Improving comprehensive care in the haemophilia community: building on the HERO Study

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Although many people with haemophilia (PWH) in developed countries lead full and productive lives, haemophilia has a significant impact on the quality of life of parents, siblings and PWH themselves [1]. In this short commentary, we briefly examine how the Haemophilia Experiences, Results and Opportunities (HERO) Study established the evidence base, and then generated an action plan at the subsequent HERO Summit. The main focus of this article is on what is being done within the haemophilia community to build on these initiatives, with the aim of improving all of the interrelated dimensions of disease management for those living with a bleeding disorder.

The HERO Study, sponsored by Novo Nordisk, was the first multinational, large-scale study that provided data to quantify anecdotal observations and subjective opinions surrounding the psychosocial issues of PWH and their families [2]. Analysis of questionnaire data, completed by 675 PWH and 561 parents of children with haemophilia (CWH) [2–4], highlighted several positive life experiences about living with haemophilia, but also identified six areas for improvement [5]: physical burden, psychological burden, relationships, knowledge gaps in society, employment and access to care.

The 2013 HERO Summit provided a forum for PWH, parents and healthcare professionals (HCPs) to discuss the HERO Study findings and establish steps to improve psychosocial care [5]. Participants concluded that training and educational strategies are required to narrow knowledge gaps in society and strengthen the communication skills of and among PWH, their families and HCPs. Initiatives should therefore reflect the needs of distinct haemophilia care groups (parents, HCPs or PWH), and be targeted at a national or local level, in order to achieve positive, effective change and improve outcomes.

In recent years, public funding for haemophilia support programmes – programmes that provide comprehensive care for people with haemophilia and related bleeding disorders and their families – has been reduced across many countries, placing greater responsibility on the community to drive local initiatives. Since the HERO Summit, participating countries have successfully initiated and enhanced national plans in order to improve outcomes at a national level. Such schemes include support programmes, many of which have been run in conjunction with patient advocacy.

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groups (PAGs) and the wider haemophilia community. Examples of programmes, which serve as models for meeting needs and which were created prior to HERO to help empower the haemophilia community, include: HOPE, We Start Young, Inhibitor Family Camps and ‘A Bright Future’ workshops.

HOPE and We Start Young were created in Italy with the support of Novo Nordisk and Pfizer, respectively, and are contrasting but concurrent means of providing support for CWH and their families. The objective of HOPE [6] is to support families during their CWH’s growing stages by bringing parents together in coordinated, facilitated support groups. HOPE empowers parents and families, and provides a network for families to share experiences, anxieties and ways of coping with their CWH’s condition, thus enhancing their own wellbeing and helping to manage any feelings of negativity as caregivers. Through HOPE, parents are therefore able to better manage their lives through knowledge sharing and mutual support. There are currently 40 families involved in HOPE groups in six Italian cities. For more information, visit www.fondazioneparacelso.it/en/programs-and-activities-en/hope.html.

We Start Young [7] is an individually tailored programme that begins from the time of diagnosis – typically a particularly stressful time – through a child’s early years. Its objective is to support the families of CWH through the help of a family mediator. After an initial meeting between the physician, CWH and their family at their haemophilia treatment centre, the mediator visits the family home and advises on practical aspects of care and the life of a CWH. Working with families at the start of their journey offers support by providing the CWH with a balanced view of their condition. Early engagement is vital, therefore, and initial knowledge gained can be retained throughout life. The mediator helps the family and CWH build a harmonious relationship with the disorder and work towards limiting their fears and anxieties. The mediator also helps families avoid perceiving that their fate is closely associated with their CWH’s illness, and also to help them avoid using defence mechanisms, such as suppressing their feelings or being in denial. There are currently more than 50 Italian families participating in We Start Young. For more information, visit www.fondazioneparacelso.it/en/programs-and-activities-en/we-start-young.html.

