

Table I. Clinical and histopathologic data of the analyzed population

Data*	Sample number
Patients	(n = 11)
Man	7 (64)
Woman	4 (36)
Age at diagnosis	70.6 ± 12
Primary ALM location	
Heel	4 (36.4)
Base of the toe	2 (18.2)
Nail bed (foot)	2 (18.2)
Nail bed (hand)	1 (9.1)
Thenar eminence (hand)	1 (9.1)
Sole	1 (9.1)
Patients with local recurrences	2 (18.2) [†]
Recurrence in the form of satellitosis	2 (18.2)
Histopathologic samples	(n = 20)
Breslow depth, mm	1.9 ± 1.6
Syringotropic Breslow depth, mm	3.2 ± 1.7
Clark level	
I	4 (20)
II	4 (20)
III	5 (25)
IV-V	7 (35)
Acrosyringium involvement	20 (100)
Glandular unit involvement	12 (60)
Duct involvement	11
Secretory coil portion involvement	0
Duct and secretory coil portion	1
Formation of eccrine melanoma nests	11 (55)
Diameter of nests, mm	0.3 ± 0.1
Microsatellitosis	2 (10)

*Continuous data are presented as the mean ± SD and categorical data as number (%).

[†]Patients experienced 2 and 5 local recurrences, respectively.

also for early human melanoma precursors.³ Zembowicz et al⁴ reported 7 syringotropic melanomas, 1 of which was an ALM. These authors suggested that melanoma spreading within the eccrine apparatus can infiltrate deeper into the reticular dermis or subcutis than conventional melanomas.⁴ Other authors have reported several cases of syringotropic ALM.⁵

Studies and reports published so far do not differentiate between the involvement of the secretory and the ductal coil portions within the glandular coil. In our series, invasion of the glandular coil seems to predict a poorer prognosis, with advanced disease development being more frequent than in cases not involving the glandular coil. In addition, the ductal component is predominantly affected, whereas the secretory coil is mostly spared. One explanation for this is that myoepithelial cells could

act as a barrier, hindering the infiltration of melanoma cells into these secretory areas.

Moreover, we believe the syringotropic Breslow depth should be taken into account in clinical practice, because in our series it was also associated with advanced disease. Further, the syringotropic Breslow depth in 5 of the 20 samples would modify the T category from T1 to T2, thus changing therapeutic management from a wide local excision to a wide local excision with sentinel lymph node biopsy.

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Optimizing presentation of expected treatment outcomes



To the Editor: Patients struggle with starting medications for various reasons, including inadequately understanding the direct personal benefits of

treatment.^{1,2} Common marketing techniques in advertising emphasize attractive imagery, rather than informative data, to convince consumers to buy their product.^{3,4} We hypothesized that physicians could use similar techniques to enhance treatment adherence by creating an emotional connection between the patient and the medication. We compared willingness to take treatment after being presented with a personally relevant treatment goal versus after clinical data (or both) in people with acne, atopic dermatitis, and psoriasis.

After Wake Forest School of Medicine Institutional Review Board approval (#IRB00057322), a randomized online survey study was performed in eligible individuals with a working knowledge of English, aged ≥ 18 years, and a self-reported diagnosis of acne, atopic dermatitis, or psoriasis. There were 305 individuals recruited through Amazon Mechanical Turk (Seattle, WA).⁵ Participants received a fact sheet and completed the survey in Qualtrics (Provo, UT) and were compensated \$0.05 for completing the survey.

Participants were randomized in a 1:1:1 double-blind, placebo-controlled ratio to assess their willingness to take treatment for acne, atopic dermatitis, or psoriasis if presented with one of the following: (1) clinical data, (2) a personally relevant treatment goal, or (3) both clinical data and a personally relevant treatment goal (Table I). Demographic information was also collected. Outcome measures were recorded as willingness to take treatment on a 10-point Likert scale. Groups were compared using one-way analysis of variance and 2-group *t* tests.

Compared with respondents presented with only clinical data (mean, 7.4; SD, 4.1), participants presented with a personally relevant treatment goal reported greater willingness to take treatment (mean, 8.5; SD, 3.1; $P = .0001$); participants presented with both a personally relevant treatment goal and clinical data also reported greater willingness to take treatment (mean, 8.1; SD, 2.9; $P = .02$) compared with clinical data alone. More participants were nearly completely or completely willing to take treatment (score of 9 or 10) in the personally relevant treatment goal group (60%) than in the clinical data group (30%) or the clinical data with a personally relevant treatment goal group (44%, $P = .005$) (Fig 1).

