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Living with Von Willebrand's Disease - A Patient's Perspective

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Introduction

Late last year I got a phone call in the middle of the night from a carer in the sheltered housing where my 85 year-old mother is now living to say that she had had a fall and an ambulance had been called to take her to the Bristol Royal Infirmary.

As I waited in A & E for the ambulance to arrive I had occasion to reflect on the number of occasions that I had visited A & E with her as a child and the progress that had been made in the treatment of the disease that she and I (and a good number of other family members) suffer from - Von Willebrand's Disease.

Family History

I was the first in our family to be diagnosed in the late 1950's when, as a 3 year old, I bled for several days from a cut to the lip and the casualty department of our local hospital in Slough referred me to the Hammersmith Hospital for tests. What seemed like numerous trips up to London were followed by further tests that confirmed that I had Von Willebrand's, but my mother and sister, who were also tested, were told (wrongly) that they did not have the condition.

As a child I seldom managed to attend a scout camp, or any other activity, without some kind of mishap and played a single game of Rugby at secondary school before ending up with a knee injury that took several weeks to get over and a decision by the school that it was not an experiment to be repeated! The condition was not one that most medics had heard of in the 1960's and 70's, but few were prepared to admit that they did not know what to do. I remember when I was about 10 going to casualty bleeding profusely having bitten my tongue. A nurse cross-examined me and told me I was very stupid to have bitten my tongue and then a doctor arrived, placed a wedge of lint in my mouth and told me to leave it there for the rest of the day.

I also remember when I was around the same age my mother going into hospital for a hysterectomy. She was in hospital for the best part of 10 weeks, largely because they could not control the bleeding. One Sunday lunch time the hospital, who had been unable to reach my father by phone, sent a policeman to our house to say that she was in real danger. Soldiers from a local barracks were drafted in to give blood when one of the doctors concluded that mum was deficient in some factor associated with clotting and thought that fresh blood might be more effective than stored blood. Fortunately the treatment worked and she survived.

Even when I was in my twenties and had my wisdom teeth out I bled for two weeks - the various concoctions that were available at the time were simply unable to deal with persistent bleeding. In the early 90's I was living in Sussex and had a haematoma in my leg that led to my being hospitalised in Brighton for 10 days. At that time a new treatment was being trialled in one of the London hospitals and my consultant arranged for a consignment to be sent to Brighton by Red Star. It arrived at about 10.00pm and a house officer came to give it to me, only to discover that all the instructions were in French! Fortunately there was a Filipino nurse who was able to translate - and the treatment proved effective.

When each of our three children were born, someone was on hand to take a blood sample to test for vWD. Our eldest son and our daughter both have the condition, but our second son does not. One of the challenges with children has been to make sure that school and youth groups have known what to do in an emergency without causing panic. We have had our ups-and-downs with this, but they have generally been able to live fairly normal lives (albeit with more hospital trips, and occasionally stays, than most of their peers!)

Problems still remain:

• When my mother was hospitalised in Slough a few years ago she had to fight off doctors who were determined to administer warfrin in case she had a clot. This is a problem I also experienced in Switzerland a few years ago when it was (mistakenly as it turned out) assumed that I had broken my ankle and my foot was put in plaster. I eventually had to phone my sister (who teaches medicine in Manchester) to persuade them that this was not a wise course of action to take.

• Doctors in A & E Departments who have not responded quickly enough to an incident involving bleeding, or have not taken sufficient notice of the complications to a fracture of Von Willebrand's. It seems we have often ended up in A & E either at the weekend or the evening when records are not readily been available, but we did eventually get clear protocols agreed with both paediatric and adult A & E.

• We have occasionally been subject to the inappropriate use of students and other less experienced medics. I appreciate the need for training and the fact that everyone needs to gain experience, but someone with Von Willebrand's Disease is not the best person for a trainee dentist to gain experience of extractions on. More recently I have been treated by consultants, with much better outcomes. We have had the same experience with taking blood or putting in lines – some professionals are a lot better at this than others and failed attempts can lead to serious bruising.

The drug situation today is very different from the regime that existed when I was younger. The treatment that I am now regularly given is called Alphanate and it is easily administered, raises levels quickly and so far has been successful in dealing with a wide range of bleeding problems that I have experienced. The organisation of care for Haemophiliacs has also improved - certainly in Bristol, where we now live. The Haemophilia Centre has an excellent team of doctors and specialist nurses who know our family well and also has a specialist physiotherapist who has provided excellent care for us. We are very grateful for the confidence that this provides us with.

Editor's Note

Von Willebrand's Disease is the most common of the hereditary bleeding disorders although it can also be acquired.

It arises from a deficiency of Von Willebrand factor (vWF) a protein that prolongs activity of factor VIII by binding to it and is required for platelet adhesion. Von Willebrand's Disease affects humans and dogs (particularly Doberman Pinschers) and rarely cattle, horses, swine and cats.

There are three types of hereditary Von Willebrand's Disease (Types vWD I, II and III) and various sub-types. The gene is located on chomosome twelve. Types I and II are inherited as autosomal dominant traits whilst Type III is the most severe form being homozygous for the defective autosomal recessive gene.

There are a number of causes of acquired Von Willebrand's Disease including production of autoantibodies, aortic valve stenosis and thrombocytopenia.

Alphanate (antihaemophilic factor) is concentrated factor VIII and also contains albumin as a stabilizer. It is prepared from pooled human plasma.

As described above modern medical techniques have transformed the prognosis in this condition. For comparison with the historical outcome please see the accompanying archive articles on Haemophilia in this issue of WEMJ.

References

1)http://www.ncbi.nlm.nih.gov/pubmedhealth/PMH0001571/
2) http://en.wikipedia.org/wiki/Von_Willebrand_disease