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Lupus Nephritis in Children

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Introduction: Childhood-onset systemic lupus erythematosus (cSLE) presents unique diagnostic and therapeutic challenges compared to its adult-onset counterpart. This review aims to elucidate the complexities of cSLE management, highlighting recent advancements and emerging therapeutic strategies.

Epidemiology and Clinical Characteristics: cSLE is a rare disease, with a prevalence of approximately 1:10,000, varying by ethnicity and more frequently observed in Asian, African-American, and Hispanic populations. Onset typically occurs before the age of 18, with an average age of onset around 12 years. Notably, cSLE exhibits higher disease activity and a greater burden of damage at diagnosis compared to adult-onset SLE. The gender distribution is skewed towards females, with a ratio of 4.5-5:1 (female to male).

Prognosis and Treatment Challenges: Untreated cSLE carries a grim prognosis, with a 5-year mortality rate of 95%. However, achieving satisfactory outcomes in cSLE extends beyond short-term survival, targeting a 50-60-year lifespan. Despite improvements, the 10-year survival rate remains at 81-92%, with up to 15% mortality observed between ages 22-27. Compliance with treatment regimens emerges as a critical determinant of outcomes, compounded by the psychological impact of chronic illness and medication side effects. Multidisciplinary family education and support are crucial components of holistic cSLE management.

Renal Involvement and Therapeutic Strategies: Renal involvement is common in cSLE, affecting 50-80% of patients. While renal outcomes have improved over time, achieving renal remission remains challenging. Kidney transplantation may be considered in refractory cases, with optimal timing and post-transplant management strategies evolving. Steroid-related damage underscores the importance of steroid-sparing strategies, particularly in childhood, to mitigate long-term consequences.

Emerging Therapeutic Modalities: Proposed treatment protocols align with published recommendations. Initial multitargeted therapy, including voclosporin or belimumab, is increasingly recommended as a standard treatment option. However, Mycophenolate Mofetil (MMF) and Cyclophosphamide (CPM) remain effective first-line treatments for childhood-onset lupus nephritis. Rituximab (RTX) is generally utilized for refractory cases, while additional treatments should be considered for coexisting thrombotic microangiopathy (TMA). Among various promising new treatments, CD19 CAR-T therapy holds significant promise as a potential paradigm-shifting treatment. Coexistence of serious systemic conditions like Macrophage Activation Syndrome (MAS) complicates renal treatment, posing a threat to life. Careful attention is crucial in such cases to mitigate risks and ensure appropriate management.