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Childhood Autoimmune Hemolytic Anemia: A Scoping Review

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Introduction
Autoimmune hemolytic anemia (AIHA) is a disorder characterized by excessive premature red blood cell breakdown due to the presence of autoantibodies. It is rare in children, with an estimated incidence of 0.2 per one million individuals younger than 20 years. There are no guidelines on the investigation and management of pediatric AIHA, and contemporary approaches are derived from adult guidelines. AIHA is an important cause of morbidity in pediatric hematology patients, and a review of pediatric AIHA investigation, diagnosis, and treatment is warranted. This scoping review will summarize the current landscape of diagnosis and management of pediatric AIHA to inform future studies aimed at formulating a pediatric specific approach to the investigation and management of this small but complex patient population.

Methods
This review searched MEDLINE, EMBASE, and the Cochrane Central Register of Controlled Trials (CENTRAL) from inception to July 27, 2021. All screening and data extraction was done in parallel by two reviewers. Experimental and observational studies reporting on diagnostic criteria, laboratory work up, or treatment/management of AIHA in populations with at least 20% of patients ≤18 years were included. Study characteristics, population characteristics, and study outcomes were extracted and synthesized narratively and descriptively using counts (percentages).

Results
After removing duplicates, the titles and abstracts of 1742 studies were screened and 109 studies were selected for full-text review. Forty three studies, published between 2003 and 2021, met inclusion criteria and proceeded to data extraction. Forty cohort studies (35 retrospective, 5 prospective) and 3 case-control studies were included. No randomized controlled trials were identified. Diagnostic criteria for AIHA was provided in 29 (67%) studies, with 4 (9%) studies classifying the severity of AIHA. All but one study defined AIHA with at least one of: positive direct antibody test, evidence of anemia, and evidence of hemolysis (including increased lactate dehydrogenase or bilirubin, or decreased haptoglobin). Patients with Evan's syndrome were included in 31 (72%) studies, and 30 (70%) studies included special populations such as transplant recipients or patients with underlying autoimmune conditions. AIHA treatments in pediatric patients were reported in 41 (95%) studies, with 26 (60%) studies dividing the treatments into first- and second-line therapies. Common first-line therapies included steroids (unspecified) in 18 (42%) studies, prednisone in 9 (21%) studies, methylprednisolone in 9 (21%) studies, intravenous immunoglobulin (IVIG) in 17 (40%) studies, and/or combinations of these therapies. Common second-line therapies included rituximab in 16 (37%) studies, cyclosporine in 16 (37%) studies, IVIG in 13 (30%) studies and/or combinations of these therapies.

Discussion
Compared to pediatric immune thrombocytopenia, there is substantially less information available describing pediatric AIHA. Most studies are retrospective and include both patients with isolated AIHA and Evan's syndrome. Data on diagnosis, investigation, and management are inconsistently reported. Only two studies reported their diagnostic approach for identifying secondary causes of AIHA. Although most studies described diagnostic criteria for AIHA, the specific criteria were variable, and only 4 studies classified AIHA according to severity. Specific treatments were variable. First-line treatments were most commonly a combination of corticosteroids and IVIG in keeping with guidelines for the management of AIHA in adults. Second-line treatments typically consisted of an immunosuppressive or immunomodulatory agent, with the specific medication or combination of medications varying depending on past therapies attempted and the patient's underlying disease.

Conclusion

Our review identified a deficit of high quality, prospective studies into pediatric AIHA. Prospective studies evaluating the treatment of pediatric AIHA are needed. A standardized definition and classification of pediatric AIHA will help guide much needed future studies of the appropriate investigation and management of pediatric AIHA.

Disclosures No relevant conflicts of interest to declare.

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