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KD50: COMPREHENSIVE CARE

Gynaecological and obstetric care for women with inherited bleeding disorders

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The issue of women with bleeding disorders was first reported by Professor von Willebrand in 1926, but it is only from around the mid 1990s that the issue has been fully recognised. Much of this is due to the vision and hard work of Professor Christine Lee and colleagues at the Katharine Dormandy Haemophilia Centre. This work has led to better diagnosis, better quality care and improved quality of life for women with bleeding disorders.

Keywords: women, haemophilia, Katharine Dormandy

My interest in the gynaecological and obstetric management of women with inherited bleeding disorders began with a simple audit in 1995. Utilising the excellent records kept in the Katharine Dormandy Haemophilia Centre, we reviewed the outcome of all the pregnancies over a 10-year period. This led to a series of recommendations that continue to be the foundation for managing carriers of haemophilia today [1]. This includes:

- Pre-pregnancy counselling
- Multidisciplinary team approach
- Prenatal diagnosis of female gender during anomaly scan at 18-20 weeks
- Clotting factor level – at booking, 28 and 34 weeks
- Clotting factor level in labour – if < 50 IU/dL treatment
- Unknown or male sex – avoid invasive foetal monitoring and instrumental delivery
- Epidural analgesia is not contraindicated provided that the level of clotting factor is normalised
- Clotting factor level of cord blood
- Vitamin K by mouth if haemophilic baby or status unknown
- Registering the haemophilia baby at haemophilia centre.

The next milestone came when we looked at all the women who came into the gynaecology clinic with heavy bleeding during menstruation but no obvious gynaecological cause. We found that 13% had von Willebrand's disease [2]. Groups in Europe and North America reproduced our work, and subsequently we conducted a systematic review that confirmed that overall 13% of women who have heavy menstrual bleeding may have an undiagnosed von Willebrand's disease [3].

The next step was setting up the first multidisciplinary clinic for women with inherited bleeding disorders. Since then, our service has grown and now includes representatives from gynaecology, midwifery, genetics, physiotherapy and family planning. We provide a truly comprehensive service and regular patient surveys show high levels of satisfaction.

In association with The Haemophilia Society, the Katharine Dormandy team developed the "Women Bleed Too" campaign, the forerunner of "Talking Red"

What is unique about the service is that it is comprehensive and also looks after the patient's family. We are there to support our patients, not only for medical care but also for social support and psychological support. We work closely with the team at Great Ormond Street Hospital to provide a smooth transition of care for the adolescent girls as they move into our adult service.

The clinic has also been a tremendous resource to learn about women's bleeding disorders. We have looked at blood loss during periods in our women and clearly seen that these women have very heavy and longer duration of menstruation that really impairs their quality of life. We designed an algorithm for the treatment of abnormal menstrual bleeding that has been widely adopted nationally and internationally.

In pregnancy, we have looked at the antenatal changes in factor level as the pregnancy progresses and these studies have been critical for managing patients with these disorders.

We are very keen to promote the provision of regional block for pain relief in labour because we saw that often our women were denied the option of effective pain relief in labour. We published our studies showing that this option can be safely offered, provided the coagulation deficiency is corrected, a multidisciplinary approach is adopted and each case is individually assessed [4].

Awareness of bleeding disorders in women

The other area that we were also very interested in is increasing awareness of bleeding disorders. So, we did a survey with the Royal College of Obstetricians to see how aware gynaecologists were of underlying bleeding disorders in menorrhagia. Our results showed that very

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Rezan Kadir is a consultant obstetrician and gynaecologist with a special interest in women with bleeding disorders and foetal medicine at the Royal Free Hospital. She is also an honorary reader with the University College School of Medicine. She has set up a multi-disciplinary clinic in the haemophilia centre at the Royal Free Hospital, which is jointly run with a haemophilia specialist, haemophilia nurse, family counsellor and therapists. This clinic has been unique in providing comprehensive care for women in families with bleeding disorders. The clinic is renowned nationally and internationally, and provides training for visiting clinicians from across the world to help develop the service in their hospitals. Her work in the field of women and bleeding disorders has been well recognised internationally and she is regarded as a world expert in the field. In this article Dr Kadir discusses the milestones in the recognition and improving management of women with inherited bleeding disorders.

few gynaecologists test or screen for von Willebrand's disease and underlying bleeding disorders in their patients with menorrhagia [5]. Similar results were published in the United States [6]. As a result of this, we worked with the Haemophilia Society and set up the "Women Bleed Too" project, with the aim of raising awareness and providing support to women with bleeding disorders.

We then wanted to understand the priorities and needs of our patients. We conducted a simple survey, which highlighted a number of problems, including poor recognition of bleeding disorders in women and the lack of information for women. Many suffered at school and at work because their teachers and employers did not know about or understand their problems. We also found that their GPs knew little about bleeding disorders and were not able to provide support.

In 2006 we published the national UKHCDO guideline [7] followed by our review of our multidisciplinary clinic, [8] which was significant in raising awareness and helping to increase the number of multidisciplinary clinics across the country.

In 2008 we re-audited our obstetric care and found a significant improvement not only in intrapartum care but in the post-partum bleeding rate in the patients with bleeding disorders, which was not higher than the general obstetric population [9].

Our clinic and this service for women with bleeding disorders has attracted some very talented people from the UK and internationally to come and work with us.

What does the future hold?

We have achieved a great deal of progress in the management of women with bleeding disorders at the Royal Free. One area in which we are keen to make further progress is prenatal diagnosis. In 2006 we introduced a test in early pregnancy to determine foetal sex [10]. This was significant as it changed the pathway of prenatal diagnosis and helped avoid invasive testings and their complications.

Following this we wanted to find a prenatal diagnosis for male babies with haemophilia. We collaborated with the team at the Chinese University of Hong Kong and developed a PCR analysis for foetal DNA in maternal plasma to diagnose whether the baby is affected or not. Our published pilot study included 12 samples from seven carriers with a male baby, taken at the different stages of pregnancy. We correctly diagnosed foetal genotype in all samples, including early gestation samples [11].
But as we know, haemophilia is heterogeneous in its mutations, so every time we have a new patient we have to produce a new PCR. So we are now focusing our efforts on finding a prenatal diagnosis for the intron 22 inversion and are aiming to have a universal test that will provide prenatal diagnosis for 50% of severe haemophilia.

One area that I feel passionate about is what is the safest mode of delivery for babies with haemophilia. We have conducted a systematic review of the literature and meta-analysis (to be published shortly), which found that the risk of intracranial haemorrhage in babies affected with haemophilia is 44 times higher than the general population, and the risk of extracranial haemorrhage is eight times higher. Delivery by instruments such as forceps and ventouse increases the risk by four times, while planned caesarean section reduces the risk by one third.

The issue of women and bleeding disorders has become a priority in the agendas of national and international societies such as the International Society for Thrombosis and Haemostasis and the World Federation of Hemophilia. We can confidently say that the work carried out by the Katherine Dormandy Centre has led to better diagnosis, better quality care and improved quality of life for women with bleeding disorders.

Disclosures

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