inflammatory cytokines in keratinocytes) and their anti-
collagenase activity via inhibition of the dermal matrix
metalloproteinases. Metalloproteinase 9 and its inhibitor
have been implicated in HHD and in Darier disease.\textsuperscript{7,8}

The recent publication of 6 cases of HHD with a dramatic
response to doxycycline,\textsuperscript{9} with ease of access and manage-
ment, low cost, and few side effects, led us to use this
drug. The treatment achieved an excellent response never
before experienced by our patient either spontaneously or
with other topical treatments (dermal corticosteroids and
fusidic acid).

**Conflicts of Interest**

The authors declare that they have no conflicts of interest

**References**

   Mutations in ATP2C1, encoding a calcium pump, cause Hailey-
2. Dhitavat J, Fairclough RJ, Hovnanian A, Burge SM. Calcium pumps
   and keratinocytes: lessons from Darier’s disease and Hailey-
3. Leinonen PT, Hägg PW, Peltonen S. Rerevaluation of the normal
   epidermal calcium gradient, and analysis of calcium levels and

**Importance of educational
sessions on cardiometabolic
comorbidities. Awareness among
psoriasis patients**

**Importancia de las sesiones educativas sobre
comorbididades cardiometabólicas. Conciencia entre los pacientes con psoriasis**

**Dear Editor:**

There is strong evidence that psoriasis is associated with sev-
eral cardiometabolic comorbidities, and that patients with
psoriasis are at a higher risk of cardiovascular morbidity and
mortality.\textsuperscript{1,2} Understanding this is of crucial importance, not
only for physicians but also for patients, as it can impact
prognosis and patient quality of life.\textsuperscript{4}

It has been shown that few patients with moderate
to severe psoriasis are aware of their increased risk of
atherothrombotic disease and metabolic syndrome.\textsuperscript{5} Edu-
cational sessions are a recognized tool for informing and
helping patients to understand the nature and course of their
disease and the different treatments available, and can also help
them to develop coping strategies.\textsuperscript{6,7}

We performed an observational study to evaluate the
impact of an educational session designed to promote knowl-
dge among patients with psoriasis about their disease,
lifestyle changes, and management of cardiometabolic
comorbidities.

The educational session was held in the psoriasis unit
of a Portuguese tertiary hospital. Briefly, it consisted of sev-
eral oral presentations (30 min each) explaining the
nature of psoriasis, introducing the various treatment
options, exploring the association between psoriasis and
cardiometabolic comorbidities/cardiovascular disease, and
underlining the importance of monitoring and treating
these. A questionnaire was created for the patients to com-
plete before, immediately after, and 6 months after the
session. The questionnaire included demographic informa-
tion, questions regarding the association between psoriasis
and cardiometabolic comorbidities/cardiovascular disease,
and assessment of lifestyle and comorbidity management.
Seventy patients participated in the session and 53 com-
pleted all 3 questionnaires correctly. The demographic data,
characteristics of disease, and treatments received are pre-
sented in Table 1. Regarding cardiometabolic comorbidities,
35.8%, 13.2%, and 35.8% of patients had a respective diag-
nosis of hypertension, diabetes mellitus, and dyslipidemia;
20.8% were obese (body mass index > 30); and 18.9% were
active smokers (Table 1).

The McNemar test was used to assess significant improve-
ments in knowledge between the different time points. A
P value of less than or equal to .05 was considered sta-
tistically significant. A significant increase was observed in
the percentage of correctly answered questions about the
association between psoriasis and cardiometabolic comor-
bidities/cardiovascular disease on comparing the answers

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We performed an observational study to evaluate the
impact of an educational session designed to promote knowl-
dge among patients with psoriasis about their disease,
before and immediately after the session and before the session and 6 months later (Table 2). When logistic regression models adjusting for age, sex, and level of education were applied, the results were not altered.

Six months after the session, 60.4% of respondents stated that they had started to exercise or had increased their level of physical activity; 49.1% stated that they had lost weight (mean loss, 2.3 kg; maximum loss, 7 kg; minimum loss, 1 kg); and 50% stated that they had cut down on smoking, although none had quit.

After the session, over 90% of the participants stated that they were willing to consult their general practitioner to screen for cardiometabolic comorbidities or monitor those already diagnosed. This attitude led to newly diagnosed comorbidities (7 cases of dyslipidemia, 2 cases of hypertension, and 2 cases of diabetes mellitus) and therapeutic adjustments (in 4 cases of hypertension, 4 cases of dyslipidemia, and 1 case of diabetes mellitus).

Our results show that an educational session held with patients with psoriasis had a real impact on their health, leading to lifestyle changes (physical activity, weight loss, reduction of smoking), increased awareness of their disease and associated comorbidities, and new diagnoses of cardiovascular risk factors or therapeutic adjustments. Repeated educational sessions with the same group of patients might further improve these positive results.

Some limitations of this study must, however, be mentioned, mainly the limited number of participants and a potential response bias, either intentional or due to comprehension factors, associated with self-reported surveys.

In brief, this educational intervention proved to be a useful tool that had significant repercussions on patients’ health, life, and ability to cope with the disease, and it should therefore be repeated with a larger number of patients.

