THE SCOTSMAN

Frightening ITP is manageable with right help

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A FLURRY of headlines appeared early last year when the young Scots tennis star Jamie Baker, a close friend of Andy and Jamie Murray, was "struck down by a deadly blood disorder" at a tournament in America and rushed to hospital.
The alarm bells started ringing the instant we heard the news because our son, Gregor, had been "struck down" by the same blood problem a few months earlier and nobody had mentioned the word "deadly" once in all the numerous times we visited Stirling Royal Infirmary's haematology department.

The disorder is called immune thrombocytopenic purpura (ITP) and it's a disease where the immune system goes haywire and attacks the platelets in your blood. Platelets are small particles that help blood to clot and are counted in thousands.

ITP is all about numbers – a healthy adult will have a platelet count of somewhere between 150 (thousand) and 400 (thousand) per millilitre of blood, while an ITP sufferer can have a count lower than five. Fortunately, it's very rare, affecting only a few people in every million.

The symptoms vary considerably, from internal bleeding, including (very rarely), bleeding of the brain, constant exhaustion, excessive bruising, increased exposure to disease and difficulty in stopping bleeding after an injury. No-one knows what causes it.

Gregor's main symptom was heavy nose bleeds – frighteningly heavy – but ITP is so uncommon that none of our GPs made the link until they exhausted all the more likely causes and tried out all the more common cures, such as cauterising. Looking back, the most obvious sign was that he was often so tired he couldn't get out of bed in the morning to go to school, which affected his exam performance. But show me a teenager who can't get out of bed…

The year 2009 has been declared the year of ITP awareness, and the ITP Support Association holds its annual conference in Edinburgh this week when keynote speakers will include some of the world's foremost experts.The association is an enormous help to sufferers and carers, who often get conflicting advice from doctors and consultants who have a very basic knowledge of ITP, which, at the moment, has no known cure.

The experts will tell you it isn't "deadly" and can be controlled, or even, as Gregor has proved over the last nine months, lived with. When he was initially diagnosed we were shocked, but not frightened, or not as frightened as we probably should have been. We knew nothing of ITP and were given no real indication of how serious it could be. The only things we had to go by were the numbers – his platelets were worryingly low.

The first thing a doctor faced with a patient suffering from ITP will do is reach for the steroids. Steroids suppress the immune system so it no longer kills the platelets, ergo the platelet count rises. Great! But what do you do when you discover that the cure is worse than the disease? When your son's weight balloons from ten stones to more than 17 in a few weeks? When his face and his body become distended by the effect of the drugs, he suffers crippling depression and his concentration suffers so he falls further and further behind his Higher class year?

What can you do? You trust his doctors. Gregor was on steroids for more than six months – that's as long as you can take them because the long-term side effects are more horrendous than the short – and the way he carried that burden, smiling through when all he wanted to do was collapse in a heap, made me as proud as a father can be.

There had been a slim chance that the disease could simply have gone away – it does that sometimes in younger sufferers – but it hadn't. So it was back to the hospital for the fortnightly blood test and platelet count, which was about 30.

Before he was taken off the steroids, we had our next big shock. The consultant who treated Gregor casually announced the next step was to remove his spleen, which controls the immune system. We were horrified: the steroids apart, Gregor was no worse off than when he'd first come to the hospital. The consultant explained it would save a lifetime of blood tests, which, in the light of what we later learned, still doesn't seem a good enough reason for removing an organ.

We talked it over at home, and Gregor, who had received a bigger blow than any of us, announced he wanted to have the operation. He was at his lowest then, and would do anything to get rid of the ITP. That was when the support association and its magazine, The Platelet, came into their own. We read that other people who'd had their spleens removed hadn't been cured of the disease and on top of ITP, they'd been condemned to a lifetime of taking antibiotics. Further research showed the percentage of patients who were helped by a splenectomy was worryingly low. There were no guarantees this radical step would succeed.

That was when we did what would have previously been unthinkable – and asked for a second opinion. Gregor's SRI consultant accepted our concerns and our reasoning and arranged an appointment at the Royal London Hospital. We discovered "The London" is the centre of excellence for ITP research and treatment in the UK and we couldn't have been in better hands. His new consultant was a Scot, Dr Drew Provan, one of Britain's acknowledged experts on ITP. He quickly arranged for Gregor to be part of a UK-wide DNA database which it is hoped will lead to greater awareness of what causes ITP and possibly provide a step towards a cure.

The first thing Dr Provan said was that he should keep his spleen; the chance of success just wasn't high enough. Next, he decided to take Gregor off the steroids, which was almost as big a relief. Afterwards, he talked us through all possible treatments, including a couple of new drugs – such as Rituximab – that other doctors had seemed suspicious of, but which he assured us could be effective in the right doses. We discussed blood counts, and for the first time learned the numbers weren't as significant as we thought. "Some people can live ordinary lives with a count of about 20, even as low as single figures," he explained.

His conclusion was Gregor shouldn't take any more drugs and we'd "see how he gets on". Nine months later and he's getting on fine. His platelets have been as high as 60, but usually hover at about 30.

We did have one scare. A worrying bout of internal bleeding, a cancelled holiday and a boy on blood and plasma transfusions for a week, but since then things have settled down. He accepts the limits imposed by ITP, but like any teenager he'll push them as far as he can. He can't play rugby or other contact sports, but he likes to play football with his mates.

He's still exhausted a lot of the time, but the nose bleeds are less frequent. He'll always carry the physical scars caused by the steroids, but, generally, he's a normal, cheerful young man who says the key is that he now understands what he has and knows he can live with it. For us, that's enough.

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Page 1 of 1

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