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Aquagenic Wrinkling of the Palms in Patients with Cystic Fibrosis

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Dear editor,

Aquagenic wrinkling of the palms (AWP) is a condition characterized by oedema, confluent white papules and excessive wrinkling of the palms after few minutes exposure to water. The phenomenon may be associated with pain, numbness and pruritus. It was first noticed and described in 1974 in children with cystic fibrosis (CF) by a paediatrician R.B. Elliott. Case reports and some smaller studies have later confirmed this association. CF is an autosomal recessive inherited abnormality caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene leading to abnormal transport of chloride over epithelial membranes.

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Our primary aim with this study was to determine the frequency of AWP in an adult cohort of CF patients in Denmark as the phenomenon seems to be underestimated and unreported among CF patients and physicians, who may focus more on critical symptoms during clinical visits.

CF patients were recruited from the Copenhagen Cystic Fibrosis Centre. In total, 103 patients with CF were included and asked to complete a questionnaire. Data on AWP, sex, age, body mass index (BMI), drug intake, hyperhidrosis, other skin diseases and CFTR mutation was collected. A hand immersion water test was carried out in 10 patients. For statistically analysis, chi-squared test was used for binary variables.

Eighty of 103 CF patients (78 %) described a present or former history of AWP. Demographics in the study population showed that CF patients with AWP are slightly younger (30.6 vs. 38.8 years), while sex and BMI did not show any difference in the two groups. A description of the demographics for the CF patients with and without AWP is available on request. Most patients (51 %) described AWP emerging between 5-10 minutes after contact with water, while 15 % and 24 % described the phenomenon after 1-5 minutes and 10 minutes respectively.

Fifty-seven patients (55 %) reported a history of hyperhidrosis with sweaty hands or feet, and in this group 93 % had AWP compared to 59 % of CF patients without hyperhidrosis (p < 0.001).

Seventy-three individuals were homozygous for ΔF508 and 54 of these (74 %) described AWP. Among 30 non-homozygous patients, 26 (87 %) described AWP. The difference in occurrence of AWP in CF patients homozygous for ΔF508 compared with non-homozygous was not statistically significant (p = 0.16).

The hand immersion water test was performed in 10 CF patients, of which 9 patients described AWP before the test. Of the 9 self-reported AWP subjects, 5 demonstrated extensive palmar wrinkling after 10 minutes while 4 subjects demonstrated minor palmar wrinkling. Two male patients with excessive palmar wrinkling experienced symptoms including pruritus after 2-3 minutes (Figure 1).

We investigated one fifth of the total Danish CF population† and found the prevalence of AWP to be 78 %. Previous studies documented a prevalence of AWP between 41% (43/105) and 84 % (43/51) in CF patients. Patient interviews revealed a variety of accompanying symptoms such as pain and pruritus and the phenomenon seemed to be underestimated and unreported in the medical records. Some patients described swimming as impossible, due to pain in hands and feet.

Fifty-five % of CF patients reported hyperhidrosis, which is significantly higher than the background prevalence of ~3 %. It was clear, that patients with self-reported hyperhidrosis
experienced more frequent AWP in contrast to patients without hyperhidrosis (93 % vs. 59 %) (p < 0.001). This might be due to the higher sweat rate or hyperosmolality. Treatment of hyperhidrosis might be helpful in reducing AWP, which should be confirmed in prospective studies.

Regarding potential aggravating medication, only a small group of patients (6 out of 80) had an occasional intake. We found no plausible association between medication and AWP. On the other hand one CF patient reported a significant reduction in palmar hyperhidrosis after treatment with ivacaftor, which is a systemic CFTR modulator proposed to decrease the sweat chloride concentrations and used for her specific genotype CFTR G551D mutation. The hand immersion water test of this patient showed no sign of AWP, although she described a former history of AWP.

This study has some limitations, as we only included one fifth of the CF population in Denmark and only adult patients. We also performed the water test in only 10 patients since the study initially was designed as a questionnaire study.

In conclusion, this study confirms that AWP is frequent among CF patients and show a correlation between AWP and palmoplantar hyperhidrosis. Most CF patients stated the skin symptom to be a severe problem with pain and pruritus, which highlights the relevance to recognize and relieve AWP.

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References


**Figure 1** Two 35-year old male patients with cystic fibrosis (CF) and aquagenic wrinkling of the palms (AWP), both homozygous for ΔF508. Patient 1 after 10 minutes of water immersion of his left hand (A). Close-up (B) showing spongy white skin with excessive palmar wrinkling, particularly around the fingertips. Patient 2 after a 10 minute water test (C).