Medical Xpress

Rare disease challenges ICT researchers

September 28 2016

While some children need daily reminders to take their medicines, others have a more pressing need simply to talk to like-minded people about their disease. Credit: Thor Nielsen/SINTEF

People with cystic fibrosis (CF) need help to ensure they are getting correct nutrition and the right amount of enzymes. They also need constant reminders. Researchers are now developing a digital support

device to promote autonomy, but are finding that this is no easy task.

CF is a rare, chronic and hereditary condition. Sufferers often develop mucus in their lungs and have problems with nutrient uptake.

As part of the international project 'MyCyFAPP', researchers are developing a digital system (app) designed to help CF sufferers and their next of kin to understand the disease and monitor treatment. Interviews were conducted recently among 71 people from seven European countries to obtain an overview of the needs that such an app must meet. Sufferers of different ages took part, together with parents, professionals and a variety of special interest organisations.

"We knew that this work wasn't going to be easy", says Jacqueline Floch, who is leading the work being carried out at SINTEF. "However, the complexity of the problem still surprised us", she says.

Different needs

The researchers came up with a total of 466 different and complex needs – an overwhelming number to begin dealing with.

"Firstly, we observe that all CF sufferers are different", says Floch. "Some are very organised when it comes to monitoring their illness, while others are far less scrupulous. Some children may also be in denial because they don't want to appear different. Some get a lot of support from their parents, who lay out the medicines and teach the kids how to take them, while others get almost no help at all.

This means that while some need daily reminders to take their medicines, others have a more pressing need simply to talk to like-minded people about their disease", she says.

CF is a condition in which an individual's needs are influenced by the gradual progression of the disease and the sufferer's situational circumstances. The everyday needs of life at home are one thing, but in other situations, such as at school or on holiday, a sufferer's needs will change.

The researchers discovered that there are also differences between circumstances in the countries taking part in the MyCyFAPP project. Norway has an efficient CF user association (the NFCF) that has set up popular closed groups on Facebook. It also organises a service providing peer mentors who are trained to support others in the same situation as themselves. Such a system is not found outside Norway.

Needs of doctors and patients

In some cases, doctors, CF sufferers and parents have vastly different requirements, as well as different priorities. For example, the doctors taking part in the MyCyFAPP project say that it is important to measure an individual's enzyme doses accurately because nutrition affects all aspects of a CF sufferer's health. (Enzymes in tablet form help the body to absorb proteins and fats when digesting food). In contrast, adults with CF and parents of children with the disease believe that they become proficient at controlling enzyme doses once they become more experienced.

"However", says Floch, "the project found that in Europe there are large variations in the enzyme dose recommendations provided by doctors, and there is reason to believe that overdosing is being carried out in some places. We're also seeing that many CF sufferers are looking for MORE than just a support device to help them calculate their enzyme doses. For example, some want an intelligent system that can make recommendations about treatment based on their own specific symptoms", she says.

"The doctors taking part in the project are sceptical to this suggestion. They fear that such a system will deliver incorrect advice", continues Floch. "Our view is that the doctors' knowledge and experience must be taken seriously and that it is important that they carry out quality assurance of the work linked to the project. At the same time, it's important that the system we develop is perceived by its users as beneficial. If not, no-one will use it", she says.

The researchers also revealed many needs that were common to all participants. The doctors think it is important that sufferers understand their disease and that good understanding will better enable them to monitor their treatments. Parents want their children to learn more about the disease, and the young people taking part in the project agree. As well as learning aspects, a wish to record a daily log of health status and symptoms also emerged. The aim of this is to enable sufferers to better understand the disease and their own health status.

Differences in treatment support

Being prepared to take responsibility for one's own treatment is an excellent launch pad when it comes to using digital devices. The results from the MyCyFAPP project may indicate that a digital device for promoting autonomy has greater potential in Norway than in some other European countries, such as Belgium, for example. There are about 300 CF sufferers in Norway and they live all across the country.

The Norwegian Expert Centre for Cystic Fibrosis (NSCF) in Oslo is the only centre of its kind in the country. It commonly provides sufferers with annual check-ups, while additional monitoring is provided by local hospitals. Participants in the project stated that while they were satisfied with the services provided by the NSCF, they would prefer all their treatment to be carried out there if they were given the choice.

In Belgium, about 1200 CF sufferers are monitored exclusively by one of a total of seven expert centres spread around the country. A digital device may help to reduce the distance between the patient and health personnel.

Good nutrition is key

The researchers have now categorised the needs into a set of functional groups based on the calculation of enzyme doses, nutritional monitoring, a daily symptoms log, treatment monitoring support, reminders, learning, digital fora and communication between CF sufferers and their doctors.

Doctors taking part in the MyCyFAPP project have been working to assign priorities between these groups, and the symptoms log and learning have been rated with high priority. However, enzyme doses, correct nutrition, the intake of calorie-rich foods and a balanced diet are without doubt the most important factors – even though the need for high-calorie foods may represent a dilemma. If a sufferer eats too much fat, he or she may incur cardiac and blood circulation conditions.

On the basis of these priorities, the researchers have organised so-called "co-design workshops". Here, a handful of children aged between 4 and 15, together with their parents and adult CF sufferers, work together to develop ideas for games and make sketches for an app designed to assist sufferer autonomy. Health personnel, including doctors, nutritional physiologists and nurses, have also taken part in making design sketches for a monitoring device.

"Software development is well underway, and an initial assessment is planned for this autumn", says Jacqueline Floch. The aim is to produce software ready for a clinical evaluation in the summer of 2017.

Cystic fibrosis (CF) is a serious hereditary condition that disrupts the

normal transport of salts and water in the body's cells. This disruption causes thick mucus to accumulate in the lungs, creating favourable conditions for infections to develop. The genetic disorder means that patients suffer inadequate uptake of proteins and fats from food in the digestive tract. Many sufferers take enzymes in tablet form in order to supplement their digestive enzymes. This can result in problems with maintaining weight and height, and may cause a general deterioration in condition. New developments in medical technology and medicines have enabled improved diagnoses, treatment and survival rates linked to CF.

There are 300 people in Norway registered as having CF. The Norwegian Expert Centre for Cystic Fibrosis), which is annexed to Oslo University Hospital, is the only centre of its kind in Norway. The Norwegian Cystic Fibrosis Association (NFCF) has set up popular Facebook groups that enable people to share their experiences. The NFCF also organises a system of peer mentors who assist others in the same situation as themselves.

The EU project MyCyFAPP is running from 2015 to 2019, and involves both medical personnel and ICT experts. The project partners come from Spain, Germany, Portugal, Italy, Belgium, the Netherlands and Norway.

The aim of the project is to design a digital device (app) to promote autonomy, with input from CF sufferers, doctors and the parents of children suffering from CF. The app is intended to help both adults and children suffering from CF to determine the enzyme doses they must take with their meals, and to assist with treatment monitoring. A game will be integrated to provide children with CF with a gradual learning programme about the disease.

Provided by SINTEF