Scope of managements of Saudi patients with interleukin-36 receptor antagonist deficiency (DITRA): Case series from a tertiary care hospital.

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Introduction: Monogenic form of Generalized Pustular Psoriasis presents in childhood with variable degrees of severity that might leads to life threatening presentation. This disorder is due to loss of function mutation in *IL-36RN* gene leading to recurrent episodes of fever, generalized erythematous skin, sterile pustular eruptions, hemodynamic instability, leukocytosis & elevated inflammatory markers.

Methods: We recruited four patients with Interleukin-36 receptor antagonist deficiency who were managed at a tertiary care Hospital in Riyadh, Saudi Arabia. The diagnosis was based on Clinical presentation, skin biopsy finding and genetic confirmation. In this report we provide description of their clinical presentation, laboratory investigations, and treatment modalities as well as outcome.

Results: Four patients presented in childhood or early adolescent age with extensive pustular eruptions covering most of the body surface, erythroderma, fever , hemodynamic instability requring ICU admission high inflammatory markers and leucocytosis. Skin Biopsy showed psoriasiform epidermal hyperplasia with neutrophilic pararkeratosis and intradermal pustular formation. Multiple treatment modalites were tried including disease-modifying anti- rheumatic drugs, Anti-Interleukin 12/23, Calcineurin inhibitor. These medications were used alone or in combination with minimal or loss of response over time .Subsequently, we managed our patients

with Anti-tumor necrosis factor alpha monoclonal antibody (Adalimumab) that leads to excellent and sustainable response.

Conclusion:

DITRA continues to be a challenging disease especially in the aspect of managemnt but Anti-tumor necrosis factor Alpha is currently showing promising results that possibly would improve quality of life and decrease the burden of the disease.