I don't remember my second awakening clearly either. I was being ventilated. The staff were extremely kind and reassuring. I was aware that something had happened, that I was big and couldn't speak, but it didn't bother me. My lips were burning and I couldn't move a finger. My only requests were for a pencil and paper and for a clock to track time.

My mother and my husband came every day and talked to me. I knew it was close to Christmas; the radio was on, and all the Christmas songs are now buried in my head.

On Saturday 20 December, the doctors planned to take me off the breathing machine. I was petrified. I kept asking the nurse, by writing, if it would be safe, and if she could check with the doctors. That afternoon they took the big step. The ventilation was removed and I was breathing on my own. My voice was very thin.

The following Monday, I was moved back to St Mary's Hospital. I was very excited but still did not realise what had happened. All I knew was that I was big and had had surgery. The ambulance came in the evening and the crew were so nice. We passed Big Ben and the Houses of Parliament, and they gave me a little sightseeing tour.

Back in St Mary's, I met my dermatologist. She was so kind and told me that everything would be OK. I started to become really thin and lose all my fluid. I couldn't sleep. Every time I went to sleep I woke up again, brutally. I had terrible diarrhoea and was unable to control myself. I was so weak I couldn't move. I was totally dependent on the medical staff and my family. I was terrified I was going to have a breakdown.

My husband brought me a small television, which was amazing. I began to rediscover the world.

On 24 December, my dermatologist explained to me what had happened (see the doctor's perspective box for her explanation). I felt really lucky and grateful. I asked the doctors when I would be able to go home and they said, when I could walk on my own and if I had some help at home. I was determined to go back to my place as soon as possible, to begin my own battle, the one with my body. The physiotherapist showed me some easy exercises to start my muscles functioning again. It was hard work; I felt as if all my physical and mental energy had been taken away.

Eventually, on 16 January, I was allowed home under supervision. I was so happy and proud of myself. I weighed 40 kg, was very weak, but was standing. This was my victory.

Contributors: EP wrote the main article, and JH supplied the box.

Competing interests: None declared.

Provenance and peer review: Not commissioned, not peer reviewed.

Accepted: 6 February 2007