Presenting a medication with a personally relevant treatment goal increases reported willingness to take treatment for patients with acne, atopic dermatitis, or psoriasis. This might be because real-life concrete treatment goals provide patients with a sense of a direct personal benefit, which they can better appreciate than facts and statistics alone. Thus,

Table I. Survey questions for presentation of treatment for acne, atopic dermatitis, and psoriasis*

Question 1 (clinical data)
Your doctor presents a medication to you by saying the following, "This treatment has a 60% chance of nearly completely alleviating your skin disease (acne, eczema, psoriasis)."
How willing would you be, on a 1 (not willing) to 10 (completely willing) scale, to take this medication?

Question 2 (treatment goal)
Your doctor presents a medication to you by saying the following, "This treatment is most likely going to clear your skin disease (acne, eczema, psoriasis) to the point you are comfortable enough to wear your favorite swimsuit to the pool this month."
How willing would you be, on a 1 (not willing) to 10 (completely willing) scale, to take this medication?

Question 3 (clinical data with a treatment goal)
Your doctor presents a medication to you by saying the following, "This treatment has a 60% chance of nearly completely alleviating your skin disease (acne, eczema, psoriasis), and there is a similar chance it would help you feel comfortable enough to wear your favorite swimsuit to the pool this month."
How willing would you be, on a 1 (not willing) to 10 (completely willing) scale, to take this medication?

*Responses were recorded on a 1 (not willing) to 10 (completely willing) scale.

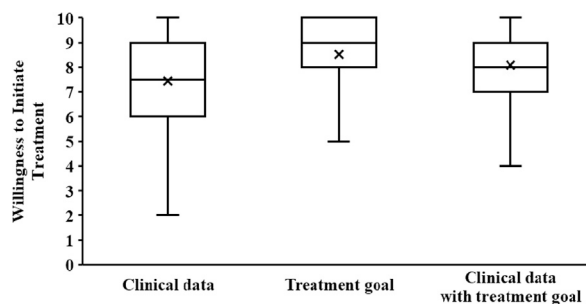


Fig 1. Presenting treatment with a personally relevant treatment goal improves individuals' willingness to initiate treatment. Respondents' mean willingness scores on a 10-point Likert scale to initiate treatment in the clinical data, treatment goal, and clinical data with treatment goal groups were 7.4, 8.5, and 8.1, respectively; median scores were 7.5, 9.0, and 8.0, respectively. Boxes depict 25th and 75th quartiles, and whiskers indicate maximum and minimum scores. The horizontal line in each box indicates median scores, and the "X" in each box indicates the mean score.

providing patients with a personally relevant treatment goal may be a simple and cost-effective technique to improve patient outcomes. Presenting

a personally relevant treatment goal alongside clinical data did not have as great of an effect as presenting a treatment goal alone. We cannot, however, conclude that we should not give patients clinical data. The potential benefit of improving adherence must be balanced with a patient's right to be informed of all pertinent information.

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Lack of genotype-phenotype correlation in basal cell nevus syndrome: A Dutch multicenter retrospective cohort study

To the Editor: Basal cell nevus syndrome (BCNS), is a rare autosomal dominant disorder (Mendelian Inheritance in Man no. 109400). The most common underlying cause is a heterozygous germline mutation in the Patched-1 (*PTCH1*) gene (chromosome 9q22-31).¹ The mutation detection rate of *PTCH1* in BCNS in clinically confirmed individuals is 40% to 85%.² Diagnosis requires 2 major criteria, 1 major criterion and 2 minor criteria, or 1 major criterion and molecular confirmation.³ To date, no clear genotype-phenotype correlation has been detected. In this study, 83 Dutch patients with a confirmed *PTCH1* mutation were analyzed.

A multicenter retrospective cohort study was conducted at the Maastricht University Medical Center and the VU University Medical Center in the Netherlands. From 1999 until 2015, *PTCH1* variant analysis was performed in 673 patients. In 141 (21.0%) individuals, a *PTCH1* variant was detected.⁴ After applications from hospitals outside the Netherlands and nonpathogenic variants were excluded, a pathogenic *PTCH1* mutation was detected in 95 patients. Clinical data could be obtained for 83 patients (37 male, 46 female) from 77 families (mean age, 25.2 years).

Two heat maps were created to investigate a genotype-phenotype correlation. To create robust heat maps, only patients with 2 or more symptoms were included. In the first heat map, the mutations were sorted according to their location within the gene, plotted against 6 clinical symptoms (Fig 1). In the second heat map, the mutations were sorted according to the type of mutation (truncated or missense) against the clinical symptoms (Fig 2). Other symptoms were excluded because specific data were missing for many cases. No specific clustering of symptoms in relation to mutations in a certain exon or part of the *PTCH1* gene could be observed, nor could phenotype similarities be found within families in our population. In accordance with Evans et al,⁵ we found that patients with a missense variant in *PTCH1* were diagnosed later with BCNS, were less likely to develop rib anomalies, and had their first basal cell carcinoma at a 3-year older age