surveillance problems and nutritional deficiencies caused by the disease may play a role.

Marjolin ulcer is a rare aggressive cutaneous malignancy with a high rate of regional metastases that arises on previously traumatized or chronically inflamed skin, most commonly in the scalp or extremities, usually a highly aggressive squamous cell carcinoma (SCC) secondary to chronic burn wounds. However, although rare, Marjolin ulcers containing BCC and baso-SCC have also been reported.[7] Interestingly, a Marjolin ulcer developed in response to chronic allergic contact dermatitis caused by an orthopaedic implant, suggesting that prolonged allergic dermatitis can promote the development of skin cancer.

As far as we are aware, no study has reported an association between CD and Marjolin ulcer. We report three cases of BCC located in the perineum of patients with familial seronegative CD, suggesting that CD may predispose patients to developing skin malignancies in this region, possibly due to chronic exposure of perineal skin to inflammatory cytokines or other irritants or antigens/triggers from the GI tract.

Case 1, a 63-year-old white man who was obese with chronic congestive heart failure presented with a 14-cm mass in his perineum which had appeared 10 years earlier as a pinpoint nodule and gradually enlarged. A biopsy of the lesion revealed nodular BCC. The patient had also experienced marked GI distress for the past two decades, which worsened when he travelled away from home. Notably, his favourite dessert at home was gluten-free tapioca pudding. His HLA type, which was previously obtained for an unrelated reason, was HLA-DQ2. A diagnosis of CD was made based on a GI biopsy of the ileum. The patient died of heart failure before receiving this diagnosis. Although the patient was never formally diagnosed with seronegative CD, family members also affected by the disease were negative for circulating antitissue transglutaminase antibodies. Interestingly, the age of onset decreased with successive generations.

Case 2, a 54-year-old woman with a 12-year history of familial seronegative CD that went undiagnosed for 11 years, presented with a 4-cm mass in her intergluteal region present for 4 years. She had hypertension but no other known comorbidity. A diagnosis of nodular BCC with neighbouring ulcer was made. The patient declined GI biopsy. Her HLA type was HLA-DQ2.

Case 3, a 58-year-old white man with a 10-year history of familial seronegative CD that went undiagnosed for 9 years, presented with a 6-cm lesion in his perineum that had gradually enlarged over the previous 5 years. A diagnosis of superficial disseminating BCC with reactive changes was made. His HLA type was found to be HLA-DQ2. Notably, testing for CD was undertaken only following the suggestion by one of us (W.C.L.) based on experience (patients 1 and 2). He had no other known comorbidity.

These associations may, of course, have been due to chance. However, we suggest that, based on our findings, physicians should consider CD or other inflammatory bowel disease in patients with skin cancer located in or near the perineum.

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Funding sources: none.

Conflicts of interest: the authors declare they have no conflicts of interest.

Data availability statement: Data available on request due to privacy/ethical restrictions.

Optimizing shared decision making in older adults with basal cell carcinoma: experiences from a specialized outpatient clinic

DOI: 10.1111/bjd.20833

Dear Editor, Balancing the risk for under- and overtreatment in older adults with basal cell carcinoma (BCC) frequently proves challenging.[1,2] As BCCs are slowly growing tumours and initially asymptomatic, patients with limited life expectancy (LLE) might frequently be overtreated,[2] leading to unnecessary and avoidable treatment burden, while little or no improvement in quality of life might be achieved. To improve patient-centred care for older patients with BCC, it was hypothesized that a more holistic approach, with specific attention to patient-related factors and treatment goals, would lead to different BCC management choices, better aligning with patient preferences and resulting in less overtreatment.

A prospective, observational pilot study was conducted at the Radboud University Medical Center (the Netherlands), evaluating BCC management in a specialized outpatient clinic for (frail) older adults. General practitioners and dermatologists were asked to refer patients where there was a dilemma regarding optimal BCC management (e.g. frail patients with LLE). Longer time slots were available to provide more time for evaluation of patient-related factors [e.g. frailty, including the 'Geriatric-8' (G8) frailty screening tool],[3,4] treatment goals and a multidisciplinary approach (e.g. with radiotherapists, plastic surgeons, primary care physicians or nursing home specialists).

A standardized shared decision-making model was used,[5] in which physicians and patients discussed their medical and personal preferences. All possible BCC management options were discussed, including the risks (e.g. complications, recurrence risk, treatment burden) and benefits (e.g. prevention of progression, symptom relief). Both active BCC therapies (including management regimens deviating from guidelines) and watchful waiting or active surveillance (no active BCC treatment) were discussed.[2] BCC management decisions were made by patients and/or their legal representatives after careful consideration of potential risks and benefits. Follow-up was carried out at least once, if feasible for the patients. The outcomes were compared with the expected regular practice management (ERPM) for each BCC, according to current guideline recommendations.[6] χ^2-tests were used to compare groups; missing values were excluded from analyses.

Between January 2018 and December 2018, 85 patients were seen with a mean age of 86·4 (SD 5·8) years. After initial consultation, 59 patients (69%), 26 (44%) men and 33 (56%) women, were diagnosed with 125 BCCs (116 histologically confirmed). Of the 55 patients with available data, 23 (42%) had asymptomatic BCCs, while 48 (87%) were classified as '(potentially) frail' (G8 score $\leq$ 14). Treatment goals and preferences other than curative treatment were mentioned by 37 (63%) patients (e.g. symptom relief, least burdensome treatment, cosmetic goals, no treatment). In 24 (41%) patients with 58 (46%) BCCs, BCC management differed from ERPM; these included 21 (44% differed) frail patients and two (29% differed) robust patients ($P = 0{\cdot}69$; one missing). During a median follow-up of 29·5 months (interquartile range 11–33·5), ERPM was initiated after all for 16 (28%) BCCs (Figure 1).

