Comment on: "Clinical and functional evaluation of the joint status of hemophiliac adults at a Brazilian blood center"

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In this issue of the Revista Brasileira de Hematologia e Hemoterapia, Ferreira et al. present a study entitled "Clinical and functional evaluation of the joint status of hemophiliac adults at a Brazilian blood center"[1]. The authors are to be commended for presenting their paper on the outcomes of the clinical and functional evaluation of adults with hemophilia from a Hemophilia Centre in Brazil. The findings identify the importance of evaluating this specific group of patients with hemophilia in order to develop a deep understanding of their physical status and the problems that this can pose for the individuals concerned. The findings will also provide evidence to begin to address the issues raised in the study and it is hoped that this will ultimately improve patient outcomes. The use of standardized measures is essential in this regard and the authors have used two well established tools to assess the adults in the study.

This scientific comment will make three observations on the findings outlined in the paper

- The importance of primary prophylaxis
- The use of standardized measures
- The lack of engagement with physiotherapy

The importance of primary prophylaxis

The use of blood products to treat bleeding episodes is very costly and there is no doubt that this poses significant challenges for all health care providers and particularly so in developing countries. There is a wealth of evidence in the literature that the use of primary prophylaxis is crucial in the prevention of hemophilic arthropathy. It is pleasing to note that the Brazilian Ministry of Health is intending to implement primary prophylaxis for children under three years of age. This will help to avoid the many physical problems that are apparent in this patient cohort who have only had access to blood products on demand. Ultimately it is hoped that in the future effective primary prophylaxis will be available to all as necessary throughout life.

The use of standardized measures

This paper has used two commonly used and established standardized measures to evaluate joint status, the World Federation of Hemophilia Physical Examination (WFH-PE) scale and the Functional Independence Score for Hemophilia (FISH). The WFH-PE scale is a measure of joint and muscle impairment and was the first scale to be developed to assess joint status. More recently other measures have been developed such as the Hemophilia Joint Health Score (HJHS)[2] as it was found that the WFH-PE Scale was not sensitive enough to detect the more minor joint changes that are often more apparent in children and adults who have had access to factor replacement on a regular basis. In this paper the physical manifestations of arthropathy are profound and so the original impairment scale is still of value. One of the limitations of impairment measures is that they may be more relevant to the health care provider than the individuals concerned who may be more interested in their ability to undertake functional activities rather than, for example, their actual range of movement. Combining the use of the WFH-PE Scale with a functional measure such as the FISH therefore provides a broader perspective on the individual's physical status. The FISH is a hemophilia-specific measure of performance of functional activities as assessed by the therapist. This can be further supported by functional tools such as the Hemophilia Activities List (HAL). The HAL is a self administered questionnaire that provides a measure of the patient's perceived ability to undertake a range of functional tasks[3]. A combination of both measures may be appropriate in ascertaining the overall functional ability of the patient as they measure different constructs. In addition, quality of life (QoL) measures are increasingly being used to explore the impact of hemophilia and this would be a useful area to explore in this group of patients in the future.

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An evaluation of some of the main hemophilia standardized tools can be accessed at the World Federation of Hemophilia Site[4].

The lack of engagement with physiotherapy

It is interesting to note that, despite the majority of the severe and moderately affected patients demonstrating manifestations of hemophilic arthropathy, only 8/31 had received any physiotherapy in the previous 12 months. The reasons for not seeking treatment from a physiotherapist included being thought to be unnecessary, that it would make the condition worse, that physiotherapy was not available and not being referred for physiotherapy. This suggests that there is considerable work to be done to both educate the patients and the health care providers on the value of physiotherapy. Physiotherapists who develop expertise in the physical treatment of hemophilia have much to offer patients in managing and improving their joint status. Physiotherapy management can include advice on adopting an appropriate lifestyle and treating the effects of bleeding episodes such as a lack of range of movement, poor proprioception and muscle weakness. This is particularly important where regular factor replacement is not readily available as appropriate physiotherapy modalities and techniques carried out with due regard to avoiding exacerbation of symptoms and any rebleeding can have significant benefits in maintaining/restoring optimal joint function.

The Musculoskeletal Committee of the World Federation of Hemophilia[5] provides a vehicle for the sharing of experiences of health care providers including physicians, orthopedic surgeons and physiotherapists who are involved in the management of hemophilia. The World Federation of Hemophilia also produces a range of publications on the management of individuals with hemophilia. Drawing on best practice and appropriate evidence, the physical status of the patient with hemophilia can be optimized enabling a fulfilling and enjoyable quality of life.

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