Telehomecare in Cystic Fibrosis: a review of the literature and the state of the art.†

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Cystic Fibrosis (CF) is the most common hereditary genetic disease among populations of Caucasian origin, with an incidence in Italy of 1/5.510 living births in 2016.[1] Mucous secretions are more dense than normal, with negative effects especially on the respiratory and digestive systems. The main complications are chronic liver disease, chronic pancreatitis with diabetes and recurring episodes of bronchitis or bronchopneumonia due to repeated bacterial lung infections with progressive organ damage. The clinical course is characterized by progressive reduction of respiratory function overtime. The trend of spirometry is 2%/year decrease of FEV1.[2] In case of infectious relapse, changes in pulmonary function (< FEV1) often precede clinical symptoms.[3] An early antibiotic treatment can prevent more serious complications, allow less invasive therapies (orally) and limit pulmonary damage in the long term.[4]

In 2005, Magrabi et al., in Australia, first exposed the possibility of using remote telecare in 5 CF adolescents,[5] concluding that home telecare may be a feasible intervention for monitoring CF.

In Rome, in Cystic Fibrosis Centre of Pediatric Hospital Bambino Gesù (OPBG CF Centre), since 2001 telehomecare (THC) was used in follow-up of patients at home, in order to prevent or early treat respiratory exacerbations. A first prospective, open-label study was carried out on a total of 45 patients enrolled between 2001 and 2005, with the aim to investigate the effect of THC in CF patients.[6] 17 CF subjects were enrolled (11 f, 6 m; mean age 5.74±5.8) and telemedicine was administered in addition to standard therapy. As controls 28 CF-patients were enrolled (13 f, 15 m; mean age 14.77±5.22) matching for respiratory function, bacterial colonization, sex, age, and complications, who received standard therapy only. Follow-up duration was 29.30±13.32 months. The endpoint was the trend of monthly FEV1 mean values overtime. In THC-treated subjects, a statistically significant decrease of outpatient accesses and an increase of therapy cycles were observed. THC seemed to increase in general the rate of access to health care, without a clear effect on pulmonary function. Moreover, in the THC-treated group were noted lower SD values of monthly mean FEV1 overtime. This trend might suggest an improved clinical stability and therefore a better quality of life. The results were confirmed by further similar studies covering the following years up to 2016.[7]

This study was later included in a systematic review appeared in 2012 on the use of telehealth in adult and pediatric CF patients for monitoring symptoms, assessing adherence to prescribed therapies or providing a therapeutic intervention.[8] The work was also selected, among eight studies, among a total of 293 references from four electronic databases. The Authors concluded that in considered studies available evidence is insufficient to reach a solid conclusion about the benefits of telehealth in people with CF, but this remains a promising area for future investigation.

These results were confirmed by a later work by Lechtzin et al.:[9] a multicenter, randomized trial including a total of 267 patients from 14 CF Centers in USA aimed at determining whether the early detection of pulmonary exacerbations using home spirometry and symptom monitoring could result in a slower decline in lung function. The primary outcome was the 52-week change in FEV1. Secondary outcomes included time to first exacerbation and subsequent exacerbation, quality of life, and change in weight. There was no significant difference between study arms in 52-week mean change in FEV1 slope, meaning THC administration did not result in slower decline in lung function, despite exacerbations were detected more frequently in the early intervention arm subjects, compared to usual care arm. This study was commented by Martelli et al.,[10] who expressed some concerns about possible bias associated with lack of blinding, low adherence rates, possible poor spirometry technique, different antibiotic administration in the two study arms. Lechtzin answered to Martelli[11] stressing the importance of letting clinicians select and administer treatment when seen fit, giving paramount priority to the patient wellness. As regards poor sampling techniques, individual pilot data displayed good agreement between home and lab spirometry; moreover, supplementary analyses failed to demonstrate differences in outcomes whether individuals adhered to the intervention more or less; last, the Authors expressed the believe that adding health technologies may allow for further tailoring of exacerbation treatment.

Some opinions were also recently expressed either in favor[12] or against[13] the use of THC in the follow-up of chronic diseases, but a definitive agreement is still to be reached.

Recently, following the progress and wider diffusion of innovative technologies, new horizons seem to have opened up.

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Wood et al. developed a smartphone application for adults with CF to allow reporting of symptoms suggestive of exacerbation. Sixty participants were included in the study, among whom 29 (48%) allocated to the intervention group.[14] The primary outcome measure was the number of courses and days of intravenous (IV) antibiotics, to be used in the most serious exacerbations. Over a 1 year follow-up, the number of courses of oral antibiotics increased, without any clear effect on the number of courses of IV antibiotics. The median time to detection of exacerbation was shorter in the intervention group. The conclusion was that the use of a smartphone application reduced time to detect respiratory exacerbations that required antibiotics, but did not demonstrate a clear effect on the number of courses of IV antibiotics.

In conclusion, the application of THC in the follow-up of CF-patients although can not lead to radical treatment of the disease, seems however to improve the stability of respiratory function and, through a faster identification of relapses, could possibly reduce the lung damage secondary to respiratory exacerbations overtime.

In the study of Wood, the use of an application that reported symptoms was enough to improve the detection of exacerbations. Medical history and symptom collection are both essential parts in every diagnostic procedure. An accurately standardized anamnesis could likely improve the effectiveness of the whole detection procedure of exacerbations. Further studies are needed to achieve a standardization of the queries, in order to balance the diagnostic significance of every medical news. Instrumental data as Fev1 measurement, on the other hand, are to be considered only as a part of the physical examination. Its clinical significance depends on and must be considered only in the context of the entire clinical framework, of which the anamnesis and subjective symptoms are essential parts.

It remains to be established whether and how long the application of telemonitoring can be reflected in the long term on life expectancy of individuals with CF. At today, the prognosis of CF has improved significantly over the last two decades and may improve further as newer medications can be used. From 1958 on, many life expectancy indicators in CF were developed, including several markers of disease and composite clinical and radiological prediction tools.[15]

To evaluate a possible positive effect of THC application at a long term, further case/control studies on life expectancy of CF subjects treated with THC are to be planned in the future.

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