Personal Paper

This disease of mine

MARCI ANN EPSTEIN

Idiopathic autoimmune haemolytic anaemia. And at age 8 not an easy phrase to recite over and over to relatives, intrigued and anxious residents, as well as curious friends. It is a condition that, partly, is still not clear—not clear to doctors, my parents, and, of course, me. The questions “why” and “how” have not ceased in the minds of many and the answers seem quite unsatisfying. An experience such as mine is not soon to be forgotten for it has affected me for life. Exactly how it has affected me I do not know. Surely my outlook on life and my feelings toward my family and friends are unconsciously marked by this experience.

November 1970, the beginning—the beginning of a plight lasting just about two years. I remember having a virus infection, as did my brother, Andy, and sister, Ileen. We all recovered well and continued our plans to visit cousins in New York. It was there that my parents noted that I had become jaundiced, though feeling well. On returning home, my parents and I made a visit to my paediatrician. The date was 23 November 1970. Pallor, enlargement of the spleen, anaemia, and a positive Coombs test were all noted. At that point, my haematocrit reading was 20.

My memory is blurry. Next, I recollect meeting my “soon-to-be haematologist.” It was late on a Friday night. My parents called him and he agreed to meet us at his office. Quiet, dark, almost morbid hallways come to mind. The place was deserted except for all-night security guards and several janitors. Precisely what took place is not clear, besides, of course, a thorough physical examination. I do not think that I felt frightened by this, only lost and empty. But, strongly gripping my parents’ hands, I was safe: safe from this strange building and kind, yet mysterious man.

Three days later I returned to his office. At that time my haematocrit reading was 22. This Monday afternoon was probably the worst one of my life. Though an 8-year-old could not fully comprehend the mere thought of excruciating pain, I quickly learnt after a bone marrow aspiration.

Surely the pain felt is something I wish on no one. The cold, shiny, even clammy examining table put shivers up my spine. There was not much for me to see since I was told to lie on my stomach. The reason was obvious—a needle was to be injected into my lower back in order to withdraw marrow that was to be evaluated. I remember a nurse having the responsibility of preparing me, almost like a specimen, for my doctor. Emotion welled up inside me. In walked my mother—a small feeling of relief. She held my hand and I was told to squeeze it when the pain was unbearable. Then the high piercing scream—one that must have scared everyone in the waiting room. Even the secretary outside was taken off guard by the sound. It was done. No pain was felt when the test was completed. The results

were in: the marrow seemed normal except for erythroid hyperplasia. The diagnosis—idiopathic autoimmune haemolytic anaemia. “OK. So what does this mean?” I remember questioning. The answers made no sense to me, though the long big words were impressive. With time these terms and others became important, and, yes, gained a certain special respect as well.

Less than a week after my first visit, my doctor started me on drug treatment. Horrible thoughts come to mind when I think of those bitter tasting, grainy, and white pills. They tasted so bad when I tried swallowing them that I developed my own “pill-taking technique.” Buried beneath a heaped tablespoon of homemade apple sauce so that they could not be seen, this ritual occurred three times daily. After the apple sauce I had to swallow a teaspoonful of chalky, milky tasting “soup.” My parents told me that it was good for me, but I really wondered.

My initial treatment was in the form of prednisone, 10 mg three times daily. My doctor thought that I stood a 50% to 60% chance of having a good response to it. Soon the dosage was increased to 30 mg and I was feeling better, except for the side effects.

Dealing with side effects

As I look back on this entire experience, I can vividly recall the emotionally stressful times of dealing with the side effects of prednisone—an increased appetite and retention of fluid. This meant only one thing: I had gained a lot of weight, retained a lot of fluid, and looked like a balloon. At this time during my treatment I returned to school, to what once was a happy third and fourth grade classroom. But now things were different. Snickering and teasing could be heard from all around and even from those who were my friends. Surely none of them knew why I looked the way I did and no one bothered to ask. Daily I was forced to face these cruel immature “friends.” Coming home only meant the retelling of the horrible encounters of the schoolday. With the advice of my family, I learnt quickly to cope and to walk away without being hurt. And, though this school trauma was one that has left a mark, it definitely has given me the strength, even now, to deal successfully with others and to understand how, unfortunately, even innocent children can be cruel.

My initial response to the prednisone was good, as shown by an increased haematocrit reading of 34. The plan was to continue the treatment for one more week at this level and if the haematocrit value continued to rise we could then gradually taper the prednisone. But because of a fever for three days on 4 February 1971, and a discouraging haematocrit value of 27, the possibility of returning to the original high dosage of prednisone seemed probable. At this time a spontaneous remission could be hoped for if the haematocrit value hovered around 30. Now splenectomy was first mentioned.

Confused, discouraged, frustrated—this is all I remember during the corticosteroid treatment. I have forgotten how many

times we played games with those small powerful pills, often begging them to respond. Week after week, Friday after Friday, we went for blood tests, with the hope and thought that maybe this is "the day." We lived by those haematocrit numbers determined by the blood technician. The mechanism of corticosteroid response was not clear. How did those little pills make such changes in my body?

After seven months of corticosteroid treatment and additional confirmations by a paediatric haematologist, the outcome was still nothing more than a decreasing haematocrit. So it was on Tuesday, 13 July 1971 that I had a splenectomy. Not only was I to have surgery, but I did not even know what they were taking out of me.

Anaesthesia, incision, operating room. The night before my operation, the surgeon visited me to explain the general procedure for the following day. I remember being inquisitive, asking questions of most importance to me. "What kind of design will I have on my stomach? And how big?" The surgeon replied with confidence that the scar would not be larger than the length of his index finger. What a relief. Soon after, this friendly warm man left my stark, white hospital room almost as mysteriously as he had come.

