

SECTION 1

Introduction

André van Niekerk

Extraordinary Professor of Paediatric Pulmonology and Inborn Errors of Immunity, Department of Paediatrics and Child Health, Faculty of Health Sciences, University of Pretoria, Gauteng

Immunoglobulin (Ig) administration is indicated in several medical conditions. Preparations of human Ig are fractionated from donated human plasma and often form the cornerstone of treatment in primary immunodeficiency diseases (PIDs) and in some secondary immunodeficiencies (SIDs). They are especially helpful in those conditions affecting the humoral immune system. The preparations used for Ig replacement therapy (IRT) are often lifesaving, prevent hospital admissions, improve quality of life (QoL) and protect against end-organ damage. They should be administered timeously, are often the only form of treatment and chronic administration should not be interrupted. IRT preparations are listed on the South African and international essential drug lists.

PIDs result from genetic disorders affecting the immune system. The exact prevalence of PIDs is unknown^[1] but occurs more frequently than previously thought. Over 350 forms of PID have been described so far.^[2] The timely diagnosis, and appropriate administration of IRT in PID and SID remain keys to successful management.^[1] The indications for IRT are likely to broaden with better understanding of the underlying defects.

A potential scarcity of human plasma and Ig, the cost, the need for further research in some applications, and the potential for adverse effects (AEs) all warrant careful consideration regarding the indication and method for administration.

The objective of this document is to develop a strategy for the use of IRT in immune deficient patients in South Africa. It specifically refers to IRT in PIDs and in SIDs and aims to guide healthcare professionals, policy and decision makers, and funders on the current indications, and thereby to inform them on effective management of IRT in the South African context.

The Chairman of the Allergy Society of South Africa (ALLSA) invited members of the Primary Immunodeficiency Diseases South Africa Working Group of ALLSA (PIDDSA) to form an editorial committee to draft this strategy. Each section editor recruited opinion leaders to conduct systematic reviews of the literature on IRT in immune deficit, and to collate the evidence into a strategy.

ALLSA, PIDDSA, and the Clinical Immunodeficiency Foundation of South Africa (AFSA) endorse this strategy. PIDDSA will review the strategy on an annual basis and publish updates as needed. The process was initiated with funding received from ALLSA. All suppliers of IRT products were invited by the editor to contribute towards an unrestricted grant for the development and the dissemination of the strategy. The sponsors remained unknown to the contributors.

References

1. **Rezaei N, et al.** *Primary Immunodeficiency Diseases: Definition, Diagnosis, and Management*. Second edition, Springer, 2017.
2. **Picard C.** International Union of Immunological Societies: 2017 Primary Immunodeficiency Diseases Committee Report on Inborn Errors of Immunity. *J Clin Immunol*, 2018; 38:96–128.

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