

**Background and Importance** Epidermolytic ichthyosis (EI) is a skin genetic disorder that predominantly affects joints and friction areas with a limited number of therapeutic options (topical and systemic treatments). In some severe EI subtypes, the off-label use of ustekinumab is based on marked elevation of cytokines in the Th17/IL-23 pathway, similar to inflammatory diseases like psoriasis.

**Aim and Objectives** To evaluate the efficacy and security of ustekinumab in an 8-year-old female patient with severe EI with KRT1 mutation, refractory to topical treatment and oral retinoids.

**Material and Methods** Firstly, she underwent keratolytics and emollients with scarce clinical response. Due to the absence of effectiveness of these topical therapies, she received oral retinoids (acitretin) up to maximum tolerated dose, but some erythematous lesions and hyperkeratosis rapidly appeared on her skin. For this reason, the treatment had to be discontinued several times. In April 2022, Dermatology Service requested the off-label use of ustekinumab with a dosage of 0.75 mg/kg at weeks 0, 4, 8 and 12, and then administered each 12 weeks to the Pharmacy therapeutics committee.

Afterwards, an extensive review was carried out by Pharmacy Service. A report was made with a positive assessment for approval of treatment. This decision was supported by some case reports showing clinical results of ustekinumab in some EI subtypes and the lack of available alternative therapies in this case.

**Results** At the beginning of treatment with ustekinumab, extensive scaly erythematous lesions with circinate margins were observed, affecting the facial area, trunk and extremities, accompanied by diffuse palmoplantar keratoderma.

After three months of the first administration of ustekinumab, the patient was examined by a dermatologist. An excellent clinical response was observed with resolution of the facial lesions and almost complete on the trunk, with hyperkeratotic lesions persisting in folds, without underlying erythema. Moreover, no adverse events related to ustekinumab were registered.

**Conclusion and Relevance** Ustekinumab is suggested to be an alternative therapy in some severe EI subtypes refractory to topical and systemic treatments. In spite of being safe and effective in this patient, longer studies are needed to consider ustekinumab in the therapeutic management of EI.

#### REFERENCES AND/OR ACKNOWLEDGEMENTS

**Conflict of Interest** No conflict of interest.

#### 6ER-028 ASSESSMENT OF SYMPTOMS AND SIGNS SEVERITY IN PSORIASIS PATIENTS

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**Background and Importance** Psoriasis is a chronic inflammatory skin disease in which moderate to severe forms can be treated with monoclonal antibodies (mAbs). Assessment of disease improvement is usually made by the Psoriasis Area Severity Index (PASI) and Body Surface Area (BSA). However, sometimes patients' feelings do not correlate with these scores.

**Aim and Objectives** To describe the symptoms and signs in patients with moderate-to-severe psoriasis by using Psoriasis Symptoms and Signs Diary (PSSD).

**Material and Methods** Prospective observational study conducted between 1-February-2022 and 30-September-2022 in a university hospital. Patients treated at least 3 months with mAbs were included. Data collected: age, sex, diagnostic, mAbs prescribed, previous treatments, PASI and BSA scores. PSSD was used to measure patient-reported outcome (PRO). It assesses the severity of psoriasis symptoms (itching, tightness, burning, pain) and signs (bleeding, cracking, dryness, scaling, shedding, redness) using a 0–10 numerical rating scale. Summary scores were derived using a scale of 0–100. Patients with a PSSD score  $\geq 20$  were referred to the dermatology service to assess the mAb switching. Data were obtained from electronic medical records and patients' interviews.

**Results** Thirty-eight patients completed the PSSD (50% women) with a median age 51.3 (37.8–61.4) years, 84.2% with psoriasis and 15.8% also with psoriatic arthritis (PA) as comorbidity. mAbs prescribed: adalimumab (44.7%), ustekinumab (13.1%), guselkumab (10.5%), tildrakizumab (10.5%), risankizumab (7.9%), secukinumab (5.3%), brodalumab (5.3%), ixekizumab (2.7%). Twenty-four patients (63.2%) received mAbs as the first line, 21.0% as the second-line and 13.1% as the third or more lines. Eight patients had PASI  $> 2$ . The PSSD average score was: itching  $1.9 \pm 2.9$ , dryness  $3.5 \pm 2.8$ , cracking  $1.2 \pm 2.4$ , tightness  $1.4 \pm 2.2$ , scaling  $1.2 \pm 2.2$ , shedding  $2 \pm 2.9$ , redness  $2.3 \pm 2.7$ , bleeding  $0.5 \pm 1.5$ , burning  $1.2 \pm .2$ , pain  $0.5 \pm 1.6$ . Dryness was the highest rated and bleeding the lowest score. Twelve patients (31.6%) had a PSSD score of  $\geq 20$  and the main treatment was adalimumab (41.6%). Three patients switched the mAb. Only in five patients the PASI and BSA scores were correlated with PSSD.

**Conclusion and Relevance** PSSD is a reliable and valid PRO instrument for assessing psoriasis-associated symptoms and signs in patients treated with mAbs in clinical practice. This score, together with PASI and BSA, could be used to guide mAbs switching.

#### REFERENCES AND/OR ACKNOWLEDGEMENTS

**Conflict of Interest** No conflict of interest.

#### 6ER-030 USING A PHARMACIST-LED ASTHMA SERVICE TO ASSESS THE CONCORDANCE BETWEEN PATIENT-REPORTED ICS ADHERENCE AND OBJECTIVE E-MONITORING OF ICS THERAPY

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**Background and Importance** The prevalence of asthma and scale of sub-optimal inhaled corticosteroid (ICS) use, demands efficient detection of non-adherence. The easy to administer Test of Adherence to Inhalers<sup>1</sup> (TAI) questionnaire asks patients to rate their agreement with 10-items, and the subsequent score classifies adherence as good, intermediate or poor. A more objective, though expensive tool, is the electronic monitor (eMonitor) that when attached to the inhaler, records the date/time of each actuation. If the person receives  $> 75\%$  of doses, this is good adherence. The biomarker Fractionated expired Nitric Oxide (FeNO) decreases following sustained ICS use. Thus, if eMonitor ICS adherence is good and there is a significant decrease in FeNO ( $> 42\%$  from baseline), pre-