



A mobile app for patients with Pompe disease and its possible clinical applications

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Abstract

In recent years, the potential of *smart technology* to provide innovative solutions for disease management has raised high expectations for patients' and healthcare professionals' community.

We developed a mobile app, called *AIGkit*, specifically designed for adult patients with Pompe disease, with the aim to help them manage the burden of illness-related factors, and also to provide clinicians with continuous tracking of each patient in real-time and ambient conditions of everyday life. We present the *AIGkit* as an innovative approach exploiting cutting-edge technology to improve quality of care and research into neuromuscular disorders.

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1. Introduction

E-health technologies have been recognized as high-potential tools in enhancing healthcare quality, accessibility and delivery. Notably, in recent years, the potential of *smart technology* to provide innovative solutions for disease management has progressively raised high expectations for patients' and healthcare professionals' community. Smartphones have been a “revolution” in the way in which people can communicate. The possibility to be “always on-line” and to carry them everywhere provide real-time on-demand communication, while their rich multimedia touch-displays operate with increasing speeds and deliver data services to document and improve the networked lives of their owners. These elements offer a unique opportunity to provide medical support *when and where people need it*. To date, a large number and variety of health-related apps (more than forty thousand)

have been released in the market, ranging from basic apps characterized simply by text message alerts, to sophisticated apps, helping patients in managing chronic conditions and lifestyle management [1]. Some apps cover a broad spectrum of general medical knowledge, others can be tailored to specific purposes. Many apps are developed for healthcare workers, including physicians, nurses and assistants. There are also patient-centered apps capable of performing a wide array of functions [2].

Here we present a mobile app specifically designed for adult patients with diagnosis of Pompe disease, called *AIGkit*. Pompe disease, or glycogen storage disease type II, is an inherited lysosomal storage muscle disorder caused by a deficiency of the enzyme acid alpha-glucosidase (GAA). The adult form of disease, complained by the majority of patients, is characterized by a variable age at onset and degree of severity and is mainly associated with a progressive limb girdle and respiratory muscles impairment. Since 2006, enzyme replacement therapy (ERT) with alglucosidase alfa (Genzyme, Cambridge, MA, USA) has been available, improving overall survival. In patients with adult form of disease, ERT has resulted in disease course

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stabilization with motor and pulmonary improvements. However, a multidisciplinary approach to care remains a crucial factor to handle the whole impact of the disease and clinical outcome. As in other chronic and progressive illness, especially if these are rare diseases, and also for Pompe disease, it has become helpful to define conceptual disease models [3] that can provide a comprehensive overview of various clinical aspects, supporting the plan of innovative healthcare strategies and decision-making; such models allow the extrapolation of outcomes based on relatively short-term follow-up data of a limited number of patients, by combining available data with known disease-specific correlations [4]. A mobile app designed for patients with Pompe disease, who are individuals with special healthcare needs and with considerable levels of psychosocial challenges, can be a technological tool useful to help them manage illness-related issues, thus reducing its burden, and to provide clinicians with continuous tracking of each patient at real-time and ecological conditions of everyday life.

2. Material and methods

2.1. Study design

The project was born under the collaboration between the Italian patients Association of Glycogenoses (Associazione Italiana Glicogenosi, AIG, <http://www.aig-aig.it>) and the scientific Italian Society of Myology (AIM, <http://www.miologia.org>). As first step (Fig. 1), we defined a multidisciplinary working group, composed of three neurologists, one pneumologist, one psychologist, three computer engineers and two members of AIG, including a caregiver and a patient affected by a late-onset form of Pompe disease. The kick-off meeting was held in September 2015 in Florence (Italy), during which we discussed the aims and the endpoints of the app, that was then called *AIGkit*, and we defined the timeline of our project. The engineers collected the clinicians' and patients' requirements and implemented a wireframe of *AIGkit*. After then the physicians and the patients checked again the wireframe giving their final feedback, and the app was started to develop. The beta version of *AIGkit* was for the first time presented at the 16th AIM Congress [5] and it was released in June 2016 in the Italian language version. We then set up a usability testing phase in September 2016 in which 8 volunteer subjects affected by Pompe disease (4 females, 4 males, age range 22–57 years, more details in [Supplementary Table 1](#)), recruited through the AIG association, downloaded and tested the app from a private store and used the *AIGKit* for two weeks. At the end of these two weeks, the users were invited to complete a questionnaire, focused to measure the capacity to meet its intended purpose, further adding any comments or suggestions. Overall, the result of the users testing phase was quite positive: all the recruited subjects stressed the good usability of the app, which they defined intuitive and functional for its purpose. The patients' feedback, including their comments and suggestions, was then discussed by the working group during a meeting in October 2016 to further refine

AIGkit. We released on the online stores the updated *AIGkit* in Italian language version in October 2016 (Fig. 1).

3. Methods

The application was created using Sencha Touch framework based on web standards such as HTML5, CSS3 and JavaScript. The goal of Sencha Touch is to facilitate quick and easy development of mobile apps that run on iOS, Android and Windows. In this way, web apps work almost as native applications. Only for iOS user that provide permission to read and write health and activity data to Health app, *AIGkit* can become a valuable health data source and it can use the shared data to bring a powerful health solutions. The exchange of data between application and web portal uses the HTTPS protocol that protects the privacy and integrity of the data. All the data stored on the Amazon servers (personal, real life and follow up data) are backed up daily, saved on different databases and encrypted in case of sensitive values.

3.1. Store links

Apple – <https://itunes.apple.com/it/app/aigkit/id1156925493?mt=8>

Android – <https://play.google.com/store/apps/details?id=it.vidiemme.dist.aig.aigkit>

Windows – <https://www.microsoft.com/it-it/store/p/aigkit/9nblggh554lp>

4. Results

We aimed to design a very easy and intuitive app. The architecture includes four main sections: a) the section *To know my disease*, that contains useful details for patients regarding clinical features of Pompe disease, diagnosis and management; b) the section *News*, that updates news flash about medical or welfare issues of the disease, through the synchronization by internet connection with AIM and AIG websites; c) the sections *My clinical paperwork* and d) *Motor training plan*, which are illustrated below. The home page also includes two additional functions: *My helpline*, that contains AIG association contacts and e-mail address for questions and support, and *My emergency contacts*, in which through a single button the user can call the phone number that it has previously set or send a short message service showing automatically her/his geographical position on the Google map.

The section “*My Clinical paperwork*” is featured as a diary in which the patient can schedule her/his therapeutic infusions (by setting the first infusion appointment the app will synchronize and remind the following ones) and, importantly, collect several data related to her/his health status in real-world framework, as shown in Fig. 2. In order to simplify the daily collection of data, some information such as performed number of steps and distance meters can be registered automatically by allowing *AIGkit* to synchronize data from the operating system (OS) of the phone, but at the moment it is feasible only for iOS devices users. In a similar way breathing parameters can be added by synchronizing a bluetooth pulse oximeter.

“*My Clinical paperwork*” also contains a brief questionnaire designed to capture patients' feelings and emotions of

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