### Table 1
Characteristics of 53 patients who completed full questionnaires before and after a psoriasis education session.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Before session</th>
<th>Immediately after session</th>
<th>Improvement</th>
<th>6 Months after session</th>
<th>Effect size</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, mean (SD), y</td>
<td>51.3 (11.2)</td>
<td>50.9 (27)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sex, No. (%) of patients</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Level of education, No. (%) of patients</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No studies</td>
<td>1.9 (1)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Basic education (4th grade)</td>
<td>37.7 (20)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Basic education (9th grade)</td>
<td>17 (9)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Full secondary education</td>
<td>22.6 (12)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Third-level education</td>
<td>20.8 (11)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Body mass index, mean (SD)</td>
<td>26.7 (4.4)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Comorbidities/history</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hypertension</td>
<td>35.8 (19)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dyslipidemia</td>
<td>35.8 (19)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Diabetes mellitus</td>
<td>13.2 (7)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Obesity</td>
<td>20.8 (11)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Smoking</td>
<td>18.9 (10)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Family history of cardiovascular disease</td>
<td>15.1 (8)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Family history of psoriasis, No. (%) of patients</td>
<td>49.1 (26)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Disease duration, mean (SD), y</td>
<td>24.6 (13.2)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Psoriatic arthritis, No. (%) of patients</td>
<td>60.4 (32)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Current treatment, No. (%) of patients</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>1.9 (1)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Topical</td>
<td>18.9 (10)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Phototherapy</td>
<td>7.5 (4)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Systemic</td>
<td>26.4 (14)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Biologic</td>
<td>39.6 (21)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Biologic + systemic</td>
<td>5.7 (3)</td>
<td></td>
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</tr>
</tbody>
</table>

### Table 2
Improvements in awareness of association between psoriasis and cardiometabolic comorbidities/cardiovascular disease. a

<table>
<thead>
<tr>
<th>Disease</th>
<th>Before session</th>
<th>Immediately after session</th>
<th>Improvement</th>
<th>6 Months after session</th>
<th>Effect size</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertension</td>
<td>50.9</td>
<td>98.1</td>
<td>&lt;.001</td>
<td>84.9</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Dyslipidemia</td>
<td>47.2</td>
<td>98.1</td>
<td>&lt;.001</td>
<td>81.1</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Diabetes mellitus</td>
<td>39.6</td>
<td>94.3</td>
<td>&lt;.001</td>
<td>75.5</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Obesity</td>
<td>39.6</td>
<td>92.5</td>
<td>&lt;.001</td>
<td>66</td>
<td>.004</td>
</tr>
<tr>
<td>Acute myocardial infarction</td>
<td>32.1</td>
<td>92.5</td>
<td>&lt;.001</td>
<td>79.2</td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Stroke</td>
<td>30.2</td>
<td>88.7</td>
<td>&lt;.001</td>
<td>73.6</td>
<td>&lt;.001</td>
</tr>
</tbody>
</table>

a Results shown as the percentage of questionnaire respondents who were aware of the association between psoriasis and the comorbidities shown.

b Improvement immediately after the session.

c Maintenance of improvement 6 months after the session (McNemar test).
Conflict of interest

The authors declare no conflict of interest.

References


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A case of Birt-Hogg-Dubé syndrome presenting with a single pedunculated fibrofolliculoma and a novel FLCN gene mutation

Un caso de síndrome de Birt-Hogg-Dubé presentando con un único fibrofolliculoma pediculado y una novedosa mutación en el gen FLCN

To the Editor:

In 1997, Birt, Hogg, and Dubé described multiple firm papules on the scalp, forehead, face, and neck in 15 persons belonging to a family of 70 individuals spanning 3 generations.  

The skin lesions were classified as 3 benign variants of hair follicle hamartomas: fibrofolliculomas, trichodiscomas, and acrochordons. This triad was later designated as the Birt-Hogg-Dubé syndrome (BHDS) (Online Mendelian Inheritance in Man catalog number, #135150).

A number of cases of BHDS have been identified since the original description was published, and significant associations have been reported, particularly with multiple lung cysts and renal cancer. A study in 2002 found that patients with BHDS have a 9.3-fold risk of developing renal tumors and a 32.3-fold risk of spontaneous pneumothorax.

Case description

A 28-year-old man presented to our department with a 1-year history of skin lesions on the nose. The patient’s past medical history was unremarkable, except for 2 spontaneous pneumothoraces at the age of 19 and 23 years. The patient also mentioned that his mother underwent right nephrectomy for a renal tumor at the age of 38 years.

Physical examination revealed 2 wine-colored, pedunculated papules, 1 measuring 3 mm on the right side of the columella, the other measuring 2 mm on the left alar rim (Fig. 1A and B). No other significant alterations were observed on examination.

Both lesions were excised. Histological examination of the papule from the left alar rim was compatible with a vascular hamartoma. The lesion from the columella showed features compatible with a fibrofolliculoma (Fig. 2A and B).

Sequencing of the coding exons (exons 4–14) and the intron-exon boundaries of the FLCN gene revealed a heterozygous FLCN:c.50G>C missense variant (p.Arg17Pro) in exon 4, both in the patient and in his mother. Numerous small cysts were observed in the basal regions of both lungs on computed tomography (CT) of the chest (Fig. 1C).

Applying the diagnostic criteria of the European BHD Consortium (Table 1), we made a diagnosis of BHDS based on 1 major criterion (an FLCN germline mutation) and 2 minor criteria (multiple lung cysts and a first-degree relative with BHDS).

Currently, the patient remains under regular follow-up because of the increased risk of developing renal cancer. Additionally, the patient’s first-degree relatives have been referred for gene analysis.

BHDS is an inherited autosomal dominant disorder caused by germline mutations of the folliculin (FLCN) gene located on chromosome 17(17p11.2). BHDS-associated renal tumors display inactivation of the wild-type FLCN allele (for example, loss of heterozygosity, mutation, methylation), confirming that FLCN is a tumor suppressor gene that fits the classic 2-hit model. This gene encodes folliculin, a 579 amino acid protein expressed in a variety of tissues