Event Report: EHC Round Table of Stakeholders on 'Pain Management in Haemophilia'

About the event

On Tuesday 11 June 2019, the European Haemophilia Consortium (EHC) organised a Round Table of Stakeholders on the topic of *'Pain Management in Haemophilia*.' The event took place in the Renaissance Hotel in Brussels, Belgium and brought together over 40 participants representing clinicians, industry, patients and academia. The event's agenda, list of speakers, presentations and a video can be consulted online on the EHC website. Pictures from the event are available on the EHC Facebook page.

Pain Management in Haemophilia

Haemophilia is a rare bleeding disorder caused by a genetic defect preventing normal blood clotting. In affected individuals, this can cause bleeding in the joints and soft tissues, which if left untreated, can lead to disability and premature death. Bleeds are extremely painful, and the damage they cause to joints and soft tissues leads to chronic pain. For these reasons, the primary focus of haemophilia treatment has been, for a long time, to prevent and treat bleeds. At the event, we considered European countries with both low and higher access to treatment. In countries with lower access to treatment, bleeds are more frequent, as is the experience of everyday pain. In European countries with higher access to treatment, we see a generation of people with haemophilia on prophylaxis since early childhood. Despite experiencing fewer major bleeds, these patients are still experiencing *micro*-bleeds and *unrecognised* bleeds. As these types of bleeds are less pronounced and harder to diagnose, they can be left untreated and equally damage joints, cause pain and negatively impact patients' quality of life. In sum, pain is ever-present in the life of people with haemophilia. Unfortunately, for a long time pain management has not been adequately addressed. For this reason, the EHC decided to organise a Round Table on this topic.

Findings and discussions

What is pain?

Pain is a complex mechanism, which arises from different centres in the brain and has two pathways: ascending and descending. In simple terms, in the ascending pathway tissue damage sends signals to the brain, which the brain translates into pain signals, and which are, in turn, sent through the descending pathway to the body. These pain signals manifest themselves as *pain symptoms*. It is essential to know that different parts of the brain are responsible for managing pain signals. This fact can make it more challenging to identify which part of the body generates the pain signal.

Also, several factors may inhibit or facilitate the pain signal in the descending pathway. Emotions like fear, depression, stress and hypervigilance will enhance the feeling of pain while, counterintuitively, physical activity will inhibit the perception of pain. Therefore, experts reiterated several times during the event that: *'There is no bleed without pain, but there may be pain without a bleed*.'

Speakers noted that when assessing pain, clinicians consider bio-psychological criteria because, for example, an increase in pain may not necessarily equal an increase in tissue damage. Traditionally, both patients and clinicians have linked acute pain to *bleeds* and chronic pain to *arthritis*. However, acute and chronic are only adjectives referring to the intensity and length of the pain experienced by the patient. These terms do not explain the cause of the pain.

Nowadays, technological advances such as more portable and user-friendly ultrasound devices allow non-specialist medical professionals to make a more accurate diagnosis. These devices can clearly establish whether a patient is experiencing pain due to a bleed or due to arthritis. This distinction will be invaluable in informing the pain management strategy.

Pain is important to manage because, if left untreated, it can feed into a vicious cycle, which leads to chronic pain a lack of and poor quality of sleep, and altered quality of life. All of these outcomes will continuously feed into an increased sensation of pain.

How much does pain impact patients' quality of life?

During the presentations, it became clear that for a long time both the clinical and patient community didn't focus on pain management as part of the standard haemophilia treatment routine. Recent studies, such as the Patient-Reported Outcomes, Burden and Experiences (PROBE) and the overall Cost of Haemophilia in Europe: a Socioeconomic Survey (CHESS), showed that patients with severe haemophilia overall experience more pain compared to the general population. For instance, haemophilia patients report more pain when performing simple and everyday activities such as walking, climbing stairs, sleeping, resting and bearing weight. These studies also show a correlation between pain experienced, joint damage and higher costs to manage and treat haemophilia. This led the audience to understand that pain not only needs to be prevented and treated early to improve and maintain a patients' quality of life but also to contain treatment costs.

These studies also busted the myths that current prophylaxis regimens lead to zero bleeds and prevent pain. In fact, they showed that over 40% of respondents, who were on prophylaxis since childhood, reported pain. Speakers also stated that the mean age of joint arthropathy in haemophilia is 37 while in the general population it is over 70. Therefore there is still much to be done to close the pain gap between the general and the haemophilia population.

How can pain be managed?

There is a multitude of methods and techniques to manage pain. This Round Table focused primarily on four approaches: surgery, physiotherapy, the use of pain killers and psychological interventions.

The best way to tackle pain management is to have a multidisciplinary approach. The level and severity of the pain experienced by the patient will inform the type of intervention needed, and multiple interventions can be carried out simultaneously. Furthermore, speakers noted that patients may decide to use non-medical methods to alleviate pain. For example, a survey referenced during the event showed that physiotherapy was considered, by surveyed patients, to be as effective in managing pain as meditation and prayer. The experience of pain is very personal. It is linked to the patient's emotional status and his or her history, which will vary in each individual. This historical experience can affect how patients perceive pain. Patients also have the best knowledge about what works and what doesn't work for them in pain management. Therefore, it will be crucial that clinicians listen to patients' experiences and identify multi-factorial causes of pain when designing pain management plans. This approach will also ensure individualised treatment.

Conclusions

This Round Table highlighted the fact that we are only at the beginning of a conversation in pain management in haemophilia. Haemophilia is lagging compared to conditions that also affect the musculoskeletal system like rheumatoid arthritis. This conversation needs to arise from both clinicians and patients. They both need to understand that pain is a phenomenon resulting from physical, emotional and

psychological problems. Pain needs to be considered as part of a larger picture and not just limited to tissue damage.

It was also clear from presentations and discussions that more research in this field is needed and that there needs to be a more systematic approach to researching pain. Participants suggested that clinical trials in novel therapies should include pain assessment. Participants also noted that there is a need for the development of better tools to measure pain, in particular to understand the effects of micro- and unrecognised bleeds, which affect patients who have been on prophylaxis since early childhood.

Speakers agreed that pain management should be individualised and based on a multidisciplinary approach. However, they also stated that this is often a significant challenge. In particular, in centres that do not have access to all medical specialities or in centres with limited face time with patients due to personnel or budget constraints.

Finally, participants agreed on the need for active patient involvement, as well as an individualised strategy for pain management. Firstly, patients will help to inform the cause of and the best approach to manage pain. Secondly, patients need to be included in the pain management process to ensure that they will adhere to the treatment provided.

The Round Table on Pain Management clarified that pain is a complex issue, which demands proper management to achieve satisfactory treatment for the patient. Participants asked for concrete European policies to make improved pain management accessible for all people with bleeding disorders in Europe.