How odd and slightly silly it all seemed when "the boys in green" arrived, only to bring me, my stretcher, and the rest of my paraphernalia down to the operating room. I have vivid memories of waiting for the elevator with my Dad. Though I know that he tried to conceal his feelings as he leaned over, to me they were all over his face. It was not difficult to realise that he wanted to go in my place. But I do remember quite clearly that I was not afraid; it was something that only I could face. And it was to be confronted alone. With a smile and small wink I looked up to my Dad and said, "Dad, you don't worry about me. I'll be fine." The elevator arrived, I was wheeled in, and the doors shut behind us.

I had not even remembered waiting on the stretcher outside the operating rooms with other patients. It is funny how it seemed like they were giving prizes away as patients waited for, what seemed to be, ever. I was incapable of explaining then, and most certainly now, the feeling after the operation. Waking up to my parents, I asked when I was going down for the operation. A tube up my nose, needle in my arm, and looking like a mummy bandaged in white, I lay there motionless. It was over. Before long though, I was told of my scar.

It was not the length of an index finger—probably twice that size. What had happened? Was there a mistake or complication with the operation? No, nothing of that sort. Because of the idiopathic autoimmune haemolytic anaemia it is difficult to give blood transfusions. My antibodies not only attack my red cells, but it is dangerous to give me anyone else's for the same reason. As a result, an incision had to be made that would minimise blood loss. Because of an enlarged spleen located underneath my ribs a "sigmoid-shaped curvilinear left subcostal incision" was made instead.

All I can remember is that awfully annoying tube. One day when the surgeon came for a postoperative visit he could see

that it was bothering me. I remember him saying, "We'll have to take care of that." With one quick pull the tube was out. The air, fresh and cool, never felt so terrific. I thought that my surgeon was "the greatest," until he forced me to start walking around. I walked, with much pain, leaning over my left side. I was convinced that the stitches would come out and I would break open. The nurses, observing my walking, would smile and tell me to walk straight. I smiled back and tried.

Five days after the operation I could be found in McDonald's eating a Big Mac with my family. A Big Mac never tasted so good. The steroid treatment continued, but now the dosage was being tapered. Everything seemed to be going well until October 1972.

Another set-back

It came suddenly like a lightning bolt. Recurrent fevers without any obvious cause plagued me. Assuming that this condition was nothing more than the average run-of-the-mill fever, I began taking penicillin. This treatment was to no avail. I was again seen by my haematologist with a haematocrit value back down to 25. Again a bone marrow examination. Besides the sharp excruciating pain, the examination detected erythroid hyperplasia, an increased red cell production. Certainly not what we had hoped for.

Lethargic, drowsy, frightened. I remember that night in the hospital very well. The thought of lifting a limb seemed too tiresome. So I remained frozen in bed as my Dad leaned over the bed rail. My eyes were so heavy and my eyelids, too, could not be lifted. A tough decision needed to be made: was it advantageous to inject a unit of packed red blood cells?

Within minutes, energy flowed through my body. It was magical. Drip by drip the deep red liquid of life trickled into my veins. Penetrating warmth stirred in me. My eyes, even, expressed a sense of life again. It seemed that I had an incomplete remission with either an "aregenerative crisis" because of an infection or possible low folate deficiency because of continuing haemolytic anaemia. Sent home to be continued on folic acid supplementation daily for the rest of my life, I was cured.

But was I really cured? Truly cured from this experience? No, definitely not, though no physical complications have arisen since then. Every day I am confronted with the difficulties of this ordeal and, as a result, challenged by it for life.

The need to study medicine surely stems from this experience. I find myself constantly driven by an unknown force to climb and always reach without fear. I now realise that life itself is so uncertain and the prolongation of a worthwhile, comfortable, and happy life is the ultimate challenge. I want professionally to face this venture with the hope of seeking a satisfying existence for others. I live with a non-stop feeling of indebtedness to those professionals who have in some way helped me through and beat this trauma. For life I remain with an IOU to medicine, and, overall, to humanity.

For almost all prepared foods and soft drinks the list of contents finishes up with the words "colour, flavour, preservative." How "safe" are all these foreign substances, which are undoubtedly useful to the food trade?

All substances added to foods¹ have to satisfy two basic criteria. Firstly, are they necessary? This entails assessing whether the additive has an effective role with regard to the sensory quality, physical properties, and stability or safety of the food in which it will be used.² Secondly, are they safe in that they are not associated with any toxicological (including allergenicity) effects. Additives come under the close scrutiny of the food additives and contaminants (FAC) committee of the Ministry of Agriculture, Fisheries, and Food. This committee is composed of independent members and has to be satisfied on both counts. In addition to evidence obtained in the United Kingdom, the committee also takes into account evidence

relating to the additives from other countries, especially the United States, Canada, and the EEC. The committee includes scientists with a wide knowledge of the relevant scientific publications. The FAC Committee makes recommendations to the various government departments on what regulations relating to additives should be implemented. The consumer is, therefore, protected by a carefully designed and conscientiously operated system. Like all human institutions, however, it is fallible, and it is important not to minimise the real difficulties inherent in the toxicological testing of food components and the problems of interpreting the results obtained with animals and the prediction from them of effects in man.—DAVID SOUTHGATE, head, division of nutrition and food quality, Norwich.

¹ British Nutrition Foundation. *Why additives?* London: Forbes Publications, 1977.
² Taylor RJ. *Food additives*. New York: John Wiley and Sons, 1980.