17744

Bart syndrome: Case report and systematic review of the literature

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Background: Bart syndrome was initially described as congenital absence of skin (CAS), nail abnormalities, and epidermolysis bullosa (EB). Further reports of patients with CAS and EB have been made with wide clinical heterogeneity among them. Current guidelines recommend the elimination of eponyms and use of the descriptive term EB with CAS.

Methods: We performed a PubMed and Medline database search of patients with Bart syndrome or EB with CAS. We included case reports or case series that contained clinical and demographic information.

Results: After review, we included 55 articles, reporting 96 patients. The CAS involved the lower extremities in all patients, with additional upper limb, trunk or head involvement in 17%. In all patients, healing was achieved between 2 weeks and 6 months, most received only conservative treatment. Regarding the subtype of EB, 73% of patients had dystrophic EB, 21% had junctional EB and 6% had EB simplex. Other extra-cutaneous features were present in 29 patients; where pyloric atresia and ear malformations were the most common. The prognosis seems to depend on the subtype of EB and the presence of additional comorbidities; 80% of the patients with junctional EB and CAS died during the first months of life, while in those with dystrophic EB, only 4% died.

Conclusions: EB with CAS is a clinically heterogeneous disorder. It is mainly associated to DEB but other subtypes are also related. Further information is required to better establish a pathologic mechanism for CAS, and its association to EB.

Commercial disclosure: None identified.

17749

Investigating the relationship between rosacea and use of vasodilatory medications in a hospital-wide population

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Frequent and intense flushing is known to play a role in the pathogenesis of rosacea. Though a common side effect of antihypertensive medications includes flushing, to date the association between the effects of antihypertensive medications on peripheral vessels and rosacea is not well understood. A retrospective cohort study of data on patients seen at the University of Iowa Hospitals and Clinics with a diagnosis of hypertension and/or a diagnosis of rosacea was undertaken. Out of a total of 162,480 patients diagnosed with hypertension, 2200 patients also had a known diagnosis of rosacea. From these 2200 with rosacea, it was found that 1750 (79.5%) had received this diagnosis within 6 years of starting vasodilator therapy for hypertension. For the purpose of this study, vasodilator therapies included calcium channel blockers, beta-blockers, alpha-blockers, ACE inhibitors, and other miscellaneous drugs with vasodilatory effects. Further analysis showed that individuals taking vasodilator medications had a significantly increased likelihood of developing rosacea over a 1-year period (RR 1.25, 95% CI 1.2-1.29, $P < .0001$), and an even higher risk over a 6-year period (RR 1.41, 95% CI 1.37-1.46, $P < .0001$). Understanding this relationship may help in guiding antihypertensive selection via risk stratification for dermatologic complications.

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17753

Analysis of 24-week response to ruxolitinib cream for the treatment of vitiligo based on patient demographics and clinical characteristics

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Ruxolitinib cream, a Janus kinase inhibitor, is under investigation for vitiligo treatment. Factors including skin type and disease duration may contribute to treatment efficacy. This 24-week component of a 104-week, phase 2, randomized, double-blind study (NCT03099304) enrolled adult patients with vitiligo that included depigmentation ≥0.5% of body surface area (BSA) on the face and ≥3% of BSA on nonfacial areas. 157 patients were equally randomized to receive ruxolitinib cream (1.5% twice daily [bid], 1.5% once daily [QD], 0.5% QD, or 0.15% QD) or vehicle bid for 24 weeks. The primary end point was the proportion of patients achieving ≥50% improvement in facial Vitiligo Area Scoring Index (F-VASI50) at week 24. This subgroup analysis investigated response by patient demographics and baseline characteristics; results were generally similar across treatment groups at week 24. Among patients who received ruxolitinib cream 1.5% BID (n = 33; F-VASI50 responders, 45.5%), a larger proportion of patients in the following subgroups were F-VASI50 responders: patients ≤50 years old (58.8%); female patients (60.0%); patients with skin type I–III (50.0%), ≤1.5% affected baseline facial, BSA (52.6%), baseline F-VASI scores of 0.75 to 20 years (60.0%); and previous recipients of topical corticosteroids (50.0%). There were no substantial differences between responders who were white (44.8%) vs nonwhite (50.0%), who had stable (46.2%) vs progressive disease (45.0%), or those with total, BSA ≤20% (45.0%) vs >20% (46.2%). Ruxolitinib cream was effective for the treatment of vitiligo across demographics and clinical characteristics, including in patients with longstanding and extensive disease.

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17850

Predicting the long-term outcomes of biologics in psoriasis patients using machine learning

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Background: Real-world data demonstrate that ~50% of psoriasis patients treated with a biologic agent will discontinue the drug because of a loss in efficacy. History of previous therapy with another biologic, female sex, and obesity were identified as predictors of drug discontinuations, but their individual predictive value is low.

Objective: To determine whether machine learning algorithms can produce models that can accurately predict outcomes of biologic therapy in psoriasis on an individual patient level.

Results: All tested machine learning algorithms could accurately predict the risk of drug discontinuation and its cause (eg, lack of efficacy vs adverse event). The learned generalized linear model achieved a diagnostic accuracy of 82%, requiring under 2 seconds per patient using the psoriasis patients dataset. Input optimization analysis established a profile of a patient who has best chances of long-term treatment success: biologic-naive patient under 49 years, early-onset plaque psoriasis without psoriatic arthritis, weight <100 kg, and moderate to severe psoriasis activity (DLQI ≥16; PASI ≥10). Moreover, a different generalized linear model is used to predict the length of treatment for each patient with mean absolute error (MAE) of 4.5 months. However, Pearson correlation coefficient indicates 0.935 linear dependencies between the actual treatment lengths and predicted ones.

Conclusions: Machine learning algorithms predict the risk of drug discontinuation and treatment duration with accuracy exceeding 80%, based on a small set of predictive variables. This approach can be used as a decision making tool, communicating expected outcomes to the patient, and development of evidence-based guidelines.

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