Web Platform to Support the Portuguese National Registry of Haemophilia and Other Inherited Blood Disorders

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ABSTRACT

Patient registries are essential tools for identifying and tracking people with a particular disease and for collecting epidemiological information, having a special role in rare and chronic diseases, where haemophilia and other inherited blood disorders (HoIBD) are classified. Web-based technologies represent an excellent solution to support different types of registries, due to the benefits that they can promote in the management of disease data. This work presents the web platform developed in a joint initiative between the Portuguese Association of Congenital Coagulopathies (PACC) and the University of Aveiro (UA), with the purpose of creating the first National Patients Registry (NPR) with HoIBD in Portugal. This application is hosted in the data centre of the UA, and at this moment it is already used by clinicians of the different Haemophilia Treatment Centres (HTC) located in Portugal, with the next challenge being the increase in the number of users.

Keywords: Chronic and Rare Diseases, Haemophilia, Inherited Blood Disorders, National Patients Registry, Portugal, Web Application

INTRODUCTION

In recent years there has been significant advancement in using Information and Communication Technologies (ICT) in the field of rare and chronic diseases, where a large part of inherited blood disorders are classified. Haemophilia is the most common type of rare and chronic inherited bleeding diseases, characterized as an X-linked congenital disorder caused by a deficiency of coagulation factor VIII (in the case of haemophilia A) or factor IX (in the case of haemophilia

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B), affecting the population in a ratio of 1 case of haemophilia (PWH) per 10000 people born (Srivastava et al., 2013).

The vision of the World Federation of Haemophilia (WFH) with ‘Treatment for all’ is that all people that suffer with haemophilia or other rare inherited blood disorder (HoIBD) must have access to medical care and appropriate treatments (Skinner, 2006). Rare diseases, also known as orphan diseases, affect a small percentage of the population, and their low prevalence requires special efforts to deal with them so as to improve diagnosis, care and hence prevent complications (Teixeira, Saavedra, Ferreira, & Santos, 2013). To this end, the first important step for improving care is the identification of all patients who suffer from this disease, being the National Patient Registry (NPR) the fundamental tool for this identification (Srivastava et al., 2013). Registries for rare and chronic diseases are broadly accepted for their usefulness in monitoring the identification and diagnosis of people suffering from that disease and evaluating their health. According to the report of the WFH, an important measure for improving the management of HoIBD is the existence of patient registries, which, in a national context, could lead to the NPR (Evatt, 2005).

A registry is a database or a collection of records of people identified with HoIBD (Evatt, 2005). In the case of people suffering from rare and chronic diseases such HoIBD, these kinds of registries are particularly useful since important research questions cannot be answered without knowing a set of information about the prevalence and evolution of the disease. In fact, registries of patients have the invaluable potential to provide an understanding of the disorder, to provide useful information for planning health care services and to identify suitable groups of patients for enrolment in clinical trials (Viviani, Zolin, Mehta, & Olesen, 2014).

The practical purpose of a registry in the context of HoIBD is to define the population demographics and collect observational data on specific health concerns. In addition to the demographic characterization of the population, the existence of those registries also allows the collection of data for statistical analysis related to the specificity of the disease, such as the prevalence of viral infections, existence of inhibitor factors, implementation of prophylactic treatments and joint evaluation, among others, to assist the decision-making process.

Given the importance of these data for disease management, the consistent definition of these data, their proper storage, and possibility of subsequent extraction of information are important factors for a more objective view of the practice of HoIBD care, with a profound impact on the health and quality of life of these patients.

The lack of a NPR for HoIBD in Portugal, associated with the difficulty that clinicians working in this field are facing in order to manage the specific patient information at a national level, has motivated a group of physicians to look for a technological solution in order to facilitate and optimize the information management process.

This article presents the newly developed web platform to support the Portuguese NPR of haemophilia and other inherited blood disorders (HoIBD), a project which arose from a joint initiative between haemophilia healthcare professionals, represented by the Portuguese Association of Congenital Coagulopathies (PACC) and a group of researchers from the University of Aveiro (UA) responsible for analysing, developing and implementing the technological solution.

**NATIONAL PATIENT REGISTRY IN CHRONIC AND RARE DISEASES**

In chronic and rare diseases, the patient registry is often the first step to know the incidence and estimate the prevalence of the disease (Richesson & Vehik, 2010). The registry represents
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