No significant differences were seen between the BCC subtypes (micronodular/infiltrative vs. superficial/nodular) and whether or not ERPM was initiated after all ($P = 0{\cdot}36$). One patient with one BCC (2%) needed more extensive surgery than the estimated treatment at first presentation. Overall, in 19 (32%) patients with 37 (30%) BCCs, it was estimated that fewer hospital visits were needed than after ERPM. Most commonly this was due to treatment directly after consultation, a shortened treatment schedule or multidisciplinary consultations during one hospital visit. In eight (14%) patients with 29 (23%) BCCs it was estimated that more hospital visits were needed than after ERPM. During follow-up, 21 (36%) patients

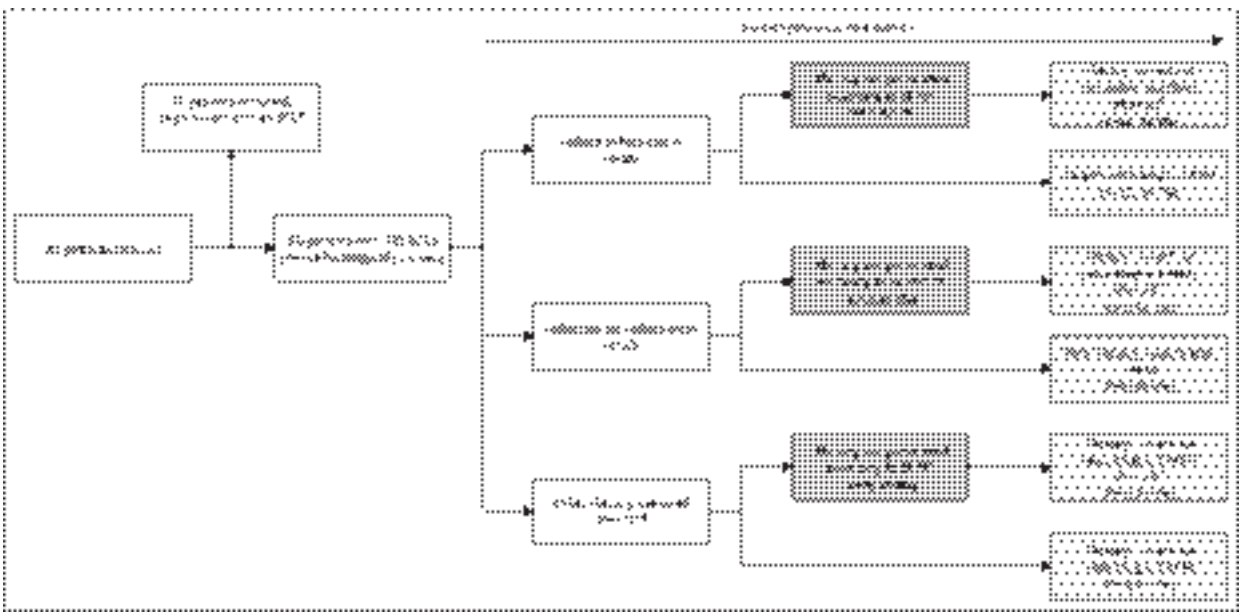

Figure 1 Treatment choices after shared decision making in a specialized outpatient clinic for older adults with basal cell carcinoma (BCC). n represents the number of BCCs. All possible treatment options, including watchful waiting and those with higher recurrence risks (e.g. electrodissection, curettage, cryotherapy), were discussed with the older adults referred to this specialized outpatient clinic for geriatric patients with BCC. In case of multiple treatment options with roughly equal recurrence risk, patients and tumours were classified as 'have received treatment according to the expected regular practice management (ERPM)'. *Diagnoses other than BCC included squamous cell carcinoma, actinic keratosis or eczema. **Other therapies indicated, for instance imiquimod or fluorouracil; note that no tumours in this category were excised. ***A shortened radiotherapy course was considered not according to ERPM. #In case of recurrences or reconsideration of previously made management decisions.

died [after a mean 14·1 (SD 9·2) months, none BCC related], 10 (48%) of whom were not treated with ERPM.

These experiences indicate that adapted management regimens after thorough shared decision making can be less burdensome and provide more appropriate care for a substantial proportion of frail patients with BCC. Our experiences are in line with those seen in other medical fields, where geriatric assessments led to less intensive treatment in more than one-third of geriatric patients.[7] Individual treatment goals other than curative treatment were highly relevant among frail older adults, considering that all management options were advised, which might include deviation from clinical guidelines. A decision aid can be considered to ensure feasibility in daily care.[8] Incorporation of integrated, holistic care is not always easy in daily practice, although the use of predictive instruments (e.g. the G8 or other frailty screening tools)[4] might assist in identifying patients in need of multidisciplinary approaches and/or more extensive counselling around BCC management.

Limitations of this study are the small population and its observational design. Therefore, more research on these tools and the clinical consequences (e.g. mortality or complications) is needed. In case of higher mortality rates, the time to benefit from BCC treatment might exceed life expectancy, and frail patients might consequently benefit from active surveillance or symptomatic treatment. However, medical decision making based solely on age should be avoided, as the heterogeneity in functional status, resilience and frailty at the same high ages underscore the need for an individualized approach.

Acknowledgments: we would like to thank all of the physicians, patients and proxies who were involved in this project.

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Funding sources: this pilot study was conducted with financial support from the health insurance company VGZ within the project 'Betaalbaar Beter', a project striving to achieve affordable and better healthcare with high patient value. The funding source had no influence on the medical choices, data collection or analysis, or contents of the manuscript.

Conflicts of interest: the authors declare they have no conflicts of interest.