In the US, the Inhibitor Family Camp (IFC) [8], supported by an education grant from Novo Nordisk and sponsored by Comprehensive Health Education Services, is the only national family camp experience for children (aged 6–18 years) with an active inhibitor. Its objective is to provide education, along with enjoyable activities, in a relaxed atmosphere that helps children with inhibitors and their families to establish meaningful bonds with families facing similar challenges. During the camp, the children are able to meet other children with inhibitors and learn to assimilate new skills and appropriate physical activities. The IFC camps allow children to feel ‘normal’ and part of a larger community. To date, 170 children have participated in the camps, and their feedback shows they found the camp to be a ‘priceless opportunity’: 94% considered the camp was useful and enhanced their relationships with others in the community, and 93% believe they learned from the Faculty and participants. For more information, visit www.comphealthed.com/index.php/inhib/.

The ‘A Bright Future’ workshops [9], sponsored by Baxalta, are a series of customised workshops developed with input from patients and professional members of the bleeding disorder community. The workshops target adults, parents, couples, men, women, teens and community leaders and the goal is to address the emotional and informational needs of people living with bleeding disorders. The workshop setting allows families to share their stories, experiences and needs in an intimate environment. Approximately, 30 workshops are led in the US every year, with about 30 participants in each workshop. Based on participant feedback, 98% of the audience believe the information imparted was highly relevant, and 99% that they are very likely to incorporate the learnings into their lives. For more information, visit www.inalex.com.

While these examples highlight some of the work being done with children and their families – which represents one of the areas of greatest need – there is clearly a need to build on this and develop programmes that support other areas of the lives of PWH and their families, beyond the early stages following diagnosis.

HERO has established a platform to advocate for improved care that reflects the needs of patients – moving beyond disease management, and extending into holistic care of the individual. Working together, we have already started to strengthen the understanding and awareness of psychosocial issues in haemophilia, and provide support for PWH and their families, but improvements are still needed. More community-led programmes are needed to further empower PWH and their families worldwide, with the aim of providing comprehensive support throughout all life stages.

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Laparoscopic surgery in a woman with factor V deficiency: revisiting platelet factor V

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Inherited coagulation factor V deficiency (FVd), first described in 1947 by Owren is a rare autosomal recessive bleeding disorder with a prevalence of 10−6. Patients with severe FVd generally present with easy bruising, epistaxis, menorrhagia and posttraumatic bleeding [1]. In contrast to severe haemophilia, patients with FVd with undetectable levels of plasma factor V (FV) generally have moderate bleeding diathesis. This has been attributed partly to the modifying effects of low levels of tissue factor pathway inhibitor (TFPI) associated with FVd [2], and residual levels of platelet FV/Va [3].

The half-life of FV is reported to be 12–36 h with significant individual variation [1,4,5]. Major surgery in FVd patients requires once or twice daily transfusions of fresh frozen plasma (FFP) to achieve FV levels of 15–20% and maintain FV trough levels of >10% [1,5]. In low-resource countries it appears reasonable to monitor the PT and PTT in assessing the efficacy and guiding therapy.

The duration of FFP support following surgery in FVd remains unclear. Several authors recommend continuing FFP transfusions until wound healing is complete following surgery in FVd [5]. Reports of good outcomes in FVd patients managed with FFP support for 2 or 3 days after major surgery raise questions about the optimal use of postoperative FFP transfusions in FVd. This is in marked contrast to haemophilia where it is standard practice following surgery to administer factor replacement therapy until wound healing is complete.

We report the case of a woman with severe FVd (FV <1%) who underwent a successful laparoscopic assisted vaginal hysterectomy and bilateral salpingo-oophorectomy [LAVH-BSO] with FFP support administered for 72 h postoperatively.

Case
A 44-year-old woman was evaluated for severe menorrhagia of 6 weeks’ duration. She was born out of a consanguineous marriage. She had a history of prolonged bleeding from minor cuts, heavy periods with clots during most of her adult life and recurrent spontaneous bruising over the thighs, abdomen and forearms. She denied epistaxis or oral mucosal bleeding. She had significant bleeding following teeth extraction, and required perioperative FFP transfusion for undiagnosed coagulopathy during Caesarian delivery, breast

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