aggressive intervention, about a quarter of patients continue to progress to end stage renal disease (ESRD).¹

There is considerable diversity in the definition of 'refractory lupus nephritis' depending on the treatment protocol, physician's subspecialty, and histopathologic findings. Molecular biomarkers further show potential as surrogates to kidney biopsies in predicting renal outcomes and long-term prognosis.² The factor of drug adherence, however, may draw the fine distinction between refractory (or resistant) and relapsing LN, with the latter substantially more common than the former.³ Nearly half of patients with proliferative LN who initially achieve a complete response to immunosuppressive therapy will have a relapse or renal flare following cessation or reduction of immunosuppression. Other risk factors for refractoriness include genetics and comorbidities like hypertension, diabetic nephropathy and antiphospholipid antibodies, each of which must be effectively addressed in the overall management of these patients.

Treatment options for refractory LN include switching or multitargeted therapy with immunosuppressives cyclophosphamide, mycophenolate derivatives and calcineurin inhibitors (cyclosporine A, tacrolimus, and recently voclosporin).⁴ Literature abounds with the use of rituximab in refractory LN including trials exploring the sequential use of rituximab plus cyclophosphamide followed by belimumab.⁵ Other modalities such as extracorporeal treatment (plasma exchange or immunoadsorption) and stem cell transplantation may be tried in special situations. Novel insights of LN pathogenesis have led to the development of new or re-purposed drugs including obinutuzumab, itolizumab, and igturatimod which can significantly prevent progression to ESRD. Finally, the timely, aggressive and highly individualised management of LN which reinforces strict patient adherence is key to enhanced outcomes.

Learning Objectives

- Define refractory and/or relapsing LN
- Identify risk factors for refractoriness among LN patients
- Describe the management approach to refractory LN
- Describe advances in drug development for LN

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REFRACTORY LUPUS CYTOPENIAS

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The aim of this presentation is to provide concise information regarding diagnosis and management of immune mediated hematological manifestations of systemic lupus erythematosus (SLE), specifically autoimmune hemolytic anemia (AIHA), immune mediated thrombocytopenia and immune mediated leukopenia.^{1–5}

Studies on hematological manifestations of SLE are scarce and with low quality of evidence, therefore most existent guidelines are supported by hematological data from non-SLE patients. Of note, SLE patients have some peculiarities that must be taken into consideration when choosing the best treatment approach for their hematological conditions.

The key points of each cytopenia are:

- a. Autoimmune hemolytic anemia (AHAI) in SLE is usually mediated by warm antibodies (IgG). First-line treatment is glucocorticoid, but there is no consensus to guide second-line therapy. Rituximab and immunosuppressive drugs can be used in refractory cases.
- b. Immune thrombocytopenia is an important prognostic indicator of survival in SLE patients. Treatment is usually indicated for patients with a platelet count $<30 \times 10^9/\text{mm}^3$. First-line therapy remains glucocorticoids (dexamethasone is the hematologists 1st choice), but sustained response after glucocorticoid discontinuation in SLE patients is unlikely to happen. Rituximab and immunosuppressive drugs can be used in refractory cases. Thrombopoietin agonists and splenectomy should be avoided due to high risk of infection and thrombosis
- c. Neutropenia may be severe and can lead to morbidity and mortality from increased susceptibility to infection. Severe neutropenia can be successfully treated with granulocyte colony-stimulating factor. Lymphopenia is most often related to drug toxicity and disease activity. Severe lymphopenia may require the use of prophylactic therapy to prevent opportunistic infections. For all cytopenias an extensive evaluation of other causes, particularly drugs and infection should be performed.

Learning Objectives

- Identify and manage immune mediated anemia in lupus
- Discuss diagnosis and treatment options for thrombocytopenia in lupus
- Explain the diagnosis and management of leukopenia in lupus

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REFRACTORY MUSCULOSKELETAL MANIFESTATIONS

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Musculoskeletal involvement is one of the most common clinical manifestations of systemic lupus erythematosus (SLE),