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SERIES: PRIMARY IMMUNODEFICIENCIES IN LATIN AMERICA (I)

Advancing the management of primary immunodeficiency diseases in Latin America: Latin American Society for Immunodeficiencies (LASID) Initiatives

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Abstract Primary immunodeficiency diseases (PIDD) are associated with significant morbidity and mortality and result in a significant public health burden. This is in part due to the lack of appropriate diagnosis and treatment of these patients. It is critical that governments become aware of this problem and provide necessary resources to reduce this impact on health care systems. Leading physicians in their respective countries must be supported by their own governments in order to implement tools and provide education and thus improve the diagnosis and treatment of PIDD. The Latin American Society of Primary Immunodeficiencies (LASID) has initiated a large number of activities aimed at achieving these goals, including the establishment of a PIDD registry, development of educational programmes and guidelines, and the introduction of a PIDD fellowship programme. These initiatives are positively impacting the identification and appropriate treatment of patients with PIDD in Latin America. Nevertheless, much remains to be done to ensure that every person with PIDD receives proper therapy.
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Introduction

Patients with primary immunodeficiency diseases (PIDD) most commonly present with recurrent bacterial infections, although some may also be affected by systemic and organ-specific autoimmunity, chronic inflammation, and higher susceptibility to certain types of cancer.[1] The treatment of most primary immunodeficiencies with impaired antibody production involves immunoglobulin replacement therapy (IRT), either via the intravenous (IV) or subcutaneous (SC) route. These interventions have been demonstrated to be highly effective for the prevention of bacterial and viral infections and reduction of complications and sequelae.[2–8] In addition, IRT is effective as adjunctive therapy in the management of selected neurological, haematological, and autoimmune disorders.[9–13]

Not all PIDD require immunoglobulin therapy replacement. However, antibody deficiency disorders represent up to 60% of all PIDD and appear to be the most common forms in Latin America, based on results from the Latin American Society of Primary Immunodeficiencies (LASID) registry.[14] These disorders require IRT. Other types of PIDD affecting antibody production, such as severe combined immunodeficiency (SCID) and other well-defined medical situations, including immunosuppression after stem cell transplantation, may also require immunoglobulin administration. Thus, approximately 85% of all PIDD have some form of antibody deficiency and are therefore eligible for IgG replacement therapy. Guidelines for appropriate use of IgG therapy have been published for Brazil[15] and are in advanced-phase preparation for all LASID countries. Effective management of patients with PIDD requires accurate diagnosis and prompt delivery of the appropriate therapy. However, this is not achieved in a substantial percentage of patients. It has been estimated that more than 180 unique genetic diseases are associated with various immunological defects currently defined as PIDD by the International Union of Immunological Societies' (IUIS) expert committee for PIDD.[16] The diagnosis of many of these conditions is often delayed, or patients are misdiagnosed.[17–19] Results from one national registry indicated that the interval between the onset of clinical symptoms and diagnosis was significant even in PIDD such as IgA deficiency, which is relatively easy to diagnose.[20] There is also evidence suggesting that PIDD are undertreated even after accurate diagnosis. Results from a recent US survey carried out by the Immune Deficiency Foundation (IDF) indicate that approximately 250,000 people in this country have the diagnosis of a PIDD; the majority are antibody deficiencies, either alone or in combination with other immune defects. In each of these PIDD, intravenous immunoglobulin (IGIV) therapy is the standard of care. However, only 22% of the patients surveyed were receiving this treatment.[21] These results indicate that increased physician and patient education are needed to improve both the diagnosis and effective treatment of PIDD. Results from small-scale studies have shown that educational interventions can improve both diagnosis and referral to specialists.[22–24] Historically, diagnosis and appropriate treatment of PIDD in Latin America has been hampered by a lack of resources and educational opportunities for healthcare professionals. However, a wide range of new initiatives developed and/or supported by LASID is addressing these gaps.

The Latin American Group for Immunodeficiencies (LAGID) was created in 1993 to study the prevalence of PIDD in Latin America and to promote awareness of these diseases. In 2009 this group evolved into the society now named LASID, and created an advisory board that has published four reports and proceedings.[25–28] The first two papers focused on the prevalence and characteristics of patients with PIDD in Latin America,[25,26] while the third and fourth summarised shortfalls in PIDD diagnosis and treatment in Latin America and described the features of an educational outreach programme, an immunology fellowship programme, and a laboratory network aimed at closing these gaps.[27,28]

This paper summarises the proceedings from a recent meeting that brought together the members of the LASID advisory board and experts from other Latin American countries and the United States. The aim of said meeting was to consider and discuss the following topics related to the diagnosis and treatment of PIDD in Latin America:

- What resources are available to assist physicians in the diagnosis of PIDD?
- What are current policies regarding governmental support for immunoglobulin treatment in patients diagnosed with PIDD?
- What support and advocacy groups exist in Latin America to assist patients with PIDD, and how can patients and physicians work together to improve diagnosis and treatment of PIDD?
- What role should the industry play in improving the diagnosis and treatment of PIDD in Latin America?

This paper also summarises actions that have been initiated to address these issues.

Resources available to physicians for the diagnosis of PIDD

Advancing the diagnosis and treatment of PIDD requires improving physician training and providing better resources for those currently in practice. At present, many specialists who should be concerned about PIDD pay relatively little attention to these diseases. For example, a survey of allergists and immunologists in Brazil indicated that only 5% devoted more than 50% of their practice to PIDD and that 70% devoted less than 10% of their total practice to the diagnosis and management of these patients. Furthermore, over 50% never followed patients that needed immunoglobulin replacement.[29] This problem is being addressed by the establishment of the LASID Online Registry programme.[14] This programme also includes an educational initiative, as physicians have the opportunity to receive education about PIDD diagnosis before registering patients. Additional educational programmes for healthcare professions have been proposed and initiated by LASID, including a continuing medical education (CME) programme focused on PIDD warning signs, a LASID fellowship programme, a Latin American laboratory network, summer school programmes, and LASID scientific meetings.[14]

LASID has also taken a lead role in promoting the importance of training in PIDD for allergists/immunologists, pulmonologists, otolaryngologists, rheumatologists, and

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